International Child Health Care:

A Practical Manual for Hospitals Worldwide

edited by David Southall, Brian Coulter, Christiane Ronald, Sue Nicholson, and Simon Parke

BMJ Books

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Child Advocacy International

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Child Advocacy International is a humanitarian aid agency and registered charity (UK 1071486) which aims to assist healthworkers in poor countries to improve hospital care for children in the government sector. It helps build medical capacity by supporting and educating local staff particularly in the case of the critically ill and seriously injured children and in the neglected areas of pain control and palliative care.
All of its work is based on the United Nations Convention on the Rights of the Child.

Child Advocacy International
79 Springfields Road, Trentvale
Stoke-on-Trent ST4 6RY
United Kingdom

www.childadvocacyinternational.co.uk
www.childfriendlyhealthcare.org
E-mail: cai_uk@compuserve.com

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Viral infectious diseases and infections

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Contributors

Mohammed Arzomand
Associate Specialist in Community Child Health, Epsom and St Helier NHS Trust, Carshalton, Surrey, UK

Wendy Bailey
Principal Experimental Officer, Liverpool School of Tropical Medicine, Liverpool, UK

Alastair Baker
Consultant Paediatric Hepatologist, King's College Hospital, London, UK

Surya Bhate
Consultant Child and Adolescent Psychiatrist, Kolvin Unit, Newcastle General Hospital, Newcastle Upon Tyne, UK

Zulfiqar A Bhutta
Professor of Paediatrics and Child Health, The Aga Khan University, Karachi, Pakistan

Paula Blurton
Sister on the Children's Unit, North Staffordshire Hospital NHS Trust, Stoke on Trent, UK

Bernard Brabin
Professor of Tropical Paediatrics, Academic Medical Centre University of Amsterdam; Senior Lecturer in Tropical Paediatrics, Liverpool School of Tropical Medicine, University of Liverpool, Liverpool, UK

John Bridson
Consultant Paediatrician, Barnsley District General Hospital, Barnsley, UK

Kathy Brookes
Consultant Child and Adolescent Psychiatrist, Malborough Family Services, London, UK

James Bunn
Clinical Lecturer in Tropical Child Health, Liverpool School of Tropical Medicine, University of Liverpool, Liverpool, UK

Susan K Bunn
Clinical Lecturer in Paediatrics, Department of Child Health, University of Aberdeen, Aberdeen, Scotland

Sue Burr
Nursing Advisor to the Royal College of Nursing, London, UK

William Carroll
Specialist Registrar in Paediatrics, North Staffordshire NHS Trust, Stoke on Trent, UK

Eddie Chaloner
Honorary Lecturer, Leonard Cheshire Department of Conflict Recovery; Specialist Registrar in Surgery, University College NHS Trust, London, UK

Pornthep Chanthavanich
Associate Professor and Head, Department of Tropical Paediatrics, Mahidol University, Thailand

Luz Marina Lozano Chavarria
Consultant, Compleje de Salud "Concepcion Palacios", Centro Nacional de Toxicologia, Managua, Nicaragua

Andrew Clarke
Project Officer: Child Friendly Healthcare Initiative Child Advocacy International, Stoke on Trent, UK

Nicholas Coleman
Consultant in Anaesthesia and Intensive Care, North Staffordshire Hospital NHS Trust, Stoke on Trent, UK

Ed Cooper
Consultant Paediatrician, Newhan Community Health Services, Plaistow Hospital, London, UK

Brian Coulter
Senior Lecturer in Tropical Child Health; Honorary Consultant Paediatrician, Liverpool School of Tropical Medicine, Liverpool, UK

Malcolm Coulthard
Consultant Paediatric Nephrologist, Royal Victoria Infirmary, Newcastle Upon Tyne, UK

David Cundall
Consultant Paediatrician in Community Child Health, Leeds General Infirmary, Leeds, UK

Sally C Davies
Professor of Haematology; Director of Research and Development North Thames NHS, Central Middlesex Hospital, London, UK

Anil Dhawan
Consultant Paediatrician in Hepatology, Kings College Hospital, London, UK

Christopher Duke
Consultant Paediatric Cardiologist, Department of Congenital Heart Disease, Guy's Hospital, London, UK

Tim Eden
Professor of Paediatric Oncology, Christie Hospital, Withington, Lancashire, UK

Andrew Freeland
Consultant Paediatric ENT Surgeon, Radcliffe Infirmary, Oxford, UK

G S Gabra
Lead Consultant Haematologist and Head of WHO Collaborating Centre for Training and Development in Transfusion Medicine, Birmingham Blood Transfusion Centre, Birmingham, UK

Michael H N Golden
Professor in Medicine, Department of Medicine and Therapeutics, University of Aberdeen, Aberdeen, Scotland

Nicholas Guerina
Director of Perinatal Infectious Diseases, Associate Professor of Neonatal Medicine, New England Medical Center, Tufts University School of Medicine, Boston, USA

Devendra Kumar Gupta
Head, Department of Paediatric Surgery, All lndia Institute of Medical Sciences, New Delhi, India

Angela O'Higgins
Lecturer in Paediatric Nursing, Keele University, UK; Consultant in Paediatric Nursing, Child Advocacy International, Prishtina, Kosovo

James I Hagadorn
Associate Professor of Neonatal Medicine, Division of Newborn Medicine, Department of Pediatrics, The Floating Hospital for Children, Boston, USA

Prudence Hamadé
Clinical Medical Officer, Ynys-Y-Plant
Community Health Services, Newtown, Powys, UK

Rod J Hay
Consultant Dermatologist and Professor, St John's Institute of Dermatology, Guys, Kings and St Thomas' School of Medicine, London, UK

Jasmine Heslop
Consultant in Paediatric Nursing, Child Advocacy International, Mulago Hospital, Kampala, Uganda

Frank Hill
Consultant Paediatric Haematologist, Birmingham Children's Hospital, Birmingham, UK

Monica Hopkins
Advanced Nurse Practitioner, Royal Liverpool Hospital, Liverpool, UK

Sue James
Chief Executive, Barnsley District Hospital, Barnsley, UK

Roberto Jiminez
Consultant Paediatric Nephrologist, Childen's Hospital, Manuel de Jesus Rivera, Managua, Nicaragua

Lynne Jones
Child and Adolescent Psychiatrist; Director, Child Psychiatry Project Kosovo, Child Advocacy International, Stoke on Trent, UK

Ismeta Kalkan
Consultant Paediatrician, Pedijatrijska Klinika, Kosevo Hospital, Sarajevo, Bosnia-Herzegovnia

Leela Kapila
Consultant Paediatric Surgeon, Queen's Medical Centre, University Hospital, Nottingham, UK

Anupam Lall
Senior Research Associate, Department of Paediatric Surgery, All India Institute of Medical Sciences, New Delhi, India

David Lalloo
Senior Lecturer in Tropical Medicine, Liverpool School of Tropical Medicine, Liverpool, UK

Alice Leahy
Specialist Registrar in Paediatrics, University Hospital, Lewisham, London, UK

James Leonard
Professor of Paediatric Metabolic Disease, Institute of Child Health, University of London, London, UK

Michael Levin
Professor of Paediatrics, Department of Academic Paediatrics, Imperial College of Science Technology and Medicine, London

Mary Limebury
Specialist Registrar in Paediatrics, Great Ormond Street Hospital, London, UK

David Mabey
Professor of Communicable Diseases, London School of Hygeine and Tropical Medicine, London, UK

Steve Mannion
Consultant Surgeon, Medecins Sans Frontieres UK and World Orthopaedic Concern

M N Mansour
Senior Neonatologist, Kuwait University, Kuwait

Waghi El Masri
Consultant Physician and Director, Midlands Centre for Spinal Injuries; Oswestry, Shrophshire; Senior Lecturer University of Keele, UK

Kieran McHugh
Consultant Paediatric Radiologist
Great Ormond Street Hospital, London, UK

Elizabeth Molyneux
Professor of Paediatrics, College of Medicine, Blantyre, Malawi, C. Africa

Allie Moosa
Professor of Paediatrics, Kuwait University, Kuwait

Sarah Morley
MRC Clinical Training Fellow, Department of Academic Paediatrics, Imperial College of Science Technology and Medicine, London, UK

Rob Moy
Senior Lecturer in Community Child Health, Institute of Child Health, University of Birmingham, Birmingham, UK

Charles Newton
Senior Lecturer in Paediatric Neurosciences, Kemri/ Wellcome Trust Unit, Kilifi, Kenya

Sue Nicholson
Project Director, Child Friendly Healthcare Initiative, Child Advocacy International, Stoke on Trent, UK

Oscar Nunez
Consultant Paediatrician; President, Nicaraguan Society of Paediatrics, Children's Hospital, Manuel de Jesus Rivera, Managua, Nicaragua

Peter Oakley
Consultant in Anaesthesia and Trauma, North Staffordshire Hospital NHS Trust, Stoke on Trent, UK

Bernhards Ogutu
Consultant Paediatrician, University of Nairobi, Nairobi, Kenya

Deb K Pal
Neurosciences Unit, Institute of Child Health, The Wolfson Centre, London, UK

Simon Parke
Specialist Registrar in Paediatrics, Birmingham Children's Hospital, Birmingham, UK

Silvia Patrizi
Consultant Neonatologist, New England Medical Center, Tufts University School of Medicine, Boston, USA

Barbara Phillips
Consultant Paediatrician in Accident and Emergency, Royal Liverpool Children's Hospital, Alder Hey, Liverpool, UK

Barry Pizer
Consultant Paediatric Oncologist, Royal Liverpool Children's Hospital, Alder Hey, Liverpool, UK

Jonathan Punt
Senior Lecturer in Paediatric Neurosurgery, Queen's Medical Centre, University Hospital, University of Nottingham, Nottingham, UK

Shakeel A Qureshi
Consultant Paediatric Cardiologist, Department of Congenital Heart Disease, Guy's Hospital, London, UK

Sajjad ur Rahman
Consultant Neonatologist, Khyber Teaching Hospital, Peshawar, Pakistan

Anthony Roberts
Director, Oxford Regional Burn Unit, United Kingdom
Consultant Plastic Surgeon, Stoke Mandeville Hospital, Aylesbury, UK

Joan Robson
Consultant in Paediatric Accident and Emergency Medicine, Royal Liverpool Children's Hospital, Alder Hey, Liverpool, UK

Christiane Ronald
Specialist Registrar in Paediatrics, New Cross Hospital, Wolverhampton, UK

Oliver Ross
Fellow in Paediatric Intensive Care, Great Ormond Street Hospital for Children, London, UK

Terence J Ryan
Emeritus Professor of Dermatology, Oxford Centre for Health Care Research & Development, The Churchill Hospital, Oxford, UK

Martin Samuels
Consultant Paediatrician and Senior Lecturer in Paediatrics, North Staffordshire Hospital NHS Trust, Stoke on Trent, UK

Sarah Samuels
Paediatric Physiotherapist, North Staffordshire Hospital NHS Trust, Stoke on Trent, UK

Felix Sanchez
Consultant in Paediatric Pulmonology, Children's Hospital, Manuel de Jesus Rivera, Managua, Nicaragua

John Sandford-Smith
Consultant Ophthalmologist, Leicester Royal Infirmary, Leicester, UK

Ivette Sandino
Consultant Paediatrician, UNICEF, Nicaragua

Sudhir Sethi
Consultant Paediatrician, Stafford General Hospital, Staffordshire, UK

Neela Shabde
Consultant Paediatrician, Northumbria Healthcare NHS Trust, Albion Road Clinic, North Shields, Tyne and Wear, UK

Manoj Shenoy
Clinical Fellow in Paediatric Urology, Nottingham City Hospital, Nottingham, UK

Sheila Silva
Consultant Paediatrician in Infectious Diseases, Children's Hospital, Manuel de Jesus Rivera, Managua, Nicaragua

Alan Smyth
Consultant in Paediatric Respiratory Medicine, Nottingham City Hospital, Nottingham, UK

Anthony Solomon
Clinical Research Fellow, KCMC, Moshi, Tanzania

David Southall
Professor of Paediatrics, Keele University; Honorary Medical Director, Child Advocacy International, Stoke on Trent, UK

Taunton R Southwood
Professor of Paediatric Rheumatology, Department of Rheumatology, Birmingham Children's Hospital, Birmingham, UK

Peter Sullivan
Lecturer in Paediatrics, Consultant Paediatrician, University of Oxford, Department of Paediatrics, John Radcliffe Hospital, Oxford, UK

R D G Theakston
Professor in Medical Biology, Liverpool School of Tropical Medicine, Liverpool, UK

Gareth Tudor-Williams
Consultant Paediatrician, Imperial College School of Medicine, St. Mary's Hospital, Department of Paediatrics, London, UK

James Tumwine
Professor of Paediatrics, Makerene University, Mulago Hospital, Kampala, Uganda

Mark Twite
Paediatric Critical Care Fellow, The Children's Hospital, Denver, Colorada, USA

Sirijitt Vasanawathana
Professor of Paediatrics, Khon Kaen Hospital, Khon Kaen, Thailand

Janet Vickers
MacMillan Nursing Sister, Royal Liverpool Children's Hospital, Alder Hey, Liverpool, UK

Jerry Wales
Senior Lecturer in Paediatric Endocrinology, Institute of Child Health, University of Sheffield, Sheffield Children's Hospital, Sheffield, UK

Mary Warrell
Centre for Tropical Medicine, John Radcliffe Hospital, Oxford, UK

Anthony Williams
Consultant Neonatologist, St George's Hospital Medical School, London, UK

Bridget Wills
Clinical Senior Lecturer in Paediatrics, University of Oxford–Wellcome Trust CRU, Centre for Tropical Diseases, Ho Chi Minh City, Vietnam

Dankwart F Wittenberg
Professor and Head of Department of Paediatrics and Child Health, University of Pretoria, South Africa

Beatrix Wonke
Consultant Haematologist, The Whittington Hospital, London, UK

George Wyatt
Senior Lecturer and Clinical Director of Tropical Medicine, Liverpool School of Tropical Medicine, Liverpool, UK

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Preface

On 20 November 1989 the General Assembly of the United Nations declared a Convention on the Rights of the Child. All but two countries have to date ratified this Convention. However, despite over 10 years of its existence, the Tables below illustrate the unacceptable mortality rates for children under 5 years of age. Although the situation in rich countries continues to improve, poor countries are hardly progressing and in some instances there is a deteriorating level of healthcare resulting in an increase in mortality and morbidity of children.

Table 1 Changes in under-5-year mortality rates from 1988 to 1997

Under-5-year mortality	1988	1997	Reduction
□40/1000 (1 in 25)	73% of the world's children	69% of the world's children	4%
□100/1000 (1 in 10)	37% of the world's children	33% of the world's children	4%
25 most advantaged (richest) countries	11.3/1000 live births	6.7/1000 live births	41%
25 most disadvantaged (poorest) countries	208.5/1000 live births	176.7/1000 live births	15%

UNICEF Data: *State of the World's Children*

Table 2 Comparison of maternal and child mortality between poor countries and one rich country (the UK)

Country	Number of deaths per year adjusted to the number that would occur if the adult plus child population was 58 million (the population of the UK)	
	Children <5 years of age	Mothers during pregnancy or delivery
United Kingdom (population 58 million)	5411 (15/day)	70
Countries in extreme poverty*	236 198 (647/day)	12 682
Afghanistan	777 200 (2129/day)	17 697

*Defined as those countries where there is an under-5-year mortality rate of ≥50/1000 (46% of the world).

Article 25 of the Universal Declaration of Human Rights states the following:

> "Everyone has the right to a standard of living adequate for the health and well-being of themselves and their family, including food, clothing, housing and medical care."

This manual has been written for paediatricians and nurses caring for children all over the world. It is primarily designed to ensure that a minimum standard of care is applied to every individual child, regardless of how poor or rich is their country or their family. There is no question that some of the worst suffering endured by children occurs in hospitals and other healthcare institutions. Not all of this is related to a lack of funds; much concerns a deficit in the education of healthcare professionals. Often, the training of doctors and nurses is a low priority and even after training they are not provided with adequate salaries, professional recognition or decent teaching materials. Standard medical textbooks are usually too expensive for doctors and nurses in disadvantaged countries, hampering their continuing medical education.

Editing and writing this book has been challenging for the authors. We have attempted to identify what we regard as the minimum standards of treatment for all major diseases and injuries that affect the newborn baby, infant and child, wherever they are cared for in the world.

But we also wanted to offer a set of ideal standards for care where resources are adequate. Therefore we have incorporated the essential minimum standard of care alongside the best standards currently available. The book describes both a gold standard and a minimum standard of care. However, readers will notice that for most of the treatments recommended, the minimum and gold standards are identical. The reason for this is that there are certain treatments that we could not sanction cutting in the provision of hospital care for children, whatever the pressures. Equally, the undertaking of high-quality medical education can attain much, particularly those programmes that include scenario-based teaching, such as the paediatric and neonatal life support courses. This manual includes reference to these courses and we are extremely grateful to the Advanced Life Support Group in the UK for letting us work with them to provide these courses in poorly resourced countries. The beneficial effects have been truly amazing with major improvements in the quality of, for example, neonatal resuscitation which otherwise was being done so badly that many children were ending up either dead or permanently disabled.

This manual is as up-to-date as we can make it and we plan, in close collaboration with our publishers, for it to be sold in rich or advantaged countries. The royalties from the sale of the book will be used to fund its translation, where necessary, and its dissemination, at low cost, to as many paediatricians as possible in the government-run hospitals of poorly resourced countries.

Acknowledgements

The editors are extremely grateful to all professionals who have contributed to this manual. They have undertaken their chapters willingly and to tight deadlines and have done their best to identify the most important issues. We are also grateful to the Department of Child and Adolescent Health and Development of the World Health Organization for a number of sections in this manual, which are taken from their own excellent publication entitled *Management of the child with a serious infection or severe malnutrition* (WHO/FCH/CAH/00.1).

How to use this book

This is a comprehensive text for all paediatricians caring for children in hospital. It can be used by those with limited resources and also where greater resources are available. We have identified the different levels of care in the following ways:

- **Minimum standards requirements** are given in a highlighted box at the the beginning of each clinical chapter.
- ***A standard of care*** where resources are not limited appears as bold, italicised text.
- **Key points** of particular importance in management of children are identified by a tick in the margin and bold text.

In this way we hope the book will act as a user-friendly, speedy reference on any paediatric ward.

Foreword

Around the world, tens of thousands of children receive little or no medical care when they are ill. But these huge numbers numb the mind, whereas the stories and the images of individual children stay with us for ever. For me, the memories include a child with sickle cell disease, disabled by a series of strokes and abandoned outside an African village by her family; a little boy with acute osteomyelitis, moaning in agony because no-one understood about pain relief in children; a baby dying of gross dehydration, while a leaking iv fluid bottle emptied itself unnoticed onto the floor.

These things are happening now, in the 21st century. The skills and the technology are available to improve health care and reduce suffering, even in the poorest countries, but for a variety of reasons they are not reaching the children's wards and clinics where they are needed.

David Southall has assembled an impressive array of experts who together have created *International Child Health*. Within its pages you will find information, guidelines and suggestions about health care in virtually every situation and setting imaginable – peace and war, emergency and chronic, hospital and clinic. The advice is up to date and authoritative. It aims to be realistic, but at the same time does not compromise on standards. The authors believe that the health-care professions around the world should be satisfied only with the best care for children. In the UK the Government is mounting an assault on so-called postcode prescribing, arguing that the quality of health care ought not to depend on where you happen to live within the country. Yet for many children around the world, whether or not they receive appropriate treatment – or indeed any treatment at all – depends entirely on where they live.

As well as professional expertise, there is an undercurrent of frustration, even anger, in this book. Frustration at the difficulties in implementing standards of care that we in the West have taken for granted for many years; anger at the needless suffering that results.

The authors hope that this book will find a home in the big teaching centres, the rural hospitals, and the village clinics of many countries around the world. Its success will be judged not only by how many copies are sold, but by its impact on the quality of paediatric practice.

David Hall
Professor of Community Paediatrics,
University of Sheffield;
President, Royal College of Paediatrics and
Child Health

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Introduction

This manual is written with the aim of helping healthcare staff who are treating children admitted to hospitals, particularly secondary and tertiary referral hospitals, in countries all over the world. It is especially aimed at healthcare staff in countries where financial and human resources may be limited, and borders and infrastructures insecure. Where there are these difficulties staff are already making considerable on-going advances in improving children's healthcare.

This manual attempts to build on existing efforts and seeks further to identify an **internationally applicable minimum standard of healthcare for children in hospitals**. It is intended to complement the World Health Organization's guidelines on the Integrated Management of Childhood Illness (IMCI) for community use, and their manual *Management of the Child with a Serious Infection or Severe Malnutrition*, which is aimed at first referral level, the district hospitals.

Although no one can doubt the wisdom of putting resources into primary care with the aim of preventing admission to hospital in as many children as possible, there will always be a need for high quality, accessible out-patient services and for secondary and tertiary care (first and secondary referral levels). As primary care improves, less and less of a country's health budget will be spent on rescuing children from illnesses and injuries that should not have progressed or happened.

However, of key importance is the degree of suffering that occurs in individual children who cannot be rescued by good primary healthcare and are managed in poorly financed and equipped hospitals with scarce human resources. Much of this suffering is not related to a lack of expensive equipment or other material resources but to a lack of attention to basic human rights.

Many hospitals in poorly financed countries are not clean, do not have basic water and sanitation, a reliable electricity supply or even minimal security. The staff, both clinical and non-clinical, are often underpaid and sometimes undervalued by their communities. There may be minimal if any pain control and the indiscriminate use of powerful antibiotics leads to multiresistant organisms that have the potential to spread to other communities or even countries.

Unnecessary suffering due to lack of respect for human rights, inadequate pain control or acquisition of hospital acquired infection can be alleviated with additional staff training, a change in attitude and a redistribution and/or more effective use of existing resources.

In some countries there is a two-tier system of healthcare with private hospitals and poorly funded government hospitals. The private hospitals are often run by multinational organisations. They have the most sophisticated and up-to-date equipment and facilities, which are available for the richest minorities. They tend to attract the most competent doctors and nurses from the state sector because of their huge financial resources. Because they provide care for the country's elite there is sometimes little incentive for those who control resources to improve care for the majority of the population.

The vision of our authors relates to the provision of this basic minimum standard of care in all state hospitals and its availability to all children regardless of their families' ability to pay. These hospitals would be run by highly competent, well-trained, well-paid and valued professional managers and support staff. They would employ at appropriate rates of pay, the best hospital-trained doctors and nurses in the country, form the basis for medical and nursing education and be closely integrated with the primary healthcare system. The resources in these hospitals as compared with the community (primary) health service, would be proportionate to the wealth of the country as a whole.

Children and their families should not be frightened to come to the hospital or regard it as a place to die. In many countries, children suffer much more before dying in hospitals than they do at home. Children deserve the best possible medical care delivered safely, with equal attention paid to their own and their families' emotional and psychological needs. This care should be "Rights Based" and provided within an appropriate ethical framework. It should avoid discrimination for any reason and should attempt to identify and support those children who for social, financial, family or other reasons are particularly disadvantaged or vulnerable.

Provision of this minimum standard is dependent on an infrastructure which includes:

- Secure prioritised financial provision for children's healthcare
- Appropriate non-clinical support facilities and services
- Appropriate clinical services, mainline and support, including services for children in need of protection and those with a disability
- Appropriate clinical and non-clinical staffing and expertise
- Good management, maintenance, care and protection of resources, facilities and staff

- A staff-training strategy that provides appropriate training opportunities for all health professional staff to internationally accepted standards and includes opportunities for on-going professional development
- Induction training for all new staff (professional and non-professional) concerning the policies and procedures of the hospital that is employing them, particularly those relating to their specific role. This should be followed by on-going refresher training opportunities
- A structure for the safe, appropriate processing, prioritisation and treatment of children who use the service
- An understanding and everyday practice of the articles relating to the delivery of healthcare as detailed in the United Nations 1989 Convention on the Rights of the Child (UNCRC).

Children must not endure unavoidable suffering. All healthcare staff, in partnership with parents, have a duty to act as advocates to ensure children receive a standard of care that is appropriate to their vulnerability.

The authors would welcome suggestions for the new edition. Please write to:

Child Advocacy International
79, Springfields Road, Trentvale
Stoke on Trent
ST4 6RY
United Kingdom
E-mail: cai_uk@compuserve.com

Section 1

Philosophies of care: hospital facilities, services and their organisation

How to use this book

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1.1 The UN Convention on the rights of the child – 1989: A healthcare worker's brief guide

Sue Nicholson

The Convention contains 52 articles. These provide a set of principles and standards that can be used for the planning and practice of children's health services for **all children and young people up to the age of 18 years**. The Convention has been ratified by most countries in the world.

The articles that relate to health are:

Article 3
Provides that any decision or action affecting children, either as individuals or as a group, should be taken with "their best interests" as the most important consideration.

Article 9
Holds that children have a right not to be separated from their parents or carers unless it is judged to be in the child's best interests.

Articles 5 and 18
Note that while the main responsibility for bringing up children lies with parents and carers, they, in their turn, may need support, advice and services in order to fulfil these obligations.

Article 12
Obliges health professionals to seek a child's opinion before taking decisions that affect her or his future … "the child who is capable of forming his or her own views has the right to express those views freely… the view of the child being given due weight in accordance with age and maturity".

Children should be consulted over all kinds of treatment, and over all aspects of management in general. Children should be informed of the nature of the treatment and should be involved in choices where alternatives exist. These considerations are particularly important in children with chronic illnesses such as cystic fibrosis, diabetes, cancer or renal failure.

Article 16
Details the child's right to privacy and confidentiality.

Articles 17 and 24
Outline the duty to provide information aimed at promoting physical and mental health, and appropriate information for health promotion being given to parents on matters such as breastfeeding, nutrition in general, hygiene, environmental sanitation, accident prevention, contraception, pre- and postnatal health.

Article 17
Additionally notes that all children have the right to information being presented in such a way to take account of their linguistic and communication needs.

Article 19
Holds that children's physical and personal integrity be respected and supports the view that health examinations should only be carried out in the child's best interests.

Articles 19 and 39
Require that appropriate support, care and rehabilitation services be available for those who have suffered abuse or neglect.

Article 23
Provides that children with disabilities and learning difficulties have the right to services and facilities provided in such a way that they are able to achieve the fullest possible social integration and individual development.

Article 24
Says that all children have the right "to the enjoyment of the highest attainable standard of health". It also describes the rights of all children to services which prevent ill health and promote good health and to the facilities for the treatment of illness and for rehabilitation after illness.

Article 30
Delineates the rights of children from indigenous populations and ethnic, linguistic, or religious minorities to have their rights safeguarded in enjoying and practising their own cultures, religions and languages.

Article 31
Holds that children in hospital, or receiving long-term treatments, have the right to equal opportunity in play, recreational and cultural activities.

Article 33
Describes the right of children to appropriate facilities and services protecting them from drug and solvent abuse.

Article 42
Holds that in order to benefit from their rights children must know and understand the rights to which they are entitled, and it is the duty of the state (and a duty which paediatricians may take on) to make the Convention's principles relating to children's health widely known.

12 Hospital non-clinical support services and facilities

Sue James, David Southall and Sue Nicholson

Minimum standards requirements

Hospital services

- Water and sanitation
- Security and safety
- Reliable electricity supply
- Heating and cooling
- Laundry
- Staff morale/salaries/continuing education

Introduction

For effective delivery of healthcare a secure financial strategy with robust financial and manpower controls, a properly maintained technical infrastructure, clear lines of accountability, and good management and communication lines all need to be in place. There should ideally be clearly defined written personnel procedures, good training systems and written policies and guidelines for all staff functions. The facilities and functions described in this chapter need to be in place and are as important as the quality of medical care given. The services and facilities discussed in this text are not comprehensive and well resourced countries may have many additional ones. If these services and facilities are in place, managed efficiently, supported and maintained, mainline healthcare delivery will be effective; without them clinical care, however excellent, is likely to be ineffectual.

Management

Advice on generic hospital management is difficult since the ability to deliver a minimum standard of care depends on the political, social and economic context in which the hospital is placed. Ideally there should be a named person responsible for each facility and service, in addition to an overall hospital manager or management team. The hospital manager, or management team, should have overall responsibility for finances, estates and facilities, human resources, clinical mainline and support services, training for all staff and the administrative services necessary to support all of these activities. There should also be a head nurse and senior doctor within the management team.

Essential services and facilities

Hospital security and access

The security and accessibility of the hospital are the most essential facilities of all, especially given the relative lack of police resources in many poorly resourced countries. There is also a need for governmental and international agencies to ensure that hospitals are protected and do not become targeted during armed conflict.

On a local level, the hospital should have secure entrances where all persons attending have to demonstrate a legitimate reason for entry. Clearly no weapons should be allowed into the hospital and it may be necessary in some countries to have a metal detector to screen all visitors.

A well-organised car parking system is required, with strictly policed access areas for emergency vehicles and for parents or relatives bringing very sick patients to and from the hospital.

Safety

There should be clear written evacuation and fire policies, together with appropriate equipment, for instance fire extinguishers.

Communications

Good communications systems for staff, visitors and patients are essential. In an ideal situation both outside and internal telephone systems should be available. If telephones are not possible, alternative effective and reliable systems of communication should be used. A hospital paging system for doctors, senior nurses and managers aids communication in emergency situations.

If a telephone system exists, Internet access is invaluable for information sharing and education, both within a country and globally. Provision can be sought via governmental or non-governmental donor sources. A nominated person with overall responsibility for hospital computer systems predisposes to a cohesive service both internally and externally, avoiding duplication and ensuring appropriate usage.

Utilities

Water and sanitation

Hygiene within the hospital is dependent on a constant and high-quality water supply and adequate sanitation and washing facilities (bathrooms, showers and accessible sinks), all of which are **vital if nosocomial infection** (see Chapter 1.6) **is to be minimised**.

Electricity

An electricity supply within the hospital that functions whatever the state of the service to the rest of the area is mandatory. This means a regularly maintained generator of sufficient power (calculated from bed dependency and operating theatre requirements) and special emergency circuits. Power cut simulations should be carried out regularly to test the system.

Heating and ventilation

Ideally there should be a functioning central heating system within the hospital. For this to work there will also need to be a continuous water supply. If either of these cannot be ensured, then electric heaters should be installed in areas where there are patients.

In hot weather, there should be sufficient windows allowing a comfortable temperature during the hottest part of the day. An air-conditioning system or fans, either electric or manual (to be worked by relatives), should be available in areas of the hospital which become particularly hot or for patients who must be kept cool (for example, children with high fevers or with head injuries).

Laundry service

Bedding and other items must be frequently washed. To do this the hospital must have a staffed laundry service that ideally has industrial washing machines in sufficient number and drying facilities. Clean bedding, towels and nappies need to be available. A small supply of children's nightwear and other clothing may be needed on the wards for families who do not have changes of clothes.

Cleaning services

The patients being cared for in the hospital are particularly vulnerable to nosocomial (hospital-acquired) infection (see Chapter 1.6). **To reduce this risk, sufficient staff should be employed on a rota over the 24-hour period to keep all areas of the hospital and grounds clean at all times.** Written cleaning policies should be in place and a supply of appropriate cleaning materials and disinfectants readily available.

Clean hospital grounds, pathways and entrances avoid dirt being transmitted into the ward and other patient areas by staff, relatives and other visitors.

Vermin need to be kept away from the hospital buildings.

Toilets, bathrooms and other facilities needed for personal hygiene and for equipment cleaning are of particularly importance and these areas should always be scrupulously clean.

Certain areas must be aseptic, for example operating theatres and selected equipment items (see Chapter 1.12). Ideally there should be a Central Sterilising Service, if this is not possible or feasible there should be suitable sterilisers and a supply of appropriate disinfectants at differing dilutions. Manufacturers' instructions should be followed for specific items of equipment wherever possible.

Rubbish disposal service

A powerful incinerator working 24 hours a day is essential for the safe disposal of clinical waste. **A system to handle and dispose of all waste (clinical and non-clinical), including "sharps" is also needed**. Written policies for various types of rubbish disposal, and training, should be available to all staff.

Facility and utility maintenance services

Buildings and utilities

It is essential for these to be maintained to as high a standard as possible. Suitably trained engineers, builders and other maintenance staff are necessary.

Equipment

There is no point in having expensive medical and surgical equipment if this cannot be maintained or used. Sufficient trained local bioengineers are therefore essential. All equipment used in the hospital should be robust, compatible, suitable for the conditions and level of expertise available and when new, should be purchased with accompanying staff training and servicing arrangements.

Portering service

For the functional relationships between different departments, for example the movement of patients to and from the operating theatres, a well-organised, trained and sympathetic team of porters is essential.

Catering service

Hospital food must be prepared under very hygienic conditions and by staff who do not have gastroenteritis or superficial skin infections. Ideally nutritious food should be provided free of charge for children. Special diets for malnourished children should be available (see Appendix 7.5).

Administration support services

Reception, and other administrative support staff need to be employed to aid facility managers and other non-clinical and clinical staff. There needs to be a staffed system to store and process medical and nursing records. There should be strict rules about who has access to the medical and nursing records, where they are stored and for how many years they are kept.

Human resource and personnel issues

Hiring and dismissing staff
There should be transparent procedures for advertising for, interviewing and employing staff which include non-discrimination policies, in particular on the basis of sex, age, ethnic or religious status.

Employment and financial issues
✔ **It is essential that the professions of medicine and nursing in all countries are highly regarded and respected. The salaries for doctors and nurses should reflect this**. If this does not occur many will have to undertake other jobs during the day and cannot have pride in their work. A lack of funding for salaries can also lead to corruption with some doctors borrowing supplies and equipment from their hospital to use in private clinics, which of course fails to serve the most needy and poor in the community.

There should be systems for ensuring the regular and secure recording of the time spent at work and the appropriate payment arrangements based on the prescribed number of hours worked (part- or full-time). On-call emergency work and its payment should also be prescribed.

✔ There should be a **professional registration system for each country which ensures a basic level of training as well as a system which validates experience and ability at intervals after initial registration** (see below).

Training and continuing staff education (see also Chapters 1.10 and 1.24)
Induction training for all staff concerning hospital policies should be mandatory.

Governments in well-resourced countries could encourage an exchange of education with those working in less well-resourced situations.

✔ **New teaching techniques, such as scenario based teaching for example APLS courses (see Chapter 1.19), should be introduced**.

Children's healthcare organisations in well-resourced countries could consider sponsoring individual children's doctors and nurses to work in less well-resourced countries. The selection of those for sponsorship requires great care.

Professional registration requirements for healthcare workers
These will vary from country to country. However some form of governmental registration is essential. There should also be procedures governing the employment of expatriate staff in the health service.

Vetting of healthcare workers
✔ **All staff working with children, either locally or from abroad, should be checked to ensure that they are suitable to work with children and have not been involved in the abuse of children**. This is also important with respect to expatriate staff.

Staff health (see also Chapter 1.8)
There needs to be a system to advise the hospital management about staff health problems that may affect patient care.

Needlestick injury
Although the risk of infection is very small, ✔ **a policy should be in place to deal with this issue urgently, especially in hospitals where there are many patients with HIV infection**.

One possible strategy is as follows:

1. Discuss with the child's family what has happened and ask if the child's HIV status is known. If not, discuss the possibility of testing the child if the injury occurred during normal working hours. Remember that anyone having an HIV test has the right to counselling. If out of hours, or the family decline testing proceed to step 3.
2. If the child has negative HIV ELISA and is over 18 months infection is extremely unlikely. If under 18 months a positive ELISA may reflect maternal antibody. However any positive test proceed to step 3. If negative, the professional is not at risk of HIV infection (however, they should be sure they are hepatitis immune).
3. Arrange a baseline HIV ELISA for the professional after appropriate counselling. If positive, the professional will need to discuss further treatment with his or her own doctor.
4. If the professional's baseline serology is negative and the patient is positive for HIV, antiretroviral prophylaxis should be started urgently. Current recommendations advise one month of treatment.

The professional will need a repeat ELISA after 3–6 months to check his/her status.

Confidentiality
Systems need to be in place ✔ **to ensure that patients' records and personnel files of employed staff are kept confidential**.

Other services for patients, parents/carers and visitors

Health information should be available for the parents and carers of children and for older children.

There should be toilets and when possible telephones available for visitors and facilities for those visitors with a disability.

Ideally, there should be written policies concerning the rights and responsibilities of patients, resident parents/carers and visitors widely displayed around the hospital. These should include issues such as the prevention of smoking, alcohol, violence (verbal and physical) and weapons in the hospital. Smoking is particularly important with regard to children's health but is so important in stressed parents that it may be inappropriate to ban it altogether, and instead limit it to defined areas.

Family-centred care
The role of parents in caring for their children alongside and in partnership with professional staff is vital but must be handled extremely carefully. Parents must not be exploited but equally in poor countries hospital care would not be possible without them. Good understanding of roles

and good communication is of paramount importance (see also Chapter 1.4).

Conclusions

The provision, organisation, and financing of these services, facilities and functions and the management of the human resource needed to service them is as important as those needed to provide the clinical and clinical support services. A sound hospital infrastructure and management is of paramount importance for the provision of good care.

Further information on other work-related issues concerning health care staff can be found in Chapters 1.3, 1.4, 1.5, 1.6, 1.7, 1.8, 1.9, and 1.10.

13 UNICEF/WHO baby friendly hospital initiative (BFHI)

Sudhir Sethi and Ivette Sandino

Minimum standards requirements

Breastfeeding

- UNICEF/WHO Baby Friendly Initiative
- Mothers' rooms and facilities

- Breastfeeding has important health benefits both for the mother and the child.
- All mothers have the right to make a fully informed choice as to how they feed their babies and staff should not discriminate against any woman due to her chosen method of infant feeding, fully supporting her once she has made her choice.
- The potential health risks from formula feeding should be discussed with all women.
- Hospitals providing maternity services should adopt a breastfeeding policy which covers all ten steps to successful breastfeeding enabling mothers to breastfeed exclusively for 4–6 months.

The ten steps of the BFHI

1. **Have a written breastfeeding policy that is routinely communicated to all healthcare staff**.
   - All staff to adhere to this policy to avoid conflicting advice.
   - New staff to be orientated to the policy.
   - The policy should be available in all areas of the maternity unit and children's wards.
   - The policy should also be accessible as audio or videotapes in appropriate local languages.

Comment:
Formulation and implementation of breastfeeding policy can encourage the implementation of other policies that protect children. **For example the registration of the newborn is a universal human right.** ✔

2. **Train all healthcare staff in the skills necessary to implement the policy**.

Comments:
   - Although BFHI involves only health personnel in training, all administrative staff in training such as secretaries and security personnel should be included.
   - This can encourage community involvement and milk bank development.

3. **Inform all pregnant women about the benefits and management of breastfeeding**.
   - Every pregnant woman should have an opportunity to discuss infant feeding on a one-to-one basis with a midwife or health visitor.
   - Aim to give women confidence in their ability to breastfeed.

Comments:
   - **To sustain a culture of breastfeeding, it is necessary to begin teaching children about breastfeeding; this may be most effective during the pregnancy or at school**. ✔
   - It is equally important to teach breastfeeding techniques such as hand extraction, since 40–50% of the women have problems related to technique and lack the knowledge to overcome them.

4. **Help mothers initiate breastfeeding soon after birth**.
   - **Encourage mothers to hold their babies in skin-to-skin contact as soon as possible after delivery** in an unhurried environment, regardless of their intended feeding method (usually within 30 minutes after uncomplicated vaginal delivery and within 3–4 hours of Caesarian section). ✔
   - Offer the first breastfeed when mother and baby are ready.
   - Help must be available from a midwife if needed.

5. **Show mothers how to breastfeed and how to maintain lactation, even if they are separated from their infants**.
   - Explain positioning and attachment of the baby to the mother, who must be helped to acquire the skill for herself.
   - All breastfeeding mothers should be shown how to hand express their milk.
   - If the baby is separated for medical reasons it is the shared responsibility of the neonatal nurse and midwife to ensure the mother is given help to express her milk to maintain lactation at least 6–8 times in a 24-hour period.

Comment:
Although electric breast pumps are helpful, hand expression is as effective.

6. **Give newborn infants no food or drinks other than breast milk unless medically indicated.**
   - No water or artificial feed should be given to a breastfed baby unless prescribed by a midwife or paediatrician in consultation with parents and the reason explained to them.
   - Parents who request supplementation should be made aware of its implications on baby's health and breastfeeding. This will allow them to make a fully informed choice.
   - Supplements when prescribed should be documented in the notes with the reasons.

Comments:
UNICEF/WHO have defined the medical reasons for prescribing supplements. These are infrequent and uncommon.

7. **Practise rooming-in – allowing mothers and infants to remain together 24 hours a day**.
   - **Separation should only be done for medical reasons (rare)**.
   - **Babies should not be routinely separated from mothers at night, including those who are formula-fed**.

Comments:
- To include the neonatal intensive care and newborn services.
- May result in quicker recovery of the newborn and decreased duration of stay of infants in hospital.

8. **Encourage breastfeeding on demand**.
   - Demand feeding should be encouraged.
   - Mothers should be informed that it is acceptable to wake their baby up if their breasts become overfull.

9. **Give no artificial teats or pacifiers (also called dummies or soothers) to breastfed infants.**
   - Staff should not recommend these.
   - Parents wishing to use them should be advised of the possible detrimental effects on breastfeeding.
   - Cups but never bottles should be used.
   - Any mother wishing to use a nipple shield must have the disadvantages explained and should be supervised by a skilled practitioner.

10. **Foster the establishment of breastfeeding support groups and refer mothers to them on discharge from the hospital or clinic.**
    - The hospital should support co-operation between healthcare professionals and voluntary support groups.
    - Contact telephone numbers and addresses of infant-feeding advisors, community midwives, health visitors, and voluntary breastfeeding counsellors should be issued to all mothers and be routinely displayed in all maternity areas.

Comments:
- Participation of primary healthcare professionals is essential.
- The breastfeeding message to mothers should be identical irrespective of their location (primary, secondary or tertiary care).
- No advertising of breast-milk substitutes, feeding bottles, teats or dummies should be permissible in any part of the hospital.
- The display of logos of manufacturers of these products on items such as calendar and stationery should be prohibited.
- No literature provided by infant formula manufacturers is permitted unless approved by a senior midwife or doctor.

Finally, compliance with the BFHI policy should be audited on an annual basis.

14 Child friendly healthcare
(Standards for the delivery of care and the psychological and emotional wellbeing of children in healthcare facilities)

Sue Burr and Sue Nicholson

The focus of care for health professionals has understandably concentrated on the effective management of specific childhood illnesses. In many parts of the world, the same attention has not been given to the broader psychosocial needs of children and the standard of healthcare delivery. Whilst lack of resources limit clinical activities, **much suffering can be relieved by changes in attitude which acknowledge the rights of children as stated in the articles contained in the 1989 UN Convention on the Rights of the Child (UNCRC)**. The UN Convention has been signed (ratified) by more countries than any other international treaty and provides healthcare staff with principles which if implemented would greatly enhance the care of children.

The Baby Friendly Hospital Initiative introduced by WHO and UNICEF in 1991 has been implemented in over 3500 hospitals in countries all over the world. The Child Friendly Hospital Initiative (CFHI), a programme of Child Advocacy International (CAI) which is supported by UNICEF, the UK Committee for UNICEF and WHO, builds on this and is about to be piloted in six countries. It proposes Twelve Standards of care which all relate to the Articles in the UN Convention on the Rights of the Child. The philosophies within the Standards are not new and are already practised by many individuals, in many countries and in many organisations. They are similar to those in the EACH charter for children in hospital and those put forward in 1996 by NAWCH, now called ACTION for Sick Children, a parent's organisation that originated in the UK.

When, after wide consultation, criteria for each of the Twelve Standards have been finalised, implementation and accreditation tools will be developed. It is hoped that the project will be interactive and that ideas and suggestions for these tools, with examples of good practice, will originate from many different countries.

The Standards place the rights of the child at the forefront of healthcare. They recognise and emphasise that:

- Children are individuals with rights, whose physical and psychological needs are unique (the UN Convention on the Rights of the Child (UNCRC) and the World Summit for Children (1990)).
- Healthcare staff can demonstrate an acceptance of the articles of the UN Convention by incorporating them into their practice and by acting as a role model to others.
- **With improved clinical knowledge and technology there has been a tendency to focus on the systems of the body ignoring the effects of the treatment, including hospitalisation, on the child's emotions and future wellbeing.** Many children are exposed to psychological and physical trauma that is not only unnecessary but may have permanent consequences.
- Admission of a child to hospital has the potential to cause considerable psychological harm and we must adapt practice to minimise the risk.
- Implementation should incur minimal additional finances as they are mostly about changing attitudes, and incorporating those changed attitudes into practice.
- Written policies and training developed to address the issues need to be sensitive to cultural norms and audited regularly.

A policy concerning the philosophies of care within the Standards should be displayed in *all areas* of the hospital/healthcare facility that serve children.

Discussion

Inevitably, difficulties will be encountered in attaining some of these Standards, particularly where resources are limited, staff are overworked and sometimes undervalued with associated lack of morale. The specific recommendations made within the Twelve Standards will need to take account of local cultural norms, concepts about illness and death, and educational practices.

One of the most difficult to achieve will be Standard 10. **Most countries lack the resources needed, or social service structure necessary, to protect children from abuse within the family, even if the legal framework exists. This initiative aims to encourage governments to develop systems and laws to protect children**.

Child friendly healthcare initiative standards

Child = newborn infant, child or young person up to 18 years of age – WHO

1. Children will be admitted to and kept in hospital or other residential institution only when this is in their best interests (*children best at home with family*)

2. The hospital/healthcare facility will provide an internationally acceptable standard of care and treatment (*care*)
3. Child- and family-centred care will be delivered in partnership with parents, in areas dedicated to children and young people that are child and family friendly, by staff with "children's" qualifications, or who are experienced, thus minimising fear and anxiety for the child (*care delivery*)
4. The environment will be secure, safe and scrupulously clean (*safety*)
5. Parents will be kept fully informed and children will be involved in all decisions affecting their care (*communication*)
6. Children will be approached without discrimination as individuals with their own age-appropriate and developmental needs and rights to privacy and dignity (*rights/equity*)
7. The hospital or healthcare facility will have a multidisciplinary team to establish and maintain guidelines for the assessment and control of the physical and psychological pain and discomfort of children (*pain control*)
8. When children are severely ill, undergoing surgery or have been given systemic analgesia and/or sedation, there will always be healthcare staff trained and experienced in the resuscitation of children immediately available (*resuscitation*)
9. Children will be able to play and learn while in a hospital or other healthcare institution (*play/learning*)
10. Healthcare staff will be familiar with the signs and symptoms of child abuse and be capable of instigating appropriate and clearly defined procedures to protect the child (*child protection*)**
11. Health will be promoted by example, education, immunisation, growth monitoring and developmental assessment when a child is admitted to or attending a hospital/healthcare facility (*health promotion*)
12. The hospital or healthcare facility will comply with the appropriate best practice standards on the support of breastfeeding (*breastfeeding*)

** It is recognised that the legal framework required to institute child protection is likely to vary within different countries. In some there may be no framework despite ratification of the UN Convention of the Rights of the Child.

✔ **The pivotal importance of pain control in the management of sick and injured children is at last being recognised.** Standard 6 will aid the appropriate use of pharmacological analgesia and complement this with non-pharmacological strategies such as use of physical comfort, distraction games and the presence of a family member or other trusted carer.

Some health staff will find changing the emphasis from issuing instructions or providing all the hands on care difficult. Encouraging, teaching and supporting parents and older children to fully participate in the planning and provision of their care will be a challenging experience. If a "rights-based child- and family-centred care" is to be achieved globally, **partnership between families and staff will need to be flexible and mutually supportive. Good communication will be essential**. ✔

Healthcare professionals and all healthcare staff working with children have a responsibility to act as advocates for their rights, needs and protection.

Further reading

Southall DP, Burr S, Smith RD, Bull DN, Williams A, Nicholson S. The Child-Friendly Healthcare Initiative (CFHI): healthcare provision in accordance with the UN Convention on the Rights of the Child. *Pediatrics* 2000;**106**:1054–64

www.childfriendlyhealthcare.org

15 Ethical systems within the hospital

Oscar Nunez, William Carroll, Monica Hopkins and David Southall

Introduction

In writing this chapter, we recognise that the majority of countries may not have any choices regarding treatment other than perhaps as treatment versus limited treatment. In its own right this is an ethical issue. There are ethical issues regarding:

- Availability of resources.
- Distribution of resources.
- "Caste" systems and their impact on resources.
- Whether health professionals are permitted or should be obligated to campaign against unethical practices.

One of the most important debates may concern the ethics of giving treatment to palliate symptoms when there is insufficient financial support for therapy, available in other well-resourced countries, to provide a cure.

There is no universally accepted model for what is ethical:

- The "duties of a doctor" should combine with systems which consider the "rights of the patient".
- Each ethical question can be examined under a number of different, but not necessarily exclusive principles.
- The culture and society within which the child lives may have expectations about the relative values of particular ethical principles. Some societies expect a beneficent or non-maleficent approach to healthcare, whereas many Western cultures morally and legally require an overriding respect for autonomy. It is essential that as well as working within professional ethics, doctors respect and abide by the law within the countries in which they practise (providing the law does not harm the child as defined by the UN Convention on the Rights of the Child). **If a law or laws do harm children or fail to protect them from harm, the paediatrician or children's nurse should advocate for appropriate changes.** ✔

Professional ethical codes

Professional medical ethics are almost as old as the science of medicine, having existed in one form or another since the conception of the Hippocratic Oath almost 2500 years ago. *The Declaration of Geneva* (1948, revised 1968 and 1983) is an updated version of the Hippocratic Oath and forms a reasonable basis for what is widely accepted as professionally ethical. It requires the doctor to consecrate his or her life to the service of humanity; to make "the health of my patient" his or her first consideration; to respect his or her patient's secrets (even after the patient's

Table 1.5.1 Medical ethics terminology

Term	Definition
Autonomy	Children, or if below the age of competence, their parents have the right to choose actions that are consistent with their situation.
Beneficence	Doctors must act in the best interest of the individual child.
Confidentiality	Respect the child's privacy.
Informed consent	The process by which a child or parent has received all pertinent information necessary to make a rational, autonomous choice. There are five elements of informed consent: • Disclosure of information • Comprehension • Voluntariness (free of control of others) • Competence • Choice
Justice	Fairness of healthcare delivery.
Non-maleficence	"*Primum non nocere*": above all, do no harm.
Veracity	Truth telling and honesty.

death); to prevent "considerations of religion, nationality, race, party politics, or social standing [intervening] between my duty and my patient"; to "maintain utmost respect for human life from its beginning"; and not to use his or her medical knowledge "contrary to the laws of humanity".

In many countries, there are huge divisions between rich minorities and the mass of the poor, exploited and disadvantaged. Professionals should be aware and acknowledge how their actions support such divisions and aim to provide high standards of healthcare which are independent of a child's family's ability to pay for it.

The ethics of consent

The issue of consent and the different practices enforced by a wide range of different laws across different countries, highlight the importance of ethical consideration of our actions. It is not enough simply to be guided by the law, because the minimum standards imposed by such an inexact and unwieldy machinery can fall well short of what is truly ethical for a given patient in a given situation. The fundamental principle underlying consent is said to be the right of self-determination: the principle or value choice of autonomy of the person. If this is so, how does it apply to those apparently incapable of such decisions? Furthermore, how do the principles of beneficence, non-maleficence and justice fit into our concepts of ethical consent.

"Every patient has the right to make his or her own decision regarding medical treatment and care and, in order to make that decision, is entitled to have full information about the material risks. The clinician's duty is to supply the information in sufficient detail to enable the patient to make that decision."

Definition of consent from "Consent to Treatment" (1993)

"A child who is capable of forming his/her view has the right to express those views freely on all matters affecting the child, the views of the child being given due weight in accordance with the age and maturity of the child."

Article 12 of the United Nations Convention on the Rights of the Child (1989)

"In an emergency, where consent cannot be obtained, you may provide medical treatment to anyone, provided the treatment is limited to what is immediately necessary to save life or avoid significant deterioration in the patient's health."

UK General Medical Council (1998)

The child's competence and parental involvement – respect for autonomy

✔ **The healthcare team and parents must enter a partnership of care whose function is to serve the best interests of the child.** This duty of care also involves respecting the wishes and views of children. Where a child is not competent to give or withhold consent to treatment, a person with parental responsibility must act as an advocate for the child to authorise investigations or treatment which are in the child's best interests. Parents have the right to be involved in the decision-making process and this right is protected in law in most countries.

It is the doctor's responsibility to assess a child's capacity to decide whether to consent to or refuse a proposed investigation or treatment before providing it. A competent child must be able to understand the nature, purpose and possible consequences of the proposed investigation or treatment, as well as the consequence of non-treatment. Competence is presumed at different ages in different countries. Children's competence is related to experience as well as to age and young children can often clearly demonstrate that they have the competence to make decisions about treatment. Such competence has a legal standing in some countries (so-called "Gillick competent" in English law).

Providing the information – an essential component of consent

In many poor countries there is obvious paternalism. How can this be ethically grounded?

In some countries disease and pain are interpreted in terms of sin and retribution. How can health professionals advocate for the child and educate the parents in such cultures?

Ideally before making a decision about whether to undergo a particular investigation or treatment, children and parents require adequate and understandable information about the condition and the options available to them. This information should include details of the diagnosis and prognosis, uncertainties about the diagnosis, options for treatment or management of the condition, the purpose of a proposed investigation or treatment and the likely benefits and probabilities of success as well as a discussion about any serious or frequently occurring risks. Wherever possible the information should be given in a way that is best understood and remembered. This may include information in writing, possibly accompanied by audio or visual aids if available.

Although all this information may be an essential prerequisite for a truly autonomous choice, it may also be truly bewildering and even overwhelming for some patients and families. The principles of beneficence and non-maleficence might suggest that a more paternalistic and less forthright doctor might be behaving more ethically. However it is important not to impose our own views, likes and dislikes about how and how much we would like to be told about an illness or treatment on our patients. A distinction must be drawn between how we would like to be treated if we were a child (or parent of a child) with a particular illness and how that child (and that parent) would like to be treated.

When providing information it is therefore essential that professionals do their best to find out about the child's

(and parent's) individual needs and priorities. This is often the most difficult part of the communication process and involves responding honestly to any questions the child or parent raises and, as far as possible, answering these as fully as possible. It is for the patient, not the doctor, to determine what is in the patient's own best interests. Nonetheless, you may wish to recommend a treatment or a course of action to patients, but you must not put pressure on patients to accept your advice.

Finally, you must consider what to withhold from the patient and their family. You should not withhold information for decision-making, unless you judge that disclosure of some relevant information would cause the child or family serious harm (the principle of non-maleficence). However, in certain settings it may be inappropriate to discuss expensive, unproven or unavailable treatments.

Summary of consent

- Practise within the limits of the law (unless harmful to the child).
- Assess the level of competence of the child before deciding how much and how to tell them.
- Children and families can tell you how much information they need to make a decision.
- Decisions must be free from coercion.

Withholding or withdrawing medical care

A report in 1997 from the Royal College of Paediatrics and Child Health concluded that there are five situations where the withholding or withdrawal of curative medical treatment for a child might be considered:

- The brain dead child (see Appendix 7.9).
- The permanent vegetative state.
- The "no chance" situation – the child has such severe disease that "life-sustaining treatment" delays death without significant alleviation of suffering.
- The "no purpose" situation – although survival is possible, the predicted degree of physical or mental impairment will be so great that it is unreasonable for the child to be expected to bear it.
- The "unbearable" situation – the child and/or family feel that in the face of progressive and irreversible illness further treatment is more than can be borne.

In addition to legal requirements, there are several axioms on which it is possible to base best practice:

- Although there is no significant ethical difference between withdrawing and withholding treatments there are significant practical differences.
- Optimal ethical decision-making concerning children requires open and timely communication between members of the healthcare team, child and family.
- Parents must decide on behalf of children who are unable, for whatever reason, to express preferences unless they are clearly acting against the child's best interests. Cultural practices and religious beliefs may impact on this.
- The wishes (antecedent if known) of a child who has sufficient understanding and experience should be given substantial consideration.
- Resolution of disagreement should be by discussion, consultation and consensus.
- The duty of care is not an absolute duty to preserve life by all means.
- A shift from life-sustaining treatment to palliation represents a change in aims and objectives and does not constitute a withdrawal of care (see Chapter 1.28).
- Doctors should never withdraw treatments that alleviate pain or promote comfort.
- There is a distinction to be drawn between treatment of the dying patient and euthanasia. When a dying patient is receiving palliative care, the underlying cause of death is the disease process (see Chapter 1.28).
- Treatments that may incidentally hasten death are justified but only if their primary aim is to relieve suffering.

Child rights (family rights in the case of young children)

- To participate in developing an individual plan of treatment.
- To receive an explanation of how components of treatment in accordance with the plan will be provided.
- Confidentiality to be broken only under the following conditions: knowledge/suspicion of child abuse, intent of patient to harm themselves or others, court order, a communicable disease which may harm others.
- To receive clinically appropriate care and treatment.
- To be treated in a manner which is ethical and free from abuse, discrimination, mistreatment and/or exploitation.
- To be treated by staff who are sensitive to the family's cultural background.
- To be afforded privacy.

Relationships

The physician–patient relationship

The physician's primary inviolate role is to be an active advocate for each patient's care and wellbeing. He/she should always place the interests of his or her patients first. The physician has a duty to accept ultimate responsibility for his/her medical decisions.

The physician should treat each patient with honesty, compassion, dignity and respect for individual autonomy. The physician should not exclude or discriminate against any patient because of ethnic origin, race, sex, creed, age, socioeconomic status, diagnosis, physical or mental disability or sexual orientation.

The physician's commitment to patients includes health education and continuity of care.

The American Medical Association Code of Medical Ethics contains helpful advice.

Code of Medical Ethics: American Medical Association.

- A physician shall be dedicated to providing competent medical service with compassion and respect for human dignity.
- A physician shall deal honestly with patients and colleagues, and strive to expose those physicians deficient in character or competence or who are engaged in fraud or deception.
- A physician shall respect the law and recognise a responsibility to seek changes in those requirements which are contrary to the best interests of the patients.
- A physician shall respect the rights of patients, of colleagues, and of other health professionals, and shall safeguard patient confidentiality within the constraints of the law.
- A physician shall continue to study, apply, and advance scientific knowledge, make relevant information available to patients, colleagues, and the public, obtain consultation, and use the talent of other health professionals when indicated.
- A physician shall, in the provision of appropriate patient care, except in emergencies, be free to choose whom to serve, with whom to associate, and the environment in which to provide medical services.
- A physician shall recognise a responsibility to participate in activities contributing to an improved community.

The physician–physician relationship

Physicians have a responsibility to maintain moral integrity, intellectual honesty and clinical competence. Physicians should be aware of the limitation of their expertise and seek consultation or assistance in clinical situations in which they are not expert.

Physicians, as stewards of medical knowledge, have an obligation to educate and share information with colleagues, including physicians-in-training. Physicians should be committed to a lifetime of learning and continuously improve the knowledge base and clinical skills relevant to their practice.

The relationship of the physician to system of care

The physician's duty of patient advocacy should not be altered by the system of healthcare delivery in which the physician practises.

Physicians should resolve conflicts of interest in a fashion that gives primacy to patients' interests.

The relationship of the physician to society

Physicians have a responsibility to serve the healthcare needs of all members of society.

Physicians have an ethical obligation to participate in the formation of healthcare policy.

Physicians have an ethical obligation to preserve and protect the trust bestowed on them by society.

Policies and procedures of a hospital ethics committee

Despite the growth of medical ethics and the publication of many professional codes of practice in recent years, there remains no consensus on how individuals, teams, and hospitals might obtain specific guidance in resolving the ethical dilemmas they face. One successfully employed approach has been to establish and use a hospital ethics committee or clinical ethics forum.

- **Function**
  Education: The committee should provide members of the hospital/medical staff with access to the language, concepts, principles and body of knowledge about ethics.
  Policy review and development: the committee can assist the hospital and medical staff in the development of polices and guidelines regarding recurrent ethics issues and questions or problems which arise in the care of individual patients.
  Case review: the committee will be a forum for analysis of ethical questions that arise in the care of individual patients.
- **Appointment and membership**
  Multidisciplinary. It should include the following disciplines: paediatrics, surgery, nursing, social work, pastoral care. It should also include the director of the hospital and chief of paediatric medical staff. A 30% lay membership has been suggested to ensure breadth of perspective and clarity of output. A lawyer experienced in family law can also be helpful.
- **Purpose**
  The jurisdiction of a paediatric ethics committee includes clinical situations involving infants and children <18 years of age.

16 Prevention of hospital-acquired infection

Angela O'Higgins and Sue Nicholson

Minimum standards requirements

Prevention of hospital infection

- Water and sanitation
- Handwashing
- Cleaning materials, soap and disinfectant
- Waste disposal including sharps
- Sterilisers and laundry services
- Isolation facilities

Nosocomial or hospital-acquired infection is a major cost to the hospital and more importantly increases morbidity and mortality in children. It may affect up to 10% of all patients. Nososcomial infection requires a source of microorganisms and a chain of transmission. It is essential that all individual healthcare staff examine their practice to ensure that they are not part of this chain of transmission.

✔ **The combination of using powerful antibiotics and poor hygiene predisposes to the development of antibiotic-resistant microorganisms** which are difficult to eradicate from the environment and difficult to treat.

While children with chronic and debilitating illness are particularly at risk of infection, not all infections are related to their particular disease process but to failure of hospital management to ensure services and facilities, and failure of individual healthcare workers to introduce and adhere to good infection control policies.

Every research study relating to the prevention of infection and cross-infection during the last hundred years has emphasised the importance of hygienic conditions in the whole of the hospital.

The following are essential if infection and cross-infection risks are to be minimised:

- **A good, clean, adequate water supply**. Just as in an emergency refugee camp where water and sanitation are of central importance in the prevention of cross-infection, so is water and sanitation in hospitals, particularly where there are vulnerable infants and children. Running water, both hot and cold is preferable. Hot water should be stored at 65°C, distributed at 60°C and then reduced to 43°C to be used from the taps. This process helps to ensure that waterborne diseases such as Legionnaire's disease are not passed on to staff or patients.
- **Accessible sinks in all areas**. Preferably with non-hand-operated taps, and effective hand-drying facilities.
- **Effective cleaning policies**. The whole of the hospital, including the grounds, should be kept clean. Entrances should screen visitors' shoes for dirt and corridors need to be cleaned at least twice a day with a disinfectant (see below). Floors, window-sills, light fittings and curtains in ward areas need to be kept scrupulously clean but **the priority is the adequacy and state of the toilets and bathrooms. These should be kept scrupulously clean by frequent cleaning and disinfection. Staff appointed as cleaners should be given adequate status and salaries as benefits the importance of the work they are doing.** ✔
- **Effective human and other waste-disposal services**. Human and other waste should be collected and disposed of separately. Foot-operated bins are preferable and frequent rubbish collections. Ideally the hospital should have its own incinerator.
- **A laundry service**. All bedding/curtains/towels/flannels must be regularly washed with a detergent/disinfectant. Industrial quality washing machines are essential in every children's unit.
- **Strict hand-washing policies**. Viruses and bacteria can survive on hands for two to three hours. **Correct hand-washing technique for all staff, visitors and patients is the most important factor in the prevention of cross-infection.** ✔ This is easily taught and frequently an improvement in practice is demonstrated in the short term. However, when examined over a longer period of time, old habits and shortcuts reappear. Good hand-washing techniques are dependent on adequate supplies of clean water, ideally elbow-operated taps, a liquid soap supply and an effective method of hand-drying. Where it is impossible to provide liquid soap and paper towels some ingenious solutions have been attempted. Bar soap suspended in a net bag over the sink area and individual cloth towels for each child, changed every 24 hours or at the discharge of the child and kept within their bed space can be effective. Added emollient protects hands from chafing. Antiseptics can be added to liquid soap to improve antimicrobial activity. Chlorhexidine is a cheap effective antiseptic that is widely available

throughout the world. However, there is no good evidence that this increases the effectiveness of hand-washing substantially. Antiseptics should be used before invasive procedures and where there is heavy soiling with potentially contaminated body fluids or other human waste. Povidone-iodine should be reserved for use as a surgical scrub.

When running water is not available or hand-washing is difficult, 70% Alcohol Gel is useful. This is a new but fairly expensive product which has a significant part to play in prevention of introduction of cross-infection in high-risk areas. When rubbed on and allowed to dry, it will disinfect hands. After initial conventional hand-washing it can be used between each patient contact, but further hand-washing is still recommended after every five or six rubs.

All the above-mentioned items may be regarded as a considerable extra cost for a health service but are cost saving when balanced against an increase in hospital stay due to infection, the additional medications and sometimes unnecessary deaths.

All staff should have a personal responsibility for hygiene but every children's unit should also identify someone (ideally a nurse with the support of a microbiologist) to be responsible for the education of all staff in techniques that will prevent the spread of infection, particularly good hand-washing and drying. This education programme will need to be ongoing, as even in the best centres, these programmes are effective for relatively short periods of time. The organisation needs to support this staff member to reinforce that all grades and members of staff have responsibility for their practice (especially senior doctors who should act as role models). Additionally, it needs to become the norm for this identified staff member, no matter how junior, to be recognised as the expert in his/her unit and anyone who is asked to carry out hand-washing must instantly comply and accept the damage to their pride.

- **Disposal of body fluids**. Each ward or unit needs an area set aside for this purpose. This area, and all the equipment it contains, needs to be kept scrupulously clean and body fluids disposed of quickly with spillage removed instantly. When there is likelihood that there is a danger of body fluids being contaminated with life-threatening organisms, additional precautions should be taken. After hand-washing, disposable clean gloves should be used by all staff and parents who will be assisting with the toileting of children. Care must be taken with sharp objects such as hypodermic needles to protect the child, their family, other unit visitors and staff. An empathetic approach is necessary to ensure the child and family does not feel labelled and unworthy of due care and attention.
- **Cleaning, disinfection and sterilisation of equipment and furniture**. Manufacturers' instructions must be followed for individual items of equipment. These will usually clearly state which items need sterilising and where disinfection will surfice. They will also indicate appropriate dilutions for disinfectants. All equipment should be cleaned before being sterilised or disinfected.
- **Sterilisation** is the complete elimination and destruction of all forms of microbial life. This is frequently achieved by steam under pressure, dry heat, gas and liquid chemicals. A system providing this must be available in every children's unit and such systems are also required for instruments and towels used in the operating theatre.
- **Disinfection** is a process, which eliminates many microorganisms with the exception of the most resistant endospores. It is usually accomplished using liquid chemicals called disinfectants. Hypochlorites are inexpensive and effective disinfectants. They are active against most microorganisms including HIV and hepatitis B. However, they do have a corrosive effect on metals and if used on fabric or carpet can bleach out colours. Hypochlorites in dilution (usually 0.1% solution) are contained in household cleaners available in markets throughout the world for domestic use. These household cleaners can be used in the hospital environment for general cleaning but stronger solutions need to be available particularly for the disposal of body fluids and for cleaning following outbreaks of notifiable infections. A 1% solution is recommended for the treatment of blood and body fluid spills and 0.1% solution can be used for all surfaces. Hypochlorites are available as tablets which make the process of dilution easier.
- **Cleaning** is often the most neglected of the three processes and must precede sterilisation and disinfection. When undertaken using a disinfectant detergent, cleaning alone will effectively reduce the number of microorganisms and allow items which come into contact with intact skin to be safe. These items include, blood pressure cuffs, bedrails and intravenous poles.
- **Isolation of children with specific infections**. For isolation procedures to be effective they need to be instituted early. Two or more children with the same infection can be isolated together. Differing isolation techniques will be needed and the use of gowns, gloves and masks can at times be necessary (see Chapter 4.23). In some cases nursing the child in a cubicle or single room until medical tests are complete is all that is required. When there is a need for gowns, gloves and masks, these will require frequent changing and washing to ensure their efficacy and to be used by everyone who comes in contact with the child, medical personnel and carers. Ideally they should be used once only and must be removed and discarded or sent for laundering on leaving the isolation area. An area will need to be set aside for changing, with supplies of gowns, gloves, aprons and masks. Gowns made of cotton material will need to be worn with plastic aprons. Children's compliance with isolation techniques will improve if the element of fear is removed. This can be achieved by all personnel allowing the child to see their face (through a window) before the mask is applied.
- **Infection control measures following the death of a child**. When a child dies the amount of time the parents and other family members are able to spend with the child will vary according to the facilities that are available. Rituals and beliefs concerning the death

of an individual and the management of the body, usually involve religious or cultural observance. There are many beliefs surrounding the distinction between physical and spiritual life, and that something of the individual survives death, either to be reborn through reincarnation or to fulfil its spiritual destiny in the afterlife. In most religions and cultures it is important that the correct funerary procedures are followed, in order to assist the passage of the dead person from this world to the next. All societies, religious or not, have to deal with the problem of the death of their children and the bereavement of parents and other close family members. Like other transitions in an individual's life, death is usually marked by a rite of passage in which central values are restated and important social bonds re-emphasised. Precise customs vary in different religions and traditions, but common features include the washing and laying out of the corpse (which may be embalmed), the wake, or watching over the dead body. These customs may need to be modified to prevent the spread of infection to other members of the community or because of the need to carry out postmortem examinations to establish an exact cause of death. Good hand-washing procedures remain of paramount importance.

In countries where extremes of temperature are a part of the climate, then refrigeration for dead bodies until they can be returned to the family is essential. Each hospital should have a mortuary building adjacent to, but separate from the hospital. To prevent the spread of infection, those working in the mortuary will need to be provided with separate clothing for use in the department. The use of two pairs of gloves, or thick rubber gloves and protective clothing will be necessary for the postmortem examination of bodies where there is a suspicion of life-threatening bacteria or viruses.

This department will need to have facilities for families to see and spend time with their dead child and a separate comfortable area where documentation can be completed and to facilitate any necessary interviews with local government officials. This department can also provide facilities for postmortem examination. In large centres this department can also be part of government facilities for forensic postmortem which may provide additional resources for this hospital. Having these centres within a hospital may improve services for families but care needs to be taken that there is a culture of openness involving parents in the consenting procedures for all examinations undertaken after death.

Conclusions

Each member of the hospital has a role to play in the prevention of nosocomial infections. The greatest responsibility lies with the health professionals, particularly nurses and doctors, who in the hospital setting are in constant contact with the different children and families over the complete 24-hour period and because of this are the main perpetrators for cross-infection. However they can also show good practice by example, be the catalysts for change and for improving the education of other hospital personnel and families. In the presence of very poor resources, parents can be asked to keep toilets and washing areas clean. They will need appropriate disinfectants, clean water, and basic equipment.

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17 Nursing and sick children

Andrew Clarke

Minimum standards requirements

Nursing

- Input/output charts
- TPR, blood pressure, O_2 saturation charts
- Handwashing
- Uniforms
- Morale and payment
- Continuing education

The role and status of nurses, together with the education they receive varies. Many countries have no specific training in the nursing of sick children. Although good care is often delivered, **optimum care can only be consistently attained by education and professional accreditation that relates specifically to the holistic physiological, physical and psychosocial care of the child and family.**

The broad and increasing demands of healthcare results in an overlap of roles undertaken by nursing and medical staff. In many situations, the nurse will be the most experienced or skilled in a certain procedure and for much of the time, especially at night, be the leading healthcare professional present.

Hospitalisation often represents a frightening experience in an unfamiliar environment where control over everyday lives has gone. Nurses should aim to return much of that control back into the hands of the child and family and work to ensure that they receive the best care possible, given the environment and resources at their disposal.

The potential benefit and contribution of parents cannot be overestimated. **If the child's family is available, they should be encouraged and supported to participate in care.** Parents provide valuable information about what is normal for their child and how they may have changed. Since they know their child better they are often the first to notice small changes in his/her condition that later may prove significant.

Nurses are central to communication between the multidisciplinary team and the family. They have a key role in being an active advocate for the child, although this is often easier to say than to do. The hierarchical cultures in some healthcare systems can conspire against this, often to the detriment of the child. It is the responsibility of all healthcare professionals to promote an environment where the views of all involved in the child's care, especially that of the child and their parents, are heard.

Inappropriate and needless painful interventions and investigations, long hospitalisation without good reason and unnecessary separations from family cannot be justified. In representing a place of safety, it is the responsibility of every healthcare facility to facilitate this ideal.

Children are unique, both as individuals and as a group, with rights enshrined by the UN Convention on the Rights of the Child 1989 that underpin the standards of care outlined in Chapter 1.1. They are not "small adults" and have physical, psychosocial and physiological needs and responses that are different. Effective nursing requires knowledge of these differences (recording vital signs means little without understanding their importance).

The spectrum of nursing care is vast, and ranges from meeting basic human needs to complex case management. However, much can be attained by a skilled "child friendly" approach that leads to minimal intervention and trauma and includes the family and where appropriate the child in decision-making.

As part of providing a minimum standard of care, children's nurses should receive education in the following:

- Child growth, development and basic psychology
- Communicating with children (including the role of play) and families of sick children (to include listening skills)
- Children's physiology/normal ranges for vital signs and basic laboratory values
- Children's rights
- Resuscitation
- Pain assessment and management
- The special needs of children with disability/learning difficulties
- Immunisation
- Recognition and management of non-accidental injury and abuse
- Basic pharmacology, calculation of drug dosages and the safe administration of drugs and fluids

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18 Other "work-related" issues for healthcare staff

Angela O'Higgins and Sue Nicholson

Attention to the following issues will improve healthcare delivery

Morale

- The care of sick and disabled children can be fulfilling and stressful. **Maintaining good staff morale is essential: all healthcare staff should be valued, supported and know their roles and responsibilities.**
- Multidisciplinary teams in which leadership moves from professional to professional, depending on the problem, utilises specific skills effectively and further enhances feelings of worth.
- Individual job descriptions and responsibilities should be agreed between professionals, their professional organisations and hospital management.
- Adaptation to the needs of children and their families leads to the provision of efficient services.

Responsibilities

- Care for children must be available 24 hours a day every day of the year.
- All staff need to remain vigilant in the prevention of cross-infection and personal hygiene.
- Staff numbers should be calculated according to the dependency of the children admitted using country norms for different categories of care.
- Parents who contribute to care should ideally be additional to professional staff numbers.
- All healthcare staff, particularly professionals, have a responsibility to act as advocates for the child and family, particularly when differences of opinion arise concerning management. Nurses through their constant contact with the child and family are ideally placed for this role.
- Communication with children and their families should be at a level appropriate to their understanding, in their first language (via an independent interpreter if possible) and given empathetically, respecting privacy and confidentiality.
- Ideally treatment plans should be made in partnership with the child and family.
- Continuing education is a professional responsibility following qualification as a health care worker (see Chapter 1.10).

Ward rounds

- Although regular ward rounds can be an excellent way for all staff to meet with the child and family to discuss progress and potential treatment in partnership, numbers present at a bedside should be limited to avoid fear and anxiety and decrease the risk of potential cross-infection. Privacy and confidentiality must be respected at all times.
- All children who require hospitalisation should be assessed by a multidisciplinary group, not necessarily at the bedside, at least daily.
- In acute areas (intensive and high-dependency care) there need to be at least three rounds daily, ideally once every eight hours, preferably to coincide with staff changes; plans of care can be decided at these times.
- All concerned with care should have the opportunity to make a contribution.

Meetings

- Regular meetings should discuss individual patients to debrief following death/clinical incidents and to audit particular aspects of clinical and unit management such as infection control.
- Such meetings need to be educational and must not be used to apportion blame.
- The outcome of audit, particularly any changes of practice, need to be available to those staff it affects. Written guidelines need regular review following related audit and new international and local recommendations.

Staff problems

- Concern about individual performance should be addressed by a senior staff member, preferably following

written guidelines, on a one-to-one basis and a period of supervised practice and/or retraining organised.

- Ideally staff with health-related problems affecting their performance need to have access to a supportive occupational healthcare system. There should be systems in place to protect patients from staff who are ill; this is a difficult but vital issue particularly with respect to illnesses such as tuberculosis, HIV infection and hepatitis.

19 Fundamental needs of sick children and their families

Paula Blurton, Angela O'Higgins, Andrew Clarke and Janet Vickers

Although children may sometimes require complex treatment, there are basic needs that will always need to be met. Unfortunately these are sometimes viewed as being of secondary importance, but often this care reduces the need for further intervention and can enhance other therapies (for example, the beneficial influence of improved nutrition on wound healing and the adverse effects of stress on respiratory function). Where therapeutic options are limited due to availability, finance or perhaps where no further curative treatment is possible, the provision of good basic personal care is particularly relevant if not paramount. It is invariably viewed as part of the nursing role. However, the underlying approach required is common to all healthcare professionals and there should be an understanding that its provision is rarely a simple task. It requires understanding, skill and patience, and is a subject that justifies a separate or expanded publication. Only the bare essentials are covered in this text.

Hydration and feeding

Children can quickly become dehydrated particularly in the event of diarrhoea, vomiting, fever or when they are too tired and lethargic to drink. Any caregiver should be able to recognise the observable signs of dehydration (see below and Chapter 3.25 for signs and symptoms).

A quick response to a child who either is or is likely to become dehydrated can prevent a further deterioration and the need for intravenous fluids. ORS powder to be mixed with clean water (boiled and cooled) should be available in all hospitals, but in its absence the following will be suitable:

- Into 1 litre of boiled (and cooled) water add:
  - $\frac{1}{2}$ teaspoon of salt (solution should taste no saltier than tears)
  - 8 teaspoons of sugar/honey.
- If available also add:
  - $\frac{1}{2}$ cup of orange juice or mashed banana (potassium source)
  - $\frac{1}{4}$ teaspoon sodium bicarbonate (baking soda)
- Dhal water, soups, rice water, bean broth and thin porridge cereal are also good.

Trying to get a child to drink is not always easy:

- Encourage children to drink small amounts and often.
- Give a good explanation of why it is important – especially to older children.
- Involve children and decide together how much they will try to drink each hour.

For small children or children too tired to drink by themselves try:

- Using a small cup, spoon or syringe.
- Encouraging them to play/participate by using the syringe themselves.
- Using small rewards.
- Praising when fluid is taken.

Children often lose their appetite when they are sick. In short acute episodes, the main priority is for fluid.

Table 1.9.1 Dehydration

Parameter	Mild	Moderate	Severe
Mental state	Alert/thirsty	Restless/lethargic/thirsty	Drowsy/cold/sweating
Skin turgor	Normal	Absent	Absent
Breathing	Normal	Fast	Fast
Pulse	Normal	Fast	Fast, weak, may be unpalpable
Blood pressure	Normal	Normal/low	Very low/may be unrecordable
Urine	Normal	Reduced/concentrated	Very little/none
Eyes	Normal	Sunken	Very sunken
Fontanelle	Normal	Sunken	Very sunken
Tears	Present	Absent	Absent
Lips/mouth	Moist	Dry	Very dry

Adapted from J Insley (1990) and S Ryan and E Molyneux (1996).

However, in longer periods of sickness, it is essential to ensure an adequate nutritional intake.

Anxiety caused by separation from family can cause children to lose their appetite and when placed in an unfamiliar environment such as a hospital, a choice over whether or not to accept food or drink may represent the only control the child retains.

Feeding difficulties often cause distress to parents for whom providing nourishment is a very important part of care. This can be the case for parents of a child that is dying, where they may feel that a lack of nutrition will contribute to or quicken their child's death. These anxieties should be understood. It should also be explained that loss of appetite is sometimes part of the deterioration process and the child should be encouraged to eat what they want.

It is important to recognise and manage other factors that have an effect on oral intake such as sore mouth, nausea, vomiting and constipation.

- Provide familiar food for the child and let the child choose the food if possible.
- Encourage children to feed themselves when able and parents to help.
- Avoid invasive procedures immediately before or during the meal.
- Try small amounts of food often, not two or three large meals a day.
- Avoid foods that are highly spiced or have strong smells unless that is the cultural norm.
- Give food at familiar times for the child and try to make it fun.
- Praise children when they eat but do not punish when they cannot.

Keep an accurate, written, record of all fluid/diet that a child takes and also their output (urine, stool, vomit). See Appendices 7.6, 7.13 and 7.14 for examples of charts. Compare the two total amounts over a period (usually 24 hours) and add an amount for insensible losses through perspiration and breathing (approximately 15 ml/kg/day; more in the case of fever or hot environment). Alongside clinical observations, this will give a good indication of a child's hydration level and will also give warning of a child who is becoming dehydrated. In reality, written records are often inaccurate and frequent weighing (for example once each day at the same time) provides a valuable guide to fluid balance.

Elimination

For the management of constipation and diarrhoea see Chapters 3.31 and 3.25.

The elimination habits of children vary with the individual but are often changed when sick or in hospital. There are many reasons that can cause this, including the disease process, surgery, injury and medication. Anticipating problems can do much to help. However, awareness of quite basic issues is always important:

- Maintain an adequate level of hydration.
- Obtain information from the family about the child's normal elimination pattern.
- Children can be too frightened to go to the toilet in a strange place – reassure.
- Pain (for example from a urine infection or anal fissure) may cause a child to retain and deny the need to go to the toilet. Analgesia and simple measures such as sitting the child in warm water can help ease the pain and encourage urination or defaecation.
- A child that is passing bloodstained stools may be frightened and need reassurance.
- Praise and encouragement are important and effective.

Personal hygiene

This can contribute significantly to the way a sick or dying child feels, and can prevent further problems such as sores at pressure areas. Attention to personal hygiene needs should always be performed in a manner that protects the dignity of the child. Even very small children often feel shy or uncomfortable when being attended to by an unfamiliar person and this is particularly the case with older children and teenagers. It is another very good reason for involving and encouraging parental involvement in the care as well as helping to fulfil many parents' natural wish to participate.

When washing, extra attention should be paid to skin folds, the neck, the back, the ears and the genitalia, and children should be encouraged to do as much as they are able. Children that are malnourished, have been sick for a long time or who have malignancies can have very fragile skin that can easily break down and require special attention. An effective method that can be undertaken by both professional and family carers is to gently change the child's position at frequent intervals. This relieves the pressure on any given part and prevents reddening and breakdown of the skin. Pain relief should be given to prevent discomfort (see Chapters 1.27 and 1.28) and where available, ***pressure-relieving mattresses are also helpful***. Good mouth care is very important and children should be encouraged to maintain this when in hospital. For children who have not brushed their teeth before, this is a good opportunity to start. Help needs to be given to those unable to achieve this themselves.

In children that are very sick or who are dying, mouth care can help prevent many problems such as bad breath, bleeding, infection, ulceration and pain that can significantly add to their suffering. For these children the following can help:

- Using a soft toothbrush or mouth sponge to clean a child's teeth and mouth regularly.
- Flavoured ice cubes or mouth swabs if a child's oral intake is low.
- Vaseline and lip balms are useful for dry lips.

Communication and comfort

Hospitalisation often represents a traumatic change in the life of a child. Much that was previously reliable in their lives has gone and been replaced by an unfamiliar environment and fears of an uncertain immediate (and

perhaps long-term) future over which they have little control. Our words and expressions convey a stream of messages to those around us. However, we often have little understanding of how we are perceived. With skill and thought, effective communication plays a vital part in the care of any sick child and has a dramatic impact on their experience and subsequent response.

Both the child and family have a need to trust those that are caring for them, to know what is wrong and also what is going to happen to them. It is a major factor in helping them adjust to the situation, develop coping strategies and make decisions about their own care.

The "information needs" of children are often neglected, sometimes on the pretext that their understanding is limited. However, even young children have a need for information in a language understandable to them. In the absence of reliable information, a child's fantasy may well be far more distressing than the reality.

These issues are also very important in preparing a child (and their family) for a procedure. They need to be told truthfully and sensitively what will happen, particularly if it might be painful. If not warned, a child's trust in those around him/her can be destroyed, future procedures will be feared and anxiety increased.

Communication is a two-way process, involving both the conveying of the message effectively and an understanding of the thoughts and feelings of the other person. To do that requires an ability to actively listen (so that the person *knows* you're listening), and to interpret non-verbal signs that often tell a different story to what is actually said. Children need to express their worries and anxieties. Young children do not always have enough language to convey how they feel, so play and other activities such as drawing are useful ways to gain expression and give control.

Factors that may appear trivial to an adult may be extremely important to a child. There may be issues causing them distress that can be easily resolved and, in cases where there is not a solution, the sense of isolation often felt by these children can still be helped by having someone to share their anxieties with.

Where the situation is grave, families sometimes find it difficult to discuss distressing subjects with their child. Although these are hard and emotional situations, even very young children are extremely perceptive to the distress and anxiety of their family. They often have a much greater level of awareness than is realised and not to acknowledge the reality of their position may make their feelings of isolation worse by denying them the opportunity to express how they feel or ask questions.

Much of the above takes time, which may well be in short supply (another reason for encouraging parental involvement) so it is important to make the little time you may have with a child count. **The way you approach, talk to and touch a child can make a big difference to the way a child feels. It helps to build trust and influences their compliance. Touch is a powerful tool and can convey more than words in terms of comfort and reassurance.** Attending to their personal needs may unfortunately be one of the few times during the day that a child has the opportunity for human touch so it is important to be kind, gentle and thoughtful in your approach. ✔

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http://mailbase.ac.uk/lists/paediatric-nursing-forum/

110 Continuing medical education (CME) for healthcare professionals

John Bridson

Minimum standards requirements

Continuing medical and nursing education

- Pocket manuals
- Evidence-based materials
- Postgraduate medical centre including library
- Internet access if possible

A manual available at all times is one of the best ways of accessing up-to-date evidence-based information. Every healthcare professional needs continuing medical education (CME) to keep up with the pace of change. How? You may be a long way away from a university. You may have no library within reach. You may not be sent any journals to read. You may not be able to go away for further education.

The solution is a postgraduate education centre that can be accessed by all healthcare staff. This education centre could, as a minimum, consist of a reasonably sized, comfortable room containing a library and if possible at least one computer. This could be the area where regular departmental meetings can be held. All non-governmental organisations (NGOs) are concerned with sustainable development and most of them regard education as an issue requiring major investment. If you are unable to obtain funding from your health service, cultivate a relationship with an NGO or similar organisation. Try to gain investment from them or other sources.

- The library should contain the basic textbooks, as up to date as possible.
- If possible, subscriptions for the major paediatric journals should be obtained or accessed online.
- It may be that in your language no significant textbooks are published. English is the major international scientific language. It seems reasonable to concentrate on English language texts.
- If your centre has an electricity supply, if you are able to obtain a computer with CD ROM, and if it is possible to install a telephone line, then e-mail allows you to communicate with specialists in advantaged countries and the Internet can provide up-to-date information on diseases and their treatments. Computers are now being designed only for the Internet, these are less expensive than the standard PC. This investment, for the price of perhaps six good books, can make available a mass of up-to-date information. A subscription is required in most countries for access to the Internet and there will be telephone line usage charges. These charges will need funding. English is likely to be the language of choice for your global communication.
- Security may be a problem. The education centre, if developed, will need to be designed with this in mind. You do not want books and computers stolen.

Continuing medical education takes many forms. Many departments will need regular meetings. Cases can be presented and discussed. A journal club can be organised. It is a good idea for each department to hold one of their weekly meetings in English.

The Internet has many millions of pages. Below is a list of a few websites, which will give you an introduction to online Child Health information

Child health on the Internet
http://www.emmadw.ukgateway.net/

PIER: Paediatric Information Education resource
http://www.pier.shef.ac.uk

International Paediatric Association (IPA)
http://www.ipa-france.net

Hong Kong College of Paediatrics
http://medicine.org.hk/hkcpaed/home.htm

The American College of Paediatrics
http://www.aap.org/default.htm

The Canadian Paediatric Society
http://www.cps.ca/english/about/about.htm

Regional and General Paediatricians Society (RGPS) the Royal Australasian College of Physicians
http://www.racp.edu.au/rgps/index.htm

British Paediatric Surveillance Unit
http://bpsu.rcpch.ac.uk

Archives of Disease in Childhood
http://www.archdischild.com/

Developmental Medicine and Child Neurology
http://www.cup.cam.ac.uk/journals/jnlscat/dmc/

Ambulatory Child Health
http://www.radcliffe-oxford.com/ach.htm

Pediatrics
http://intl.pediatrics.org/

European Society for Paediatric Research
http://www.espr.dk/

Neonatal and Paediatric Pharmacists (NPPG)
http://www.nppg.demon.co.uk

OMNI: Organising Medical Networked Information
http://omni.ac.uk/

OMNI is the UK gateway to quality biomedical and health information on the Internet, maintaining a searchable directory of reviews and links to quality Internet-accessible resources. OMNI also provides access to several more focused biomedical resource databases, as well as services such as training, education, and Continuing Medical Education and biomedical news.

The Institute of Child Health of London
http://www.ich.ucl.ac.uk

British Medical Journal
http://www.bmj.com/

Systematic Revue Training Unit
http://www.ich.ucl.ac.uk/srtu/homenofr.htm

OMIM (genetic syndromes)
http://www3.ncbi.nlm.nih.gov/Omim/searchomim.html

Text books on-line: Pediatrics
http://www.emedicine.com/ped/index.shtml

Bibliography of on-line texts,
http://www.drsref.com.au/books.html

CME Virtual Hospital
http://vh.radiology.uiowa.edu/

Pediatric *X* rays
http://www.uhrad.com/pedsarc.htm

W3EMRS
http://www.emrs.org/medweb/

American Academy of Pediatrics
http://www.aap.org/

Facts for Families
http://www.aacap.org/info_families/index.htm

British Society of Paediatric Endocrinology
http://bspe.shef.ac.uk

ISPAD
http://www.ispad.org

Pediatric evidence-based medicine
http://depts.washington.edu/pedebm/

BASPCAN
British Association for the Study and Prevention of Child Abuse
http://www.baspcan.org.uk

Child protection interest group
http://www.cpig.co.uk

Child Trauma Academy
http://bcm.tmc.edu/cta/

General paediatrics
http://www.generalpediatrics.com/

National Electronic Library for Health
http://www.nhs.uk/nelh/

Search Yahoo health sites http://dir.yahoo.com/Health/

Acknowledgements

Sites from various sources.; Royal College of Paediatrics and Child Health web site, Jerry Wales, Lavleen Chadha

1.11 Essential imaging facilities

Kieran McHugh

Minimum standards requirements

Imaging

- Simple radiography (WHIS-RAD)
- Ultrasound
- (***Neuroimaging if available***)

It is over a hundred years since *X* rays were first discovered. However, more than half of the world's population has no access to any form of radiographic examination. In the context of severely limited resources, health planners need to be selective in their choice of imaging technology. Some radiographic equipment is so expensive that its purchase would be to the detriment of other important components of a basic health service. ✔ **Nevertheless, simple radiographic and ultrasound equipment should be available in every institution that merits the title of Hospital.**

- The majority of studies are plain radiographic examinations. In a small rural or suburban hospital plain radiography will account for 90% of all the necessary examinations and where available ultrasound would satisfy most of the other 10 %.
- There is little point in expensive imaging equipment (which is difficult to maintain) if there is no expertise locally to treat the conditions diagnosed. A computed tomography (CT) scan of the brain, for example, is worthless in the absence of a neurosurgeon to treat the traumatic cerebral injuries, abscesses or tumours diagnosed.
- Good diagnostic imaging frequently leads to less hospitalisation, allows for quick and accurate diagnosis, and results in less suffering and pain. WHO recommends that small hospitals and clinics with only one doctor need imaging equipment. Access to simple radiography should be the right of any child when his or her doctor believes it would help with more accurate diagnosis and treatment.
- Access to at least the following should be available to all sick and injured children.

X ray equipment

- Radiographic equipment that is easy to operate and maintain, as defined by the World Health Imaging System for Radiography (WHIS-RAD), is based on much practical experience and is ideally suited to radiology departments in disadvantaged countries.
- WHIS-RAD was fully specified in 1995 and has replaced the WHO – Basic Radiological System (WHO-BRS). It is noteworthy that some manufacturers continue to market substandard so-called BRS machines that do not meet the WHO specifications and have not been evaluated by the WHO Collaborating Centre in Sweden.
- A list of the machines and manufacturers that meet the specified requirements is available from any WHO office.
- WHIS-RAD is suitable for use in children and small infants.
- Radiation doses from a WHIS-RAD unit are lower than many conventional radiographic machines which is particularly appealing where standards of radiation protection, use of cones, lead protection and dosimetry may be variable and often non-existent.
- The *x* ray generator specified for WHIS-RAD may be used with almost any power supply, however variable.
- Patients can be examined standing, sitting or in a recumbent position.
- WHO have produced two manuals that make the equipment easy to use, even by those with no formal training in radiography (manuals of dark room and radiographic techniques). Operators can be trained in a matter of months.
- A WHIS-RAD unit is easily applicable to children, is surprisingly inexpensive, is safe for patients and operators and produces high quality images.
- Donation of old but functional radiographic equipment from rich countries, although laudable, is often worthless. Bulky out-of-date equipment frequently cannot be installed, operated nor maintained. The service manuals are often missing and spare parts are a major problem.

A comprehensive list of the indications for diagnostic radiography in children is too long to include here. Suffice it to say that all children with a serious pneumonia or suspected tuberculosis, for example, or a fractured limb merit radiography. In practice, chest and skeletal examinations are the most frequent indications for diagnostic imaging in all parts of the globe.

Ultrasound

The wide range of applications of ultrasound in children, its versatility and safety probably make it ✔

better suited to disadvantaged countries than any other imaging modality. As far as we know, sonography is harmless – it does not generate ionising radiation thus making it particularly suitable for imaging children.

- Ultrasound machines are simple to operate but the images are also as easy to misinterpret.
- It is critically important that the person performing and interpreting ultrasound studies in children is suitably trained and competent (usually a doctor). **Sonography must be taught on a supervised practical basis in a local environment.** This training must include paediatric scanning. WHO recommends a six-month minimum training for diagnostic sonography.
- WHO recommends minimum specifications for a general purpose ultrasound scanner.
- The scanner should be able to operate from the local electrical power supply.
- Servicing should be available locally.
- It must be possible to store the unit safely under adverse conditions.
- When scanning children, a 5-MHz transducer is desirable.
- Doppler techniques do not generally warrant extra expenditure, as vascular diseases are uncommon.
- A permanent, hard-copy record is recommended for patient follow up in particular and in the interests of teaching/training in general.
- Mobile ultrasound scanners can be operated at the bedside or in the Accident & Emergency department for the most severely ill children.
- Abdominal and pelvic ultrasound where available have a well-established role in the assessment of the paediatric abdominal emergency. Sonography can quite simply resolve mass lesions from organomegaly and can be a useful tool in guiding interventions (aspiration, drainage or biopsy).
- Ultrasound studies frequently reduce the need for plain abdominal radiographs.
- Sonography of the infant brain is possible and can provide useful information in the febrile or unconscious patient.
- The role of ultrasound in the paediatric musculoskeletal system is steadily expanding.

1.12 Essential operating theatre resources

Leela Kapila, Devendra Kumar Gupta and Anupam Lall

Design and contents of operating theatres (OT)

- Ideally located in the most inaccessible area of the hospital to reduce the chances of infection.
- Should be of adequate size (minimum 7 m × 7 m) for the placement of essential equipment and the unobstructed movement of staff.
- It should not be used for storage, for which a separate side room should be available and also used for handwashing.

Essential equipment

- Ordinary OT table with a facility for the lithotomy position and lowering and raising the height of the table.
- A good focussing OT light is very important.
- ✔ **A simple Boyle's apparatus with an uninterrupted oxygen and nitrous oxide supply is the most essential equipment for the anaesthetist. Reserve cylinders for both oxygen and nitrous oxide should always be there.** If nitrous oxide is not available, then the patient can be maintained on ether or halothane but the level of anaesthesia has to be deep requiring more intensive postoperative monitoring.
- Suction machine (both electrical and manual functions – in case of electrical failure) should be periodically emptied and cleaned with antiseptic solution. It must be constantly checked.
- Fumigation machine is essential for the sterilisation of the OT.
- Anaesthetic equipment and supplies (see Chapter 1.13 for list of essentials).
- All emergency drugs like lidocaine, epinephrine, atropine, sodium bicarbonate, 25% glucose, morphine, etc. should be readily available in the OT with syringes (again see Chapters 1.13 and 1.17 for full list).
- A boiler is essential for sterilisation if an autoclaving facility is not available. A heater of some kind is also essential for warming up saline to be used during surgery to prevent hypothermia.
- Monitoring equipment: see Chapter 1.17 for essential list.
- Room heaters are essential especially for surgery on infants. The OT temperature should be in the range of 28–32°C to prevent hypothermia in babies. Hot-water bottles can provide heat to the infant and are inexpensive. Radiant warmers, incubators and electric blankets are helpful if available.
- A cautery machine is essential for reducing blood loss during surgery. An ordinary unipolar cautery will suffice for most procedures. Heating a probe red-hot using a bunsen burner can provide thermocoagulation on touching the bleeding sites. It is a low cost-effective method when a cautery machine is not available.
- Instruments. The minimum instruments required for minor surgery are:

Artery forceps	
Mosquito	6
Kelly's	6
Towel clips	
Bulldog	6
Scissors	
Metzembaum's	1
Mayo's	1
Thumb forceps	
tooth	1
non-tooth	1
Intestinal clamps	
non-crushing	2 (4)
Martin artery forceps	2
Right-angled forceps	1
Needle holders (paediatric) which can hold 3.5 to 5.0 sutures	2
Retractors	
Right-angled	2
Zerneys	2
Devers	2
Malleable	2
Suction tip	1

- Other instruments for specific surgical procedures include:
  Retractors for chest size (Finochieto)
  Retractors for oral surgery (Boyle-Davis mouth gag)
  Retractors for urological procedure (Denis-Browne retractor)
  Bone instruments and dental scalers may be required for rib resection and palate surgery.

OT personnel

Apart from the paediatric surgeon, an adequately trained anaesthetist (preferably with paediatric experience) is essential.

- Nursing staff should be adequately trained in the care and handling of instruments and equipment in the OT. They should be made responsible for the proper functioning of all equipment and trained in the sterilisation of the OT and the instruments used.
- OT assistants are important for moving patients to and from the ward. They should be aware of the function of the equipment in the OT. They should be counselled about the hazards of contact with blood and other patient secretions, especially an awareness about AIDS and hepatitis B.
- OT cleaners should also be aware of the threat of these communicable diseases. It is essential to clean the OT between every case to prevent nosocomial infections.

Procedures and practices to reduce the chances of infection in the OT

- The floors, walls, table and all equipment in the OT should be cleaned and disinfected at least once a day and after every case involving infection.
- Autoclaving is the standard method of sterilisation but if not available boiling for 1 hour should be done.
- Spirit flaming of the instruments (all instruments are placed in a kidney tray and spirit is poured in it and a matchstick is used to flame it) can be undertaken where the barest of means are available.
- There must be restricted entry to the OT and this should only be permitted after a complete, except for underwear, change of clean clothes and shoes and with the wearing of a proper clean head covering and mask (these items should be used once only before discarding or washing). Religious hand-washing for at least five minutes utilising an antiseptic soap solution will reduce the incidence of infection.
- The OT should be situated in the most inaccessible part of the hospital so that there is a minimum encroachment by the general hospital patients.
- Ideally there should be an air purifying/conditioning system in the OT.
- All tubing (suction, oxygen, anaesthetic) should be regularly and frequently cleaned and disinfected, according to individual manufacturer's instructions, to reduce the chances of nosocomial infection.

Set-up of the recovery room

- Should be adjacent to the OT so that the surgeon and the anaesthetist have immediate access to the patient.
- Nursing care, oxygen, suction and emergency medicines should be available. Resuscitation equipment should also be available (see Chapter 1.17).
- **An adequately trained doctor, proficient in resuscitative measures and critical care management, should be present whenever patients are in the recovery room.** Frequent evaluation and monitoring of surgical patients should be undertaken during the first 24 hours following a major operation. This should include observations/measurements of hydration, urine output, output from drains, soakage from the wound and blood pressure. Postoperative pain management is most important and a relatively pain-free patient has a better outcome (see Chapter 1.27). ✔
- All day-case surgical patients should be sent home after an evaluation has ensured that the effect of the anaesthesia has weaned off. Ideally the child should have taken at least one feed in the hospital uneventfully.

Major operations that require intensive care should be referred when feasible to a care centre equipped with all the sophisticated equipment and expertise manpower required.

113 Paediatric anaesthesia

Oliver Ross

Minimum standards requirements

Paediatric anaesthesia

- Local anaesthesia
  – Lidocaine with and without epinephrine
  – Blunt needles
- Oxygen
- Ketamine
- Inhalational anaesthesia (ether/nitrous oxide/halothane)
- Boyle's machine
- Intubation equipment plus Ayre's T-piece and Jackson-Rees modification
- Suction manual/electric
- Pulse oximeter
- Essential drugs: epinephrine, atropine, ketamine, diazepam/midazolam, opiates, thiopentone, suxamethonium, pancuronium/atracurium
- Intubation: laryngoscopes, blades, spare bulbs, McGill's forceps, ETT tubes, reinflating bags and masks with reservoirs, tape, suction tubes (including Yankauer)

Before you start

- Create a child-friendly environment; if possible, parents should be present through to the time of induction of anaesthesia (see Chapter 1.4).
- Be aware of anatomical, physiological and pharmacological concepts from neonate through to adolescent.
- Know normal values of main physiological variables (see Appendices 7.1 and 7.8).
- Know ABC of assessment and resuscitation of sick child (see Chapters 1.17, 1.19, and 1.20).
- Know hourly fluid and blood requirements for every child (see Chapter 1.14 and Appendix 7.1).

Preoperative assessment

- Ensure adequate resuscitation equipment is present.
- Define physiological status: airway, oxygenation and ventilation, cardiovascular stability, hydration.
- Assess airway and ease of intubation – chin (look from side), mouth opening, visibility of uvula and posterior pharyngeal wall.
- Past medical history (including anaesthetic): in particular cardiorespiratory illness, presence of respiratory tract infection which increases adverse respiratory events during and after anaesthesia.
- Medication and allergies.
- Nil by mouth guidelines:

Clear fluids	2 hours
Breast milk	4 hours
Formula milk/food	6 hours

- Plan fluid requirements:
  - **Avoid hypotonic solutions where possible** ✔
  - **Sodium is the most important ion**
  - **Give glucose (10%) as required; check blood glucose (especially in neonates, infants)**
  - Basic maintenance fluids:
    4 ml/kg/hour for first 10 kg of body weight then add
    2 ml/kg/hour for next 10 kg of body weight then add
    1 ml/kg/hour for each kilogram after
    Additional fluids: Judge clinically: cardiovascular status and urine output (> 0.5–1ml/kg of body weight/hour)
- Premedication: limit to paracetamol, minimise sedative and vagolytic medications
- Explanation to child and parents

Intraoperative considerations

Plan the anaesthetic

- Maintenance of normal physiological status is part of balanced anaesthesia.
- General anaesthesia: involves hypnosis (reflex inhibition), muscle relaxation and narcosis (analgesia). Anaesthetic drugs rarely provide all of these three, for example ketamine is a poor muscle relaxant, ether is not analgesic, local anaesthetics provide no hypnosis. Therefore, modern anaesthesia uses combinations of drugs to provide balanced anaesthesia.
- Avoid general anaesthesia wherever possible. Most operations can be performed using one or all of the following: sedation, local anaesthesia and ketamine.

These techniques should be the basis of anaesthesia for the non-specialist anaesthetist.

- General anaesthesia is indicated where other methods are precluded – lack of knowledge, lack of drugs, nature of surgical procedure (for example abdominal surgery) or contraindication to ketamine/local anaesthetic drug.
- Inhalation anaesthesia with or without muscle relaxant and local anaesthetic/opioid as analgesia is the standard combination.
- Induction can be achieved by inhalation of anaesthetic gases (particularly where there is an empty stomach, particularly valuable in acute upper airway obstruction) when there is adequate expertise and equipment. Ketamine (particularly if there is cardiovascular instability) or thiopentone (if there is raised intracranial pressure or fits) can also be used to induce anaesthesia. Ketamine can be the sole anaesthetic agent with a loading dose followed by supplemental boluses or an infusion (see below).
- Neonates and infants form a special group. Sedation and ketamine anaesthesia are more difficult to perform safely. Under general anaesthesia, they do not breathe well (difficult airway maintenance, unfavourable chest wall/lung mechanics and limited reserve in face of hypoxaemia); thus in general, ventilation must be controlled. Caution must be exercised with drug doses (opioids and local anaesthetics due to side effects, suxamethonium required in higher dose) and postoperative hazards are enhanced. Ketamine or inhalational anaesthesia with controlled ventilation is the technique of choice. DO NOT UNDERTAKE ANAESTHESIA LIGHTLY IN THIS AGE GROUP.

For all anaesthesia

- Always give oxygen if available (especially at altitude).
- Use all monitoring available (essential).
- Maintain normothermia (warm fluids, high ambient temperature).
- Ensure good intravenous access; fluids as maintenance and as indicated clinically; optimal haemoglobin depends on age, >10 g/dl generally.
- Humidification of inspired gases good if available.
- Be aware of anaphylaxis.
- Analgesia: paracetamol, non-steroidal anti-inflammatory drugs, local anaesthetic infiltrations and blocks, opioids (codeine, morphine and pentazocine).
- Plan to maintain spontaneous ventilation wherever possible; never use muscle relaxants without knowledge of or experience in how to intubate.

When to intubate

To protect airway/lungs

All acutely ill children have poor gastric emptying; if in doubt or there is a strong indication of "full stomach" (acute abdomen) you MUST protect lungs with an endotracheal tube. Intravenous induction with application of cricoid pressure prior to intubation is technique of choice; prolong nil by mouth times post-trauma to minimise risk of regurgitation into the airway.

To ensure safe maintained airway

Difficult airway or potentially difficult intubation: NEVER give muscle relaxant until airway is secure with endotracheal tube, i.e. keep breathing! Be cautious with ketamine.

To provide positive pressure ventilation

- Prolonged surgery, where muscle relaxant is essential (abdominal surgery).
- Neonates and infants.
- To improve left ventricular function in septic shock.

To improve oxygenation

- Can administer 100% oxygen and maintain better lung volumes.

Postoperative care

- Basic recovery care: attention to ABC, maintenance of normothermia, continued fluid therapy and provision of safe and effective analgesia.
- Commence oral fluids as soon as possible.
- Regular oral/rectal analgesia (paracetamol, NSAIDs) with opioid analgesia early and when needed; OPIOID INFUSIONS (see Chapter 1.27).
- Pain assessment scores to titrate analgesia.
- Parents have an important role.

Techniques

Sedation (see Chapter 1.27)

- Conscious sedation through to general anaesthesia: based on loss of airway self-maintenance, gradual loss of protective reflexes and decreased responsiveness.
- All drugs can have unpredictable and prolonged effects.
- Cardiorespiratory compromise is the biggest danger.
- **Avoid absolutely in upper airway obstruction**. ✔
- Prepare as for general anaesthesia.
- Drugs: chloral hydrate, midazolam, diazepam.
  - Chloral hydrate 25–50 mg/kg oral dose
  - Midazolam 200 micrograms/kg intranasal/sublingual 500 micrograms/kg oral dose
  - Diazepam 200 micrograms/kg intravenous/oral dose (500 micrograms/kg rectal dose)

Local anaesthesia

Why?

- Cheap, simple, minimal equipment required
- Can avoid general anaesthesia
- For simple, brief procedures
- Post-trauma analgesia/postoperative analgesia

Beware!

- Slow onset, prolonged effect
- Each block has potential major complications
- Toxicity: central nervous system (seizures) and cardiovascular (arrhythmias)

- All techniques can be lethal
- Not sedative!

Safety
- Always ensure sterility
- **NEVER exceed maximum doses of local anaesthetics: lidocaine 3 mg/kg (7 mg/kg with 1 in 200 000 epinephrine)**
- Be cautious with doses in neonates
- Know the anatomy
- Use fine needles (ideally 25–29 gauge)
- Use blunted needles (easier to identify layers)
- Always aspirate before any injection (not 100% guarantee of avoiding intravascular injection)
- All injections should be easy i.e. no resistance to injection (resistance indicates intraneural injection)
- Treat toxicity: ABC, diazepam for fits

Applications: topical, infiltration, nerve blockade, central neural blockade

Safe use of local anaesthetic blockade
- Main danger is intravascular injection
- Systemic uptake can be high from certain techniques – for example intercostal infiltration
- Intraneural injection will cause permanent nerve damage
- Always ensure sterility of all equipment and drugs
- Respect contraindications: allergy to drug, local sepsis, bleeding diathesis
- Be cautious with doses in neonates (decreased protein binding, reduced hepatic clearance)
- Know the anatomy for each block; look in a book!
- Use blunted needles (easier to identify layers)
- Always ASPIRATE before any injection (not 100% guarantee of avoiding intravascular injection)
- All injections should be easy, i.e. no resistance to injection (minimises intraneural injection)
- Any limb block: USE WITH CAUTION if at risk of compartment syndrome
- Early symptom of toxicity is tingling of the lips
- ✔ **Do not use with epinephrine in digital or penile blocks**
- Know how to treat toxicity: ABC, diazepam for fits

Topical: easy to do and can be very effective, for example Emla, local anaesthetic soaked dressings, eye drops.
Infiltration: small needle (25–29 gauge), slow injection, warm, add 8.4% sodium bicarbonate (1 in 10 for 8.4%: lidocaine 1%).

Nerve blocks

- Explain to patient and carers; gain consent.
- Warn patient of motor blockade and sensation of sensory blockade.
- Apply principles of safe use of local anaesthetics.
- Onset can be slow: 30–60 minutes.
- Effect can be prolonged: 24 hours.
- Be aware of distribution of analgesia for each block.
- Use equivalent volume to those stated if using different local agent.

Femoral/"3 in 1"
- Femoral shaft fractures, burns, grafts from anterior thigh
- Medial calf only blocked below the knee
- Lie supine; femoral nerve lies lateral to vascular sheath just below inguinal ligament (Lateral to medial = nerve, artery, vein)
- Sterilise skin, give skin analgesia
- Identify artery; injection point 0.5 – 1 cm lateral to artery
- Advance 1–inch 21gauge blunted needle at 45° to skin until two "pops" felt (fascia lata, fascia iliacus)
- Aspirate, inject lidocaine 1% with epinephrine 1 in 200 000 at a dose of 0.5 to 0.7 ml/kg
- Larger volume blocks obturator and lateral cutaneous nerves in addition; hence "3 in 1"

Brachial plexus block (axillary approach)
- Easiest and safest approach
- Blocks whole arm except upper arm and shoulder
- Lie supine, abduct arm to 90°, rotate externally, forearm to 90° (that is hand level to or close to back of the head)
- Identify artery, sterilise skin, skin analgesia
- Advance 1–inch 22 gauge needle aiming for apex of axilla, over and parallel to artery
- One pop, let go needle – it will bounce with arterial pulsation if correctly sited
- Support needle, aspirate and inject 0.5 ml/kg lidocaine 1% with epinephrine 1 in 200 000

Intercostal
- Useful for fractured ribs, upper abdominal surgery
- Risk of complications high but effective block
- Identify posteromedial curve of rib
- Sterilise skin, give skin analgesia
- 22–24 gauge needle perpendicular to skin, hit rib
- "Walk" needle just under rib, aspirate and inject
- Repeat at each rib [Beware maximum dose (usually 0.5 ml/kg of lidocaine 1% with epinephrine 1 in 200 000) in total to be limited by toxic level of the anaesthetic combination used]
- Intravascular uptake is high from this site

VRA (intravenous regional anaesthetic)/Bier's block
- Distal limb excisions and fracture manipulations
- Elevate arm
- Tourniquet (double if available)
- Two intravenous cannulae – one in the limb to be blocked as distal as possible, the other for safety in another limb
- Inflate tourniquet (twice systolic arterial pressure)
- Lidocaine 1% (not with epinephrine and not use bupivicaine) 0.3 ml/kg into cannula in limb to be blocked
- 10 minute onset, safe to release tourniquet after 30 minutes

Ilioinguinal/iliohypogastric block (field block)
- Hernia repair, orchidopexy
- Lie supine and identify anterior superior iliac spine
- 1 cm medial, 1 cm caudal

- Sterilise skin, give skin analgesia
- 22 guage blunted needle perpendicular to skin
- Advance until one pop felt (after skin), aspirate and inject
- Two pops is acceptable, three pops or "feels too far" runs risk of femoral nerve block
- Infiltrate 0.5 ml/kg 1% lidocaine with 1 in 200 000 epinephrine after aspiration
- Withdraw to skin and infiltrate

Central blocks including caudal: refer to a specialist anaesthetic textbook

Ketamine

Ketamine is an analgesic and dissociative anaesthetic, inducing a trance-like cataleptic state dissociated from the environment.

- Advantages: airway maintenance, cardiovascular stability, and good for short procedures, particularly on extremities.
- Disadvantages: airway not guaranteed especially in neonates and interference risks laryngo- and bronchospasm. The accompanying cardiovascular stability is no alternative to good resuscitation. Desaturation and apnoea (especially after bolus administration); hypertonus especially with prolonged anaesthesia (greater than 1 hour); resistance unpredictable especially in developmentally delayed child; raised intracranial and intraocular pressure are all potential problems. Raised intracranial pressure is an absolute contraindication. Emergence phenomena – hallucinations, perhaps less in children, can be minimised with benzodiazepines.
- Dose: see below.
- Use as low a dose as possible.
- Recovery may be prolonged.
- Caution in neonates.

Ketamine doses

- 1 mg/kg slow IV bolus (over 2–5 minutes)
- Repeat using half dose (500 micrograms/kg) after 15 minutes
- For IM induction use 7 mg/kg
- For infusion purposes aim to make up a solution of 1 mg/ml (for example 500 mg in 500 ml bag of 5% glucose or 0.9% saline)

  As a rough guide for maintenance of anaesthesia use the following regime based on a standard giving set where approx. 15–20 drops = 1 ml (a micropipette giving set from a burette system gives 60 drops = 1 ml). A dose of 2–4 mg/kg/hour is usually appropriate for anaesthesia.

  1 drop/second if 60 kg body weight is equivalent to 3–4 mg/kg/hour

  1 drop/2 seconds if 30 kg body weight is equivalent to 3–4 mg/kg/hour

  1 drop/4 seconds if 15 kg body weight is equivalent to 3–4 mg/kg/hour

  1 drop/8 seconds if 7.5 kg body weight is equivalent to 3–4 mg/kg/hour

  For analgesia a lower dose of 500 micrograms – 1mg/kg/hour is usually effective.

  Marked tachyphylaxis can occur with infusions lasting > 30–60 minutes.

Inhalational anaesthesia

- DO NOT UNDERTAKE LIGHTLY.
- Need airway maintenance skills and ability to recognise appropriately anaesthetised patient as bare minimum for safe practice.
- Best simple guide to depth of anaesthesia is the level of sympathetic nervous system arousal.
- Equipment is generally more specialised.
- Spontaneous ventilation via mask or endotracheal tube and breathing system is the safest application.

Ether

Ether is a relatively safe drug to use. It can be given by open method or by a breathing system and vaporiser, usually of draw-over type. Induction of anaesthesia is slow and relatively predictable. Respiratory depression is late and cardiovascular stability is well maintained. Recovery can be prolonged. **It has no analgesic effect**.

Halothane

Halothane is a potent but highly effective inhalational anaesthetic agent. **It can only safely be given via a vaporiser. Overdose easy and dangerous**.

Trichloroethylene

Trichloroethylene (Trilene) has advantages of slow onset, high potency and analgesia. Tachypnoea and postoperative nausea are seen.

Triservice apparatus

The Triservice apparatus, incorporating draw over OMV vaporisers, is well established and can be used for inhalational anaesthesia without the need for compressed gases. Refer to a specialist anaesthetic textbook for further details.

Essential equipment

Minimum equipment: Ketamine and local anaesthesia provision

- Intravenous cannulae
- Syringes
- Needles

Desirable equipment: Ketamine, inhalational anaesthesia and resuscitation equipment

- Oxygen masks with/without reservoir bags, paediatric or adult sizes
- Oxygen supply-cylinders with oxygen flowmeter
- Intravenous fluids (isotonic > glucose solutions)
- Intravenous administration sets (burettes ideal)
- Anaesthetic facemasks (ideally clear masks with inflatable rims providing an airtight seal with minimal deadspace)

- Guedel-type oropharyngeal airways – sizes 0–4
- "Bag-valve-mask" incorporating non-rebreathing valve (paediatric), reservoir tubing/bag and self-inflating bag (neonatal and child sizes)
- Ayre's T-piece, with Jackson–Rees modification (open-ended 500–ml bag)
- Endotracheal tubes: 2.0–9.0 mm internal diameter cuffed and uncuffed, PVC
- Laryngoscopes – straight bladed, curved bladed
- Magill forceps
- Fixation tape
- Suction apparatus (manual-foot/hand pump/electric)
- Suction catheters
- Yankauer suckers
- Means of administering inhalational anaesthetic agents
  - Boyle's machine
  - Anaesthetic circuits (refer to a specialist anaesthetic textbook)
  - Vaporisers
  - Triservice apparatus

Essential monitoring

Improves patient safety, reducing morbidity and mortality

- Use in any location and for any technique including sedation
- Use from induction through to recovery
- Documentation essential

✔ **The best and universally available monitor is the presence and vigilance of the person administering the anaesthetic.**

Minimum monitoring

- At everyone's disposal
- Colour, pulse rate and volume, capillary refill time, respiratory rate and auscultatory findings, pupil size

Minimal monitoring standards

Anaesthetist and all the above signs

Remember to always check:

- Equipment prior to use
- Is there enough oxygen?
- Is oxygen flowing into the patient?
- Patient: electrocardiogram, non-invasive blood pressure, arterial oxygen saturation (pulse oximeter: expensive but very useful), temperature (ideally core), blood glucose, urine output, ***end-tidal carbon dioxide (expensive but useful)***

Essential drugs

The drugs highlighted in bold below must be available to provide safe anaesthesia whatever the technique.

- Local anaesthetics (lidocaine)
- Ketamine
- **Atropine** 10–20 micrograms/kg IV
- **Diazepam** 100 micrograms/kg IV/midazolam 200 micrograms/kg IV
- Paracetamol
- Morphine
- Codeine
- **Suxamethonium** (bromide if no refrigeration) 1–2 mg/kg IV (lasts 5 minutes, higher dose in neonate, salivation, hyperkalaemia, masseter spasm, anaphylaxis)
- Pancuronium 100 micrograms/kg (or atracurium, vecuronium)
- Neostigmine, edrophonium
- **Epinephrine** (resuscitation doses: 1 in 10 000 = 100 micrograms/ml and 1 in 1000 = 1 mg/ml)
- **Oxygen**
- **Intravenous fluids**
- Thiopentone 2–5 mg/kg (TAKE CARE IF NEVER USED BEFORE, apnoea, hypotension)
- Inhalational agents

Intubation

Endotracheal tube sizes

Age/4 plus 4 = internal diameter in mm

- 3 × internal diameter = length at lips in cm, add 2 cm at nares
- Uncuffed tube under 25 kg (narrowest part of larynx is non-distensible circular cricoid ring)

Why intubate?

- Secure airway
- Protect airway
- Prolonged ventilation
- Intraoperative ventilation
- Tracheobronchial toilet
- Application high airway pressures and PEEP
- Cardiopulmonary resuscitation (all of the above)
- In raised intracranial pressure to maintain normal oxygenation and normocapnia

Which tube?

- Uncuffed under 25 kg: larynx narrowest below the glottis at the circular non-distensible cricoid ring.
- Correct tube is that which passes easily through the glottis and subglottic area with a small air leak detectable at 20 cm H_2O (= sustained gentle positive pressure).
- Size of tube is one that can just fit into the nostril.
- In preterm neonates 2.5–3.5–mm internal diameter.
- In full-term neonates 3.0–4.0–mm internal diameter.
- In infants after neonatal period 3.5–4.5–mm internal diameter.
- Children over 1 year:
  Internal diameter in mm = age/4 + 4
  Length of tube in cm = age/2 + 12 for oral tube
  = age/2 + 14 for nasal tube

Aids to intubation

- Laryngoscope: blade (straight for neonates and infants because of long, floppy epiglottis, curved for older children), bulb and handle
- Magill forceps
- Introducer (not protruding further than end of tube itself)
- Gum elastic bougie (over which tube can pass)
- Cricoid pressure (can help visualisation of larynx)
- Suction apparatus must be available plus Yankaur and other catheters
- Syringe (cuffed tube)

Predicting difficulty

- Likely to be difficult:
  difficulty in opening mouth
  reduced neck mobility
  laryngeal/pharyngeal lesions
- Congenital:
  Pierre Robin syndrome, small mandible, mucopolysaccharoidoses, Down's syndrome
- Acquired:
  burns, trauma
- Look from side: small chin means difficulty

Complications

- Displacement: oesophageal, right (usually) or left main bronchus rather than trachea
- Obstruction: kinking, secretions
- Trauma: lips to larynx
- Hypertensive response
- Vagal response
- Spasm: laryngeal, pharyngeal
- Aspiration: gastric contents

How to do it

Prepare and check equipment

- Choose appropriate tube size with one size above and below available
- Get tape ready to fix tube
- Suction must be available
- Induce anaesthesia and give muscle relaxant unless completely obtunded
- ✔ **Do not attempt intubation in semi-conscious child**

Position child:

- > 3–4 years: "sniffing morning air" position (head extended on shoulders, flexed at neck: a pillow under the head)
- < 3 years (especially neonates and infants): neutral position (large occiput no pillow but towel under shoulders may help)
- keep in neutral position with in-line immobilisation if unstable cervical spine (trauma, Down's)

Oxygenate child

Introduce laryngoscope into right side of mouth, sweep tongue to the left, advance blade until epiglottis seen.
Curved blade:
advance blade anterior to epiglottis, lift epiglottis forward by moving blade away from own body
Straight blade:
advance blade beneath epiglottis into oesophagus, pull back, glottis will "flop" into view

Recognise glottis

- Insert endotracheal tube gently through vocal cords. McGill's forceps may be required
- Stop at predetermined calculated length (usually marked on the end of tube)

Confirm correct placement:

- Chest moves up and down with ventilation
- Listen to breath sounds in axillae and anterior chest wall
- Confirm no breath sounds in stomach
- Oxygen saturations do not go down AND COLOUR DOES NOT DETERIORATE

Secure tube. Proceed to nasal intubation if skilled (best for long-term ventilation). **Contraindicated in base of skull fracture**

Fresh gas flow

- Fresh gas flow through T-piece circuit to prevent rebreathing carbon dioxide
- Minute ventilation = 1000 ml plus 100 ml/kg
- For spontaneous ventilation, 3 × minute ventilation
- For positive pressure ventilation, 1.5 × minute ventilation

Ventilator rates and tidal volumes (by hand or mechanical)

Tidal volume is that which is **enough to see the chest expand adequately**: usually 5–10 ml/kg
Rates:

Neonates	30–40 per minute
Infants	25–30 per minute
Child	20 per minute
Adolescent	15 per minute

Minute ventilation = rate × tidal volume

1.14 Safe paediatric and neonatal transfusion practice

G S Gabra and M N Mansour

Minimum standards requirements

Blood transfusion services

- Blood (fresh frozen plasma, platelets, clotting factors, cryoprecipitate)
- Small volume packs
- Screening for hepatitis B and C, HIV, syphilis, malaria
- Cross-matching
- Burette giving sets

Introduction

✔
- **Blood or blood products should be transfused only when needed to save life or prevent major morbidity.**
- Risk of transmission of infection is a major concern in countries with limited resources and poorly organised blood transfusion services.

Situations where transfusion is required

In neonates

After birth, haemoglobin drops to less than 100 g/litre in term infants at 8–12 weeks of age, but in premature infants, it can drop to 70 – 100 g/litre, even earlier at 6 weeks (oxygen delivery is well maintained because of a rising haemoglobin A).

- Hypovolaemic shock can result from acute blood loss, such as premature separation of the placenta or feto-maternal haemorrhage, twin-to-twin transfusion and other causes of fetal or neonatal haemorrhage.
- Neonates may lose considerable blood volume as a result of sampling for laboratory tests. Samples should be minimised.
- Transfusion in neonates can be reduced by adequate antenatal care to reduce the risks of premature delivery and when possible prevention of nutritional anaemia in the mother.
- Additional measures to reduce transfusion include: encouraging breastfeeding and early provision of vitamin K prophylaxis, iron, vitamins and other haematinics especially in premature babies.

In children

- Malaria and other chronic infections including HIV
- Major trauma/surgery
- Haematological malignancies
- Sickle cell disease
- Other congenital haemolytic anaemias such as thalassaemia and severe haemolysis of G6PD deficiency
- Burns (see Chapter 5.11)

Transfusion policies and guidelines

- In hypovolaemic shock, erythrocyte-free volume expanders may be used to maintain tissue perfusion. Oxygen and top-up blood transfusion (10–20 ml/kg over 5–10 minutes) may be required when tissue oxygenation is compromised.
- Transfuse for anaemia only when there are clinical signs, such as tachycardia, tachypnoea, recurrent apnoea, failure to thrive or early signs of heart failure.
- When possible provide malaria prophylaxis particularly in pregnant women and children with sickle cell disease. Early treatment of clinical malaria reduces the profound haemolysis which is a major reason for transfusion in endemic areas.
- Anaemia due to malaria responds to treatment with antimalarials and folic acid.
- Blood transfusion is not required for sickle cell disease in the steady state: may be indicated in severe anaemia with incipient or established cardiac failure, acute splenic enlargement, sequestration crisis with rapidly-falling haemoglobin, aplastic crisis, acute chest syndrome, stroke and sometimes as exchange transfusion for severe priapism (see Chapter 3.22).
- National programmes for thalassaemia and other congenital haemolytic disorders, such as G6PD deficiency help reduce transfusion requirements.

In situations where safe blood is not readily available the following recommendations have been made:

- Transfusion is not necessary if haemoglobin is more than 50 g/litre.
- Transfusion may be necessary if haemoglobin is less than 50 g/litre and there is incipient cardiorespiratory

distress (air hunger, hypotension, tachycardia and oedema).
- Transfusion may be necessary if haemoglobin is less than 40 g/litre and complicated by malaria or bacterial infection, even without incipient cardiac failure.
- Transfusion may be necessary if haemoglobin is less than 30 g/litre with no apparent complications.

In situations where there is safe blood:
- **Neonates and infants <4 months old**
  - Blood loss of >15% over 2 days.
  - Haemoglobin <70 g/litre with clinical manifestations of anaemia.

- **Infants 4 months or older:**
  - Acute blood loss unresponsive to crystalloid and colloid infusions.
  - Intraoperative blood loss >15% of total blood volume and postoperative haemoglobin <80 g/litre with clinical symptoms.
  - Haemoglobin <110 g/litre with severe pulmonary disease.
  - Acute haemolysis with haemoglobin <80 g/litre with signs of anaemia.
  - To suppress endogenous haemoglobin in sickle cell disease crises and thalassaemic syndrome.

Frozen fresh plasma is only recommended when a specific haemostatic defect has been identified.

Platelets are prepared from fresh blood using a special, simple centrifugation method. The remaining partially packed red cells can be used to transfuse other patients. Once extracted by this method platelets can last up to 5 days at room temperature (around 23 °C). Platelets should not be stored in a refrigerator. Transfused platelets survive only briefly and repeated infusion may be required for active bleeding or preceding essential procedures such as a lumbar puncture in a child with severe thrombocytopenia.

Provision of blood

- Ideally by routine whole blood collection from an established panel of blood donors with quality standards for testing, processing and distribution.
- Most transfusions are required and given as an emergency procedure. Emergency collection of blood for paediatric use should ideally not be necessary.
- Safe transfusion in children is enhanced by the following measures:
  - Collection of blood from repeat regular donors screened using a standard health-check questionnaire and whose donations are found negative for all markers for transfusion-transmissible infection.
  - Collection in a multipack which allows each donation to be divided into small volumes, in a closed sterile system to reduce wastage and donor exposure.
  - Multiple, small-volume packs can be used for multiple transfusions in one child or neonate without having to repeat the pretransfusion tests.
- Group O Rhesus negative small-volume packs facilitate transfusion across the ABO barrier. They must be checked for high-titre anti-A or anti-B by a suitable antiglobulin method.
- Emergency collection of blood for paediatric use should be discouraged.
- Establish a routine procedure for collection, testing and processing which should cover routine and emergency transfusions

Maternal blood is not recommended even in an emergency although in theory it can be used after compatibility testing with the recipient's serum.

Pretransfusion testing

- Minimum acceptable tests on blood prior to transfusion
  - ABO and Rhesus D grouping
  - Screening for hepatitis B antigen and antibodies to HIV-1 and -2, hepatitis C virus and syphilis
  - Additional tests for locally prevalent infections; such as malaria and Chagas disease
- 0.1–0.2 ml blood in an EDTA bottle is required for grouping and 2 ml of clotted blood in a plain bottle for compatibility testing cross-match.
- In infants less than 4 months of age, maternal blood is always required: 4 ml EDTA plus 5 ml clotted.
- Blood grouping of the neonate can be performed using a cord, capillary or small venous sample (2–3 drops and specific standard reagents anti-A, anti-B, anti-A + B and anti-RhD. Red cells only are used, because antibodies in the sera of neonates are too low to be of significant value).
- The inclusion of control A, B, O, RhD-positive and -negative cells in the procedure is part of good laboratory practice, and should be part of the testing method.
- Two methods should when possible be used for grouping to ensure reliabilty
- For neonates and infants up to 4 months, compatibility testing is not required if the mother's serum is negative for alloantibodies. Compatibility between the mother's serum and red cells to be transfused is required only if the mother has antibodies or if there is a previous history of haemolytic disease of the newborn.
- The most suitable method for compatibility is by the anti-human globulin technique at 37°C for one hour. Agglutination should be read before and after the addition of the antihuman globulin reagent.

Exchange transfusion

- For haemolytic disease of the newborn with severe anaemia and/or severe hyperbilirubinaemia (see Chapters 3.48 and 6.9). Exchange of double the neonate's blood volume is often required using 160–180 ml/kg of whole blood and/or plasma reduced red cells. The latter is prepared by removing from a bag of whole blood (450 ml) approximately 100 ml of plasma to obtain a haematocrit of 0.5–0.6.

- Patients with sickle cell anaemia and acute chest syndrome or impending cerebrovascular episodes may benefit (see Chapter 3.22).

Blood for exchange transfusion

- Rhesus-negative blood, Group O or same ABO group as infant and compatible with maternal or infant's serum.
- Fresh, never more than 5 days old.
- Warmed with heating coil or stood for 1 hour at room temperature or under the mother's dress.
- Exchange 160–180 ml/kg of blood with partially packed cells.
- Additionally screened for HbS if exchange is used to treat sickle cell disease

Bedside transfusion

- Calculate blood replacement volume by taking into account the total blood volumes (80–85 ml/kg in full-term neonates and children and 100 ml/kg in premature infants).
- Venous access should be chosen to allow a 23–25 gauge needle or 22–24 gauge vascular catheter.
- Giving sets and filters – blood is usually cleaned and filtered in the lab, so when transfusing it to a patient no special filter is required other than the on-line filter of the standard giving set.
- Blood should be treated like any other IV fluid and be given using an accurate measurement of rate and time. ✔ **A burette should be used for infants or in children where too rapid an input could be dangerous, for example in incipient or actual heart failure**.

Dosage

Acute blood loss

- Give 10–20 ml/kg of whole blood as rapidly as possible or a similar volume 4.5% albumin (if possible through a wide bore cannula or central venous line).
- Estimate infusion rate for continuing transfusion using:
  - an estimate of existing blood loss
  - an estimate of continuing blood loss
  - vital signs

Top-up transfusion

- Give 20 ml/kg, partially packed cells, over 4 hours.
- Ideally blood should be < 7 days old.
- Furosemide is not routinely given.

Other points

- For neonates, 2–3 ml/kg/hour is considered to be a safe rate and the total volume to be given varies between 10–15 ml/kg.
- A lower infusion rate with diuretic cover may be required in cardiac failure.
- A record of the volumes used should be kept in the bedside file, along with details of any adverse reactions.
- Blood for transfusion should NOT be exposed to room temperature for > 6 hours from the time the pack was removed from storage (2–6 °C).
- If a transfusion reaction occurs send remainder of blood pack for immediate testing.
- All blood packs should be checked by at least two staff before being given (usually the nurse and doctor in charge). The name of the recipient, the blood group of the donor and the recipient, the date of collection and expiry and identification number of the donation should be carefully documented.

Reactions to blood transfusions

- ✔ **Before transfusion the blood group, expiry date and details of whom the blood has been issued to should be checked, ideally by two trained nurses or the nurse and the doctor. Then it should be checked against the recipient's details, that is name, blood group, date of birth, hospital number (if recorded) and prescription chart. Medical staff should prescribe all blood products.**
- Baseline observations of the child's temperature, pulse, respirations and blood pressure should be recorded before the commencement of a blood transfusion. These observations should then be recorded every 30 minutes whilst the child receives the blood transfusion. Any deviation from the normal, such as a rise in temperature, pulse, or respirations, could indicate a reaction to the transfusion and consideration that the blood transfusion should be stopped.
- Blood should be given at the prescribed rate, and be given over a maximum of 4 hours. If the amount of blood prescribed will take longer than 4 hours, another unit of blood should be used. Once the unit of blood has been removed from the refrigerator, it should be used within 6 hours. The warming of the blood is only needed when the transfusion rate exceeds 15 ml/kg/hour.
- A fluid balance chart should be maintained throughout the transfusion. The child's urinary output should also be monitored.

The following types of reactions are possible when giving a blood transfusion:

- Febrile
- Allergic
- Haemolytic
- Circulatory overload
- Air emboli
- Hypothermic
- Electrolyte disturbance

As well as recording the observations a visual observation should be noted. The nurse/doctor should look for symptoms such as: fever, chill, nausea, vomiting, flank pain, headache, rash, wheeze, cyanosis, dyspnoea, as other indicators of a reaction to the transfusion.

If a severe reaction is identified, then the transfusion should be stopped and medical help sought. The remainder of the pack should be sent to the laboratory for immediate testing.

Each reaction will have to be treated on an individual basis, but anti histamines are often used for mild reactions to blood.

Maternal blood

This is not recommended, even in an emergency mainly because of the risk of graft–versus–host disease although in theory it could be used, particularly if the mother is known to be blood group O Rhesus negative.

Further reading

Guidelines for Administration of Blood Products: Transfusion of Infants and Neonates. British Committee for Standards in Haematology Blood Transfusion Task Force. *Transf Med* 1994;**4**:63–9.

Guidelines for the Appropriate Use of Blood. Geneva: WHO, Global Blood Safety Initiative, 1989.

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1.15 Essential laboratory services for sick and injured children in hospital

Wendy Bailey

Basic services provided in the laboratory and on the ward

Whenever possible, the regional or central laboratory should procure the chemicals, prepare the reagents and standards, and distribute them with the necessary controls and appropriate testing procedure to district laboratories. Details on how to prepare the required reagents, standards and controls can be found in the 1995 WHO publication *Production of Basic Laboratory Reagents*.

For all small hospitals, WHO recommends six basic investigations as an absolute minimum. They are:

- Haemoglobin or packed cell volume
- Blood smear for malaria
- Blood glucose
- Microscopy of cerebrospinal fluid (CSF) and urine
- Blood grouping and cross-matching
- For newborn care, blood bilirubin

Tests that can be performed on the ward

- Blood grouping
- Urgent blood film for malarial parasites
- Urine microscopy (see Chapter 6.25)
- HIV rapid screening test
- HBsAg screening test
- Hot stool examination (for *E.histolytica*)
- Rapid Hb (WHO paper method)
- CSF/gland/chancre aspirate/wet preparation for trypanosomes

Tests to be performed in laboratory

- Thick and thin blood films for malaria and other parasites
- Smears for *Leishmania* amastigotes
- DAT for *Leishmania* antibodies
- TB sputum smears
- Slit skin smears for leprosy
- Gram–stained smears
- Hb estimation and platelets
- Total and differential white cell count
- ESR
- Sickle test
- HIV and hepatitis screening tests
- Blood grouping and cross-matching
- Urine deposits
- Saline smears and formol ethyl acetate concentration for stool parasites

Essential equipment

A decent microscope is essential and also saves time and therefore salary costs. The following is recommended: Tropical Health Technology Binocular Microscope from Doddington March, Cambridgeshire, PE15 0TT, UK.

Haematological investigations

- Haemoglobin

  Haemoglobinometer (BMS meter, Haemo-Check) – a useful method for single investigations. No dilution or measurement of sample required.

  Cyanmethaemoglobin method – useful for multiple sampling, requires colorimeter with 540 nm filter, Drabkins and standard Hb reference solutions.

  Microhaematocrit centrifugation for estimation (note: may get raised value due to plasma loss, for example burns, dehydration)

  Haemoglobin colour scale (WHO) filter paper method – simple, portable (from WHO, Geneva)
- White blood cell count

  Improved Neubauer haemocytometer, diluting fluid and hand tally counter required
- ESR Westergren method recommended
- Differential white cell counts

  Thin blood film stained with Leishmans or Giemsa stain (pH 6.8). Tally counters required. Film may also be used to examine red cell morphology for cases of suspected nutritional anaemia, for example iron deficiency
- Sickle test

  Slide test using 2% sodium metabisulphite (prepared daily) will enable sickled RBCs to be

seen. Sickle solubility test is an alternative if microscopy is not available

- HIV test
  Rapid tests easy to use for blood transfusion purposes
- Hepatitis B and C Ag kit
  Simple kits for blood transfusion purposes

Blood grouping/cross-matching sera should be available.

Biochemical investigations

- Tests on whole blood, serum or plasma
  - urea, creatinine and electrolytes
  - glucose
  - albumin
  - bilirubin
  - amylase
  - AST
- In specialised hospitals the following can be measured: alkaline phosphatase, ALT (alanine aminotransferase), calcium, cholesterol, cholinesterase, iron and triglycerides.
- Urine clinical chemistry tests
  - protein
  - glucose
  - bilirubin and urobilinogen
  - ketones
  - haemoglobin
  - nitrite
  - specific gravity
- Faecal clinical chemistry tests
  - occult blood
  - lactose
  - excess fat
- Cerebrospinal fluid clinical chemistry tests
  - protein (approximately 1 ml of CSF)
  - glucose (0.5 ml into a fluorite oxalate bottle)

Investigations for specific diseases

Malaria

- Thick blood film stained with Field's (or Giemsa) stain, made from finger prick or venous blood.
- Field's is preferred as the film can be stained in 20 seconds.
- For a diagnosis of the species a thin blood film (hand stained at pH 7.2) with Leishman's or Giemsa stain is required.
- Thick blood films may also reveal *Borrelia*, microfilariae and trypanosomes.

African Trypanosomiasis

- A thick blood film stained as above is the simplest way of diagnosing *T.b. Rhodesiense* (if a chancre is present a sample may be taken from between the edge and centre of the lesion).
- Gland fluid from a swollen posterior cervical gland may be examined (particularly useful in *T.b. Gambiense*).
- If these tests are negative up to four microhaematocrit (MHCT) tubes of blood should be taken, centrifuged for 5 minutes, attached to microscope slides and the buffy coat area examined for motile trypanosomes.
- All samples must be examined as soon as possible to avoid parasite lysis.
- **If the blood is positive for trypanosomes or it is suspected that the patient has late stage (stage II), CNS disease, a lumbar puncture must be taken and CSF examined microscopically within 10 minutes for trypanosomes and/or WBCs.** ✔

Leishmaniasis

- For cutaneous leishmaniasis a smear taken from the raised, red edge of a lesion may be taken and stained with Leishman's stain (20 minutes, pH 7.2) or Giemsa to demonstate amastigotes.
- For suspected visceral leishmaniasis, haematological investigations plus a serological test such as the direct agglutination test (DAT) are the most useful and safest investigations.

TB/Leprosy

- *For suspected TB*
  - If possible, up to 3, consecutive, morning sputum samples should be examined.
  - The Ziehl–Neelsen (ZN) method of staining should be used.
  - The addition of bleach to liquefy the sample may improve sensitivity and lowers the risk of laboratory infection.
- *For suspected leprosy* ZN stain should be used on slit skin smears.

Diarrhoeal diseases

- *For suspected parasitic cause*

Direct microscopy using saline/iodine smears. Formol ether (or ethyl acetate) concentration of use particularly in parasites such as *S. mansoni* where egg output by the female worm is low.

- *For suspected bacteriological cause*
  - Ideally the specimen should be sent for culture and sensitivity testing.
  - Culture may not be possible due to the need for sterile facilities and supplies of media.
  - If possible, samples could be sent to a reference laboratory for culture.
  - Supplies of transport media and sterile swabs should be available:
    Stuart's or Ames medium for suspected *S. typhi and Shigellae*
    Alkaline peptone water and Cary-Blair medium for *V. cholerae*
  - If microscopy of a fluid stool containing blood shows RBC's, WBC's/macrophages and numerous bacteria then bacterial dysentery is likely.

- If the sample is loose with blood and mucus then a hot stool (examined within 30 minutes of voiding) should be examined for *E. histolytica* trophozoites.

Urinary infections and renal diseases: urine examination

- Urine dipstick tests are useful to detect blood/protein/glucose/bilirubin/urobilinogen.
- A midstream urine (MSU) may be examined microscopically (see Chapter 6.25) for:
  - *S. haematobium* ova
  - Pus (WBC) cells
  - Erythrocytes
  - Casts
  - Bacteria (suspected UTI) (see Chapter 3.7)
- The addition of a drop of 1% methylene blue in physiological saline may aid microscopical examination.
- If a urine is to be sent for culture, 20 ml of an MSU should be mixed with 3 mg of boric acid (preservative).
- It is important to give instructions on how to obtain an MSU/bag urine/suprapubic aspirate.

Ulcers/exudates

For suspected bacterial (and fungal) infections a smear of the pus or exudate should be stained with Gram stain.

Meningitis

- A Gram-stained CSF deposit may be useful in cases of suspected meningitis.
- India ink stain for cryptococcal meningitis.

Further reading

District Laboratory Practice in Tropical Countries. Cheesbrough M. Cambridge: Cambridge University Press, 1998.

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116 Management of drugs

Angela O'Higgins and David Southall

Storage of medicines

Hospital and community personnel involved in the care of sick children have struggled for many years to ensure that appropriate medicines are available when needed despite the problems of controlling the abuse and illegal use of some drugs. Medicines which create the major areas of concern are **narcotics and sedatives. Supplies of these drugs need to be available for the treatment of acutely ill children, at the point of admission, in intensive care and postsurgical areas and in all areas involved in the care of children with terminal illness**. Tragically, many care settings have solved the problem of storage by refusing to have stocks of these drugs readily available, either through a belief that children do not feel pain due to their physiological immaturity or through fear of abuse by the children (misplaced) and their families or staff members.

Custody and storage of medicines in wards and departments

The responsibility for the safe custody and storage of all medicines in a ward or department is that of the nurse in charge at any one time. Designated cupboards for the different types of drugs should be available. All cupboards, which should be permanently fixed to an inside wall, should have secure locks making them inaccessible to unauthorised staff and visitors.

- Narcotic drugs, which may be controlled by law within the country should have a separate cupboard again permanently fixed, ideally within the normal drug cupboard. The keys to drug cupboards should be kept separately to all other keys and be carried by a qualified nurse for the period of each shift.
- Two books are necessary for recording the ordering and use of narcotic drugs. One is completed to order stocks, using one page for each order. The other book is to record the use of each ampoule, tablet or dose of liquid. The name of the child, hospital identification, date and time given and whether or not any portion of the drug was discarded, is entered in the register. Then each entry is signed by two staff members. Both must hold a nursing/medical qualification and one must be a member of the ward or unit staff.
- In addition, two members of unit staff must check the stock levels once in every 24-hour period and sign that the stocks are correct. Any discrepancy must be reported immediately to the Senior Nurse Manager for the hospital.
- Each hospital should have a policy to deal with unauthorised use of narcotic drugs and in some countries this will involve national law enforcement agencies.
- When new drug stocks are required, the order book is sent to the central pharmacy in a container with a tamper-proof seal. Once the pharmacist has placed the order in the container, it is sealed and must not be opened until its arrival in the receiving ward or department.
- When the stock arrives in the unit the seal is broken in the presence of the messenger and the contents checked against the order book, which is then signed by both. Drugs are then entered in the drug register, two staff members checking and signing. The drugs are placed in the appropriate cupboard, which is then relocked.

In most hospital wards and units these simple precautions will ensure that adequate narcotic drugs are available when they are needed by sick children and prevent provision of supplies to those who may be guilty of abuse.

Correct storage of drugs is paramount for prolonging shelf-life of the drug, safety and legal issues. All medication should be kept locked at all times, with the drug keys being the responsibility of the nurse in charge.

Due to the shelf-life of some drugs they need to be stored in a refrigerator, with the temperature set to store the drugs at between 2°C and 8°C. Drugs that require this sort of storage are as follows:

- Reconstituted oral antibiotics
- Eye drops
- Rectal paracetamol
- Some vaccinations
- Insulins (although can be stored for up to one month at room temperature)
- Oral midazolam
- Pancuronium/vecuronium

Calculating and giving the correct dose

The child should be weighed naked and their weight recorded in kilograms on the prescription chart. The use of a drug formulae should be considered when calculating

the therapeutic dose. To ensure that the correct amount of drug is given from the stock bottle/vial the following calculation should be used:

$$\frac{\text{Dose required}}{\text{Stock dose}} \times \text{Dilution of stock bottle}$$

$$\frac{125\,\text{mg (required)}}{250\,\text{mg (stock dose)}} \times 5\,\text{ml (dilution of stock bottle)}$$

Amount given would be 2.5 ml (see Chapter 6.3).

Medical staff should change prescribed doses if, after using the above calculation, the dose is not easily measurable, for example 1.33 ml, or 2.46 ml. To ensure that the calculated dose is given accurately, a premarked syringe should be used. The smaller the required dose, the smaller the syringe that should be used as it will give a more accurate measurement (that is a 1- or 2-ml syringe and not a 10-ml syringe). Other forms of measurement can be used for larger doses such as 5 or 10 ml. These include a premeasured medicine pot or a 5 ml premeasured medicine spoon. For safety, the calculation should be done by two trained nurses and the amount dispensed checked by the same two nurses. Although it is recognised in some hospitals that one trained nurse can check oral medication on their own, **ideally, intravenous and intramuscular drugs should be checked by two trained nurses or a nurse and a doctor**.

Prescription chart

NAME		WEIGHT		AGE				
DATE	TREATMENT	DOSE	REGIMEN	SIGN	NURSES: TIME GIVEN/SIGN DOSE: 1	2	3	4

117 Essential equipment, supplies and drugs for treating children in hospital

Martin Samuels and David Southall

A For resuscitation

Airway and breathing

- Suction apparatus
  - Wall, electrical or manual suction
  - Yankauer and soft suction catheters
  - Manual suction device, as used by midwives
- Facemask with reservoir bag (for delivering 100% oxygen)
- Self-inflating resuscitation bag with 500 ml (infants) and 1600 ml (child and adult) reservoir bags and face-masks in a range of sizes (masks that are too large may be used inverted)
- Nasal cannulae
- Airway devices
  - Oropharyngeal airways – range of sizes (000, 00, 0, 1, 2, 3)
  - Endotracheal tubes – range of sizes (2.5–7.5 mm) and connectors*
- Laryngoscopes *
  - adult curved and paediatric straight-bladed
  - spare bulbs and batteries
- Magill forceps *
- Cannulae for cricothyoidotomy

Vascular access

- Peripheral vascular cannulae range of sizes: 18–25 gauge
- Intraosseous needles 16–18 gauge
- Sterile catheter (suction, feeding, etc.) for umbilical access (for newborn); umbilical vessel dilator; artery forceps
- Central venous catheters
- Syringes, incl 50 ml for fluid boluses + three-way tap
- Intravenous giving set and graduated burette

Trauma

- Hard cervical collars and sandbags/foam blocks
- Peripheral vascular cannulae, three-way taps and syringes
- Scalpels, sutures, needle holders and scissors
- Splints
- Chest drains: range of sizes; dissecting forceps; underwater drainage system, or flap valves
- Nasogastric tubes – range of sizes

Drugs, fluids, etc.

- Oxygen supply
- Saline 0.9% – vials and bags/bottles
- Colloid – for example albumin 4.5%, gelatin, hetastarch
- Epinephrine 100 micrograms/ml (1 in 10 000) and 1 mg/ml (1 in 1000)
- Glucose 10%, 25% and 50%
- Mannitol 20%
- Diazepam rectal solution
- Lorazepam or diazepam for IV
- Phenytoin phenobarbitone, paraldehyde
- Atropine
- Sodium bicarbonate
- Broad-spectrum antibiotic, for example cefotaxime, ceftriaxone
- Quinine
- Morphine/diamorphine
- Naloxone
- Insulin
- Local (for example lidocaine) and general anaesthetic agents (for example ketamine) *
- Paralysing agents *
- Skin cleansing solution (for example chlorhexidine, alcohol)
- Steroids (prednisolone, hydrocortisone)
- Salbutamol
- Furosemide

Monitoring and other equipment

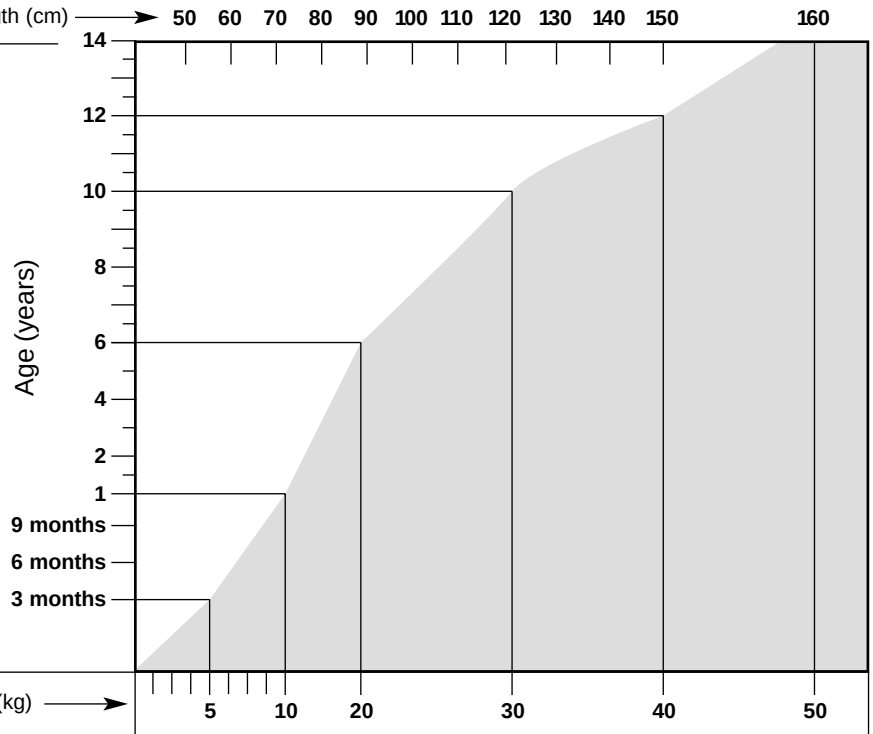
- Pulse oximeter
- ECG monitor – defibrillator
- Sphygmomanometer or blood pressure oscillometry
- Thermometer (ideally low reading)
- Blood/urine glucose testing kits
- Urinary catheters (silicone, rubber or soft feeding tubes) of various sizes

* These items are to be used only if facilities exist for intubation and assisted ventilation, for example on an intensive care unit.

Paediatric resuscitation chart

Endotracheal tube

Oral length (cm)	Internal diameter (mm)
18–21	7.5–8.0 *cuffed*
18	7.0 *cuffed*
17	6.5
16	6.0
15	5.5
14	5.0
13	4.5
12	4.0
	3.5
10	3.0–3.5



	5	10	20	30	40	50
Epinephrine (ml of 1 in 10 000) *initial* *intravenous of intraosseous*	**0.5**	**1**	**2**	**3**	**4**	**5**
Epinephrine (ml of 1 in 1000) *subsequent* *intravenous or intraosseous (or intial endotracheal)*	**0.5**	**1**	**2**	**3**	**4**	**5**
* **Atropine (ml of 100 micrograms/ml)** *intravenous or intraosseous (or double if endotracheal)*	**1**	**2**	**4**	**6**	**6**	**6**
Atropine (ml of 600 micrograms/ml)	**–**	**0.3**	**0.7**	**1**	**1**	**1**
Bicarbonate (ml of 8.4%) *intravenous or intraosseous (dilute to 4.2% in infants)*	**5**	**10**	**20**	**30**	**40**	**50**
Diazepam (**ml of 5 mg/ml** *emulsion)* *intravenous or rectal*	**0.2**	**0.5**	**1.0**	**1.5**	**2**	**2**
Diazepam (mg *rectal tube solution*) *rectal*	*2.5 mg*	*5 mg*	*10 mg*	*10 mg*	*10 mg*	*10 mg*
Glucose (ml of 10%) *intravenous or intraosseous*	**25**	**50**	**100**	**150**	**200**	**250**
* **Lidocaine (ml of 1%) 10 mg in 1 ml** *intravenous or intraosseous*	**0.5**	**1**	**2**	**3**	**4**	**5**
Naloxone *neonatal* (ml of 20 micrograms/ml) *intravenous or intraosseous*	**2.5**	**5**	**–**	**–**	**–**	**–**
Naloxone adult (ml of 400 micrograms/ml)	**–**	**0.25**	**0.5**	**0.75**	**1**	**1.25**
* **Salbutamol** (**mg** *nebuliser solution*) *via nebuliser (dilute to 2.5–5.0ml in normal saline)*	*–*	*2.5 mg*	*5 mg*	*5 mg*	*5 mg*	*5 mg*
Initial DC defibrillation (J) *for VF or VT with no pulse*	*10*	*20*	*40*	*60*	*80*	*100*
Initial DC cardioversion (J) *for SVT with shock (synchronous) or VT with shock (non-synchronous)*	*5*	*5*	*10*	*15*	*20*	*25*
Initial fluid bolus in shock (ml) *crystalloid or colloid*	**100**	**200**	**400**	**600**	**800**	**1000**

* ***CAUTION!*** *Non-standard drug concentrations may be available:*
*Use **atroprine** 100 micrograms/ml or prepare by diluting 1 mg to 10 ml or 600 micrograms to 6 ml in normal saline.*
*Use **lidocaine** (without epinephrine) 1 % or give twice the volume of 0.5 %. Give half the volume of 2 % or dilute appropriately.*
***Salbutamol** may also be given by slow intravenous injection (4–6 micrograms/kg), but beware of the different concentrations available (for example 50 and 500 micrograms/ml).*
From: Oakley et al. *Advanced Paediatric Life Support*, 2nd ed. London, BMJ Books 1997.

Figure 1.17.1

B For care and treatment

Table 1.17.1 Essential equipment for the wards

To achieve minimum standards	To improve standards
Basic ward cleaning	
Brooms, buckets, mops, flannels	
Elbow operated taps	
Hand-washing resources; liquid soap dispensers, hand-driers or towel dispensers	
Incinerator	
Industrial washing machines	
Sharps disposable boxes/tins	
Waste bins and plastic bag liners	
Anaesthetics, respiratory support and resuscitation	
Ayre's T-piece with Jackson-Rees modification and open ended 500 ml bags	Blood warmer
Chest tube drainage kit (underwater seal)	Blood gas machine with electrolyte and glucose measurement
Clocks	Boyles machine
CPAP systems	CNEP
Emergency drug and equipment boxes	Cooling blanket
Guedal oropharyngeal airways of various sizes 0–4	Expired carbon dioxide monitor
Gum elastic bougies	Transillumination system
Hard cervical collars	Triservice anaesthetic apparatus
Heimlich valves	Transcutaneous Pco_2 monitors
Laryngoscopes with different blades	Vaporisers
Magill forceps	Ventilators (volume and pressure limited)
Nebulisers	
Oxygen concentrators	
Oxygen cylinders with high (1 to 15 litres per minute) and low (0 to 2 litres per minute) flowmeters	
Oxygen masks	
Oxygen tubing (bubble type)	
Pulse oximeters	
Resuscitaires	
Self-inflating resuscitation bags (infant and adult) with non-rebreathing valves and reservoir bags (500 and 1600 ml)	
Spare laryngoscope bulbs	
Suction (manual and electric)	
Imaging	
Ultrasound scanner	CT scanner
X ray equipment	Echocardiography
	Endoscopy kit plus ability to undertake sclerotherapy
	MRI scanner
	Portable *X* ray
Cardiovascular support	
Autolets	Automatic blood pressure machines
ECG monitor	Defibrillator

To achieve minimum standards	To improve standards
ECG machine	Portable ECG machine
IV stands	Syringe drivers
Tourniquets	Syringe pumps
Simple blood pressure machines with cuffs of a range of different sizes infant to adult	
Examination aids	
128 Hz tuning forks	Audiometry
Bright torches	Bed scales
Examination lights	Binocular indirect ophthalmoscope
Height/length measures	EEG
Icterometer	EMG
Kitchen scales	Nerve conduction equipment
Microscope	Slit lamp
Ophthalmoscope	Tonometer
Otoscope	
Polaroid camera	
Scales for infants and for children	
Vision testing charts	
Stethoscopes	
Tape measure	
Tendon hammers	
Thermometers (including low-reading)	
Toys for skill assessment	
Basic ward equipment	
Beds with polythene covered mattresses	Impregnated mosquito nets
Bowls	
Chairs for mothers to sleep next to their children	
Cold boxes	
Fans	
Infant cots with safety sides	
Potties	
Refrigerators	
Room heaters	
Sheets	
Towels	
Trolleys with protective sides	
Washable blankets	
Special needs	
Aids for disabled children	
Glasses for refractive errors of vision	
Hearing aids	
Limb prostheses	
Low cost visual aids	
Special aids for disabled children, for example, ankle splints, foam wedges, footboards, Piedro boots, wheelchairs, parallel bars, toilet aids, hand function aids	
Vision testing charts	
Wheelchairs	
Ward instruments	
Curved and straight artery forceps	Jejunal biopsy

To achieve minimum standards	To improve standards
Needle holders	
Scalpels	
Scissors	
Neonatal equipment	
Closed incubators	Breast pumps
Infant eye shields	Convective heated blanket
Phototherapy unit	
Platform incubator	
Head boxes	
Theatre equipment	
Autoclaves	Air purifier and conditioner for operating theatre
Basic surgical instruments including burr hole system	Binocular telescopic magnifying lenses
Biopsy needles (Trucut or Menghini)	ICP monitoring equipment
Cautery machine	Neuroendoscopy
Operating theatre light	
Operating theatre table with lithotomy position	
Sterile drapes	
Sterilizer	
Surgical hats and boots	
Miscellaneous	
Internal hospital communications system	Computer data base systems
Log books for narcotics	Digital camera
Safe storage for narcotics	Haemodialysis
Library, overhead projector, computer with Internet access, blackboard, chalk, flipcharts, and pens	Multimedia projector
Emergency generator	Radiotherapy
Medical records system	Slide projector
	Video recorder and television

Table 1.17.2 Essential supplies

To achieve minimum standards	To improve standards
For the ward	
Alcohol swabs	Infusion sets for syringe pumps and volume infusion pumps
Blood collecting systems, transfusion screening kits, cross-match kits	Long-term central venous catheters (Broviac or Hickman)
Bone marrow needles	Tenckhoff catheters
Bubble wrap	
Butterfly needles 18–24 gauge	
Charts for drugs, fluid balance, growth, weight, head circumference, temperature/pulse/blood pressure and oxygenation	
Disposable razors	
In-line blood filters (16–20 μm)	
Intraosseous needles infant and paediatric	
IV cannulae 25–14 gauge	
IV giving sets with and without in-line infusion burette chambers	
Laminated charts for the wall	
LP needles 22–18 gauge	
Lubricant jelly	
Medical and nursing records	
Multilumen central venous catheters	
Name bands	
Nasogastric tubes sizes 6, 8, 10, 12, 14, 16 FG	
Needles 14 to 25 gauge	
Rubber gloves (sterile and non-sterile) of different sizes (ideally disposable)	
Seldinger wires	
Sharps boxes	
Soap (preferably liquid)	
Spatulae	
Silk sutures and needles 3/0 and 4/0	
T-pieces with extension	
Three-way taps	
Towels for hand-drying (paper ideally)	
Tracheostomy kits and tubes sizes from 3 mm to 8 mm i.d.	
Urine catheters sizes 6, 8,10,12 FG	
Urine collection bags	
Vaccine vial monitors	
Cleaning materials	
Floor, wall, sink, toilet disinfectants and detergents	
Theatre materials	
Isolation gowns, aprons, eye protection, gloves	
Plastic bags for waste	
Specimen pots	
Shunts for hydrocephalus	
Sterile and non-sterile latex gloves of different sizes (ideally disposable)	
Surgical masks	
Ward investigations	
Blood glucose sticks	
Blood sample bottles	

To achieve minimum standards	To improve standards
CSF sample bottles	
Litmus papers	
Microbiology swabs	
Microscope slides with chambers, grids and cover slips	
Sterile fluoroscein strips	
Throat swabs	
Tongue depressors	
Urine stick tests for protein, bilirubin, glucose, urobilinogen, and ketones	
WHO haemoglobin strips	
Tapes, dressings and bandages	
Adhesive tape (elastic and non-elastic)	Alginate dressings
Butterfly wound-closing plasters	
Crêpe bandages	
Eye pads	
Gauze bandages	
Plaster of Paris bandages	
Plasters	
Splints	
Sterile cotton wool	
Sterile pads (including non-adherent)	
Vaseline gauze pads	
Emergency supplies	
Chest drains (sizes 8–32 FG)	
Cricothyroidotomy kits (infant and paediatric)	
Electrode gel	
Endotracheal tube introducers of different sizes	
Endotracheal tubes 2.5–8.0 mm cuffed and uncuffed	
Nasopharyngeal catheters for oxygen (8FG)	
Oxygen face masks (paediatric and adult)	
Short nasal cannulae for oxygen (infant, paediatric and adult)	
Sterile umbilical cord clamps	
Suction catheters sizes (6–16 FG)	
Yankauer suckers (paediatric and adult sizes)	

Table 1.17.3 Essential drugs for the hospital (derived from chapters in this manual)

To achieve minimum standards	To improve standards
Antimicrobials	
Acyclovir IV or oral and ointment	Aminosidine*
Albendazole	Amoxycillin-clavulanic acid
Amiodaquine	Azithromycin
Amoxycillin (oral and IV)	Cefixime
Amphotericin B	Cefotaxime
Ampicillin	Clarithromycin
Artemether*	Clindamycin
Artenusate* IV	Eflornithine*
Ceftazidine	Folinic acid
Ceftriaxone	HAART
Cefuroxime	Halofantrine*
Cephalexin	Imipenem
Chloramphenicol	Kanamycin
Chloramphenicol oily for IM	Levamisole
Chloroquine (IV and oral)	Liposomal amphotericin B
Ciprofloxacin	Mefloquine*
Clofazimine*	Meropenem
Cotrimoxazole	Minocycline*
Dapsone*	Nalidixic acid
Doxycycline	Ofloxacin*
Erythromycin IV and oral	Proguanil*
Ethambutol	Quinidine*
Ethionamide	Rifabutin
Flucloxacillin or cloxacillin or oxacillin IV and oral	Streptomycin
Fluconazole	Sulphadiazine
Fusidin (IV and oral)	Thiacetazone
Ganciclovir IV	Tinidazol
Gentamicin	
Griseofulvin	
Isoniazid (oral and IV)	
Ivermectin*	
Ketoconazole	
Mebendazole	
Melarsoprol*	
Metakelfin*	
Metriphonate*	
Metronidazole	
Nevirapine	
Nitrofurantoin	
Penicillin (oral, IV, long-acting)	
Pentamidine*	
Pentavalent antimony (sodium stibogluconate or meglumine antimoniate)*	
Praziquantel*	
Pyrantel*	
Pyrazinamide	
Pyrimethamine with sulfadoxime	

To achieve minimum standards	To improve standards
Quinine IV and oral	
Rifampicin	
Suramin*	
Tetracycline	
Trimethoprim	
Vancomycin	
Steroids	
1% hydrocortisone cream	Methyl prednisolone
Betamethasone 0.1% drops and ointment	Nebulized beclometasone
Dexamethasone	Stanozolol
Prednisolone	Triamcinolone joint injection
Hydrocortisone	
Fludrocortisone	
Hydrocortisone 1% eye drops and 0.5% eye ointment	
Inhaled beclomethasone	
Prednisolone enema	
Gases	
Ether	Halothane
Nitrous oxide	Nitrous oxide
Oxygen	Trichlorethylene
Vaccines and specific immunoglobulins	
Diphtheria antitoxin	Botulinum antitoxin
Rabies antitoxin	Hepatitis C vaccine
Tetanus antitoxin (ideally human)	IV gamma globulin
EPI vaccines	Measles IgG
Haemophilus influenzae vaccine	Meningococcal vaccines
Hepatitis B vaccine	Rabies vaccine
Mantoux tuberculin	Typhoid vaccine
Pneumococcal vaccine	Varicella vaccine
	VZIG
IV and other fluids plus blood products	
0.9% Saline	
10% Glucose	20% albumin
10% Glucose in 0.9% saline	Blood clotting factors
50% Glucose	Cryoprecipitate
Blood cross-matched, infection screened	Fresh frozen plasma
O–negative	Platelets
Packed cells	
Dextran 40	
Gelofusin	
Glucagon	
Hartmann’s	
Heparin	
Peritoneal dialysis fluid (1.36 and 3.68%)	
Diuretics	
Chlorthiazide	
Furosemide	

To achieve minimum standards	To improve standards
Mannitol	
Spironolactone	
Paralysing drugs	
Atracurium, pancuronium or vecuronium	
Suxamethonium	
Local anaesthetics	
1% Lidocaine	
1% lidocaine plus epinephrine 1 in 200 000 (5 micrograms/ml)	
Analgesics and anti-inflammatory drugs	
Aspirin	Fentanyl (IV and transdermal)
Codeine	Hydromorphine
Diamorphine	Methadone
Dihydrocodeine	Mesalazine enema
Diclofenac (tablets and suppositories)	
Ibuprofen	
Ketamine	
Morphine (oral short and long acting, IV, subcutaneous)	
Paracetamol (tablets, suspension and suppositories)	
Sulphasalazine	
Broncho-dilators and respiratory stimulants	
Aminophylline	Theophylline
Caffeine	
Inhaled salbutamol	
Salbutamol IV injection	
Salbutamol nebuliser solution	
Cardiovascular drugs	
Adenosine	Dobutamine
Atenolol	Hydralazine
Atropine	Nifedipine
Captopril	Prostaglandin E
Digoxin (oral and IV)	
Dopamine	
Epinephrine (1 in 1000 =1 mg/ml and 1 in 10 000 =100 micrograms/ml)	
Glyceryl trinitrate	
Labetalol (oral and IV)	
Prazosin	
Antihistamines	
Chlorphenamine	Promethazine
Drugs applied to body surfaces	
1% Crotamiton cream	Calamine lotion
1% Gamma benzene hexachloride lotion	
1% Gentian violet	Ephedrine nose drops
1% Potassium permanganate solution	5% Permethrin cream

To achieve minimum standards	To improve standards
10–25% Podophyllin	70% alcohol handwash
15% aminasidine and 12% methyl benzathonium chloride ointment*	Alcohol spray
25% Benzyl benzoate emulsion	
50% White soft paraffin plus	
50% liquid paraffin	
5–10% Sulphur in white soft paraffin	
95% Alcohol	
Acyclovir 3% eye ointment	
Acyclovir cream	
Amphotericin B suspension or lozenges	
Antifungal eye drops	
Antiseptic ointment, for example iodine	
Aqueous cream	
Atropine 0.5% eye drops and 1% ointment	
Cetrimide	
Chloramphenicol 0.5% eye drops and 1% ointment	
Chloramphenicol eye drops	
Chlorhexidine	
Chlortetracycline 1% eye ointment	
Ciprofloxacin 0.3% eye drops	
Clotrimoxazole 1%	
Collodion	
Cyclopentolate 0.5% eye drops	
Fluorescein 1% eye drops	
Fucidin 1% eye drops	
Fucidin cream	
Gentamicin eye drops 0.3%	
Ketoconazole shampoo	
KY jelly	
Miconazole 2% gel or cream	
Mupirocin cream	
Nystatin cream	
Nystatin oral suspension	
Petroleum jelly	
Povidone iodine solution (10%)	
Saline nose drops	
Selenium sulphide shampoo	
Tetracaine 0.5 and 1% eye drops	
Tetracycline eye drops	
Tetracycline eye ointment	
Topical thiabendazole*	
Trichloracetic acid	
Tulle gras	
Vioform	
White soft paraffin	
Zinc and castor oil ointment	

To achieve minimum standards	To improve standards
Vitamins	
1 α-hydroxyvitamin D	
Multivitamin drops	
Nicotinic acid	
Pyridoxine	
Thiamine (oral or IV)	
Vitamin A (oral and IM)	
Vitamin D (calciferol, cholecalciferol)	
Vitamin E	
Vitamin K	
Electrolyte solutions	
10% Calcium gluconate	
15% KCl	
30% NaCl	
Buffered phosphate solution	
Calcium carbonate	
Calcium gluconate	
Copper	
Ferrous fumarate syrup	
Ferrous sulphate	
Ferrous sulphate with folic acid	
Folic acid	
Hypertonic 10% saline	
Hypostop gel	
Magnesium	
ORS	
Phosphorus solution 4 mmol/ml	
Potassium chloride (IV and oral)	
ReSoMal	
Sodium bicarbonate (8.4% and tablets)	
Sodium chloride	
Zinc	
Special foods	
F75 and F100 foods	Cholestyramine
Pancreatic enzyme supplements	Elemental diet
	Gluten-free diet
Anticonvulsants	
Carbamazepine	Ethosuximide
Clonazepam	Thiopentone
Diazepam (oral, IV and rectal)	Lorazepam
Paraldehyde	
Phenobarbital (oral and IV)	
Phenytoin	
Sodium valproate	
Sedatives	
Chloral hydrate	Chlormethiazole
Chlorpromazine (oral and IV)	Methotrimeprazine
Flupenthixol	
Haloperidol	
Midazolam (oral and IV and rectal)	

To achieve minimum standards	To improve standards
Hormones	
Carbimazole	DDAVP
Insulin	Growth hormone
Levothyroxine	
Lugol's iodine, potassium iodide	
Testosterone esters	
Drugs for treating poisoning	
Activated charcoal	Ferrifox
Calcium Resonium	Flumazenil
d-penicillamine	Kelfer
Desferrioxamine	
EDTA	
Methionine	
Mono- or polyspecific antivenoms	
N-acetylcysteine	
Naloxone (neonatal and adult preparations)	
Neostigmine	
Paediatric ipecacuanha	
Pralidoxime	
Laxatives	
Docusate sodium	Co-danthramer/co-danthrusate
Enemas (Microlax, phosphate, arachus oil)	
Glycerol suppositories	
Lactulose	
Picosulphate sodium	
Senna	
Disinfectants	
Bleach (1 in 10 and 1 in 100)	
Chlorhexidine 0.5 %	
Hibitane	
Anti-emetics	
Cyclizine	Metoclopramide
Domperidone	Ondansetron or other $5HT_3$ serotonin antagonist
Prochlorperazine (tablets, suspension and suppositories)	
Antacids	
Aluminium hydroxide	Omeprazole
Cimetidine	
Magnesium carbonate	
Ranitidine	
Sucralfate	
Antidepressants or other psychotropic drugs	Clozapine
Amitriptyline	Lithium
SSRIs (for example fluoxetine)	
Miscellaneous	Baclofen
Barium	Clonidine
Buscopan	Edrophonium

To achieve minimum standards	To improve standards
	Hyoscine hydrobromide
	Pizotifen
	Sodium benzoate
	Sumatriptan
	Thalidomide
	Tranexamic acid
	Ursodeoxycholic acid
Chemotherapy drugs	Azathiaprine
	Cyclophosphamide
	Cyclosporin
	Methotrexate
	Specific anticancer drugs
	Vincristine

*Only in countries where the diseases treated by the drug are endemic.

118 Records, history taking and examination

Christiane Ronald

Records

- These can be held by parents, by the hospital or both.
- If parent held, they can be developed into health booklets containing advice on how to manage illnesses (possibly in the form of pictures for illiterate parents). Information in them about immunisations should comply with national immunisation programmes.
- Hospital records should be kept safe and confidential. They should also be stored in a logical system for audit purposes and for easy access.
- Discharge information should be entered in the parent held booklet.
- If possible, diagnoses should be coded and entered according to the International Classification of Diseases.

History taking

- Medical history taking should, when age appropriate, include the child. The source of the information, maybe the mother, the father or the child itself, should be documented.
- It is important to listen, especially to the mother's worries about her child, taking into account her general frame of mind, experience with previous children and her ability to communicate.
- Time can be a restricting factor due to the workload, but it is important to ask about:
  - Pregnancy and previous deliveries – including stillbirths
  - Feeding history
  - Immunisation record (best kept by the mothers)
  - Previous admissions or visits to hospital
  - Social circumstances at home and family history
  - Cultural belief/religion/tribe
  - Medication and allergies
  - Presenting complaints and current treatment, if any
- Most mothers are anxious when their child is ill. They need reassurance, kindness and understanding.

Examination

- A triage nurse (see Chapter 1.19) can be helpful in making a preliminary assessment of patients. They can assess each child and decide with the help of recording temperature, weight, general condition and pain score how urgently he/she should be seen by the doctor.
- Do not rush the examination. A thorough examination is often needed and taking time can help gain the confidence of parent and child.
- If the child is critically ill, quick action is required and questions can be asked later.
- Try to be gentle and avoid palpating a painful body part before everything else is done. The last thing you want is a crying child whom you cannot examine or auscultate.
- Small children and infants are best examined on the parent's lap, bigger ones can be asked to lie down.
- In general, the examination of a child will follow the same systematic approach as in adults, however, you may need to be more opportunistic.
- Essential examination checklist (see PLS, Chapter 1.19)
  - **A**irway
  - **B**reathing
  - **C**irculation
  - **D**isability
  - **E**xposure
- Critically ill child: proceed to paediatric life support.

Child not in need of immediate resuscitation

- General inspection: dysmorphism, skin rashes or bruises, nutritional status, weight and height for age, jaundice, pallor, clubbing, relationship with parent, state of consciousness.
- Respiratory system: document retractions, respiratory rate, cyanosis, palpation, percussion, auscultation, rhythm (presence/episodes of apnoea?)
- Cardiovascular system: do not forget to feel all pulses, particularly the femoral pulses, look for clubbing, measure the blood pressure (cuff MUST cover two-thirds of the upper arm circumference), examine jugular venous pressure (in child), palpate the cardiac impulses (that is for left and right ventricles), auscultate apex, left sternal edge, pulmonary and aortic areas and carotids and over the back.
- Abdominal system: in neonates, inspect the umbilicus, check genitals for cryptorchidism, hernias, gender. Rectal examinations are occasionally necessary but

need to be explained to the parent and child (where appropriate). Inspect mouth and teeth

- Neurological system: use the modified Glasgow coma score (see Chapter 3.36). Observe infants for their degree of responsiveness and rapport appropriate for age, social and motor skills, look for neurocutaneous stigmata. Test for age appropriate reflexes and saving reactions when assessing developmental delay. Leave sensation testing until last. Ideally fundoscopy needs mydriatics, a dark room and occasionally sedation of the child.
- Motor system: always examine infants for dislocated/dislocatable hips. Check gait.

Parents have the right to be told any abnormal findings if these are relevant to the child's illness or important. The actual process of the examination should be explained to the child in age-appropriate language.

The history and examination findings including the child's weight and height should be recorded, with daily entries on management and progress. When the child is discharged parents should have discharge information about the child's admission and any further treatment needed to share with their primary care health workers.

A detailed description of a paediatric examination is beyond the scope of this manual and we can recommend the books listed in the Further reading section.

See Section 7 for samples of various charts including vital signs, fluid balance, growth and body mass index.

Further reading

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McMaster P, McMaster H, Simunovic V, Selimovic N, Southall DP. Parent and young person held child health record and advice booklets and their use in Bosnia and Herzegovena. *International Child Health* 1995; **6**: 121–31.

Stephenson T, Wallace H. *Clinical Paediatrics for Postgraduate Examinations.* 2nd ed. Edinburgh: Churchill Livingstone, 1995.

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119 Essentials of triage and paediatric life support

Mary Limebury

Seeing the sickest first: triage

Triage all sick children

If any sign positive:
- give treatment(s),
- call for help,
- draw emergency labs (glucose, malaria smear, Haemoglobin)
- then assess response to initial treatment

Priority signs:

These children need prompt assessment and treatment:

- Visible severe wasting
- Oedema of both feet
- Severe palmar pallor
- Any sick young infant (< 2 months)
- Lethargy
- Continually irritable and restless
- Major burn
- Any respiratory distress
- An urgent referral note from another facility

If a child has trauma or other surgical problems, get surgical help or follow surgical guidelines

Non-urgent

Then proceed with care and directed assessment and further treatment according to the child's needs and priority. Cards given to parents containing their triage classification and timing of assessment can be useful. Different colours and time codes can be an easy way of efficiently scoring the patients.

Minimum equipment required

- Bag and mask system with various mask sizes and baby and child sized self-inflating bags
- Oxygen and masks/tubing
- Magill forceps
- Endotracheal tubes, 2.5–8.0 mm internal diameter
- Laryngoscope and blades: straight/curved
- Suction equipment (mechanical not just mucus extractors hand/foot-operated) ***electrical/wall***
- Guedal airways: various sizes
- Intravenous access/intraosseous access

Table 1.19.1 Emergency signs, assessment and treatment (adapted from WHO)

Area of assessment	Clinical signs	Result	Treatment: if any sign positive or coma or convulsing, give treatment(s), call for help, draw emergency bloods (glucose, malaria smear, Hb, electrolytes) then assess response to initial treatment
Assess: Airway and breathing	• Obstructed breathing or • Central cyanosis or • Severe respiratory distress	**any sign positive**	Manage airway Give oxygen Make sure child is warm
Assess: circulation	Shock • Capillary refill >2 sec after 5 sec pressure on sternum	**any sign positive**	Stop any bleeding Give oxygen Make sure child is warm
→	• Cold/pale/sweating • Weak and fast pulse • Bradycardia? (listen with stethoscope)	Check for severe malnutrition	**If no severe malnutrition**: Insert IV and give 10–20 ml/kg 0.9% saline or colloid as rapidly as possible. If not able to insert peripheral IV, insert external jugular, femoral venous, long saphenous cut down or intraosseous line

Table 1.19.1 *Continued*

Area of assessment	Clinical signs	Result	Treatment: if any sign positive or coma or convulsing, give treatment (s), call for help, draw emergency bloods (glucose, malaria smear, Hb, electrolytes) then assess response to initial treatment
			If severe malnutrition: • Give IV glucose 10% 5 ml/kg • Proceed to full assessment and treatment (see Chapter 3.16)
Assess Neurological State →	• Agitated or depressed conciousness • Coma or • Convulsing (now)	**If Coma or convulsing**: Check for head/neck trauma before treating child – stabilise neck if cervical spine injury possible (see Chapter 5.4)	Manage airway Position child • If convulsing, give diazepam rectally (500 micrograms/kg) 2.5 mg if <1 year, 5 mg if 1–3 years and 10 mg >3 years • If no response give IV lorazepam 100 micrograms/kg or IV diazepam 250 micrograms/kg • Give IV glucose 10% 5 ml/kg or 2 ml/kg of 25 % Make sure child is warm

Minimum fluids

- 0.9% saline plus ability to add K/Ca
- 10% glucose
- Plasma expander (colloid/blood/plasma/4.5% albumin)
- Hartmann's (Ringer lactate)

Minimum drugs required

- Epinephrine 1 in 1000 (1 mg/ml), 1 in 10 000(100 micrograms/ml)
- Sodium bicarbonate 8.4%/4.2%
- Antibiotics: third generation cephalosporin/benzyl penicillin/fluoxacillin/metronidazole/amoxycillin
- Steroids: IV hydrocortisone/dexamethasone
- Morphine IV
- Diazepam/Midazolam IV
- Antimalarials (quinine)
- Mannitol
- Phenobarbitone/phenytoin/paraldehyde
- Salbutamol for nebulising
- Pancuronium/atracurium IV
- Atropine IV
- Insulin
- 10%/25%/50% glucose

Introduction

✔ **Paediatric cardiac arrest is rarely sudden but is usually the end result of respiratory failure, shock or neurological dysfunction.** Babies and children die mostly from respiratory, circulatory or neurological failure secondary to illness or trauma. Death is usually preceded by a period of illness during which time intervention and support may prevent deterioration and death. Recognition of this illness and appropriate intervention form the basics of Paediatric Life Support (PLS). PLS skills must be learnt by all medical/nursing personnel involved in the treatment of sick babies and children wherever they are being cared for.

ABC

- Intervention to support the seriously ill or injured child always follows the same scheme:
  Airway (plus cervical spine control in trauma)
  Breathing
  Circulation
- Without an airway the child cannot breathe, without breathing the child cannot be oxygenated, and without oxygenation the circulation is useless.
- The child who is sick may not have respiratory disease but may die of respiratory failure secondary to exhaustion.
- Providing good oxygenation and an adequate circulation prevents secondary damage (particularly to the brain) no matter what the primary disease process.

Airway

Must be opened and controlled first.

- If there is any possibility of cervical spine trauma then the neck should be immobilised for instance using bags of fluid and tape.
- Airway opening is performed using positioning of the head and neck – neutral in infant (<1 year), "sniffing" in the child (>1 year) – plus chin lift (not in trauma) or jaw thrust manoeuvres.
- Suction of the oropharynx under direct vision and of the nasophanynx may be required.

Breathing

- If the child is not breathing then give five "rescue" breaths – watch for movement of the chest to ensure

Table 1.19.2 Assessment for emergency conditions requiring immediate treatment (ETAT vs APLS)

	ETAT guidelines*	APLS course**
Airway and breathing	Obstructed breathing, or central cyanosis, or severe respiratory distress	Not breathing Work of breathing, assessed by: respiratory rate/rhythm recession (intercostal, subcostal or sternal) grunting accessory muscle use nasal flaring Auscultation: for wheeze and stridor to estimate air movement Skin colour: pallor or cyanosis
Circulation	Cold hands with: capillary refill >3 secs and weak and fast pulse	Skin temperature Heart rate Pulse volume Capillary refill >2 secs
Disability	Coma or Convulsing (now)	Mental status/conscious level Posture (hypotonia, decerebrate or decorticate) Pupils
Severe dehydration (only in child with diarrhoea)	Diarrhoea, plus any two of these: sunken eyes, lethargy, very slow skin pinch	

* ETAT, emergency triage assessment and treatment (*WHO management of the child with a serious infection or severe malnutrition*. WHO/FCH/CAH/00.1)
** APLS, Advanced Paediatric Life Support. Oakley et al. *Advanced Paediatric Life Support*, 2nd ed. London BMJ Books 1997

this is effective. If available use a bag and mask system with oxygen to inflate the child's chest, otherwise use mouth-to-mouth or mouth-to-mouth and nose.
- If breathing adequately, then give high flow oxygen by mask.
- If the airway requires protection then intubate the child.

Circulation

- Assess circulation by palpating for pulse – brachial or femoral easiest especially in small child/infant.
- Check capillary refill time: press on sternum for five seconds then release – pink coloration should return within two seconds.
- Check blood pressure.
- If no pulse or too slow and shock is present then commence cardiac compressions on the midsternum – two fingers, one fingerbreadth below the internipple line for babies, heel of one hand one fingerbreadth above the xiphoid for small children, two fingerbreadths above xiphoid for older children.
- Depress the sternum by at least one-third and give a rate of compressions of between 40 and 60 per minute – this allows time for the heart to fill between compressions. Most professionals fail to apply effective cardiac massage because they are frightened of damaging the baby.
- If compressions are effective femoral or brachial pulses should be palpable then follow the protocol for cardiac arrest.
- If pulse is present and adequate rate, but circulation is poor – capillary refill prolonged, tachycardia, low blood pressure (**warning**: this is a late sign) – then give boluses of fluid: use either 0.9% saline or a colloid preparation (4.5 % albumin, plasma, blood).
- Boluses should be given as 20 ml/kg (10–20 ml/kg in neonates) syringed in as quickly as possible and repeated until capillary refill is normal, tachycardia is improved and blood pressure maintained.

If circulation remains poor despite adequate fluid resuscitation then inotropes may be useful (dopamine, epinephrine).

120 Cardiorespiratory arrest

Simon Parke

Initial actions

- GET HELP
- "Are you alright?" (if old enough to understand)
- Airway opening
- LOOK, LISTEN, FEEL – for respiration
- Give five breaths with bag and mask if breathing absent or ineffective (ideally of 100% oxygen)
- Check pulse – brachial in infant, femoral in child and/or heart rate with stethoscope

Management

The management of a cardiac arrest can be divided into three distinct phases.

- Phase 1 Getting control – ABC
- Phase 2 Establishing cardiac output – treat any rhythm abnormality
- Phase 3 Consolidation or withdrawal – postresuscitation care

Preliminaries

- GET HELP
- Check time
- Open resuscitation box/bag

Phase one: ABC

First priority – establish airway and ventilation

- Open airway – head tilt, chin lift and jaw thrust (beware risk of cervical spine injury and if possible do only jaw thrust)
- Only do suction if airway blocked or filled with blood or vomit – thin secretions are not important
- Self-inflating bag with 100% oxygen through face mask – five initial breaths
- In absence of severe upper airway obstruction, adequate ventilation should be obtained
- If unable to inflate chest – check position
- Still unable to inflate chest – try Guedel airway
- Still unable to inflate chest – consider intubation

INTUBATION SHOULD BE ONLY DONE BY AN EXPERIENCED RESUSCITATOR

Age	Tube size
Full-term infant	3–3.5 mm
<1 year	4–4.5 mm
In children >1 year use this formula to calculate internal diameter in millimetre	Tube size $\pm 0.5 = \left(\frac{\text{Age}}{4}\right) + 4$

- Ensure the tube is passed only 2–3 cm below the vocal cords to avoid ventilating only one lung – the black line on the ET tube should *just* pass through the cords

After intubation check that lung inflation is occurring and is equal:

- Chest movement most useful sign
- Auscultate in axillae
- Chest *X* ray to check position – if prolonged ventilation needed

Failure to ventilate effectively may be due to:

- Incorrectly placed tube (oesophagus or right main bronchus)
- Pneumothorax

Ventilate at approximately 30 breaths per minute. This rate will be slower when external massage is required.

Do not forget that mouth to mouth/mouth-and-nose is effective if there are problems in obtaining equipment. ✔

Second priority – establish cardiac output

Reassess when ventilated. Pulses will often be palpable at this stage.

Cardiac massage

- Place the child on a firm surface (board, floor, examination couch).

  Infants: use two fingers to compress the sternum one fingerbreadth below the internipple line

  Small children (<9 years): use heel of one hand to depress the sternum, the area of compression is one fingerbreadth above the xiphisternum.

  Larger children (>8 years): the heels of both hands are used to depress the sternum, the area of compression is two fingerbreadths above the xiphisternum.

- Start with about 100 compressions/minute.
- Coordinate with ventilation: five compressions to one ventilation – i.e. 20 cycles/minute
- Aim to compress chest by about one-third of antero-posterior diameter.
- Effective massage always produces femoral pulses.
- Usual reason for ineffective massage is insufficient compression.
- Tamponade is a rare cause.

Third priority – establish IV line

This method depends on the expertise of the operator.
- Peripheral.
- Intraosseous.
- Internal jugular puncture/femoral vein.
- Cut down long saphenous vein.

Fourth priority – attach patient to ECG (if available)

- Commonest presenting rhythm: asystole: flat line, occasional P-wave.
- Pulseless electrical activity (PEA): absence of a palpable pulse despite the presence of recognisable complexes on the ECG monitor, most commonly caused by profound shock.
- Ventricular fibrillation.

Phase two

- Many children will be effectively resuscitated during Phase 1.
- If there is no monitor treat as pulseless electrical activity (PEA).
- Treat rhythm according to flowchart (see end of chapter).
- The "panic stage" should now be over.
- Correct "correctable" factors before starting something else.

  Severe dehydration: start 0.9% saline 20 ml/kg boluses
  Haemorrhage: start O-negative blood, 20 ml/kg boluses
- ✔ **Always give drugs time to work before starting something else.**

Drug therapy

Epinephrine
Give 0.01 ml/kg of 1 in 1000 solution (or 0.1 ml/kg of 1 in 10 000) IV or intraosseus and flush with 3–5 ml 0.9 % saline or give 0.1 ml/kg of 1 in 1000 solution via ET tube, then follow flowchart. For subsequent doses multiply the IV/intraosseous dose by 10 (i.e. 0.1 ml/kg of 1 in 1000).

Bicarbonate
The routine use of alkalising agents has not been shown to be of benefit. These agents should be administered only in cases where profound acidosis is likely, and should be considered if spontaneous circulation has not returned after the first dose of epinephrine. It should be remembered that good basic life support is more effective than alkalising agents at raising myocardial pH.

Use 1 mmol/kg (1 ml/kg of an 8.4% or 2 ml/kg of an 4.2% solution). ✔ **Do not use the intratracheal route. Bicarbonate must not be given in the same IV line as calcium because precipitation will occur. Sodium bicarbonate inactivates epinephrine and dopamine**, therefore flush the line with saline if these drugs are subsequently given.

Calcium and atropine are not recommended in the treatment of most cardiac arrests.

Antibiotics
Commence on broad-spectrum antibiotics.

Ventricular fibrillation (see flowchart)

- DC shock with defibrillator.
- ✔ **If you have not been trained in defibrillation find someone who is! The defibrillator is potentially very dangerous and must never be used by someone who is not competent.**
- Do not smear jelly between electrodes.
- Straddle heart with electrodes – i.e. right upper sternal border and over apex.
- Do not touch child or bed when shock delivered.
- Consider underlying cause (for example for hypothermia, commence active rewarming).

Phase three

Consolidation: postresuscitation care

Resuscitation successful following genuine cardiac arrest:
- Arrange for child to be ventilated for 24–48 hours if possible.
- Transfer to intensive care unit (if available).
- Treat, if possible, the underlying cause of the arrest.

All children should be monitored for:
- Pulse rate and rhythm (ECG monitor)
- Oxygen saturation (pulse oximeter)
- Core temperature (low-reading thermometer)
- Skin temperature
- Blood pressure (non-invasive monitor)
- Urine output (urinary catheter)

The following investigations should be performed as soon as possible following successful resuscitation:

Postresuscitation investigations

- Chest *X* ray: do not send child to *X* ray department unless accompanied by resuscitation team
- Haemoglobin, haematocrit and platelets
- Group and save serum for cross-match
- Na^+ K^+, urea and creatinine
- Bedside clotting time
- Blood glucose
- Liver function tests (if possible)
- 12-lead ECG – do not send child to department

When to stop resuscitation

If there have been no detectable signs of cardiac output, and there has been no evidence of cerebral activity despite up to 30 minutes of cardiopulmonary resuscitation, it is reasonable to stop resuscitation. The decision will be taken by the team leader, in consultation with the rest of the resuscitation team.

In many hospitals it may not be possible to ventilate children, particularly those who are small perhaps less than 15 kg. Therefore if these children have no respiratory effort *and* fixed dilated pupils after 30 minutes, even with good cardiac output, it may be reasonable to discontinue.

✔ **In these circumstances the child is not dead, so should not be moved or wrapped. The parents must have the situation explained to them.**

Exceptions to this rule include the hypothermic child (in whom resuscitation must continue until the core temperature is at least 32 °C or cannot be raised despite active measures) and children who have received overdoses of cerebral depressant drugs such as phenobarbital. In these cases prolonged resuscitation attempts will be necessary.

Further reading

Mackway-Jones K, Molyneux E, Phillips B, Wieteska S, eds. *Advanced Paediatric Life Support,* 3rd ed. London: BMJ Books, 2001.

Figure 1.20.1

Protocol for drugs in cardiac asystole

✔ **Before the administration of any drug the child must be receiving continuous and effective basic life support**

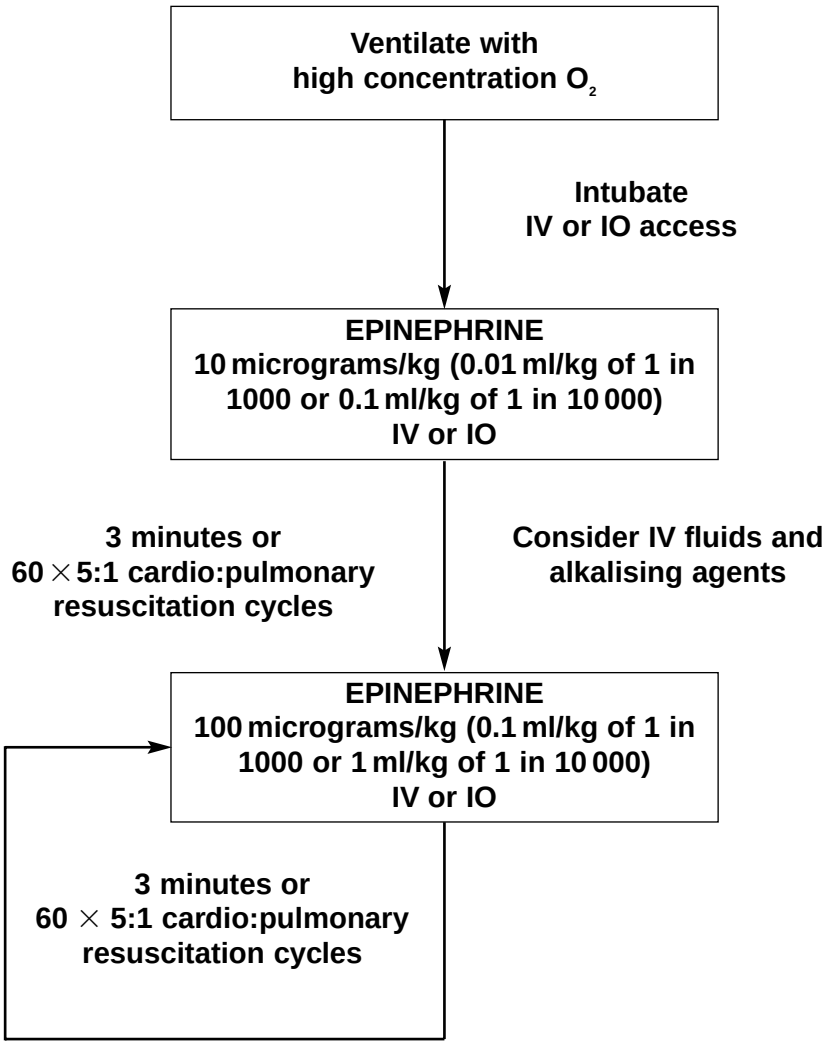


Figure 1.20.2

Protocol for pulseless electrical activity (PEA)

The ECG may look normal or show bradycardia and no pulse is palpable

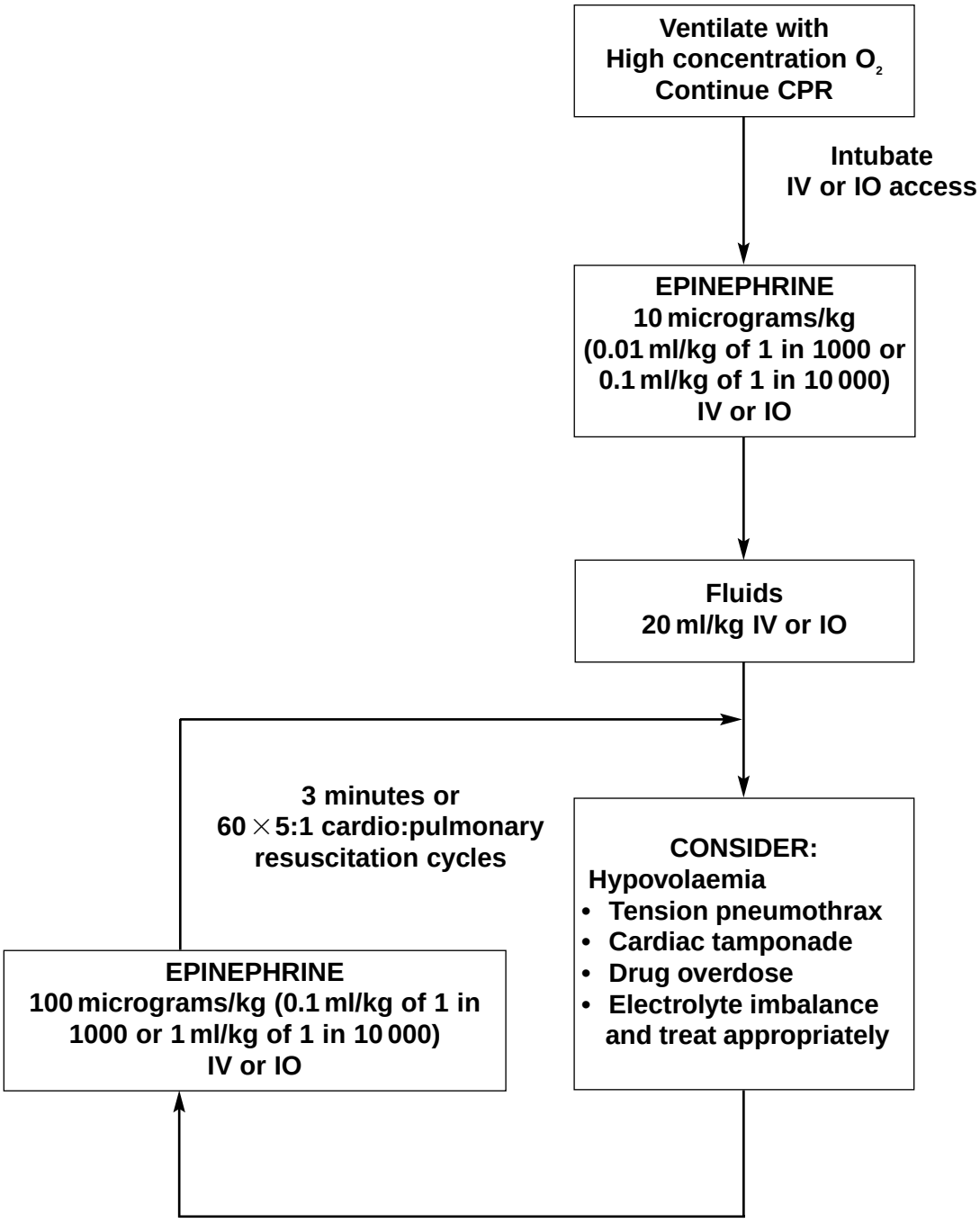
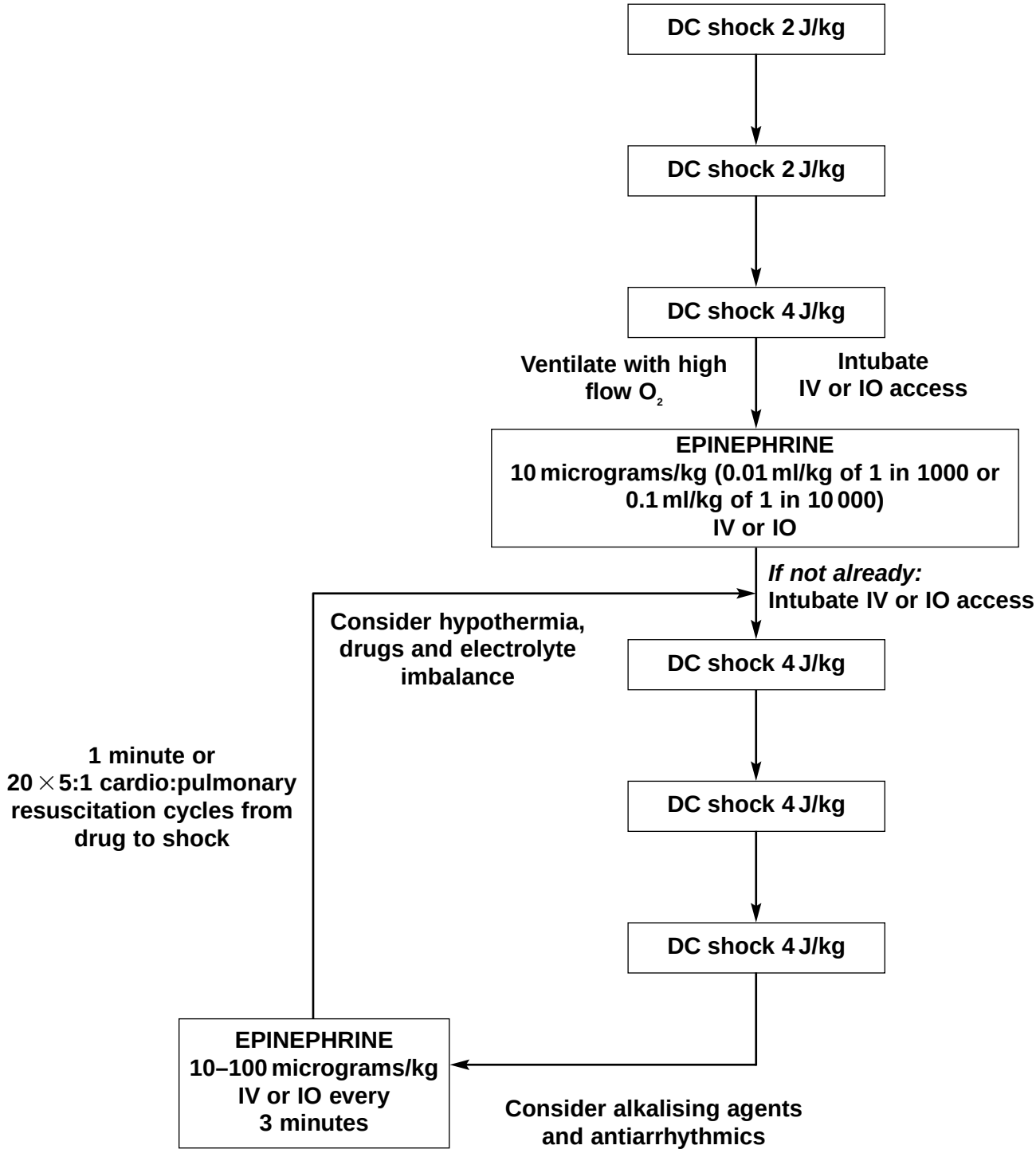


Figure 1.20.3

Ventricular fibrillation and pulseless VT



1.21 Intraosseous infusions for circulatory collapse

Simon Parke

(See also Chapter 6.10)

In children presenting in profound shock, normally secondary to profound dehydration or septicaemia, the siting of an intravenous line may prove to be very difficult. In these circumstances an initial intraosseous infusion will allow rapid expansion of the circulating volume. This will buy time for obtaining intravenous access and facilitate the procedure by increasing venous filling. If no intraosseous needles are available, you can use an 18 gauge bone marrow needle, a 21 gauge lumbar puncture needle or a 16 or 18 gauge plain needle (for example needle from blood giving set). **Sterility is very important**, the insertion site should be well sterilized with alcohol or iodine. **If the patient is conscious you MUST use local anaesthetic before insertion**.

The preferred sites are:

- Proximal tibia

Identify anteriomedial surface of tibia, 2–3 cm below the tibial tuberosity. Direct the needle distally from the epiphyseal plate at an angle of 90° to the skin. Continue to advance the needle until a 'give' is felt as the cortex is penetrated. Attach a 5-ml syringe and aspirate to confirm correct positioning. If using a needle rather than a 'proper' IO needle, flush with saline before insertion. Check position by gently flushing, watching insertion site closely (see Chapter 6.10).

- Distal tibia

Access site is on the medial surface of the tibia proximal to the medial malleolus.

Administration of a fluid bolus via an intraosseous needle

Purpose

To quickly administer an intraosseous fluid bolus to an infant or child who requires fluid resuscitation. (Fluid cannot be given by gravity alone.)

Equipment

Note: It may be necessary to improvise; remember sterility is important.

- 50-ml syringe (or largest available)
- IV giving set
- Three-way tap
- Wide-bore extension set

} these should all be found in the resuscitation box

- Resuscitation fluid (usually 0.9% saline, blood, 4.5% albumin or plasma)

Procedure

1. **Wash hands.**
2. Assemble the giving set, syringe and wide-bore extension set.
3. Turn the three-way tap off to the child and on to intraosseous fluid.
4. Draw up the required amount of fluid.
5. Turn off three-way tap to the fluid and on to the child.
6. Deliver the bolus (some pressure is usually required), returning the three-way tap off to the child after fluid is delivered.
7. Reassess the need for a further bolus.
8. Repeat the procedure.

Further reading

Harte FA, Chalmer PC, Walsh RF, Danker PR, Sheikh FM. Intraosseous fluid administration: a parenteral alternative in paediatric resuscitation. *Anaesth Analges* 1987;**66**:687–9.

Mackway-Jones K, Molyneux E, Phillips B, Wieteska S, eds. *Advanced Paediatric Life Support,* 3rd ed. London: BMJ Books, 2001.

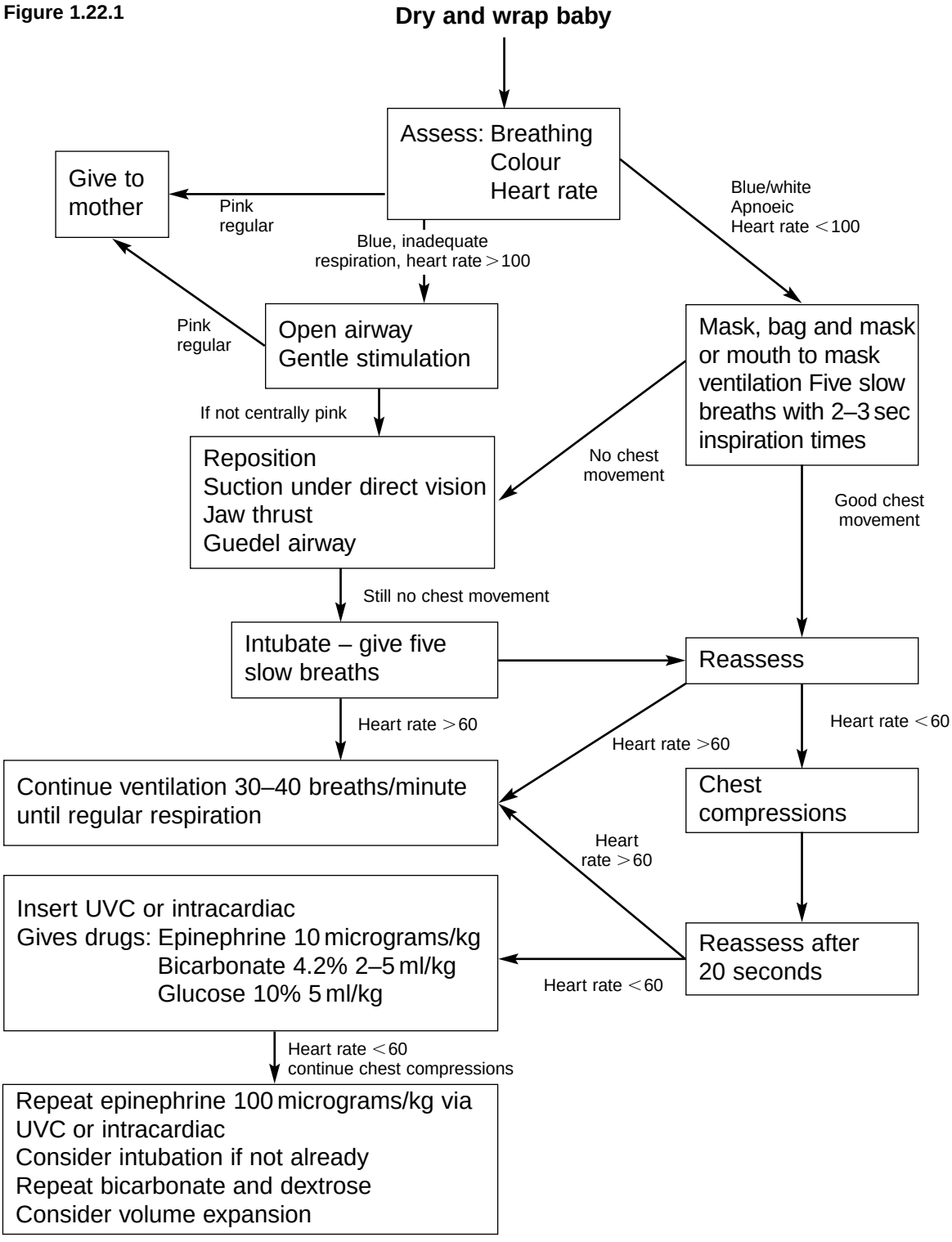
McNamara RM, Spivey WH, Unger HD, Malone DR. Emergency applications of intraosseous infusion. *J Emerg Med* 1987;**5**:97–101.

Spivey WH. Intraosseous infusions. *J Paediatrics* 1987;**111**:639–43.

1.22 Resuscitation of the newborn

Christiane Ronald and Simon Parke

Figure 1.22.1



- The place for resuscitation is the place of birth – do not move the baby until stable. If you need help, get someone from the baby unit.
- ✔ **Remember it is very traumatic for parents to witness their baby being resuscitated**, so provide explanations, reassurance or, in the event of death, meet and talk with them.

Before delivery

Most resuscitation in infants is unexpected – however it can be predicted in some circumstances.

- High risk deliveries in which a trained resuscitator should be present:
  - Breech delivery
  - Preterm delivery and severe IUGR
  - Multiple pregnancy
  - Fetal distress, for example ***persistent late decelerations***, cord prolapse, abruptio placentae, placenta praevia
  - Prolonged rupture of membranes (>24 hours)
  - Severe toxaemia
- Check the resuscitation equipment:
  - Warm dry towels ready – heat source on if possible
  - reinflating bag and appropriately sized masks available
  - Oxygen supply working and connected
  - Laryngoscope working
  - Suction device functioning
  - Endotracheal tubes available
  - Stethoscope

At delivery

- Note the time at the moment the baby is free from mother (start clock)
- Dry the baby and keep baby warm by wrapping and using radiant heat
- Assess condition – breathing/colour/heart rate

Normal respiration

First breath within seconds, regular respiration, heart rate >100

- Dry baby, wrap him/her in towels and give baby to mother.

Breathing impaired (see Appendix 7.10)

Respiration is shallow or irregular, heart rate >100

- Dry, place under heat source
- Facial oxygen and gentle tactile stimulation
- If improves hand to mother, if fails to improve bag and mask or mask and flow of gas at controlled pressure ventilation

Not breathing

GET HELP

No/weak respiratory movement, poor peripheral perfusion, heart rate <100

- Dry, wrap, place under heater
- Open airway – neutral position
- Ventilate with bag and mask or mask and flow of gas at controlled pressure with blow-off pressure release valve – use correct size mask – from above chin to bridge of nose.
- Give five initial breaths – each breath should take 2–3 seconds, at a pressure of about 30 cm H_2O
- Ensure the chest is moving by the fifth breath
- If no chest movement check position for airway
- Still no chest movement:
  - Inspect airway and suck under direct vision
  - Try jaw thrust
  - Consider Guedel airway
- If still no chest movement, the baby will need intubation
- Reassess after breaths – may need continued ventilation at rate of 30–40/minute

No respiratory movement, white/blue, heart rate <60

- Dry, place under heater
- Treat as above
- Check heart rate with stethoscope after 20 seconds IPPV, if still <60 start cardiac massage
- ✔ **Do not start chest compressions until ventilation achieved**
- Cardiac massage
  Two fingers or circle the chest with both hands and press the sternum with both thumbs 1 cm below internipple line:
  - rate 100/minute;
  - ratio – three to five compressions to one breath;
  - check that an output is achieved (heart rate > 100);
  - depress the sternum by about-one-third of the A–P diameter (usually 3 cm).

Table 1.22.1 Drug doses and indications

Drug	Indication	Dosage and route
Sodium bicarbonate 4.2%	Asystole Prolonged resuscitation	2–4 ml/kg IV/via UVC or intracardiac (see Chapters 6.8 and 6.16) (Not by ET tube)
Epinephrine 1 in 1000 (1 mg/ml)	Asystole	First dose: 10 micrograms/kg (0.01 ml/kg) IV/UVC or intracardiac or 30 micrograms/kg via ET tube Repeat dose: 100 micrograms/kg (0.1 ml/kg) IV, UVC or via ET tube if intubated or intracardiac
Glucose	Prolonged resuscitation Asystole	5 ml/kg 10% glucose IV, UVC or intracardiac (not by ET tube)

Remember – you only need to move oxygenated blood a few centimetres to the coronary arteries; the response to chest compressions is usually rapid.

- Assess heart rate and respiration every 30–60 seconds. Stop compressions when the heart rate is >100.

Drugs

If you need to use drugs the prognosis is very poor – there is no particular evidence for any order of drugs

- Drugs – Epinephrine, sodium bicarbonate, glucose
- Epinephrine can be given by UVC, IV, or intracardiac. *Only* epinephrine can be given endotracheally.
- When there is a strong suspicion of hypovolaemia due to blood loss in a neonate unresponsive to other resuscitative methods, uncross-matched O-negative blood may be used.
- If mother has received opiates consider naloxone 10 micrograms/kg IV or 100 micrograms/kg IM. (After breathing and circulation supported.)
- If no heart rate after 15 minutes then resuscitation should be terminated.
- If no spontaneous respirations after 30 minutes then resuscitation should be terminated.

Failure to inflate chest or oxygenate despite intubation

- ET tube in oesophagus
  Sometimes auscultation can be misleading, but genuine chest expansion will not be seen and auscultation is louder over the stomach than the lungs. Air will bubble up from mouth – heard in the neck with stethoscope (entry of air is less in left axilla than in right).
- Tube in right main bronchus
  Inflation improves as tube is withdrawn by 1–2 cms.
- Lung pathology
  - Pneumothorax (especially tension, see Chapter 6.11 for emergency thoracocentesis)
  - Diaphragmatic hernia (clue: scaphoid abdomen)
  - Hypoplastic lungs (clue: Potter's facies)
  - Hydrops fetalis or pleural effusion
- Technical problems
  - Oxygen may not be connected

No improvement in baby with good chest movement

Fetal haemorrhage

- Suspect if large APH or abruption, cord compression, vasa praevia, accidental incision of placenta during Caesarean section
- Give 0.9% saline plasma expanders or O negative blood 20–30 ml/kg via umbilical vein

Very severe asphyxia

- Acidosis may preclude recovery because it causes pulmonary hypertension and has a negative inotropic effect. If circulation is poor after 10 minutes of IPPV then give sodium bicarbonate 4.2% IV or intracardiac over 3–5 minutes: 2–4 ml/kg.
- IV glucose is not routinely given in asphyxiated infants, but the blood glucose should be checked as soon as the baby is resuscitated and again 30 minutes later. Hypoglycaemia must be treated promptly.

Meconium

Meconium is rarely a problem at birth – when meconium is present at delivery, try to aspirate the pharynx as the head is delivered. Only proceed to resuscitation if there is evidence of poor or absent respiratory effort. If there is quickly move the baby to the resuscitation table and check for meconium aspiration by direct laryngoscopy. When the trachea and the cords are clear, resuscitate in the usual way. Only worry about thick meconium – thin meconium is not a problem.

When meconium is present in the trachea:

- Meconium should be aspirated before the first inflation.
- Electively intubate with a large suction catheter (size 10 FG) or ET tube and apply suction directly to trachea.
- When meconium is very thick it will not pass through the ET tube or suction catheter but can be removed by suction as it is withdrawn.

Table 1.22.2 APGAR score

Clinical feature	Score		
	0	1	2
Heart rate	0	<100	>100
Respiration	Absent	Gasping or irregular	Regular or crying
Muscle tone	Limp	Diminished or normal with no movements	Normal with active movements
Response to pharyngeal suction	Nil	Grimace	Cough
Colour of trunk	White	Blue	Pink

✔ **NEVER DO LAVAGE**
NEVER DO BLIND SUCTION

✔ **Remember babies who aspirate can also be asphyxiated and therefore lung inflation should not be delayed**.

- If the chest moves the airway is not obstructed.
- If the baby is crying the airway is not obstructed.
- If the chest does not move it is usually a position problem of the airway.

✔ **AIRWAY POSITION NOT SUCTION**

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1.23 Anaphylactic shock

Martin Samuels

Minimum standards requirements

- ABC of resuscitation
- Epinephrine
- Oxygen
- Salbutamol nebulisers
- Hydrocortisone
- Aminophylline

Anaphylaxis is an immunologically mediated reaction to ingested, inhaled or topical substances, which may progress to life-threatening shock and or respiratory distress. Common causes include allergy to penicillin, to ***radiographic contrast media***, and to certain foods, especially nuts.

Consider the diagnosis with any of the following symptoms when there is a history of previous severe reaction, rapidly progressive or increasingly severe symptoms, a history of asthma, eczema or rhinitis (atopy), or in the presence of treatment with beta-blockers.

This situation is potentially life-threatening and may result in change in conscious level, collapse, respiratory or cardiac arrest.

Management

Remove allergen if possible.

- Assess **A**irway:
  - If no problem, assess **B**reathing
  - If stridor there is obstruction (usually at larynx):
    10 micrograms/kg epinephrine IM, then
    5 ml epinephrine 1 in 1000 nebulised
    Give 100% oxygen
    Consider intubation. Call for anaesthetic and ENT assistance
  - If stridor with complete obstruction: intubate or create a surgical airway (see Chapter 6.14).
- Assess **B**reathing:
  - If no problem, assess **C**irculation
  - If no breathing, five rescue breaths with 100% oxygen and assess circulation
  - If wheeze, 10 micrograms/kg epinephrine IM and salbutamol inhaled dose (either 2.5 mg nebulised with 100 % oxygen or 1000 micrograms (5 puffs) via spacer and repeating or continuously as required).
- Assess **C**irculation:
  - If no problem, observe
  - If no pulse, start basic life support, assess rhythm and treat
  - If shocked, 10 micrograms/kg epinephrine IM and 20 ml/kg 0.9% saline/colloid (4.5% albumin).

Table 1.23.1 Features of anaphylaxis

	Symptoms	Signs
Mild	Burning sensation in mouth Itching of lips, mouth, throat Feeling of warmth Nausea Abdominal pain	Urticarial rash Angio-oedema Conjunctivitis Red throat
Moderate	Coughing/wheezing Loose bowel motions Sweating Irritability	Bronchospasm Tachycardia Pallor
Severe	Difficulty breathing Faintness or collapse Vomiting Uncontrolled defaecation	Severe bronchospasm Laryngeal oedema Shock Respiratory arrest Cardiac arrest

From Advanced Life Support Group. *Advanced Paediatric Life Support,* 3rd ed. London: BMJ Books, 2001.

- Re-assess **ABC and continue 100% oxygen:**
  - If there is airway deterioration, repeat IM epinephrine 10 micrograms/kg with or without intubation or airway surgery
  - If still wheezy, repeat IM epinephrine 10 micrograms/kg and hydrocortisone 4 mg/kg IV (over 15 minutes). Consider aminophylline 5 mg/kg slow IV injection over 20–30 minutes followed by a 1 mg/kg/hour IV infusion or salbutamol 4–6 micrograms/kg IV slow injection followed by an IV infusion of 0.5–2.0 micrograms/kg/minute
  - If still shocked repeat IM epinephrine 10 micrograms/kg and 20 ml/kg 0.9% saline/colloid (4.5% albumin) and add an inotropic infusion (dopamine or epinephrine (see Chapter 1.25)
  - If no problem, then observe
  - If no symptoms, other than rash or itching
    give oral antihistamine (chlorpheniramine 250 micrograms/kg)
    give oral steroids (0.5–1 mg/kg oral prednisolone).

Epinephrine

Epinephrine is given intramuscularly unless intractable shock or cardiac arrest when it should be given intravenously or by the intraosseous route. If intubated it can be given at 10 times the dose down the endotracheal tube, that is epinephrine 100 micrograms/kg.

If repeated IM injections of epinephrine are not effective or lasting only a short time start giving it IV. For treating children in severe shock:

- Place 1 mg (1 ml of 1 in 1000 epinephrine) in 50 ml of 0.9% saline
- Then give 2–5 ml (40–100 micrograms) in a child (depending on size) and 1 ml (20 micrograms) in an infant <1 year. Give IV slowly using a peripheral (ideally central) vein, ideally with EGG monitoring
- Repeat as required.
- An infusion of epinephrine at 0.05–2.0 micrograms/kg/min may be needed (preferably via a central vein).

Severe anaphylactic shock

Intubation and ventilation are required for severe cases.

1.24 Self-instructional educational programmes in neonatal resuscitation and perinatal care

James I Hagadorn, Nicholas Guerina and Ismeta Kalkan

Neonatal resuscitation program (NRP)

✔ **Between 5 and 10% of newborn infants may require resuscitative efforts in the delivery room, primarily in response to respiratory depression.** The majority of these infants may be successfully managed provided that basic cardiopulmonary resuscitation is administered with consideration given to the unique transitional physiology of the newborn and the environmental stresses present at delivery. The Neonatal Resuscitation Program (NRP) was launched in 1988 with the basic premise being that at least one of the persons in attendance at any newborn delivery should be trained in newborn resuscitation. The American Academy of Pediatrics currently recommends renewing the course every two years.

Table 1.24.1 Lesson topics from the self-instructional *Neonatal Resuscitation Textbook*

Lesson 1:	Introduction to the Program
Lesson 2:	Initial Steps in Resuscitation
Lesson 3A:	Use of a Resuscitation Bag and Mask
Lesson 3B:	Use of a Resuscitation Bag and Mask: Ventilating the Infant
Lesson 4:	Chest Compressions
Lesson 5:	Endotracheal Intubation
Lesson 6:	Medications

Course description

Basic aspects of physiology are reviewed, but each lesson focuses primarily on practice rather than theory. The first four lessons provide the minimum skills required for successful newborn resuscitation in any circumstance. Successful completion of the course depends upon passing a written test and competent demonstration of practical skills from each lesson.

Even if individuals have training and experience in neonatal emergencies, the course may be beneficial in that it provides a standardised approach to resuscitation. The course is easily adapted for different personnel operating in different medical settings. A course limited to the first four lessons would be appropriate for midwives and general practitioners attending deliveries in small village clinics or homes, whereas all six lessons would be appropriate for maternity hospitals. At maternity hospitals, all personnel, including obstetricians, midwives, pediatricians, and nurses, should complete the NRP course since any of these individuals may find themselves in the position of being the first person available to initiate the resuscitation of a newborn.

Healthcare workers interested in establishing an NRP course may obtain information from: The American Academy of Pediatrics, 141 Northwest Point Boulevard, Elk Grove Village, IL 60007-1098 USA [AAP General Information at (001) 847/434-4000 or www.aap.org/; NRP information at (001) 888/227-1770 or www.lifesupport@aap.org/. International aid workers seeking to introduce the NRP course into a foreign country should also obtain guidelines from the American Academy of Pediatrics.

The perinatal continuing education program (PCEP)

PCEP is a community hospital-based self-instructional programme that reviews a broad array of topics in perinatal and neonatal care over a 40-week period.

Hospital implementation – the PCEP coordinator

A member of the hospital's staff (doctor or nurse) acts as the course coordinator. Course participants typically include physicians, nurses, and ancillary staff involved in obstetric or neonatal care. Because not all hospitals provide comprehensive services for newborn infants, the specific units used in the course may be tailored easily to the hospital's capabilities and personnel training needs. The course encourages use of materials actually in use at the hospital to review bedside care techniques.

Each participant receives a set of instructional texts, and completes the self-instructional materials over a period of approximately 40 weeks under the supervision of their PCEP coordinator. Assessment of learning occurs frequently during this period. For each of the 26 units, the participants complete a pretest, review the unit materials at their own pace, then finish with a post-test to assess cognitive learning. There is also a pretest completed upon starting the programme, and a post-test following completion of all 26 units.

The section of the PCEP text dealing with neonatal resuscitation is based on the NRP.

Regional implementation – the regional coordinator

The PCEP programme is designed for implementation on a regional basis. It assumes that obstetric and newborn services in a region are delivered by a group of relatively small community hospitals or clinics served by a large, full-service referral hospital. The referral centre usually takes the initiative to begin the course at the region's smaller hospitals by training and assisting each hospital's PCEP coordinator. The regional coordinator helps local PCEP coordinators keep their courses on schedule and participates in teaching workshops at selected times during the local courses. Upon completion of the local hospital's course the regional coordinator usually meets with programme participants at the local hospital to exchange feedback.

Table 1.24.2 Topics covered in the perinatal continuing education program

- Fetal Evaluation and Immediate Newborn Care
  - Assessment of fetal well-being
  - Identifying and managing high risk deliveries
  - Newborn assessment
  - Resuscitation of the newborn infant
- High-Risk Newborn Care: General Concepts and Procedures
  - Controlling temperatures of sick and at risk infants
  - Administering and monitoring oxygen for sick and at risk infants
  - Identifying and caring for infants with respiratory distress
  - Recognising and treating pneumothorax
  - Identifying and caring for infants with low blood pressure
  - Identifying and caring for infants with hypoglycaemia
  - Giving intravenous therapy to sick and at risk infants
  - Feeding sick and at risk infants
  - Identifying and caring for infants with hyperbilirubinaemia
  - Identifying and caring for infants with infection
- Complex/Specialised Newborn Care
  - Preparing the baby for transport
  - Providing continuing care for at risk infants
  - Using umbilical catheters with sick and at risk infants
  - Direct blood pressure monitoring
  - Using exchange, reduction and direct transfusions for sick and at risk infants
  - Delivering continuous positive airway pressure
  - Providing assisted ventilation with mechanical ventilators
  - Transcutaneous monitoring
  - Providing surfactant therapy

Available translations of PCEP

PCEP has been implemented successfully in Poland on a large scale. The programme was well received, rates of completion were high, and there were statistically significant improvements between pre- and post-unit test scores for all professional disciplines. It was felt that the success of the programme was enhanced by its self-instructional format, participation of instructors and students from a variety of healthcare disciplines, an organised implementation strategy coordinated by local personnel, and the fact that the programme's content was oriented toward practice rather than theory.

The PCEP programme has since been translated into Spanish in Mexico, where it has been successfully implemented in several hospitals in Yucatan. A South African programme based on PCEP has also been developed and has been used in Zimbabwe, Namibia and Botswana. A partial Chinese translation exists, but few details about its use are available. Recently, the programme has been translated into Bosnian, and will be underway in Bosnia-Herzegovina this year.

Developing self-instructional programmes abroad

Successful implementation of self-instructional programmes abroad depends on the support and advice of local administrators and organisations. Courses should be offered through official health channels, and sponsored by a non-government organisation registered in the selected country. Healthcare providers considering implementation of NRP, PCEP or similar programmes abroad should carefully match the course with local requirements and resources, and encourage local control of the project. Local personnel willing actively to coordinate the program are essential. When programme recommendations conflict with local custom or are impractical due to local physical conditions, the programme should be adapted as necessary, but fundamental content should be preserved to as great an extent as feasible. Special consideration must be given to establishing mechanisms for continuation of the programme and tracking trainees for recertification. The success of an initial implementation of a newly translated programme should be assessed systematically.

Questions regarding PCEP and further details regarding translated editions of the programme may be directed to: Perinatal Continuing Education Program, Department of Pediatrics, University of Virginia Health Sciences Center, Charlottesville, VA 22908 USA, or by contacting the PCEP US national coordinator, Lynn Cook RNC MPH (tel 610-617-7373, lcook@netreach.net).

Further reading

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125 Intensive/high-dependency care of critically ill and injured children

Mark Twite

Definition

✔ Paediatric intensive care is **a service provided for children with potentially recoverable pathological processes who** can benefit from more detailed observation and treatment than is generally available on the standard hospital ward. Intensive care is usually for children with threatened or established organ failure, which may have arisen as a result of:

- An acute illness
- Trauma
- A predictable phase in a planned treatment programme for example after major surgery.

Organisation

The health needs of children are best met through an integrated approach of several agencies including primary and secondary healthcare, education and social services. ✔ **Together such services may help prevent some of the conditions that lead to children requiring intensive care.** For example vaccination programmes will decrease the number of children developing respiratory failure due to preventable diseases such as pertussis and measles. Education and legislation are important in reducing the number of children seriously injured in road traffic accidents and in accidents in the home.

Paediatric intensive care is a low-volume, high-demand specialty. More than half the children requiring intensive care are under two years of age and the majority of these have respiratory failure. There also may be a seasonal variation with a peak in winter months again associated with respiratory related illness.

Dedicated paediatric intensive care units, in large tertiary care centres have been shown to have the best outcomes. ✔ **Ideally every country in the world should have units which provide this service.** However, the majority of children requiring intensive care will present to smaller peripheral hospitals rather than the large tertiary centre. ✔ **It is absolutely essential therefore that paediatric staff in smaller peripheral hospitals are able to recognise and appropriately treat a sick child in the early stages of their illness.** Medical and nursing staff should all be well trained in Paediatric Life Support (see Chapter 1.19) so as to be able to stabilise a sick child, initiate appropriate medical therapy, which may involve intubation and ventilation, until the sick child can be safely transferred to an intensive care unit if still appropriate. Often with good initial resuscitation and early diagnosis and treatment, the need for intensive care can be avoided. In a child requiring intensive care there should be early consultation with the regional/ national paediatric intensive care unit, usually by telephone or radio, so that further management can be jointly decided until a retrieval team arrives to collect the child.

Transportation of sick children, particularly ventilated children, requires appropriately trained staff and equipment. It is best thought of as "an intensive care bed on wheels" and the aim should be that the child does not deteriorate during the transport. Proper resuscitation and stabilisation is essential before moving the child.

Children are not small adults. They exhibit fundamental differences in many ways that influence the training of staff and the type and size of equipment available. The differences extend across anatomy, physiology, pharmacology, and behaviour. Children have less reserve than adults so tend to decompensate early and quickly. However, they also have a greater capacity to make a full recovery. Provision of intensive care is not just about equipment and facilities, the surrounding environment and contact with their families are crucial to promote a child's recovery.

Levels of high-dependency/ intensive care

There are three levels of care that are designed to make the most appropriate use of staff and equipment resource (Figure 1.21.1). During a single stay a child may progress through the different levels. The majority of children can be managed at level one with close monitoring, good nursing care and appropriate medical therapy. By providing optimal therapy it is often possible to prevent the deterioration of the child, for example good fluid management, early but appropriate antibiotics and the use of adequate amounts of oxygen. Hospitals with poor resources may have to attempt Level 2 care but children will suffer greatly if they are ventilated with poorly maintained equipment, with an unreliable oxygen supply and absence of non-invasive or invasive blood gas monitoring. As far as possible in countries with limited resources, intubation and ventilation should be avoided until it is absolutely necessary. Many children can tolerate high P_{CO_2} levels with a

compensated pH – it is hypoxia that damages and kills children. It may be appropriate to develop and have available non-invasive modes of ventilatory support such as nasal mask or prong CPAP, nasal or face mask IPPV (for example BIPAP), or negative-pressure ventilation (CNEP or INPV) (see Chapter 1.26). Similarly, the more invasive a procedure or the monitoring, the more risk there is of complications.

Infusions used in PICU

- **High K intravenous infusions for hypokalaemia**
  Maximum concentration to be given IV is:
  4 mmol/100ml (either in 5% glucose or 0.9% saline).
  This is given at a rate not exceeding 0.5 mmol/kg/hour, ideally with ECG monitoring
  If acidotic, bicarbonate should only be given if serum K > 3 mmol/litre
  Watch serum magnesium levels if possible
  A urine K > 25 mmol/litre confirms renal potassium loss

- **Epinephrine solution for treating children in severe shock**
  Place 1 mg (1 ml of 1 in 1000) in 50 ml of 0.9% saline
  Give 2–5 ml (40–100 micrograms) in a child and 1 ml (20 micrograms) in an infant <1 year
  Give IV slowly, ideally with ECG monitoring
  Repeat as required

Figure 1.25.1 Levels of high-dependency and intensive care.

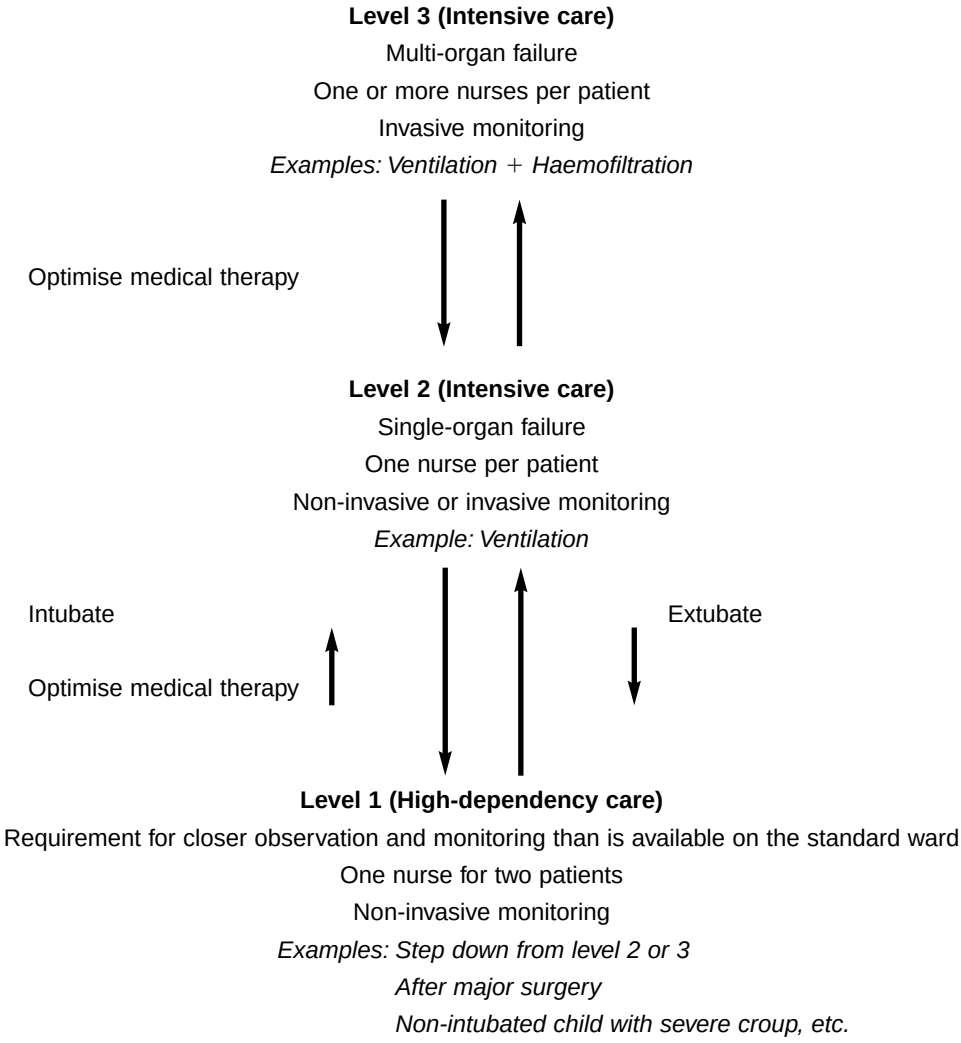

Table 1.25.1 Minimum standards of a lead centre providing paediatric intensive care

Medical staff
- Consultants with appropriate training in paediatric intensive care medicine
- Training programme for junior medical staff specialising in paediatric intensive care
- Provision of 24-hour cover at a senior and junior level
- Resident junior cover for 24 hours by staff with advanced PLS skills, whose only clinical responsibility is to the intensive care unit
- Access on site to other specialist paediatric consultants, for example cardiology, surgery

Nursing staff
- Trained in paediatric intensive care and advanced PLS
- On-going training and support for nursing staff
- Continuous 24-hour observation of each child at all times by a nurse qualified in intensive care with observations documented

Support staff
- Availability of 24-hour physiotherapist
- Available pharmacist and dietician with paediatric experience

Equipment
- Medical staff and nursing staff trained in how to use all equipment
- Equipment maintained on a regular basis and according to manufacturers' guidelines

Retrieval service
- Available 24 hours
- Does not take staff from the unit leaving it uncovered
- Usually an experienced doctor and nurse
- Able to provide phone or radio advice
- Equipped with portable monitors, infusion pumps and suction. Possible to provide hand bag ventilation rather than have a transport ventilator

Clinical effectiveness and management
- Protocols for admissions, discharges, retrievals, resuscitation, stabilisation and for treating major conditions
- Data collection and audit to improve care provided

Facilities for families
- Access of parents at all times
- Accommodation and food for parents
- Child-friendly environment (see Chapter 1.4)

Table 1.25.2 Drug infusions in severely ill or injured children*

Aminophylline: 5% glucose or 0.9% saline
Loading dose (*do not give if theophylline has been received in the last 24 hours*)
IV infusion over 20–30 minutes. 5 mg/kg for <12 years and 250–500 mg total if >12 years
Then 1 mg/kg/hour if <12 years and 500 micrograms/kg/hour if >12 years or <1 year: this is equivalent to
50 mg/kg in 50 ml run at 1 ml/hour for 1–12 years and 0.5 ml/hour for >12 years or <1 year

Atracurium: 0.9% saline (*reversed by atropine 20 micrograms/kg followed by neostigmine 80 micrograms/kg*)
500 micrograms/kg initial loading dose then 200 micrograms/kg supplements as required
or
200–600 micrograms/kg/hour
Maximum concentration 500 micrograms/ml

Diamorphine: 0.9% saline or water; use within 24 hours (*after loading dose of 50 micrograms/kg over 30 minutes*)
Intravenous 10–30 micrograms/kg/hour: this is equivalent to
2 mg/kg in 50 ml run at 0.25–0.75 ml/hour (*lower volume than morphine, useful in neonates and SC*)
Subcutaneous 20–100 micrograms/kg/hour: this is equivalent to
2 mg/kg in 50 ml run at 0.5–2.5 ml/hour

***Dobutamine*: 5% glucose or 0.9% saline. Do not mix with bicarbonate**
2–20 micrograms/kg/minute: this is equivalent to
30 mg/kg in 50 ml run at 0.2–2 ml/hour (maximum concentration of 5 mg/ml)

Table 1.25.2 *Continued*

Dopamine: 5% glucose or 0.9% saline or neat (ideally via a central line). Do not mix with bicarbonate. Can be mixed with dobutamine.

2–20 micrograms/kg/minute (renal = up to 5 micrograms/kg/minute): this is equivalent to
30 mg/kg in 50 ml run at 0.2–2 ml/hour

Epinephrine: 5% glucose or 0.9% saline. Do not mix with bicarbonate.

0.05–2 micrograms/kg/minute: this is equivalent to
0.3 ml/kg of 1 in 1000 (300 micrograms/kg) in 50-ml run at 0.5–20 ml/hour

Fentanyl: 5% glucose or 0.9% saline or neat

1–8 micrograms/kg/hour: this is equivalent to
200 micrograms/kg in 50 ml at 0.25–2 ml/hour
or
Neat (50 micrograms/ml): run at 0.02–0.16 ml/kg/hour. Doses >3 micrograms/kg/hour only needed when to tolerance has developed

Ketamine: 5% glucose or 0.9% saline

10–45 micrograms/kg/minute: this is equivalent to
50 mg/kg in 50 ml run at 0.6–2.7 ml/hour (maximum concentration 50 mg/ml)

Midazolam: 5% glucose or 0.9% saline or neat

1–6 micrograms/kg/minute (60–360 micrograms/kg/hour): this is equivalent to
6 mg/kg in 50 ml run at
0.5–3 ml/hour
or
Neat: (5 mg/ml): run at 0.012–0.072 ml/kg/hour. Doses >3 micrograms/kg/minute only needed when tolerance has developed

Morphine: 5% glucose or 0.9% saline (after loading dose: see Chapter 1.27)

10–60 micrograms/kg/hour: this is equivalent to
1 mg/kg in 50 ml run at 0.5–3 ml/hour

Nitroprusside: 5% glucose only

0.2–8 micrograms/kg/minute: this is equivalent to
3 mg/kg in 50 ml run at 0.2–8 ml/hour
Protect infusion from light. Discard after 24 hours

Pancuronium: 0.9% saline

Loading dose of 100 micrograms/kg and then 50–100 micrograms/kg supplements as required. (Reversed by atropine 20 micrograms/kg followed by neostigmine 80 micrograms/kg)

Propofol*: *Neat (Beware – older children >3 years only). Can be diluted with 5% glucose.
Neat (10 mg/ml): run at 0.2 ml/kg/hour (= 2 mg/kg/hour) increase as required to a maximum of 10 mg/kg/hour

Prostacyclin (Epoprostenol)*: *0.9% saline only. Incompatible with glucose.
5–20 nanograms/kg/minute: this is equivalent to
12 micrograms/kg in 50 ml run at 1.25–5 ml/hour

Prostaglandin: E_2 (Dinoprostone): 5% glucose or 0.9% saline. Use separate IV line.
5–10 fold higher doses of prostaglandin E_2 have been used to re-open the ductus arteriosus but this commonly causes apnoea
5–20 nanograms/kg/minute: this is equivalent to 12 micrograms/kg in 50 ml run at 1.25–5 ml/hour

Salbutamol: 5% glucose or 0.9% saline

0.6–5 micrograms/kg/minute: this is equivalent to
3 mg/kg in 50 ml run at 0.6–5 ml/hour

Thiopental – reconstituted with water to give 25 mg/ml. Can be further diluted with 5% glucose or 0.9% saline

2–8 mg/kg/hour: this is equivalent to
25 mg/ml: run at 0.08–0.32 ml/kg/hour

Vecuronium: 5% glucose or 0.9% saline

1–2 micrograms/kg/minute: this is equivalent to
3 mg/kg in 50 ml run at 1–2 ml/hour (shorter duration of action than pancuronium)

* Start at lowest rate of infusion and build up.

126 Non-invasive respiratory support

Martin Samuels

See Chapter 6.18.

Respiratory support is needed when the patient fails to sustain an adequate airway, oxygenation or ventilation, despite treatment of the condition leading to respiratory failure. Respiratory failure may result from:

- Respiratory illnesses
- Severe shock
- Coma
- Convulsions
- Meningoencephalitis
- Neuromuscular disorders
- Raised intracranial pressure, for example from trauma

Infants and young children are more likely to progress to respiratory failure because:

- They are more susceptible to infection;
- The airways are smaller;
- The thoracic cage is more compliant;
- The ribs are (nearer) horizontal;
- Respiratory muscles are more prone to fatigue

Presenting symptoms of respiratory illness

- Breathlessness
- Cough
- Noisy breathing (stridor, grunting)
- Hoarseness
- Drooling
- Inability to drink
- Abdominal pain

Respiration rate in relation to age is shown in the table below.

Table 1.26.1 Respiration rate

Age (years)	Respiration rate (bpm)
<1	30–40
2–5	25–30
5–12	15–20
>12	15–20

As respiratory failure progresses, it will ultimately lead to cardiorespiratory arrest and death. Thus recognition of the severity of the conditions that lead to respiratory failure, followed by appropriate treatment will reduce morbidity and mortality.

Signs indicating the adequacy of breathing:

- Intercostal, subcostal and suprasternal recession
- Respiratory rate
- Inspiratory and expiratory noises
- Use of accessory muscles
- Adequacy of breath sounds and chest expansion
- Heart rate
- Skin colour
- Mental status

To help assess the development of respiratory failure, it is necessary to assess *changes* in the clinical signs. In the following situations, however, these signs are less useful because there is absent or decreased work of breathing:

- with fatigue or exhaustion, for example after prolonged respiratory effort;
- with cerebral depression from raised intracranial pressure, poisoning or encephalopathy;
- in children with neuromuscular disease

In these cases, pay more attention to the chest expansion, heart rate, skin colour, mental status and if available, SaO_2 measurement.

Pulse oximetry is of additional value to measure the arterial oxygen saturation through the skin (SpO_2 or SaO_2); values of SpO_2 < 92–94% in air at sea level (see Chapter 3.19 for values at altitude) are abnormal and would warrant at least initial treatment with additional inspired oxygen. **It is essential to remember that in respiratory failure normal SpO_2 values whilst receiving additional inspired oxygen are likely to be associated with significant hypoventilation or intrapulmonary shunting.** ***Measurement of transcutaneous, end-expired or blood carbon dioxide levels will confirm this hypoventilation.***

When respiratory fatigue is severe, oxygenation is poor or deteriorating, or carbon dioxide levels are rising, respiratory support should be used, if available.

Table 1.26.2 Modes of respiratory support showing the patient interfaces, nursing care and medical treatment needed and the conditions for which the support is used

Mode of support	Interface with patient	Level of nursing care	Associated medical treatment	Clinical use	Examples of conditions treated
Continuous positive airways pressure (CPAP)	Nasal prongs or nasopharyngeal tube	High-dependency	Sedation or analgesia may be needed	To keep upper and lower airways patent and maintain adequate lung volume (oxygenation)	Neonatal respiratory distress syndrome, *bronchiolitis**
	Nasal mask or face mask	Home, ward, high-dependency	Nil		Sleep-related upper airway obstruction
		Intensive care	Sedation or analgesia may be needed		Acute upper airway obstruction before, *instead of** or after extubation
	Nasal mask or pillows, face mask (NIPPV)	Home to intensive care	Nil	To treat hypoventilation (raised CO_2) when airway control and clearance are adequate	Chronic, for example central neuromuscular Acute, for example after surgery
Intermittent positive pressure ventilation (IPPV)	Endotracheal tube	Intensive care	Anaesthesia for intubation Sedation or analgesia will be needed	To treat hypoventilation when clearance/support of airway(s), or close control of ventilation needed	Procedures or surgery needing anaesthesia Severe respiratory illnesses, raised intracranial pressure
	Tracheostomy	Home to intensive care	ENT surgical procedure	Long-term ventilation where day and night support needed	Brainstem/high spinal injury or neuromuscular disease
Continuous negative extrathoracic pressure (CNEP) Intermittent negative pressure ventilation (INEP or INPV)	Chamber or jacket	Home to intensive care	Nil	To keep lower airways patent and maintain adequate lung volume To treat hypoventilation where airway control and clearance are adequate or kept adequate by CPAP	Bronchiolitis and other severe lower respiratory infections Central hypoventilation, for example apnoea of prematurity or neuromuscular disease

Shaded areas are those that require a lower dependency of care, for example have been used in the home setting, but may be useful in acute conditions.
* High-risk situation: CPAP may be ineffective and intubation may be required.

The use of positive pressure ventilation with particular reference to non-invasive support

- Monitoring of patient status and either airway or extrathoracic pressures are necessary when undertaking any form of respiratory support (see below).
- Positive airway pressure involves a flow of air or other gas mixture to the patient's airways. This flow may be continuous (as in CPAP) or intermittent (as used in IPPV). It may vary with inspiration and expiration (BiPAP), or to accommodate for the leaks or variable compliance of ventilator tubing, airways or lung units.
- Mask ventilation can be well tolerated by children, but it may be more difficult for infants and young children to tolerate appliances on their face.
- In the presence of excess airway secretions or an open mouth, nasal masks and nasal prongs may not produce as effective airway pressures as ventilation with tracheal intubation (or relatively higher pressures may be needed for the same effect).
- The pressures used with masks and prongs may be higher than that used with tracheal intubation because of the greater potential for air leaks and other volume loss in compliant upper airway structures.
- Infants and young children will sometimes tolerate masks and prongs only with the use of sedation, in which case close monitoring of respiratory failure must be undertaken in case full intubation and ventilation is needed.
- Endotracheal intubation should be undertaken with rapid sequence drug or gaseous induction, and subsequent analgesia, anxiolysis and sedation be provided.
- Positive pressure ventilation administered through an endotracheal tube must be accompanied by adequate humidity of the inspired gases.
- Oxygen may be administered either using a built-in mixer in the ventilator, or by entraining a supply in the ventilator tubing nearer to the patient.
- Positive pressure ventilators should be able to provide manipulation of either the pressure or volume administered, and the time intervals for inspiration and expiration. There should be alarms for failure to cycle, and excessive pressure/volume administered.

127 Pain control in children

David Southall

Introduction

- ✔ **It is ethically wrong and a failure of professional duties for a child to suffer uncontrolled pain**.
- Uncontrolled pain has adverse cardiovascular, respiratory, immunological and metabolic consequences as well as long-term psychological effects.
- Both pharmacological and non-pharmacological approaches are valuable, especially in chronic pain.
- ✔ **Attempts should be made to anticipate and prevent pain rather than trying to relieve it when it is established**. "As required" regimens should be avoided. Analgesics should be used in regular and adequate doses.
- There is little place for intramuscular pain relief, particularly as a repeated treatment. Many children would rather suffer and hide their pain than receive intramuscular analgesia.
- If a **conscious** child has to be restrained for a procedure, it must be done kindly but firmly by a person or persons (ideally a parent) and not by contraptions such as strait-jackets or the tying down of limbs.
- It is important to ask for and value the child's judgement concerning the adequacy of pain relief provided.
- When beginning a course of treatment for pain it is important to realise that it may be for a long time.
- ✔ **Pain control must therefore be of the highest quality from the onset**, with an emphasis on preventative measures.

Assessment of pain in children

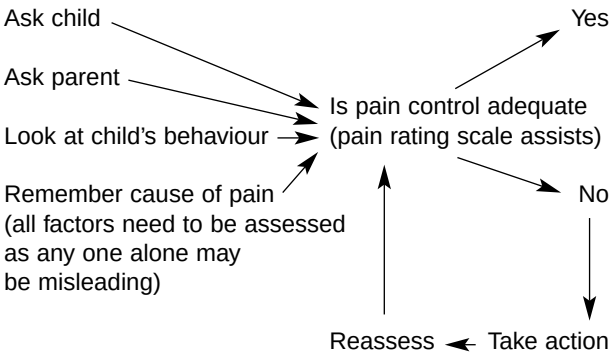

Figure 1.27.1 Pain assessment

Problems with assessment

- Hidden suffering by child.
- Differentiating anxiety from pain.
- Recognition that parents (and professionals) may underestimate or overestimate pain.
- Recognise that preverbal and non-verbal children (for example, those with learning difficulties or with sensory handicap) may not be able adequately to express their need for pain control.

Methods for assessing pain

- By what is to be expected.
- By discussion.
- By using a self-report scale.
- By observing, ideally in collaboration with parents, the child's behaviour.
- By observing physiological measures in the presence or absence of behavioural changes.
- By using a diary.

Methods for reducing pain without drugs (however drugs are mandatory in severe pain of organic origin)

Environmental

- Negative aspects of the environment should be minimised or removed. These include an overly "clinical" appearance and evidence of invasive instrumentation. Needles should be kept out of sight (see Chapter 1.4).
- An attractive, decorated environment with toys, mobiles and pictures.
- Privacy is desirable.

Supportive and distractive techniques

✔ **Parents should be present with their child during invasive procedures, unless there are very good medical reasons why they should be excluded**.

Age-appropriate distraction strategies include:

- Presence of familiar objects (comforters) for example, pillow, soft cuddly toy
- Singing, concentrating on nice things, jokes, games and puzzles

- Moving images on a nearby wall, for example, fish swimming, birds flying
- Imaginary journeys
- Blowing out air or bubbles
- Reading pop-up books
- Kaleidoscope
- 3D viewers
- Breathing out, but not hyperventilation which may increase anxiety
- Mirror to show view through a nearby window
- TV, video, interactive computer games
- Listening through headphones to stories or music

Relaxation exercises, for example tensing muscles in sequence and then relaxing them.

Guidelines for analgesia in children

Procedure-related pain control

✔ - **Procedures are often painful, undignified or both. Ideally they should be undertaken in a treatment room so that other children are not frightened and so that their bed-space is a sanctuary where such events will not happen**.
- They often have to be repeated. Therefore provide optimal treatment on the first occasion in order to minimise a dread of future procedures.
- The child's fear is often the major emotion to address.
- Pharmacological and non-pharmacological methods should be used to control the pain and help the child.
- For major procedures requiring powerful analgesia/sedation, an anaesthetist should be present in addition to the paediatrician/nurse/surgeon undertaking the procedure. These include chest drain insertion, repeated lumbar puncture, bone marrow aspiration, and central venous cannulation. ***Such procedures may be best undertaken under general anaesthesia if this can be given safely*** (often not the case in disadvantaged countries).
- For venous cannulation, size appropriate catheters must be available. It is not for example appropriate to have to use an 18 or 20 gauge cannula in a neonate. Although local anaesthetic creams such as Emla represent best practice, they are expensive. Ice or an alcohol spray may relieve some of the pain providing that they do not make veins less visible.

Pain induced by operations

Preoperative management
This should include patient and family assessment, including history of previous painful experiences and child's response.
- What sort of painful things have happened to your child in the past?
- How does your child usually react to sudden pain? To chronic pain?
- Does your child tell you (or others) if he/she is in pain?
- What does your child do to get relief from pain?
- Which actions appear to work the best?

Pain management during surgery
- Opiates/NSAIDs can reduce postoperative pain
- Wound infiltration with a local anaesthetic such as bupivacaine
- Use local or regional anaesthetic as part of overall strategy (see Chapter 1.13)

Prophylactic antiemetics for children of four years and older when opioids are part of the pain-control plan.

Postoperative pain management
- Provide analgesia before the pain becomes established.
- Use safe and effective doses of opioids along with other analgesics to reduce the amount of opioid required.
- Avoid intramuscular injections.
- Assess – check response – reassess.
- Most at risk of poor pain control are children with limited/absent verbal ability.
- If pain seems out of proportion to surgical trauma consider complication and reassessment by surgeons.
- If asleep, assume pain is acceptable – do not wake up to make assessment but check regularly to ensure still asleep. If awake and lying quietly, do not assume comfortable without enquiring.

Special issues regarding pain in the newborn infant

- **Most studies, some controlled, have shown that neonates (premature and full term) react to pain**. ✔
- Infants can easily be forced to put up with suffering.
- Small doses should be measured and given with an oral syringe.
- Adequate general anaesthesia, using opioids when needed, should be given for all surgical procedures on neonates.
- Local anaesthetics must be used when they would be used in an older child undergoing the same procedure.

Pain control during procedures in neonates

- A sugar dummy coated with 2 ml of 25–50% sucrose two minutes before the procedure can be helpful.
- Breastfeeding during procedures may be equally as valuable.
- In all cases comfort and containment (swaddling) should be provided by a parent or a nurse.

Pain management in intensive care

- Where possible all invasive procedures should be elective. Every effort should be made to avoid unexpected emergency procedures, such as intubation, by

adequate monitoring and provision of airway and blood oxygenation, adequate lung volume, and airway care.

✔ ● **Emergency procedures are frequently extremely painful, dangerous to the child and represent a failure of intensive care**.

✔ ● **Muscle relaxants should never be used unless the child is pain free and sedated**.

● Aim to provide a child- and family-friendly environment.
  - Provide a day/night cycle (uninterrupted natural sleep can lessen the need for analgesia/sedation).
  - Ensure minimal noise/low lighting from 8 pm to 8 am.
  - Emergency admissions at night should occur away from sleeping patients.
  - Monitors should be set to alarm audibly only when essential.
  - Consider ear plugs – especially when the child is paralysed.
  - Provide human input through voice, touch, music, cuddling, rocking, holding, pacifying.
  - Consider distraction, play therapy, relaxation, behavioural techniques, hypnosis, massage, and aromatherapy – particularly in patients undergoing long-term intensive/high-dependency care.
  - Provide privacy when possible.
  - Watch for depression after prolonged intensive care. Consider seeking help from a child psychiatrist, child psychologist or child psychiatric nurse.
  - Consider methadone and clonidine for the control of opioid withdrawal after prolonged treatment.

Use of local anaesthesia

Indications

Consider for any painful procedure.

Pharmacology

Lidocaine: normal concentration 0.5–2%. Rapid onset (minutes)/short duration (1–2 hours). Maximum dose 3 mg/kg (plain).

● Local anaesthetics come as water-soluble salts. They dissociate in the tissues liberating the active base. Inflamed tissue is slightly acidic and dissocation is inhibited thereby making the anaesthesia unreliable – hence local infiltration into an abscess is not recommended.

Table 1.27.1 Oral analgesia for mild or moderate pain

Drug	Preparation	Comments
Paracetamol **Oral** loading dose 25 mg/kg • Maintenance dose 24 mg/kg 6 hourly • Maximum dose 100 mg/kg/24 hour (60 mg/kg/24 hours < 3 months)	Oral suspension: 120 mg/5 ml 250 mg/5 ml Tablets/soluble 500 mg	The maximum daily dose should not be given for > 3 days Consider measuring drug levels (if available)
Rectal loading dose 40 mg/kg • Maintenance dose 20 mg/kg 6 hourly (15 mg/kg if < 3 months; 8 hourly if > 36 weeks and 12 hourly if < 36 weeks gestation) • Maximum dose 90 mg/kg/ 24 hours (60 mg/kg/ 24 hours if < 3months)	Suppositories: 60, 125, 250, 500 mg and 1 g or oral suspension can be given rectally	Caution with liver impairment No anti-inflammatory effects Avoid repeated rectal administration Can combine with NSAIDs
Ibuprofen (NSAID) 4–10 mg/kg 6–8 hourly	Oral suspension: 100 mg/5 ml Tablets: 200 mg and 400 mg	Do not use if less than 1 year old **Caution in asthmatics and patients with renal impairment**
Diclofenac (NSAID) *Oral or rectal* 500 micrograms to 1 mg/kg 8–12 hourly Maximum dose 3 mg/kg/24 hours	Tablets: 25 mg and 50 mg Suppositories: 12.5 mg, 25 mg, 50 mg, 100 mg	**Contraindications:** **SHOCK** **Bleeding disorders and hypersensitivity to aspirin**
Dihydrocodeine 500 micrograms/kg/dose 4–6 hourly **Codeine phosphate** 500 micrograms to 1 mg/kg/dose 4–6 hourly	Tablets: 30 mg Elixir: 10 mg/5 ml Tablets: 15 mg, 30 mg Elixir: 25 mg/5 ml	**Caution with liver impairment and head injuries** May lead to constipation. Consider prophylactic lactulose CODEINE MUST NOT BE GIVEN IV AS IT CAN REDUCE CARDIAC OUTPUT THROUGH HISTAMINE RELEASE

* NSAIDs/dyhydrocodeine or codeine/paracetamol can be used in combination.

Table 1.27.2 Oral analgesia for severe pain in infants and children

Drug	Preparation	Comments
Oramorph ● 1–3 months 100 micrograms/kg every 4 hours Maximum of five doses in 24 hours ● 3 months–12 years 200–500 micrograms/kg/dose every 4 hours ● Over 12 years 10–15 mg every 4 hours Single dose prior to painful procedure may be useful For long-term severe pain, give as the total daily dose in two divided doses (usually 200–500 micrograms/kg every 12 hours) (see Chapter 1.28)	Mixture: 10 mg/5 ml, 30 mg/5 ml, 100 mg/5 ml Tablets: 10 mg, 20 mg, 50 mg Suppositories 15 mg, 30 mg Slow-release tablets: 5 mg, 10 mg, 30 mg, 60 mg, 100 mg Slow-release suspension: Sachets 20 mg, 30 mg, 60 mg, 100 mg, 200 mg	**Side effects:** ● Respiratory depression. IF RESPIRATORY RATE IS **Age** / **Rate** $<$6 months / $<$20 breaths/min $>$6 months–2 years / $<$16 breaths/min 2–10 years / $<$14 breaths/min 10–16 years / $<$12 breaths/min ALERT MEDICAL STAFF AND ENSURE NALOXONE IS AVAILABLE Monitor SaO_2 as appropriate (should not be $<$94% in air at sea level) ● Constipation therefore use prophylactic docusate sodium or other laxatives ● CAUTION with head injuries/liver/renal impairment

Response of individual patients is very variable and it is essential to individualise treatment.

Table 1.27.3 Parenteral analgesia for severe pain Subcutaneous intermittent morphine in infants and children

Technique	Dose	Monitoring
• 22/24 gauge subcutaneous cannula • Suitable sites: uppermost arm, abdominal skin. • Give dose slowly over 5 minutes Flush with 0.3 ml 0.9% saline • Can be sited at the time of surgery	100–200 micrograms/kg $\times$ 4–6 hourly Maximum 6 $\times$ 24 hours.	Side effects: ● Respiratory depression. IF RESPIRATORY RATE IS **Age** / **Rate** $<$6 months / $<$20 breaths/min $>$6 months–2 years / $<$16 breaths/min 2–10 years / $<$14 breaths/min 10–16 years / $<$12 breaths/min ALERT MEDICAL STAFF AND ENSURE NALOXONE IS AVAILABLE Monitor SaO_2 with pulse oximeter as appropriate (SaO_2 should not be $<$94% in air at sea level (see Chapter 3.19) ● Constipation therefore use prophylactic docusate sodium or other laxative ● CAUTION with head injuries/liver/renal impairment

Response of individual patients is very variable and it is essential to individualise treatment.

Table 1.27.3 *Continued*

Technique	Dose	Monitoring
		● Naloxone to reverse respiratory depression at 10 micrograms/kg should be immediately available (neonatal ampoule 40 micrograms/2 ml *or* adult ampoule 400 micrograms/ml. Give IV (SC or IM if not possible). Repeat after 2–3 minutes if no response when second dose may need to be much higher (up to 100 micrograms/kg). An IV infusion may be needed if protracted depression of respiration occurs.

Table 1.27.4 Intermittent intravenous bolus morphine in infants and children*

Age	Loading dose	Subsequent doses (Give slowly over 10 minutes)	Monitoring
● 1–3 months	100 micrograms/kg over 30 minutes	25 micrograms/kg/dose × 6 hourly	Side effects: ● Respiratory depression. IF RESPIRATORY RATE IS **Age** / **Rate** < 6 months / < 20 breaths/min > 6 months–2 years / < 16 breaths/min 2–10 years / < 14 breaths/min 10–16 years / < 12 breaths/min ALERT MEDICAL STAFF AND ENSURE NALOXONE IS AVAILABLE Monitor SaO_2 with pulse oximeter as appropriate (SaO_2 should not be < 94% in air at sea level) ● Constipation therefore use prophylactic docusate sodium or other laxatives ● CAUTION with head injuries/liver/renal impairment. ● Naloxone to reverse respiratory depression at 10 micrograms/kg should be immediately available (neonatal ampoule 40 micrograms/2 ml *or* adult ampoule 400 micrograms/ml). Give IV (SC or IM if not possible). Repeat after 2–3 minutes if no response when second dose may need to be much higher (up to 100 micrograms/kg). An IV infusion may be needed if protracted depression of respiration occurs.
● 3–12 months	100 micrograms/kg over 30 minutes	50 micrograms/kg/dose × 6 hourly	
● 1–12 years	100–200 micrograms/kg over 5–20 minutes	100–200 micrograms/kg/dose × 4 hourly	
● > 12 years	2.5–10 mg over 5–20 minutes	2.5–10 mg × 4 hourly	

* Experienced staff only. Response of individual patients is very variable and it is essential to individualise treatment.

Table 1.27.5 Intravenous infusion of morphine* in infants and children

Loading dose	Technique	Continuous infusion	Monitoring
● 1–12 months: 100 micrograms/kg over 30 minutes ● Over 1 year 100–200 micrograms/kg over 5–20 minutes	Use dedicated cannula Requires one-to-one nursing	10–40 micrograms/kg/hour For most situations start at 10 micrograms/kg/hour and increase in 5 micrograms/kg/hour units Major surgery: start at 20 micrograms/kg/hour adjust according to pain control	Side effects: ● Respiratory depression. IF RESPIRATORY RATE IS Age / Rate: <6 months: <20 breaths/min >6 months–2 years: <16 breaths/min 2–10 years: <14 breaths/min 10–16 years: <12 breaths/min ALERT MEDICAL STAFF AND ENSURE NALOXONE IS AVAILABLE ● Monitor SaO_2 with pulse oximeter as appropriate (SaO_2 should not be <94% in air at sea level) ● Constipation therefore use prophylactic docusate sodium or other laxative ● CAUTION with head injuries/liver/renal impairment. ● Naloxone to reverse respiratory depression at 10 micrograms/kg should be immediately available (neonatal ampoule 40 micrograms/2 ml *or* adult ampoule 400 micrograms/ml). Give IV (SC or IM if not possible). Repeat after 2–3 minutes if no response when second dose may need to be much higher (up to 100 micrograms/kg). An IV infusion may be needed if protracted depression of respiration occurs. ● Monitor syringe movement. ● Monitor signs of inflammation at site of infusion. ● Monitor for urinary retention.

* Morphine dose 1 mg/kg made up to 50 ml 0.9% saline or 5% glucose.
Then 1 ml/hour = 20 micrograms/kg/hour, 2 ml/hour = 40 micrograms/kg/hour, 3 ml/hour = 60 micrograms/kg/hour, etc.
Response of individual patients is very variable and it is essential to individualise treatment.

Table 1.27.6 Prevention of nausea and vomiting due to morphine using prochlorperazine (Stemetil)

Age	Oral dose (8–12 hourly)	Rectal dose (8–12 hourly)	IM dose (8–12 hourly)
<1 year	100–250 micrograms/kg		
1–4 years (>10 kg)	1.25–2.5 mg	2.5 mg	1.25–2.5 mg
5–12 years	2.5–5 mg	5–10 mg	5–6.25 mg
Over 12 years	5–10 mg	12.5–25 mg	12.5 mg

* Tablets 5 mg. Liquid 5 mg in 5 ml. Suppositories 5 mg and 25 mg.
Injection 12.5 mg in 1 ml.

- Duration of action prolonged using epinephrine (as vasoconstrictor) at a concentration of 1 in 200 000.
- ✔ **Vasoconstrictors must not be used in tissues with end arteries, for example finger, toes, penis, where tissue necrosis may occur.** Lidocaine maximum dose 7 mg/kg when combined with epinephrine.

Toxicity

- Related to dose.
- **If accidentally administered intravenously.** ✔ Therefore draw back before infusing and ensure needle is not in a vein.

- Can be absorbed through mucous membranes in sufficient concentrations to be toxic.
- Systemic effects
  Neurological: nausea, restless, convulsions
  Cardiovascular: bradycardia, hypotension

Guidelines for the sedation of children

General comments

Sedation and anaesthesia are a spectrum. If you give enough "sedation" you can induce anaesthesia, i.e. **loss of consciousness and the inability to feel pain**. The fine distinction lies in the ability of the patient to maintain vital functions without assistance, and to respond to being roused.

Minimum information required to prescribe sedation

✔ **Anyone giving intravenous sedation could inadvertently produce anaesthesia and must be able to deal with the following possible consequences:**

- **Support respiration**
- **Manage the upper airway**
- **Use suction appropriately**
- **Intubate if necessary**

One-to-one nursing is required.

A combination of drugs may give better effects with fewer side effects than continually repeating doses of the same drug, for example morphine combined with benzodiazepine. Each should be given separately and doses adjusted.

- Some children are difficult to sedate for predictable reasons, for example treatment for epilepsy may make dose required much higher than normal. Some children just refuse.
- Some children respond in a paradoxical manner to sedation and become unmanageable and agitated, for example children with "behavioural disorders" and some children with cerebral palsy.
- Some children are very resistant to sedation, possibly due to excess anxiety, but are otherwise normal. The first attempt at sedation may not succeed and an increased dose may be tried next time.
- Children needing heavy sedation should have oral intake restricted as for anaesthesia.
- Some children are more vulnerable to the effects of sedation, particularly those with problems within respiration or the upper airway.

The minimum information required to prescribe sedation includes:

- Child's age
- Child's weight
- Procedure for which sedation is required
- Previous sedation history
- Other drugs taken
- **Other major illnesses affecting respiratory function and upper airway competence** ✔
- Current status/coughs/colds/pyrexia
- Oral intake status

Table 1.27.7 Differences between sedation and anaesthesia

Vital function	Sedation	Anaesthesia
Response to being roused	Present	Absent (usually)
Respiration	Rate and depth may be slightly reduced	Rate and depth markedly reduced or absent
Swallowing reflex	Present	Absent (usually)
Gag reflex	Present	Absent
Cough reflex	Laryngeal spasm unlikely, but possible with sufficient provocation	May be present at lighter levels but unlikely Hypersensitivity and laryngeal spasm may occur
Cardiovascular stability	Mild hypotension may occur	Severe hypotension may occur

Loss of any of the above reflexes is routine in anaesthetic environments but should *not* occur when providing sedation.

Table 1.27.8 Patients at risk from the effects of sedation or analgesia

Risk factor	Underlying cause
Impaired upper airway Obstruction	Croup Foreign body Congenital stridor. For example Pierre-Robin syndrome or cleft palate Baby with very blocked nose
Impaired reflexes	Pre-existing neuromuscular problems Swallowing difficulties Known bulbar problems, especially if combined with reflux.
Impaired central respiratory drive	Head injury Drug effects (opiates) Raised intracranial pressure (RICP) Impaired level of consciousness Encephalopathy (hypoxic, metabolic, infective)
Impaired respiratory muscle function	Neuropathy and myopathy Chronic illness and weakness Malnutrition Prematurity Infancy
Impaired lung function	Chest infection Pleural effusions Chronic lung disease
Impaired cardiovascular function	Haemorrhage Sepsis Drugs

Table 1.27.9 Sedative drugs

Drug	Route	Onset	Duration	Dose (Single)
Chloral hydrate Liquid 100 mg/ml Suppositories 100 mg and 500 mg	Oral or rectal	30 minutes–1hour	1–2 hours	30 mg/kg for night sedation 50–70 mg/kg for procedures 70–100 mg/kg for scans Maximum dose: 1 g
Comments: Chloral hydrate is better for younger children $<$18 months. Do not give $>$75 mg/kg to a child $<$6 months or $<$15 kg but may paradoxically worsen agitation (for example in Down's syndrome)				
Midazolam	Slow IV injection (over 5 minutes)	Immediate	30 minutes–2 hours	100–200 micrograms/kg/hour
Injection 10 mg in 5 ml	IV infusion	Immediate		30–300 micrograms/kg/hour
Injection 10 mg in 2 ml	Intra-nasal or sublingual oral	10–30 minutes	1–2 hours	200 micrograms/kg (max. 10 mg)
		20–30 minutes	1–2 hours	500 micrograms/kg (max. 10 mg)
Comments: Benzodiazepines are more suitable for children older than 18 months				
Ketamine – see chapter on paediatric, anaesthesia, 1.13 (only to be used by staff experienced in airway maintenance and full resuscitation)				

128 Palliative care for the dying child

Janet Vickers

Although palliative care actually means relief of symptoms in all care, the term is usually associated with the relief of symptoms when the emphasis is no longer on curative treatment. The decision to stop or withdraw curative treatment will never be easy for parents or professionals and may evolve over a period of time. It is important however to state that even when we cannot cure the body, it is never true that nothing more can be done.

Like all of us, children have personal needs, and careful attention must be given to the physical, social, emotional and spiritual needs of the child and their family. Staff too should have support through what can be a distressing time.

Choice for the family and child in the setting for palliative care is of great importance. In the presence of effective care and support networks, **home has been frequently demonstrated to be the best place for palliative care for both the child and the family.** However, it is recognised that the necessary resources may be minimal or absent in many locations and conditions locally will determine what options are available.

Helping a dying child

- Include parents in care
  - This matters at all times.
  - Their familiar presence will comfort.
  - Even unconscious children may still know their voices.
  - Parents invariably want to be able to provide care for their child. It is a natural wish and can aid their own coping strategies.
- Set realistic goals
  - The art of terminal care is to know when both goal and treatment must change.
  - The goal is to help the child enjoy what is left of life.
  - Resuscitation is not usually appropriate in a terminally ill child where curative therapy no longer has a place. A non-resuscitation policy should be discussed and agreed as the way forward with all involved.
  - Our aim is not now to cure, and never to kill, but always to comfort and relieve suffering.
  - The social needs and goals of a dying child include access by siblings and friends to play with and talk to. They should be made welcome.
- Listen and explain
  - It should be clear from the child's deteriorating condition that the goals are changing and death is on the way. This must be gently explained and the parents' (and the child's) questions answered. It is wise, especially with children, to clarify the real question being asked. Replies should be honest, but the truth shared sensitively little by little.
  - Explanations are very important for both parents and children and appropriate, understandable terms should be used.
  - Forewarning of procedures, with hugs and praise afterwards, will reduce fears and fantasies.
  - Honesty earns better trust and cooperation than saying something will not hurt when it will.
  - All involved, from a young child to an elderly grandparent, will harbour fears and anxieties. Active listening is a major part of caring for a dying child and his/her family. Great comfort can be gained from the acknowledgement and expression of anxiety and the frequent sense of isolation dissipated.
  - Adolescents will also have particular concerns, worries and maybe spiritual needs.
- List and treat symptoms
  - In palliative care symptom intervention and practical care is paramount.
  - Even with limited resources symptoms can often be helped.
  - The availability of drugs does not guarantee their skilful use but when used effectively medication will make life and death more bearable.
  - Child and carer together should list all symptoms to guide palliation, even when the cause is incurable.

The duration and nature of palliative care will be individual to each child and their particular disease. **For those children who cannot be cured, high-quality symptom control is paramount to enable a good quality of life for the time that is remaining.**

It is essential to approach the management of any symptom systematically.

Principles of pain control

(See Chapter 1.27)
Pain is probably the most common symptom in palliative care and is frequently seen in both malignant and non-malignant disease. It is a complex sensation related to the physiological insult to the tissues, but is also influenced by psychological, social and cultural factors.

It is helpful to think of severe pain in terms of response to opioids:

- Opioid-responsive, i.e. pain relieved by opioids.
- Opioid-semiresponsive, i.e. pain relieved by the concurrent use of an opioid and an adjuvant drug.
- Opioid-resistant i.e. pain not relieved by opioids.

Neuropathic or nerve pain is more likely to fall into the semiresponsive or unresponsive groups.

Analgesic approaches to pain relief

The optimal approach to pain management in children includes drug therapy with analgesics usually the mainstay of treatment. Correct use of analgesic drugs will relieve pain in most children and should be based on the four key concepts recommended by World Health Organisation (WHO).

- By the ladder
- By the clock
- By the appropriate route
- By the child

By the ladder

Use the "three-step" approach to analgesia as proposed by the WHO, illustrated in Figure 1.28.1. Pain is classified as mild, moderate or severe and analgesic choices are adjusted accordingly. The ladder approach is based on drugs that should be widely available in all countries. The sequential use of analgesic drugs is based on the child's level of pain, with a non-opioid analgesic usually the first step. Importantly, however, assessment of a child's pain may indicate immediate use of a strong opioid.

There should be no hesitation in moving on to the next step of the analgesic ladder if pain control is inadequate. Only one drug from each group should be used at the same time. For example if a weak opioid, for example codeine, ceases to be effective, then a strong opioid should be prescribed, not an alternative weak opioid. Strong opioids can be increased until pain is relieved. Occasionally an alternative strong opioid may be substituted if its side effects are intolerable.

The aim is for the child to be:

- Pain-free on movement
- Pain-free at rest
- Pain-free at night

By the clock

Analgesia should be given regularly, for example every 4 hours. **There is no place for "as required" prescribing of analgesics in palliative care**. The dosing interval should be determined according to the severity of the child's pain and the duration of action of the drug being used. Additional "rescue" doses for intermittent and breakthrough pain should be prescribed. Effectiveness of analgesia should be regularly reviewed, as effective titration of analgesia demands reassessment.

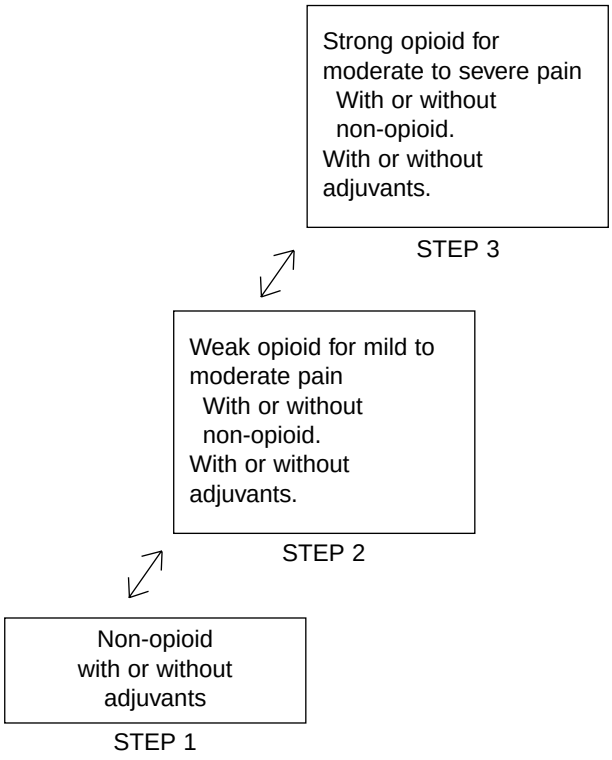

Figure 1.28.1 WHO three-step analgesic ladder.

By the appropriate route

Children should have drugs by the most simple, effective and least painful route; the oral route being the preferred route. IM injections should not be used; they are painful and there is a risk of abscess and/or haematoma formation particularly with children who may have low platelet counts or other clotting problems. Considerations in selecting the best route of analgesic administration include the nature and severity of the pain, the potency of the drug, the required dosing interval and the compliance of the child.

By the child

Doses of all analgesics must be based on each child's symptoms and circumstances; there is no single dose that will be appropriate for all children.

Regular reassessment of a child's pain and effectiveness of the analgesia is essential, in order that that the drugs can be adjusted accordingly to maintain a child pain free.

Some children, particularly with malignancy induced pain, may require very large doses of opioids to achieve satisfactory pain control, hence it should be noted that some of the suggested dosage recommendations included in this section are different to that

specified elsewhere in the manual. This is appropriate in palliative care and reflects the different goals and priorities in the acute and palliative setting.

Analgesics

Non-opioid analgesics

Non-opioid analgesics are used to relieve mild pain or, in combination with opioids, to relieve moderate and severe pain. **Paracetamol** is the drug of choice because it has a very high therapeutic ratio for children and can be given orally or rectally. It is available as an elixir, tablet or suppository form and can be given 4–6 hourly. For dosage see Chapter 1.27.

Weak opioid analgesics

The two most commonly used weak opioids are

- **Codeine**
- **Dihydrocodeine** (see Chapter 1.27 for doses)

Weak opioids have a "ceiling effect", which means their analgesic power does not increase beyond a certain dose. However, with increasing doses, side effects such as respiratory depression and constipation can occur. Laxatives should generally be prescribed. It is sometimes appropriate to progress straight from paracetamol, to morphine.

Strong opioid analgesics

Strong opioids are required alone, or in combination with non-opioid analgesics and/or adjuvant drugs, to provide effective pain relief. Strong analgesics do not have an analgesic "ceiling effect", i.e. there is no maximum dose and children may require extremely large doses to obtain pain relief.

Morphine

The strong opioid of choice internationally is oral **morphine**. ***Alternatives include fentanyl, hydromorphone, oxycodone and methadone.*** Cost and availability may/will dictate the choice(s) of strong opioid.

Morphine is preferred for the oral route, but if the subcutaneous or IV route is required, **diamorphine** (where available) is the preferred drug, as it is more soluble in water (for example only 1.6 ml of water is required to dissolve 1 g of diamorphine). Diamorphine and morphine can both be given by a slow continuous infusion, which will give a steady level of analgesia and is preferred to intermittent SC or IV administration.

Children peaking at age 1–7 years have been found to rapidly eliminate morphine metabolites. This group of children may require more frequent dosing and relatively higher doses to achieve pain relief.

✔ **Morphine must be available in all countries**. Ideally it should be available as immediate and sustained release preparations including immediate release suppositories. ***Once-daily preparations are commercially available, but there is little experience of their use in children.***

Immediate release

- Morphine tablets (Sevredol) 10 mg, 20 mg, and 50 mg.
- Morphine sulphate mixture (Oramorph) 10 mg/5 ml.
- Morphine sulphate mixture (Oramorph concentrate) 100 mg/5 ml.

Oral morphine starting doses

1 mg/kg/24 hours if not previously receiving opioids and 2 mg/kg/24 hours if already on a weak opioid. Immediate release morphine should be given regularly every four hours; it may be useful to increase the night-time dose by 50–100% to eliminate night time waking in pain.

Sustained release

- Morphine tablets (MST Continus) 5 mg, 10 mg, 30 mg, 60 mg,100 mg and 200 mg.
- Morphine granules for suspension (MST Continus) 20 mg, 30 mg, 60 mg, 100 mg and 200 mg.

Conversion to sustained release morphine is usually useful once a child is established on morphine. This prevents fewer peaks and troughs in drug levels, reduces the amount of drugs children have to take and minimises the need for night-time waking. The first dose of sustained release morphine should be given at the same time as the last dose of immediate release morphine.

Starting dosage

2 mg/kg/24 hours or divide current total daily dose of immediate release morphine by two. The sustained release morphine should be given in two divided doses at 12-hourly intervals. Occasionally younger children may require 8-hourly dosing.

Breakthrough pain

For both preparations immediate release morphine should be prescribed in a dose equivalent to the four hourly dose (i.e. one-sixth of the total daily dose). This can be given up to hourly for breakthrough pain and parents should be advised to keep a record of all extra doses given so that the regular dose of morphine can be titrated accurately.

Titration of morphine dose

Pain relief should be reviewed regularly. The morphine dose should be titrated against the level of pain. If frequent breakthrough analgesia is required, the total dose of morphine taken during the day (regular plus "breakthrough" doses) is assessed. Usually increments of 20–50% of the previous total daily dose are required. Regular review allows the regular dose of morphine to be adjusted according to the level of breakthrough pain. *Remember:* to increase the dose of breakthrough morphine accordingly, when the regular dose is increased.

Alternatives to oral route of administration

Indications
- Persistent vomiting
- Non-compliance with oral medication
- Dysphagia
- Bowel obstruction
- Physical deterioration preventing oral intake
- Unsatisfactory response to oral medication

Subcutaneous
Many drugs are well absorbed subcutaneously and this route of treatment can be easily established in children without venous access.

Oral morphine to subcutaneous or intravenous diamorphine or morphine
For both SC and IV routes the dose should be titrated to overcome breakthrough pain in increments of 20–50%.

IV infusions are best in children with indwelling central venous lines, for example Hickman OR where death is judged to be imminent (within a few days).

Diamorphine
- Diamorphine (if available) is the drug of choice for both SC and IV use because it is more soluble and lower volume
- Divide the total daily dose of oral morphine by 3 to obtain the equivalent daily dose of diamorphine
- Starting dose of diamorphine in those where oral morphine has not been given is 12.5–25 micrograms/kg/hour by continuous infusion.

Morphine
- The potency of morphine by infusion or injection is approximately twice that of oral morphine. Therefore use one-half of the total daily dose of oral morphine as the equivalent 24 hour morphine dose for SC or IV infusion.

If syringe drivers are not available, parents can be trained to give regular boluses of morphine SC or IV. The total daily dose of either morphine or diamorphine is divided by a practical number that coincides with the number of individual doses to be given by the parents (usually 1–2 hourly in the day and 4 hourly at night).

Rectal
This route may be acceptable for some children who are unable to take oral medication.
- Paracetamol can be given as suppository.
- Morphine suppositories can be given if available.
- Use the same dose and interval, i.e. 4 hourly.
- Anecdotally MST tablets may be used rectally.

Epidural/Intrathecal
- ***Occasionally indicated in uncontrolled neuropathic pain.***
- ***This method of administration should only be considered with the help of a suitably experienced anaesthetist.***

Side effects of opioids

All opioid drugs cause similar side effects. These problems are well known and should be **anticipated and treated whenever children are given opioids, so that pain control is not accompanied by unacceptable side effects**. Parents and children, when appropriate, should be informed about the possible side effects and their management. Children on strong opioids should be assessed regularly. ✔

Constipation
Is a common side effect and laxatives, such as co-danthramer and docusate, should always be prescribed with strong opioids (see below). Advice should be given to increase fluids and fibre (vegetables, fruit and cereals) in the child's diet where appropriate.

Nausea and vomiting
Routine antiemetics are not commonly required, but should be prescribed in case of opioid induced nausea and vomiting (see section below on nausea and vomiting). When it does occur it normally resolves within 3–4 days.

Drowsiness and confusion
Daytime drowsiness, dizziness and mental clouding can occur at the start of treatment and sometimes following a dose increase. This almost always resolves within a few days. Cognitive and psychomotor disturbances are minimal once patients are receiving a stable dose of opioid.

Pruritus
Itching is not an uncommon side effect of opioids in children. Simple skin care may be effective alone:
- Avoid hot baths and avoid using soap
- Add oilatum to the bath water and use aqueous cream as a soap substitute
- Massage skin gently using aqueous cream, palm or olive oil
- Avoid overheating and sweating
- Cool cotton clothing and bedding
- Keep nails short to discourage scratching

If itching is persistent, review medication. If itching is opioid related and the opioid cannot be changed, the addition of a systemic antihistamine such as chlopheniramine may be beneficial.

Pruritus associated with obstructive jaundice will require good skin care plus systemic medication such as ***stanozolol or ondansetron or levomepromazine, if available.***

Respiratory depression
Respiratory depression is uncommon in the conscious patient with severe pain. If it does occur, management will be dictated by the child's overall condition.

Nightmares and hallucinations
Nightmares can occur. If they are distressing and not resolved by reassurance or resolution of other anxieties, try haloperidol at night (50–100 micrograms/kg). Hallucinations are rare and again haloperidol at night may be useful.

Urine retention
Urine retention may be a problem particularly after rapid dose escalation. Most children respond to simple measures such as a warm bath, warm packs or relief of constipation. Catheterisation may be required but is usually only needed as a temporary measure.

Morphine toxicity

Can occur with:
- Too high dose
- Too rapid dose escalation
- Pain that is not morphine responsive
- Renal impairment
- Following therapeutic intervention to relieve pain, for example radiotherapy or nerve block

Warning signs include:
- Drowsiness
- Confusion
- Pin-point pupils
- Myoclonic jerks
- Hallucinations (auditory and visual)
- Vomiting
- Nightmares

If toxicity occurs, consider reducing morphine dose (several doses may need to be missed) then restart at a lower dose or stop morphine. However, WATCH CAREFULLY FOR BREAKTHROUGH PAIN. Address side effects as discussed above. Escalating doses of opioids and metabolic disorders can exacerbate myoclonic jerks. Oral diazepam can be useful. If a child is unable to swallow, rectal diazepam or subcutaneous midazolam are effective.

Consider an alternative strong opioid if available, for example hydromorphone or transdermal fentanyl. Consultation with personnel experienced in palliative care is recommended.

Addiction and tolerance

Addiction and tolerance are common unfounded fears. Psychological addiction and tolerance are not a problem when opioids are used correctly in palliative care.

Prescribing opioids in renal impairment

The active morphine metabolites are renally excreted and accumulate in renal impairment causing toxicity. When prescribing any opioid analgesics in children with renal failure, care must be taken, as they are extremely sensitive to opioids.

Alternatives to oral morphine for severe pain

✔ **Few children are truly morphine intolerant, and if the pain is not responding to morphine always consider the aetiology of the pain and consider the use of adjuvant therapy.**

- ***Opioid rotation should be considered if the side effects of morphine are intolerable at a dose required for adequate analgesia.***
- ***Opioid rotation will depend on the availability and cost of alternative strong opioids and should be undertaken in consultation with personnel experienced in palliative care.***
- ***Alternative drugs include:***
  - ***Transdermal fentanyl***
  - ***Hydromorphone***
  - ***Methadone***
  - ***Oxycodone***

Transdermal fentanyl will be the only drug discussed in depth here.

Transdermal fentanyl (Durogesic)
Can be considered in situations of stable opioid requirements but unacceptable:
- ***constipation***
- ***nausea and vomiting***
- ***mood disturbance***
- ***unable to swallow***
- ***poor oral compliance***
- ***drowsiness***

Fentanyl is unsuitable in children with:
- ***Rapidly changing opioid requirements***
- ***Chronic skin disorders***
- ***Renal or hepatic impairment***
- ***Intolerance to removal of adhesive dressings***

Four patch strengths are available, each delivering the patch strength in micrograms/hour:
- ***Durogesic 25***
- ***Durogesic 50***
- ***Durogesic 75***
- ***Durogesic 100***

The appropriate dose of fentanyl is determined from the child's total daily dose of morphine.

The first fentanyl patch needs to be applied at the same time as the last dose of MST. Each patch will last 72 hours. When converting from four-hourly oral morphine, children will usually need 3–4 four-hourly doses of morphine after the patch has been applied.

Parents should be informed that breakthrough doses of oral morphine may be required during the first 24–48 hours of patch application (to allow the subcutaneous depot of fentanyl to build up). Breakthrough dose of immediate release morphine must be prescribed with fentanyl, at the breakthrough dose that would be needed if the patient were taking the equivalent dose of morphine.

Children should have their dose titrated according to breakthrough pain, normally in dose increments of 25 micrograms/hour.

If the patches are discontinued it is important to remember that a subcutaneous depot of fentanyl persists for up to 24 hours after patch

removal, when prescribing a replacement analgesic.

Table 1.28.1 Conversion chart for oral morphine and fentanyl patches

24-hourly dose of oral morphine (mg)	*Equivalent fentanyl dose (micrograms/hour)*
30–134	25
135–224	50
225–314	75
315–404	100
405–494	125
495–584	150
585–674	175
675–764	200
765–854	225
855–944	250
945–1034	275
1035–1124	300

Hydromorphone

Hydromorphone is similar to morphine in its pharmokinetics, efficacy and toxicity, but it is about six times more potent on parenteral administration and eight times more potent when given orally. It is available (in some countries) for oral, rectal and parenteral administration and its oral: parenteral ratio is 5:1.

Methadone

Methadone is a synthetic, long-acting analgesic, which is recommended for children unable to tolerate morphine and hydromorphone because of side effects. The oral route is preferred. The prolonged half-life of methadone necessitates extremely careful titration to achieve pain control. A starting oral dose of 200 micrograms/ kg (WHO) is recommended, but effective dosing schedules may range from 4–12 hourly. Accumulation can occur over a few days and cause marked drowsiness and respiratory depression. The methadone should be withheld until the child is easily rousable and the dose recommenced at 50% or at longer dosing intervals.

Methadone should initially be given as required until a child's requirement is established and regular dosing can be established, during this period (24–48 hours) a child requires careful monitoring for side effects.

Methadone is not an appropriate drug to use for a child with a rapidly changing clinical condition and analgesic requirements.

Oxycodone

Oxycodone is a strong, oral, opioid indicated for use in moderate to severe pain. This means that it can be started at level two of the analgesic ladder and titrated upwards with no "ceiling" dose and no need to change to morphine. There is little experience of its use with children.

Adjuvant therapy

Types of pain

Neuropathic pain

Co-analgesics such as an anticonvulsant or tricyclic antidepressant are essential, because this pain is only semi-responsive to opioids. It should be considered if the pain has a burning or stabbing/shooting component.

Nerve compression pain

May arise from compression of a nerve root and morphine plus a trial of oral steroids should be tried. The steroid should relieve pain within 48 hours, probably through reduction of oedema around the tumour. If no improvement occurs, steroids should be discontinued.

Nerve injury pain

May arise from tumour invasion of a nerve or as a side effect, for example radiotherapy.

Bone pain

Non-steroidal anti-inflammatory drugs (NSAIDs) have analgesic, antipyretic and anti-inflammatory properties. They are often effective in musculoskeletal pain that is associated with bone metastases or soft tissue inflammation.

- Regular dosing is required for their full effect, but the maximum is usually seen within two weeks.
- It is worth trying another NSAID if there is no response to one type.
- Damage to the gastrointestinal mucosa is the most frequent side effect.
- NSAIDs are not usually appropriate for children with thrombocytopaenia.

For common dosages see Chapter 1.27.

Difficult cases

Consider referral to a pain specialist, for consideration of, for example, spinal analgesia or regional nerve blocks.

Anticonvulsants

Useful for pain that is shooting or stabbing. Carbamazepine and sodium valproate are commonly used, ***with clonazepam and gabapentin more recent additions.*** Start at a low dose and gradually increase to avoid sedation and toxicity.

- Carbamazepine
  Starting dose: 2.5 mg/kg twice daily increasing by 2.5–5 mg/kg/day at weekly intervals.
  Maintenance dose: 10–20 mg/kg/day in 2–3 divided doses, increasing gradually as above.
- Sodium valproate
  Starting dose: 20 mg/day in two divided doses, increasing if required by increments of 5 mg/kg at weekly intervals.
  Maintenance dose: 20–30 mg/kg/day in two divided doses.

Tricyclic antidepressants

- Useful for pain that is burning in nature.
- Give at night to avoid excessive sedation during the day. Can cause constipation.
- Analgesic effect begins after about 5–7 days of treatment but may take longer.
- Starting dose: Amitriptyline 0.5 mg/kg at night increasing if needed to 1 mg/kg/day. Increase carefully to avoid excessive drowsiness.

Steroids

Steroids have specific benefits for palliative care because of their ability to produce euphoria, improve appetite and weight gain. They also have an anti-inflammatory effect, which may help with nerve compression and raised intracranial pressure. However, they should be used with caution in children as the side effects of steroids can far outweigh the benefits, for example rapid weight gain, change in appearance, mood swings and behaviour changes and insomnia, which can be distressing for both the child and parents. If there is no improvement in symptoms within a short period of time (for example 5–7 days), steroids should be discontinued. If initial symptom relief is not maintained, long-term use should be avoided.

- Dosage: dexamethasone should be taken before 6 pm to minimise insomnia.
- High dose: normally used to relieve raised intracranial pressure or nerve compression pain.
  - 2–5 years: 2 mg twice daily
  - 6–12 years: 3 mg twice daily
  - over 12 years: 4–8 mg twice daily
- Low dose: normally used to improve appetite and well being.
  - 2–5 years: 0.5 mg–1 mg daily
  - 6–12 years: 1–2 mg daily
  - over 12 years: 2–4 mg daily

Radiotherapy

Radiotherapy, if available can be particularly useful in treating isolated sites of a radiosensitive tumour. This may include bony metastases, spinal cord compression, and relief of nerve compression from a solid tumour and isolated cerebral metastases. Radiotherapy can also be used in the management of fungating tumours. Single treatments or short courses are often appropriate and effective in palliative care.

Non-pharmacological approaches

Non-drug therapies must be an integral part of the management of children's pain, complementing, but not replacing appropriate drug therapy.

A combination of non-pharmacological approaches, used in conjunction with analgesics, may be extremely effective. These approaches include:

- Progressive relaxation
- Hypnosis and guided imagery
- Massage and reflexology
- Play and distraction therapy
- Heat or cold pads
- ***Transcutaneous electrical nerve stimulation (TENS)***

Other symptom management

Nausea and vomiting

These are common symptoms in palliative care. The causes may be multifactorial and it is important to try to determine the cause in order to implement an effective treatment plan.

- Cancer related
  - Raised intracranial pressure
  - Abdominal mass
  - Irritation of upper gastrointestinal tract
  - Gastric outflow obstruction
  - Anxiety
  - Uraemia
  - Pain
  - Blood in stomach
- Treatment related
  - Opioids (side effect and as a result of constipation)
  - Chemotherapy
  - NSAIDs
  - Corticosteroids (IV)
  - Carbamazepine
  - Antibiotics

Management

- Identify cause(s) as above and implement appropriate management, for example for constipation, raised intracranial pressure.
- Consider stopping gastric irritants if possible, for example antibiotics, NSAIDs, steroids. Prescribe H_2-receptor antagonist (ranitidine 2–4 mg/kg 12 hourly.
- Prescribe an appropriate antiemetic according to cause
- Review therapy regularly and adjust as required
- If treatment is unsuccessful, consider:
  - Was the cause of the vomiting correctly identified and the appropriate antiemetic prescribed?
  - Has the antiemetic had time to work at maximum dose?
  - Is the route of administration appropriate for the child?

Antiemetic therapy

- Antiemetic choice depends on the cause of vomiting and the site of the antiemetic action.
- Combinations of drugs with different sites of action are sometimes required.
- To avoid side effects, avoid combining drugs of the same class.
- Severe nausea and vomiting may require initial management by SC or IV infusion and then switching to oral medication when control is gained.

1 For Drug induced and metabolic causes of vomiting

Dopamine antagonists
Avoid combining two of these drugs, which may increase risk of extrapyramidal side effects.

Haloperidol
Antiemetic of choice for opioid-induced vomiting. It acts on the chemoreceptor trigger zone.

Dosage: 12.5–25 micrograms/kg/twice a day, oral, SC or IV.

Methotrimeprazine (levomeprazine)
Powerful broad-spectrum antiemetic, acting at several sites. Highly sedative at high doses, increasing evidence that it is an effective antiemetic at low doses without sedation. It is often effective as a single night-time dose.

Oral dosage: 75–250 micrograms/kg 6 hourly or 500 micrograms/kg as a single night-time dose

IV or SC dosage: 50–125 micrograms/kg 6 hourly or 250 micrograms/kg as a single night-time dose

2 For vomiting due to raised intracranial pressure/intestinal obstruction

Cyclizine
Antihistamine, acts on the vomiting centre.
Oral dosage:
All ages 1 mg/kg three times daily to a maximum 50 mg per dose
IV or SC dosage:
All ages 1 mg/kg/8 hourly

Dexamethasone
Dosage: use moderate doses, i.e. 100 micrograms/kg/12 hourly.

3 For vomiting due to gastrointestinal problems
Metoclopramide
Acts both on the upper gastrointestinal tract and the chemoreceptor trigger zone. Speeds up gastric emptying. Extrapyramidal side effects more common in children.

Useful for: oesophageal reflux, gastric stasis, gastric irritation and squashed stomach; **avoid in complete bowel obstruction**.

Oral dosage:

1–12 years:	100 micrograms/kg	2–3 times a day
Over 12 years:	5–10 mg	2–3 times a day

SC/IV dosage:

Maximum of	500 micrograms/kg	over 24 hours

Domperidone
Acts both on the upper gastrointestinal tract and the chemoreceptor trigger zone, speeds up gastric emptying.
Oral dosage:

1–12years:	200–400 micrograms/kg	3–4 times a day
Over 12 years	10–20 mg	3–4 times a day

Rectal dosage:

1–12 years	15 –30 mg	3–4 times a day
Over 12 years	30–60 mg	3–4 times a day

Constipation

Constipation is common in paediatric palliative care and the causes may be multifactorial. The prevention and relief of constipation for the terminally ill child is very important, as unresolved it can cause abdominal pain and discomfort and nausea and vomiting.

Consider the following causes:
- Drug induced, i.e. opioids, anticholinergics and antidepressants
- Reduced activity
- Poor oral intake of fluids and food and general debility
- Dehydration
- Bowel obstruction
- Spinal cord compression

Management
- Treat underlying cause where appropriate and possible.
- Constipation should be anticipated when opioids, anticholinergic or antidepressant drugs are being used and laxatives prescribed prophylactically.
- Use laxatives appropriately and at the right doses and avoid mixing two drugs of the same group, for example two stimulants.
- A good first choice is the combination of stimulant laxative with a softening agent, for example senna plus lactulose or docusate, ***or co-danthramer or co-danthrusate***.
- Titrate doses up as required, rather than adding a new laxative.
- If oral therapy fails, consider rectal measures such as suppositories/enemas.

Bowel obstruction

Bowel obstruction may be mechanical or functional or both. In children with advanced disease, surgical management is not usually indicated. The aim of treatment is the palliation of symptoms. Nasogastric tubes and IV fluids are rarely indicated.

Management
Eliminate pain and colic
- For constant background pain, administer diamorphine/morphine by continuous IV or SC infusion, using a portable syringe driver. If this is not available, parents can be trained to give frequent (hourly or even ½ hourly boluses).
- If colic is present, avoid prokinetic antiemetics (for example metoclopramide, domperidone).
- Discontinue bulk-forming, osmotic and stimulant laxatives.
- Relieve associated constipation, continue softening agents if possible and use rectal measures to relieve faecal impaction.
- If colic persists add hyoscine butylbromide (Buscopan) 10–20 mg orally 8 hourly or 5–20 mg IV as a single dose over at least 1 minute (5 mg 2–5 years, 10 mg 6–10 years, 15 mg 11–14 years, 20 mg 15–18 years). Repeat 8 hourly as required.

Eliminate nausea and reduce vomiting to once or twice a day
- The choice antiemetic depends on whether colic is present.

- If colic is present, cyclizine is the first-line drug. Add haloperidol if nausea persists. If this is ineffective ***substitute for both with methotrimeprazime if available***.
- If colic is absent and flatus is present a trial of SC or IV metoclopramide is indicated. If ineffective instigate management as above.
- Dexamethasone may be of benefit in second-line management.

Dyspnoea

Shortness of breath associated with pulmonary complications in advanced paediatric cancer can be very distressing for both child and parents and requires effective management. The underlying pathophysiology needs to be considered when deciding the management.

Common causes of dyspnoea are:
- Metastases
- Effusions
- Pulmonary fibrosis
- Anaemia
- Infection
- Superior vena cava obstruction
- Anxiety/fear
- Increased secretions
- Cardiac failure
- Chest wall pain/constriction
- Pulmonary embolus

Management
- Identify cause
- Give explanation to parents and child
- Treat specific cause(s) or modify pathological process, for example high-dose steroids and ***radiotherapy*** for superior vena cava obstruction
- Non-drug measures
  - A calm approach
  - Breathing exercises
  - Position
  - Calm environment
  - Cool draught of air for example by fanning
  - Play therapy
- Drug measures: these are outlined below:

Opioids
Have a complex action on respiration, which are not fully understood. They often reduce respiratory rate to a more comfortable level. Opioids should be prescribed regularly in children with continuous breathlessness at standard starting doses. If already on opioids increase dose by 30–50%.

Benzodiazepines
Anxiolytic and sedative effects also cause relaxation of the smooth respiratory muscles.

May be helpful if the child or teenager is very anxious, as a single dose and then at night or twice daily.

Diazepam (oral)
Dosage:

4 weeks to 1 year:	
200 micrograms/kg	2–3 times daily
1–12 years:	
2 mg	2–3 times daily
>12 years:	
5 mg	2–3 times daily

Lorazepam: well absorbed sublingually (good for panic attacks), short acting and rapid onset of relief.
Dosage:

1–12 years	***50 micrograms/kg***	***per dose***
>12 years	***1–2 mg***	***per dose***

May be repeated after 12 hours

Corticosteroids:
- May be useful particularly in superior vena cava obstruction and multiple lung secondaries.
- Use moderate doses of dexamethasone.
- Benefit should be apparent in 5 days.
- Reduce to lowest effective dose.

Oxygen
- May be helpful for patients with hypoxeamia.
- May be helpful if a child is very anxious.
- Nebulised saline or salbutamol may provide subjective relief.

Cough

Consider the following causes:
- Respiratory infection
- Airways disease
- Obstruction of the airway by tumour
- Drug induced
- Oesophageal reflux
- Aspiration of saliva
- Treatment related

Wherever possible the cause of the cough should be treated.

Symptomatic management should follow the guidelines for the management of dyspnoea.

Drug management may include:
- Simple linctus
- Codeine linctus
- Opioids
- Nebulised saline
- Oral antibiotics: indicated if symptomatic chest infection with productive cough affecting quality of life

Anxiety

This is common in palliative care.
- Try and identify the cause of a child's anxiety; for example is it related to symptoms or fears about what is happening
- Simple explanations, reassurance and a calm environment are important
- Relaxation and massage may help
- Diazepam, ***lorazepam***, or midazolam, as required or regularly, may help if other measures fail
- Anxiety and discomfort go together so reassess pain

Management of bleeding

Massive external bleeding

Death from massive external bleeding is uncommon in children, but the risk of this is frightening and distressing for both parents and the child and prevention of this should be the aim of management, though this may not always be possible.

Causes of external bleeding include:

- Low platelets
- Clotting deficiencies
- Primary or secondary liver disease
- Disease progression
- Previous treatment, for example radiotherapy/ chemotherapy

Management

If there is a risk of massive haemorrhage, it is extremely valuable to have diamorphine and an appropriate sedative readily available at home, for example IV/SC or rectal diazepam or midazolam.

Persistent surface bleeding

This is common in the child with leukaemia and can be alarming to both child and family. It can be managed in the home environment.

Management

- Epinephrine 1 in 1000 solution can be used topically soaked in gauze and applied directly to the bleeding point.
- Other haemostatic dressings can be used for persistent surface bleeding, for example in fungating tumours, these include ***Kaltostat, Sorbsan, Oxycell***.
- ***Tranexamic acid can be useful and can be used topically undiluted, direct to bleeding gums or nostrils, or as a mouthwash. It can also be given systemically as prophylaxis.***
- ***Platelet transfusions during palliative care are not routine practice, but should be considered to maintain quality of life and prevent distressing haemorrhage, where appropriate and available. Home platelet transfusion may be possible with appropriately trained personnel.***

Spinal cord compression

Consider with the following signs and symptoms:

- Localised pain in spine, radiating around chest
- Sudden onset of weakness, for example of legs
- Sensory disturbance
- Sphincter dysfunction

Investigations

- History and clinical findings
- ***Urgent MRI scan of spine or CT myelogram***

Management

- Patients with paraparesis do better than those who are totally paraplegic.
- Loss of sphincter function is a poor prognostic sign.
- Rapid onset of complete paraplegia has a poor prognosis.
- Main therapeutic options are:
  - Corticosteroids
  - Radiotherapy
- Steroids should be given in high doses initially and then reduced according to response. The steroids often bring about an early improvement and relief of pain by reducing the peritumour inflammation.
- ***Referral for concurrent radiotherapy should be considered if prognosis is not very poor.***
- ***Surgery, such as laminectomy is only occasionally indicated.***
- Consider:
  - Pressure-relieving mattress
  - Pressure area care
  - Bowel function
  - Physiotherapy to prevent contractures
  - Urinary catheterisation
  - Avoid danthron-containing aperients if the child is catheterised or incontinent, because of the risk of danthron burns

Fitting

Fitting may be a potential or existing problem for children with brain tumours or other neurological and metabolic disorders. For emergency management of seizures in palliative and terminal care diazepam given rectally is the drug of choice.

Diazepam dosage: rectally

< 1 year:	2.5 mg (half 5 mg rectal tube)
1–4 years:	one 5 mg rectal tube
5–12 years:	5 –10 mg rectal tubes
12 years	10 mg rectal tube

For continuing severe seizures, consider midazolam by SC or IV infusion (see Chapters 3.37 and 3.38).

Muscle spasm

Muscle spasm can be severe in children with neurological and neurodegenerative disorders. Muscle spasm can occur alone or can also be triggered by pain elsewhere, for example constipation.

Useful drugs for muscle spasm include: diazepam and ***baclofen***.

Incontinence

Incontinence can be the source of much discomfort and anxiety for both children and their families, as well as presenting difficulties in keeping the child clean and protecting the skin. Children with some degenerative conditions may have had faecal or urinary incontinence for a long time, whereas for others it may become a feature towards the end stage of their disease (for example due to local tumour, neurological/spinal cord damage to bladder control, laxative imbalance).

For children with long-standing difficulties, intermittent catheterisation or the use of an indwelling catheter may be a well established, successful and accepted method; however, where the problem occurs at end-stage disease this can sometimes be an unwelcome and inappropriate

intervention. It may also be that sufficient supplies are not available for the choice to arise.

Some suggestions that might be useful:

- Review of laxatives where appropriate
- ***Consider intranasal desmopressin dose 20–40 micrograms at bedtime if nights are disturbed by urinary incontinence.***
- Keep urinal or bedpan close to bedside.
- Use pads or towels (with plastic underneath) on top of bed sheets to lay child on. This will avoid the need to change all the sheets and therefore minimise disturbance to the child.
- Keep area well ventilated (or window open if appropriate).
- Try to ensure the skin is kept clean and use dimethicone based barrier creams where available.
- Help the child to wash regularly.
- Try to preserve and maintain the child's dignity at all times. Give reassurance and support to both child and parents.

Fungating wounds

Fungating wounds are extremely rare in paediatric palliative care. They may occur with soft tissue sarcomas, often of the head and neck, which can be very distressing for the child and family.

Useful tips for management (where available) include:

- Soak any dressings with saline to ease removal, as these tumours may be friable and prone to bleeding.
- Have available if possible topical epinephrine 1 in 1000, ***or an alginate dressing, for example Kaltostat, or tranexamic acid, to apply topically to the tumour if it bleeds profusely, for example during a dressing change.***

These tumours can cause offensive smells due to anaerobic organisms, which can be distressing to the child and family. ***Topical preparations of metronidazole, such as Anabact or Metrotop, may be useful.*** Oral metronidazole may be used. ***Charcoal dressings, if available may be helpful to absorb the smell.*** Simple measures such as using aromatherapy oils around the home may be helpful.

Final days/ hours of life

Terminal restlessness and agitation

These symptoms are not uncommon in the final stages of life; useful drugs include midazolam, ***methotrimeprazine*** and rectal diazepam.

Midazolam

- Sedative of choice.
- Mixes with other commonly used drugs in syringe drivers or syringes used manually by parents.
- ***Can be used with methotrimeprazine.***

Dosage:

Initial single loading dose:	100 micrograms/kg by SC or IV injection
Initial regime:	30–100 micrograms/kg/hour by SC or IV infusion, titrate upwards as required, (upper dose may be limited by volume).

Methotrimeprazine (levomepromazine)

- ***A phenothiazine – antipsychotic***
- ***Very sedating at higher doses***
- ***Drug of choice if antiemetic effect also required***
- ***Can be combined with midazolam***
- ***Available orally***
- ***Use with caution in children with cerebral metastases or epilepsy***

Dosage:

All ages:	***15–100 micrograms/kg/hour by SC or IV infusion***
If oral is possible:	***250–1000 micrograms/kg/ 24 hours.***

Rectal Diazepam

May be useful if IV or SC infusions are not possible or the drugs not available.

Dose range: Doses: see for "fitting" above. Dose may be repeated if child remains very agitated and restless.

Increased secretions

- Can be very distressing for parents and carers ("death rattle").
- Good mouth care is essential.
- Antisecretory agents are useful but can cause drowsiness and anticholinergic side effects.
- Start drug treatment early to avoid build up of excessive secretions.

Hyoscine hydrobromide (scopolamine)

- An anticholinergic
- Reduces pharyngeal secretions
- Use prophylactically at first signs of excess secretions
- Mixes with other commonly used drugs
- Potential routes for administration: oral, ***transdermal***, SC or IV
- ***Hyoscine hydrobromide may be available as a transdermal patch releasing 1000 micrograms/72 hours (Scopoderm TTS) or as a sublingual tablet (Kwells) 300 micrograms.***

Dosage:

Oral/sub-lingual:

1–12 years:	10 micrograms/kg/dose
>12 years:	400 micrograms/dose

Patch/72 hours:

1–4 years:	***quarter of 1000 micrograms patch***
>4 years:	***half patch/72 hours***

Infusion (SC, IV):

All ages:	10–50 micrograms/kg/24 hours

Loss of the oral route (use of SC (or) IV medication)

As a child's condition deteriorates the oral route for medication may become difficult. As discussed earlier, other

routes that can be used at this point are rectal, SC and where already established, the IV route. Children who have been treated for cancer may have central IV access, which can be used effectively in palliative care.

Drugs, which can be given SC or IV, include analgesics, antiemetics, sedation, anxiolytics and anticholinergics; these can be combined together in an infusion, as long as they are compatible with each other.

Where available, it is possible to use small, portable infusion pumps at home, to deliver combinations of medication over 24 hours, (for example the Graesby MS 26 or the Walkmed). If they are not available, parents can give frequent SC or IV boluses using standard syringes and indwelling catheters.

Additional points

- Avoid high concentrations of drug combinations, especially when using cyclizine.
- Avoid mixing dexamethasone with other drugs if possible.
- Never use chlorpromazine, prochlorperazine and diazepam subcutaneously.
- More than two drugs can be combined in portable syringe drivers or manual syringes, although there is little supporting clinical data. Consult your local pharmacist before using any unusual combinations.

Psychological support for the child, parents and siblings

Care that is child and family centred is an essential principle of palliative care. The availability of an experienced key worker to coordinate a child's care with community healthcare professionals and parents is essential. Good communication between professionals and between professionals and the family is paramount.

Initially parents may need a lot of support around making the decision to withdraw curative treatment and where to care for their child. Whether in hospital or at home, parents will have many questions, fears and anxieties during this time and the opportunity to discuss their worries, changes in the child's condition and symptom management, should be available if possible 24 hours a day. Commonly asked questions include. "How long will it be?" and "How will my child die?" These are not easy questions to answer and will also depend on the nature of the child's illness. For example, children with leukaemia may have a very short period of palliative care, whilst a child with a brain tumour or neurodegenerative disease may live for several months. It is probably best to give an indication of time span, but highlighting that every child is different and guiding parents as the disease progresses. "Days or weeks", "weeks or months" or even "hours rather than days" give adequate warning without being too precise.

Parents may also worry about their child being in pain, but also have anxieties about using strong medication like morphine; a clear explanation of the use of analgesics is essential in this situation.

Talking to the dying child and siblings is a subject that many parents may want advice about. Preparing brothers and sisters will depend very much on their age, understanding and parental beliefs. For older children and teenagers it is probably best to be honest with them, preparing them gradually for what is happening and allowing them to ask questions and participate in their sibling's care if appropriate. With younger children, the language used must be very simple and clear, for example avoid using "going to sleep" as the euphemism for death, and it is probably more appropriate to prepare them for a sibling's death when the end is obviously very close.

Talking to the child that is dying is very personal for parents and will also be influenced by the child's age and understanding of their illness. Examples of this include a teenager with cystic fibrosis who may have always known that they may anticipate death in adolescence or young adulthood, or the teenager who has had multiple relapses of cancer since they were three years old and realises that curative treatment is no longer working. Where possible and appropriate, it is important that children and teenagers are given the opportunity to express their wishes and anxieties. When children are not enabled to express themselves they can become very anxious and agitated or even withdrawn. Professionals can only try and encourage parents to have an open and honest approach to their child's questions and wishes during this time.

Preparation for death

Parents commonly have many questions about the time and nature of death and what happens afterwards. It can be very helpful to try and prepare parents for what may happen at the time of death if they wish to have this information. Changes in breathing are commonly distressing. Simple explanations of, for example, Cheyne-Stokes respiration or the 'death rattle' can avoid unnecessary distress. A sighing respiration after death if the child is moved is not uncommon and it should be explained to the parents that their child is not still alive. Explanations of the changes in colour and the very cold feel of the skin are important for parents and siblings. Parents have been distressed that their child was incontinent at the time of death. For some diseases, for example leukaemia, parents will need warning that their child may bleed from the nose or mouth at the time of or after death, and given simple practical measures to manage this situation.

Some families will require professional support around the time of death and it is essential that this provision is available.

After their child has died, parents need to know that they need not rush to do anything, but spend time with their child. However, it is important that any specific cultural or religious requirements are acknowledged and undertaken. Parents should be encouraged, if they wish, to hold their child and to wash and dress their child in their favourite clothes. Some may want to take photographs, or locks of hair or hand- and footprints and organise favourite toys, photographs and letters, etc. for

the child "to take with them". The participation of siblings in these activities can be very helpful.

In most countries, it is usually required for a child's death to be confirmed by a medical practitioner. In this situation it is very rare for a postmortem to be required. The death certificate then gives the authority for the death to be registered (according to the country's prevailing law) and the funeral arrangements to be made.

The various cultural and religious beliefs of different races and countries will play an important role in a child's funeral. However, parents may need advice about the choices of burial or cremation or about the funeral service itself.

Support after death

Support for parents, siblings and extended family around death and in the weeks and months afterwards, will be very much influenced by the family's culture and family network as well as the support provided during the child's terminal care. Bereavement contact from the professionals involved with the family should be offered where possible.

On-going bereavement support should be based on the family's particular needs and requests and the availability of appropriate bereavement support for both parents and siblings. Bereavement literature and parent support groups may be helpful where available.

Recognition of the child's birthday and anniversary are often important times for professionals and friends to remember.

Further reading

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1.29 Immunisation

Bernard Brabin

In 1974, WHO initiated the Expanded Programme on Immunisation (EPI). This aims to develop widespread national commitment to achieve high vaccination coverage. Choice of the original six EPI vaccines was based on importance of the disease and availability of safe, efficacious and low-cost vaccines.

Table 1.29.1 Expanded programme on immunisation schedule

Age	Vaccines	Hepatitis B vaccine**	
		Scheme A	Scheme B
Birth	BCG, OPV0	HB 1	–
6 weeks	DPT1, OPV1	HB 2	HB 1
10 weeks	DPT2, OPV2	–	HB 2
14 weeks	DPT3, OPV3	HB 3	HB 3
9 months	Measles, Yellow fever*	–	–

* In countries where yellow fever poses a risk.
** Scheme A: where perinatal transmission frequent. Scheme B: where perinatal transmission less frequent.

Polio

Oral live poliovirus vaccines (OPV) and inactivated injectable polio virus (IPV) are effective but have important differences.

- IPV produces neutralising antibodies to all three polio serotypes after two injections and can be included in the same vial as DPT.
- OPV requires at least a four-dose schedule which immunises most of the population, but additional doses are required to ensure immunity to all serotypes.
- The WHO target to eradicate poliomyelitis within the next ten years is dependent on high infant immunisation coverage and National Immunisation Days (NIDs) which aim to eradicate the circulation of wild virus. NIDs are designed to complement routine immunisation by targeting the most vulnerable in as short a period as possible. OPV is given over a two-day period, one month apart and the NIDs are repeated annually for at least three years.

Pertussis

Fever and mild local reactions are common. Consider two-dose schedule for those areas where services can be provided only twice a year.

Measles

Accelerated implementation of strategies to reduce the burden of measles are required including national immunisation days. Targeting children <5 years in major cities is a priority. Strategies to reduce impact of infant measles include:

- Increasing coverage to 9–23 months age group.
- Two dose schedule at 6 months and at an older age.

Tetanus

All health workers at antenatal clinics should guarantee that no women attending will have a child dying of neonatal tetanus by giving immunisation and advice about umbilical cord care. Vaccination of young adolescent girls is recommended in areas with poor antenatal coverage. Vaccinate mothers not seen antenatally when they bring their infants to clinic. ✔

BCG

Offers good protection against disseminated tuberculosis and also affords some protection from leprosy.

New vaccines and EPI

Hepatitis B and yellow fever vaccines have been recommended since 1992. Addition of Hib vaccine is also a priority and Hepatitis B and Hib should be given whenever possible alongside the DPT1–3 doses. Hepatitis B is especially important in endemic areas.

Conditions which are *not* contraindications to immunisation.

- Minor illnesses such as upper respiratory infections or diarrhoea, with fever $<38.5\,°C$
- Allergy, asthma, or other atopic manifestations, hayfever or "snuffles"
- Prematurity, small-for-date infants
- Malnutrition
- Child being breastfed
- Family history of convulsions
- Treatment with antibiotics, low-dose corticosteroids or locally acting (for example topical or inhaled) steroids
- Dermatoses, eczema or localised skin infection
- Chronic diseases of the heart, lung, kidney and liver
- Stable neurological conditions, such as cerebral palsy and Down's syndrome
- History of jaundice after birth

HIV infection and vaccination

Individuals with known or suspected asymptomatic HIV infection should receive all EPI vaccines according to nationally recommended schedules. Because of the risk of early and severe measles, infants should receive a standard dose at 6 months with a second dose as soon after age 9 months as possible.

Individuals with symptomatic HIV infection may receive all EPI vaccines except BCG and yellow fever.

BCG should not be given to children with symptomatic HIV infection (i.e. AIDS). In asymptomatic children, the decision to give BCG should be based on local risk of tuberculosis.

- Where risk of tuberculosis is high, BCG is recommended at birth or as soon as possible thereafter.
- In areas where risk of tuberculosis is low but BCG is recommended as a routine immunisation, BCG should be withheld from individuals known or suspected to be HIV infected.

Table 1.29.2 WHO/UNICEF recommendations for the immunisation of HIV-infected children and women of childbearing age

Vaccine	Asymptomatic HIV infection	Symptomatic HIV infection	Optimal timing of immunisation
BCG	Yes	No	Birth
DPT	Yes	Yes	6, 10, 14 weeks
OPV*	Yes	Yes	0, 6, 10, 14 weeks
Measles	Yes	Yes	6 and 9 months
Hepatitis B	Yes	Yes	As for uninfected children
Yellow fever	Yes	No**	–
Tetanus toxoid	Yes	Yes	Five doses***

*IPV can be used as an alternative for children with symptomatic HIV infection.
**Pending further studies.
***Doses of tetanus toxoid for women of childbearing age as for non-HIV infected persons.

Table 1.29.3 Recommended vaccine storage time and temperature

Vaccines	Shelf life	Transport state/district	State/district	Transport to PHC	PHC
DPT/TT and typhoid	1–1½ years (4–8°C)	+4 to +8°C	3 months (4–8°C)	4–8°C	1 month (4–8°C)
BCG	8 months (4–8°C)	+4 to +8°C	3 months (4–8°C)	4–8°C	1 month (4–8°C)
Measles and OPV	2 years at −20°C	−20°C to +8°C	3 months (−20°C)	−20°C to +8°C	1 month (4–8°C)

PHC = Primary Healthcare Facility

Immunisation instruments recommended by EPI include:

Sterilisable glass or plastic syringes and steel needles should after use, be immediately cleaned by placing them in a proprietary cleaning solution (for example Vizkon viricidal solution), rinsed with sterile water and stored in a sealed metal container until autoclaved. Handling precautions include either autoclaving or pressure kettle for 20 minutes, or boiling at 100 °C for 20 minutes. Disposable instruments are not currently recommended in disadvantaged countries because the temptation for their re-use is too great. Used needles should be placed in a hard container for disposal, sealed, autoclaved and ideally incinerated.

- Opened vials of OPV, DPT and hepatitis B vaccines may be used in subsequent immunisation sessions until a new shipment arrives (provided the expiry date has not passed, vaccines are kept in the cold chain and the vials have not been used outside the health centre).
- Opened vials of measles, yellow fever and BCG vaccines *must* be discarded at end of each immunisation session.
- Vaccines vial monitors (VVMs) will enable field staff to reject vials of vaccine which are heat damaged.
- Screen and immunise at every contact. Non-immunisation of eligible children at clinics may be as high as 30%.
- Reduce wastage by choosing correct vial size.
- ✔ **Ensure appropriate use of vaccine cold box.** Cold boxes only work if they are kept cold with the lids tightly shut. Ice packs are placed in the bottom and around the sides of the box and on the top of the vaccines. Newspaper should be placed between the vaccines and the icepacks to protect DPT and tetanus from the ice. A thermometer should be placed with the vaccines to record vaccine temperature when they are removed from the cold box. Diluent used to reconstitute measles and BCG vaccine must also be kept cold.
- Effective supervision requires focus on the essentials.
- Evaluate and monitor the programme.

Care of refrigerators

A constant supply of electricity, gas or kerosene is required. The electric plug can be taped to its socket to ensure it is not inadvertently removed. For gas and kerosene fridges a reserve full bottle of gas or can of fuel should always be present. A regulator valve should be used with the gas bottle. Kerosene tanks should be filled daily using a funnel and filter to remove dirt. The fridge should be positioned in a completely upright position with a draught blowing on the temperature exchanger to keep it cool. A fan can be used to achieve this if there is no wind and the ambient temperature is high.

130 Recognition of vulnerable children and the management of child ill-treatment and abuse

Neela Shabde and David Southall

- The first General Declaration on the Rights of the Child was in 1924. The United Nations Convention on the Rights of the Child was passed in 1989. Article 19 of the Convention states that legislative, administrative, social and educational measures should be taken to protect children from all forms of physical and mental violence, injury and abuse (including sexual abuse) and negligent treatment.
- ✔ **Child abuse is a worldwide problem and occurs in every country.**
- Child protection should be within the legal framework of the country; if not present, health professionals caring for children should lobby their government to pass and implement suitable laws.

Categories of child ill-treatment

Category 1:

- *Ill-treatment from universal human weakness*
  - Ranges from smacks to derogatory remarks.
  - Danger that introduction of child protective legislation against minor degrees of ill-treatment universal in a population might undermine support for strategies to develop effective frameworks which address severe abuse (see Category 3).
  - Best addressed through education, religious or other community initiatives.

Category 2:

- *Ill-treatment resulting from stress*
  - Involves excessive violence, physical or mental.
  - Parent is unhappy, depressed, dependent on drugs or alcohol, unsupported, may have been inadequately parented in their own childhood.
  - After ill-treatment parent is distressed; they do love their child.
  - Needs professional support not punitive legislation.
  - Appropriately led by Local Social Services Staff.

Category 3:

- *Abuse undertaken for gain*
  - Most serious involving great suffering.
  - Perpetrator usually suffering psychopathic personality disorder.
  - Immune/insensitive to suffering of others.
  - May enjoy inflicting emotional or physical pain.
  - Mental illness is not responsible for the abuse.
  - Aware that what they are doing is wrong.
  - Aim to avoid detection by elaborate and plausible lies.
  - Characteristically weave objects of truth into a latticework of deceit.
  - Perpetrators are dangerous and appropriately frighten local social workers, health visitors, doctors and teachers who need to be involved in a protected way.

Kinds of ill-treatment (Category 1)

- Scape-goating
- Neglect
- Gender bias

Kinds of ill-treatment (Category 2)

- Bruises from single, violent lashing out (usually one site)
- Neglect
- Some forms of child labour

Kinds of abuse (Category 3)

These include:

- Deliberate burns, for example cigarettes, scalding, holding against hot objects
- Multiple fractures, often at different times
- Ritual punishments (for example regular and savage beatings usually with implements)
- Deliberate starvation as distinct from neglect as in Category 2
- Fabrication/inducement of illness
- Sexual abuse/exploitation
- Female genital mutilation
- Severe neglect or killing of female children
- Use and indoctrination of children as soldiers
- Deliberately induced deformity to use children for begging

Most difficult issue is to separate this form of abuse from the stress-related child ill-treatment (Category 2).

The possibility of child ill-treatment/abuse must be considered in the differential diagnosis of all children who have suffered an injury. **All professionals working with** ✔

children need to be aware of the manifestations of child/ill-treatment abuse and do everything they can to protect children from harm. Some cultural practices are abusive. Child abuse/ill-treatment occurs across all social classes.

The possible features of parenting known to be associated with child ill-treatment/abuse include:

- Is the relationship between the parent(s) and the child loving and caring?
- Has one or both parents been abused themselves as children?
- Are the parent(s) young and/or unsupported?
- Is the parent single or substitutive?
- Has the parent learning difficulties?
- Do parents have a poor or unstable relationship?
- Is there domestic violence, drug or alcohol dependence?
- Is there mental illness, for example postnatal depression?

Possible factors in the child that make them vulnerable to abuse and ill treatment:

- Prematurity
- Separation and impaired bonding in the neonatal period
- Physical or mental handicap
- Behavioural problems
- Difficult temperament/personality
- Soiling and wetting past developmental age
- "Hyperactivity" and attention deficit
- Screamers (crying interminably and inconsolably)

Critical threshold for concern

Child abuse or ill-treatment can be defined as any act of commission or omission by an adult or institution responsible for the child which either directly or indirectly harms the child or damages their prospects of a safe and healthy development into adulthood.

Arriving at the critical threshold may be immediate and straightforward, for example finding of bruising on a small infant, or a direct disclosure of abuse from the child. In some circumstances the situation is less clear, for example if there are a number of non-specific signs or indicators or in cases of neglect. At some point a balanced assessment is required between the provision of family support for a child judged to be "in need" as opposed to action taken to directly protect the child.

The "critical threshold" is that point beyond which behaviour(s) towards a child can be considered to be ill-treatment or abuse and beyond which it becomes necessary to take action. That is when to raise concerns with the parents/carers and when to refer to the Statutory Agencies (Social Services or Police depending on the local legislative system).

1.31 Facilities for children with special needs and learning difficulties

David Cundall, Prudence Hamadé and Mohammed Arzomand

This chapter outlines the types of aids for disabled children which should be available in all hospitals.

✔ ● **The most valuable asset is staff who can spend time with the children and their families (preferably also able to visit them at home).**

● For children presenting at the hospital with established disabilities, the challenge is to ensure they make best use of their abilities and do not develop further disabilities.

Assessment kit:

For physical examination	For skills assessment
• Tape measure	• 20 brightly coloured wooden cubes (2.5 cm)
• Auriscope	• Threading beads of various sizes
• Ophthalmoscope	• 20 pictures (culturally appropriate) of common domestic objects (some of which have similar sounding names in the local language)
• Tendon hammer 128-Hz tuning fork	• Soft ball (approximately 10-cm diameter)
• Simple audiometer	• 'Denver Developmental Screening Test'

Aids for disabled children

There are many conditions which can cause disability and the ✔ **aim of the aids listed is to minimise disability and maximise independent function**. It should be noted that some positions which may appear desirable, for example the upright walking posture in a child with excessive extensor tone, may adversely affect the child's ultimate mobility. The advice of a trained paediatric physiotherapist is invaluable.

The book *Disabled Village Children* by David Werner is an excellent source of ideas and advice.

General principles

● "Look first at my strengths and not at my weaknesses": the preservation of best function is *primarily* achieved by education of the child and his carers, but aids and appliances can be very useful.

● Always consider the developmental stage of the child.

● An understanding of the home environment of the child is essential – a donkey may be a more useful mobility aid than a wheelchair once the child leaves hospital

● The prevention of secondary disabilities, for example contractures or pressure sores is a major priority in the care of disabled children.

● Always consider the purpose of the aid you think will help and ask yourself the following questions:
  - How will this aid help this child to function in his daily life?
  - Will the use of this aid reduce this child's abilities to do other things?
  - Will the use of this aid improve the way the child feels about him- or herself?
  - Who will review this aid to ensure that it is still helping the child and is still the right size for the growing child?
  - Who will maintain this aid to ensure that it still works?

Developmental aids

These are primarily used with children with delayed development but may also be useful for children who have suffered a neurological insult, whether or not they are showing signs of recovery. Most children function better if they can experience a variety of positions and can be part of activities with others.

Lying aids

Many children who are ill or recovering from illness spend most of their time lying on their backs or on their sides.

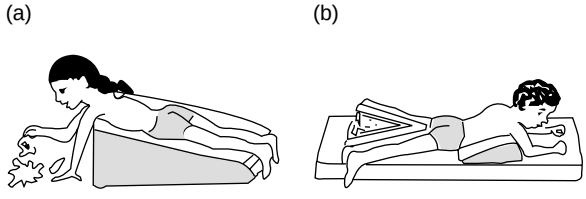

Figure 1.31.1 (a) Wedge, (b) leg separator

Lying on their front helps to develop trunk and arm strength and stretches muscles in the hips, knees and shoulders. A **pillow** under the chest helps to release the arms and hands for play.

A **wedge** is a more substantial version of the same idea and can be made from material like stiff foam plastic. Some children who need their legs separating because of adductor spasm will need a leg separator, also made of similar material.

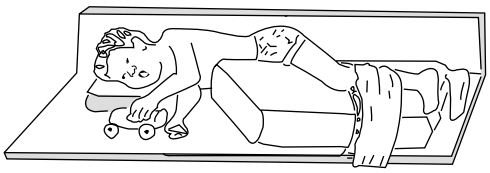

Figure 1.31.2 Side-lying frame

Some children with severe spasticity, usually due to cerebral palsy, benefit from a **side-lying frame** as this may be the position in which their arms are most functional.

Sitting aids

The type of sitting aids used will depend on the particular difficulties and developmental stage of the child. Most children with cerebral palsy benefit from being seated in a position where ankles, feet and hips are at 90° and where legs are kept apart (abducted). There are many varieties of seats available, for a young child a **corner seat** is often helpful. Special seating can also be fun! **(the "steam engine")**
Children with spasticity also often benefit from a slight tilt backwards, the position and amount of head support needed depends on the amount of head control and extensor tone.

Standing aids

These may be useful for children who are improving in their motor skills and can be expected to learn to stand independently but are also useful for children who may never do so because the standing position helps with circulation, bone growth and strength, particularly of the hip joints.

Walking aids

There are a wide variety of these aids, perhaps the most useful is a walking frame which goes behind the child and which can have a variety of attachments depending on the child's balance and strength of his arms. Some **parallel bars** are also useful and will need to be set at different heights depending on the size of the child.

A selection of **underarm crutches, elbow crutches, and tripod sticks** will be useful.

These can often be made locally and will need to be of various sizes.

Note that underarm crutches can cause nerve damage if the child hangs off the crutches when attempting to walk.

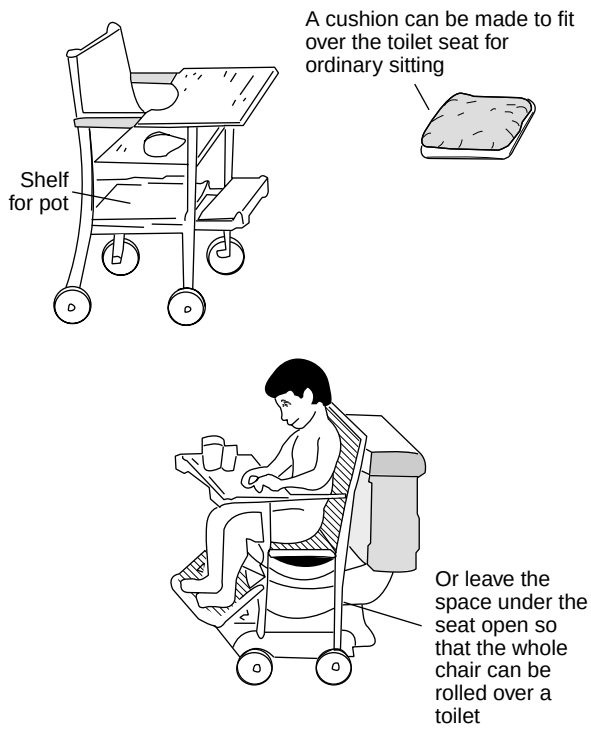

Figure 1.31.3 Toileting aids

Wheelchair technology

This is beyond the scope of this book. The general principles listed at the beginning of this section apply. Remember that a wheelchair is not the only solution for an otherwise immobile child. If the child has no sensation in his or her buttocks as a result of spinal cord damage he/she will be at risk of developing pressure sores if he/she remains seated in the same position for long periods of time. He/she can learn ways of taking the pressure off his/her buttocks. If pressure sores have developed, getting around the hospital may be better using a **gurney**.

Communication aids

Children who are unable to communicate verbally, either because of deafness or an inability to use their oromotor muscles, will often be able to use a **communication board** or book with pictures of objects, people and actions. If the child is unable to point using a finger, hand, toe or foot they may well be able to "eye point". An attentive carer will be aware that the child is eye pointing and the use of a communication aid may "unlock" the child who had previously been assumed to be unable to communicate beyond indicating pleasure or distress. More technological solutions are available using computers with specialised software which enables children to "speak" but the basic principle of being able to select a pictorial representation of an object or an idea is the same.

Aids to prevent common secondary problems developing in hospital

Preventing foot drop

One of the commonest preventable complications for children with weak legs is the development of foot drop. This should not happen in hospital. Regular exercises to move the ankles through their full range of movement should be done at least twice a day. It is best for the feet to rest with the ankles at 90°. This is easily ensured by positioning a roll of blanket or similar material bracing the feet in this position.

Preventing knee and hip contractures

Regular exercises taking the joints through as full a range of movement as possible are the mainstay of prevention. If possible, the child should spend some time each day lying on their front with hips and knees extended.

Preventing scoliosis

This is achieved by symmetrical positioning of the child so attention must be paid to both lying and sitting positions. With excessive and asymmetrical muscle tone it is often difficult to prevent scoliosis and once it has developed, it often gets worse, particularity at times of rapid growth.

Preventing pressure sores

Pressure sores develop anywhere in the body where skin is kept under pressure for too long. This commonly happens in areas where sensation has been lost and will happen more quickly if the circulation is poor. There is no substitude for good nutrition, regular moving and turning of the child but there are some aids which are helpful.

Further reading

Werner, D. *Disabled Village Children*. A guide for community health workers, rehabilitation workers and families. Berkeley, California: The Hesperian Foundation, 1999.

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Section 2

Presenting symptoms and clinical signs of illness and aspects of management

This section is a problem-oriented approach to clinical diagnosis, based on presenting symptoms, signs and laboratory tests. For more details, reference should be made to other sections of this manual.

How to use this book

This is a comprehensive text for all paediatricians caring for children in hospital. It can be used by those with limited resources and also where greater resources are available. We have identified the different levels of care in the following ways:

- **Minimum standards requirements** are given in a highlighted box at the the beginning of each clinical chapter.
- ***A standard of care*** where resources are not limited appears as bold, italicised text.
- **Key points** of particular importance in management of children are identified by a tick in the margin and bold text.

In this way we hope the book will act as a user-friendly, speedy reference on any paediatric ward.

2.1 Abdominal pain

David Southall

Important causes of acute and chronic abdominal pain

- Idiopathic — Irritable bowel syndrome; Migraine
- Psychogenic
- Gastrointestinal — Appendicitis; Peptic ulcer; Gastroenteritis; Intussusception; Oesophagitis; Inflammatory bowel disease; Constipation; Bowel obstruction; Food intolerance, for example milk protein, gluten; Meckel's diverticulum; Henoch-Schönlein purpura; Sickle cell disease
- Urinary tract — Infection, calculi, hydronephrosis
- Liver — Hepatitis
- Pancreas — Inflammation
- Malignancy — Lymphoma
- Gynaecological — Dysmenorrhoea; Pelvic inflammatory disease; Ovarian cyst; Pregnancy
- Respiratory — Pneumonia/pleurisy
- Trauma
- Poisoning — Lead

Table 2.1.1 Investigations

Investigation	Looking for
Full blood count	Anaemia, eosinophilia, infection
ESR/CRP	Inflammation
Urea, electrolytes	Renal disease
Amylase	Pancreatitis
Liver function tests	Liver dysfunction, hepatitis
Urine stick test – blood, protein, glucose	Infection, glomerulonephritis, diabetes
Microscopy – WBC, organisms, casts, culture	
Stool, ova, cysts, parasites, WBC and RBC	Infestation, dysentery, inflammatory bowel disease
Pregnancy test if appropriate	
Ultrasound – abdomen and pelvis, *X* ray – straight abdominal film	Bowel obstruction, constipation, lead poisoning, ovarian cyst, pregnancy and calculi
Barium studies and ***endoscopy***	Peptic ulcer, inflammatory bowel disease

Table 2.1.2 Differentiating organic from non-organic pain

	Organic	Non-organic
Nature of pain	Day and night	Periodic – good in between often periumbilical
History	Weight loss/reduced appetite Lack of energy Fever Change in bowel habit Urinary symptoms Intestinal symptoms Vomiting bile stained continuous blood Rectal bleeding	Migraine School and family problems Isolated vomiting – not bile stained
Examination	Ill appearance Weight loss Distension Absent /accentuated bowel sounds Shock Abdominal mass constipation other	Normal, thriving

2.2 Anaemia

Christiane Ronald

Anaemia, especially due to iron deficiency, is very common in poor communities. Anaemia can be due to a combination of inadequate nutrition and recurrent infections, for example malaria. Intestinal parasites such as hookworm are important causes. Genetic disorders such as sickle cell disease and thalassaemia should always be considered in relevant ethnic groups. Acute worsening of anaemia may present as heart failure in young children.

- If <6 years normal Hb >9.3 g/dl (haematocit >27%)
- Moderate anaemia = 6 to 9.3 g/dl
- Severe anaemia = <6 g/dl, severe pallor (palmar/conjunctival), heart failure (gallop rhythm, enlarged liver and pulmonary oedema (fine basal creps))

Causes of anaemia

Decreased production

- Prematurity: 6–8 weeks
- Hypochromic: Iron deficiency (diet, blood loss, chronic inflammation)
- Normochromic: Chronic infection or inflammation
  Nutritional: malnutrition, scurvy
  Infiltration: for example leukaemia, malignancy
  Metabolic: renal and liver disease
- Megaloblastic: Folic acid deficiency; infection, coeliac disease, anticonvulsants, haemolysis; Vitamin B_{12} deficiency: intestinal resections, Crohn's disease, vegan diet
- Hypoplastic: Sickle cell crises, drugs (for example chloramphenicol), malignancy

Increased haemolysis

- Haemoglobinopathies: Sickle cell disease, thalassaemia major
- Non-immune: Drugs, infection, hypersplenism, burns, haemolytic uraemic syndrome, DIC, porphyria, venoms
- Enzyme deficiency: Drug-induced and spontaneous G6PD deficiency, glutathione synthetase deficiency, pyruvate kinase deficiency
- Immune: Rhesus and ABO incompatibility, autoimmune (for example reticuloses), *Mycoplasma* infection, SLE, drugs
- Membrane defects: spherocytosis, elliptocytosis, stomatocytosis, erythropoietic porphyria, abetalipoproteinaemia

Blood loss

- Perinatal: Placental and cord accidents
  Fetomaternal, twin-to-twin transfusions
  Birth injury for example cephalhaematoma, subaponeurotic haemorrhage, severe bruising
  Haemorrhagic disease of the newborn
- Epistaxis
- Trauma
- Alimentary tract: Haematemesis, rectal bleeding, hookworm
- Blood clotting disorders: For example haemophilia, thrombocytopenia
- Renal tract: Haematuria

History

- Symptoms of anaemia: lethargy, tiredness, shortness of breath on exertion, poor growth
- Obvious blood loss: epistaxis, haematemesis, haematuria, blood in stools
- Assess the diet, for example inadequate weaning diet
- Steatorrhoea
- Chronic infection, inflammation
- Drugs: especially antibiotics, antimalarials, anticonvulsants, analgesics, cytotoxic agents

Examination

- Chart growth/nutritional state
- Conjunctivae, nails and palms for pallor
- Stomatitis

- Jaundice
- Bruising, lymphadenopathy, or petechiae
- Hepatosplenomegaly
- Tachycardia, flow murmur, cardiac failure

Table 2.2.1 Investigations

Investigation	Looking for
Full blood count	Hb, white cells, platelets
Blood film	Red cell morphology, malaria, target cells, haemolysis
Hb electrophoresis	Sickle cell disease and thalassaemia
MCV, reticulocytes	Iron deficiency, haemolysis
Coombs test	Haemolysis
Bone marrow	Leukaemia, malignant infiltration, aplasia
Bilirubin, ***Liver function tests***	Direct/indirect bilirubin
Urinalysis	Red cells, casts, bacteria, WBC, protein, culture
Serum ferritin	Iron stores
Barium meal/***endoscopy***	Inflammatory bowel disease
Platelets and clotting	Coagulation disorder
Stool microscopy, culture and occult blood	Hookworm (egg count), occult blood loss

2.3 Diarrhoea

Brian Coulter

Causes

There are a number of causes of diarrhoea. The management of acute, persistent and chronic diarrhoea is covered in Chapters 3.25 and 3.26.

Common causes of diarrhoea

- **Infection**
  - Acute (<14 days) — Viruses, bacteria, parasites
  - Persistent (>14 days)
- **Secondary**
  - Malnutrition
  - HIV
  - Disaccharide intolerance
  - Malaria
- **Chronic** (non-infectious)
  - Food intolerance — Milk protein, soy protein; Coeliac disease; Multiple food intolerance
  - Inflammation — Crohn's disease; Ulcerative colitis
  - Pancreatic disease — Cystic fibrosis, Schwachman syndrome (cyclic neutropenia)
- **Miscellaneous** — Non-specific "toddlers" diarrhoea; Irritable bowel; Excessive squash/fruit drinks

History

- Duration
- Nature of stool, for example fatty/floats/watery? with blood
- Number per day
- Dietary intake
- Foreign travel, food poisoning

Examination

- Chart growth/nutritional status
- Document degree of dehydration
- Look for fever, anaemia, lymphadenopathy, hepatosplenomegaly, finger clubbing
- Look for signs of vitamin or mineral deficiency, oral ulcers and anal fissures
- Look for candidiasis

Table 2.3.1 Investigations

Investigations	Looking for
Stool	Infection
Microscopy (warm stool for *E. histolytica*)	
WBC, RBC ova, parasites	
Culture	
PH (<5.5)	Lactose intolerance
Clinitest tablets or Benedict's solution	
Fat globules	Pancreatic disease
Breath hydrogen test	
Blood	
Culture (high temperature, convulsions)	Septicaemia, for example *Salmonella*
Urea, creatinine, electrolytes (if oliguria)	Haemolytic uraemic syndrome
FBC	Hypo/hypernatraemia
Albumin	Chronic diarrhoea
X ray abdomen, ultrasound	Ileus, perforation
Urine microscopy	Haemolytic uraemic syndrome

2.4 Jaundice

David Southall

Causes

- Depending on age (for neonatal jaundice see Chapters 2.16 and 3.48)
- Excess haemolysis (prehepatic)
  - Sickle cell disease
  - Thalassaemia
  - Hereditary spherocytosis
  - Malaria
- Liver disease (see Chapters 3.9, 3.10, and 4.16)
  - Hepatocellular
  - Obstruction to bile secretion
    Infective hepatitis
    Acute liver failure
    Chronic liver disease

History

- Family history of hereditary haemoglobinopathy or liver disorder
- Blood transfusion?
- Anorexia
- Abdominal pain
- Pruritus
- Colour, nature and contents of stools and urine

Examination

- Assess growth/nutritional state
- Look for skin signs of chronic liver disease, for example spider naevi, clubbing, leukonychia, liver palms, scratches from pruritus
- Assess liver and spleen – enlarged? tender?
- Anaemia?
- Ascites?
- Look for frontal bossing or maxillary overgrowth (sickle cell disease or thalassaemia)
- Look at colour of stool and urine

Table 2.4.1 Investigations

Investigation	Looking for
Full blood count and film	Anaemia
Reticulocytes	Haemolysis
Haemoglobin electrophoresis	Sickle cell disease and thalassaemia
Urine	Bilirubin and urobilinogen
Liver function tests:	Bilirubin conjugated (liver disease or biliary obstruction) or unconjugated (haemolysis)
Transaminases	Hepatitis
Serology	Identification of viral causes
Coagulation	Liver failure
Autoantibodies	Chronic active hepatitis

2.5 Lymphadenopathy

Brian Coulter

Common causes of generalised lymphadenopathy

- HIV infection
- Infectious mononucleosis
- Tuberculosis
- Leukaemia
- Hodgkin's and non-Hodgkin's lymphoma
- CMV, toxoplasmosis
- African trypanosomiasis

Infective causes of local lymphadenopathy

- Local skin (especially scalp) infections
- TB
- Environmental mycobacteria
- Cat scratch disease

History

- Known epidemiology for HIV and trypanosomiasis in the area
- Contact with TB
- Chronic ill health (malignancy, HIV or TB)
- Nodes static or increasing in size?

Examination

- Chart growth and nutritional status
- Fever?
- Liver or spleen enlargement?
- Purpura or anaemia?
- *Candida?*

Table 2.5.1 Investigations

Investigations	Looking for
Full blood count	Atypical lymphocytes, leukaemic picture
Thick blood film	Trypanosomiasis
Bone marrow	Malignancy
HIV antibodies	HIV
Paul-Bunnell	Infectious mononucleosis (+ve 60%)
ESR/CRP	Infection, TB
Mantoux test	TB, environmental mycobacteria
Serology	EB virus, CMV, toxoplasmosis
Chest *X* ray	TB, malignancy
Lymphnode biopsy	Diagnosis

2.6 Fits, faints and apparent life threatening events (ALTE)

David Southall

Common causes

- Febrile convulsions
- Epileptic seizures
- Hypoglycaemia
- Infantile apnoea/hypoxaemic events:
  - Premature birth
  - Respiratory infection, for example bronchiolitis, pertussis
- Sleep-related upper airway obstruction (see Chapter 3.19)
- Vasovagal episodes (simple faints)
- Cardiac arrhythmias
- Cyanotic breath-holding
- White breath-holding (reflex anoxic seizures)

History

- Cyanosed
  - Occurs with infant apnoea
  - Some febrile convulsions/epileptic seizures
- Extreme pallor
  - Vasovagal
  - Cardiac arrhythmia
- Trauma related (especially to head) = white breath-holding
- Emotional upset/anger/temper = cyanotic breath-holding
- Snoring/inspiratory stridor during sleep often with chest recession and restlessness = sleep related upper airway obstruction
- Drug abuse
- Fabricated or induced illness (see Chapter 5.2)

Examination

- Growth and nutritional status
- Respiratory infection?
- Anaemia (associated with cyanotic breath-holding and infant apnoea)
- Fever?
- Neurological examination (to exclude coma or epilepsy); see Chapters 3.36 and 3.37
- Cardiac disorder?
- Blood pressure lying and standing for vasovagal episodes
- Mouth, throat for enlarged tonsils or retrograde/small mandible for predisposition to sleep-related airway obstruction (the latter is also common in sickle cell disease and Down's syndrome.

Table 2.6.1 Investigations

Investigation	Looking for
Full blood count	Anaemia, infection
Blood glucose	Hypoglycaemia
Hb electrophoresis	Sickle cell disease
ECG	Wolf-Parkinson-White and long QT syndromes Structural lesion of heart
Oxygen saturation during sleep	Low baseline SaO_2 predisposes to infant apnoea/hypoxaemic events Should be $>94\%$ (at sea level, see Chapter 3.19) Especially in preterm infants and infants <6 months with respiratory infection
Video (if available) during sleep	Sleep related upper airway obstruction
EEG	Epileptic cause
Chest *X* ray	Lung disease in infantile apnoea/hypoxaemic events

2.7 Generalised oedema

Brian Coulter

The major differential diagnosis relates to the presence or absence of hypoalbuminaemia.

Common pathophysiology

- Heart failure
  - Jugular vein pressure ↑, liver enlarged, triple rhythm, murmurs
  - Cardiovascular disorders
  - Anaemia
- Acute glomerulonephritis
- Low serum albumin
  - Nephrotic syndrome
  - Liver disorders
  - Protein-losing enteropathy, for example malabsorption, intestinal lymphangiectasis
  - Malnutrition
- Increased vascular permeability
  - Anaphylaxis (history)
  - Shock
- Over-hydration (particularly excessive IV solutions such as 5% glucose)

History

- Short of breath, chest pain (pericarditis)
- Blood in urine (nephritis)
- Facial swelling (nephrotic syndrome or acute glomerulonephritis)
- Nutritional history
- Gastrointestinal symptoms
- Exposure to allergen or sting (anaphylaxis)
- Excess IV fluids

Examination

- Chart growth and nutritional status, look for features of kwashiorkor and vitamin deficiencies.
- Cardiovascular system including blood pressure.
- Jaundice or other signs of liver disease.
- Anaemia, lymphadenopathy?
- Enlarged liver and/or spleen.
- Ascites (especially nephrotic syndrome). Ascites may be transudate, (for example nephrotic syndrome) or inflammatory (for example TB, peritonitis). Abdominal malignancy may cause ascites and obstructive oedema of lower limbs.

Table 2.7.1 Investigations

Investigation	Looking for
Full blood count	Anaemia
Urinalysis	
Dipstix: protein, blood	Nephrotic syndrome, nephritis
Bilirubin	Liver disease
Microscopy: RBC, casts	Nephritis
Stool	Hookworm
Serum albumin	Low albumin
Imaging	
Abdominal ultrasound	Hepatosplenomegaly Malignancy Ascites – transudate/ inflammation
Echocardiogram	Cardiac disorders
Ascitic fluid	
Colour: clear, cloudy, bloody, chylous	Inflammation, for example TB
Cells: WBC, malignant cells	Malignancy
Ziehl–Neelsen stain	
Protein:	< 25 g/litre transudate > 25 g/litre exudate
Culture: TB/general	

2.8 Rash

Brian Coulter

Causes

- Macular — Viral for example exanthema, sometimes meningococcal infection
  - discrete — Juvenile chronic arthritis
  - morbilliform — Kawasaki disease
  - erythema multiforme
  - erythema marginatum — Rheumatic fever
- Papular — Chickenpox
  - vesicles — H. simplex
  - pustules — Impetigo
  - bullae — Scabies
- Purpuric — Meningococcal disease
  - petechial — Henoch-Schönlein purpura
  - ecchymosis — Thrombocytopenia
- Desquamation with or without mucosal involvement — Scalded skin syndrome; Stevens–Johnson syndrome; Toxic epidermonecrolysis; Post-Kawasaki; Post-scarlet fever; Post-toxic shock syndrome; Epidermolysis bullosa
- Erythema nodosum — Streptococcal disease; Tuberculosis; Connective tissue disorders; Sarcoidosis; Drugs

Table 2.8.1 Investigations

Investigation	Looking for
Full blood count ESR, CRP	Systemic bacterial infection, for example meningococcal disease
Blood culture	Kawasaki disease Thrombocytopenia
Skin swab	Bacterial infection
Skin scraping	Scabies
Throat swab and ASOT	Streptococcal infection
Urinalysis RBC, casts Protein	Nephritis, for example Henoch-Schönlein purpura, connective tissue disorders
Skin biopsy	Epidermolysis bullosa
Autoantibodies	Connective tissue disorder

2.9 Respiratory distress

Alice Leahy

Presenting features

- Tachypnoea
- Increased work of breathing: tracheal tug, inter/subcostal recession
- Poor feeding, sleep disturbance
- Grunting
- Unable to speak in sentences
- Positioning: sitting up/forward, neck extension, splinting chest
- Tachycardia
- Altered mental state: agitation (hypoxaemia)/drowsiness (hypercapnia)
- Pallor/cyanosis (late sign)

Table 2.9.1 Causes

Common cause	On examination
Upper airway obstuction	Stridor, hoarse voice, drooling, sitting up, head held forward
Inhaled foreign body	Suggestive history, tracheal deviation, unilateral hyperexpansion on chest *X* ray
Asthma	Hyperexpansion, wheeze, reduced air entry, reduced peak flow, hypoxaemic ($SaO_2 < 94\%$*)
Bronchiolitis	Inspiratory crackles, wheeze, hypoxaemic ($SaO_2 < 94\%$*)
Pneumonia	Fever, grunting, pleuritic or abdominal pain, signs of consolidation or effusion. Clubbing indicates chronic disease, for example bronchiectasis
Tuberculosis	Contact history lymphadenopathy, fever, weight loss
Pneumothorax	Unilateral hyper-resonance on percussion, tracheal deviation, apex displacement
Heart failure/ pulmonary oedema	Sweaty, gallop rhythm, hepatomegaly, murmurs
Sickle cell disease chest syndrome	Hypoxaemia ($SaO_2 < 94\%$*), chest pain

* at sea level, see Chapter 3.19.

Table 2.9.2 Investigations

Investigation	Looking for
Oxygen saturation (pulse oximeter)	Hypoxaemia ($< 94\%$ SaO_2)*
Chest *X* ray	Lung disorder
ECG, ***echocardiogram***	Cardiac disorder
Mantoux	TB
ESR, CRP	Inflammation
Full blood count	Infection
Hb electrophoresis	Sickle cell disease
Bronchoscopy	Foreign body

* at sea level only.

210 Vomiting

Brian Coulter

History of acute, recurrent or chronic nature of this symptom indicates approach to the diagnosis.

Common causes (depending on age)

Infants

- Gastroenteritis
- Gastro-oesophageal reflux (distinguish from possetting)
- Overfeeding
- Bowel obstruction
  - pyloric stenosis
  - intussusception
  - congenital gut anomalies
- Infection – especially urinary tract
  - meningitis
  - otitis media
  - pertussis
- Poisoning

Childhood

- Gastroenteritis
- Appendicitis (with pain)
- Infection – especially urinary tract
  - meningitis (including TB)
  - malaria
- Bowel obstruction
- Ingestion of drugs or poisons
- Migraine
- Pregnancy
- Bulimia (but rarely does child admit this)
- Raised intracranial pressure (RICP)
- Hypertension
- Diabetic ketoacidosis

History

- Accidental drug ingestion?
- Is it vomiting or regurgitation or possetting (especially in an infant)?
- Is it associated with coughing? whoop?
- Is it projectile?
- Does it contain blood or bile?
- Any diarrhoea or constipation?
- Is there abdominal pain?
- Is there a family history of migraine?
- Are there difficulties in coordination during physical activity?

Examination

- Does the child look ill?
- Assess growth and nutritional status
- Examine vomit
  - bile stained suggests bowel obstruction
  - blood (coffee ground)?
- Fever?
- Full examination (include blood pressure, funduscopy and anorectal examination as indicated)
- Dehydration?
- Abdomen
  - test feed for pyloric stenosis: swelling or visible peristalsis
  - tenderness or mass
  - are bowel sounds present? what are they like?

Table 2.10.1 Investigations

Investigation	Looking for
Urine microscopy	Urinary tract infection
Full blood count	Infection
Thick film	Malaria
Urea and electrolytes	Prerenal or renal failure, pyloric stenosis
Blood culture	Infection
Lumbar puncture	Meningitis
Stool microscopy and culture	Ova, cysts, parasites, bacteria and viruses
Liver function tests	Hepatitis
Abdominal ultrasound	Masses, obstruction, free fluid
Straight abdominal *X* ray/chest *X* ray	Bowel obstruction, free air
Barium studies ***and/or endoscopy***	Specific diagnosis
Pregnancy test	Pregnancy
Mantoux test	TB, meningitis
Brain imaging	Raised intracranial pressure

211 Wasting/failure to thrive

Brian Coulter

Table 2.11.1 Causes of failure to thrive

Mechanism	Systems involved
Inadequate intake	Anorexia Breastfeeding failure Feeding mismanagement * Swallowing disorders
Loss	* Vomiting * Diarrhoea * Malabsorption
Structural dysfunction of organs	Brain – cerebral palsy, learning difficulties Respiratory Cardiac Urinary tract * GIT
Increased requirement for nutrients or metabolites	Infection Connective tissue disorders Immune disorders
Failure of end-organ response	Metabolic for example amino acid and organic acid disorders Endocrine, for example thyroid disorder Malignancy Chromosomal abnormalities
Emotional/ psychological	Parent *neglect abuse family dysfunction Child *feeding/behaviour disorders anorexia nervosa bulimia

* Common causes.

Failure to thrive: gastrointestinal disorders

Oropharynx	Cleft palate
Oesophagus	Incoordination of swallowing, for example cerebral palsy
Stomach	Gastro-oesophageal reflux Pyloric stenosis
Digestion	Pancreas – cystic fibrosis Liver – cirrhosis
Small gut disorders	Milk protein intolerance Coeliac disease Carbohydrate malabsorption Protein-losing enteropathy Short gut syndrome Crohn's disease
Large gut disorders	Ulcerative colitis Crohn's disease Hirschsprung's disease

Approach to failure to thrive

- Failure to thrive is due to inadequate delivery of nutrients to developing tissues.
- Usually manifest by failure to gain weight as expected. In extreme circumstances height (length) and head circumference may be affected. Plot mid-parental height.
- Majority of cases are related to gastrointestinal disorders: poor intake/malabsorption.
- Observe feeding, mother's interaction, child's behaviour, vomiting, diarrhoea and weight gain before embarking on investigations.
- Investigations: when likely system/disorder is identified. See relevant chapters on gastroenterology (Section 3).

2.12 Headaches

Allie Moosa

- Headaches are common in children.
- Should be taken seriously if they persist.
- Prevalence increases with age.

Acute headache

Common causes of acute headache include:
- Febrile illness.
- Meningitis/encephalitis.
- Acute sinusitis: pain and tenderness (elicited by gentle percussion) over the maxilla; there is usually a history of preceding upper respiratory tract infection and a postnasal discharge may be present.
- Head injury.
- Raised intracranial pressure.
- Intracranial haemorrhage (severe, sudden headache, with rapid loss of consciousness).

A careful history and physical examination will usually reveal the cause.

Raised intracranial pressure (RICP)

- Headache may be sudden or gradual in onset, often occipital in location and becomes progressively more severe.
- Made worse by lying down (compared to migraine and tension headache which are relieved by lying down), by coughing, stooping and straining and may wake the child from sleep.
- Worse in the morning and often associated with nausea and vomiting.
- Other signs of raised intracranial pressure may be present such as impaired consciousness, bilateral abducens nerve palsies (false localising sign) and, when severe, bradycardia and hypertension.
- Papilloedema is a late sign.
- Localising neurological signs may be present depending on the site of the lesion: ataxia indicates a posterior fossa tumour; cranial nerve palsies a brainstem lesion; visual field defect a craniopharyngioma; and unequal pupils a suptratentorial lesion such as subdural haematoma.
- In endemic areas, cerebral malaria and neurocysticercosis are important causes.

Benign intracranial hypertension

- Raised intracranial hypertension without any space occupying lesion or obstruction of the CSF.
- Can be caused by drugs (corticosteroids especially during withdrawal, ampicillin, nalidixic acid) and sagittal sinus thrombosis.
- Most without cause, especially in young adolescent girls.

Recurrent or chronic headaches

Two common causes are anxiety (tension) and migraine.

Tension headaches

- Affect about 10% of school children.
- Typically headache is symmetrical and described as hurting or aching over the cranial vault.
- Develop gradually and are not associated with other symptoms.
- Induced by stress (school examinations, assignments, etc.) and can co-exist with migraine in the same child.
- May be caused by isometric contraction of head and neck muscles in anxious children.

Migraine: see Chapter 3.43.

Conversion (hysterical) headache

- Headache can be a conversion symptom used by the child to gain attention.
- Initial headache may have been due to an organic cause (for example febrile illness) but its persistence and recurrence is due to psychological factors.

Management see also section on coma for acute onset of headache (Chapter 3.36)

- Detailed history and careful examination to rule out serious underlying causes.
- Investigations are rarely needed.
- *X* ray of the sinuses will confirm sinusitis and CSF examination meningitis/encephalitis.

✔
- **A CT scan of brain is essential if raised intracranial pressure is suspected or if there are localising neurological signs**.
- Treatment is directed at the underlying cause and pain relief.
- Benign intracranial hypertension can be controlled with corticosteroids (dexamethasone 0.6 mg/kg/day in two divided doses) and/or acetazolamide (8 mg/kg/ 8 hourly increasing to a maximum of 32 mg/kg/day) and repeated lumbar puncture.

For tension and conversion headaches, counselling and stress management is important.

Relief of pain

For most headaches, simple analgesics alone or together with non-steroidal anti-inflammatory agents will suffice, for example paracetamol with or without ibuprofen.

213 Limp, joint pains and swelling

Brian Coulter

Causes

The differential diagnosis is broad.

Common causes of limp, joint pains and swelling

Acute

- Injury/snake bite
- Arthritis
- Rheumatic fever
- Osteomyelitis
- Fracture
- Tumours
- Leukaemia
- Poliomyelitis
- Sickle cell disease
- Haemophilia
- Spinal disease
- Osteochondroses, for example Perthes' disease
- Slipped femoral epiphysis
- Rickets
- Henoch–Schönlein purpura
- Meningism

Chronic

- Juvenile rheumatoid arthritis
- Cerebral palsy
- Congenital malformation

History

- Duration of symptoms
- Family history of connective tissue disorder
- Known sickle cell disease

Examination

- Full general examination
- Chart growth and nutritional status (see Chapter 6.1)
- Signs of acute infection, for example septic arthritis or osteomyelitis
- Evidence of vitamin D deficiency? (Chapter 3.15)
- Anaemia?
- Fever?
- Rash?

Table 2.13.1 Investigations

Investigation	Looking for
Full blood count	Infection, anaemia, thrombocytopenia
ESR, CRP	Inflammation
Blood culture	
Bone scan	Osteomyelitis
Sickle cell test	Sickle cell disease
Haemoglobin electrophoresis	
ASOT	Rheumatic fever
ECG	
Rheumatoid factor	Chronic juvenile arthritis
Antinuclear factor	Connective tissue disorders
Double-stranded DNA	
Mantoux test, Chest *X* ray	Tuberculosis
X ray Osteomyelitis	
Ultrasound	Osteochondrosis Chronic arthritis
Aspiration/biopsy	Infection Connective tissue disorder

214 Fever of unknown origin

Alice Leahy and Sheila Silva

Definition

Minimum temperature of at least 38.3°C for 1–3 weeks with at least 1 week of hospital investigation. It is very important to determine whether fever is continuous or recurrent by plotting it on a chart (see Appendix 7.13).

Baseline investigations

- Full blood count and film
- ESR/CRP
- Blood cultures
- Thick film for malaria (endemic areas/recent foreign travel)
- Mantoux test
- ***Epstein–Barr and other viral serology***
- Urine microscopy/culture
- Chest *X* ray
- Lumbar puncture (if meningeal signs present)

Table 2.14.1 Causes of fever of unknown origin

	Causes	Specific investigations
Bacterial infection	Tuberculosis	Chest *X* ray, Mantoux test, lumbar puncture
	Campylobacter	***Serology***/culture
	Legionellosis	***Serology***/culture
	Typhoid	Culture (blood/stool)
	Brucellosis	***Serology***
	Cat scratch disease	Lymph node biopsy
	Rheumatic fever	Throat swab, ASOT
Localised infection	Abscess	Abdominal ultrasound
	Endocarditis	Echocardiogram
	Osteomyelitis	*X* ray, ***bone scan***
	Pyelonephritis	Urinalysis
	Cholangitis	Abdominal ultrasound
Spirochaete infection	Lyme disease	***Serology***
	Syphilis	***Serology***
	Leptospirosis	Urine culture, serology
Viral infection	Cytomegalovirus	***Serology***
	Epstein–Barr virus	***Serology***
	HIV	Serology
Chlamydial infection	Psittacosis	***Serology***
Rickettsial infection	Q fever	***Serology***
Fungal infection	Histoplasmosis	Culture/***serology***
Parasitic infection	Giardiasis	Stool microscopy
	Malaria	Thick/thin blood film
	Trypanosomiasis	Thick blood film
	Toxoplasmosis	***Serology***
	Toxocariasis	***Serology***, eosinophil count
	Leishmaniasis	Bone marrow, ***serology***
Connective tissue disorders	Juvenile chronic arthritis	***Autoantibodies***
	Systemic lupus erythematosus	***Autoantibodies***
Neoplasia	Lymphoma	Node biopsy
	Leukaemia	Blood film/bone marrow
	Neuroblastoma	***Urinary VMA***
	Wilms' tumour	Ultrasound or ***CT scan***
Miscellaneous	Kawasaki disease	ESR, platelets, clinical findings
	Imflammatory bowel disease	Barium studies/***endoscopy***
	Chronic active hepatitis	Liver biopsy
	Thyrotoxicosis	Thyroid function tests
	Factitious fever	Surveillance

215 Cough, wheeze and stridor

Felix Sanchez

Major differential diagnoses include acute causes such as pertussis, chronic conditions such as bronchiectasis and recurrent conditions such as asthma.

Table 2.15.1 Causes of upper respiratory system disorders

Disease	Symptoms	Investigations
Acute causes		
Common cold	Time of the year, acute onset, other relatives affected, variable fever, nasal dribbling.	
Viral pharyngitis (most common cause)	Rhinitis, conjunctivitis, diarrhoea, morbiliform rash.	
B-haemolytic streptococci, Group A pharyngotonsillitis (15% of pharyngitis)	Fever, nausea, vomiting. Tender lymphadenopathy. Congested uvula. Yellow exudates. Impetigo in nares, scarlet fever-like rash, petechiae on palate. Cough infrequent.	Throat swab ***ASOT***
Acute sinusitis	Flu-like syndrome that does not improve, persistent fever, purulent nasal discharge or headache and facial pain.	Nasal secretions: leucocytes, mainly polymorphs. Sinus *X* ray: opaque sinuses, fluid levels and mucosal thickening.
Chronic causes		
Allergic rhinitis	Positive family history, dermatitis, personal history of other allergic disorders, nasal pruritus, sneezes, non-purulent rhinorrhoea, postnasal dribbling.	Nasal secretions: contain eosinophils.
Vasomotor rhinitis	No family history of allergy. Predisposing factors like environment or irritants, cold, damp.	Nasal secretions: no eosinophils.
Chronic sinusitis	Purulent nasal discharge, nasal obstruction, headache, bad breath, facial pain.	*X* ray of paranasal sinuses: opaque sinuses, fluid levels and mucosal thickening.
Adenoid hypertrophy	Snoring, open mouth breathing.	*X* ray: enlarged adenoid tissue with narrowing of air column.

Table 2.15.2 Causes of lower respiratory system disorders

Disease	Other features	Investigations
Acute causes		
Laryngotracheobronchitis (croup) Agent: Parainfluenza virus	Between 6 months and 3 years. Epidemic peak. Initial coryza, barking cough, stridor, breathing difficulty, fever, not septic looking.	
Epiglottitis Agent: *Haemophilus influenzae*	Stridor, drooling, hypoventilation with hypoxaemia and hypercapnia. "Septic"	Examination under anaesthetic with CPAP and subsequent intubation or tracheostomy.
Bacterial tracheitis Agent: *Staphylococcus aureus*	Barking cough, stridor, breathing difficulty, high fever, very septic looking with rapid deterioration.	
Foreign body aspiration	Previously well, choking episode, sudden onset of breathing difficulties.	AP and lateral chest *X* ray Air trapping or lobar/segmental atelectasis. Bronchoscopy under general anaesthesia.
Bronchiolitis	Variable respiratory distress, tachypnoea, wheeze, crepitations, hyperexpanded chest. Rainy season. Variable fever.	AP chest *X* ray: bilateral air trapping.
Pneumonia	Worsening respiratory distress, intercostal/subcostal recession, crepitations or wheezes, consolidation, fever, pleuritic pain, purulent sputum with or without blood. Cough may be absent.	AP chest *X* ray: alveolar infiltrates. Shadows progressing to consolidation, segmental or lobar, indicates bacterial pneumonia. Air trapping, interstitial pattern or atelectasis suggests viral pneumonia.
Chronic causes		
Bronchial asthma	Family history of asthma or allergy, personal history of allergic illnesses (rhinitis, dermatitis), wheeze, exercise induced and/or nocturnal cough, improved by bronchodilators.	AP chest *X* ray: bilateral air trapping. Bronchodilator challenge: improvement of cough and other signs and symptoms.
Pulmonary tuberculosis	Contact with TB, long-term fever, weight loss, asthenia	Mantoux test AP and lateral chest *X* ray: pneumonia or atelactasis of middle lobe, perihilar lymph nodes, diffuse alveolo-interstitial infiltrate, micronodular pattern. Cough swab or gastric aspirate: Ziehl–Neelsen stain and culture for acid–alcohol-resistant bacilli (see Chapter 4.10)
Pulmonary malformations	Recurrent pneumonia in first few months of life.	AP chest *X* ray: persistent opacity in same place at different dates, not clearing after weeks; presence of cysts or persistent atelectasis.
Irritant exposure	Cigarette/wood smoke, chemicals	

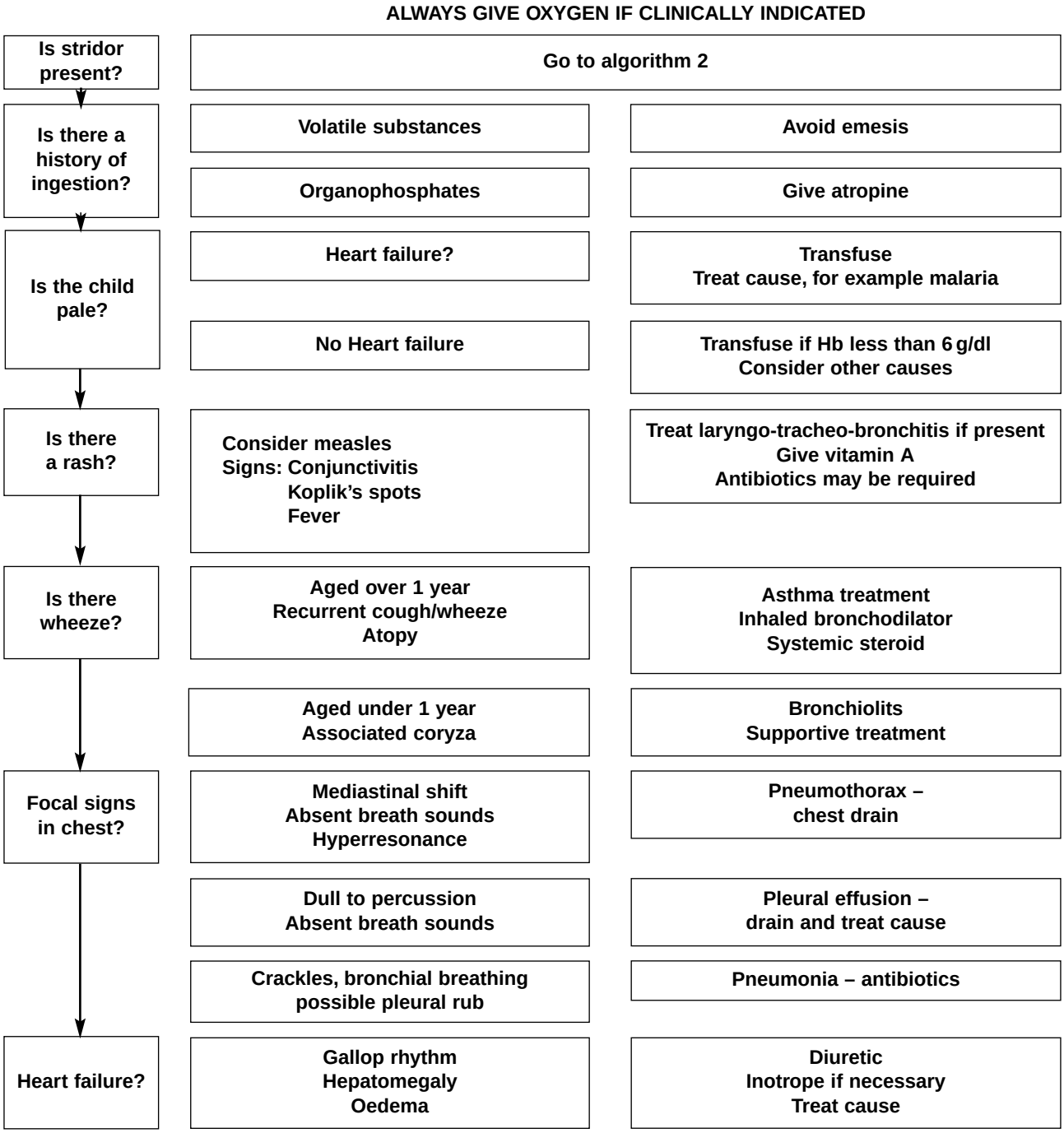
Table 2.15.3 Causes of specific infections

Disease	Other features	Investigations
Chlamydia trachomatis	Maternal genital infection, neonatal conjunctivitis, apyrexial.	FBC: eosinophilia. Chest *X* ray: diffuse interstitial pattern
Bordetella pertussis	Paroxysmal cough with stridor. Apnoea in infants. Sub-conjunctival haemorrhages. Well in between coughing episodes. Not immunised	FBC: leucocytosis, mainly lymphocytes (30×90^9/litre)
Adenovirus	Pharyngitis, conjunctivitis, rash, haemorrhagic cystitis	Chest *X* ray: interstitial pattern
Respiratory syncytial virus (RSV)	More severe in preterm or with congenital heart lesion. Family contact. Prolonged expiratory phase, crackles and wheeze	Chest *X* ray: diffuse interstitial pattern, hyperinflated
Loeffler's syndrome	Family and personal history of allergy	Stool: *Ascaris lumbricoides, Trichiuris, Toxocara,* uncinarias, hookworm, filariasis. Blood: eosinophilia Chest *X* ray: peripheral alveolo-interstial infiltrates
Bronchiectasis	Past history of severe pneumonia TB, pertussis, measles Bad breath, profuse morning sputum, clubbing	AP chest *X* ray: bronchial dilation, gross interstitial pattern, localised shadowing, honeycomb pattern
Aspiration	Cough and choking on feeding, vomiting, excessive salivation, neurological disorders	Barium swallow. Chest *X* ray ***Oesophageal pH studies***
Immunodeficiency	Failure to thrive, recurrent infections, TB, diarrhoea, pyodermitis	FBC: leucopenia, lymphopenia, neutropenia Sputum culture: opportunistic infections
Airway compression Vascular rings Mediastinal masses	Onset in first months of life. Murmurs	Barium meal: indentations or deviations in barium column AP and lateral chest *X* ray: cardiomegaly or presence of masses
Cystic fibrosis	Family history of chronic chest infections. Meconium ileus, sinusitis, steatorrhoea, failure to thrive, clubbing	***Sweat test***
Psychogenic cough	Teenager. "Noisy" cough. Stops during sleep	

2.16 Diagnostic algorithms

Simon Parke and Jasmine Heslop

The following algorithms are intended to be guidelines for the recognition of common conditions, based on their mode of presentation. The diagnostic lists are not intended to be exhaustive but rather a quick check list to enable the identification and therefore management of common emergencies in paediatrics.



Note
If none of these present the patient requires investigation, with full blood count and chest *X* ray at least. Empirical antibiotic therapy should be considered. Always give oxygen unless mild.

Figure 2.16.1 Difficult breathing

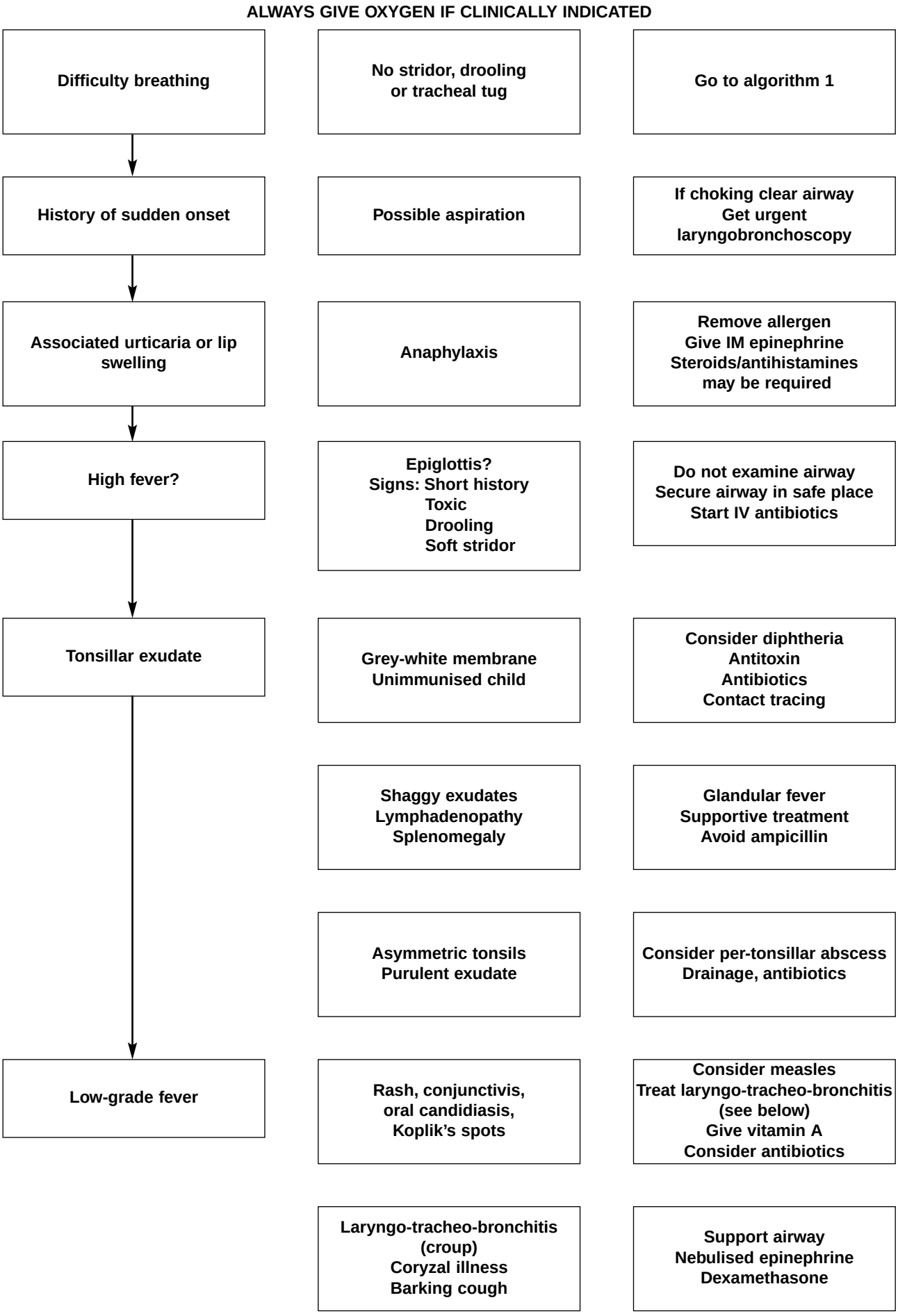
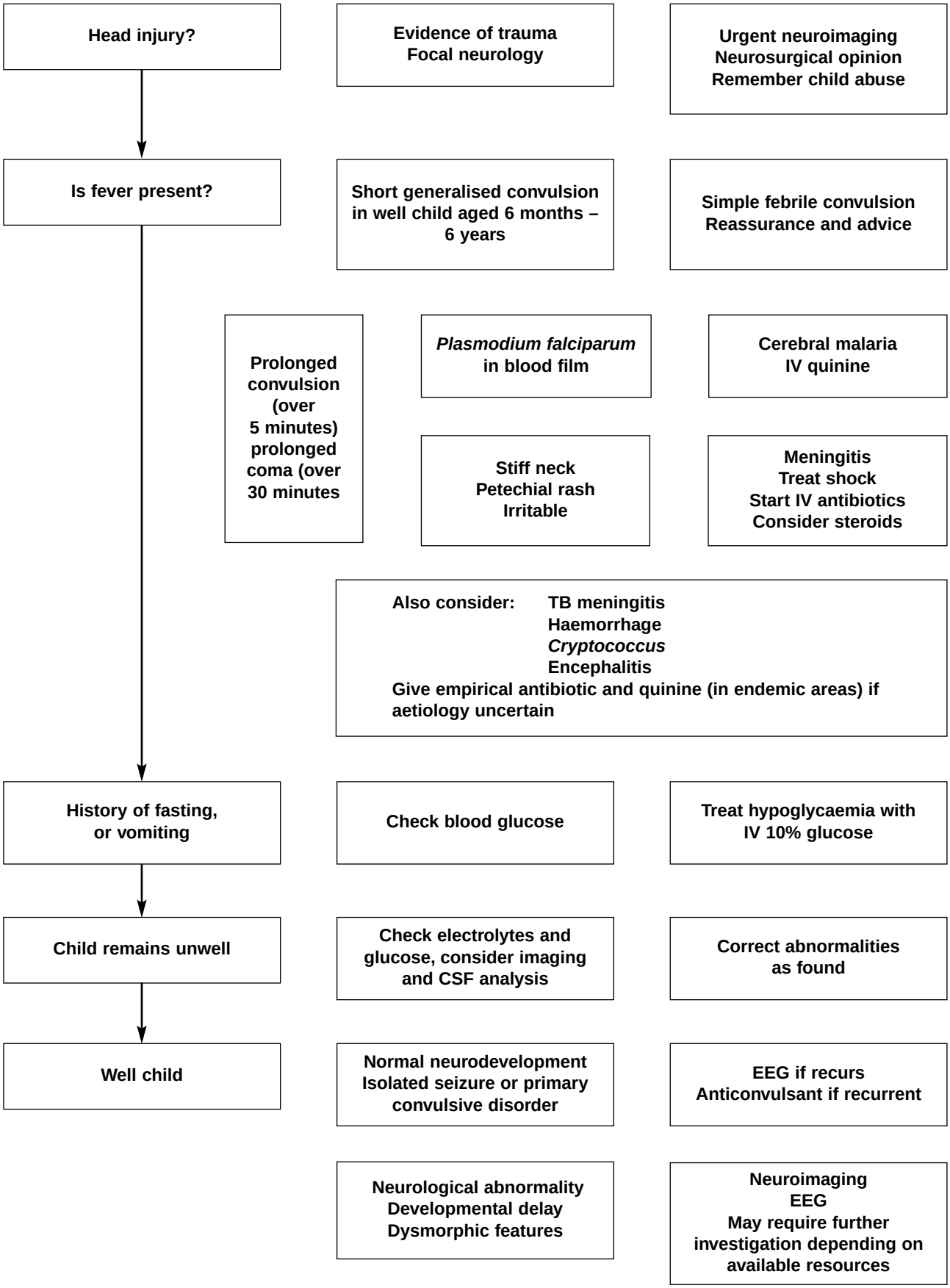
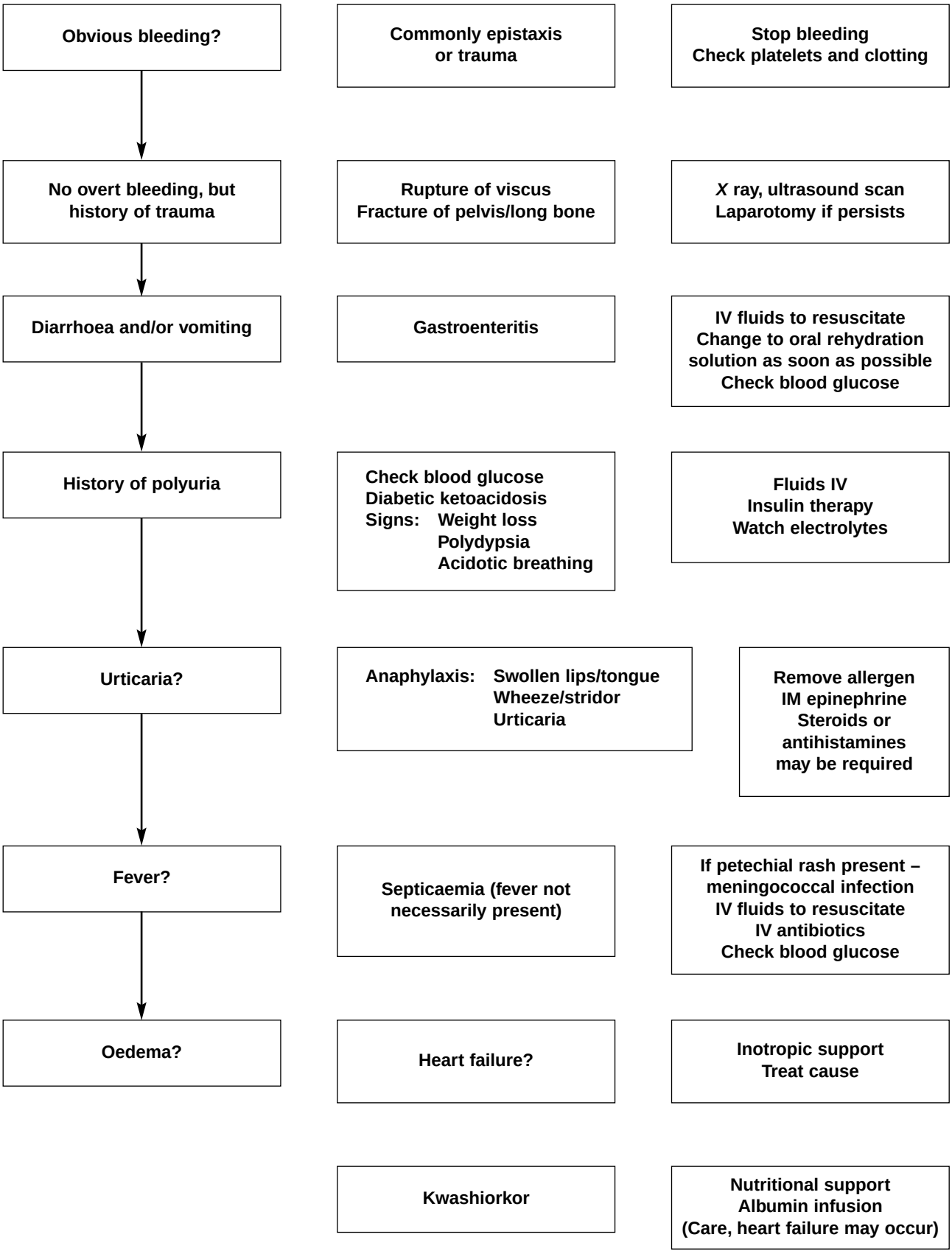


Figure 2.16.2 Upper airway obstruction



Notes
1 CSF analysis should not be done immediately after a convulsion, or where there are focal neurological signs or GCS <9.
2 Before considering diagnosis, stop convulsion and secure ABC.

Figure 2.16.3 Convulsion



Note
Treat shock immediately – if doubt over diagnosis, do not delay treatment.

Figure 2.16.4 Shock

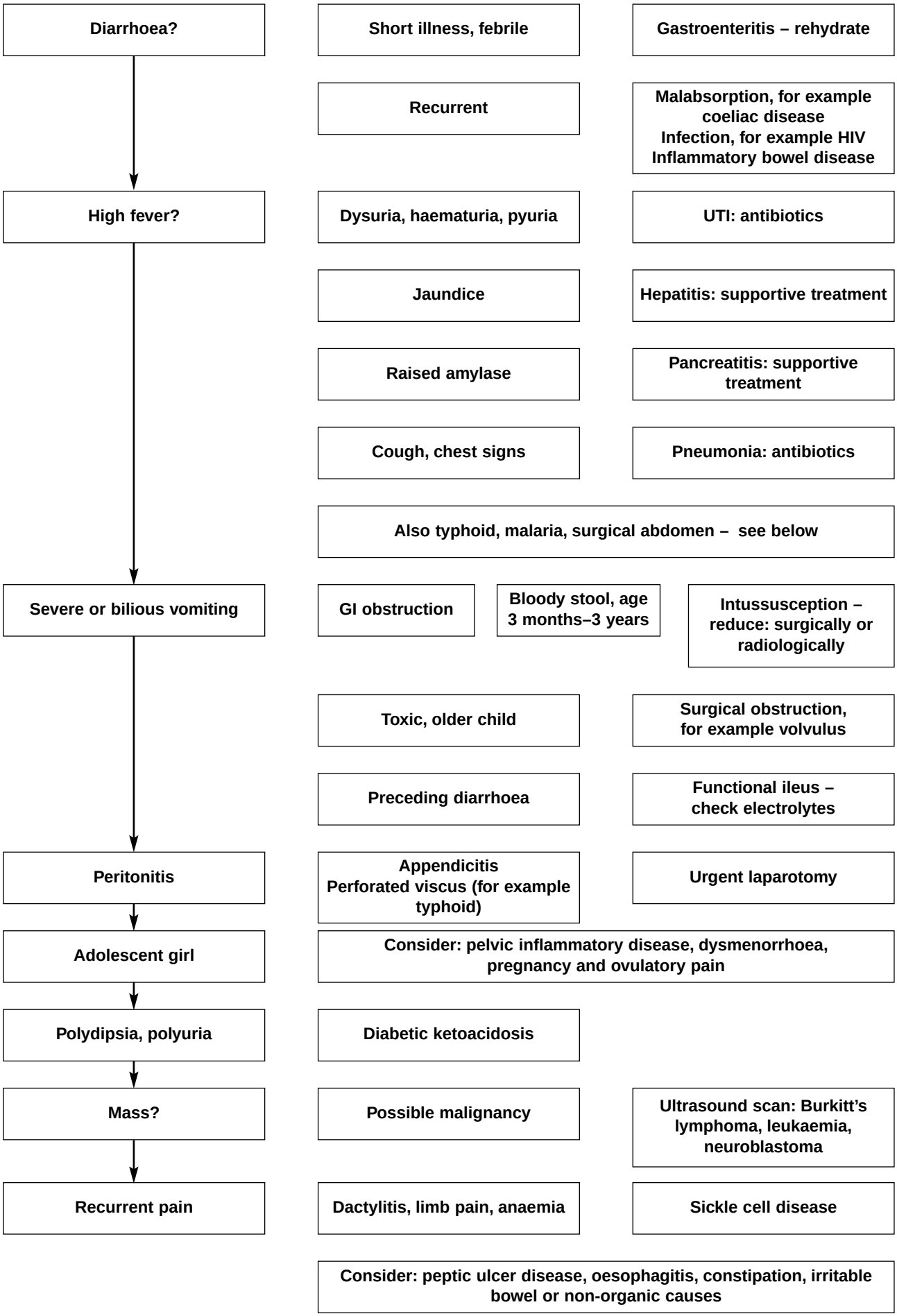


Figure 2.16.5 Abdominal pain

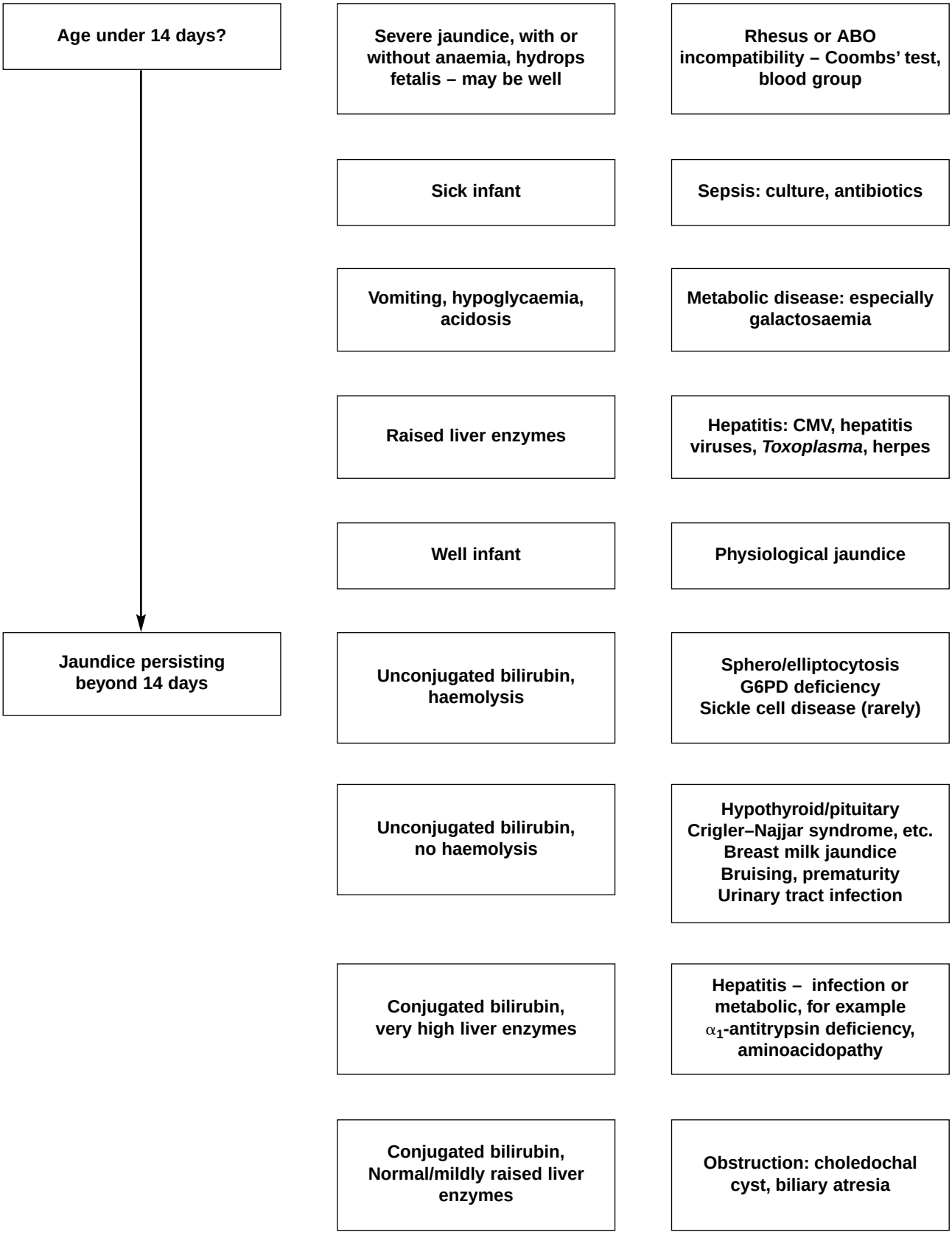
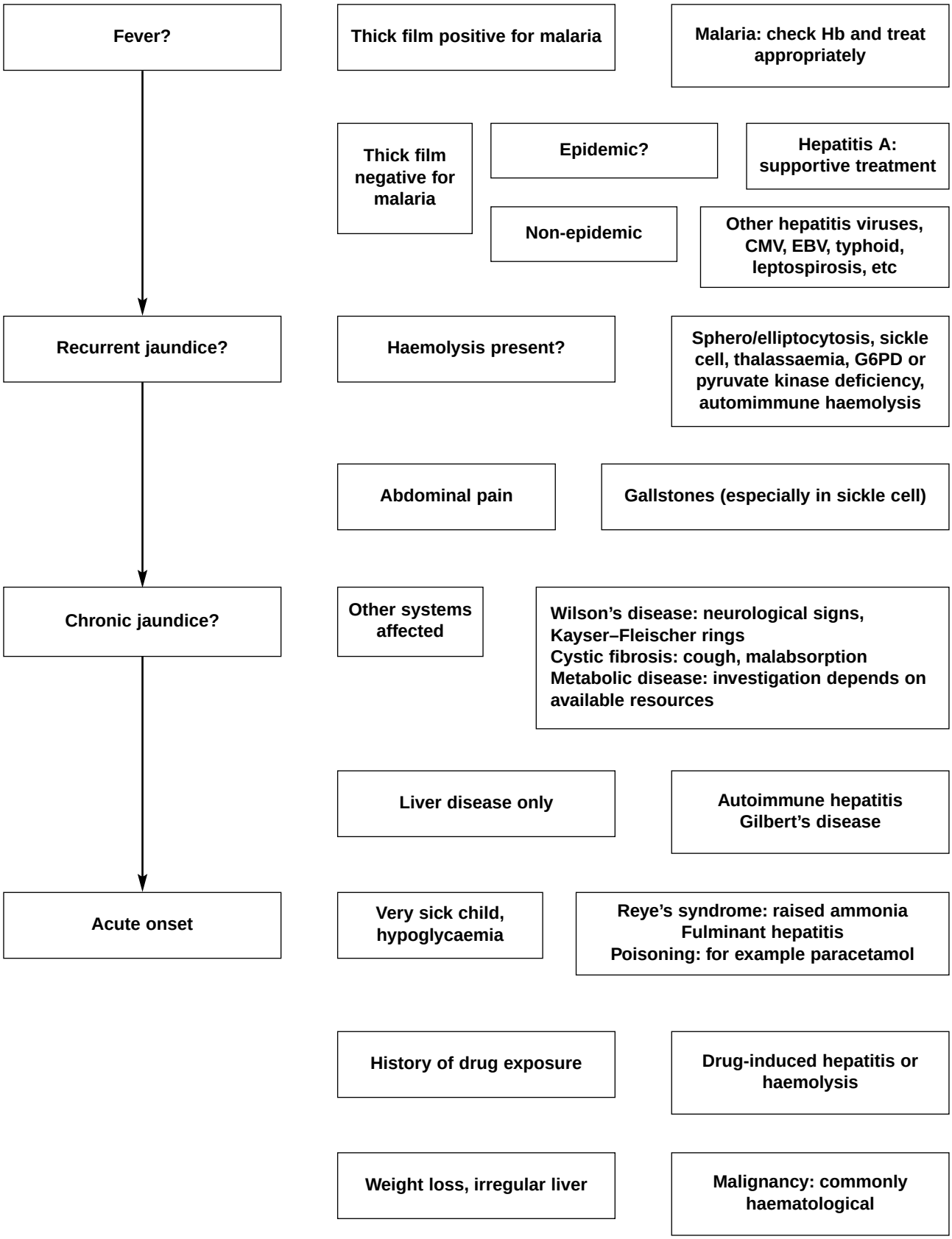
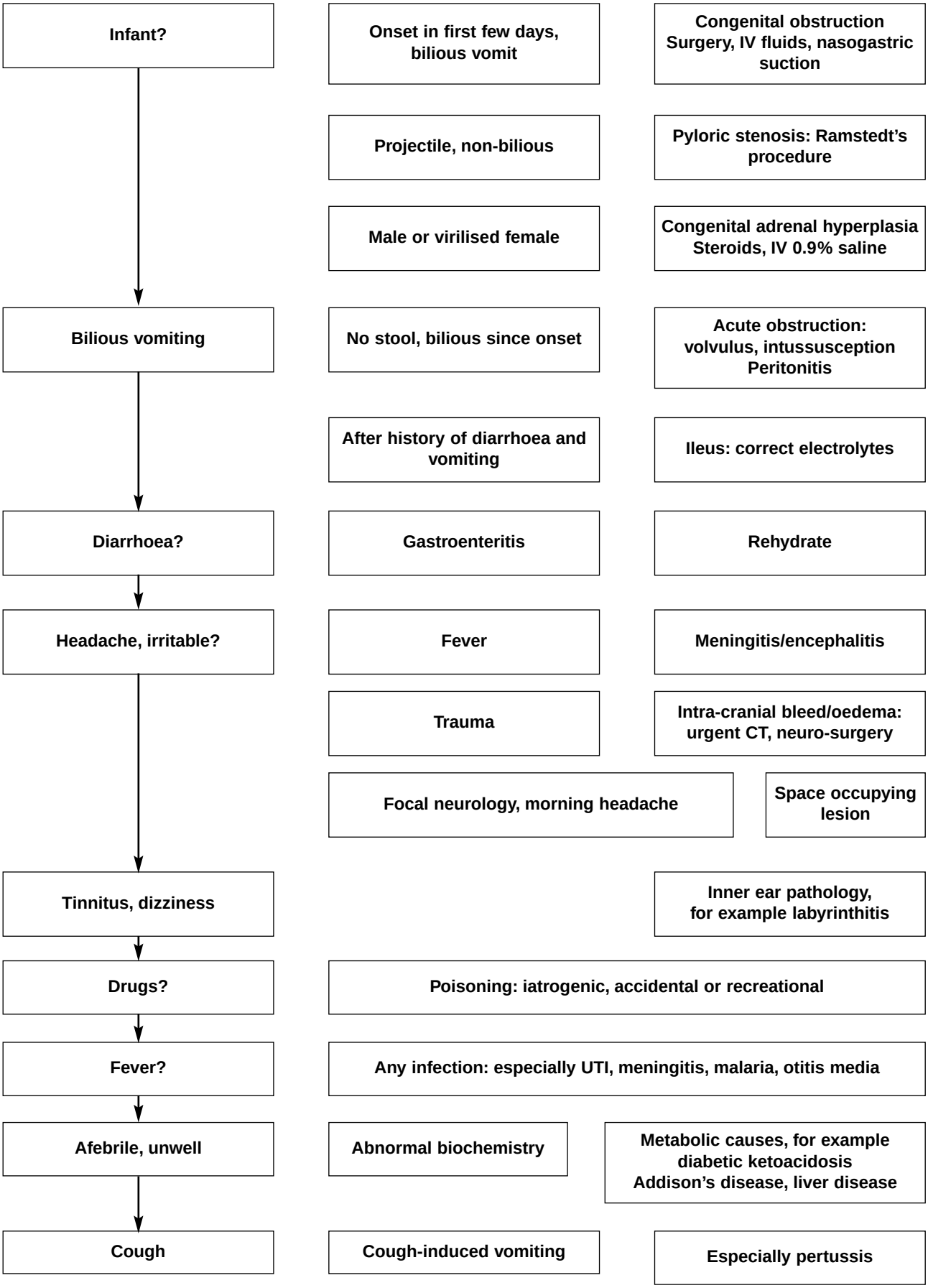


Figure 2.16.6 Neonatal jaundice



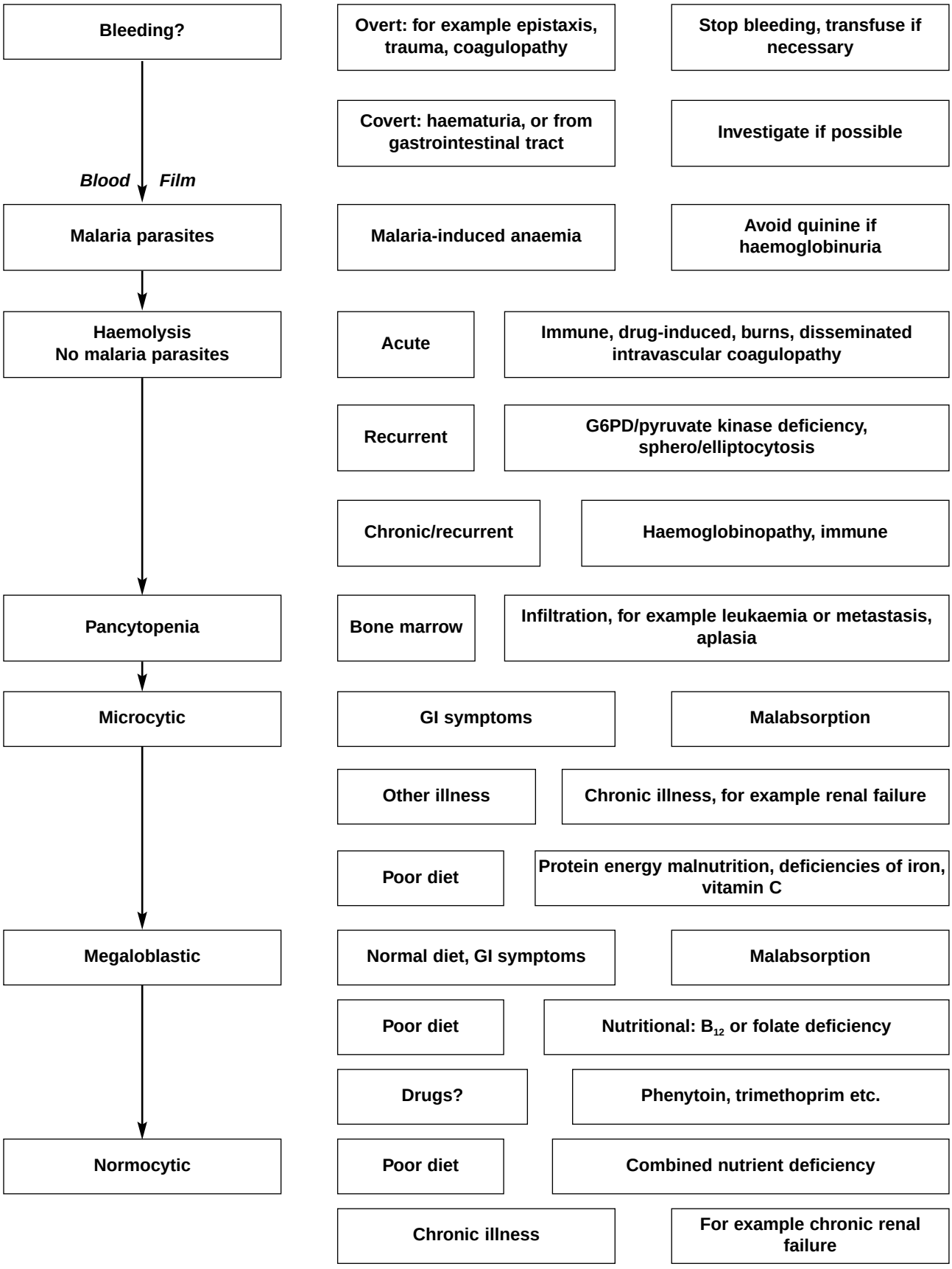
Notes
1 In any jaundiced child the following investigations should be done: blood film and malaria screen (endemic areas), liver enzymes, clotting factors, blood glucose, urine for bilirubin.
2 Features described above are not always consistent.

Figure 2.16.7 Jaundice



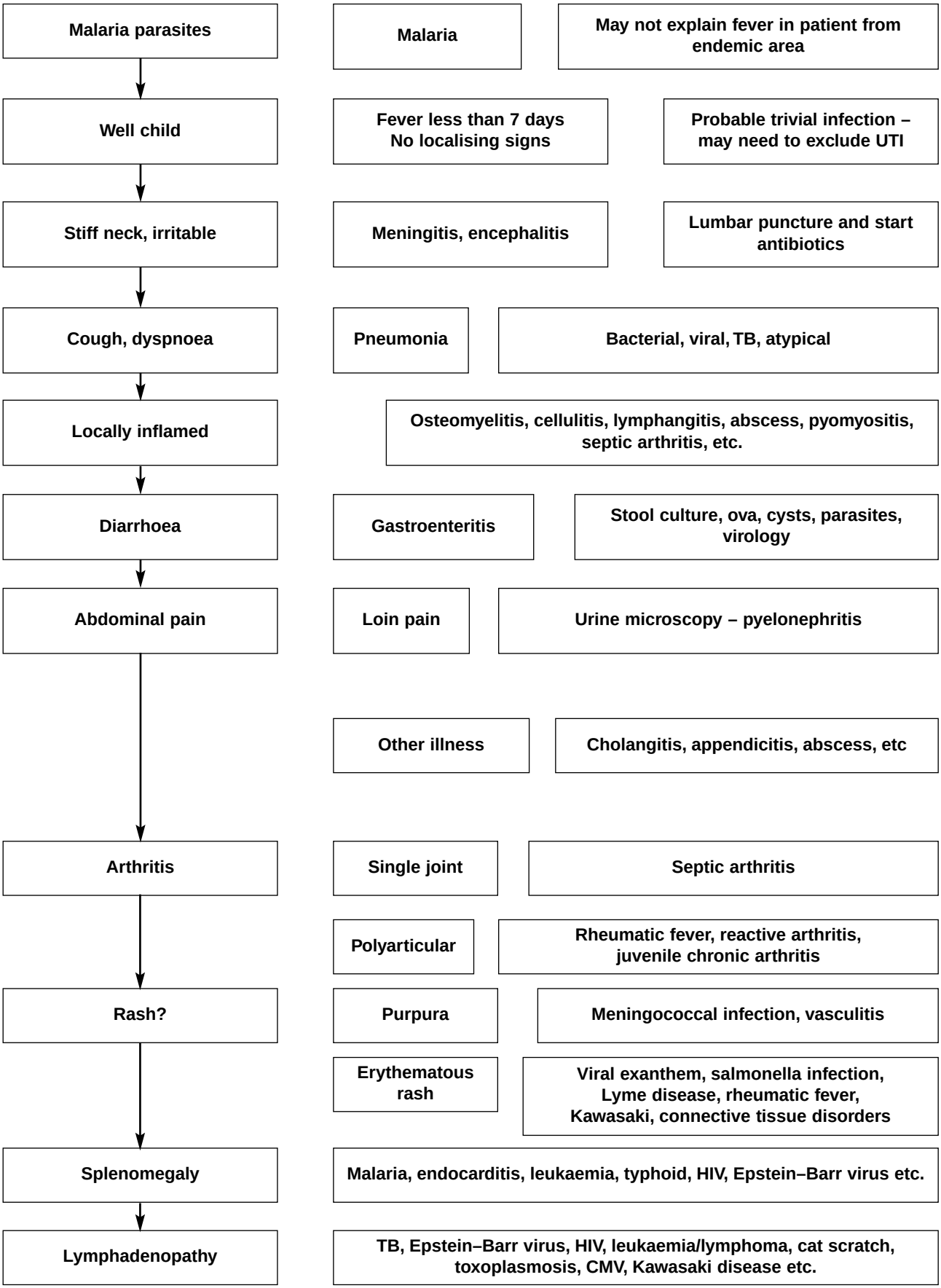
Notes
1 Vomiting is very non-specific. Almost any childhood illness or drug may cause vomiting.
2 Bilious vomiting is very uncommon in simple gastroenteritis.

Figure 2.16.8 Vomiting



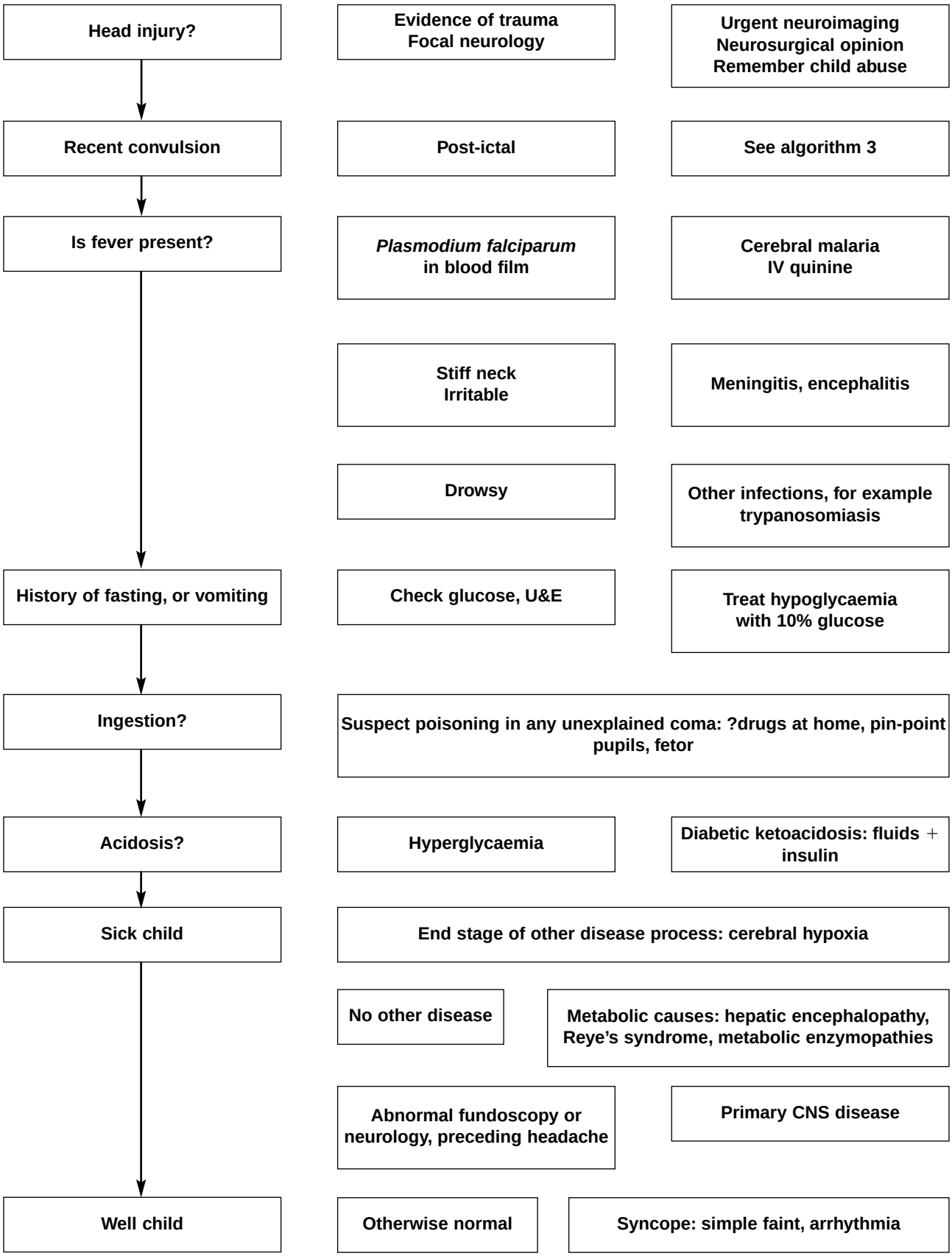
Notes
1 Anaemia is often multifactorial.
2 In endemic areas consider tropical splenomegaly syndrome if large spleen even if no malaria parasites seen.

Figure 2.16.9 Anaemia



Note
If fever prolonged >1 week, needs investigation (see Chapter 2.14).

Figure 2.16.10 Fever



Notes
1 CSF analysis should **not** be done immediately after a convulsion, or where there are focal neurological signs or GCS <9.
2 If in doubt or glucose monitoring unavailable give glucose 10% IV.

Figure 2.16.11 Coma

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Section 3

Management of system and organ dysfunction

How to use this book

This is a comprehensive text for all paediatricians caring for children in hospital. It can be used by those with limited resources and also where greater resources are available. We have identified the different levels of care in the following ways:

- **Minimum standards requirements** are given in a highlighted box at the the beginning of each clinical chapter.
- ***A standard of care*** where resources are not limited appears as bold, italicised text.
- **Key points** of particular importance in management of children are identified by a tick in the margin and bold text.

In this way we hope the book will act as a user-friendly, speedy reference on any paediatric ward.

3.1 Acute respiratory infection

Alan Smyth

Minimum standards requirements

- Oxygen, masks, nasal cannulae
- Antibiotics: chloramphenicol, penicillin, gentamicin, co-trimoxazole, amoxycillin, erythromycin, flucloxacillin
- Non-invasive respiratory support (Chapter 1.26)
- Chest drain kit (includes local anaesthetic)
- Physiotherapy

There are two categories of acute respiratory infection (ARI):

- **Acute upper respiratory infection (AURI):** above the vocal cords and epiglottis. These infections include colds; tonsillitis and otitis media. They are not life threatening but may lead to disability (for example otitis media is a leading cause of deafness in disadvantaged countries) and complications (for example rheumatic fever following streptococcal pharyngitis).
- **Acute lower respiratory infection (ALRI):** below and including the vocal cords and epiglottis. Including: croup (and other infectious causes of upper airways obstruction); pneumonia and bronchiolitis. Acute upper airways obstruction has been dealt with in Chapter 3.17.

Importance of ARI

Acute respiratory infections are responsible for 4 million of the 15 million deaths occurring annually in children under 5. Most of these deaths are from pneumonia. In poor countries, most of these infections are bacterial and the most common causative bacteria are *Streptococcus pneumoniae* and *Haemophilus influenzae*.

Immunisation

Currently available vaccines against *Streptococcus pneumoniae* do not produce good immunity in children under 2 years. The HiB vaccine (against encapsulated *H. influenzae* type B) will not protect against unencapsulated *H. influenzae* which causes some cases of pneumonia in the disadvantaged world. **Nevertheless, the HIb vaccine is very effective against other very serious infections due to *H. influenzae* (for example meningitis and epiglottitis) and should be given to all infants in every country.** ✔

Children at greatest risk of dying

These are the risk factors:

- Age under 1 year
- Malnutrition
- Where pneumonia is a complication of infection with measles, pertussis, malaria or HIV.

Diagnosis of ALRI

In many hospitals in poor countries special tests (such as microbiology of blood or respiratory secretions and *X* rays) may be limited or unavailable. However, because the prevalence of bacterial pneumonia is high, the diagnosis can be made clinically. This will not be 100% accurate, and so a few children may receive antibiotics unnecessarily (i.e. clinical diagnosis has less than 100% **specificity**). However it is more important not to miss children who do need antibiotics (i.e. clinical diagnosis should have a good **sensitivity**). Clinical diagnosis may be as accurate as an *X* ray and more helpful in deciding whether such treatments as oxygen are indicated. The clinical features will also help decide how severe the child's infection is and what treatment is appropriate. The following clinical features should be recorded and compared with Table 3.1.1:

- The presence of **cyanosis** which is best seen in the lips or tongue. It may be missed if lighting is poor or if the child is anaemic (for example due to co-infection with malaria). It can be difficult to detect in black children (this is a relatively late sign and when possible the state of oxygenation should be assessed with a pulse oximeter; at sea level SaO_2 is normally $>94\%$).
- An inability of the child to drink.
- The presence of chest indrawing – an inward motion of the lower chest wall when the child breathes in.

(The terms "subcostal indrawing," and "subcostal retractions" mean the same thing.) Intercostal retractions occur when the tissue between the ribs is "sucked in" as the child breathes in. This is a less severe sign of respiratory distress than chest indrawing. In infants under 2 months of age some indrawing during REM sleep is normal and it is only significant when it is severe. Chest indrawing signifies an increased work of breathing and is usually accompanied by flaring of the nose and the use of accessory muscles of respiration: **remember that a child who is exhausted at the time of presentation may not show this sign**. It is also important to note that infants less than 6 months of age may show hypoventilation and apnoea rather than increased work of breathing.

- An elevated respiratory rate. Respiratory rate is measured over 1 minute, using a suitable timing device. The respiratory rate in children varies with age. The following values define abnormally fast breathing for various age groups:
  - <2 months: >60 breaths/minute
  - 2–12 months: >50 breaths/minute
  - 12 months–5 years: >40 breaths/minute
- The presence of grunting (expiratory braking).
- The presence of hyperinflation (asthma or broncholitis).

Auscultation should always be undertaken, **but only after first looking for cyanosis and at respiratory pattern and the other signs as above.** Important clinical signs include:

- Evidence for consolidation or effusion/empyema
- Evidence of wheeze
- Evidence of bronchiolitis (widespread crepitations with or without wheeze)
- Evidence of alveolitis (for example, in HIV-induced *Pneumocystis* pneumonia) with end-inspiratory crepitations.
- Evidence for pericardial involvement (rare)
- Evidence for pneumothorax (rare).

A chest *X* ray may be helpful if there is a doubt about the diagnosis or if the child is seriously ill.

Additional features of ARLI usually include a fever and a cough. Pleuritic chest pain (which may radiate to the abdomen) may also be present in older children if the diagnosis is pneumonia.

Table 3.1.1 gives guidelines for the assessment and treatment of ARI. Children with the following features should be managed differently and this is dealt with elsewhere:

- Stridor (see Chapter 3.17)
- Wheeze (see Chapter 3.3)
- Severe undernutrition (see Chapter 3.16)
- Signs suggesting meningitis. (see Chapter 4.1)

For children with no evidence of pneumonia **but with signs suggesting a chest infection**, look for ear and throat infections or infections in another system and treat accordingly.

The management of a child with ALRI

Oxygen

Which children should have oxygen?

Children with severe pneumonia (central cyanosis or unable to drink) are likely to be hypoxaemic. However cyanosis is a late sign of hypoxaemia. **Oxygen should not be kept in reserve and given only when the child is dying.** This is useless and may create a mistaken belief amongst mothers that oxygen is harmful to sick children.

Giving oxygen

Give oxygen if:

- **Cyanosis**
- Restlessness (if oxygen improves the condition)
- Severe chest indrawing
- Breathing rate of >70 breaths per minute (child 2 months–5 years)
- Grunting (child <2months)
- A pulse oximeter is available and SaO_2 is <92–94% (at sea level; lower values will be present at high altitude and normal values of SaO_2 should be known for healthy local children).

Oxygen administration

The most reliable source of oxygen is an oxygen concentrator. This is a durable piece of equipment, but it requires a continuous supply of mains electricity to provide oxygen. It works on the "molecular sieve" principle, removing nitrogen from room air. The alternative is cylinder oxygen, but cylinders must be replenished regularly, which is expensive and may raise transport difficulties. The concentrator or cylinder should be connected to a low flow meter. The use of a flow splitter will allow up to four children to receive oxygen from one source. The oxygen should be delivered to the child using a nasal cannula or nasopharyngeal catheter (see Figures 3.1.1 and 3.1.2).

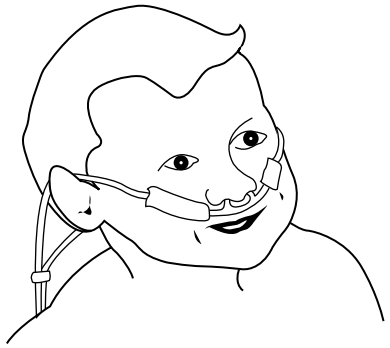


Figure 3.1.1 Oxygen being administered through a nasal cannula. The nasal prongs should only be 2–3 mm to avoid nasal irritation. The cannula has been taped to the child's cheeks, close to the nostrils. The tubing is run under the child's shirt to stop them pulling it and leads to the low flow meter and oxygen concentrator or cylinder. (A flow splitter may be used.)

Table 3.1.1 Recognition and management of ALRI (modified from WHO)

Classification	Most relevant clinical signs	Summary of treatment instructions
Pneumonia (severe ALRI)	Cyanosis or SaO_2 <92–94%	Admit
	Not able to drink	Give oxygen (sufficient to remove cyanosis or to keep SaO_2 >94%)
	Chest indrawing	Consider respiratory support if available (see Chapters 1.26 and 6.18)
	Expiratory grunting	Give an antibiotic: chloramphenicol (or benzylpenicillin and gentamicin)
	Tachypnoea or in infants less than 6 months hypoventilation or apnoea	Give supportive care Reassess frequently as dictated by state of child and response to treatment
Mild ALRI	None of the above except tachypnoea	Consider home care Give an antibiotic (at home): co-trimoxazole, amoxycillin or procaine penicillin.
	Cough and fever	Advise mother to return immediately if the child is getting worse.

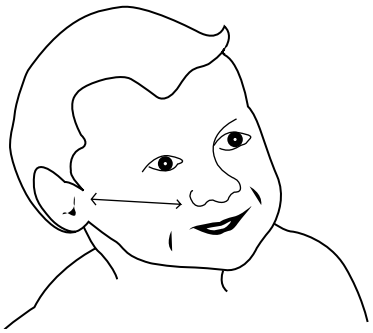


Figure 3.1.2 When a nasal catheter is used, the distance from the tragus (ear) to the nostril is first measured. This gives an estimate of how far the catheter should be inserted into the nostril. When inserted to this distance, the catheter should sit just at the edge of the soft palate.

The diameter of the nasopharyngeal catheter should be small (8F gauge is ideal) so that it does not obstruct the nasal airway. (Young infants are obligate nasal breathers.) The catheter should be clean and kept free of secretions. When the NP catheter is placed properly (Figure 3.1.3), an oxygen flow rate of 1 litre/min gives an oxygen concentration of approximately 50% in children up to 1 year.

In school-age children a mask may be used.

Antibiotics

See Table 3.1.1 for which antibiotic to use. Children who are vomiting or who require intravenous fluids should have their antibiotics given intravenously (preferably) or intramuscularly for the first 48 hours **if vascular access is difficult to achieve or maintain**. Some antibiotics such as gentamicin are always given parenterally. Some antibiotics are reserved for specific circumstances, for example high-dose co-trimoxazole for suspected *Pneumocystis carinii* pneumonia and flucloxacillin for pulmonary abscess or bacterial tracheitis. These are described at the end of the section on antibiotics.

Doses of antibiotics (give for a total of 7–10 days: IV plus oral)

- Chloramphenicol
  Intravenous (or IM if access is not possible): 25 mg/kg 6 hourly*
  Oral: 20 mg/kg 6 hourly

  *These are high doses used for severe infection. Intramuscular chloramphenicol is poorly absorbed – give it intravenously if possible and change to the oral route as the child improves. **Do not give chloramphenicol to infants <2 months old (risk of "grey baby syndrome").** In this age group give benzylpenicillin and gentamicin (*Listeria monocytogenes* will be effectively treated by penicillin). ✔
- Benzylpenicillin
  Intravenous (or IM if access is not possible): 50 000 U/kg 6 hourly (=30 mg/kg 6 hourly)
- Gentamicin
  Intravenous (or IM if access is not possible): 2.5 mg/kg 8 hourly* (see page 394)
  ***This drug is toxic to the kidneys and the inner ear. Make sure the dose is calculated correctly.**
- Amoxycillin
  Oral: 15 mg/kg 8 hourly
- Procaine penicillin
  Intramuscular only: 50 000 U/kg once daily

(when possible IM injections should be avoided (see page 393))

Antibiotics used in special circumstances

- Co-trimoxazole
  Oral: 24 mg/kg 12 hourly
  *High-dose co-trimoxazole: 60 mg/kg 12 hourly

  *If *Pneumocystis carinii* pneumonia (PCP) is suspected. Children in whom PCP is suspected and who are hypoxaemic should receive prednisolone 2 mg/kg/day while they are receiving co-trimoxazole. (Wean gradually if given for more than 3 weeks.)
- Erythromycin
  Prescribe if *Mycoplasma pneumoniae* infection or *Bordetella pertussis* is suspected.
  Oral: 125 mg 6 hourly (1 month–2 years)
  250 mg 6 hourly (2–9 years)
  500 mg 6 hourly (>9 years)

- Flucloxacillin (cloxacillin or oxacillin):
  Use if pulmonary abscess or bacterial tracheitis are suspected.
  Intramuscular or intravenous: 25 mg/kg 6 hourly
  Oral: 12.5 mg/kg 6 hourly

Symptomatic and supportive treatment

Fever
Remember fever may not be due simply to the child's pneumonia. Consider other diagnoses such as malaria.

Paracetamol
Oral or rectal: 10–15 mg/kg 4–6 hourly

Dehydration
See also Chapter 3.25 for the treatment of diarrhoea and Chapter 3.6 for the management of shock. Look for signs of dehydration or shock (capillary refill time prolonged >2 seconds).

- Shocked: Site an intravenous line and give a bolus of crystalloid, for example 0.9% saline, Ringer's lactate or colloid 20 ml/kg (10 ml/kg in a neonate).
- Not shocked but clinically dehydrated: see Chapter 3.25. ORS 15–20 ml/kg/hour for 2 hours orally or via nasogastric tube. Encourage breastfeeding.

Other supportive treatment

- Nurse the child in a thermoneutral environment (lightly clothed in a warm room at around 25 °C).
- Gently clear any secretions which are obstructing nasal breathing.
- When the child is recovering nutritional rehabilitation may be necessary (see Chapter 3.16).

Management of ALRI in special circumstances

Management of the child under 6 months of age
Young infants with severe ALRI/pneumonia may not cough, rather they may present with apnoea, poor feeding or hypothermia. Remember, in infants under 2 months, the respiratory rate cut off is higher (>60 breaths/minute). For ages 2–12 months the cut off is >50 breaths/minute.

- Some chest indrawing is normal during REM (dream) sleep.
- ✔ **All infants with severe ALRI/pneumonia should be admitted to hospital for treatment.**
- Bronchiolitis is a frequent cause and usually involves hypoxaemia due to V/Q mismatch. Oxygen is usually required. Additional respiratory support (see Chapter 1.26) may also be necessary especially if there is apnoea or severe respiratory distress leading to exhaustion.
- Grunting (a short, gruff noise at the start of expiration) is **common and usually an indication for oxygen**.
- Avoid using chloramphenicol in infants **under 2 months of age** (risk of "grey baby syndrome"). Use benzylpenicillin and gentamicin instead.
- Respiratory infection in neonates may rapidly develop into septicaemia, shock and death so it is essential to act quickly.

Management of the child with stridor
See Chapters 2.16 and 3.17.

Management of the child with wheeze
Wheezing is a whistling noise heard during expiration. The child who has cough or difficulty breathing *and* wheezing will fit into one of the following categories:

- Pneumonia complicated by wheezing **(be careful there is no inhaled foreign body – see below)**
- Asthma (see Chapter 3.3)
- Bronchiolitis.

In the first two categories, a bronchodilator will be an essential part of treatment. An aerosol and large volume spacer (which may be improvised) is the best way of administering a bronchodilator (see Chapter 3.3 on asthma).

Bronchiolitis is very prevalent in rich countries (much more prevalent than bacterial pneumonia) but does occur in poor countries particularly in the rainy season.

- Infants with bronchiolitis will be coryzal, have a troublesome cough and may feed poorly.
- On examining the chest, there may be hyperinflation, wheeze and fine crackles at the lung bases.
- Bronchiolitis is a viral infection. Only supportive treatment (oxygen and fluids) is effective and antibiotics and bronchodilators do not help. However, unless **certain** that pneumonia is not present, it is safer to give antibiotics (as shown in Table 3.1.1) and a trial of a bronchodilator (stop it if it is not helping). Non-invasive respiratory support may be valuable (Chapter 1.26).

Management of the child with acute upper respiratory infection AURI

- Cold or pharyngitis: symptomatic treatment only.
- Acute otitis media: The tympanic membrane is red and bulging or may have already perforated, discharging pus. Give 5 days of oral antibiotics (co-trimoxazole or amoxycillin) and then reassess. Continuing fever and a swelling behind the ear indicates mastoiditis (see Chapter 3.18).
- Tonsillitis (see Chapter 3.18) Many of these infections are viral, but a streptococcal infection may be complicated by rheumatic fever (Chapter 3.5). Look for enlarged tonsils with a white exudate and enlarged, tender, tonsillar nodes. Many rheumatic fever prevention programmes recommend:
  Benzathine penicillin
  Intramuscular (single injection):
  children <5 years 600 000 U
  children >5 years 1 200 000 U

Inhaled foreign body

Diagnosis of an inhaled foreign body

Any small object that can get into the trachea or large bronchi, such as a seed, a peanut or an eraser from the top of a pencil. There may be a clear history from the parent or child of an episode of coughing or choking, followed by difficulty breathing. On examination of the child's chest

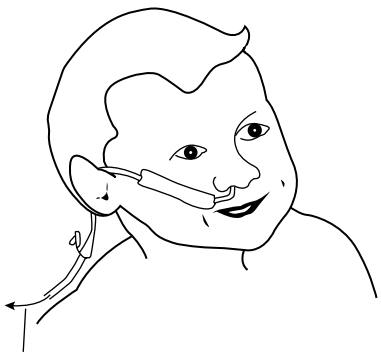


Figure 3.1.3 The nasal catheter in position. An 8 FG feeding tube has been used and taped to the cheek. If an additional tube is used for nasogastric feeding use the same nostril so that the child can breathe through the other side. *Label the 2 tubes to avoid confusion*. A flow rate of 1 l/min gives an inspired oxygen concentration of around 50%. This is appropriate for a child over 2 months. Use 0.5 l/min for infants under 2 months. Keep the catheter free of mucus. Oxygen should be humidified and the water must be clean and changed daily.

is there less chest expansion on one side when breathing in? Feel the trachea – it may be pushed away from the midline by air trapping on the side affected by the foreign body. This may also be seen on a chest *X* ray, if this is available; ideally an expiratory and inspiratory film. An inhaled foreign body in a young child can go down the right or left side. In older children and adults, a foreign body on the right is much more common. There may be a harsh wheezing noise, heard on the side of the chest where the foreign body has lodged.

Treatment

Air may be trapped in the lungs beyond the point where the foreign body has lodged, or this part of the lung may become infected. Give the child antibiotics: chloramphenicol is a good first choice but add flucloxacillin if there is a suspicion of proven infection with *S. aureus* (see above for doses). Removal of a foreign body is a specialised procedure which must be carried out using a rigid bronchoscope. Treat infection until the child can be transferred to a hospital where this procedure can be performed. Some gentle physiotherapy may help but take care not to dislodge the foreign body, causing infection and obstruction in another part of the lung.

Emergency treatment of a large foreign body obstructing the airway

Attempt to dislodge and expel it. (See Figure 3.1.4) Large foreign bodies stuck in the oesphagus can also inhibit breathing and produce hypoxaemia.

Pleural Effusion

Diagnosis

A pleural effusion is a collection of fluid between the chest wall and the lung. A small effusion of clear fluid is common in children with pneumonia. This should be suspected when one side of the chest sounds very dull to percussion and the breath sounds are very quiet. It can also be seen as shadowing on the *X* ray. Usually this fluid will quickly disappear once the infection has been treated. However, if treatment is started late, or the child is unlucky, this clear fluid can become infected too. This leads to pus accumulating in the chest cavity (an empyema).

Treatment

If a pleural effusion is suspect, *X* ray the chest if possible. If this confirms your suspicions then perform a diagnostic tap.

- The best way to do this, with young children, is with the child sitting on mother's lap, facing her. The mother then holds the child tightly in a bear hug. Older children can sit on a stool but it is important to explain carefully to them what is being done and have a helper to hold the child steady.
- Percuss out the area of dullness, put on sterile gloves and clean the skin with alcohol.
- Gently inject some local anaesthetic (1% lidocaine) under the skin, down to the rib using an orange (25 gauge) or blue needle (23 gauge).
- Then take a fresh 20 gauge needle or butterfly needle connected to a syringe and press the needle though the chest wall just below the level where the percussion note becomes dull. **Remember to go just above the rib (to avoid intercostal blood vessels)** and aspirate all the time. Ultrasound support is ideal if available. ✔

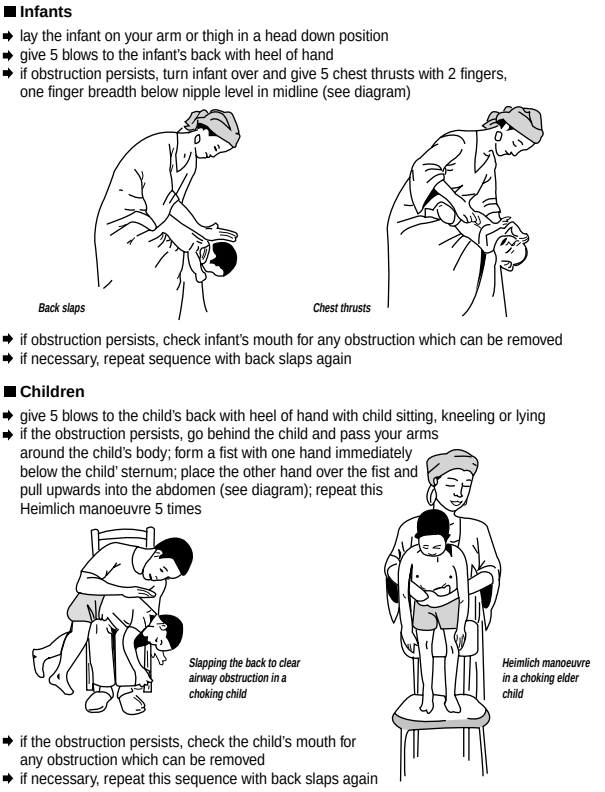


Figure 3.1.4 How to manage the airway in a choking child (foreign body aspiration with increasing respiratory distress).

- When fluid appears, aspirate a diagnostic specimen and send this for microscopy, protein level, cell count, Gram and Ziehl-Neelsen stain (low yield for AFB), and culture for bacterial and tuberculosis. Aspirate as much fluid as possible during the procedure to allow the child to breathe more comfortably. A three-way tap connected to the catheter can be helpful. ENSURE AIR CANNOT ENTER THE PLEURAL SPACE.

If clear fluid is aspirated (straw coloured or brown) remove sufficient fluid to relieve distress and then remove the needle.

If more than a few millilitres of fluid containing pus (opaque) is aspirated and this does not easily pass down the needle, then a chest drain will be required. This is best done under a general anaesthetic by a surgeon because he or she can break down any fibrous tissue in the chest cavity that could slow healing. If there is no surgeon available, then it will have to be placed under local anaesthetic. This must be a sterile procedure.

- Position the child and locate the effusion in the same way as for the diagnostic tap.
- Use sufficient local anaesthetic 1% lidocaine as outlined in Chapter 1.13.
- Make an incision in the skin and part the underlying muscle with artery forceps.
- Avoid the neurovascular bundle on the inferior part of the rib by keeping the incision and passage of the drain on top of the rib.
- When the pleura is reached, puncture the pleura with the forceps and thread the chest drain, the largest that will comfortably pass through the intercostal space into the cavity by holding the tip of the tube in the forceps. Do not use the stylette as this can damage the lung.
- Ensure all the drainage holes of the catheter are inside the chest.
- Fix the drain with a gauze dressing, tape and a suture.
- Connect to an underwater seal. If the drain has been placed correctly, fluid will flow out and the fluid level will "swing" with respiration.

A prolonged course of antibiotics will be needed (2–4 weeks). Use chloramphenicol (with flucloxacillin if possible). (See above for doses.) If possible give the first week intravenously before changing to oral. Remember a clear fluid aspirate can suggest another diagnosis such as tuberculosis or lymphoma (especially if blood stained).

Lung abcess

Diagnosis

A lung abscess is collection of pus in the lung. This can result from an untreated foreign body, aspiration of other material such as vomit, infection with *S. aureus* or as a complication of bronchiectasis. When examining the child, the findings may be similar to the child with pneumonia, though he/she will often have been ill for longer. A chest *X* ray will be helpful. Ultrasound can show whether the abscess lies close to the posterior chest wall.

Treatment

Antibiotics are the most important form of treatment and a long course (4–6 weeks) must be given. Use chloramphenicol and flucloxacillin as in empyema. If certain the abscess lies close to the posterior chest wall then it can be aspirated it in the same way as an empyema (see above). **On no account should a chest drain be placed in a pulmonary abscess as this will create a fistula**. If the child has been ill for weeks, then ensure good nutrition. ✔

Bronchiectasis

Diagnosis

Broncheictasis occurs when the bronchi become baggy and full of mucus and pus. Bronchiectasis may follow infection such as tuberculosis, pertussis and measles. It may be due to a congenital problem such as cystic fibrosis (see Chapter 3.4) or abnormal cilia.

Sometimes a child who has had lobar pneumonia does not recover fully and develops bronchiectasis in the affected lobe. There are other rare causes such as some viral infections.

Children with bronchiectasis usually cough and produce sputum every day. Their symptoms may become much worse at times due to secondary infection. The child may have finger clubbing and a hyperinflated chest and coarse crackles in many parts of the lung. Look for thickened bronchi and areas of consolidation on the chest *X* ray.

Treatment

Bronchiectasis cannot be cured though ***occasionally symptoms can be improved by removing the lung lobe that is most severely affected***. The child and their parents must understand that daily treatment with chest physiotherapy and frequent courses of antibiotics will be needed. The use of antibiotics (above) and physiotherapy is described in Chapter 3.2.

3.2 Physiotherapy for suppurative lung disease

Sarah Samuels

This chapter describes therapy for bronchiectasis, cystic fibrosis and other conditions with excess airway secretions.

Postural drainage

Positioning to allow drainage by gravity from lung segments to central airways.

Infants: use a maximum of five positions in 10 minutes, progressing to older children: (two to three positions in up to 30 minutes).

- upper lobe
  - apical segments – sitting (Figure 3.2.1.1)
  - posterior segments – prone, one pillow below affected side (Figure 3.2.1.2)
  - anterior segment – supine (Figure 3.2.1.3)
- middle lobe/lingual
  - chest tipped 15° below horizontal, lying supine, with a pillow supporting ipsilateral hip and shoulder (Figure 3.2.1.4)
- lower lobe
  - apical segments – prone (Figure 3.2.1.5)
  - anterior basal – chest tipped 20° below horizontal, lying supine (Figure 3.2.1.6)
  - lateral basal – chest tipped 20° below horizontal, lying on unaffected side Figure (3.2.1.7)
  - posterior basal – chest tipped 20°, below horizontal, lying on unaffected side (Figure 3.2.1.8)

Equipment

Carer's lap (infant)/bean bags/pillows/tilted bed.

Adjuncts to postural drainage

The following may be combined with postural drainage:

- Chest clapping – done over area to be cleared with a cupped hand
- Chest shaking – fine manual shaking in line with rib motion during the expiratory phase of breathing
- Active cycle of breathing – relaxed tidal breathing, four deep breaths to maximal inspiration with hold, relaxed expiration. Huff – forced expiration at mid to low lung volumes with glottis open (as if misting glass), cough to clear secretions and repeat cycle until chest clear. Note where bronchoconstriction is an issue: (i) increase time spent doing tidal volume breathing, (ii) omit percussion, or (iii) increase tidal volume breathing and omit percussion.

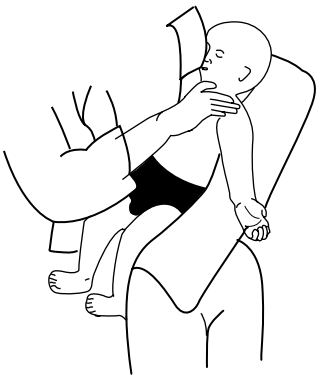
Relative contraindications

- Raised intracranial pressure
- Severe hypertension
- After abdominal surgery
- Major haemoptysis
- Pulmonary oedema
- Surgical emphysema
- Tension pneumothorax
- Cardiac arrhythmias
- Gastro-oesophageal reflux

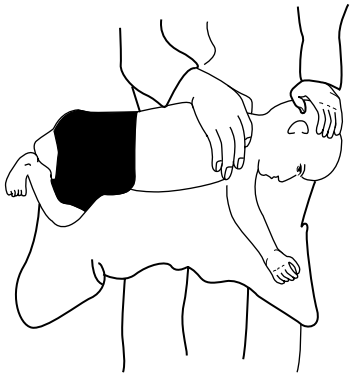
Positioning

- To maximise ventilation-perfusion matching (for example pneumonia, asthma, pneumothorax) in self-ventilating patients, position with better ventilated lung uppermost.
- In severely breathless patients, use sitting with a forward lean, or recovery position. Use pillows to raise and support the chest where patients cannot tolerate lying flat.

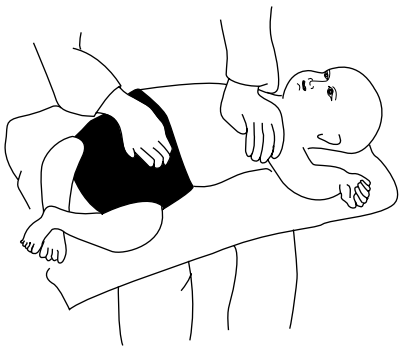
1 Apical segments upper lobes



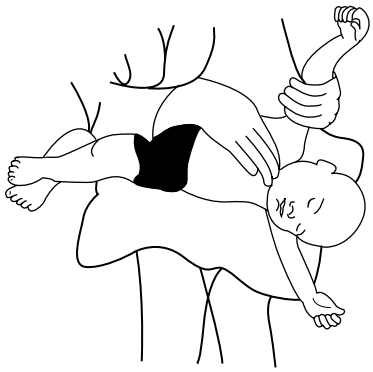
2 Posterior segment right upper lobe (reverse for left)



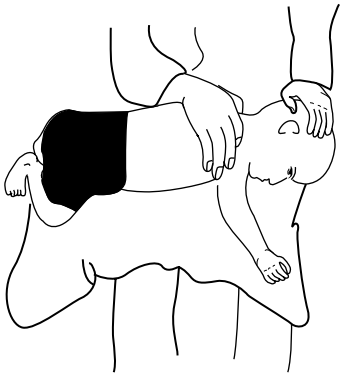
3 Anterior segments upper lobes



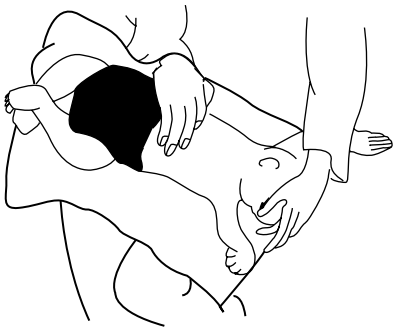
4 Right middle lobe (reverse for left)



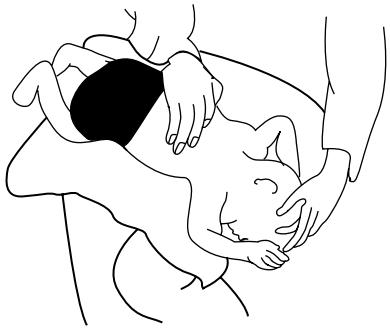
5 Apical segments of lower lobes



6 Anterior segments of lower lobes



7 Posterior segments of lower lobes



8 Lateral segments of right lower lobes (reverse for left)

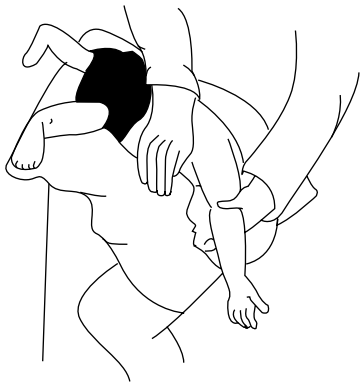


Figure 3.2.1 Postural drainage positions

3.3 Asthma

Martin Samuels

Minimum standards requirements

- Inhaled bronchodilators (with spacers)
- Inhaled steroids
- Prednisolone/hydrocortisone
- Nebulised bronchodilators
- Oxygen
- Epinephrine/IV salbutamol/IV aminophylline
- Pulse oximetry

Asthma is a condition characterized by episodic or recurrent symptoms of cough, wheeze, chest tightness and shortness of breath. It is due to variable and reversible airway obstruction associated with chronic airway inflammation. Asthma has become more prevalent over the last 20 years, along with the other atopic conditions such as allergic rhinitis and eczema. This is particularly so in rich countries where it is reported to occur in up 10–15% of children.

Young children (<5 years) often have "asthma-like" symptoms (cough, wheeze and shortness of breath) in response to respiratory infections, but with no demonstrable problem between infections. This tendency often stops in early school years. In these children, treatment of episodic symptoms with asthma therapies may still provide relief of symptoms. However, in young children (<2–3 years old) with severe episodes or symptoms continuing between infections (interval symptoms), it is necessary to consider or exclude other diagnoses, such as bronchiectasis, tuberculosis, foreign body and cystic fibrosis.

Diagnosis

The diagnosis is clinical, and based on a history of:

- recurrent cough, wheeze, shortness of breath, or chest tightness
- symptoms worse at night, and on exertion
- symptoms worsened by respiratory infections, inhaled irritants (for example cigarette smoke), cold air, animal furs, excitement or upset
- personal or close family history of eczema, rhinitis or asthma.

Examination may identify:

- no abnormalities
- slow growth
- overinflation of chest, Harrison's sulci
- wheeze, particularly on forced expiration
- rhinitis or eczema.

Investigations are not usually needed, but may help support the diagnosis or exclude other conditions:

- Chest *X* ray – is normal or shows overinflation (flat diaphragms and hyperlucency, particularly when severe or acute), or increased peri-hilar linear markings
- ***Peak flow (in 7–8 year olds and over)*** – this may show >15%:
  - variability from morning to night (keep a peak flow diary)
  - fall after 5–10 minutes hard exercise
  - rise after a dose of inhaled bronchodilators (for example salbutamol)
- ***Spirometry – will show FEV1:FVC of < 85% and concavity in flow-volume loop, at least partially reversed by a dose of inhaled bronchodilators.***

Skin prick tests, or IgE RASTs do not help in the diagnosis, and infrequently in the management.

Symptoms that resolve with bronchodilators with or without steroids also support the diagnosis.

Management

- Avoid allergic/irritant factors, for example smoke, chemical fumes, house dust mites, animal fur. Discourage cigarette smoking and new pets at home.
- Do not stop child from exercising, but predose 5–10 minutes beforehand with a dose of inhaled bronchodilators (for example salbutamol, terbutaline).
- Occasional symptoms (for example on 2–4 days per week) may be relieved with a beta-2 agonist bronchodilator (a reliever).
  - Use inhaled where possible, apart from in acute severe or life-threatening attacks when the intravenous route may be used.
  - Use an aerosol spray (metered dose inhaler) with a spacer (first choice):
    (i) A commercial medium to large volume spacer, for example Volumatic, Nebuhaler, Aerochamber, or

(ii) A plastic bottle or polystyrene cup with the aerosol sealed into one end, and the open end held closely over the nose and mouth
(iii) Use 200–1000 micrograms (2–10 sprays): more may be needed in younger children, or if acutely breathless (and repeated)
(iv) Each spray/puff should be inhaled individually in turn with 4–5 breaths, rather than filling the spacer device with multiple sprays
(v) For children <5 years old, attach a facemask (for example inverted adult mask) to the mouthpiece of a spacer
(vi) clean spacer with soapy water and leave to dry naturally to reduce static electrical charges on inside.

- Alternatively use a dry powder inhaler such as a rotahaler, turbohaler, diskhaler (these are more portable) or nebuliser (less portable).

✔ **Children with asthma should always have immediate availability to their usual reliever inhaler device: over 7–8 year olds may keep their device with them.**

- More frequent symptoms, regular nocturnal symptoms or daily use of a bronchodilator should be treated with regular medication aimed to control airway inflammation (preventer): cromoglycate or steroids. Use inhaled, preferably through a spacer (first choice).
  - sodium cromoglycate: 10 mg (two puffs of MDI) four times daily (expensive)
  - beclomethasone: 200–400 micrograms twice daily
    – rinse mouth after each dose of inhaled steroid
    – aim for rapid control of symptoms, and then tail down dose slowly
    – gaining control may be helped by a short course (7–10 days) of systemic steroid (for example prednisolone 500 micrograms/kg once daily after food or milk, maximum daily dose 40 mg)
    – continue with bronchodilator use for symptom relief
- For regular or severe symptoms, consider:
  - if diagnosis is correct
  - if there are other aggravating factors, for example rhinitis, stress, gastro-oesophageal reflux
  - if medication is being taken correctly
  - increasing inhaled steroid dose (beclomethasone to 400–800 micrograms twice daily) or
  - ***adding long-acting inhaled (for example salmeterol) or long-acting oral beta-2 agonists*** or
  - oral methylxanthines (for example theophylline 5 mg/kg 3–4 times a day)
  - **as a last resort**, use of alternate-day oral prednosilone (start at 500 micrograms/kg on alternate days and reduce rapidly to 100 micrograms/kg on alternate days [to nearest 1 mg or 5 mg tablets]). Stop as soon as possible.

✔ **Children on inhaled or oral steroids should have regular checks of their growth and be watched for steroid side effects (for example oral thrush)**

The control of asthma should be regularly reviewed (for example three-monthly) and medication stepped up or down dependent on symptoms ± peak flow measurements ***or spirometry***. Families should have written instructions and may learn to change treatment themselves, with support.

Acute asthma

Initial treatment of a mild to moderate acute attack of asthma includes:

- Reassure child and parents and avoid upset which may exacerbate respiratory distress
- Give regular inhaled beta-2 agonist bronchodilator, for example salbutamol aerosol 200–1000 micrograms via spacer or 2.5 mg for <5 years and 5 mg for >5 years via nebuliser 2–4 hourly (use oxygen to drive the nebuliser if possible)
- Give systemic steroids: oral prednisolone 500 micrograms/kg (maximum of 40 mg) with food or milk to avoid gastric irritation or IV hydrocortisone 4 mg/kg 12–24 hourly
- Treat or remove any exacerbating factors (see "Diagnosis" above).

Features of severe or life-threatening asthma include:

- too breathless to feed, drink or talk
- marked recession/use of accessory muscles
- respiratory rate >50 breaths/min
- pulse rate >140 beats/min
- poor chest movement/silent chest
- exhaustion/agitation/altered conscious level.

Treat immediately (use 'ABC' approach): ✔

- **100% oxygen** via facemask with reservoir bag at 10–15 litres/min ✔
- Inhaled beta-2 agonist **bronchodilator** given **continuously at first** and then 30 minutes to 2 hourly, for example salbutamol aerosol 1–2 mg (10–20 puffs) via spacer, or 2.5 mg for <5 years and 5 mg for >5 years via nebuliser, and repeated as required. ✔
- If nebulised or inhaled bronchodilators not available, give subcutaneous epinephrine 10 micrograms/kg of 1 in 1000 to a maximum dose of 300 micrograms. If no improvement after 20 minutes, repeat dose
- **Systemic steroids** (prednisolone or hydrocortisone – see above) ✔
- Consider intravenous beta-2 agonist salbutamol (loading dose 5–15 micrograms/kg over 10–15 min, followed by 100–500 nanograms/kg/min (that is 0.1–0.5 micrograms/kg/min) by IV infusion. Severe and life-threatening hypokalaemia may occur with IV salbutamol, potentiated by steroids. If possible monitor the ECG continuously and check K^+ 12 hourly. ECG signs of hypokalaemia are: ST depression, T wave reduction and prominent U waves. Ensure maintenance potassium intake is given.
- An alternative is aminophylline (loading dose 5 mg/kg over 20 minutes, followed by 1 mg/kg/hour by IV infusion if 1–12 years and 500 micrograms/kg/hour if

>12 years or <1 year of age. Severe nausea may occur with this drug.

Monitor above clinical features regularly and also monitor oxygen saturation, by pulse oximeter if available. Keep SaO_2 >94% by the administration of oxygen, either by face mask or by nasal cannulae. Use oxygen to drive nebulisers.

Transcutaneous PCO_2 monitoring is valuable in severe asthma.

In cases not responding to above measures, obtain chest *X* ray and ***consider mechanical ventilation (slow rate, long expiration). A blood gas measurement showing respiratory acidosis can be valuable at this time,*** **but remember that invasive procedures can worsen respiratory distress.** ***If intubation and ventilation becomes essential, ketamine induction followed by inhalational anaesthetic gases (for example halothane) may assist bronchodilatation.*** ✔

Following any acute episode, review asthma control and management, including correct use of medications and need for step up in "preventive" treatment.

3.4 Cystic fibrosis

Alan Smyth

Minimum standards requirements

- Pancreatic enzyme supplements
- Fat-soluble vitamins (A, D, E)
- Daily chest physiotherapy
- Early antibiotics: flucloxacillin, cephalosporins, amoxycillin, chloramphenicol, ciprofloxacin, gentamicin, ceftazidime

Cystic fibrosis (CF) is a genetic disorder (autosomal recessive) affecting lung, digestive system, sweat glands, liver, pancreas and reproductive system. Most deaths from cystic fibrosis are from respiratory failure. In rich countries, many patients now survive well into adult life.

Incidence

The incidence, in countries such as the United Kingdom and the USA, is around 1 in 2500 live births, and around 1 in 25 of the population are carriers. Very little is known about its frequency in poor countries. ***Diagnosis relies on the sweat test, which is difficult to perform where laboratory facilities are limited.*** The incidence of CF amongst black South Africans is thought to be between 1 in 700 and 1 in 14000 with between 1 in 14 and 1 in 60 being carriers amongst the general population. In some rich countries there is ***routine screening of newborn infants from heel prick blood samples***.

The CF gene

The CF gene is on chromosome 7. The commonest mutation causing disease is DF508 and it occurs all over the world. (It is as common in CF patients in North Africa as it is in Northern Ireland.) Over 600 other mutations have been found, many of which are rare. The gene product is a protein which sits on the apical membrane of epithelial cells and regulates the movement of chloride ions. When there are two abnormal genes, the protein is defective and chloride transport is disrupted.

Pathophysiology

In the cells lining the airways of CF patients, chloride ions cannot leave the cell to enter the bronchial lumen. The cell cytoplasm is high in salt and water moves from the airway lumen into the cell by osmosis. The mucus within the lumen becomes dehydrated. Sticky mucus interferes with the action of the respiratory cilia, leads to bacterial colonisation of the airway, with chronic inflammation and neutrophil damage. There are also viscid secretions in the biliary tract, pancreas and reproductive system.

Presentation

Meconium ileus

In the newborn period babies may present with a triad of:
- Failure to pass meconium in the first 24 hours
- Abdominal distension
- Vomiting.

This picture may also occur in surgical conditions (Hirschsprung's disease, imperforate anus) and the sick newborn infant may develop non-specific abdominal distension. Around 15% of babies with CF present with meconium ileus; difficulty passing thick, sticky meconium, leading to small bowel obstruction.

Presentation in older children

- Malabsorption (pale, greasy stools) and failure to thrive
- Rectal prolapse
- Chronic and recurrent chest infections
- Partially digested material, with a high fat content may block the ascending colon ("distal intestinal obstruction syndrome").

Differential diagnosis

The differential diagnosis of chronic cough and failure to thrive includes the following:
- Pulmonary tuberculosis
- Bronchiectasis (especially following measles, which may also cause chronic diarrhoea)
- HIV infection

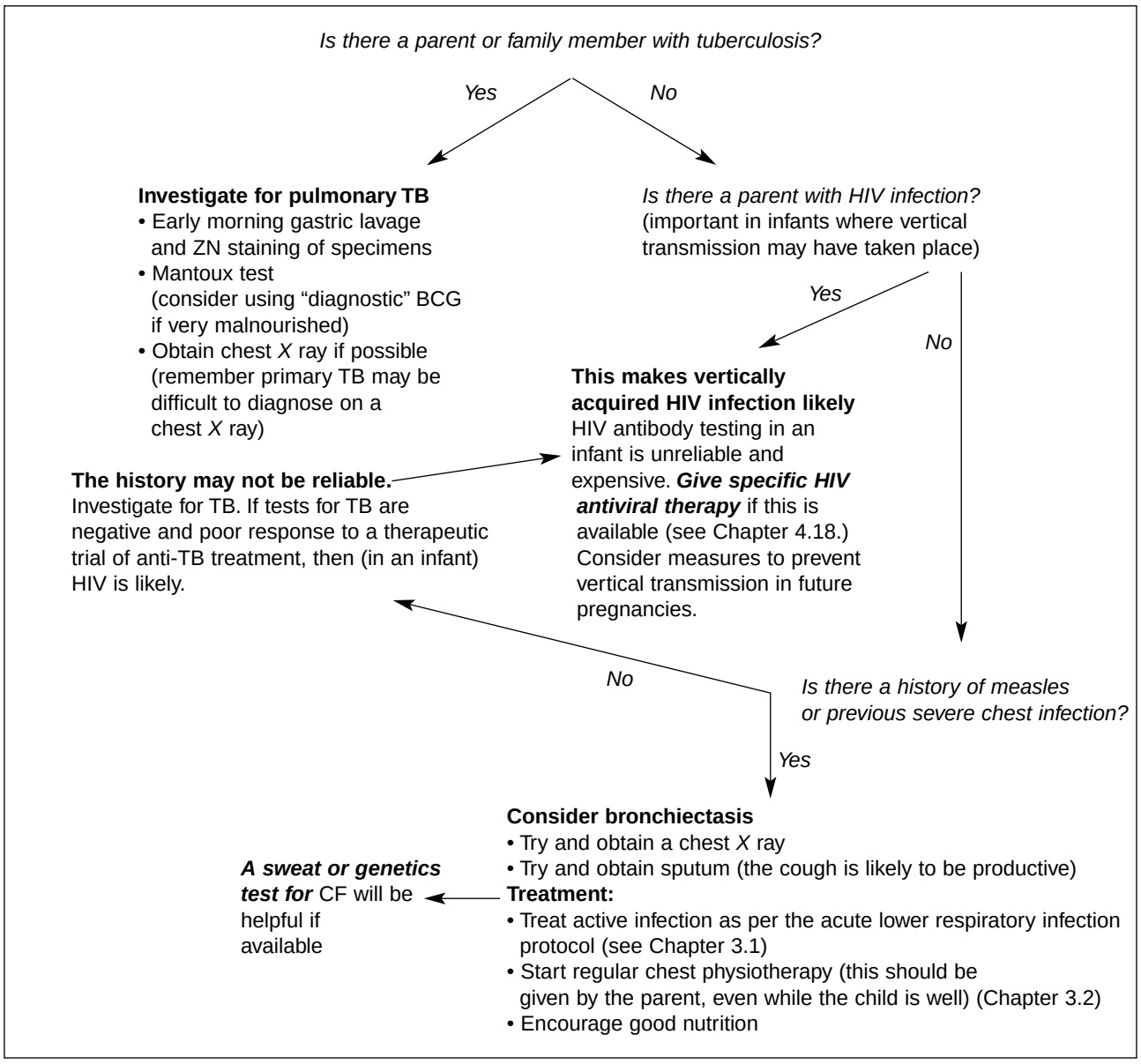


Figure 3.4.1 The differential diagnosis of the child with chronic cough and failure to thrive.

Figure 3.4.1 gives a flow diagram for the investigation of a child with chronic cough and failure to thrive, in areas where pulmonary tuberculosis and HIV infection are prevalent.

Diagnosing cystic fibrosis

The sweat test

This detects the high levels of chloride and sodium in sweat found in CF patients. Ideally, the specialised laboratory and experienced technician needed should be available in at least one hospital in every country. The principle of the test is to allow pilocarpine to diffuse into the skin of the forearm using an electric current (pilocarpine iontophoresis) which stimulates sweating via cholinergic receptors in sweat glands. The sweat is collected on filter paper and the weight, chloride and sodium concentrations are calculated. At least 100 mg of sweat is needed. Values highly suggestive of cystic fibrosis are concentrations of chloride and sodium of greater than 60 mmol/litre and a chloride concentration greater than the sodium.

Genetic tests

These can be performed on very small amounts of blood, collected as a dried blood spot on filter paper. It may be possible to send dried blood spots to a genetics laboratory for analysis. A negative genetic test does not rule out CF (only common genes are tested).

Management

Treatment of children with CF in poor countries has been identified as a priority area by WHO. For practical reasons, children with CF can be seen regularly in a clinic alongside children with bronchiectasis. It is important that the child's parents understand that CF cannot be cured.

However many children can lead active lives with minimal symptoms, provided daily treatment is given and deteriorations are treated promptly.

Pancreatic enzyme supplements

Most children with CF will require pancreatic enzyme supplements (for example Creon, Solvay Healthcare; or Pancrease, Jansen-Cilag). Young infants are given half a capsule per milk feed. Older children may have over 10 capsules per meal. The capsules contain protease, lipase and amylase. The lipase is the most important component in preventing malabsorption. Most brands contain between 5000 and 10 000 units of lipase/capsule. The correct dose of pancreatic enzyme supplements is not necessarily related to age but rather that which is required to control symptoms of steatorrhoea and achieve normal growth. The maximum dose (expressed as units of lipase) is 10 000 U/kg/day.

Fat-soluble vitamins

Give extra fat-soluble vitamins. Appropriate doses are vitamin E 50 mg once daily in infants/young children (0–5 years); 100 mg/day older children (5–12 years); 200 mg/day >12 years. Vitamin E may be given as vitamin E suspension 100 mg/ml or 50 mg tablets. Multivitamin drops, such as Abidec, which contains vitamin A 4000 units/6 ml and vitamin D_2 400 units/6 ml are also required. Give Abidec:

- 0.3 ml/day for newborn infants
- 0.6 ml/day for infants 1–12 months
- 1.2 ml/day for children >1 year

Remember good calorie intake is vital. **Do not restrict fat in the diet.**

Chest physiotherapy (see Chapter 3.2)

Routine, daily chest physiotherapy should be started as soon as the diagnosis is suspected. The most common method is percussion and postural drainage. In young infants this can be performed with the child across their parent's lap whereas in pre-school children a foam "wedge" helps the child achieve the correct position for postural drainage (see Chapter 3.2). The percussion element of the treatment involves firm "clapping" movements with the flat of the hand against the child's chest. This imparts vibrational energy to the chest that helps loosen bronchial secretions. Older children and teenagers should be encouraged to take a more active part in their physiotherapy. A technique incorporating periods of diaphragmatic breathing followed by a forced expiration or "huff" is suitable at this age.

Antibiotics

Children with CF often have intermittent infection with *Staphylococcus aureus* in the first two years of life. In pre-school and primary school age children, infection with *Haemophilus influenzae* may take its place. Finally, in the early teens, chronic infection with *Pseudomonas aeruginosa* may become established. If the results of sputum or "cough swab" cultures are available, then these will allow you to choose an appropriate antibiotic. If not, the likely organisms in the age groups described above, will be a rough guide.

Prophylaxis

In rich countries, a continuous prophylactic antibiotic is often given to children with CF, up until 2 years of age. An antibiotic active against *S. aureus* is usually chosen. In poorer countries this may not be an option, either because the diagnosis is made when the child is over 2 years or because continuous antibiotics are too expensive. However one of the following antibiotics should be prescribed if possible for children under 2 years of age:

Flucloxacillin (oral): 125 mg 4 times daily
Cephadrine: 250 mg twice daily

Treatment of exacerbations

If the cough worsens or the child produces more sputum, a full course of antibiotics (different to the prophylactic drug) should be started and continued for at least two weeks. Longer courses of antibiotics are given than in most other conditions. The following are suitable antibiotics:

- Flucloxacillin, combined with amoxycillin, has good activity against *S. aureus* and *H. influenzae*

Table 3.4.1

Oral flucloxacillin

Age	Dose	Doses/day
<1 year	125 mg	4
1–7 years	250 mg	4
7–12 years	500 mg	4
>12 years	500 mg–1 g	4

Oral amoxycillin

Age	Dose	Doses/day
<1 year	125 mg	4
1–7 years	250 mg	4
>7 years	500 mg	4

- An alternative is chloramphenicol that is active against *S. aureus* and *H. influenzae*. Its activity against *P. aeruginosa* is poor. Children with CF may receive many courses of antibiotics in their lifetime and it is important to limit the number of courses of chloramphenicol that they receive, because of the risk of aplastic anaemia. However, because chloramphenicol is cheap and readily absorbed when given orally, it is justified to use it sparingly in CF.

  The oral dose of chloramphenicol is 12.5 mg/kg 6 hourly

- If *P. aeruginosa* has been identified in sputum or infection is suspected then use one of the following antibiotics:

Table 3.4.2 Oral ciprofloxacin

Age	Dose	Doses/day	Duration
Up to 1 year	7.5 mg/kg/dose	2	2 weeks
1–3 years	62.5 mg	2	2 weeks
3–7 years	125 mg	2	2 weeks
7–12 years	250 mg	2	2 weeks
>12 years			
<50 kg	500 mg	2	2 weeks
>50 kg	750 mg	2	2 weeks

Table 3.4.3 Intravenous or intramuscular gentamicin*

Dose	Doses/day	Duration
3 mg/kg/dose	3	2 weeks

**** Monitor levels if possible:***
Peak: 5–10 micrograms/ml
Trough: <2 micrograms/ml

Patients with CF often have more rapid renal clearance and have lower levels for a given dose than other patients.

If possible combine gentamicin with another anti-pseudomonal antibiotic such as ceftazidime:

Table 3.4.4 Intravenous ceftazidime

Dose*	Doses/day	Duration (infusion)	Duration (course)
50 mg/kg/dose	3	30 min	2 weeks

* Maximum dose 3 g three times daily

What complications may occur in CF?

In addition to those features mentioned above under clinical presentation, the following may occur:

- Haemoptysis
- Pneumothorax
- Bronchiectasis
- Biliary cirrhosis, portal hypertension, and oesophageal varices
- Diabetes mellitus (requiring insulin)
- Infertility (men)
- Women may become pregnant but will need careful management of their chest problems.

With the best care, survival is possible into the fourth decade. Careful management will improve the quality of life greatly for children in poor countries. Sadly, most patients with CF, in any part of the world, ultimately die of respiratory failure.

3.5 Cardiac problems

Christopher Duke and Shakeel A Qureshi

Minimum standards requirements

- Antibiotics for rheumatic fever and endocarditis
- Anti-heart failure drugs
- Anti-arrhythmic drugs and DC conversion
- A centre for specialist care
- Oxygen

This chapter does not attempt to give a detailed description of the many different cardiac pathologies. Instead, it aims to describe the common ways in which cardiac disorders present and provides a means of making a practical diagnosis and starting appropriate treatment. ✔ **Every country or region should have a hospital that can surgically correct the easily curable acquired or congenital heart defects.** This is the assumption here, but the reality is very different. More than 90% of countries do not have such facilities. Investigations or treatments, which are likely to be unavailable or irrelevant in the absence of a specialist cardiac centre in the country, are highlighted in bold italics.

Heart failure

Minimum standards requirements

- Furosemide, potassium, spironolactone
- Captopril
- Digoxin

The term "heart failure" is often used to describe the clinical changes that occur when the cardiac pump cannot meet the demands of the body. This may occur either because the workload imposed on the heart is excessive or because the cardiac pump is weak. The heart may be presented with an excessive workload when it is required to pump a larger volume than usual or when it has to pump against a higher resistance. These two conditions are known as volume loading and pressure loading. In disadvantaged countries, heart failure secondary to volume loading is most commonly caused by severe anaemia or fluid overload (particularly when giving intravenous fluids as part of the treatment for infections or severe malnutrition). In advantaged countries, one of the commonest causes of heart failure secondary to volume loading is congenital heart disease with a large left to right shunt. Cardiac failure secondary to pressure loading is less common and may be caused by congenital heart disease with an obstruction to left heart outflow.

Establishing heart failure

The first priority is to establish that heart failure is present:

- Symptoms of heart failure in infancy
  - Fatigue
  - Poor feeding
  - Breathlessness (particularly during feeds)
  - Sweatiness (particularly during feeds)
- Signs of heart failure in infancy
  - Failure to thrive
  - Tachypnoea
  - Increased respiratory effort
  - Tachycardia >160 bpm
  - Sweating
  - Pallor
  - Hepatomegaly
  - Gallop rhythm
- Additional symptoms of heart failure in the older child
  - Exercise intolerance
  - Shortness of breath on exertion
- Additional signs of heart failure in the older child
  - Elevated jugular venous pressure
  - Basal crepitations on lung auscultation
  - The cut-off for tachycardia is different beyond infancy (>120 bpm 1–5 years; >100 bpm >5 years).

Potential causes

Once cardiac failure has been diagnosed the potential causes should be considered:

In first few weeks

Heart failure in the first few weeks of life is a medical emergency and may be caused by:

- Severe anaemia
- Excessive intravenous fluids
- Renal failure resulting in fluid retention
- Left heart obstruction
- Supraventricular tachycardia

- High output cardiac failure secondary to vascular malformations.

Carefully check the fluid balance to exclude fluid overload. Check renal function and exclude severe anaemia. Perform an ECG to detect supraventricular tachycardia. Examine the baby for cranial and hepatic bruits as cranial and hepatic arteriovenous malformations are a potential (rare) cause of high output cardiac failure. **If these tests are negative refer the child to a paediatric cardiologist as a left heart obstructive lesion is likely and there may be duct dependent systemic circulation (see section below on congenital heart disease).**

Beyond first few weeks

Heart failure beyond the first few weeks of life may be caused by:

- Severe anaemia
- Severe malnutrition
- Excessive intravenous fluids
- Congenital heart disease with increased pulmonary blood flow (left to right shunting lesions and more rarely cyanotic congenital heart defects with high pulmonary blood flow)
- Rheumatic fever
- Myocarditis
- Cardiomyopathy
- Infective endocarditis
- Constrictive pericarditis (rare and most often caused by tuberculosis) (see Chapter 4.10)
- The same causes that present in the first few weeks of life.

Anaemia is a common and often severe problem in disadvantaged countries (see Chapter 3.20). When the haemoglobin falls below 7 g/dl cardiac output must increase to maintain oxygen delivery. Cardiac failure may develop (usually with a haemoglobin less than 5 g/dl) secondary to high output. The treatment is transfusion, but the increase in intravascular volume may precipitate worsening heart failure. Blood must therefore be infused slowly in small boluses and an exchange transfusion may be needed if there is clinical deterioration. Furosemide 1 mg/kg IV should be given during transfusion. Protein-calorie malnutrition is also an important cause of cardiac failure in disadvantaged countries (see Chapter 3.16). Although cardiac failure is unusual at presentation, it may occur after several days of refeeding. Rapid refeeding can cause a hypermetabolic state, demanding an increase in cardiac output which cannot be met by the malnourished heart which has a decreased cardiac reserve. The problem is exacerbated by coexisting anaemia, blood transfusion, inappropriate intravenous fluid administration and high sodium diets. The other common causes of cardiac failure are dealt with individually in the sections below.

Management of heart failure

- Avoid intravenous fluids (especially those containing sodium).
- Calorie supplementation (see Chapter 3.16).
- Nasogastric feeding if inadequate oral intake.
- Bed rest for older children, in a semi-upright position with the legs dependent.
- Oxygen if there is respiratory distress or hypoxaemia due to pulmonary oedema.
- Relieve fever if >38°C.
- In an emergency where there is pulmonary oedema, give furosemide 1 mg/kg IV which should produce a diuresis within 2 hours. If the initial dose is ineffective, give 2 mg/kg IV and repeat after 12 hours if necessary.
- Furosemide 1 mg/kg orally once per day, twice per day or three times per day. Dose frequency should be adjusted to control symptoms.
- Spironolactone 1 mg/kg orally once per day, twice per day or three times per day. Give in combination with furosemide, matching the dose frequency, to enhance diuresis and prevent furosemide-related hypokalaemia.
- If furosemide is used without spironolactone, oral potassium 3–5 mmol/kg/day should be given (supplemental potassium is not required if furosemide is given for less than 4 days).

If more than twice daily diuretics are required, consider captopril. Captopril should be commenced in hospital with a 100 micrograms/kg test dose. The dose should then be increased gradually over a number of days to a maximum of 1 mg/kg three times a day. After the test dose and each increment monitor the blood pressure carefully as hypotension is common. Reduce the dose if hypotension occurs. Monitor renal function (plasma creatinine) carefully as renal failure is a well recognised side effect. Stop captopril if renal impairment develops. Stop spironolactone when the captopril dose is greater than 500 micrograms/kg as both drugs cause potassium retention. Do not give captopril if there is left heart obstruction.

Monitor heart and respiratory rates, respiratory distress and oxygenation regularly during treatment of acute heart failure. It is necessary to both control the symptoms of failure and to determine and treat the underlying cause.

Rheumatic fever

Minimum standards requirements: rheumatic fever

- Penicillin
- Aspirin
- Prednisolone
- Haloperidol/diazepam/lorazepam
- Anti-endocarditis measures

Rheumatic fever is an abnormal immune response to group A streptococcal infection in genetically susceptible individuals. It is most common between the ages of 6 and 16. Symptoms of acute rheumatic fever follow streptococcal

pharyngitis after a latent period of approximately three weeks. There is no definitive test and diagnosis depends on recognition of clinical signs known as the Jones criteria:

Jones criteria for rheumatic fever

Major criteria	Minor criteria
Carditis	Previous history of rheumatic fever
Migratory large joint polyarthritis	Fever
Erythema marginatum	Arthralgia
Subcutaneous nodules	First-degree heart block
Chorea (onset 2–6 months after pharyngitis)	Elevated acute phase reactants: erythrocyte sedimentation rate (ESR)/ C-reactive protein (CRP)

Acute rheumatic fever may be diagnosed when there is evidence of recent infection with group A *Streptococcus* and the child has two major or one major and two minor criteria. Evidence of streptococcal infection is either recent scarlet fever, a positive throat swab (poor reliability because of the high rate of chronic carriage of group A *Streptococcus* in the throat) ***or a raised ASO titre***. If the clinical findings are suspicious and the ASO titre is negative, ***further serological evidence of streptococcal infection can be sought, for example, by performing an antideoxyribonuclease B assay.***

The following may be associated with rheumatic fever:

- *Arthritis*: occurs in approximately 75% of patients (particularly in the knees, ankles, elbows and wrists) and may cause marked pain that results in the child refusing to use the affected limb. It is characterised by its migratory quality (each joint usually remains inflammed for less than a week) and its rapid response to salicylate treatment. Symptoms usually resolve immediately and rarely persist more than 2–3 days once aspirin (up to 120 mg/kg/day in 4–6 divided doses by mouth after food) has been started. It is therefore important to withold anti-inflammatory treatment until signs and symptoms are sufficient to recognise the migratory nature of the polyarthritis.
- *Carditis*: occurs in approximately 50% of patients and often follows arthritis, occuring about 1 week later. The severity of the carditis is often inversely proportional to the severity of the arthritis.
- *Erythema marginatum*: is rarely seen. It consists of irregular "snake like" erythematous macular lesions with an advancing margin that clears in the centre. The rash occurs mainly on the trunk, arms, buttocks and thighs and is neither painful nor pruritic. It occurs early in the disease but may persist for months.
- *Subcutaneous nodules*: are rarely seen unless there is chronic carditis. They are found on the extensor surfaces of large joints, over the spinous processes of the vertebrae and in the occipital region.
- *Sydenham's chorea*: occurs in 10–15% of patients but its onset lags behind the other manifestations by several months. There is rapid involuntary purposeless movement of the face, tongue and extremities, marked emotional lability and muscular incoordination. There may be irregular contractions of the hand muscles and when the hand is outstretched the wrist may be flexed and the fingers dorsiflexed, giving a "silver fork" deformity. Serial evaluation of handwriting may be useful to follow improvement.

Cardiac inflammation may involve the endocardium (valvulitis mostly affecting the mitral and aortic valves), the myocardium (impaired cardiac function) or the pericardium in severe cases (pericarditis). Carditis usually causes no symptoms and is diagnosed during examination of the patient with arthritis or chorea. Examination may reveal an apical pansystolic murmur from mitral regurgitation, a mid-diastolic murmur at the apex from narrowing of the mitral orifice by thickened oedematous cusps or an early diastolic decrescendo murmur from aortic regurgitation. If aortic regurgitation is severe there will be a wide pulse pressure. When myocarditis occurs, it is almost always accompanied by valvulitis and may be marked by tachycardia out of proportion to the fever, cardiac failure and rapid cardiac enlargement on the chest *X* ray. Pericarditis may cause chest pain and a pericardial friction rub. Despite a tachycardia there is usually a prolonged PR interval on the ECG. Severe early carditis is a poor prognostic marker.

Management of acute rheumatic fever

- Supportive care with bed rest during the acute phase.
- Eradicate streptococcal infection (for treatment regime for streptococcal pharyngitis see below).
- Commence aspirin 90–120 mg/day in 4 divided doses.
  - Reduce the dose to two-thirds when there is a clinical response.
  - When the C-reactive protein/erythrocyte-sedimentation rate normalise, taper the aspirin dose over 2 weeks.
- Give prednisolone 2 mg/kg/day (maximum 60 mg/day) in place of aspirin for moderate to severe carditis or pericarditis.
  - If prednisolone is given, continue for 3 weeks then taper dose over a further 2–3 weeks. As the prednisolone dose starts to taper, commence aspirin 50 mg/kg/day in four divided doses and stop aspirin one week after prednisolone is stopped.
- Treat heart failure as described above.
- Endocarditis prophylaxis is needed for dental and surgical procedures after carditis (see below).
- Chorea is usually self limiting but symptoms can be severe. Haloperidol (12.5–25 micrograms/kg twice daily – maximum 10 mg/day for child <12 years and 60 mg/day for child >12 years) may be given to relieve symptoms and benzodiazepines to provide sedation. There may be extrapyramidal side effects from haloperidol.

The disease may be prevented by detecting group A streptococci in cases of pharyngitis (throat swab or ***rapid antigen test***) and treating with either oral penicillin V at a

high dose of 250 mg (children) or 500 mg (adolescents) three times per day for 10 days or one dose of IM benzathine penicillin 0.6 MU for children <27 kg or 1.2 MU for children >27 kg. Once there has been one episode of rheumatic fever a recurrence is likely. The recurrence risk is minimised by giving a monthly dose of IM benzathine penicillin 0.6 MU for children <27 kg or 1.2 MU for children >27 kg or oral penicillin V 250 mg (children) and 500 mg (>12 years) twice daily, after the acute attack, (preferably for life). *For patients allergic to penicillin*, erythromycin 40 mg/kg/day in 2–4 divided doses (maximum 1 g/day) should be given to treat acute infection and erythromycin 250 mg (children <12 years) and 500 mg (children >12 years) both twice daily, given to prevent recurrence.

Long-term consequences of rheumatic fever

After an attack of acute rheumatic fever there may be permanent valve damage. Rheumatic heart disease occurs when acute valve inflammation is followed by scarring and fibrosis, resulting in various degrees of shortening, thickening, rigidity, deformity, retraction and fusion of the valve cusps. The commonest valve lesions are mitral regurgitation, mitral stenosis and aortic regurgitation.

Rheumatic heart disease is most severe and progressive in (1) children who initially have severe carditis (2) children who have recurrent attacks of acute rheumatic fever. The prognosis is more favourable if recurrences are prevented (residual cardiac disease may disappear or improve and valve damage only worsens in a few cases). It is therefore crucial to maintain continuous antibiotic prophylaxis to prevent further valve damage, particularly as children are prone to develop a recurrence after the initial attack (below).

Mitral regurgitation

Mitral regurgitation is the commonest valve lesion in children with rheumatic heart disease. Patients are often asymptomatic during childhood as symptoms are caused by left ventricular failure which may take as long as two decades to develop. However, cases may present before adolescence and mitral regurgitation may be rapidly progressive in regions where the incidence of rheumatic fever is high and recurrent rheumatic fever is common. Mitral regurgitation may be diagnosed by the presence of a blowing apical pansystolic murmur radiating to the left axilla. There may also be a third heart sound and a short low frequency mid-diastolic murmur from increased transmitral flow.

Features of severe mitral regurgitation

- Easy fatigue (caused by low cardiac output)
- Shortness of breath on exertion (caused by pulmonary oedema)
- Hyperdynamic apical impulse and pansystolic murmur
- Apical impulse displaced laterally and inferiorly
- The chest *X* ray demonstrates cardiomegaly and left atrial enlargement (a double density on the right heart border and elevation of the left main bronchus)
- The ECG demonstrates left atrial enlargement (broad bifid P waves in lead II and a prominent negative component to the P in V1) and left ventricular hypertrophy
- Signs of pulmonary hypertension (see below)

If there are features of severe mitral regurgitation, the child should be urgently referred for a paediatric cardiology opinion as surgery is likely to be necessary. ***Ideally all children with mitral regurgitation should be evaluated by echocardiography annually***, as progressive left heart dilation may result in irreversible left ventricular dysfunction if referral is delayed until symptoms develop. Medical treatment should be given for heart failure (captopril is particularly useful) but children who are unwell enough to require this often need either a ***mitral valve repair or a mitral valve replacement with a mechanical valve or bioprosthesis***.

Mitral stenosis

If there is effective antibiotic prophylaxis, mitral stenosis usually develops slowly over 5–10 years and is often not sufficiently severe to cause symptoms in childhood. The reality in countries where there is inadequate prophylaxis and recurrent attacks of rheumatic fever are common is that mitral stenosis may progress much more rapidly and symptoms may be evident 6 months to 3 years after the first attack. Mild stenosis does not cause symptoms, moderate stenosis causes shortness of breath on exertion and severe stenosis causes easy fatigue, shortness of breath at rest, orthopnoea, paroxysmal nocturnal dyspnoea and haemoptysis. Mitral stenosis may be diagnosed by the presence of a low frequency mid-diastolic murmur maximal at the apex. The murmur may be accentuated by exercise and is often accompanied by a loud first heart sound and a diastolic opening snap. The murmur becomes longer as the severity of the stenosis increases. In severe cases there are also signs of pulmonary hypertension.

Signs of pulmonary hypertension

- Left parasternal heave
- Loud second heart sound
- Early diastolic murmur of pulmonary regurgitation at the upper left sternal border
- Elevated JVP and hepatomegaly if there is right heart failure

The chest *X* ray and ECG often show left atrial enlargement when there is moderate mitral stenosis. Radiographic signs of pulmonary oedema may be evident when stenosis is severe. ECG changes of right ventricular hypertrophy and right axis deviation are present when there is pulmonary hypertension. Symptoms should be treated with diuretics and a low-sodium diet. Digoxin is only indicated in rare cases where there is atrial fibrillation secondary to left atrial enlargement. Symptomatic children and children with signs of pulmonary hypertension should be referred for paediatric cardiology review as surgery is often necessary. ***The options for treatment are open or closed mitral commissurotomy, mitral valve replacement and percutaneous catheter balloon mitral commissurotomy***.

Aortic regurgitation

Aortic regurgitation is less common than mitral regurgitation and frequently occurs in combination with mitral valve disease. Affected children usually remain asymptomatic for many years as symptoms only become evident when left ventricular dysfunction develops secondary to chronic left ventricular volume overload. Severe symptomatic aortic regurgitation may however become established within 1–2 years of the initial attack of rheumatic fever if recurrence is not prevented. Once symptoms appear deterioration is often rapid. Symptoms include exercise intolerance, shortness of breath on exertion and chest pain in a few severely affected cases. Examination reveals a blowing decrescendo early diastolic murmur maximal at the mid to lower left sternal border. The murmur is loudest sitting forward with the breath held in expiration.

Signs of moderate to severe aortic regurgitation

- The murmur lengthens and may be throughout diastole
- Hyperdynamic apex
- Apical impulse displaced laterally and inferiorly
- Wide pulse pressure
- Collapsing pulses
- Visible pulsations in the suprasternal notch and neck vessels
- Systolic murmur at the upper right sternal border (from increased aortic valve flow)

If patients are symptomatic or have signs of severe aortic regurgitation they should be referred for paediatric cardiology assessment as surgery may be necessary. Marked cardiomegaly on the chest *X* ray or multiple ventricular ectopics on the ECG should also prompt referral. ***Ideally all children with aortic regurgitation should have an echocardiogram at least annually*** as it is important to assess left ventricular dilation and function to ensure that surgery is carried out before irreversible left ventricular dysfunction develops. Exercise tolerance may be improved by captopril treatment and medical treatment for heart failure may be necessary in severe cases. Surgical options include ***aortic valve reconstruction, aortic valve replacement with an aortic homograft or mechanical valve and transferring the patient's own pulmonary valve to the aortic position (Ross procedure)***.

Cardiomyopathy and myocarditis

Dilated cardiomyopathy and myocarditis are disorders of myocardial contractility.

Both are characterised by the presence of a large poorly contracting heart and usually present as cardiac failure in infancy, childhood or adolescence. It is very difficult to distinguish myocarditis from cardiomyopathy. A history of an acute febrile illness preceding the onset of cardiac failure by about two weeks is suggestive of myocarditis.

Physical signs include poor peripheral perfusion, tachycardia, tachypnoea, increased respiratory effort, hepatomegaly, elevated jugular venous pressure, a gallop rhythm, a displaced apex beat and often a mitral regurgitation murmur. Examine the child for signs of disorders that may cause cardiomyopathy such as myopathies, mitochondrial cytopathies, metabolic disorders and connective tissue diseases.

If possible, ***arrange an echocardiogram to evaluate cardiac function and to exclude coronary artery abnormalities.*** Carry out a 12-lead ECG and a ***24-hour ECG*** to exclude incessant tachycardia as the cause of the poor contractility. If facilities are available, ***screening tests for myocarditis and the rare underlying causes of cardiomyopathy*** can be carried out. Screening tests include a full ***metabolic screen, an autoimmune screen,*** a full blood count, inflammatory markers, blood cultures, blood biochemistry, liver function, ***cardiac enzymes, and a viral screen (stool for enterovirus and serology for coxsackie virus, echo virus, HIV, influenza, parainfluenza, measles, mumps, rubella and Epstein–Barr virus)***. Often the search for an underlying cause is unproductive.

Treatment

Myocarditis and cardiomyopathy are treated in the same way.

- ***When possible refer to specialist centre.***
- Furosemide and spironolactone (see section on heart failure, above).
- Captopril (see section on heart failure, above).
- ***Intubation and ventilation if pulmonary oedema severe.***
- ***Inotropic support if available (dobutamine 5–10 micrograms/kg/minute, dopamine 5–10 micrograms/kg/minute).***
- ***Anticoagulation to prevent thromboembolism if cardiac function is very poor.***

Infective endocarditis

Minimum standards requirements: infective endocarditis

- Penicillin, gentamicin, flucloxacillin, vancomycin, fucidin
- Blood culture
- Cardiac ultrasound
- Long-term venous catheters

Endocarditis should always be suspected in a child with a cardiac defect when there is a fever without a focus. Infection develops on injured areas of endothelium or on abnormal or damaged heart valves. In some cases the onset may be sudden with obvious signs of sepsis and cardiac failure (secondary to valve damage). However, in most cases the onset is insidious and the diagnosis is unclear. There may be fever, malaise, fatigue, arthralgia, anorexia and weight loss. It may occur in a child previously thought to have a normal heart (likely to have undiagnosed congenital heart disease or valve damage from rheumatic fever).

Signs of endocarditis

- Pyrexia
- Microscopic haematuria
- Splenomegaly
- Changing heart murmur
- Petechiae
- Neurological abnormalities (caused by cerebral abscess or infarction)
- Splinter haemorrhages, Janeway lesions, Osler's nodes and Roth's spots (characteristic but rare)

The diagnosis is made by isolating microorganisms from the blood. At least three sets of blood cultures must be obtained from different puncture sites. If possible, antibiotics should be witheld until multiple blood cultures have been obtained and should only be started when the diagnosis is clear or there is a pressing clinical urgency. Blood cultures will be negative in 10–15% of cases. ***Echocardiography helps*** to make the diagnosis if vegetations are seen, but a negative echocardiogram does not exclude the diagnosis.

Organisms most commonly isolated in endocarditis

- *Streptococcus viridans* (commonest overall)
- *Staphylococcus aureus* (most cases of fulminant endocarditis)
- Coagulase-negative staphylococci (if the patient has a central line or is immunocompromised)

For *Streptococcus viridans*, IV benzylpenicillin 25 mg/kg 6 hourly and gentamicin 2.5 mg/kg 8 hourly are given for two weeks, followed by a further two weeks of oral amoxycillin. If the organism is *Staphylococcus aureus*, IV flucloxacillin 25 mg/kg 6 hourly is given for 4 weeks, coupled with IV gentamicin 2.5 mg/kg 8 hourly (or sodium fucidate) 6–7 mg/kg 8 hourly for the first two weeks. Vancomycin 10 mg/kg 6 hourly is used in place of flucloxacillin if the organism is a coagulase negative *Staphylococcus* or the patient is allergic to penicillin. The success of treatment is monitored by symptoms and inflammatory markers (WBC and CRP). ***Surgery is necessary when the organism cannot be eradicated, when there is evidence of embolisation, where there is a large mobile vegetation at risk of embolisation or when there is severe cardiac failure from valve damage.*** **(See page 394 for gentamicin dosage.)**

Arrhythmias

Minimum standards requirements

- ECG monitoring
- Adenosine
- Synchronised DC conversion
- Digoxin/propanolol

Supraventricular tachycardia

Supraventricular tachycardia (SVT) is the commonest tachyarrhythmia in childhood. It may present with poor systemic output and heart failure in infancy or palpitations and dizziness in later childhood. The tachycardia is usually narrow complex and the rate is usually faster than 230 beats/minute, distinguishing it from sinus tachycardia. The commonest cause is an abnormal bundle of muscle fibres bridging from the atrium to the ventricle (known as an "accessory pathway"). The accessory pathway may allow normal atrial impulses to conduct to the ventricles prematurely causing a slurred stroke before the QRS complexes on the ECG known as a delta wave (Wolff-Parkinson-White syndrome). The pathway also allows electrical depolarisation to pass in an abnormal circuit, usually forward across the atrioventricular node and back across the pathway, then forward across the atrioventricular node again, setting up a form of tachycardia known as "re-entry". Tachycardia secondary to abnormally rapid atrial discharge (atrial flutter or atrial ectopic tachycardia) is rarer in childhood.

Management of SVT

- Record a 12-lead ECG during the tachycardia.
- Record a rhythm strip (as many leads as possible) as attempts are made to terminate the tachycardia.
- In the infant try to terminate the SVT by facial immersion in ice-cold water for 1–2 seconds.
- If there are no symptoms and the attacks are short-lived and resolve spontaneously in the older child no treatment is needed.
- To terminate symptomatic or prolonged attacks of SVT in the older child try vagal manoeuvres such as ice-cold packs on the face, the Valsalva manoeuvre and carotid sinus massage.
- If tachycardia persists obtain IV access (via a large antecubital vein if possible) and give a rapid bolus of adenosine 50 micrograms/kg followed by a rapid saline flush. **Adenosine may cause bronchospasm and is contraindicated in asthma.** ✔
- If the SVT is not terminated give a larger dose of adenosine, increasing by 50 micrograms/kg with each dose until a maximum dose of 250 micrograms/kg is reached (maximum 12 mg).
- If the mechanism is not re-entry, transient atrioventricular block caused by adenosine should reveal underlying rapid atrial discharge (the tachycardia will then continue).
- In infancy, digoxin can be used to treat an acute SVT that does not respond to adenosine, providing the child is haemodynamically stable. A loading dose of digoxin is given (loading dose 20 micrograms/kg) immediately, then 10 micrograms/kg at 8 hours and 10 micrograms/kg at 16 hours, doses can be given orally or IV. IV doses should be infused over 30 minutes and adenosine is then repeated if the tachycardia has not terminated.
- Beyond 2 years of age and for treating acute SVT that does not respond to adenosine, give verapramil as follows: age 2–5 years 15 micrograms/kg; 5–10 years 50 micrograms/kg; 10–15 years 100 micrograms/kg. Give all doses of verapramil over 15 minutes. Stop giving when

✔ tachycardia terminates. Only give if the child is haemodynamically stable (**never give a beta-blocker and verapramil together**).

- If the tachycardia persists and there is haemodynamic instability, carry out DC cardioversion (ideally QRS synchronised but asynchronised if that is not possible).
- In infants digoxin (4 micrograms/kg twice per day after loading dose is given, ***if possible monitor plasma levels***) or propranolol (250 micrograms −1 mg/kg orally four times per day, titrate to response) can be given to prevent further SVTs.
- Propranolol (dose as above orally), atenolol (1mg/kg once per day) or flecainide (1mg/kg twice per day) can be used to prevent further SVTs in older children.

DC Conversion

Use paediatric paddles for a child under 10 kg. Place one paddle over the apex of the heart in the mid-axillary line and the other immediately below the clavicle just to the right of the sternum. If there are only adult paddles and the child is less than 10 kg place one on the back and one over the lower chest anteriorly. The first shock should be 0.5 joules/kg and subsequent shocks should be increased stepwise to a maximum of 2 joules/kg.

Congenital complete heart block

- Consider in any newborn who has a consistent bradycardia.
- P waves are dissociated from QRS complexes on the 12-lead ECG.
- Perform an ***echocardiogram*** to exclude structural heart disease.
- Monitor the heart rate for 24–48 hours.
- Assess perfusion, blood pressure and examine for signs of heart failure.
- ***Arrange for a permanent pacemaker if there is inadequate cardiac output, heart failure, structural heart disease or the heart rate is <50/min.***

Atropine 20 micrograms/kg or isoprenaline infusion 100–1000 nanograms/kg/min can be used for emergency treatment of severe bradycardia with inadequate cardiac output.

Congenital heart disease

Minimum standards requirements

- Oxygen
- Morphine/propranolol
- Inotropes: dopamine, dobutamine, epinephrine
- Antibiotic prophylaxis
- Drug treatment for heart failure

Congenital heart disease occurs in 5–8 per 1000 live births. Congenital heart disease may present as:

- Cyanosis in the newborn period
- Cyanosis in the older infant

Incidence of common congenital cardiac defects (UK data)	
Ventricular septal defect (VSD)	32%
Persistent arterial duct (PDA)	12%
Pulmonary stenosis	8%
Atrial septal defect (ASD)	6%
Coarctation of the aorta	6%
Tetralogy of Fallot	6%
Aortic stenosis	5%
Transposition of the great arteries	5%
Hypoplastic left heart syndrome	3%
Atrioventricular septal defect (AVSD)	2%

- Cardiovascular collapse in the newborn period
- Cardiac failure in infancy
- An asymptomatic murmur

This section explains how to recognise the presence of congenital heart disease in each of these clinical scenarios and provides enough information to make a working diagnosis. Management decisions can then be made when modern imaging techniques are not immediately available.

The cyanotic newborn

Is there a cardiac problem?

When a child is referred as a "blue baby" first check to see whether there is genuine central cyanosis. Examine the lips and tongue for blue discoloration and confirm the clinical impression by measuring the oxygen saturation (less than 94% is abnormal). If there is central cyanosis this may have a cardiac or respiratory cause. Differentiation between these can usually be determined from the clinical features described in Table 3.5.1.

Table 3.5.1 Features suggesting the cause of central cyanosis

Cardiac features	Respiratory features
Term baby	Premature
Tachypnoea but no respiratory distress	Respiratory distress
May have cardiac signs on examination	Chest *X* ray: abnormal lung fields
Arterial blood gas: Po_2 ↓, Pco_2 ↓ or normal	***Arterial blood gas: Po_2 ↓, Pco_2 ↑ or normal***
Fails hyperoxia test	Passes hyperoxia test

Persistent pulmonary hypertension of the newborn (PPHN) may mimic cyanotic congenital heart disease using the clinical criteria described in Table 3.5.1. However, it is usually possible to distinguish PPHN on the basis of the history. In PPHN there is often a history of fetal distress, resuscitation is frequently needed at birth, improvements in oxygenation may be possible ***after intubation and ventilation***, there may be neurological signs and saturations in the right arm may be significantly higher than in the feet (suggesting right to left shunting across the arterial duct).

How to perform the hyperoxia test

- Ensure there is good IV access.
- Monitor oxygen saturations continuously.
- Give 100% oxygen for 10 minutes.
- ***Take an arterial blood gas in the right arm (preductal).***
- ***If $Po_2 < 20$ kPa (150 mmHg), a cardiac cause of cyanosis is likely (the test is "failed").***
- ***If $Po_2 > 20$ kPa (150 mmHg), a respiratory cause of cyanosis is likely (the test is "passed")***
- Although pulse oximetry cannot reliably be used in place of an arterial blood gas, a resting saturation $< 80\%$ and a saturation $< 90\%$ after 10 minutes in 100% oxygen suggests cyanotic heart disease requiring early intervention.
- In theory, oxygen administration can cause closure of the arterial duct, precipitating profound hypoxaemia in some types of cyanotic congenital heart disease, but this is not usually observed in practice.
- ***Prostaglandin E (which opens the duct) should be available at the time of the test and should be given if oxygenation deteriorates.***

What type of cyanotic cardiac defect is present?

If cyanotic congenital heart disease is suspected, an attempt should be made to categorise the defect. It is not necessary to make an exact anatomical diagnosis to decide on the appropriate initial treatment. Cyanotic cardiac defects can be divided into three broad categories, based on physiology. The defect should be placed into one of the three categories using the available clinical information. Immediate management decisions can then be made.

1. **Low pulmonary blood flow**
   In defects where there is low pulmonary blood flow the physiology is the same whatever the precise anatomy of the defect. Deoxygenated blood returning from the systemic veins cannot flow through the right side of the heart to the lungs. Pulmonary blood supply is therefore via the arterial duct. The deoxygenated blood from the right side of the heart shunts to the left side of the heart (either via an atrial or a ventricular septal defect) and the left ventricle receives both deoxygenated blood from the right heart and oxygenated blood from the pulmonary venous return. Blood entering the aorta is therefore not fully oxygenated and the child appears cyanosed. If the duct closes the infant becomes profoundly cyanosed and is unlikely to survive unless pulmonary blood flow is rapidly restored. This is duct dependent pulmonary circulation, an example of which is shown in Figure 3.5.1 (a diagram of the circulation in pulmonary atresia with intact ventricular septum).
2. **Complete transposition of the great arteries (TGA)**
   Here the aorta arises from the right ventricle and the pulmonary artery from the left ventricle (Figure 3.5.2).

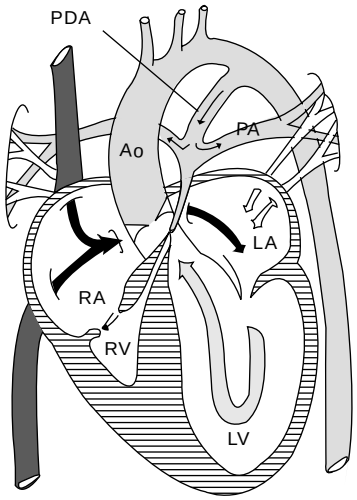


Figure 3.5.1 Circulation in pulmonary atresia with intact ventricular septum. Example of low pulmonary blood flow

RV = right ventricle; PA = pulmonary artery;
LV = left ventricle; Ao = aorta;
RA = right atrium; PDA = patent arterial duct.
LA = left atrium;

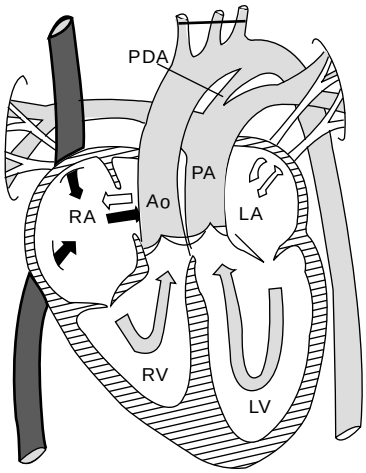


Figure 3.5.2 Transportation of the great arteries. For key, see Figure 3.5.1. Example of complete transposition

Systemic venous return enters the right side of the heart and is recirculated to the systemic arteries. Pulmonary venous return enters the left side of the heart and is recirculated to the lungs. Oxygenated and deoxygenated blood are therefore separated in two parallel circuits. Oxygenated blood enters the systemic circulation only when there is mixing between the two circuits. Mixing occurs at atrial level (across the foramen ovale) and at ductal level (while the duct remains open). Systemic oxygen saturation reflects the amount of mixing (which in turn depends on the size of these communications). If the atrial communication is small oxygenation may therefore be duct dependent.

Table 3.5.2 Features that help to distinguish the three types of cyanotic heart defect

	Low pulmonary blood flow	Complete TGA	Common mixing lesion
Po_2 at rest	Often <35 mmHg	Often <35 mmHg	Often >45 mmHg
SaO_2 at rest	$<80\%$	$<80\%$	80–90%
Po_2 hyperoxia test	Often <50 mmHg	Often <50 mmHg	75–200 mmHg
SaO_2 hyperoxia test	$<90\%$	$<90\%$	90–100%
Chest *X* ray	Reduced pulmonary vascular markings	Normal or increased pulmonary vascular markings with or without narrow mediastinum	Normal or increased pulmonary vascular markings

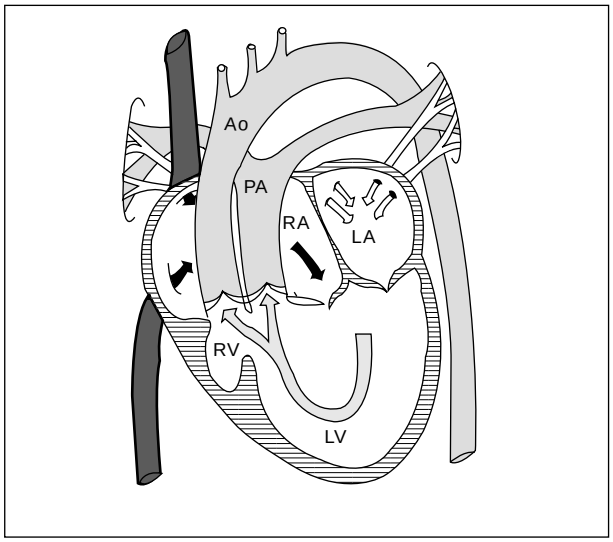


Figure 3.5.3 Circulation in double-inlet left ventricle. For key, see Figure 3.5.1. Example of common mixing lesions

3. **Common mixing lesions**
   In common mixing lesions oxygenated pulmonary venous blood and deoxygenated systemic venous blood mix in one of the cardiac chambers. An example is shown in Figure 3.5.3 (a diagram of the circulation in double inlet left ventricle). The systemic output is therefore only partly oxygenated. The relative amounts of pulmonary and systemic blood in the mixture determine the oxygen saturation and the mode of presentation. If pulmonary blood flow is high cyanosis is minimal and the child usually presents at about two months of age in heart failure. If pulmonary blood flow is low (the complex lesion may coexist with pulmonary stenosis) cyanosis is severe and is often detected early.

The defect may be placed into one of the three categories using Table 3.5.2. The table describes the typical clinical findings in each category. These guidelines assist the clinician but are not infallible and the nature of the defect is sometimes only clear after echocardiography.

Although it is not imperative to reach a more specific diagnosis, an anatomical diagnosis can sometimes be made using clinical information and simple investigations.

Cyanotic defects with low pulmonary blood flow

- Critical pulmonary stenosis
- Pulmonary atresia with intact ventricular septum
- Tetralogy of Fallot (with severe right ventricular outflow tract obstruction)
- Pulmonary atresia with ventricular septal defect
- Absent right atrioventricular connection

- **Critical pulmonary stenosis**
  There may be an ejection systolic murmur at the upper left sternal border (from right ventricular outflow tract flow) or a murmur at the lower left sternal border (tricuspid regurgitation).
- **Pulmonary atresia with intact ventricular septum**
  There is either no murmur or a murmur at the lower left sternal border (tricuspid regurgitation). There is often cardiomegaly on the chest *X* ray. The praecordial leads on the ECG usually show decreased right ventricular voltages (small R waves in V1 and V2) and dominant left ventricular voltages (prominent S waves in V1 and 2 and prominent R waves in V5 and 6). The QRS axis is usually between 0 and $+90$ degrees.
- **Tetralogy of Fallot and pulmonary atresia with ventricular septal defect**
  In both defects the cardiac silhouette on the chest *X* ray has a concavity on the left heart border where there is usually a convexity produced by the right ventricular outflow tract and pulmonary artery. The ECG shows dominant right ventricular voltages (normal neonatal RS progression). The two pathologies are similar, but in Fallot's tetralogy the right ventricular outflow tract is patent, albeit narrow, generating a high-pitched ejection systolic murmur at the upper left sternal border.
- **Absent right atrioventricular connection (also known as tricuspid atresia)**
  There is often a long harsh systolic murmur (this may arise from a restrictive ventricular septal defect or pulmonary stenosis). The praecordial leads on the ECG show decreased right ventricular voltages and dominant left ventricular voltages. The QRS axis is characteristically directed to the left and superiorly between 0 and -90 degrees.
- **Complete transposition of the great arteries**
  There is usually no murmur. The ECG shows dominant right ventricular voltages (normal neonatal RS

progression). Therefore, if a newborn is severely cyanosed and otherwise appears clinically normal, actively look for a narrow mediastinum on the chest *X* ray to help make the diagnosis.

Management of defects with low pulmonary blood flow or complete TGA

- Do not give oxygen after the hyperoxia test as it may precipitate ductal closure.
- ***Start intravenous prostaglandin E (PGE) to maintain ductal patency. There are two formulations, prostaglandin E_1 and prostaglandin E_2 which is much less expensive. Commence PGE_1 or PGE_2 at 10 nanograms/kg/min.***
- ***PGE infusion is made up by adding 30 micrograms/kg of prostaglandin to 50 ml of 5% glucose or 0.9% saline (if pump run at 1 ml/hour = 10 nanograms/kg/min).***
- ***PGE often causes hypoventilation and apnoea. If oxygen saturations start to fall after PGE has been started assess respiratory effort. If respiration is shallow or slow intubate and ventilate in air.***
- ***If oxygen saturations start to fall after PGE is started and respiratory effort appears adequate, increase PGE dose stepwise until a response is seen (maximum dose 100 nanograms/kg/min for PGE_1 and PGE_2).***
- ***If saturations are initially very low and fail to improve increase the PGE dose stepwise until a response is seen.***
- Arrange for an urgent paediatric cardiology review and transfer to a cardiac centre.
- ***Defects with poor pulmonary blood flow usually require a systemic to pulmonary artery shunt (modified Blalock–Taussig shunt) to provide a stable pulmonary blood supply.***
- ***TGA often requires enlargement of the interatrial communication by balloon atrial septostomy, followed by an arterial switch operation if surgical expertise is available.***

Management of common mixing lesions

- Monitor the child on the neonatal unit.
- Arrange for an ***echocardiogram as soon as possible to*** define the anatomy.
- ***If oxygen saturations fall progressively to less than 70% commence PGE*** and arrange for an urgent paediatric cardiology review.
- Once the anatomy is defined it may be possible to discharge the baby without further treatment (after paediatric cardiology advice has been obtained).

The older infant with cyanosis

Is there a cardiac problem?

When an older infant presents with cyanosis, cardiac pathology is likely if:

- respiratory distress is not severe
- ***there is no carbon dioxide retention***
- respiratory pathology is not evident on the chest *X* ray
- The cardiovascular examination is abnormal (see below).

What type of cardiac defect is present?

The cyanotic defects that commonly present after the neonatal period are tetralogy of Fallot and cyanotic defects with high pulmonary blood flow. They may escape detection at birth because cyanosis is initially only mild. In tetralogy of Fallot there is right ventricular outflow tract obstruction and a large ventricular septal defect (VSD) (right ventricular hypertrophy and aortic override are the other components of the tetralogy). The right ventricular outflow tract obstruction limits blood flow to the pulmonary arteries, causing deoxygenated blood to shunt right to left across the VSD, resulting in cyanosis. With time the right ventricular outflow tract obstruction usually becomes more severe, causing further reductions in pulmonary blood flow, more right to left shunting and increasing cyanosis.

In cyanotic defects with high pulmonary blood flow (mostly common mixing defects) pulmonary flow increases as pulmonary vascular resistance decreases over the first few weeks of life, resulting in progressively worsening cardiac failure.

Cyanotic defects with high pulmonary blood flow

Truncus arteriosus
Total anomalous pulmonary venous connection
Double inlet left ventricle
Absent right atrioventricular connection with large ventricular septal defect
Pulmonary atresia with large or multiple aortopulmonary collateral arteries
Transposition of the great arteries with large ventricular septal defect

- **Findings in tetralogy of Fallot**:
  - May present with increasing cyanosis
  - May present with an ejection systolic murmur at the upper left sternal border
  - Reduced pulmonary vascular markings on chest *X* ray
  - Children are often asymptomatic but there may be sudden periods of increased cyanosis known as "hypercyanotic spells".

In the management of tetralogy of Fallot, the anatomy should ideally be confirmed by echocardiography, preferably within a few weeks of presentation, and surgical correction should be carried out between 6 and 12 months of age (although it can be carried out later).

- **Characteristics of hypercyanotic spells**:
  - Spells often occur on waking from sleep or after feeding
  - The infant becomes restless and agitated
  - There is increased cyanosis and pallor
  - Respiration is often rapid and shallow
  - In severe spells crying is followed by limpness or loss of consciousness

- They usually last 1–5 mins but may last longer when severe
- The ejection systolic murmur shortens or becomes inaudible.

✔ **Hypercyanotic spells may be life threatening. If a child starts to have spells, discuss with a paediatric cardiologist immediately as this is an indication for urgent surgery.**

- If spells are more than a few minutes in duration treat urgently as follows:
  - Knee–chest position
  - Oxygen by face mask
  - IV or IM morphine 100 micrograms/kg (or IV ketamine 1 mg/kg)
  - IV bolus 0.9% saline 10–20 ml/kg
  - IV propranolol 50 micrograms/kg (have isoprenaline ready in case of excessive beta–blockade)
  - General anaesthesia in intractable cases
  - Epinephrine may make them worse.
- **Findings in defects with high pulmonary blood flow**:
  - May present with cardiac failure 2–6 weeks of age
  - Active praecordium
  - Murmur usually present (may be systolic, diastolic or continuous)
  - Increased pulmonary vascular markings on chest *X* ray.

In the management of cyanotic defects with high pulmonary blood flow, ***define the anatomy by echocardiography.*** Manage cardiac failure medically (see below). ***Surgical correction or pulmonary artery banding will be necessary in most cases.***

Neonatal cardiovascular collapse

Is there a cardiac problem?

When a child presents in shock in the first month of life, the working diagnosis is often dehydration or sepsis. The following features may suggest a cardiac rather than a non-cardiac cause for the poor systemic output:

- Collapse in the first two weeks of life
- Poor feeding, lethargy and tachypnoea prior to collapse
- Hepatomegaly
- Pulmonary oedema and cardiomegaly on chest *X* ray
- Lack of response to intravascular volume expansion.

What type of cardiac defect is present?

Left heart obstruction is the most likely cardiac cause of cardiovascular collapse with low systemic output in the first two weeks of life.

Left heart obstruction

- Critical aortic stenosis
- Hypoplastic left heart syndrome (HLHS)
- Coarctation of the aorta
- Interrupted aortic arch

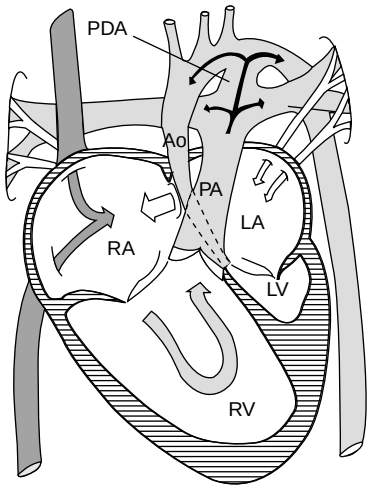


Figure 3.5.4 Hypoplastic left heart syndrome. For key, see Figure 3.5.1.

In **hypoplastic left heart syndrome** all of the left heart structures are small (Figure 3.5.4). There is insufficient forward flow through the left ventricle and the aortic valve to support the systemic circulation. Pulmonary venous return cannot pass through the left heart so crosses the atrial septum and enters the right atrium, mixing with systemic venous return. Mixed pulmonary and systemic venous blood enters the right ventricle and is pumped to the pulmonary arteries and also across the arterial duct to supply the systemic circulation. Ductal flow passes to the descending aorta and retrogradely around the aortic arch to supply the head and neck vessels and the coronary arteries. Ductal flow is not fully oxygenated so there is a degree of central cyanosis. When the duct closes the cardiac output falls precipitously, the infant becomes shocked and cardiac failure develops. This is duct dependent systemic circulation. The haemodynamics are the same in **critical aortic stenosis**.

Coarctation of the aorta consists of a narrowing in the descending aorta close to the aortic end of the arterial duct. Contractile tissue may extend from the duct into the aorta so that when the duct closes it draws in the adjacent section of aorta causing obstruction. Flow to the head and neck vessels is maintained but flow to the lower body distal to the coarctation site is dramatically reduced. The infant becomes shocked and acidotic. Cardiac failure develops secondary to high systemic afterload. This is also an example of the systemic circulation depending on ductal patency (although systemic blood flow may not directly depend on a right to left shunt through the duct). In **interrupted aortic arch** perfusion to the lower part of the body depends on right to left ductal flow and presentation is similar to coarctation.

The following features help to distinguish between the lesions:

- If all of the pulses are weak or absent consider HLHS or critical aortic stenosis
- If the right arm pulses are palpable and the femoral pulses are weak or absent consider coarctation or interrupted aortic arch (note however that all pulses may initially be impalpable if the cardiac output is poor)

- If four limb blood pressures demonstrate significantly lower blood pressures in the legs than the right arm (a gradient of more than 20 mmHg), consider coarctation or interrupted aortic arch
- Coarctation often presents towards the beginning of the second week of life
- HLHS often presents in the first two days of life
- In HLHS, the ECG shows reduced left ventricular voltages (small R waves V5 and V6).

Other cardiac causes of cardiovascular collapse in the first few weeks of life are supraventricular tachycardia (SVT) and cyanotic congenital heart disease with duct-dependent pulmonary blood flow (when the duct closes the ensuing profound hypoxaemia causes acidosis and cardiovascular collapse). SVT should be evident on the ECG and cyanotic heart disease should be suspected when the oxygen saturation remains low after instituting the management detailed below for left heart obstruction.

Emergency management of low systemic output secondary to left heart obstruction

- Check ECG (to exclude SVT as a cause of collapse).
- Peripheral intravenous access if not already established (if intravenous access is difficult, then intraosseous access should be obtained).
- Fluid bolus 10 ml/kg (0.9%) N saline if not already given.
- ***Intubate and ventilate if significant respiratory distress (high PEEP 8–10 cmH_2O)***
- ***Commence prostaglandin E_1 or E_2 at 100 nanograms/kg/min (give for 30 minutes then reduce to 25 nanograms/kg/min, reducing again to 10 nanograms/kg/min when stabilised).***
- ***Admit to paediatric ICU/HDU.***
- Check blood sugar, FBC, U&E, coagulation, ***calcium, magnesium*** and correct abnormalities.
- Take blood cultures and treat with intravenous antibiotics as sepsis cannot be excluded.
- ***Check arterial blood gas (right arm if possible).***
- Correct acidosis with IV sodium bicarbonate 4.2%; ***ideally intubated and ventilated.***
- Give IV furosemide 1 mg/kg if chest *X* ray shows pulmonary oedema.
- ***Central venous access and arterial line.***
- Reassess whether further intravascular volume needed (give if CVP low).
- Dopamine 5–10 micrograms/kg/min if perfusion remains poor or blood pressure remains low.
- Epinephrine 100 nanograms to 2 micrograms/kg/min if perfusion remains poor or blood pressure low (by central venous access only).
- ***Ask for urgent paediatric cardiology review and advice.***

Congenital heart disease causing heart failure

The causes of cardiac failure are detailed at the start of the chapter. If non-cardiac causes and acquired cardiac problems are unlikely on the basis of the clinical findings, congenital heart disease must be considered. If congenital heart disease is possibly the cause of the cardiac failure, refer to a paediatric cardiologist without delay as many congenital heart defects are eminently treatable.

In the first few weeks of life left heart obstruction is the commonest type of cardiac defect causing cardiac failure and requires urgent cardiology assessment. Beyond the first few weeks of life, left to right shunting lesions are a far more common cause. Rarely, failure may be caused by a cyanotic congenital heart defect with high pulmonary flow. Examine the child for cyanosis and take the oxygen saturation. It should be possible to detect those children with cyanotic defects immediately (note however that children with AVSD are sometimes mildly desaturated). Having excluded cyanosis, next attempt to detect the children with left to right shunts, looking for the following features which are present in significant shunts:

- Hyperdynamic praecordial impulse
- Apical impulse displaced laterally and inferiorly
- Apical mid-diastolic murmur (from increased flow across the mitral valve)
- Loud second heart sound (from increased pulmonary artery diastolic pressure)
- Cardiomegaly and increased pulmonary vascular markings on the chest *X* ray.

Common left to right shunting lesions that cause heart failure
Large ventricular septal defect (VSD)
Atrioventricular septal defect with large ventricular component (AVSD)
Large persistent arterial duct (PDA)

If there are signs of heart failure without signs of a large shunt, a left heart obstructive lesion should be considered.

The physiology of left to right shunts

A large defect between the ventricles or great arteries allows free communication between the left and right sides of the heart. Left and right heart pressures therefore equalise and pulmonary artery pressure is maintained at systemic level. The pulmonary vascular resistance then determines the pulmonary blood flow. In the newborn period the pulmonary vascular resistance remains high, limiting the pulmonary blood flow and the left to right shunt across the defect. Over the first 6 weeks of life the pulmonary vascular resistance gradually falls, allowing the pulmonary blood flow and the left to right shunt to increase. This gives rise to heart failure which usually appears at about 2–4 weeks of age. If the pulmonary arteries are exposed to high pressure and flow for a prolonged period pulmonary vascular disease develops. This normally becomes significant between 12 and 18 months of age. High pulmonary vascular resistance secondary to pulmonary vascular disease reduces the left to right shunt and symptoms gradually resolve. Eventually pulmonary resistance becomes so high that flow across the defect becomes right to left and cyanosis develops (Eisenmenger syndrome). The pulmonary artery pressure remains high throughout, it is only the amount of flow through the lungs that changes.

✔ **If there is evidence of a large left to right shunt refer the child to a paediatric cardiologist within a few weeks. These signs must not be missed as a remediable cardiac defect is rendered inoperable by delay.**

Although it is not imperative to make a more specific diagnosis, the following clinical features discriminate between the three most common left to right shunts:

- The persistent arterial duct has a continuous murmur maximal in the left infraclavicular area
- A large ventricular septal defect has a quiet pansystolic murmur maximal at the lower left sternal border radiating to the lower right sternal border. There may also be a soft ejection systolic murmur at the upper left sternal border from increased flow across the pulmonary valve
- An atrioventricular septal defect with a large ventricular component may have a pansystolic murmur like a ventricular septal defect or may have a blowing pansystolic murmur at the lower left sternal border or apex from atrioventricular valve regurgitation. The ECG shows a characteristic superior QRS axis (between −30 and −180 degrees).

Asymptomatic murmurs

When a child presents with an asymptomatic murmur, first examine for cyanosis and take the oxygen saturation. If there is desaturation refer the child for an ***echocardiogram*** as cyanotic congenital heart disease requires a detailed anatomical assessment. Tetralogy of Fallot is the most likely diagnosis. If cyanosis is excluded, the child may have an innocent cardiac murmur or one of the following defects:

- Left to right shunts:
  - Small to moderate sized VSD
  - Small to moderate sized PDA
  - Atrial septal defect (ASD)
  - Partial AVSD
- Left or right heart obstruction
  - Pulmonary stenosis
  - Aortic stenosis
  - Coarctation of the aorta

Innocent murmurs are characterised as follows:

- The Still's murmur is a vibratory short systolic murmur heard at the lower left sternal border or apex
- The venous hum is a soft continuous murmur heard best below the clavicles and is abolished by pressure over the jugular vein or lying down with the neck flexed
- The pulmonary flow murmur is a soft ejection systolic murmur at the upper left sternal border and may be confused with an ASD or mild pulmonary stenosis
- The neck bruit is an ejection systolic murmur maximal above the clavicle and may be confused with aortic stenosis, though it is often abolished by shoulder hyperextension.

The cardiac defects are characterised as follows:

- In coarctation the right arm blood pressure is often elevated, the femoral pulses are weak or impalpable and there is brachiofemoral delay.
- The PDA has a continuous murmur loudest in the left infraclavicular region.
- The VSD has a harsh pansystolic murmur loudest at the lower left sternal border radiating to the lower right sternal border.
- Aortic stenosis, pulmonary stenosis, ASD and partial AVSD all have an ejection systolic murmur at the upper left sternal border.
- In aortic stenosis the ejection systolic murmur is harsh and may be heard at the upper right and left sternal border. The murmur radiates to the carotid arteries and there is often a carotid thrill. There may be an ejection click at the apex if the stenosis is at valvular level.
- In pulmonary stenosis the ejection systolic murmur is harsh and radiates to the back. There may be an ejection click along the left sternal border if the stenosis is at valvular level.
- In an ASD there is a soft ejection systolic murmur at the upper left sternal border from increased flow across the pulmonary valve. There is sometimes a fixed widely split second heart sound and there may be a mid-diastolic murmur at the lower left sternal border (from increased flow across the tricuspid valve) when the left to right shunt is large.
- In partial AVSD there is an abnormal atrioventricular valve and a defect in the atrial septum. There may be a blowing pansystolic murmur at the lower left sternal border or apex from atrioventricular valve regurgitation. The ejection systolic murmur may mimic an ASD but the defect is distinguished by a superior QRS axis on the ECG.

Unless the murmur is clearly innocent, perform an ECG and chest *X* ray. Right ventricular hypertrophy (RVH) is indicated by an R wave in V1 > 98th centile for age (≥ 20 mm is always abnormal), a neonatal RS progression beyond the neonatal period (dominant R waves in V1 and dominant S waves in V6) or an upright T wave in V1 before teenage years. Left ventricular hypertrophy (LVH) is indicated by T inversion in V5 and V6, loss of the Q wave in V6 or the amplitude of the R wave in V6 + S wave in V1 > 98th centile for age (≥ 50 mm is always abnormal). RVH may indicate significant right heart obstruction or high pulmonary artery pressure (secondary to a large left to right shunt or pulmonary vascular disease). LVH may indicate significant left heart obstruction. Cardiomegaly and increased pulmonary vascular markings on the chest *X* ray may indicate a large left to right shunt.

Any child who is thought to have an anatomical defect on the basis of the clinical examination, or any child with an abnormal ECG or chest *X* ray should be referred to a paediatric cardiologist for an ***echocardiogram***. If there is evidence of a significant left to right shunt (see earlier section on heart failure) in a VSD or PDA, the referral should be as soon as possible as there is still a risk of pulmonary vascular disease even when the child does not present in heart failure.

If a cardiac defect is diagnosed, arrange further follow up and inform the parents of the need **for antibiotic prophylaxis for dental and surgical procedures (see Table 3.5.3).** ✔

Table 3.5.3 Antibiotic prophylaxis against infective endocarditis

Procedure	<5 years old	5–10 years old	>10 years old
Dental or surgical procedures under local anaesthetic	Oral amoxycillin 750 mg 1 hour before procedure	Oral amoxycillin 1.5 g 1 hour before procedure	Oral amoxycillin 3 g 1 hour before procedure
Dental or surgical procedures under general anaesthetic	IV amoxycillin 250 mg on induction plus oral amoxycillin 125 mg 6 hours later	IV amoxycillin 500 mg on induction plus oral amoxycillin 250 mg 6 hours later	IV amoxycillin 1 g on induction plus oral amoxycillin 500 mg 6 hours later
High-risk cases (prosthetic valve/previous endocarditis/ genitourinary procedure)	IV amoxycillin 250 mg plus IV gentamicin 2 mg/kg on induction plus oral amoxycillin 125 mg 6 hours later	IV amoxycillin 500 mg plus IV gentamicin 2 mg/kg (max. 120 mg) on induction plus oral amoxycillin 250 mg 6 hours later	IV amoxycillin 1 g plus IV gentamicin 2 mg/kg (max 120 mg) on induction plus oral amoxycillin 500 mg 6 hours later

If allergic to penicillin or the child has had more than one dose of penicillin in the last month substitute another antibiotic in place of amoxycillin for example:

- 50 mg oral clindamycin for every 250 mg oral amoxycillin that would have been given
- 75 mg of IV clindamycin for every 250 mg of IV amoxycillin that would have been given or
- 20 mg/kg IV vancomycin (max 1 g) in place of IV amoxycillin

3.6 The child in shock

Barbara Phillips and Alice Leahy

Minimum standards requirements

- **ABC** (see Chapters 1.19, 1.20, 1.21)
- Oxygen
- 0.9 % saline/4.5% albumin via IV or IO cannulae
- 10 % IV glucose
- Epinephrine
- Antibiotics: cefotaxime/ceftriaxone
- Blood and blood-clotting factors

Shock results from an acute failure of circulatory function. Inadequate amounts of nutrients, especially oxygen, are delivered to body tissues and there is inadequate removal of tissue waste products.

Maintenance of adequate tissue perfusion depends on a pump (the heart) delivering the correct type and volume of fluid (blood) through controlled vessels (arteries, veins and capillaries) without abnormal obstruction to flow. Inadequate tissue perfusion resulting in impaired cellular respiration (i.e. shock) may result from defects of the pump (cardiogenic), loss of fluid (hypovolaemic), abnormalities of vessels (distributive), flow restriction (obstructive), or inadequate oxygen-releasing capacity (dissociative).

The most common causes of shock in childhood are hypovolaemia from any cause, septicaemia and the effects of trauma.

Children in shock are usually presented by parents who are aware that their child is worryingly ill or seriously injured even though they may not be able to express their concerns clearly. The child may be presented primarily with a fever, a rash, with pallor, poor feeding or drowsiness or with a history of trauma or poisoning. The initial assessment will identify which patients are in shock.

Classification of causes of shock*

Cardiogenic	Arrhythmias Cardiomyopathy Heart failure Valvular disease Myocardial contusion Myocardial infarction
Hypovolaemic	**Haemorrhage** **Gastroenteritis** **Volvulus** **Burns** **Peritonitis**
Distributive	**Septicaemia** Anaphylaxis Vasodilating drugs Anaesthesia Spinal cord injury
Obstructive	**Tension pneumothorax** **Haemopneumothorax** Flail chest Cardiac tamponade Pulmonary embolism Hypertension
Dissociative	Profound anaemia Carbon monoxide poisoning Methaemoglobinaemia

*Common causes are bold.

Approach to the child in shock

Primary assessment

Airway

Assess airway patency by the "look, listen and feel" method.

If the child can speak or cry, this indicates that the airway is patent, that breathing is occurring and there is adequate circulation.

If there is no evidence of air movement then chin lift or jaw thrust manoeuvres should be carried out and the airway reassessed. If there continues to be no evidence of air movement then airway patency can be assessed by performing an opening manoeuvre and giving rescue breaths (see the section Basic life support in the *Advanced*

Paediatric Life Support Manual, 3rd edition, published by BMJ Books, 2001).

Breathing
Assess the adequacy of breathing.
- Effort of breathing
  - Recession
  - Respiratory rate
  - Grunting
  - Accessory muscle use
  - Flare of the alae nasi
- Efficacy of breathing
  - Breath sounds
  - Chest expansion/abdominal excursion
- Effects of breathing
  - Heart rate
  - Skin colour
  - Mental status

Oxygen saturation
Monitor oxygen saturation with a pulse oximeter (if available).

Cardiovascular status

Heart rate
A raised heart rate is a common response to many types of stress (fever, anxiety, hypoxia, hypovolaemia). In shock, tachycardia is caused by catecholamine release, and is an attempt to maintain cardiac output by increasing heart rate in the face of falling stroke volume. **Bradycardia in a shocked child is caused by hypoxia and acidosis and is a preterminal sign.** ✔

Pulse volume
Examination of central and peripheral pulses may reveal a poor pulse volume peripherally or, more worryingly, centrally. In early septic shock there is sometimes a high output state which will produce bounding pulses.

Capillary refill
Poor skin perfusion can be a useful early sign of shock. Slow capillary refill (>2 seconds) after blanching pressure for 5 seconds is evidence of reduced skin perfusion. When testing for capillary refill press on the skin of the sternum or a digit held at the level of the heart. Mottling, pallor and peripheral cyanosis also indicate poor skin perfusion. **All these signs may be difficult to interpret in patients who have just been exposed to cold.**

Blood pressure
Blood pressure is a difficult measure to obtain and interpret especially in young infants. A formula for calculating normal systolic blood pressure is:

$$80 + (2 \times \text{Age in years})$$

Children's cardiovascular systems compensate well initially in shock. **Hypotension is a late and often sudden sign of decompensation and, if not reversed, will be rapidly followed by death.** ✔

Serial measurements of blood pressure should be performed frequently.

Effects of circulatory inadequacy on other organs
- Acidotic sighing respirations
  The acidosis produced by poor tissue perfusion in shock leads to rapid deep breathing.
- Pale, cyanosed or cold skin
  A core/toe temperature difference of more than 3 °C is a sign of poor skin perfusion.

Urinary output
Urine flow is decreased or absent in shock. Hourly measurement is helpful in monitoring progress. A minimum flow of 1 ml/kg/hour in children and 2 ml/kg/hour in infants indicates adequate renal perfusion.

NOTE: Poor capillary refill, core/toe temperature difference and differential pulse volumes are neither sensitive nor specific indicators of shock when used in isolation. They are helpful when used in conjunction with the other signs described.

Mental status: agitation or depressed conscious level
Early signs of brain hypoperfusion are agitation and confusion, often alternating with drowsiness. Infants may be irritable but drowsy with a weak cry and hypotonia. They may not focus on the parent's face. These are important early cerebral signs of shock. Later the child becomes progressively drowsier until consciousness is lost.

Look for the presence of signs of heart failure
- Tachycardia
- Raised jugular venous pressure **often not seen in infants in heart failure**
- Lung crepitations on auscultation
- Gallop rhythm
- Enlarged liver

Disability
- Assess neurological function
- A rapid measure of level of consciousness should be recorded using the AVPU scale.
  A ALERT
  V responds to VOICE
  P responds to PAIN
  U UNRESPONSIVE
- Pupillary size and reaction should be noted
- Note the child's posture: **children in shock are usually hypotonic** ✔
- The presence of convulsive movements should be noted

Exposure
- Take the child's core and toe temperatures
- Look for a rash: if one is present, ascertain if it is purpuric
- Look for evidence of poisoning

Resuscitation

Airway

- A patent airway is the first requisite. If the airway is not patent an airway opening manoeuvre should be used. The airway should then be secured by intubation by experienced senior help.

Breathing

- All children in shock should receive high flow oxygen through a face mask with a reservoir as soon as the airway has been demonstrated to be adequate.
- If the child is hypoventilating, respiration should be supported with oxygen via a bag-valve-mask device and experienced senior help summoned.

Circulation

Gain intravenous or intraosseous access

- Take blood for FBC, U&Es, blood culture, cross-match, glucose stick test and laboratory test.
- Give 20 ml/kg rapid bolus of crystalloid to all patients except for those with signs of heart failure.
- The initial bolus should be colloid and an antibiotic such as cefotaxime 100 mg/kg should be used for those in whom a diagnosis of septicaemia is made obvious by the presence of a purpuric rash.
- If a tachyarrhythmia is identified as the cause of shock, up to three synchronous electrical shocks at 0.5, 0.5 and 1 joule should be given.
  - If the arrhythmia is broad complex and the synchronous shocks are not activated by the defibrillator then attempt an asynchronous shock.
  - A conscious child should be anaesthetised first or sedated.
  - If the shocked child's tachyarrhythmia is SVT then he/she can be treated with intravenous/intraosseous adenosine if this can be administered more quickly than a synchronous electrical shock.

Circulatory access

A short, wide-bore peripheral venous or intraosseous cannula should be used. Upper central venous lines are unsuitable for the resuscitation of hypovolaemic children because of the risk of iatrogenic pneumothorax, or exacerbation of an unsuspected neck injury; both these complications can be fatal. Femoral vein access is safer, if peripheral or intraosseous access is impossible. It is wise to obtain two separate intravenous and/or intraosseous lines both to give large volumes of fluid quickly and also in case one line is lost.

Techniques for vascular access are described in Chapters 6.6 to 6.8.

Antibiotics

In paediatric practice, septicaemia is the commonest cause of a child presenting in shock. Therefore, unless an alternative diagnosis is very clear (such as trauma, anaphylaxis or poisoning) an antibiotic, usually a third generation cephalosporin such as cefotaxime or ceftriaxone, is given as soon as a blood culture has been taken.

Hypoglycaemia

Hypoglycaemia may give a similar clinical picture to that of compensated shock. This must always be excluded by urgent glucose stick test and blood glucose estimation. Shock and hypoglycaemia may coexist as the sick infant or small child has poor glucose-producing reserves.

Choice of fluid

Crystalloid or colloid fluids are available for volume replacement. Colloids diffuse less readily into the interstitial space and therefore more effectively expand the intravascular volume with the use of smaller volumes. Proponents of the use of crystalloids suggest that the interstitial space is already depleted in hypovolaemia because of shifts of fluids from the interstitial space into the intravascular and intracellular compartments. It is also thought that, when colloids do leak into the interstitial space, their high osmotic pressure increases the fluid loss from capillaries into the interstitial space.

Key features in shock

While the primary assessment and resuscitation are being carried out a focused history of the child's health and activity over the previous 24 hours and any significant previous illness should be gained. Certain key features which will be identified clinically in the assessment, from the focused history and from the initial blood test results can point the clinician to the likeliest working diagnosis for emergency treatment.

- A history of vomiting and/or diarrhoea points to **fluid loss** either externally (for example gastroenteritis) or into the abdomen (for example volvulus, intussusception).
- The presence of fever and/or a rash points to **septicaemia**.
- The presence of urticaria, angioneurotic oedema and a history of allergen exposure points to **anaphylaxis**.
- The presence of cyanosis unresponsive to oxygen or a grey colour with signs of heart failure in a baby under 4 weeks points to **duct-dependent congenital heart disease.**
- The presence of heart failure in an older infant or child points to **cardiomyopathy.**
- A history of sickle cell disease or a recent diarrhoeal illness and a very low haemoglobin points to **acute haemolysis.**
- An immediate history of major trauma points to **blood loss**, and more rarely, **tension pneumothorax, haemothorax, cardiac tamponade** or **spinal cord transection** (see Chapters 5.4 and 5.7).
- The presence of severe tachycardia and an abnormal rhythm on the ECG points to an **arrhythmia** (see Chapter 3.5).
- A history of polyuria and the presence of acidotic breathing and a very high blood glucose points to **diabetes** (see Chapter 3.11).
- A history of drug ingestion points to poisoning (see Chapter 5.12).

When crystalloids are used the volume required is two to three times that of a colloid. As crystalloids leak into the interstitial space, it is not uncommon for tissue oedema to occur. There is disagreement as to whether the oedema is detrimental to tissue oxygenation or not. Clearly the decision to use a particular type of fluid to replace volume loss needs to take into account the cause of hypovolaemia. Often, the use of both crystalloid and colloid is appropriate.

If blood is needed, it may be given after full cross-match that takes about 1 hour to perform. In more urgent situations type-specific non-cross-matched blood (which is ABO rhesus compatible but has a higher incidence of transfusion reactions) should be requested. It takes about 15 minutes to prepare. In dire emergencies O-negative blood must be given.

Fluids should ideally be **warmed, but do not delay if this is not possible**. Isotonic electrolyte solution should ideally be kept available in a warmed cabinet.

Approach to the child with fluid loss (see Chapter 3.25)

Infants are more likely than older children to present with shock due to sudden fluid loss in gastroenteritis or with concealed fluid loss secondary to a "surgical abdomen" such as a volvulus. This is due both to the infant's low physiological reserve and his/her increased susceptibility to these conditions.

In infants, gastroenteritis may occasionally present as a circulatory collapse with little or no significant preceding history of vomiting or diarrhoea. The infecting organism can be any of the usual diarrhoeal pathogens, of which the most common is rotavirus. The mechanism leading to this presentation is that there is a sudden massive loss of fluid from the bowel wall into the gut lumen, causing depletion of the intravascular volume and the appearance of shock in the infant. This occurs before the stool is passed so that the diagnosis may be unsuspected. Usually during resuscitation of these infants, copious watery diarrhoea is evacuated.

Having completed the primary assessment and resuscitation and identified by means of the key features that fluid loss is the most likely diagnosis, the child is reassessed to identify the response to the first fluid bolus.

Reassess ABC

Fluid loss: emergency treatment

If the child still shows clinical signs of shock after the first bolus of fluid, give a second bolus of crystalloid. If there is clinical suspicion of a surgical abdominal problem, such as bile-stained vomiting or abdominal guarding, seek an urgent surgical opinion. An abdominal radiograph and an ultrasound scan may be helpful in showing distended bowel, intra-abdominal air or fluid.

In the case of infants with gastroenteritis, two boluses of crystalloid are usually sufficient to restore the circulating volume. If after this amount of fluid, the child is still in shock when assessed clinically, give the third bolus as colloid (human albumen gelofusine, haemaccel and pentastarch) and consider whether there is an additional or alternative diagnosis, such as an intra-abdominal surgical problem (for example volvulus, peritonitis) in the patient originally thought to have gastroenteritis or coexistent septicaemia in the patient with the "surgical abdomen".

Obtain surgical and anaesthetic advice if not already obtained and give broad spectrum antibiotics intravenously if more than two boluses of fluid have been required.

In the patient with gastroenteritis who has stabilised after treatment for shock there will still be a need to treat dehydration and electrolyte imbalance. See Chapter 3.25 for further management.

Approach to the child with septicaemia (see Chapter 4.6)

Approach to the child with anaphylaxis (see Chapter 1.23)

3.7 Medical renal problems

Malcolm Coulthard

Investigations common to all renal disorders

Biochemistry

Plasma or serum

Electrolytes

Sodium and potassium assays are essential for the logical management of children with kidney dysfunction. Bicarbonate is also extremely helpful but more difficult to measure.

Creatinine

✔ **Creatinine measurements provide the only relatively cheap and convenient measurement of glomerular renal function.** Though formulae can be used, the following guidelines allow the glomerular filtration rate (GFR) to be estimated in most clinical situations. The plasma creatinine depends on the bulk of the patient's muscle (where it is produced) and their height, so on average men have higher values than women, and older children have higher values than babies, except in the first few days of life: (Table 3.7.1)

For example, a creatinine of 150 µmol/l in a well-nourished 5-year old girl would be three times the upper limit of normal, indicating a GFR of one-third normal. The same creatinine in a very undernourished girl with little muscle bulk would imply a GFR considerably lower than one-third.

Table 3.7.1 Plasma creatinine concentrations

	Plasma creatinine concentration (upper limit)	
Subject	**micromol/l**	**mg/dl**
Well-nourished average man	100	1.15
Well-nourished average woman	75	0.85
Well-nourished average 10-year old	60	0.70
Well-nourished average 5-year old	50	0.65
Well-nourished average baby or toddler	40	0.45
Baby 3 days to 3 weeks	Variable	
Baby in first 2 days of life	Depends on maternal levels	

Urea

Though useful in managing children with renal failure, it is an inaccurate way of measuring renal function because it is also highly dependent on hydration, and the carbohydrate and protein intake.

Urine

✔ **Measurements, if available, of the urinary sodium and creatinine concentrations of a spot sample are the only reliable biochemical way to distinguish pre- from established renal failure, and to diagnose hypovolaemia.** They are used to calculate the **fractional excretion of sodium (FENa)** from the urine (U) and plasma (P) concentrations (check P and U creatinine are in the same units), using following formula:

$$\text{FENa (\%)} = \text{U/P sodium} \times \text{P/U creatinine} \times 100$$

Urine collection and examination

Collecting urine

Collecting urine from babies can be difficult. If a clean catch collection cannot be managed, it is cheaper and more comfortable to use a sterile collecting pad than an adhesive bag. For toddlers it is fine to use a potty or equivalent which has been thoroughly washed in hot water and detergent (using antiseptics or bleach, or scalding with boiling water are unreliable). Suprapubic sampling is usually only justified in desperately ill children where it is not possible to accept delay.

Stick testing of urine and protein measurement

Dip-stick testing for blood, protein, and glucose are useful and reliable. Stick testing for nitrite to identify urine infection (UTI) is unreliable because it remains negative in 50% of cases. Stick testing for white cells to diagnose UTIs is unreliable because they may occur without white cells, and because white cell numbers also increase in the urine of febrile children without UTIs. Urine protein should be <20 mg protein per mmol creatinine. In nephrotic syndrome typically > 500 mg protein per mmol of creatinine.

Microscopy

Microscopy of unspun fresh urine can provide a cheap and reliable way of rapidly diagnosing UTIs (by identifying bacteria directly, rather than by counting white cells), and of diagnosing schistosomiasis. Red blood cells can be identified as being due to glomerulonephritis (where they are

small, fragmented and of varied and distorted shapes), or due to other causes, such as trauma, stones or bladder inflammation (where they are all similar, and typically biconcave). A standard light microscope with a magnification of ×400 is sufficient. Using a counting chamber (or a microscope slide with a scratched surface) and cover slip ensures that the microscope is focused at the correct plane; otherwise it is not possible to tell when microscoping a normal urine. A counting chamber with a mirrored surface is not essential, but makes identification of bacteria easier. Phase contrast makes identification even easier; a highly reliable, almost pocket-sized microscope (McArthur) is available with phase contrast.

Urinary tract imaging techniques

All renal imaging techniques are relatively expensive, and many will have limited availability.

Ultrasound

Good information can only be obtained from ultrasound by a good operator using an adequate machine (see Chapter 1.11). It demonstrates anatomy, but not function. It is radiation free, and, when available, is now the first choice for initial imaging in most renal conditions in children. It is excellent at demonstrating obstruction and dilatation, and has a similar sensitivity to the IVU for demonstrating longstanding or extensive scarring. Nephritis causes echo-brightness of the kidneys. Tumours and cysts are easily seen, usually before they are visible by other modalities. Stones can be easily identified, but may be misinterpreted by the inexperienced because the whole stone is not seen; a bright line identifies where the ultrasound hits the front edge of the stone, and an acoustic shadow is thrown behind it. Nephrocalcinosis can be detected easily as white renal pyramids long before it can be seen on *X* rays.

Micturating cystogram (MCUG)

This is still the most reliable way to assess vesicoureteric reflux, but unfortunately depends on invasive urethral catheterisation, and requires a high dose of radiation. *(Reflux is rare in black children.)*

Plain *X* ray

Demonstrates radio-opaque stones, but these, and nephrocalcinosis are usually easier to see on an ultrasound scan.

Urinary tract infections (UTIs)

Minimum standards requirements

- Urine microscopy
- Ultrasound
- ***MCUG***
- Antibiotics: trimethoprim, cephalosporins IV and oral, amoxycillin, nitrofurantoin, gentamicin

Background

UTIs are very common. Vesicouretic reflux is a common and important cause (see Chapter 3.8). Recent studies have shown that about 10% of girls and 3% of boys will have had a UTI diagnosed by the age of 16 in the UK. They are important because they may cause scarring. Large scars may cause renal failure, but even small ones can cause hypertension, often much later. **To prevent serious sequelae of hypertension, children with scars should have life-long blood pressure monitoring.** Infants are the most vulnerable to scarring; a child over 4 without a scar is unlikely to ever develop one. Animal studies suggest that a UTI in a vulnerable individual may cause permanent scarring very rapidly, perhaps in just 3 days.

Diagnosis

Symptoms

In older children, symptoms are typically due to bladder and urethral irritation, such as frequency and dysuria, but some children have few or no symptoms. Younger children (<2 years) often only have non-specific symptoms such as anorexia, failure to thrive, unexplained fever or prolonged jaundice. Therefore, all young children with an unexplained illness, particularly with a fever, should have a UTI excluded.

Urine testing

A diagnosis is usually made by culture of a pure growth of one species of bacteria (most commonly *E. coli*) at a concentration of more than 10^5/ml. White cells (>50/microlitre) are usually considered helpful in making the diagnosis, but UTIs can occur without any white cells (sometimes because they lyse in minutes), and misleading because they occur commonly in children with fever who do not have a UTI. Any bacteria in a suprapubic urine sample suggests infection.

Microscopy

Microscopy of freshly voided unspun urine is a quick, reliable and cheap way to diagnose UTIs if a ×400 microscope is available (see Chapter 6.25), and allows an immediate diagnosis to be made. This allows the best-guess antibiotic to be started at once. Ideally, infected urines need to be cultured to obtain antibiotic sensitivities. Infected urine will have at least one bacterium, and up to thousands per high-power field, depending slightly on the depth of urine under the cover slip. They will all look the same, and are typically rods of identical length. Do not expect to see them swimming about, though sometimes they do. Occasionally UTIs are caused by streptococci which are seen as long chains of dots. Separate small dots that appear to be swimming are not streptococci, but are phosphate crystals (the shimmering movement is due to Brownian motion). Most, but not all children with UTIs will also have >50 white cells/microlitre, or at least 1 per 10 high-power fields.

Urines where no bacteria are seen in about 5 high power fields are not infected; samples need no further testing, and can be discarded. Urines containing less than 1 bacterium per high power field, or mixtures of rods and

cocci are likely to have been contaminated. Because this can be identified instantly, further samples can be collected until an uninfected or sterile one is obtained.

Imaging (see Chapter 1.11 for test details)

- **Ultrasound**. Perform on all children after their first recognised UTI, whatever their age, to identify structural abnormalities, and to try to identify scars. The chance of picking up a scar is much greater if it is large, involving multiple renal segments, or several years old so that it will have had time to shrink and distort. Negative scans in young children (under 4 years) therefore need to be interpreted with caution.
- ***Micturating cystogram. It is probably ideal to perform an MCUG on all children under 1 year that have had a UTI because vesicoureteric reflux may indicate that they are at risk of developing scars with future UTIs, which may be prevented by using prophylactic antibiotics.*** An alternative approach is to give prophylactic antibiotics (see below) to all infants who develop a UTI. ***Finding vesicoureteric reflux should make you suspicious that the child may have a scar that was not identified by ultrasound. Children aged 1 to 4, who are still at risk of developing a first scar with a further UTI, should have an MCUG if they have recurrent UTIs, and protection with prophylactic antibiotics if they have reflux. Children of any age should have an MCUG if they have a scar identified on ultrasound.***

Treatment

Encourage a high fluid intake to produce a dilute urine, and reduce the symptoms of dysuria. Treat for a week initially with oral trimethoprim (4 mg/kg twice daily), or cephalexin or amoxycillin (10 mg/kg three times daily of either), or nitrofurantoin (1mg/kg three times daily). Intravenous antibiotics may be necessary for very unwell children (particularly under 2 years of age) for as long as they are unable to tolerate oral medication. This may include gentamicin (see page 394); give a standard first dose of 2 mg/kg, and further (2 mg/kg 8 hourly) doses only after confirmation that the plasma creatinine is normal. If there is renal failure, no more should be given after the single dose, unless blood levels are available to guide dosage. If necessary, change the antibiotic according to the laboratory sensitivity testing, once and if it is available.

For children under 4 years, continue to give antibiotic prophylaxis until the results of the imaging are known. Give a night-time dose of trimethoprim (2 mg/kg) or cephalexin (about 10 mg/kg) or nitrofurantoin (1 mg/kg); do not use amoxycillin for prophylaxis because resistant organisms are likely to emerge. For children under 4 years with vesicoureteric reflux but no scars, continue prophylaxis until they are 4. For children with scars, give prophylaxis for at least two years, and restart subsequently if UTI recur.

Continent children with vesicoureteric reflux should practice double voiding at least once per day if they can do it (not all can produce a second urine). They should be encouraged to develop a routine of taking their time to void fully, and then repeat this about 5 minutes later, for example before and after cleaning their teeth at night.

Girls should be taught to wipe their bottoms backwards, and to avoid using soap to the vulva as far as possible. They should be discouraged from wearing nylon knickers. Boys old enough to pull back the foreskin should be encouraged to do this gently when washing themselves.

Urinary tract stones

Minimum standards requirements

- Analgesia: morphine or pethidine
- Chlorthiazide
- Surgery

Background

There is wide geographic variation in the frequency of stone disease, and there have been major changes in prevalence with time within populations. Generally, urinary stones are more common among socially disadvantaged people, and in the tropics. The reasons for this are not clear, but may in part be due to dietary factors, and a tendency to developing dehydration in hot climates.

Causes

There are three broad causes for urinary tract stones (which may coexist in individual children):

- **Proteus urinary tract infections**. The mechanism is twofold. The infection results in turbid urine containing cells and debris, and secondly proteus splits urea to form ammonia, which raises the urinary pH. Because calcium ammonium phosphate is relatively insoluble in alkaline urine, it will coprecipitate readily onto the urinary debris under these conditions to form thick sludge initially, and subsequently a stone. This explains why these stones take up the shape of the tract they form in ("stag-horns" in the pelvicalyceal system, "date stones" in the lower ureter, round stones in the bladder). Preschool boys are affected much more than any other groups; it is not known why.
- **Relative dehydration and possibly dietary factors**. The mechanisms of stone formation are probably similar to those in infection stones, with chemicals normally found in the urine reaching relatively high concentrations due to low urine volumes, and high dietary intakes and consequent excretion rates of relatively insoluble chemicals.
- **Rare inherited metabolic conditions** which result in excessive urinary excretion of poorly soluble chemicals. Calcium stones are most commonly caused by isolated hypercalciuria (without hypercalcaemia), and more rarely by hypercalciuria combined with hypercalcaemia in hyperparathyroidism. Cystine

stones are seen due to an (dominantly or recessively) inherited failure of the proximal tubules to reabsorb this amino acid. Oxalate stones may be due to excessive gut absorption of oxalate when the calcium is unavailable to precipitate it, such as with steatorrhoea. Rarely it is also produced and excreted in excess due to a recessive liver enzyme deficiency.

Presentation and diagnosis

Children may pass a stone or present with severe colicky abdominal pain (typically in one loin), often with frank haematuria. Ultrasound is a sensitive imaging tool, but it must be remembered that the whole of the stone cannot be seen. Rather, the front edge will be seen as a bright line, and an acoustic shadow will be thrown behind it. Nephrocalcinosis associated with hypercalciuria is easily seen as white (echobright) renal pyramids.

A plain abdominal *X* ray will show most stones, and is often very useful in distinguishing the type. Similarly, the appearance of a passed stone or fragment may aid identification. Infection and dehydration stones are usually grey, and only moderately *X* ray dense, and take up the shape of the collecting system. Calcium (white) and cystine (yellow) stones are very *X* ray dense, may grow up to 2 cm or more in diameter, and are typically smooth, and round or oval wherever they form. Oxalate stones are yellowish-buff coloured and typically grow to 5 mm, with irregular, spiky edges. A high oxalate load will result in many small stones rather than large individual ones.

If the type of stone is not clear from the history, chemical measurements can made on an untimed "spot" urine collected during the morning (but not the overnight sample). Excessive excretion of calcium results in a calcium:creatinine ratio (the ratio of the calcium and creatinine concentrations, both in mmol/litre) of >0.8 mmol/mmol. Excessive cystine excretion produces a cystine:creatinine ratio of >25 micromol/mmol, and excess oxalate causes an oxalate:creatinine ratio >0.18 mmol/mmol. These ratios will be normal in children with infection stones, or those secondary to dehydration.

Treatment

Removal of stones

Small stones may be passed spontaneously – remember ureteric colic may be excruciating, and should be treated with powerful opiate analgesia (see Chapter 1.27). ***Spasmolytics such as hyoscine butylbromide (6–12 years 5–10 mg IV or orally; >12 years 20 mg IV or orally) are unlikely to be effective but are used. Larger stones may need surgical removal by open surgery or cystoscopy. Percutaneous nephrostomy or lithotripsy have little role in children.***

Preventing recurrences

Infection stones are not likely to recur. Stones due to metabolic causes and those related to dehydration are all helped by a consistently high fluid intake, but it is probably more important to avoid episodes of acute dehydration (e.g., with vomiting or diarrhoea) than to attempt to increase the daily fluid intake.

Chlorothiazide up to 10 mg/kg twice daily reduces urinary calcium excretion; its dose can be titrated in hypercalciuria to keep the urinary calcium: creatinine ratio in the normal range. Furosemide should be avoided because it increases urinary calcium excretion.

Children with hyperparathyroidism usually require parathyroidectomy.

In cystinuria, it is essential to maintain a life-long high fluid intake. Alkalinising the urine increases the solubility of cystine. Give oral sodium bicarbonate supplements (start with 1 mmol/kg daily) until the urine pH is usually 7 or more on home testing with strip test paper.

With oxalate stones due to malabsorbtion, treat the underlying bowel problem. Inherited hyperoxaluria typically leads to renal failure and calcification of all soft tissues.

Hypertension

Minimum standards requirements

- Blood pressure measurement
- Furosemide
- Nifedipine, hydralazine/atenolol/captopril

Background

More than 95% of childhood hypertension is caused by renal problems. Often the blood pressure becomes very much higher than normal (unlike the majority of adult hypertension patients, whose blood pressure is only moderately elevated, causing a skewed frequency distribution curve). Children are also relatively intolerant of hypertension, so they are at major risk of sequelae, especially encephalopathy, blindness and death.

Measurement

Blood pressure is best measured with a simple sphygmomanometer. Automatic blood pressure machines are unnecessary, and often unreliable. Use the largest cuff that will fit onto the upper arm (forget "formulae" that relate the cuff size to the child's size). A cuff that is too large will not significantly underestimate the blood pressure, but one that is too small will overestimate it. In children, systolic blood pressure is the best to use; it is just as important as diastolic for diagnosis and treatment, and is easier and more reliable to measure. In most children, palpating the reappearance of the pulse at the wrist is as accurate as using a stethoscope at the antecubital fossa, and allows a larger cuff to be used without it getting in the way. ***A luxury is to use a Doppler at the wrist to detect the reappearance of the pulse.*** High values should be confirmed with the child relaxed to exclude the effects of

anxiety. The normal blood pressure range varies with age. Table 3.7.2 gives a guide to management.

Table 3.7.2 Management of hypertension

Value	Systolic blood pressures at different ages				
	1 month	1 year	5 years	10 years	15 years
Mean	60	80	90	105	115
Upper limit of normal	80	100	110	120	130
Needing urgent treatment	110	130	140	150	160

Causes and diagnosis

It is important to find the underlying cause of the hypertension to guide management. Sometimes the cause is clear from the history, examination or urine testing, and sometimes it requires diagnostic imaging. Ultrasound is the most useful screening technique. Hypertension can be caused by a scar that is too small to detect with ultrasound.

Treatment

If the hypertension is known to be of recent onset, as in acute glomerulonephritis, it is safe to reduce the BP quickly. Usually salt and water overload is a major factor; if so, sodium restrict and give furosemide 1–2 mg/kg (oral is as effective as intravenous).

In other cases, treat the blood pressure slowly because cerebral arterial vasoconstriction may have occurred to protect the brain parenchyme from the impact of the hypertension, and made the cerebral blood flow dependent on a high blood pressure being sustained. **A rapid fall in blood pressure may cause infarction, especially blindness.** Reduction over two or more days allows the vascular tone to return to normal. **Slow control may be achieved by introducing oral hypotensive drugs slowly, at much below the maximum dose.**

Table 3.7.4 Examples of oral hypotensive drugs for use in children

Class	Example	maximum dose (mg/kg/day)
Calcium-channel blocker	Nifedipine	1
Vasodilator	Hydralazine	10
Beta-blocker	Atenolol	1
Angiotensin-converting enzyme (ACE) inhibitor	Captopril	5

Notes: (i) These are often given in combination to achieve a powerful effect with fewer side effects.
(ii) ACE inhibitors must be started by giving a very low test dose first and building up slowly. They must be used with great care if renal artery stenosis is suspected.

Nephrotic syndrome

Minimum standards requirements

- Prednisolone
- IV albumin
- Fluid balance charts and accurate daily weights
- Penicillin
- ***Cyclophosphamide***

Table 3.7.3 Renal causes of hypertension

Diagnosis	Notes	Kidney ultrasound
Reflux nephropathy	Also called pyelonephritis. See UTIs	Focally scarred, or small shrunken
Glomerulonephritis		
postinfective	Usually have proteinuria and glomerular haematuria Typically after a streptococcal sore throat / skin infection. Give penicillin for 10 days	Echo-bright
other causes	May have clinical evidence of Henoch-Schönlein, or lupus. Unless there are clinical differences ***diagnosis usually depends on a renal biopsy.***	
Inherited polycystic disease		
infantile-type (recessive)	Kidneys usually huge at birth, and typically have severe hypertension, and renal failure in early life	Huge, homogeneous, echo-bright
adult-type (dominant)	Seldom causes childhood renal failure, but may cause hypertension; screen blood pressure in children of affected parents	Discrete cysts develop through childhood
Narrowed arterial supply		
coarctation of the aorta	Check for femoral pulses	
renal artery stenosis	May need surgical treatment ***or balloon angioplasty***	May be small (difficult to diagnose without expensive imaging)
small intrarenal arteries	Require long-term medical treatment. May occur with neurofibromatosis, so screen all patients with this	

Background and clinical features

Defined as hypoalbuminaemia due to very high urinary albumin losses, which causes oedema. It must be differentiated from other causes of hypoalbuminaemia such as protein malnutrition (see Chapter 3.16) and protein-losing enteropathy (see Chapter 3.30). It is traditionally classified as early (congenital; diagnosed under 6 months) and later onset types.

- **Early onset:** Children with Finnish (recessively inherited) nephrotic syndrome die early of protein malnutrition, infection, or thrombosis unless they are aggressively treated, ***including early unilateral or bilateral nephrectomy, dialysis and transplantation*** – unrealistic in most of the world. Diffuse mesangial sclerosis is a similar condition, but is usually less acute. Congenital syphilis can cause neonatal nephrotic syndrome which may respond to penicillin treatment.
- **Later onset:** Most children with nephrotic syndrome are steroid responsive (lose their proteinuria within a month of treatment). Steroid responsive children also tend to share clinical characteristics (Table 3.7.5). Steroid resistant nephrotics may have a range of diagnoses, including focal segmental glomerulosclerosis, Henoch-Schönlein purpura, lupus and mesangiocapillary glomerulonephritis. There is a strong association with infections, especially malaria and hepatitis B.

Table 3.7.5 Typical features of children with steroid sensitive and steroid resistant syndrome

	Typical findings	
Feature	**Steroid sensitive**	**Steroid resistant**
Sex	Male > Female	Varies with condition
Age	1–3 years	Usually older
Blood pressure	Normal	Often elevated
Speed of onset	Rapid (days or weeks)	Usually weeks or months
Urinary blood	Microscopic	Often macroscopic
Plasma creatinine	Normal or low, unless hypovolaemic	May be elevated

Acute management

In all cases, it is reasonable to attempt to induce a remission with steroids, unless their clinical picture virtually excludes the possibility of steroid sensitivity. Use prednisolone 60 mg/m^2 daily (see table in Appendix 7.7 to convert from body weight to surface area) for up to six weeks (about 95% of children that will respond do so within one month). Monitor carefully for the development of hypertension on steroids.

Limit fluid retention by imposing a tight dietary sodium restriction. Prevent secondary pneumococcal infection with prophylactic penicillin V (125 mg twice daily up to 5 years of age, 250 mg twice daily thereafter). Avoid the sequelae of hypovolaemia. Intravascular hypovolaemia is a high risk and should be monitored clinically by the appearance of cold peripheries. Hypotension may not occur until late, and blood pressure can even be high. Hypovolaemia may cause abdominal pain. The best laboratory test is a urinary sodium less than 15 mmol/l, especially if combined with a urine osmolality of over 800 mosmol/kg; blood tests are seldom helpful. Treatment of hypovolaemia should be with 1 g/kg intravenous albumin over 4 hours, preferably with 2 mg/kg IV furosemide given half way through.

Subsequent management IF STEROID SENSITIVE

This is ideally based on daily home monitoring of the morning urine protein level by stick testing. A common definition of a relapse is ++ proteinuria for 7 consecutive days, or +++ for 3 days, and should be responded to by re-introducing salt restriction, penicillin V and prednisolone.

Protocols for doses and duration of using prednisolone and steroid-sparing agents vary.

This is a proposed example:

- First presentation – give prednisolone 60 mg/m^2 daily (use Appendix 7.7), and if respond (by loss of proteinuria), then complete 6 weeks of 60 mg/m^2 daily, followed by 40 mg/m^2 on alternate days for a further 6 weeks.
- Subsequent relapse – restart 60 mg/m^2 daily until no proteinuria for 3 days, then give 40 mg/m^2 on alternate days for a further 4 weeks.
- Frequent relapses – give prophylactic, low-dose (for example 200 micrograms/kg), alternate-day prednisolone. Titrate the dose up until either relapses are prevented, or steroid side effects develop.
- If steroid prophylaxis causes unacceptable side effects, add prophylactic levamisole 2.5 mg/kg on alternate days (approximately 50% will benefit) which can be used relatively long term.
- ***If levamisole is ineffective, consider cyclophosphamide 2.5–3 mg/kg daily for 12 weeks, monitoring weekly with white blood cell count, and reducing the dose if the absolute neutrophil count falls below 1 $\times$ 10^9/litre, or stopping if it falls below 0.5 $\times$ 10^9/litre. Potentially dangerous in poor circumstances where infections are frequent.***

Subsequent management IF STEROID RESISTANT

There is a wide range of conditions that may induce steroid resistant nephrotic syndrome. These include infective agents, auto-immune diseases, some drugs and poisons, and unknown causes. Though a cause may be suggested or may even appear certain from the history and examination, ***in most cases the diagnosis relies on the accurate interpretation of a kidney biopsy.***

The infective causes include hepatitis B, *schistosoma mansoni*, leprosy, tuberculosis and malaria. These conditions should be sought in those parts of the world where

they are likely to be found, and treated appropriately. Hepatitis B typically causes a membranous nephropathy which tends to improve spontaneously. Post-streptococcal glomerulonephritis may cause nephrotic syndrome, but it is seldom the presenting feature. Though it is not the only cause of this clinical picture, **it is sensible to treat any child that develops nephrotic syndrome after an acute nephritic illness with 10 days of oral penicillin V, approximately 10 mg/kg per dose 6 hourly**.

The auto-immune causes include Henoch-Schönlein and IgA nephritis, lupus, mesangiocapillary glomerulonephritis, and some cases of membranous nephropathy. The commonest cause of steroid resistant nephrotic syndrome in many parts of the world is focal segmental glomerulosclerosis (FSGS), whose pathophysiological mechanism is unknown. In some of these conditions (including lupus, mesangiocapillary glomerulonephritis, and FSGS), some children do respond to steroids. However, many children with steroid resistant nephrotic syndrome do not respond to any treatment at all. ***Most of those that do only respond to more powerful immunosuppressants such as cyclophosphamide or cyclosporin, or require antibodies to be removed by plasmapheresis.*** These treatments are difficult to apply because they are expensive, and require close monitoring for side effects. Even under ideal medical conditions, many cases still progress to end-stage renal failure.

Children with nephrotic syndrome may lose huge quantities of protein into their urine. **If they are on a low protein diet they will quickly lose muscle mass as the body proteins are utilised to synthesise plasma albumin.** A relatively high protein diet will be muscle sparing, but will make no significant difference to the plasma albumin concentration.

Glomerulonephritis

Glomerulonephritis (GN) strictly refers to inflammation of the glomeruli with cellular proliferation, though is often used to include other glomerulopathies such as FSGS and membranous nephropathy, both of which typically cause steroid-resistant nephrotic syndrome. The commonest cause of childhood GN varies widely across the world. In disadvantaged countries acute post-streptococcal GN is the commonest. In advantaged countries this is now unusual, and IgA nephropathy predominates.

Post-streptococcal GN

This is caused by children producing antibodies in response to specific strains of streptococci. These bacteria typically cause throat and skin infections. The antibodies then form complexes and are deposited within the glomeruli along with C3. Because it takes time for antibody production to occur, the signs and symptoms of nephritis do not usually begin until 10 to 20 days after the start of the infection.

The inflamed glomeruli leak blood and protein, so the first symptom is usually the child passing smoky or frankly bloody urine. The glomerular filtration rate usually falls a little, so the plasma creatinine is typically slightly elevated. Also the tubules reabsorb sodium and water excessively which causes water retention out of proportion to the fall in glomerular filtration rate. This leads to swelling which is most easily noticed around the eyes and face, and in the legs, but which does not pit as easily as oedema does in the nephrotic syndrome. The water retention also leads to hypertension. Most children with acute post-streptococcal GN do not lose enough protein into the urine to cause nephrotic syndrome as well, though some do, producing a mixed nephrotic-nephritic picture.

A presumptive diagnosis is made by examination of the urine for the presence of protein (using stick tests) and glomerular red cells and casts (by microscopy – see Chapter 6.25), in a child with a history of a recent sore throat or skin infection. Culture of a specific strain of streptococcus from a throat or skin swab may confirm the diagnosis. ***It is not reliable to make a diagnosis from a single titre of an anti-streptococcal antibody such as the ASOT or the antiDNAse-B because many children have an elevated level from previous exposure to other strains of streptococci. Rather, confirmation requires a significant rise between two titres taken at least 10 days apart. The plasma C3 concentration is reduced, and often stays subnormal for up to 6 weeks, and the C4 may also be low. This is not diagnostic of post-streptococcal GN, however. The plasma C3 is usually low in mesangiocapillary GN, and the C3 and C4 are often both low in lupus, and both these conditions may look clinically identical to post-streptococcal GN.***

Treatment

If post-streptococcal GN is suspected immediately start penicillin V 10 mg/kg four times daily for 10 days to eradicate the organism. There is always a delay in obtaining bacteriological confirmation, either from culture or ***from paired titres***, so it is best to start the penicillin at once, and use these tests as retrospective confirmatory evidence.

It is essential to measure the child's fluid intakes and losses accurately, and restrict the amounts of sodium and water allowed. This should be to balance the losses, or to cause net fluid reduction if the child is significantly fluid overloaded. The insensible loss is about 300 ml/m^2 daily, and will obviously be higher in a hot dry climate. Estimate the surface area from the table in Appendix 7.7. **Salt restriction is far more important than water restriction**, and is sometimes all that is required for a child to maintain fluid balance. This is because the tubules retain sodium avidly, so any salt eaten will be retained in the body and cause hypernatraemia. This drives an intense thirst, and it then becomes almost impossible to stop the child drinking. By contrast, a tight salt restriction will minimise the thirst, which aids management.

If the plasma albumin concentration is normal or only slightly reduced, it is safe to give an oral dose of furosemide, 1–2 mg/kg. This will increase the urinary excretion of sodium and water and thus improve fluid overload and hypertension. It will also increase potassium

loss which is helpful if the fall in glomerular filtration has led to hyperkalaemia. It may be repeated as needed. If the child has a very low plasma albumin from a mixed nephrotic-nephritic picture, giving furosemide may precipitate hypovolaemia. Because of this, either give intravenous albumin combined with furosemide (see section on acute management of the nephrotic syndrome), or give furosemide under close observation and be prepared to give albumin if hypovolaemia occurs. Cold peripheries and abdominal pain (from splanchnic vasoconstriction) are important signs of this.

The raised blood pressure is frequently fully controlled by salt and water restriction and furosemide, but in some cases hypotensive agents are also needed (see table 3.7.4). Under such acute conditions it is safe to reduce the blood pressure rapidly.

In children with post-streptococcal GN, the kidneys almost always make a full recovery, and progression to renal failure is very rare. Therefore most will have no sequelae provided their fluid and electrolyte balance and blood pressure are carefully managed.

IgA nephropathy (Berger's disease)

The typical presentation is of a child aged 5 to 15 years who develops an acute viral illness, and simultaneously has heavy haematuria that lasts for about 5 days. Urine microscopy reveals distorted "glomerular" red cells (see Chapter 6.25). Usually the urine then clears completely, but the haematuria returns with subsequent viral illnesses. Some children with IgA disease have a more insidious illness with little or no macroscopic haematuria.

The diagnosis is usually obvious in children who present with recurrent heavy glomerular haematuria. This is strengthened if there is a family history. ***The plasma IgA concentration may be higher in affected children, but this test is a poor discriminator. In children with a less obvious clinical picture the diagnosis can only be made on a kidney biopsy. Antibody staining will show granular deposits of IgA in glomeruli that have mesangial proliferation. Histologically, IgA disease is identical to Henoch-Schönlein nephritis.***

The best prognostic indicator in IgA nephropathy is the amount of proteinuria that persists between the acute episodes of haematuria. Most children have heavy haematuria but little or no proteinuria between attacks, and virtually all of these grow out of the condition during the teenage years, usually without any sequelae. Often the ones with the most dramatic bleeding recover particularly well. The children with a more insidious onset are more likely to have persistent proteinuria, and to continue with the condition into adult life, eventually developing end-stage renal failure by middle age. There is no treatment that is known to prevent this happening. Good blood pressure control is important.

Rarely, IgA disease first presents as a severe rapidly progressing GN. The picture is one of an acute nephritis in which the creatinine rises rapidly and inexorably. It is therefore clinically indistinguishable from any other rapidly progressive GN, ***other than by renal biopsy.*** Treatment options include ***various immunosuppressive drugs and plasmapheresis***. These have not been subject to control trials, are expensive and may lead to serious complications.

Haematuria

- Urine test sticks are highly sensitive and detect the smallest traces of blood.
- **For most conditions that can cause haematuria, the clinical significance is best predicted by the quantity of protein present, so always test for that too.** ✔
- **The most important test to determine the cause of haematuria is to check the shape of the red cells, ideally under phase-contrast microscopy (see Chapter 6.25).** ✔

Macroscopic glomerular haematuria

The presence of distorted red cells may be due to any form of glomerulonephritis, as listed above. The history of a simultaneous infection may suggest IgA nephropathy, while a recent infection points to post-streptococcal GN. The presence of a rash, or joint involvement, or abdominal pain might suggest Henoch-Schönlein or lupus nephritis as causes, but most other types can only be diagnosed on renal biopsy.

Macroscopic non-glomerular haematuria

The presence of red cells with a normal biconcave appearance indicates bleeding into the urine, and excludes glomerulonephritis as a cause. This may be due to trauma, but this would have to be major, because the renal tract is physically well protected. Minor trauma will only cause bleeding if the kidney is enlarged with cysts, as in adult type (dominantly inherited) polycystic kidney disease, or if it is vulnerable by being ectopically positioned. Urinary tract stones can also cause bleeding. The cystitis caused by a urinary tract infection or by *Schistosoma mansoni* may cause frank haematuria. These can be distinguished on phase-contrast microscopy of a fresh sample, when either bacteria or ova are easily visible. Though bleeding into the urine can be due to a malignancy, this is rare in childhood, and is then usually from a Wilms' tumour. Frank haematuria in a newborn suggests a renal vein thrombosis which may be unilateral or bilateral, and the affected kidney is usually easily palpated.

Trauma, stones, an ectopic kidney, adult polycystic disease, a Wilms' tumour and renal vein thrombosis can each be identified on ultrasound where they have a characteristic appearance. In cases with no cause found, cystoscopy should be considered.

Microscopic glomerular haematuria

Blood may be detected in urine that looks completely clear. Rarely the red cells appear normal on microscopy; these children should be investigated as for frank

non-glomerular haematuria. Most children with microscopic haematuria have distorted "glomerular" red cells. In this group, management depends on how much proteinuria they have. Those with massive, nephrotic-level proteinuria should be assessed and managed as for other children with nephrotic syndrome. Those with moderate proteinuria are likely to have a form of glomerulonephritis. The vast majority of children will not have any proteinuria with their microscopic haematuria. Investigation of this group is unlikely to identify a cause. If the renal ultrasound scan is normal, these children should be monitored annually just with a urine stick test and blood pressure measurement. If the blood disappears, follow up may be discontinued. If it persists without proteinuria or hypertension developing, continue annual reviews. If proteinuria or hypertension appear, the child needs to be investigated accordingly.

Confusing findings

Blood may be present without any red cells visible on microscopy. This indicates acute haemolysis such as may occur in G6PD deficiency or malaria.

Large quantities of urate make the urine brick red. Though families may think it resembles blood, it is easily distinguished visually. Porphyria is a very rare cause of confusion. Ingestion of red vegetables and sweets is said to occasionally cause red urine, though this is of doubtful importance.

Acute renal failure

Minimum standards requirements

- Fluid balance charts and accurate daily weights
- IV N saline/4.5% albumin
- Furosemide
- IV glucose 10%
- Sodium bicarbonate
- Antihypertensive drugs
- ***Peritoneal dialysis***

Types of acute renal failure (ARF)

Acute renal failure may be caused by insults to the renal tubule cells such as poor perfusion which leaves them functioning under difficult conditions, causing pre-renal failure. More severe insults that cause actual acute damage to the kidney cells result in established renal failure. Most commonly established renal failure results from more extreme or more prolonged versions of the same insults that cause pre-renal failure, though some other causes also exist, such as poisoning by drugs and haemolytic-uraemic syndrome. The prognosis for recovery depends on whether only the tubule cells are damaged, or if the glomeruli are also involved. Acute complete obstructions to the renal tract are rare, but can cause failure of urine production, termed post-renal failure. Each type of ARF has a different management. It is therefore important to distinguish them clearly.

Diagnosis and initial management of ARF

Pre-renal acute renal failure

Pre-renal failure is essentially a reversible renal dysfunction due to the kidneys being underperfused, but where the perfusion is still sufficient to prevent necrosis of the renal tissue. Hypotension (shock: see Chapter 3.6) is the commonest mechanism, and there is usually an obvious history of a cause for this. The commonest cause worldwide is dehydration from gastroenteritis. Other causes of shock include blood loss, plasma loss from burns, severe infections including septicaemia, and heart failure.

The clinical diagnosis is made by recognising the signs of shock. Commonest are a delayed capillary refill time, cool peripheries, a weak pulse, and usually a low blood pressure. However, the blood pressure may also be unexpectedly high because of the powerful renin drive in response to hypovolaemia. An important feature is that the child may complain of abdominal pain (induced by splanchnic ischaemia as blood flow is diverted from the gut to more vital organs).

Laboratory confirmation of the diagnosis is made by measuring the fractional excretion of sodium (FENa, see above). This requires measurement of the sodium and creatinine concentrations in a sample of blood and urine. If the FENa is less than 1% this indicates that the renal tubule cells are still alive, and able to respond to shock by reabsorbing sodium avidly. This therefore confirms a diagnosis of pre-renal failure. No other tests, including measurements of osmolality, of urinary sodium concentration alone, nor urine microscopy can reliably differentiate pre-renal from established renal failure. If an ultrasound was available, it would look normal or may be echo-bright.

Treat by urgent rehydration followed by furosemide. ✔ Rehydration should be with plasma, or normal saline/4.5% albumin (which is an effective plasma substitute). Give 10–20 ml/kg as rapidly as possible initially, and repeat if necessary. Thereafter give normal (0.9%) saline to fully correct the fluid deficit within 2 to 4 hours. The deficit can be estimated by multiplying the child's weight by the estimated percentage dehydration. For example a 6 kg infant estimated to be 10% dehydrated is deficient of approximately 600 ml. By the above guidelines he would receive between 60 and 240 ml of plasma or plasma substitute very rapidly, and the rest of the 600 ml as 0.9% saline.

Once rehydration has begun give furosemide 2 mg/kg orally or IV. If the blood pressure remains markedly depressed after rehydration, it may be due to cardiogenic shock; consider administering inotropes.

Established acute renal failure

Established ARF is due to acute parenchymal damage to the kidneys. In most cases the causes are exactly the same

as for pre-renal failure, but an increased severity or duration of the insult has led to death of some of the renal cells. Thus, the pertinent history and clinical signs are usually the same as for pre-renal failure. Other cases are due to directly toxic effects of drugs such as gentamicin, or poisons to the tubular cells. Some forms of glomerulonephritis may lead to ARF (see above), as may the arteriolar disease, haemolytic uraemic syndrome (HUS) (see below).

The laboratory diagnosis of established renal failure due to under-perfusion or an ischaemic insult can only be made reliably by calculating the FENa from a measurement of the sodium and creatinine concentrations in a plasma and a spot urine sample. The FENa is typically greater than 2% because the damaged tubules are usually unable to reabsorb sodium avidly. Again, attempts to use other laboratory criteria are unreliable. The history, clinical examination and laboratory confirmation of glomerulonephritis and HUS are described elsewhere.

The most vulnerable region of the kidney is the highly metabolically active mass of proximal tubule cells. If these cells alone die from the insult, this causes acute tubular necrosis (ATN) which will fully recover in 2 to 4 weeks if the child is maintained in good health during that period of renal failure. More severe insults may result in damage to some or all the glomeruli as well, which are in the renal cortex. Glomerular damage is irreversible, and acute cortical necrosis may therefore result in chronic or end-stage renal failure.

Fluid repletion and furosemide administration will not result in recovery of renal function. If an FENa is not available to distinguish between pre-renal and established ARF, it is sensible to give a trial of furosemide. Management thereafter consists of correcting the dehydration, as for pre-renal failure, and then careful maintenance of fluid and electrolyte balance and nutrition while it is hoped that some recovery of tubule cells will lead to recovery of kidney function. ***This is likely to require dialysis***. If no recovery has begun by 4 weeks it is unlikely to. There are no reliable imaging techniques that determine whether the child has ATN or cortical necrosis.

Post-renal ARF

Post-renal causes are due to obstruction to all of the urinary flow, and are rare. This will not occur if the flow from just one kidney is blocked. Causes in a child with two kidneys include a bladder stone obstructing the urethra, and in a child with a single kidney include a ureteric stone, or a pelviureteric junction narrowing (which are congenital, but often block intermittently and present late).

All these pathologies cause severe acute colicky abdominal pain. This is well localised in older children to either unilateral pain with ureteric obstruction, or lower abdominal pain with bladder neck obstruction. An ultrasound scan will reveal stones and dilatation of the urinary tract proximal to the site of the obstruction.

The treatment of post-renal ARF is to remove or bypass the obstruction. For a bladder neck stone obstruction, catheterise the child. Giving pain relief with an opiate analgesic may allow time for an obstructing urethral stone to pass, or for the intermittent blockage from a pelviureteric junction narrowing to clear. If not, the stone may need to be removed cystoscopically or by ureterolithotomy, or the upper renal tract drained by insertion of a percutaneous nephrostomy under ultrasound guidance. Once removal of the obstruction has allowed the renal function to recover, procedures such as surgical repair of the pelviureteric junction may be performed.

Ongoing management of persistent ARF

General

The management of ARF consists of the provision of good general care to an acutely ill child, plus the specific management of fluid and electrolyte balance, blood pressure, and the adjustment of some drug dosages. In many instances the limitations that need to be imposed to keep in metabolic balance compromise the care that can be given in other areas.

To manage these children safely requires the maintenance of meticulous fluid balance. To achieve this it is necessary to accurately measure all intakes and losses. For babies, stool and urine losses are best estimated by weighing their clean and dirty nappies. Insensible water losses need to be estimated. This is done most reliably by assuming it to be 300 ml/m^2 in temperate conditions, and to be higher in hotter climates and at low humidity (see Appendix 7.7 for estimation of body surface area). The best guide to the child's overall changes in fluid balance is to weigh the child twice daily. ✔

Nutrition, fluid and electrolyte balance

Adequate nutrition is important for recovery, but may be difficult to provide. If a child is old enough and well enough to eat solid food they are relatively easy to manage because they can obtain their requirements with little water. Aim to provide their normal calorie intake from carbohydrates and fats, and limit the protein intake to about 1 g/kg/day to minimise uraemia. It is necessary to limit the salt intake to prevent sodium retention and hypernatraemia, which leads to insatiable thirst, and hence fluid overload. It may be necessary to provide some of the sodium as bicarbonate to prevent acidosis, typically at a starting dose of 1 mmol/kg/day (note, 1 ml of an 8.4% sodium bicarbonate solution contains 1 mmol, and 1 g of powder contains 12 mmol). **Dietary potassium must be restricted (avoid the following in particular: bananas, tomatoes, coconut, citrus fruits or juices, chocolate) to decrease the risk of hyperkalaemia, and the dietary phosphate restricted (restrict milk and diary products but not breastfeeding) to reduce the risk of hyperphosphataemia.** Giving calcium carbonate with the food (for example, 0.5 to 2 grams with each meal) will bind the intestinal phosphate and reduce hyperphosphataemia as well as reducing the tendency to hypocalcaemia. ✔

Young infants who normally take milk, and children too ill to eat solid food, or with gastrointestinal involvement will either need nasogastric tube feeding ***or intravenous nutrition***. Either way, their nutrition has to be delivered in a large fluid volume. If the child has polyuric renal failure, or has high non-renal water losses such as

from diarrhoea or drain fluids this can be achieved. However, if the child is oligoanuric it is not possible to give sufficient nutrition without causing fluid overload which can lead to hypertension and pulmonary oedema. Concentrated fat-based oral feeds can be made up from ingredients such as double cream. ***For parenteral nutrition it is necessary to use sophisticated intravenous fluids with a high glucose content and individually adjusted sodium (and bicarbonate) concentrations, tailored to balance losses.***

The need for dialysis

It is not possible to both provide adequate nutrition and maintain stable water and chemical balance over a prolonged period in a child with oligoanuria. If such a child does not start to regain renal function, they will die unless dialysed. Hyperkalaemia is the commonest indication to start dialysis in centres where it is available. This is discussed below.

Severe metabolic acidosis is another important reason to dialyse if available. Treatment with sodium bicarbonate is limited because this may lead to massive sodium overload, and hence to dangerous levels of hypernatraemia, and to greater fluid retention. Fluid overload is worsened if hypoglycaemia occurs and needs to be treated with intravenous glucose solutions, and if other fluids are required such as platelets. Also, if the urea rises high enough to cause clinical uraemia (above 40 mmol/litre) it needs to be reduced by providing more non-protein calories, which means more water volume.

Hyperkalaemia

Minimum standards requirements: hyperkalaemia

- ECG monitoring
- Calcium gluconate
- Nebulised salbutamol
- Calcium resonium
- Sodium bicarbonate
- IV glucose 10% and insulin

This causes life threatening arrhythmias, especially in acute renal failure where other metabolic changes may exacerbate the risk (for example, hypocalcaemia). Aim to keep the plasma potassium below 6.5 mmol/litre in an older child and below 7.0 mmol/litre in neonates who appear to tolerate hyperkalaemia better.

✔ **There are 3 pharmacological approaches to managing a child with hyperkalaemia. First, reduce the chance of it causing arrhythmias, second remove potassium from the body, and third push potassium into the cells**. This last option only results in a temporary improvement because as soon as the treatment stops the potassium moves back out of the cells. Essentially this approach is only a holding treatment while a more effective therapy is prepared.

1) Reduce the effect of hyperkalaemia by increasing the plasma calcium concentration. Give 0.5 ml/kg (0.1 mmol/kg) of calcium gluconate 10% IV.
2) Give calcium resonium 1 g/kg orally or rectally, and repeat with 0.5 g/kg/12 hourly. This ion exchange resin exchanges potassium for calcium. It is not well tolerated.
3)
   - Give a β_2-adrenergic agonist, such as salbutamol. Nebulise 2.5 mg for children under 25 kg, and 5 mg in larger children, or give 4 micrograms/kg IV. This works rapidly, but the potassium will move back out of the cells within a few hours.
   - Alternatively, infuse a high concentration of glucose. Monitor the plasma glucose concentration and be prepared to infuse insulin beginning at a dose of 0.05 Units/kg/hour if it exceeds 12 mmol/litre. It is unsafe to mix the glucose and insulin and infuse together in children as it may cause hypoglycaemia. This necessitates close monitoring, an inevitable fluid load, and only lasts while it is continued.
   - Bicarbonate infusions push potassium into the cells. A dose of 2.5 mmol/kg may be infused over 15 minutes. If a solution of 8.4% is used, containing 1mmol/ml, it will increase the plasma sodium concentration by approximately 5 mmol/litre very quickly which may be hazardous. It is better to use a solution of 1.26% which is isonatraemic, but this requires that a volume of 17 ml/kg be infused, adding to fluid overload.

Haemolytic uraemic syndrome (HUS)

Minimum standards requirements

- Acute renal failure (see above)
- Blood transfusion

HUS is a common cause of established (parenchymal) acute renal failure (see above). Children with HUS fall broadly into two groups, according to their pathophysiological mechanisms. It is important to divide them clinically into diarrhoea-associated HUS (D + HUS) and diarrhoea-negative HUS (D − HUS).

D + HUS

This much commoner type occurs in otherwise normal children, and is triggered by a toxin that is produced by some colonic bacteria, including *Shigella* and *E. coli* O157. These may result from contamination of milk or meat by cattle faeces. Public health measures to identify a source of the organism are important in preventing and limiting outbreaks. Typically the child has approximately one week of bloody diarrhoea, and then becomes pale and mildly jaundiced (from haemolytic anaemia), may bruise and have petechiae (from thrombocytopenia), and develops oligoanuria. A blood film shows fragmented red blood cells and a low platelet count.

Antibiotics are not of benefit, and may worsen the condition by causing the acute release of more bacterial toxin. Blood transfusion is virtually always needed (usually when the Hb falls below 6 g/dl). Platelet transfusion may exacerbate the condition, and should only be used in the face of uncontrolled bleeding. There is no evidence that any specific medication is of benefit. Management is as for other children with ARF (see above). ***Most children with D + HUS in centres that can offer it are dialysed.*** Not all children that are dialysed in such centres would have died without treatment, but mortality in advantaged countries is under 5%, and much higher than this elsewhere.

In a minority of cases, D + HUS can affect other organs, sometimes severely. Effects can include bowel perforations, pancreatitis with diabetes mellitus, and cerebral involvement with fits, coma and death.

The long-term outcome for children that survive D + HUS is relatively good. Most appear to fully recover renal function, though a minority have persistent hypertension or proteinuria. Few develop end-stage renal failure.

D − HUS

This much rarer type is due to an inherited (dominant or recessive) disorder, often associated with a functional or actual deficiency of factor H, so a minor trigger (such as a minor viral illness) will precipitate the typical clinical and haematological HUS picture, but without a diarrhoeal prodrome. Typically D − HUS patients fare much worse long-term than D + HUS patients.

Acute peritoneal dialysis

Indications

Children with acute renal failure can be considered for peritoneal dialysis if their biochemical control is not safe despite careful treatment (see section on management of acute renal failure above). Though the specific indications for initiating peritoneal dialysis vary from case to case, the commonest reason is a high and rising plasma potassium concentration (for example above 6.5 mmol/litre in an older child, or 7 mmol/litre in a neonate). Others include a urea above 40 mmol/litre, a phosphate above 3.5 mmol/litre, or acidosis with a bicarbonate below 12 mmol/litre, as well as hypertension or pulmonary oedema due to fluid overload.

The primary underlying cause for needing to proceed to dialysis is usually anuria or severe oliguria. This is because even a moderate urine flow will prevent fluid overload if the intake is restricted, and because it "makes space" for biochemically appropriate replacement fluid. Even poor quality urine contains potassium, so replacement with potassium-free fluid allows a net loss. Also, urinary sodium losses can be replaced with intravenous sodium bicarbonate to counter acidosis, and a high infused glucose concentration will reduce catabolism and so minimise urea, potassium and phosphate production. Take advantage of all fluid losses; diarrhoeal losses will "make space" just as well as urine losses.

Practical techniques

Catheter

Ideally, a catheter with side-holes should be inserted so that its tip lies in or near one of the iliac fossae. The ideal catheter is a cuffed sialastic Tenckhoff which has a series of side holes and an end which is cut off straight, but these are expensive and require to be inserted through a peel-away sheath (usually in the midline below the umbilicus). It is possible to dialyse adequately using other more readily available catheters that have side-holes, such as chest drains. These are usually inserted over a metal trocar, and have a tapered tip with an end-hole that is considerably smaller than the diameter of the tube lumen, which can lead to difficulties with blockage with omentum (see below).

Insertion of catheter

- ***This must be a strictly aseptic technique performed either under general anaesthetic, or under sedation/systemic analgesia (see Chapters 1.13 and 1.27) and local anaesthetic.***
- ***To prevent fluid leakage it is essential that the catheter is inserted through the skin with a very tight fit; using a larger skin hole and stitching it closed will inevitably leak in time. Cut a skin slit that is obviously smaller than the tube, and stretch it with a surgical clip or stitch holder.***
- ***Before introducing the catheter, insert an intravenous cannula through the skin cut and fill the abdomen with about 40 ml/kg 0.9% saline until the abdominal wall is fairly tense.***
- ***To insert the catheter through a tight hole requires considerable force, and this is best done by pushing the catheter and trocar tip into the dilated skin slit as far as possible, and then suddenly advancing it with a sharp forceful lunge through the tense abdominal wall. Grip the catheter and trocar tightly about 3 cm from its tip to act as a stop as it pops into the abdomen (the risk of causing damage is greatly reduced by the presence of sufficient instilled fluid).***
- ***To further minimise the risk of trauma, it is better is to enter the upper quadrant lateral to the rectus sheath, and aim towards the opposite iliac fossa, than to use an infra-umbilical approach. Be aware of a possibly enlarged spleen or liver.***
- ***Once sited, test that fluid flows rapidly in and out, before securing with a skin stitch and sterile dressing.***

Problems with omentum

It is common for omentum to wrap round the end of the catheter, and for some to enter the end hole. This slows or stops drainage because the omentum

is sucked further into the lumen, but has little effect on filling because the omentum is washed back towards the catheter tip, and the fluid exits through the side holes. Deal with it as follows:

- *The omentum can often be forced out by rapidly injecting up to 50 ml of dialysis fluid or 0.9% saline into the catheter under pressure.*
- *If this fails, withdraw the catheter from the abdomen using full aseptic technique. If the omentum has become detached, simply reinsert the catheter, and resume dialysis.*
- *If (as usually happens) the catheter comes out with the omentum attached, detach it, and gently pull more omentum out, tie round it with an absorbable suture near to the skin surface, cut off the excess, and return the omentum into the abdomen, using the stitch to obtain easy purchase, and replace the catheter.*

Fluid and cycles

- *Run the dialysis fluid in through a giving set with a burette, and with the bag held about 1 metre above the patient, and leave it to dwell for 30 minutes. Allow it to drain by gravity through a Y-connector into a sealed bag for about 10 to 15 minutes; by then, it should have drained about as much as was instilled, and the flow should have stopped.*
- *The osmolality of the dialysis fluid determines the amount of water that is drawn off (ultra-filtered) during each peritoneal dialysis cycle, and this is adjusted by varying the glucose content. Typical glucose concentrations available are 1.36% (standard) and 3.86% (high osmolality) bags. Start with 1.36% glucose.*
- *Add heparin 1000 units/litre to the fluid initially to prevent any blood from the insertion clotting the catheter; discontinue once the effluent fluid looks clear.*
- *Start with 10 ml/kg cycles of dialysis fluid for the first two days; using this small volume minimises the risk of a peritoneopleural leak of dialysate.*
- *The first cycle balances are unreliable because there is always a sump of fluid left, but after that the ultrafiltrate required is the volume of fluid that needs to be removed to correct any overload, plus an amount equivalent to the urine that would normally be passed (so just a little less than the normal fluid intake).*
- *If the ultrafiltrate is too little, increase the glucose concentration of the dialysate by giving some cycles of 1.36% and some of 3.86% glucose. Continue to review the fluid balance, and vary the proportion of cycles of each strength as necessary.*
- *Increase the cycle volume by 10 ml/kg every two days until tolerance, or a maximum of 40 ml/kg. As the cycle volume increases, it is not necessary to dialyse so intensively. Either continue with 30 minute dwells, but just for part of the day (for example, 8 hours overnight), or lengthen the dwells, eventually moving to chronic ambulatory peritoneal dialysis (CAPD) where the fluid is left in the peritoneum all the time, and exchanged four to six times per day.*

Biochemical control

The fluid sodium, calcium and magnesium contents are similar to plasma, and it contains lactate which is converted to base, so is equivalent to bicarbonate. Cycling therefore tends to keep the plasma concentrations stable. Peritoneal dialysis fluid has no potassium, urea or creatinine, so these are removed.

- *Urea equilibrates rapidly, so is cleared well, allowing children to have a normal protein intake.*
- *Creatinine is removed slowly, so peritoneal dialysis never brings the plasma levels to normal. This is useful because creatinine is not toxic, and its plasma concentration continues to provide a measure of intrinsic renal function.*
- *Sometimes the dialysis required to control fluid or urea excretion is sufficient to cause hypokalaemia. If so, reduce the potassium dialysis clearance by adding up to 3 mmol/litre potassium chloride to the dialysate bags (NEVER use more than this – if the potassium is still too low, give extra orally or intravenously).*

Peritonitis

Infection is the major hazard of peritoneal dialysis, and produces a cloudy dialysis effluent in the drainage bag due to white cells. Prevention is crucial, by scrupulous hand washing and avoiding touching the open tubing ends while changing peritoneal dialysis bags, and by changing connections as infrequently as possible.

- *Monitor constantly by inspecting the effluent fluid clarity.*
- *Do daily microscopy for white cells (should be $<$50 cells/microlitre; see section on microscoping urine).*
- *If cloudy, and microscopy confirms the presence of large numbers of white cells (over 100, but typically several hundred), culture a sample of fluid, and start treatment at once by adding heparin (to stop blockage of tube holes with fibrin) and antibiotics to peritoneal dialysis bags and revert to continuous cycling if not still doing that. Start with vancomycin and ceftazidime, and adjust according to culture and sensitivity results. Concentrations of antibiotics that may be added to peritoneal dialysis fluid are:*
  - *vancomycin 25 mg/litre*
  - *ceftazidime 125 mg/litre*
  - *ampicillin 125 mg/litre*
  - *flucloxacillin 250 mg/litre*
  - *gentamicin 8 mg/litre*
- *Continue continuous cycling until white cells $<$50 cells/microlitre on two samples, 12 hours apart, then return to previous dialysis cycles,*

adding peritoneal dialysis antibiotics for 14 days.

- ***If accidental contamination occurs, such as touching the open dialysis catheter during a bag exchange, or a fluid leak from a connection or punctured bag, add vancomycin and either ceftazidime or gentamicin to the dialysis fluid for the next 12 hours.***
- ***Fungal peritonitis is difficult to clear; it is best to remove the catheter and treat systemically until the peritonitis resolves.***

Chronic renal failure

Minimum standards requirements

- Sodium bicarbonate
- Calcium carbonate
- Vitamin D with serum calcium monitoring

Background

Chronic renal failure (CRF) is more frequent in boys than girls because its commonest cause is renal dysplasia secondary to severe antenatal vesicoureteric reflux, often associated with posterior urethral valves. It can also follow almost any form of acute renal failure.

It is relatively easy to improve the quality of life for children with milder forms of CRF by simple treatments, especially in older children. In its more severe forms, CRF is very difficult to treat effectively, requiring expensive drugs and intensive laboratory monitoring. Very young children with CRF are particularly difficult to manage as they usually have marked anorexia, and failure to thrive. Successful treatment requires a massive family and medical input, highly expensive drugs, and a complex medical infrastructure which only has limited availability worldwide at present. ***Each country should have a specialised centre which can provide care for such children.***

Progression of CRF

CRF tends to worsen progressively through childhood. This is mainly because dysplastic or damaged kidneys may not grow in parallel to body growth, and renal function becomes outstripped by demand. Deterioration is likely to be quicker if the child has hypertension, or has recurrent urine infections with continuing reflux; long-term prophylactic antibiotics are then appropriate (see above).

Management

Water, sodium and potassium

Children with dysplastic kidneys usually have **polyuric renal failure** in which they lose water and salt, and often potassium in an uncontrolled way. Consequently, they have a persistent thirst, and can become dehydrated extremely rapidly if they vomit persistently. They need intravenous fluids early particularly if there is an episode of gastroenteritis. Hyperkalaemia due to severe CRF occurs relatively late in children with polyuria.

Supplementing with sodium bicarbonate, or salt as needed, can improve wellbeing and growth. For each, start by adding about 1 mmol/kg per day. For bicarbonate, increase daily until the plasma concentration is in the normal range. The total extra sodium needed is best judged by measuring lying and standing blood pressures to detect postural hypotension; a fall in plasma sodium concentration is a very late event. Note that 1 mmol bicarbonate = 84 mg, so 1 g contains about 12 mmol, and that for intravenous use, 8.4% bicarbonate solution contains 1 mmol/ml. For sodium chloride (salt), 1 mmol = 57 mg, so 1 g contains about 18 mmol sodium. For intravenous use, each litre of 0.9% saline contains 150 mmol, and strong sterile sodium chloride solutions can be used to increase the sodium concentrations of standard intravenous fluids; for example a 30% solution contains 5 mmol sodium per ml.

Children with **oliguric renal failure** are more difficult to manage because they require salt and water restriction to prevent hypertension, and potassium restriction to prevent hyperkalaemia. ***When dialysis is available, an intolerable diet or fluid restriction are the commonest indications to begin this treatment.***

Calcium and phosphate

CRF can lead to abnormalities of the plasma calcium and phosphate concentrations, and these can cause rickets and hyperparathyroidism (renal osteodystrophy). These can result in bone pain, limb deformities, and fractures (especially slipped femoral capital epiphyses). The primary problem is phosphate retention due to a reduced glomerular filtration rate. This causes a high plasma phosphate, which in turn leads to a low plasma calcium by mass action, and by suppressing the enzyme 1α-hydroxylase, thus lowering the concentration of circulating activated 1α-hydroxyvitamin D. A primary lack of 1α-hydroxylase enzyme from destruction of kidney tissue is rare except in very severe CRF.

Treatment is therefore aimed at reducing the phosphate intake, either directly by dietary restriction (less meat, less dairy produce) or by giving **calcium carbonate** with meals. This binds with the phosphate in the gut, and prevents its absorption. The dose needed is very variable; start at about 50–100 mg/kg, divided among the day's meals, and titrate the dose (if biochemical monitoring is available) to keep plasma phosphate levels at the bottom of its normal range. This commonly also results in a rise of the plasma calcium levels into the normal range; because of this, it is seldom necessary to treat mild CRF with **1α-hydroxyvitamin D_3**. If it is needed, start with about 20 nanograms/kg once daily, and titrate the dose up until the plasma calcium is normalised. It is extremely potent, and using it without regular monitoring can easily lead to severe hypercalcaemia which can result in permanent calcification of tissues, including the renal medullae.

Anaemia
Severe CRF leads to anaemia because the kidneys fail to produce enough erythropoietin. Treatment by repeated transfusions is unsatisfactory because blood is often scarce, carries infective risks, is always expensive, and leads eventually to iron overload.

Growth
Many factors lead to growth failure in children with CRF.

- In older children, attention to fluid and electrolyte intakes, prevention of acidosis with bicarbonate supplements, and control of the bone biochemistry help considerably. Control of uraemia by encouraging a diet with about 1g protein/kg daily and a high carbohydrate intake will also contribute to good growth.
- In young children, the problems are much greater. They are often extremely anorexic; most babies with severe CRF virtually do not feed, and only survive if tube fed for months or years. Many also vomit excessively. Even when supplemented with tube feeds, very young children with CRF remain small.

3.8 Vesicoureteric reflux

Manoj Shenoy and Leela Kapila

Minimum standards requirements

- Ultrasound scanning
- Antibiotics: trimethoprim, nitrofurantoin, nalidixic acid
- ***Surgery***

Introduction

Vesicoureteric reflux (VUR), the abnormal flow of urine from the bladder into the upper urinary tract, occurs in about 1 in 100 in the general population and is more common in girls. It is the commonest cause of a documented urinary tract infection (UTI). **Hence all children with a proven UTI need further investigation.** Reflux nephropathy is among the most frequent causes of hypertension and chronic renal failure in children and young adults.

Renal scarring is an acquired phenomenon that usually occurs during the first few years of life and rarely after the age of 5 years.

Grades of VUR (International Reflux Study Group Classification)

Grade I –	Partial filling of an undilated ureter
Grade II –	Total filling of an undilated upper tract
Grade III–	Dilated calyces but fornices sharp
Grade IV–	Blunted fornices and degree of dilatation greater than in lower stages
Grade V –	Massive hydronephrosis and tortuosity of the ureters

Clinical presentation

- VUR is almost always in conjunction with an associated UTI.
- Rarely a cause of flank pain.
- Symptoms may include fever, lethargy, anorexia, nausea, vomiting and failure to thrive.
- **Fever is the single most important symptom to differentiate children with upper tract (pyelonephritis) from those with lower tract (cystitis) infections.**

Investigations

The minimal acceptable standards of investigation would include:

- Ultrasonography (US)
  Good for detection of dilatation not good at demonstrating scars or reflux
- ***Micturating cystourethrogram***
- Investigations in siblings
  VUR occurs in up to 30% of siblings and families should be made aware of this.

Medical management

- Spontaneous resolution most often in the first 2–3 years after diagnosis and then at 10–15% per year.
- The main goal is the prevention of ascending UTI and renal scarring.
- Prevent UTI through:
  - proper wiping techniques
  - frequent voiding
  - avoidance of constipation
  - ensure low-pressure voiding
  - continuous antibiotic prophylaxis usually maintained for two years
- Trimethoprim 2 mg/kg/day is the usual prophylactic agent.
- If breakthrough infections resistant to this occur, then a suitable alternative prophylactic such as **nitrofurantoin** (1 mg/kg/ day) or **nalidixic acid** (7.5 mg/kg twice daily) may be used.

If medical treatment fails the child will require ***reimplantation of the ureters.***

3.9 Acute liver failure

Alastair Baker and Anil Dhawan

Minimum standards requirements

- Vitamin K
- IV glucose 10%
- Oxygen
- Lactulose
- Blood transfusions and clotting factors
- Ranitidine/antacids
- Antibiotics and antifungal treatment
- High carbohydrate diet
- *N*-acetylcysteine
- Measurements of prolonged blood clotting times

Introduction

Unlike adults, acute liver failure (ALF) in children may not be accompanied by encephalopathy.

Prolonged prothrombin time (PTT) or Prothrombin International Normalised Ratio (INR) indicates coagulopathy from absence of coagulation factors synthesised in the liver and is a valuable indicator of liver dysfunction. However, coagulopathy in the presence of liver dysfunction can also result from vitamin K deficiency (usually due to prolonged cholestasis) and consumption of coagulation factors due to disseminated intravascular coagulation (DIC).

Definition

Based on the above, ALF is present in children when coagulopathy accompanies liver disease but is not due to DIC or a lack of vitamin K (Table 3.9.1). Administration of IV or IM vitamin K (300 micrograms/Kg aged 1 month to 12 years: 10 mg for >12 years) ensures that remaining coagulopathy is due to failed production (liver failure) or excess consumption (DIC). Markers suggesting DIC, rather than ALF, include a low platelet count, compatible blood film (fragmented cells, schistocytes) and a serum bilirubin that is predominantly unconjugated.

Table 3.9.1 Clinical Features of ALF

- Bruising/petechiae/bleeding secondary to deranged clotting (INR $>$ 4 = 90% mortality).
- Jaundice with tender hepatomegaly or a liver that is reducing in size over days.
- Encephalopathy complicated by raised intracranial pressure.
- Metabolic alkalosis from failure of the urea cycle associated with a low serum potassium.
- Failure to maintain normoglycaemia.

Diagnosis of ALF

- History may establish previous episode of shock, evidence of ingestion of toxic mushrooms or drugs, or exposure to infection such as *Salmonella typhimurium* (Tables 3.9.2 and 3.9.6).
- Examination may show features of acute portal hypertension with liver tenderness to suggest Budd-Chiari syndrome, or lymphadenopathy to suggest malignancy (Table 3.9.3).
- Urine should be tested for bilirubin, urobilinogen and reducing substances.
- Stools should be examined for colour.
- ***Tests to establish many of the causes of ALF require sophisticated laboratory facilities which may not be available.***
- The cause is more likely to be diagnosed from local epidemiology.
- A blood film and an INR or prothrombin ratio should be measured.
- Full septic screen, excluding lumbar puncture because of coagulopathy, should be performed (include fungal cultures and chest *X* ray).

Table 3.9.2 Causes of ALF

Type	Cause
Infective viral	Hepatitis A-D, HIV, parvovirus, herpesvirus, enterovirus, adenovirus, echovirus, varicella, yellow fever, Lassa, Ebola, Marburg, Dengue
Bacterial, protozoal	Leptospirosis, Typhoid, Malaria
Metabolic	Wilson's, tyrosinaemia*, urea cycle disorder, galactosaemia*, mitochondrial disorders, haemochromatosis*, Niemann-Pick C*
Drugs	Paracetamol, antituberculous drugs, halothane, carbamazepine, sodium valproate
Toxins	*Amanita phalloides*, heat stroke, shock – all causes
Autoimmune	Antismooth muscle, anti liver–kidney microsome (LKM) antibody positive, giant cell hepatitis with haemolytic anaemia
Infiltrative	Leukaemias, lymphomas, haemophagocytic lymphohistiocytosis
Vascular	Budd–Chiari syndrome, veno-occlusive disease (may follow ingestion of bush teas)
Cryptogenic	"Non-A, non-B hepatitis"

* Represent neonatal causes.

Table 3.9.3 Complications of ALF

- Encephalopathy and raised intracranial pressure, convulsions
- Hepatorenal syndrome
- High output cardiac failure
- Hepatopulmonary syndrome
- Acid–base disturbance; initially alkalosis with hypokalaemia followed by metabolic acidosis from multiorgan failure
- Gastrointestinal bleeding, including early development of oesophageal varices
- Pancreatitis
- Bone-marrow aplasia
- Sepsis, particularly Gram-negative and fungal, pulmonary (including aspiration) and septicaemia

Managing children with ALF

- Refer to a specialised centre if one exists in the country.
- Undertake frequent reviews, hourly observations and individual nursing.
- Blood tests for coagulation, electrolytes, blood glucose and blood count should be performed frequently (ideally 8 hourly).
- Hypoglycaemia and hypokalaemia must be detected and corrected.
- Children with encephalopathy should be nursed with their heads elevated at 30° above horizontal and without neck flexion (to decrease intracranial pressure and minimise cerebral irritability).
- Children with agitated encephalopathy of grade II–III represent a major management problem as they may pull out monitoring equipment and intravenous lines. Sedation will worsen their encephalopathy.
- Maintain blood glucose between 4 and 9 mmol/litre using a restricted fluid volume (Table 3.9.5) consisting of a minimum concentration of 10 % glucose (given IV or orally). 20% glucose is the preferred solution but is irritant to peripheral veins and is best given into a central vein or better, if tolerated, orally or via a nasogastric tube.
- A metabolic alkalosis resulting from a failure of the urea cycle may cause hypokalaemia as a result of a shift of potassium into cells. This hypokalaemia can worsen encephalopathy and should be corrected enterally or IV.

Table 3.9.4 Grades of hepatic encephalopathy

Grade I	Irritable. Inappropriate behaviour. Difficulty in drawing. Lethargy. Mildly depressed awareness. Tremor or flap (slow wave in outstretched extended hand).
Grade II	Aggressive outbursts. Bad language. Unable to stay still. Pulling at intravenous cannulae/ plaster, etc.
Grade III	Mood swings. Irritable, odd behaviour. Not recognising parents. Photophobia.
Grade IVa	Mostly sleeping but rousable. Incoherent. Sluggish pupils. Hypertonia with or without clonus and extensor spasm.
Grade IVb	Absent reflexes. Irregular gasps with imminent respiratory failure.* Bradycardia. Unresponsive to painful stimuli.

* Associated with raised intracranial pressure.

Table 3.9.5 Fluid balance

Weight/fluid	Normal requirements	Liver failure*
Body weight		
$<$10 kg	120–150 ml/kg/day	60–80 ml/kg/day
10–20 kg	90–120 ml/kg/day	40–60 ml/kg/day
$>$20 kg	50–90 ml/kg/day	30–50 ml/kg/day
Fluid type		Glucose $>$10% (adjust according to blood glucose levels)
K^+	1.0–3.5 mmol/kg/day	Nil while anuric
Na^+	1.5–3.5 mmol/kg/day	Nil added
Other fluids		Albumin 20% 5 ml/kg
For transfusion		Fresh frozen plasma 10–20 ml/kg

* Approximately two thirds maintenance.

- Hypoxaemia should be prevented with oxygen by nasal cannulae or face mask.

- Strict fluid balance is essential (see Table 3.9.5).
- Allowance should be made for a hot climate and 10% extra fluid given for each degree of fever.
- Strict monitoring of urinary output and fluid balance is required. Aim for a urine output of not less than 0.5 ml/kg/hour (determined by weighing nappies or measuring output).
- Daily weights are useful if the child can be moved and will allow greater precision in fluid balance.
- ***If possible/appropriate insert a central venous line and aim to provide a central venous pressure (CVP) of 6–10 cm H_2O initially to give a normal blood pressure. Increased CVP may be required to compensate for an increased cardiac output or to treat reduced cardiac performance seen as the liver failure progresses.***
- ***Patients requiring inotropes are developing multiorgan failure and have a very poor prognosis.***
- Stop oral protein initially and during recovery gradually re-introduce 0.5–1 g/kg/day in oral or nasogastric feeding.
- A high-energy intake, predominantly of dietary carbohydrate, should be promoted to prevent protein catabolism and thereby an increased serum ammonia. In the absence of products such as Maxijul, uncooked corn-starch may be used as a source of carbohydrate. It may be given up to two hourly to provide predicted energy requirements and may also help to maintain normoglycaemia.
- Lactulose 5–10 ml 2–3 times a day is given to produce two to four soft and acid stools per day (omitted if diarrhoea occurs).
- Maintain normothermia by environmental measures (but NOT with paracetamol).
- Give one dose of IV or IM vitamin K (300 micrograms/kg aged 1 month to 12 years: 10 mg for >12 years) to attempt correction of prolonged clotting time.
- If frank bleeding (gastrointestinal or other) consider fresh frozen plasma or ***cryoprecipitate*** at 10 ml/kg IV.
- A prophylactic H^2-blocking agent (for example ranitidine 2 mg/kg twice daily orally or IV) is given with oral antacid (for example sucralfate 250 mg four times a day 1 month to 2 years, 500 mg four times a day 2–12 years, 1 g four times a day 12–18 years) to prevent gastric/duodenal ulceration.
- Broad-spectrum antibiotics for example cephalosporin plus amoxycillin or penicillin plus gentamicin should be used prophylactically.
- Treat any confirmed sepsis aggressively.
- Systemic fungal infection may require IV amphotericin (250 micrograms to 1 mg/kg/day) or oral fluconazole (10 mg/kg once daily).
- Give prophylactic oral nystatin mouth washes (100 000 IU (1 ml) four times a day).
- Manage hypotension with IV colloids and ***possibly dopamine and epinephrine infusions if central venous access has been achieved*** (see Chapters 1.25 and 6.6).
- *N*-acetylcysteine 100 mg/kg/day as continuous infusion should be given in all forms of liver failure. In paracetamol poisoning it provides a source of sulphydryl groups for renewing glutathione. In other causes of ALF it may scavenge free radicals and improve tissue oxygen delivery.
- If **paracetamol overdose** is suspected or ascertained, *N*-acetylcysteine must be started immediately, whatever the time between the alleged overdose and the visit to the hospital. Histories after overdose are often misleading with multiple administrations and other drugs not immediately admitted. *N*-acetylcysteine is given IV at 150 mg/kg over 15 minutes as a loading dose then 100 mg/kg over 12 hours then 100 mg/kg/day as a continuous infusion until INR is normal.

Prognosis of paracetamol overdose

- The most important prognostic parameter for ALF is metabolic acidosis. Even in the presence of a very prolonged INR, a patient who is not acidotic will have an 80% chance of survival. A plasma pH <7.25 ***(if blood gas measurement is available)*** indicates a 95 % risk of mortality.
- Other factors predicting a poor outcome are grade III–IV hepatic encephalopathy and oliguric renal failure (usually occurring 3–4 days after onset).

Prognosis of ALF

Adverse factors are as follows:

- Age <2 years
- INR ≥4

mortality >90%

- Serum bilirubin >350 micromol/litre
- Grade III or IV encephalopathy
- Non A non B hepatitis
- Drug induced.

Table 3.9.6 Poisoning or toxic reactions associated with ALF

- Paracetamol (acetaminophen)
- Mushrooms particularly *Amanita phalloides* and similar species
- Carbon tetrachloride
- Copper
- Iron
- Halothane and other volatile anaesthetic agents
- Sodium valproate
- Carbamazepine
- Phenytoin
- Phenobarbitone
- Isoniazid
- Cytotoxic drugs
- Irradiation

Galactosaemia

- A defect of galactose-1-phosphate uridyl transferase revealed in the perinatal period when affected infants are first exposed to milk feeding.
- Infants present with vomiting, hepatitis, liver failure and DIC often with septicaemia.
- Symptoms settle when feeding is discontinued.
- Hypoglycaemia is seen in the majority.
- Fanconi's nephropathy explains the presence of galactose in the urine giving the characteristic "clinistix negative, clinitest positive" side-room test pattern when the infant is receiving feeds.
- Management consists of the ***removal of galactose from the diet*** and standard management of liver failure and sepsis.

Further reading

Badhuri BR, Mieli-Vergani G. Fulminant hepatic failure: paediatric aspects. *Semin Liver Dis* 1996; **16**(4):349–55.

3.10 Chronic liver disease

Alastair Baker and Anil Dhawan

Minimum standards requirements

- Hepatitis B vaccine
- Vitamin K
- Fat-soluble vitamins (A, D, E, K)
- Diet high in PUFAs

The liver is strategically positioned between the effluent from the gastrointestinal tract and the systemic circulation. It acts as a filter of organic and inorganic substances, microorganisms and their breakdown products including endotoxins. Its anatomical position is the key to its nutritional homeostatic role.

Although chronic liver disease (CLD) can often only be cured only in specialised centres in rich countries where transplantation is available (costs are over $100 000 per case), much can be done to relieve suffering in children with this condition.

Clinical symptoms and signs of CLD

Jaundice

When accompanied by dark urine and pale stools is characteristic of cholestatic liver disease. The urine of infants should not contain significant colour or stain the nappy, and yellow urine strongly suggests obstruction of bile flow. Yellow sclerae suggests cholestatic jaundice but are difficult to detect in small infants. Particular emphasis should be placed on examining stool and urine as the history, particularly from first-time parents, can be misleading. White stool, or stools the colour of cream cheese or uncooked pastry, are clearly abnormal, while pale yellow, pale green or pale brown stools may also raise concern about liver function.

Hepatomegaly

Healthy infants may have up to 2 cm of liver edge palpable below the costal margin but the texture is soft. An abnormally hard texture or irregular inferior margin strongly suggests established liver disease with fibrosis/cirrhosis. Changed liver conformation with prominence in the midline but impalpable right lobe suggests collapse, regeneration and the development of cirrhosis. Tenderness of a smoothly enlarged liver suggests a rapid recent increase in liver size, for example in acute hepatitis and also heart failure.

Splenomegaly

This occurs in portal hypertension but, as children get older, a larger spleen can be accommodated beneath the ribs.

Coagulopathy

In the context of cholestasis, this results from a failure of absorption of sufficient vitamin K. In infants this may present as haemorrhagic disease of the newborn. Infants should not normally suffer spontaneous bleeding and fresh blood from sites such as the umbilicus or nares should always prompt a search for evidence of vitamin K malabsorption or liver disease. Jaundice may be mild in such cases.

In liver disease and coagulopathy unresponsive to vitamin K but without consumptive coagulopathy, liver synthetic failure must be present. The degree of coagulopathy is the most sensitive index of liver impairment in children.

Hypoglycaemia

More common in acute liver failure.

Encephalopathy

Also more common in acute liver failure (see Chapter 3.9). Hepatic encephalopathy may be insidious with educational failure, poor impulse control, bizarre behaviour and absences noted intermittently over months or years. Improvement may be associated with a low protein diet down to no less than 1 g/kg/day, lactulose to give acid stools and thereby change the gut flora in favour of organisms less likely to produce amines associated with encephalopathy, and sodium benzoate 250 mg/kg/day in 2–4 divided doses (to reduce plasma ammonia levels).

Ascites

This is seen in advanced CLD.

Cutaneous manifestations

These include pruritus, liver palms, cutaneous shunts, clubbing, white nails and xanthomas.

Hepatopulmonary syndrome

This is a progressive cyanosis without lung disease, associated with low pulmonary artery pressure. Exertional dyspnoea is a frequent early feature. Type 1 implies pulmonary capillary vasodilatation and improves at least in part with inspired oxygen while Type 2 implies fixed shunts without response to oxygen. Garlic is a current anecdotal treatment (4–10 capsules per day but there is no controlled evidence).

CLD may be present without detectable symptoms or signs (for example, chronic viral hepatitis B, a highly prevalent infection in disadvantaged countries, can be present for decades proceeding to cirrhosis without any external evidence).

Investigations into CLD

Consider liver dysfunction according to three categories (Table 3.10.1). These are:

- Cholestasis: impairment of bile flow with consequent reduction of intraluminal bile salt concentration and associated conjugated hyperbilirubinaemia
- Portal hypertension (PHT) with associated hypersplenism and the effects of portosystemic shunting
- Hepatocellular impairment (cell dysfunction) with failure of synthetic and homeostatic function.

Table 3.10.1 Clinical features of CLD

Clinical feature	Cholestasis	PHT	Cell dysfunction
Jaundice	Conjugated	–	Mixed if severe
Pruritus	+	–	–
Leuconychia	+	–	–
Clubbing	+	–	–
Fat-soluble vitamin deficiency	+	–	–
Xanthomas	+*	–	–
Splenomegaly	–	+	–
Cutaneous shunts	–	+	–
Other cutaneous stigmata	–	+	+
Hypersplenism	–	+	–
Hepatopulmonary syndrome	–	+	+
Oesophageal varices	–	+	–
Ascites	–	+	+
Encephalopathy	–	+	+
Dependent oedema	–	–	+
Malnutrition	+	+	+

* But not in familial intrahepatic cholestasis.

Table 3.10.2 Laboratory features of CLD

Laboratory feature	Cholestasis	PHT	Cell dysfunction
Serum bilirubin	Conjugated	Normal	Normal or mixed
Serum albumin	Normal	Normal	Low
Serum cholesterol	High**	Normal	Low
Prothrombin time/ratio	Normal***	Normal*	Prolonged if severe

* Implies minor prolongation seen in portal vein thrombosis.

** Except in familial intrahepatic cholestasis types 1 and 2.

*** If adequate vitamin K.

Table 3.10.3 Basic investigations in liver disease

Investigation	Role
Serum bilirubin, total and conjugated	Conjugated elevated in cholestasis Unconjugated elevated in hepatocellular injury
Urine bilirubin	Present in cholestasis
Serum aspartate aminotransferase, alkaline phosphatase, gamma-glutamyl transpeptidase	Elevated in hepatocellular injury plus cholestasis
Serum sodium, potassium, urea, creatinine, albumin, glucose	Hepatocellular injury
Full blood count, prothrombin time or International Normalised Ratio (INR)	Coagulopathy in liver failure, also in cholestasis due to vitamin K malabsorption
Hepatitis A IgM, hepatitis B surface (S) antigen, e antigen, S antibody, HIV antibody, ***toxoplasma, rubella, herpes, cytomegalovirus***, syphilis antibodies	Congenital/acquired infections
Serum total protein, immunoglobulins	Abnormal in autoimmune disease
Alpha-fetoprotein	Elevated in liver tumour
Plain *X* rays – spine, cardiac silhouette	Alagille syndrome, dextrocardia rarely in biliary atresia
Ultrasound	Biliary, portal, parenchymal abnormalities
Eye review for Kaiser-Fleischer rings ***and embryotoxon***	Wilson's Disease, Alagille syndrome

Clinical findings (Tables 3.10.2 and 3.10.3) can be interpreted according to this classification although some, for example ascites, are represented in more than one category. Transudative ascites can result from a combination of increased portal pressure and low plasma oncotic pressure from low serum albumin. Serum albumin reflects liver synthetic function but also depends on nutritional status and losses, for example via the gastrointestinal tract or kidneys. Thus it is necessary to consider all clinical features supported by basic laboratory parameters when possible to evaluate the severity of liver disease in a given patient.

A precise diagnosis of the various causes of CLD is often not possible without specialised and expensive investigations.

Causes of CLD

Cholestatic CLD: diagnosis and management

Cholestasis is most frequently seen as a complication of the neonatal hepatitis syndrome. The commonest defined diagnosis causing the syndrome is biliary atresia (BA), an obliterative inflammatory condition affecting the intra- and extrahepatic biliary system exclusive to the perinatal period. Infants present typically with jaundice, pale stools and dark urine. Untreated, biliary atresia progresses to biliary cirrhosis and death from decompensated liver function or associated sepsis within two years in 95% of cases. It is the commonest individual congenital cause of severe liver disease in children in all known populations occurring in 1 in 9 000–16 000 live births.

Rare and difficult to treat causes of cholestatic CLD

- Alagille syndrome: a condition characterised by cholestasis of variable severity associated with syndromic features.
- Progressive familial intrahepatic cholestasis (PFIC): a clinical syndrome of cholestasis complicating neonatal giant cell hepatitis.
- Neonatal sclerosing cholangitis: a rare condition which may mimic biliary atresia.

Consequences of cholestasis include:

- **Pruritus**. This is a particularly troublesome symptom resulting in disruption for the whole household, especially at night. Persistent scratching can be complicated by secondary infection of broken skin and blood staining of clothes and bedclothes. Early onset, before 7 months of age, heralds profound cholestasis and a poor prognosis. Treatment is often unrewarding. First-line management should be **cholestyramine** starting dose <6 years 2 g ($\frac{1}{2}$ sachet)/day, >6 years 4 g (1 sachet)/day. In practice 1 sachet/day from diagnosis and up to 6 sachets/day rarely. According to response increase to a maximum of 24 g/day but not given within 4 hours of vitamins or other medicine, followed by second-line **rifampicin** 2–4 mg/kg twice daily then third-line **ursodeoxycholic acid** 5–7.5 mg/kg 2–3 times a day (very expensive).
- **Fat soluble vitamin deficiencies (A, D, E and K)**. These are frequent unless patients receive prophylactic treatment. Clinical features of rickets such as splayed epiphyses, especially swollen wrists, rickety rosary and craniotabes should be sought regularly. Metabolic bone disease should, ***if possible, be screened for by measurements of serum phosphate and calcium*** and regular wrist *X* rays. Prothrombin time or INR should be measured to ensure adequate vitamin K repletion.

Vitamin A replacement is 5 000–10 000 units per day or 100 000 units by deep IM injection every 2–4 months.
Vitamin D deficiency may be refractory to oral calciferol (vitamin D_2) tablets or cholecalciferol (vitamin D_3) but up to 10 000 units (250 micrograms) per day of either may help. More water-soluble preparations such as 1α-calcidol 50–100 nanograms/kg once daily are more effective but expensive. They may also cause hypercalcaemia.
Vitamin E deficiency is associated with hypotonia, peripheral neuropathy, developmental delay, and haemolysis and is the most frequently encountered vitamin deficiency in liver disease. Significant clinical complications are rarely recognised but it is unknown if a mild degree of developmental delay may result. The dose of vitamin E is at all ages 150–200 mg/kg once daily.
Vitamin K replacement for infants is 1 mg/day; for children 5–10 mg/day.
Nutritional management applies to all three categories of CLD and is discussed below.

Portal hypertension (PHT): diagnosis and management

The complications of portal hypertension can be divided into:

- those related to the increased pressure e.g. enteropathy, hypersplenism
- those related to the anatomy of any collateral circulation e.g. bleeding varices, haemorrhoids; and
- those related to the effects of substances bypassing the liver by portosystemic shunting, e.g. hepatic encephalopathy, hepatopulmonary syndrome, portopulmonary syndrome, hepatorenal syndrome. In this group complications may increase as shunting increases and portal pressure falls.

The aetiology of PHT has conventionally been divided into the following:

- Prehepatic causes, including portal vein thrombosis and other congenital and acquired portal vein anomalies, including arterioportal fistulae
- Hepatic causes, including all causes of cirrhosis, especially cystic fibrosis and other biliary diseases, congenital hepatic fibrosis, and causes of non-cirrhotic portal hypertension including portal vein sclerosis
- Posthepatic causes including hepatic venous outflow obstruction such as Budd–Chiari syndrome, various causes of veno-occlusive disease and problems of inferior vena caval flow or right heart function, particularly constrictive pericarditis.

Treatment of bleeding varices

Acute Management

1. Do not panic and stay with the child. Unless the CLD is very advanced, or the child is vitamin K deficient, the bleed will probably stop spontaneously although the child may be shocked by that time.
2. Gain intravenous access and obtain cross-matched blood.
3. Give intravenous vitamin K slowly over 5 minutes (1 mg under 1 year; 3 mg 1–4 years; 5 mg 5–12 years; 10 mg >12 years). Repeat according to results of clotting studies.
4. Start antacids (see below).
5. ***Arrange skilled endoscopy with sclerotherapy or banding (if available).***

Prevention

- Propranolol. This appears beneficial in preventing variceal bleeding, particularly when given early in the course of PHT. Give 500 micrograms/kg orally twice daily (adjust according to the heart rate; aim to reduce rate by up to 25%).
- Antacids. If there is a tendency to diarrhoea, use aluminium hydroxide (aged 6–12 years 5 ml 3–4 times/day between meals and >12 years 10 ml 3–4 times/day). If there is a tendency to constipation, use magnesium carbonate in same dosage.
- ✔ **Avoid aspirin, ibuprofen and other gastric irritants**. H_2 receptor antagonists are of no proven value but often used (ranitidine 2–4 mg/kg twice daily).

Minimum standards requirements: bleeding oesophageal varices

- Vitamin K
- Blood transfusion and blood-clotting factors
- Propranolol
- Antacids (aluminium hydroxide or magnesium carbonate)
- Ranitidine

Hepatocellular liver disease: diagnosis and management

Chronic viral hepatitis B, C and D

Millions of children are infected with hepatitis B virus (HBV) and many die from its complications, particularly decompensated cirrhosis and hepatocellular carcinoma (usually in adult life). Population prevalence may exceed 10% making HBV a major international public health problem. Spread may occur vertically at the time of birth or shortly after, but more frequently is transmitted horizontally. This is especially so in poor communities. In this context it is probably blood-borne or occasionally from other body fluids, including by sexual means. ✔ **Unlike HIV infection, surface contact with very small amounts of infected blood can result in infection** (for example, sharing toothbrushes). A neonate exposed to HBV for the first time has more than 90% chance of becoming chronically infected while a child has 25% and an adult 10%.

Factors associated with the development of cirrhosis and hepatocellular carcinoma include e Ag+, a high level of HBV DNA, and male sex. Once infected, children have about 15% chance per annum of reducing a high-risk state to a low-risk state as defined by serum markers.

Vaccines based on the antigenicity of the S Ag are highly efficacious in generating antibody response and providing protection. Protocols which involve three subcutaneous immunisations given at time zero, 1 month and 6 months, give adequate antibody levels in 95% of individuals. In neonates, vaccination with the same dose, or half the dose for economy, at birth, 1 month, 3 months, and at 1 year achieves similar protection. Up to 5% of individuals will not mount an antibody response despite repeated vaccination; but it is not clear if they have no immunity. As implied above, all neonates of HBV positive mothers should receive a course of vaccine, irrespective of the mother's infectivity, as infants of S Ag+, e Ab+ mothers may develop fatal liver failure.

✔ **WHO has recommended universal HBV vaccination**. If such a policy were implemented it is highly likely that HBV would become a rare disease of children in less than 10 years, with corresponding reduction in cirrhosis and hepatocellular carcinoma in four to five decades.

Hepatitis C

Hepatitis C virus (HCV) was responsible for at least 90% of post-transfusion hepatitis in early US studies. Five percent of sexual partners may become infected. Six percent of infants of viraemic mothers may become infected. The risk is related to the level of maternal viraemia with HIV positive mothers having highest HCV viral loads and highest risk of transmission. HCV is also a small but important risk for healthcare workers.

Following exposure, viraemia in HCV occurs in 7 days with antibody-positivity appearing from 21–28 days. Less than 10% adequately remove the virus and the remainder

Table 3.10.4 Drugs and liver injury

Injury	Associated drugs
Acute/subacute hepatocellular toxicity	Paracetamol, iron, aspirin, antituberculous drugs (especially isoniazid, pyrazinamide), anticonvulsants (especially sodium valproate and carbamazepine), chemotherapeutic agents (especially methotrexate), ketoconazole, propionic acid derivatives such as ibuprofen
Cholestasis	Rifampicin, antibiotics (especially penicillins, erythromycin estolate), oestrogens, anabolic steroids
Progressive fibrosis	Azathioprine

progress to chronic liver disease. The rate of progression to cirrhosis is unclear but factors such as liver iron content, alcohol consumption, other viral infections including hepatitis A contribute. Five percent of infected adults develop cirrhosis per year in HCV. The median timescale of developing cirrhosis in HCV is probably of the order of four decades. Hepatocellular carcinoma is a recognised complication of HCV and cirrhosis following the latter by 5–15 years typically.

Wilson's disease
An autosomal recessive disorder due to the accumulation of copper in the liver, brain, eyes, kidney and bone. Prognosis depends on early diagnosis and treatment with a low copper diet and penicillamine.

Drugs and the liver
Drugs are a major cause of liver dysfunction. Over 600 drug hepatopathies have been documented. Common examples are given in Table 3.10.4. Drug clearance may be reduced in liver disease and liver disease increases the risk of drug injury to the liver.

HIV and the liver (see Chapter 4.18)
HIV is known to be associated with worsening of hepatitis due to other conventional causes and cholangiopathy, probably related to ascending infection with low-grade organisms such as *Cryptosporidium* or cytomegalovirus (CMV) infection. Hepatitis due to a conventional cause especially CMV may be particularly severe or progressive when associated with a low CD4 count.

Metabolic liver diseases
These are rare and difficult, if not impossible, to treat without liver transplantation or expensive diets.

The management of nutrition in CLD

Malnutrition is a serious consequence of CLD. Thin limbs and a prominent abdomen are frequently seen and malnutrition will be evident in anthropometric measurements. Triceps skin-fold thickness tends to become reduced earlier in the course of progressive disease followed by a reduction in MUAC. Stunting tends to occur later, unless severe rickets is present. Weight is affected by fluid balance abnormalities and organomegaly, and is therefore an insensitive indicator of nutritional state. It is lean body mass, probably skeletal muscle in particular, that is prone to depletion as a result of progressive liver disease.

Many nutritional factors have been considered important in chronic liver disease. Anorexia is attributed to organomegaly or pressure effects of ascites, but may be equally due to a congested gastric mucosa or reduced gastrointestinal motility of portal hypertension or central effects of unidentified toxins. Malabsorption of long chain fats, including those with polyunsaturated fatty acids (PUFAs), is dependent on intra-luminal bile acid concentration. Cholestasis, even incomplete, may result in intraluminal bile acid concentration falling below that required for micelles to be formed. The resulting steatorrhoea creates faecal energy loss but also risks essential fatty acid deficiency, with possible neurodevelopmental consequences particularly in early life. Protein malabsorption may also result from functional pancreatic insufficiency with failure of protease activation by bile acids if cholestasis is profound. Malabsorption may also result from bacterial overgrowth or other unspecified effects of portal hypertension, for example, congestion of the gut may cause impaired active or passive absorption.

Thus, malnourished children with liver disease have high energy expenditure for their size. Target energy intake should be estimated from what the child's current weight for age should be. ✔

Recent work has shown that breast milk contains more PUFA than current formula milks. PUFAs are essential for normal cell membranes and for myelination, particularly in infancy. PUFAs are long chain fats and dependent on intraluminal bile acids for absorption. Breastfeeding is therefore important in children with CLD.

Treatment with intensive enteral feeding and high-dose enteral or parenteral vitamins can improve the quality of life. In the absence of specialist feeds a modular feed may be made using protein powder, carbohydrate polymer, MCT oil and long-chain fat oil preferably with essential fatty acids from walnut oil to provide 4% of fat. Up to 4 g/kg/day protein, 100–140 kcal/kg/day of energy, of which two-thirds is from carbohydrate and two-thirds of lipid as MCT is an ideal target range.

Commercial liver formulas are extremely expensive and their effect on the outcome of the liver disease is unproven. In the absence of the supplements described above, proprietary baby formula can be enriched with locally available oils and starches to give 140 kcal/kg/day with one-third of the supplemental energy as lipid and two-thirds as starch. Remember if commercial formula is given at increased concentration to increase protein intake, the electrolyte intake will increase proportionately with a risk of sodium overload and fluid retention.

3.11 Endocrinology

Jerry Wales

Minimum standards requirements: diabetic ketoacidosis

- **ABC** (see Chapter 1.19)
- Oxygen
- IV N saline and 4.5% albumin
- Insulin
- Potassium
- Mannitol
- ECG monitoring

Endocrine emergencies

Diabetic ketoacidosis

Introduction

Diabetic ketoacidosis (DKA) is the commonest endocrine emergency, usually occurring in previously diagnosed diabetes but should be suspected in any child with:

- dehydration
- abdominal pain
- ketone smell on the breath
- acidosis
- acidotic breathing
- unexplained coma.

In DKA a child may die from hypokalaemia or cerebral oedema. Cerebral oedema is unpredictable, occurs more frequently in younger children and new diabetics and has a mortality of around 80%. These guidelines are intended for the management of children who are more than 5% dehydrated and/or vomiting and/or drowsy and/or clinically acidotic. Children who are 5% dehydrated or less and not clinically unwell usually tolerate oral rehydration and subcutaneous insulin.

Emergency management of children >5% dehydrated and clinically unwell

- General resuscitation: A, B, C.
  - **A**irway: ensure that airway is patent and if child is comatose, insert an airway. If comatose or recurrent vomiting, insert nasogastric tube, aspirate and leave on open drainage.
  - **B**reathing: give 100% oxygen. Bag and mask ventilation if apnoeic.
  - **C**irculation: insert IV cannula and take blood samples (see below). If shocked (tachycardia with poor capillary filling or hypotension) give 20 ml/kg 4.5% albumin solution or 0.9% saline as quickly as possible, and repeat as necessary.
- Confirm the diagnosis:
  - History: polydipsia, polyuria
  - Clinical: acidotic respiration; dehydration; drowsiness; abdominal pain/vomiting
  - Biochemical: high blood glucose on finger-prick test; ketones or glucose in urine.
- Investigations:
  - **Weigh the child**. If not possible because of the clinical condition, use most recent clinic weight as a guideline, or an estimated weight from centile charts
  - Blood glucose
  - Urea and electrolytes (U and E) (if bicarbonate available ***arterial blood gas/ASTRUP may be omitted***)
  - PCV and full blood count
  - Blood culture
  - Urine microscopy, culture and sensitivity (see Chapter 3.7)
  - Set up cardiac monitor to observe T waves (hypokalaemia causes flat T waves and may cause cardiac dysrhythmias; hyperkalaemia causes peaked T waves)
  - Plus other investigations if indicated, for example chest *X* ray, CSF, throat swab, etc. (DKA may be precipitated by sepsis, and **fever is not part of DKA**.) ✔

Assess and record in the notes, so that comparisons can be made by others later:

- Degree of dehydration
  - 3% dehydration is only just clinically detectable.
  - 3–5 %: dry mucous membranes, reduced skin turgor.
  - 5–8 %: as above with sunken eyes, poor capillary return.
  - ≥ 8 %: with shock – severely ill with poor perfusion, thready rapid pulse, reduced blood pressure.
- Conscious level
  - Assess AVPU (**A**lert; responds to **V**oice; responds to **P**ain; **U**nresponsive).

- Institute hourly neurological observations. If less than **A**lert on admission, or there is any subsequent deterioration, record Glasgow Coma Score (see Chapter 3.36) ***and transfer to ICU***. Consider instituting cerebral oedema management.

- Full examination

Look particularly for evidence of:

- **Cerebral oedema: irritability, slow pulse, high blood pressure** and papilloedema. Examine fundi, however; papilloedema is a late sign
- Infection: look for a focus. **DKA can cause a leucocytosis but not a fever**
- Ileus.

- Observations to be carried out (ensure full instructions are given to the nursing staff).
  - Strict fluid balance and urine testing of every sample.
  - Hourly capillary blood glucose measurements.
  - Twice daily weights.
  - Initially hourly or more frequent neurological observations.
  - **Report immediately to medical staff (even at night) symptoms of headache or any change in either conscious level or behaviour.**
  - Reporting any changes in the ECG trace, especially T wave changes.

Management

Fluid volume

By this stage, circulating volume should have been restored. If not, give a further 10 ml/kg 4.5% albumin or 0.9% saline over 30 minutes. Otherwise, once circulating blood volume has been restored, calculate fluid requirements as follows:

$$\text{Requirement} = \text{Maintenance} + \text{Deficit}$$
$$\text{Deficit (mL)} = \text{Percentage dehydration} \times \text{body weight (kg)} \times 10$$

For example, if 10% dehydrated weighing 10 kg $(10 \times 10 \times 10) = 1\,000$ ml deficit. Avoid overzealous fluid replacement, which may be a risk factor for cerebral oedema. ***Calculate deficit as if the patient is no more than 8% dehydrated.*** **Ignore volume of albumin that may have been given to resuscitate**.

Table 3.11.1 Maintenance of fluids

0–2 years	100 ml/kg/24 hours
3–5 years	90 ml/kg/24 hours
6–9 years	75 ml/kg/24 hours
10–14 years	50 ml/kg/24 hours
Adult	25–30 ml/kg/24 hours

Add maintenance and deficit and give total volume evenly over the next 24 hours. Document fluid balance carefully.

Type of fluid

Initially use 0.9 % saline. Once blood glucose has fallen to 12 mmol/litre change fluid to 0.45% saline/5% glucose. If plasma sodium concentration at this time is >155 mmol/litre, consider changing back to 0.9% saline/5% glucose.

- Glucose >12 mmol/litre: give 0.9% saline
- Glucose <12 mmol/litre: give 0.45% saline + 5% glucose
- Sodium 135–155 mmol/litre correct over 24 hours
- Sodium 155 mmol/litre correct over 48 hours using no lower than 0.45% saline

Expect the sodium to rise initially as glucose falls and water is removed from circulation.

- **Cerebral oedema may be related to a plasma sodium concentration which falls or does not show expected rise as glucose levels fall. Therefore, if plasma sodium is falling, change from a 0.45% to a 0.9% saline solution.**

Bicarbonate

- **Bicarbonate is rarely, if ever, necessary**. Continuing acidosis usually indicates insufficient resuscitation. Bicarbonate should only be considered in children who are profoundly acidotic (pH < 7.0) and shocked with circulatory failure. Its only purpose is to improve cardiac contractility in severe shock. The maximum volume of 8.4 % sodium bicarbonate for **half-correction** of acidosis is calculated according to the following formula, and given over 60 minutes:

$$\text{Volume (ml 8.4\% sodium bicarbonate)} = \frac{\frac{1}{3} \times \text{weight (kg)} \times \text{base deficit (mmol/l)}}{2}$$

If no blood gas is available do not give bicarbonate unless in profound shock.

Potassium

- Commence potassium immediately unless anuria is suspected or there are peaked T waves on the ECG or the serum potassium is above 7.0 mmol/litre.
- Potassium is mainly an intracellular ion, and there is always massive depletion of total body potassium although initial plasma levels may be low, normal or even high. Levels in blood will fall once insulin is commenced.
- Add 20 mmol KCl to every 500 ml unit of IV fluid given for the first 24 hours and then reduce to 10 mmol KCl to every 500 ml unit of IV fluid.
- Check U and E 2 hours after resuscitation commenced and then at least 4 hourly, and alter potassium replacements accordingly.
- Use cardiac monitor and observe frequently for T wave changes.

Insulin

Once fluids are running, calculation of insulin infusion rate may be made at leisure, since blood glucose will already be falling. Continuous low-dose intravenous infusion is the preferred method. There is no need for an initial bolus.

Make up a solution of 1 unit per ml of human soluble insulin (for example Actrapid) by adding 50 units (0.5 ml) insulin to 50 ml 0.9% saline in a syringe pump. Attach this using a Y-connector to the IV fluids already running. Do not add insulin directly to the fluid bags. Solution should then run at 0.1 unit/kg/hour (0.1 ml/kg/hour).

- **If rate of blood glucose fall exceeds 5 mmol/litre/hour, reduce insulin infusion rate to 0.05 unit/kg/hour.**
- Once blood glucose is down to 12 mmol/litre, and a glucose-containing fluid has been started, consider reducing insulin infusion rate.
- Do not stop insulin infusion while dextrose is being infused, as insulin is required to switch off ketone production.
- If blood glucose falls below 7 mmol/litre, consider adding extra glucose to infusion.
- If blood glucose rises out of control re-evaluate (is there sepsis or other condition?), and consider starting the whole protocol again.

If a syringe pump is not available, give subcutaneous boluses of Actrapid 6 hourly at 0.6 unit/kg/dose (that is, 0.1 unit/kg/hour). Give half the dose if the blood sugar is falling too fast.

✔ **Remember you must have a glucose drip ready to avoid hypoglycaemia.**

Continuing management

Output

- Urinary catheterisation should be avoided but may be useful in a critically ill child with impaired consciousness. With or without catheterisation, documentation of fluid balance, if necessary by weighing nappies, is of paramount importance.
- Measure accurately and test all urine samples for glucose and ketones.
- Record all fluid input (even oral fluids).
- If a massive diuresis continues, fluid input may need to be increased.
- If large volumes of gastric aspirate continue, replace with 0.45% saline plus 10 mmol/litre KCl.

Laboratory results

- Check biochemistry, ***blood pH***, and laboratory blood glucose 2 hours after the start of resuscitation, and then at least 4 hourly.
- Review fluid composition and rate according to each set of electrolyte results. If acidosis is not correcting, resuscitation may have been inadequate, therefore consider giving more albumin. If blood glucose falls by more than 5 mmol/hour despite reducing rate of insulin infusion, slow down rate of intravenous fluid replacement, so that rehydration takes place over 48 hours rather than 24 hours.

Insulin management

Continue with intravenous fluids until child is drinking well and able to tolerate food. Do not expect ketones to have disappeared completely before changing to subcutaneous insulin. ✔ **Discontinue insulin infusion 30 minutes after first subcutaneous injection to avoid rebound hyperglycaemia.**

Cerebral oedema in DKA

Signs and symptoms of cerebral oedema are:

- headache
- irritability
- seizures
- increasing blood pressure, slowing pulse
- confusion
- reduced conscious level
- small pupils
- possibly respiratory impairment

Management

- Exclude hypoglycaemia.
- Give mannitol 500 mg/kg immediately (2.5 ml/kg mannitol 20 % over 15 minutes). Give as soon as cerebral oedema is suspected.
- **Restrict intravenous fluids to two-thirds maintenance and replace deficit over 72 hours rather than 24 hours**. ✔
- ***Arrange for child to be intubated and, keep $Paco_2$ 3.5–5.0 KpA.*** **Keep $Na^+ > 135$ mmol/litre. Keep head in midline and 30° elevated. Avoid fever > 38.0°C.** ✔
- Repeated doses of mannitol (above dose every 4–6 hours) should be used to control intracranial pressure.

Care of the newly diagnosed diabetic child

After treatment of DKA or in the newly presenting, well, diabetic child the process of education and treatment should commence. It is not feasible to stabilise a child's control or to teach all aspects of diabetic care while an in-patient, so ideally if resources permit, this should take place at home, although some advocate a prolonged initial admission for this process.

Ensure that the parents and child (if of appropriate ability) understand or can perform the following:

- Insulin administration
- Urine testing
- Blood testing
- Diet
- Other general issues.

Insulin

- Draw up specified dose of insulin correctly. As a rough guide a new patient will need approximately 0.5 unit/kg/day split into two-thirds in the morning and one-third in the evening. If < 10 years use medium acting human insulin alone. A 30/70 mixed insulin is usually used initially if >10 years. (30% short acting plus 70% medium acting – pre-mixed or free mixed)
- Further modification of dose will take place as an outpatient and may involve more than two injections per day of fast-acting insulin before meals with once or twice daily "background" long-acting insulin.
- It is almost never possible to achieve adequate control with a single daily dose of insulin except in very small children. However once-daily medium-acting or premixed insulin should be seen as a minimum fallback position if availability of insulin is a problem. Likewise, although it is common practice to use human or genetically modified insulins, pork or beef insulin may be substituted if necessary.

Urine testing

- Test urine for sugar using stix. Clinitest tablets for reducing substances are too cumbersome for routine use but may substitute if needed. Suggest stix testing about twice daily at home in the child too young or unwilling to use blood testing strips or if these are not available. Emphasise the value of testing the first morning urine to estimate overnight control.
- Test urine for ketones using Ketostix or tablets. This only needs to be routinely used if the urine has □3% sugar and in times of intercurrent illness when the persistence of ketonuria should prompt the seeking of medical attention for incipient DKA.
- The importance of accurate recording of the results in a control book, if possible, should be emphasised to aid in decision-making at follow up.

Blood testing

If resources allow, all parents should be able to test the blood sugar in an emergency and many older children prefer this method for routine monitoring of control. The parent (and child if appropriate) should be taught:

- To use a lancet (or automatic finger-pricking device) to draw blood from the side (**not** the pulp) of the finger
- How to ensure an adequate sample is placed on the strip
- How to read the strip visually (A meter may used if resources allow.)
- If this method of monitoring control is chosen then the importance of providing test results at staggered times through the day (ideally one or two tests per day), should be explained. Accurate recording of the values in a control book should be emphasised
- The instantaneous nature of the result obtained and the detection of hypoglycaemia should be contrasted to the urine testing
- Young children do not like blood tests and this should be weighed against the sometimes overenthusiastic monitoring of control by parents. In many young diabetics urine testing is perfectly adequate for routine purposes.

Diet

- The parents and child should ***ideally meet the specialist diabetic dietician*** and discuss the concept of carbohydrate balance and how the diet is spread through the day. They should understand the importance of fairly close adherence to the advised diet and that this may need to be revised from time to time as the child grows and the pattern of activity changes. They should understand the influence of food intake on blood sugar. Diabetic carbohydrate 10 grams "portions" are no longer recommended by most clinicians as they are difficult to comprehend or adhere to for even the most intelligent. However the diet must be adequate for growth and nutrition and contain around 50% of energy as complex carbohydrate. It is not advisable to allow "free" fatty foods as they may accentuate later macrovascular complications.
- Sweet, unrefined sugars should ideally only be taken pre-exercise or as occasional treats, but, ideally the insulin dose should be varied to take this into account.

General

- The parents and child (if appropriate) should understand how exercise, diet and insulin interact to influence blood sugar.
- The symptoms of hypoglycaemia should be explained. It is important that both parents understand the possible signs of an attack and understand what may be done to terminate the "hypo". The parents should know how to use rapid acting sweet sugary gel, drinks or tablets for the early stages of the attack. A 1 mg glucagon pack should be given to each family prior to discharge and the parents should be shown how to prepare and give the prepacked injection in an emergency to terminate a severe hypo attack with unconsciousness or fits.
- The family should have the address of any local support groups for people and families with diabetes, if they exist. If possible give the parents and child a folder containing relevant booklets on diabetes.
- The family should ideally have access to medical advice and treatment at any time of day or night if they are worried about their child's immediate health or can be seen in the next out-patient clinic for less urgent problems.

Ideal equipment checklist at discharge

- Hypostop gel – 1 box of plastic tubes.
- Disposable syringes and needles, ideally 0.3 ml low volume syringes with as small a needle as available (down to 31 gauge are available).
- Insulin.
- Glucagen Novo 1 mg pack.
- BM stix, finger-pricking device + lancets **or** urine stix
- Control book
- Ketostix
- Sharps disposal bin

Arrange to see at next diabetic clinic.

Out-patient care

The patient should be reviewed at regular intervals as frequently as resources allow. Ideally, at least once a year the child should have the following reviewed:

- Knowledge of diabetes and emergency management
- Growth
- Blood pressure
- State of injection sites
- Foot examination and discussion of foot care
- Fundoscopy: (at diagnosis (for cataracts), after 5 year's diabetes or in teenagers)
- Microalbumin/creatinine ratio in first morning urine sample for detection of renal complications (after 5 year's diabetes or in teenagers)
- ***Glycosylated haemoglobin*** for monitoring long-term control
- ***Antithyroid antibodies*** (some would argue for ***antigliadin antibodies***) at diagnosis, then every four years and, if antithyroid positive yearly thyroid function; if coeliac disease found then gluten-free diet ***after jejunal biopsy***.

Transfer to adult services should be in a planned manner, ideally in a joint handover clinic.

Adrenal crisis

Minimum standards requirements: adrenal crisis

- **ABC** (see Chapter 1.19)
- Hydrocortisone and fludrocortisone
- IV 0.9% saline
- IV glucose 10%

Diagnosis

- Most likely to be encountered in a neonate with congenital adrenal hyperplasia (CAH) or hypopituitarism (look for virilisation in the female with CAH and micropenis and cryptorchidism in the male with hypopituitarism).
- May occur in older children with adrenal destruction secondary to autoimmune process or tuberculosis.
- Suspected in severely ill child with:
  - acidosis
  - hyponatraemia
  - hypotension
  - hyperkalaemia
  - hypoglycaemia

Children receiving long-term steroid therapy

Replacement steroids given as hydrocortisone up to 15 mg/m^2/day replicate natural secretion and are free of side effects if adequately monitored. Therapeutic doses of steroids for asthma, rheumatoid, etc. will produce adrenal suppression in a manner related to dose and duration of treatment. Short five-day courses of prednisolone therapy will produce measurable adrenal suppression that almost never requires action. Longer courses up to one month should be tapered off over a two-week period to allow recovery of the pituitary adrenal axis. More prolonged treatment with high dose steroids may produce profound hypoadrenalism for months after cessation of treatment. In this case taper the steroid dose to the equivalent of 5 mg/m^2/day of prednisolone. Then convert this to an equivalent dose of hydrocortisone given in the morning (1 mg prednisolone = approx 3 mg hydrocortisone). Then reduce the hydrocortisone by 2.5 mg/week until on approximately 6 mg/m^2/day when it is probably safe to stop treatment after two weeks. ***If possible check the 9 a.m. pre-dose cortisol level and stop treatment if this exceeds 150 nmol/litre at any time.*** Severe stress, infection or injury will require increased steroid cover **during the next six months**.

Children on physiological replacement treatment or prolonged pharmacological doses of steroids should ideally carry some warning identification for medical staff advising against the abrupt cessation of steroids and the emergency stress dose of oral (usually three times replacement dose) or parenteral treatment for operative cover or at times of illness associated with vomiting (hydrocortisone 12.5 mg for infants, 25 for children, 50 mg for older children and 100 mg for adults given as an immediate IM dose or 4–6 hourly IV).

Management

- Treat airway, breathing, shock and hypoglycaemia (see above and Chapters 1.19 and 3.6).
- Continue 0.9 % saline to correct deficit and for maintenance (see DKA above).
- Give hydrocortisone IV 4 hourly as follows: dose: 12.5 mg for neonate and infant; 25 mg for 1–5 years, 50 mg for 6–12 years and 100 mg for 13–18 years.
- If diagnosis established continue maintenance hydrocortisone 15 mg/m^2 per day in three divided doses and, if salt loss demonstrated, fludrocortisone 150–250 micrograms/m^2/day in one dose with sodium chloride 1 g/10 kg/day (60 mg = 1 mmol).

Hypoglycaemia

See Chapter 3.12, and Chapter 3.48 for section on neonatal hypoglycaemia.

Neonatal thyrotoxicosis

Mothers with active thyrotoxicosis or mothers who have become **hypothyroid** as a consequence of treatment of thyrotoxicosis may still pass thyroid stimulating antibodies to the fetus during the last trimester.

The neonate (or fetus) will show:

- Hydrops in severe cases
- Tachycardia with heart failure – **this may occur at up to 1 week postdelivery** especially if the mother is on anti-thyroid drugs
- Thinness/light-for-dates
- Diarrhoea
- Hyperkinesis
- Possibly craniosynostosis

Management

- If hyperthyroidism is detected antenatally, treat mother with low-dose carbimazole 5–15 mg/day (use the lowest dose possible for control).
- Treat infant with:
  - Propranolol 1 mg/kg 3–4 times orally daily
  - Carbimazole 400–600 micrograms/kg/day (single dose)
  - Lugol's iodine (5% iodine + 10% potassium iodide: 130 mg/ml, 1 drop (0.1-0.3 ml) every 8 hours) until thyroid control achieved.
- Stimulating antibodies will clear by 3 months of age and treatment can be stopped.

Outpatient endocrinology

Diabetes mellitus

This is the commonest endocrine problem in northern latitudes and is less frequent in disadvantaged countries except for a few rare ethnic groups. Almost all diabetes in children is type 1 (insulin dependent) diabetes, although type 2 (insulin resistant) occurs in populations with extreme obesity and some rare genetic forms exist. Diagnosis is confirmed by a blood glucose > 11 mmol/ litre. No further tests are required in majority of cases.

Management (see earlier in this chapter)

- **Insulin**. Give subcutaneously at least once per day, preferably twice or more, in a starting dose of 0.5 unit/kg/day. Dose is then titrated against blood glucose to maintain levels in single figures and avoids frequent severe hypoglycaemia (especially in under-five-year children). Teenagers may need up to 2 units/kg during midpuberty.
- **Diet**. Total intake as required for normal growth but divided into three main meals and three snacks containing at least 50%, preferably unrefined, carbohydrate and around 25–30% fat. Balance these two factors against exercise (which will require less insulin/more food) and ill health (which will require more insulin/more drinks) and adjusted regularly as the child grows.
- **Education**. Teach family/child to recognise and treat hypoglycaemia (pallor, sweatiness, hunger, abdominal pain, confusion, irritability, coma, fits) by giving sweet fluids early in the attack or glucagon (see above) if consciousness is clouded. Monitor glucose level during intercurrent ill health along with urinary ketones; ***insulin should never be discontinued***. If ketosis and hyperglycaemia occur 2–4 units of soluble insulin should be given before considering intravenous fluids as described above.

Hypothyroidism

Minimum standards requirements: thyroid disorders

- Carbimazole
- Propranolol
- Iodine supplements
- Thyroxine

Congenital hypothyroidism

Between 1/2000 and 1/10 000 babies are born with a maldescended or absent thyroid gland. There are rarer cases of dyshormonogenesis (dominant and recessive) associated with neonatal goitre and very rare isolated thyroid-stimulating hormone (TSH) deficiency.

Untreated early hypothyroidism results in cretinism.

Many countries screen for this condition in the first month of life (elevated TSH (except in TSH deficiency) and/or low thyroxine or free thyroxine). (Different screening laboratories will produce different assay results.) In general TSH in high double figures (mU/litre) is unequivocally raised and will be confirmed by a total thyroxine less than 50 nmol/litre or a free thyroxine in single figures (pmol/litre). In disadvantaged countries, *X* ray of knee or wrist to detect delayed bone age in infants and young children is helpful for diagnosis where TSH, thyroxine assays are unavailable.

An affected child will develop, in this order:

1. Jaundice
2. Constipation
3. Hoarse cry
4. Umbilical hernia
5. Coarse features
6. Mental retardation
7. Poor growth

Management

Thyroxine 12–15 micrograms/kg once daily titrated to maintain TSH in normal range with normal growth and development. Adult dose is around 2–3 micrograms/kg.

Iodine deficiency

Occurs usually in inland mountainous areas. Features vary in different ethnic groups with deafness, mutism, mental impairment and poor growth being common and goitre universal. It may be prevented by adding potassium iodide to cooking salt (10 mg/kg) or as supplemented sweets and bread. Iodide as an oily suspension can be given intramuscularly every three years.

Acquired hypothyroidism

This is usually part of an autoimmune process (which may be familial) and may be associated with diabetes mellitus. It is much commoner in older girls who will usually have:

- Goitre
- Lethargy
- Poor growth rate with excess weight gain
- Pallor
- Constipation
- Hair loss/dry skin

Diagnosis is confirmed by raised blood TSH and, if possible, demonstration of antithyroid peroxisomal antibodies.

Management

Thyroxine 2–3 micrograms/kg to suppress TSH to normal range and allow normal growth and pubertal development.

Thyrotoxicosis

Much commoner in older girls often with a family history of thyroid disease. Suspected if there is:

- Fine tremor
- Weight loss
- Psychiatric disturbance
- Exophthalmos (rare in children)
- Tachycardia with a wide pulse pressure
- Loose stools
- Goitre with bruit

Diagnosis is confirmed by suppressed TSH (undetectable) with raised thyroxine level.

Treatment is low dose carbimazole 400–600 micrograms/kg/day (single dose) continued for at least two years when withdrawal should be attempted. If relapse occurs options include further medical therapy, ***surgery by an experienced thyroid surgeon or radioiodine in a specialised centre.***

Thyroid mass

Smooth goitre

Isolated, smooth goitre with or without a bruit may occur in:

- Iodine deficiency

- Acute and subacute thyroiditis (viral, bacterial, lymphocytic, other)
- Goitrogen ingestion (for example cabbage, kale, or other brassicas)
- Familial dyshormonogenesis
- Idiopathic pubertal goitre
- Thyrotoxicosis (Graves disease, thyroiditis, thyroid hormone resistance)
- Hashimoto's thyroiditis

If thyroid function is normal, no treatment is necessary, otherwise treat as described above. If iodine deficiency but thyroid investigations are not possible, treat as above.

Nodular goitre
Nodular goitre may occur in:
- Hashimoto's thyroiditis
- Adenoma (hot, cold, euthyroid)
- Lymphoma
- Non-thyroidal masses (lymph nodes, branchial cleft cyst, thyroglossal cyst)
- Isolated simple cyst
- Carcinoma
- Histiocytosis

Nodules require investigation by fine-needle aspiration and histology.

Ambiguous genitalia

Uncertainty regarding a child's sex is a distressing emergency for the family.
- Most children are well unless associated with CAH and salt loss (see above) or other major congenital abnormalities.
- ✔ **Avoid urge to decide appropriate sex of rearing of the child until diagnostic tests are available.** Support parents during this difficult time.
- Intersex may be result of overvirilisation of the female (commonest situation, usually secondary to CAH of 21 hydroxylase deficiency variety), undervirilisation of the male or, rarely, true hermaphroditism. ***Minimum investigations are chromosome analysis and plasma for 17-hydroxyprogesterone (elevated in the commonest form of CAH).***
- ✔ **If a baby with indeterminate genitalia becomes unwell with hypotension, hyponatraemia and hyperkalaemia assume an adrenal crisis and treat as described above.**
- Further investigation requires highly specialised tests, i.e. blood, radiology and ultrasound, fibroblasts, laparoscopy or laparotomy. Treatment of non-CAH intersex is also complex but can often be deferred to allow appropriate transfer of care to specialist centre.

Congenital adrenal hyperplasia (CAH)

Congenital adrenal hyperplasia (CAH) is an autosomal recessive condition, hence commoner in consanguineous liaisons. Many forms exist as several enzymes involved in synthesis of cortisol and aldosterone may be deficient, partial cases also occur within each subtype.

Salt-losing 21 hydroxylase deficiency is by far the commonest type. Most forms result in overvirilisation of the female (although undervirilisation of the male can also occur in defects near the start of the biosynthetic pathway). Salt loss occurs in several forms (see above) although the second commonest deficiency (11 β-hydroxylase) causes salt retention and hypertension.

Females usually present as intersex (see above) and males with salt loss which occurs usually after the first week of life (see above for acute and long-term management). In non-salt losing forms there will be incomplete early puberty in males (see below).

Addison's disease and Cushing's syndrome

Hypoadrenalism
Hypoadrenalism may present as an emergency (see above) or be suspected if there is:
- Unexplained lethargy
- Failure to thrive
- Pigmentation of scars and skin
- Vitiligo or other signs of autoimmune disease
- Strong family history of hypoadrenalism or unexplained sudden death
- Hyponatraemia and hyperkalaemia
- Syndrome of candidiasis and hypoparathyroidism predating the hypoadrenalism (HAM).

If confirmed by a low 9 a.m. cortisol (<150 nmol/litre) treat as outlined on page above.

Cushing's syndrome
Cushing's syndrome is usually the result of iatrogenic corticosteroid administration (>15 mg/m^2/day hydrocortisone or equivalent – see above). Oversecretion of adrenal steroids is rare. Signs of corticosteroid excess include:
- Poor (zero) growth rate
- Red cheeks
- Striae
- Glucose intolerance
- Excess weight gain (central)
- Muscle weakness
- Hypertension.

Adrenal carcinoma or adenoma may produce Cushing's syndrome. There is often accompanying virilisation and an abdominal mass. The child is usually young in contrast to the older child with Cushing's disease secondary to an ACTH secreting pituitary adenoma.

Diagnosis is supported by a ***detectable midnight cortisol level (>50 nmol/litre) or raised urinary free cortisol excretion. The 9 a.m. cortisol fails to suppress to undetectable levels in response to dexamethasone 0.3 mg/m^2 single dose given previous night. Treatment usually requires specialist surgery.***

Hypogonadism and delayed puberty

- Hypogonadism may be secondary to central gonadotrophin deficiency (hypogonadotrophism) or peripheral gonadal failure (hypergonadotrophism).

- Suspect in a male neonate with undescended testes and micropenis (<2.5 cm length shaft). Hypopituitarism may also be present.
- If undetected, failure of, or incomplete, pubertal development will occur.

Table 3.11.2 Causes of delayed puberty

LH/FSH low testosterone/oestrogen low
• Chronic ill health
• Constitutional/familial
• Starvation, low body mass index
• Genetic (i.e. Kallman's and Prader–Willi syndromes)
• Prolactinoma (rare)
• Hypopituitarism, hypothyroidism
• Thalassaemia
• Trauma/infection
• Gonadal dysgenesis or Turner's syndrome (XO)
• Klinefelter's syndrome (XXY)
• Testicular feminisation (XY, female)
• Autoimmune ovarian damage
• Galactosaemia
• Steroid synthesis defects

- Delayed puberty is often familial but may be induced by emotional or nutritional deprivation.
- There is delayed maturation of gonadotrophin secretion.
- Treat if delay is severe enough to cause psychological damage.
- Give a brief course of testosterone esters (100 mg by deep IM injection (Sustanon) at 1 month intervals three times in boys) or oral oestrogen (5–10 micrograms/day for 3 months in girls) which allows puberty to be induced and also reduces risk of later osteoporosis.
- If the hypogonadism is likely to be permanent continue and increase testosterone to 250 mg/month or oestrogen to 25–50 micrograms/day (the latter as a combined oral contraceptive medication to allow withdrawal bleeding).

Precocious puberty

- In precocious puberty early sexual maturity (<8 years in females, <10 years in males) is usually accompanied by a growth spurt and relative tall stature for age.
- In central gonadotrophin activation there is development of full puberty, i.e. breast and pubic hair with eventual menstruation or testicular enlargement plus pubic hair and penis development.
- In secondary cases there is excess peripheral sex steroid production/ingestion. Puberty will be exaggerated whilst other tissues will be normal, or may regress i.e. large penis and pubic hair plus small, hard testes in androgen excess in CAH; large breasts but no pubic hair in oestrogen-secreting tumour.
- Idiopathic central precocity is commonest in females and may be familial.
- Male gonadotrophin activation may be a sign of a CNS tumour.
- Investigation and treatment are complex and specialised and include suppression of gonadotrophin secretion in central precocious puberty and surgical removal or suppression/blocking of peripheral source of sex steroid production in peripheral causes.

Growth hormone deficiency and short stature

- Growth hormone deficiency (GHD) may be idiopathic, familial and part of hypopituitarism or isolated.
- In the neonatal period isolated GHD or hypopituitarism may cause hypoglycaemia (see Chapter 3.12). After excluding or treating hypoadrenalism (see above) ***growth hormone 200 micrograms once or twice per day may be required to maintain normoglycaemia.***
- Growth hormone is essential for normal growth and GHD should be suspected in child who is:
  - short in relation to peers and in comparison to parents
  - growing slowly
  - relatively heavy for height.

Outside the neonatal period treatment is rarely urgent. ***Growth hormone is administered as a subcutaneous injection. It is expensive and difficult to store.***

Severe short stature may be:

- Secondary to chronic ill health or undernutrition – often thin
- Secondary to chronic emotional trauma – often thin
- Endocrine (hypothyroid; hypopituitary; GHD; Cushing's syndrome) – often relatively heavy
- Syndromic (Turner syndrome, etc.) usually dysmorphic
- Disproportionate with short limbs (bony dysplasias, rickets)
- Metabolic (storage disorders, osteogenesis) with longer limbs than back.

Short stature in the latter three causes is extremely difficult and expensive to treat. ✔

Hypopituitarism

- In the neonate there will be hypoadrenalism with or without GHD leading to hypoglycaemia (see above).
- Suspect in any male neonate with cryptorchidism and micropenis.
- Onset later in life may signal an intracranial lesion such as craniopharyngioma (which may be visible as a calcified mass on plain lateral skull *X* ray). Symptoms outside the neonatal period include:
  - Poor growth/short stature (secondary to GHD)
  - Lethargy
  - Hypotension
  - Hypothermia
  - Hypothyroidism (see above)
  - Hypogonadism (see above)
  - Visual field defects/headache/raised intracranial pressure if secondary to tumour.

Treatment is outlined in sections on individual hormone deficiencies above.

Diabetes insipidus

Isolated diabetes insipidus is rare but can occur as part of hypopituitarism or secondary to infiltration of posterior pituitary by tumour or destruction by infection. Suspect in any case of:

- Dehydration with dilute (colourless) urine
- Polyuria and polydipsia not due to diabetes mellitus
- Secondary daytime wetting without obvious cause
- Familial history.

Diagnosis is confirmed by simultaneous presence of hyperosmolar serum (>290 mosm/litre) and dilute urine (<300 mosm/litre or specific gravity <1005).

Treat with replacement of antidiuretic hormone as a long-acting analogue, DDAVP which can be given intramuscularly, by nasal spray, or orally. The dose is titrated to keep urine specific gravity in 1005–1010 range. However, at present the cost is so high that it is unlikely to be available in poor countries.

3.12 Hypoglycaemia

James Leonard and Jerry Wales

Minimum standards requirements

- Oral glucose solutions
- IV 10% and 20% glucose
- Central venous access
- Glucagon

Introduction

Hypoglycaemia is an important cause of morbidity and mortality that needs to be recognised, as the complications are potentially preventable.

Definition

Hypoglycaemia is now widely defined as a blood glucose concentration of less than 2.6 mmol/litre (47 mg/dl) at any age. The measurement should only be made in a laboratory with appropriate quality control. Testing with reagent strips is much less accurate, particularly within the critical range.

Presentation and aetiology

Hypoglycaemia may present at any age from birth into adult life. Symptoms are varied and rarely specific, particularly in babies and infants. In neonates fits and apnoeic attacks may be important clues. In infants and children the most important presentation, because of the risk of complications, are also fits and encephalopathy (Table 3.12.1).

The common causes are listed in Table 3.12.2. In infants and children in advantaged countries ketotic hypoglycaemia, endocrine and metabolic disorders usually predominate. By contrast in the disadvantaged world malnutrition and infections such as malaria are much more common.

Treatment

- As symptoms are non-specific, measure blood glucose if possible.
- When not possible, treat any critically ill child presenting with suspicious symptoms such as fits, encephalopathy or with a condition known to be associated with hypoglycaemia, such as severe malnutrition or malaria.
- Give glucose orally if it can be given safely (0.5 g–1.0 g/kg) If child is conscious and able to eat, give food or sugary fluids.
- Otherwise 2 ml/kg 10% glucose IV over 3 minutes. **Never use stronger glucose solutions**. Continue with 0.1 ml/kg/minute 10 % glucose to maintain blood sugar >5 and <8 mmol/litre.
- If hypoadrenalism/pituitarism is suspected give hydrocortisone as described in Chapter 3.11.
- In diabetics or suspected hyperinsulinaemia if IV access is not possible buy time by giving glucagon IM 100 micrograms/kg (maximum 1 mg as single dose).

Longer term management:

- Appropriate endocrine management
- Avoid periods of fasting, give glucose orally when at risk during intercurrent infections or IV if comatose, vomiting and during anaesthesia.

Diagnosis

If blood sugar <2.6 mmol/litre it is important to establish a cause: take 1 ml in fluoride tube, if possible 1 ml heparinised blood and, if possible, the first urine after the attack to send for metabolic analysis in particular, ketones.

- Is there ketosis? If so, look for signs of hypopituitarism and/or growth hormone deficiency.
- ***If feasible check cortisol growth hormone level and insulin levels in blood taken at time of hypoglycaemia.***
- ***If blood lactate level raised consider organic acidaemia or defect of gluconeogenesis. (Will be high if a seizure has occurred)***
- If ketosis is absent consider hyperinsulinism (large baby) or disorders of fatty acid oxidation.

Prevention

In the neonate, every care should be taken to avoid those factors that will exacerbate hypoglycaemia including delayed feeding and hypothermia (see Chapter 3.48).

Table 3.12.1 Common symptoms and signs of hypoglycaemia

In childhood	*In neonates*
Seizures	Jitteriness/tremors
Anxiety	Lethargy
Sweating	Cyanotic episodes
Palpitations	Seizures
Headache	Apnoeic episodes
Behaviour abnormalities	Feeding difficulties
Confusion	
Visual disturbances	
Dysarthria	
Ataxia	
Pallor	
Hunger	
Nausea	

Table 3.12.2 Some causes of hypoglycaemia

Neonates
Birth asphyxia
Small for gestational age
Very preterm
Sepsis
Starvation
Hypothermia
Infant of diabetic mother
Rhesus disease
Liver disease, endocrine and metabolic disorders – (see below)
Infants and children
Endocrine disorders
- Diabetic on treatment
- Persistent hyperinsulinaemic hypoglycaemia of infancy (formerly nesidioblastosis) and other congenital and inherited hyperinsulinaemic syndromes
- Islet cell tumours
- Hypopituitarism
- Growth hormone deficiency
- Adrenal insufficiency – any cause

Metabolic disorders
- Disorders of glycogen metabolism, gluconeogenesis, fatty acid oxidation, organic acidaemias, etc.

Ketotic hypoglycaemia (“accelerated starvation”)
Liver disease – any severe acute liver disease
Malnutrition
Infections – malaria
Any severe illness
Poisoning
- Alcohol
- Salicylates
- Insulin

Drugs
- Oral hypoglycaemic agents

3.13 Psychiatric disorders

Surya Bhate and Kathy Brookes

Minimum standards requirements

- Antidepressants
- Sodium valproate
- Chlorpromazine, haloperidol and flupenthixol
- Propranolol and clonidine
- Psychotherapy

Introduction

- Between 10% and 20% of children are at risk of developing mental health difficulties. The rates are higher in urban areas and increase in adolescence.
- Prematurity, poor nutritional status, low birth weight, organic brain damage and physical handicap often bring about biological stressors.
- Disadvantaged socio-economic status of families contributes negatively to the mental health of children. Child development suffers where there is persistent marital discord, parental psychiatric ill health and/or history of substance abuse. "Protective" factors include stable care, an attractive and engaging personality, problem-solving abilities and a supportive network of family and friends.
- Psychiatric disorders arising in adolescence are different from those in children and similar to those in adults. The vulnerability of adolescence relates to difficulty in establishing an identity, during which there may be alienation from parents. There is also intense emotional interaction with friends and issues of sexuality.
- Emotional disorders include anxiety states, depression, hysteria and circumscribed phobias. Conduct disorders occur in about the same proportion. About 20% of the teenagers may present with a mixture of disorders.

Acute psychiatric emergencies: suicide and deliberate self-harm

In rich countries there has been a persistent rise in suicide rates in young males, especially amongst those with a history of substance abuse, conflict with the law and personal and mental illness. Suicide is extremely rare in pre-pubescent children but the frequency rises sharply during the teenage period.

Deliberate self-harm is a non-fatal act in which a child/young person deliberately ingests noxious substances in excess of therapeutic doses or causes self-injury.

Assessment and questions to be asked

- Is there a risk of suicide or of repeated attempt?
- Was a suicide note left?
- Was there pre-planning?
- What was the prospect of being found?
- Did he/she know how toxic the substance was?
- Quantity of substance taken?
- Was it impulsive in the context of a conflictual relationship?
- Was it to attract sympathy, for example, following disciplinary crisis or loss of a friend?
- Is there a psychiatric disorder?
- What is the family and developmental history, including educational functioning?
- How well does the child solve problems, cope with difficulties, social/parental support, including supervision?

It is important to clarify to the family (preferably in the presence of the child) that information given by the child is confidential.

High-risk factors

- Risk of repetition is higher in the next four weeks. It also increases with history of past self-harm attempts.
- Male sex.
- Lack of support and easy access to a means of committing suicide, for example a firearm.
- Presence of depressive illness with loss of sleep, appetite, depressed mood, agitation, particularly continued suicidal ideas (hopelessness – inability to enjoy life, "what's the point?").

Treatment

- Treatment of the medical consequences of self-harm is the priority (see Chapters 5.12 and 5.4).
- Assessment of the child/family should be undertaken when the child is free from the after effects of the drug

overdose/self-inflicted injury. Those with low risk of repetition can be offered support during subsequent crises and arrangements made to assist the child/family to develop coping strategies. A psychologist, social worker or trained psychiatric nurse (if available) can assist the family in this way. Children at high risk of death need a major input although in-patient facilities are generally sparse and often unavailable. Depending on resources (or lack of them) the physician needs to improvise and involve social agencies and the family (particularly the extended family if there are immediate parental problems) to provide appropriate supervision, support and treatment.

- Those with a history of substance abuse will need specific counselling.
- Nothing predicts behaviour better than past behaviour. The presence of mental illness merits specific treatment (psychological therapies/medication) and intervention (see below).

Depressive disorders

Sadness, unhappiness and misery are common experiences. When sadness is extreme in intensity and duration, it needs urgent attention. A depressed mood (dysphoria) is accompanied by loss of emotional involvement (withdrawal), feelings of guilt and self-deprecation, and an inability to cope effectively. A "depressive disorder" refers to an observable depressed mood, tearfulness, suicidal thoughts, disturbance of sleep and appetite, and a lack of energy.

When the above symptoms persist for two weeks or more and functioning is impaired, a diagnosis of depressive disorder is made. It is worth noting that around 40% of children with conduct (behaviour disorders) have associated mood disturbances.

How to assess

It is important to include both the child/adolescent and the family. This may be impossible for the teenager who has no family or for the older teenager that refuses to have his/her parents involved.

At the beginning of the assessment it is helpful to clarify the bounds of confidentiality. The parents and the child need to understand that what each of them says will not be freely shared without consent. However it should also be made clear that there are limits to confidentiality in situations in which the law requires reporting, for example, abuse and also in situations where the child's safety is at serious risk for example of suicide.

Assess the degree of dysfunction and distress the symptoms are causing to the child and the family.

Risk factors

- Family, adoption and twin studies have demonstrated the importance of genetic factors.
- The process of puberty can precipitate depression.
- Family conflict, broken family relationships, and a lack of support contribute negatively.
- Stressful life events, such as the death of a loved one, can also precipitate a degree of depression which fulfils the definition of "depressive disorder".

Principles of management

- The diagnosis of a depressive disorder (as opposed to transient sadness) should be made following careful history-taking and information from the family, school and when possible close friends.
- **Medical conditions which can present with depression should be excluded:** in particular vitamin or mineral deficiencies (full blood count), thyroid dysfunction (TSH), tuberculosis (chest *X* ray), and HIV infection. ✔
- If possible, address any stressful factors in the child's environment.
- Cognitive therapy can be effective. It requires specialist training and is usually delivered by psychologists or psychiatrists. Since the depressed child will tend to blame him/herself, there should be an attempt to enable the child to deal with issues without such negative feelings.
- In more severe forms of depressive disorder, antidepressants are effective. ECT (electroconvulsive therapy) is only indicated in severe adolescent depression with psychotic features, if other forms of treatment are not effective and/or the patient is at high risk of death. The effectiveness of antidepressants in children and adolescents is variable. Occasionally they may precipitate hypomania in children/adolescents with bipolar disorder (manic-depressive illness). If the latter happens, sodium valproate represents the first-line maintenance treatment for disadvantaged countries.

If the child/adolescent has a history of hypomania/mania **or** these symptoms are precipitated by use of an antidepressant the diagnosis of a bipolar affective disorder (manic depression) should be made. This is characterised by alternating episodes of hypomania/mania and depression. Some people will only have manic episodes. Mania is characterized by an elevated mood with an increase in physical and mental activity. Associated symptoms include increased energy, decreased need for sleep, increased activity, racing thoughts, pressure of speech, decrease in social inhibitions, and grandiosity. The mood is labile and may be very irritable and at times aggressive.

Mania can present with psychotic features and needs to be differentiated from schizophrenia or a drug induced psychosis.

Mania is treated with a mood stabiliser such as ***lithium carbonate*** or **sodium valproate** (start at 10 mg/kg/day and increase to a maximum of 30 mg/kg/day in two divided doses).

Antidepressant medication

- Tricyclic antidepressants may not be clinically effective and have major side effects including cardiovascular complications. They therefore offer little benefit.

MAOIs should not be considered as a first line treatment due to dietary restrictions.

- ***Selective serotonin reuptake inhibitors (SSRIs) such as fluoxetine are effective but expensive***. They should be prescribed following baseline measurements of blood pressure, heart rate, physical examination for extrapyramidal symptoms etc. The suggested baseline laboratory investigations are: complete blood count, liver function tests, pregnancy tests for females, and ECG.

The common side effects of SSRIs are dizziness, sweating, diarrhoea, headaches, fatigue, restlessness, initial insomnia, and weight loss or gain. Uncommon side effects may be delayed micturition, blurred vision, skin rashes etc. and should be explained to the patient and family. Usually fluoxetine is best tolerated given in the morning after breakfast but in situations in which it causes drowsiness it can be taken in the evening. Changes in symptoms do not occur for at least 3–6 weeks.

Fluoxetine
Test dose: 5 mg daily
Starting dose: 5 mg daily
Target dose range: 5–40 mg daily

✔ **It is vital that the child is informed that his/her symptoms and their effects on behaviour and educational function are due to a mental illness.** The child and their family need to learn how to differentiate and distinguish between the normal range of feelings and those, including sadness, that suggest the onset/presence of the depressive disorder.

Hysterical conversion disorder

This is a subgroup of somatoform disorders. It refers to loss or alteration of physical functioning without organic cause. The child presents with physical symptoms (often referred to the central nervous system), which appear to result in apparent disability in the absence of consistent physical signs or evidence of a physical illness. The most frequent symptoms are pseudo-seizures, loss of sensation and loss of limb function. These are common in postpubertal female adolescents. They usually arise when the adolescent is facing a predicament he/she cannot resolve. The assessment should include full physical examination to assess symptoms that do not correlate with known neurological pathways (for example a gait that is inconsistent and varying, etc.). Once a psychiatric disorder is diagnosed, emphasis should move from medical investigations to amelioration of presenting symptoms and appropriate psychological intervention. The latter should incorporate "face-saving" formulae to allow the young person to come to terms with the absence of physical disease, but the presence of an "illness" which is "real" as far as the young person is concerned.

Drug and alcohol: use and abuse

- Drug use and availability has changed radically in the last decade or two. The substances abused depend on availability and supply. As children get older, the proportion who have ever tried them increases.
- Reliable figures of drug abuse in children are scanty and not validated. However there is increasing acceptance that the rates are increasing, particularly in inner city areas amongst children who are deprived, for example "street children".
- There is a strong association of substance abuse with conduct disorder in the previous 12 months. Conduct disorders include a repetitive and persistent pattern of behaviour in which societal norms or rules are violated, for example fighting, bullying, cruelty to people and animals, destruction of property, stealing and deceit. Delinquency is a legal concept essentially implying breaking the legal codes of the country.

Assessment

- It is important to establish the extent, frequency and severity of drug abuse. Additionally, information needs to be elicited concerning behavioural patterns, social competency, educational functioning, peer relationship and psychiatric status.
- Physical examination should include a check for fresh injection marks, old scars or the physical sequelae of drug use.
- A child's abuse of drugs is contextual, i.e. societal norm, family history, etc. Parental criminality or substance abuse increases the risk.

Management

- There are very few, if any, specialised treatment centres for children who abuse drugs. Treatment outcome will vary according to the chronicity and/or the substances abused. For example, limited impact is made on alcohol or marijuana abuse, whilst heroin or cocaine treatment programmes are more successful in reducing the use of these drugs.
- Volatile substance abuse is common in children but seldom persists into adulthood. Solvents are easily available, i.e. butane gas, lighter fuel, aerosols, etc. are most commonly abused through a plastic bag to maximise the effects.
- Stimulants like cocaine and amphetamines are taken in powder form, intranasally or injected. They produce elevation of mood, energy, reduction in appetite and hallucinations.
- Opiates can give rise to nausea and vomiting. Withdrawal symptoms include restlessness, irritability, increased bowel activity with abdominal pains.
- Methadone is the commonest approach to managing opiate dependence. Initially for children >15 years 10–20 mg daily, increasing by 10 mg/day until there are no signs of withdrawal or toxicity (usual dose 40–60 mg/day). Methadone is not appropriate for those with a short history of opiate dependence.

Schizophrenia

This is a serious mental illness characterised by abnormalities of thinking, perception and emotion, usually diagnosed

first in late adolescence though rarely the onset can be seen in childhood. Diagnose if two or more of the following symptoms are present for a month or more:

- Delusions – beliefs which are unshakeable
- Hallucinations
- Disorganised speech (incoherent)
- Grossly disorganised or catatonic behaviour
- Negative symptoms (flat affect).

Mood disorders may present with schizophrenic like symptoms and are sometimes difficult to disentangle. Careful physical examination is advised, as the symptoms of both schizophrenia and depressive disorder occasionally arise on the basis of a disease of the brain. If possible, an EEG should be undertaken to rule out temporal lobe epilepsy.

Assessment

- Diagnosis is dependent on detailed history-taking and examination, and schizophrenia is no exception.
- To evaluate the progress it is important to define the baseline symptoms, functioning and problems in various aspects of the child's life, i.e. education, family and social functioning.

Management

- Children and young people with schizophrenia present a challenge as they are seriously ill and often social and educational progress is seriously disrupted.
- Treatments so far lessen the frequency of relapse and disability, but do not cure the illness. Negative symptoms such as blunting of emotions, impoverished thinking and lack of motivation are particularly distressing to relatives.
- Pharmacological treatment to control symptoms is the important initial management. Psychoeducational, social, cognitive and family intervention programmes are important in long-term management.
- Oral neuroleptics provide the patient with a sense of control, any adverse effects are quickly apparent but the medication must be administered daily and the patient may not always be compliant.
- Depot neuroleptics provide regular opportunities for monitoring, but may delay adverse side effects for a few days following injection.
- Under normal circumstances treatment is initiated with chlorpromazine (orally 500 micrograms/kg every 6–8 hours starting dose progressing to control of symptoms or a maximum daily dose of 75 mg for aged 6–12 years and 300 mg/day for ages 12–18 years) or an equivalent but less sedative drug such as haloperidol (starting dose 25 micrograms/kg twice daily and progressing to control of symptoms or a maximum daily dose of 10 mg for aged 6–12 years and 20 mg for ages 12–18 years). Premature changes in drug choice should be avoided, as response time may be 30 days or more. Poor response may be due to inadequate dose or poor compliance. Depot medication is suitable for long-term treatment (flupenthixol by deep IM injection – test dose of 20 mg then after 7 days 20–40 mg repeated 3–4 weekly).
- Atypical antipsychotic drugs are now the treatment of choice if available. Risperidone in children >12 years starting at 1–2 mg per day increasing by 1 mg per week to a maximum daily dose of 6 mg.
- ***In refractory patients clozapine may be considered. It is however an expensive drug and carries a substantial risk of agranulocytosis and on initiation there is a risk of hypotension. The dose is 12.5 mg once or twice on day one, then building up by 25 mg/day to a final dose at three weeks of 150 mg twice daily.***
- It is important to work closely with the family. Highly expressed emotions and negative feelings increase the risk of relapse. The family will need support and help to reduce the intensity. The techniques to reduce highly expressed emotions require specialist training.

Post-traumatic stress disorder

Introduction

Post-traumatic stress disorder (PTSD) is a relatively new diagnostic category first officially created by DSM III in 1980. Between 25–35% of those exposed to traumatic events develop PTSD. Individual differences in response to trauma depend on:

- Stressor severity and degree of exposure to the stressor
- The child's perception of the event
- The child's appraisal of the threat to their survival, degree of human accountability
- For younger children, the response and functioning of adults, particularly close family, around them can be important
- Anxiety disorders, abnormal grief reaction, somatic complaints and diminution in school functioning can all occur.

Diagnostic criteria

- The child has experienced an event that is outside the range of usual experience, that is life-threatening to him/herself or those close to him/her.
- There is persistent re-experiencing of the traumatic event, that is distressing recollections, dreams or flashbacks.
- There is avoidance of the stimuli associated with the trauma.
- There is a range of signs of physiological arousal, such as difficulty in sleeping, irritability or poor concentration.
- In younger children, repetitive play related to trauma may be present.

Assessment

It must first be established that the child has experienced a traumatic event that preceded the onset of symptoms. The traumatic event may not necessarily lead to development of PTSD. Instead, the child may develop acute stress disorder or depression. Comorbidity is common and may

reflect symptom overlap. In other words, an additional diagnosis may be made.

In assessing the child, the interviewer will need to take account of the child's maturation, his/her verbal facility and functioning. Details of the traumatic event, the child's perception of the event, his/her response immediately and later, should be evaluated.

The differential diagnoses include obsessive compulsive disorder, schizophrenia or anxiety disorder. Flashbacks may need to be distinguished from intrusive and unwanted thoughts that are unrelated to the traumatic event, which occur in obsessive compulsive disorder.

Treatment (see Chapter 3.14)

- Little is known about the pharmacological treatment of PTSD in children and adolescents. If used it should be an adjunct to psychosocial treatments involving the child and family.
- Drugs with serotonergic actions (SSRIs) have achieved the most positive effects.
- Adrenergic-blocking agents such as **propanolol** (500 micrograms/kg to 1 mg/kg orally 2–3 times daily) and **clonidine** (2–3 micrograms/kg/day in three divided doses) may reduce significantly the avoidance, startle response and persistent arousal in traumatised children. Propranolol must not be given to children with asthma. Clonidine may cause hypotension.
- If medication is prescribed, care needs to be taken to explain to the child and family the purpose of the treatment, potential side effects, what to do if the child gets side effects, and other drugs/foods to avoid.
- Psychological treatments include cognitive behavioural treatment programmes. These in the main are based on a combination of classical and operant conditioning. The goals of treatment are reduction of symptoms, development of coping skills and helping the individual to gain a sense of well-being and control. Education and gradually increasing goal-setting helps the child to relax, solve problems and gradually achieve mastery over fearful thoughts. The help of a clinically trained psychologist or psychiatrist may be needed to plan the treatment.

Panic attacks

These are common in children and adolescents and can mimic physical illnesses. Hyperventilation, sometimes with tetany, is a key feature as well as the fear of "going crazy" or dying. The best way of controlling these is to explain the physiological features of panic to the child and the family and to teach proper breathing techniques (namely to breathe slowly at a rate normal for the child's age).

3.14 Grief and loss in war affected societies

Lynne Jones

These notes provide a brief introduction to working with grieving families in situations of political violence. The main experience for almost all those affected by political violence is massive loss. The range of losses are illustrated in Table 3.14.1. Their effect can be overwhelming. Understanding the reactions to loss, and how to distinguish between normal and abnormal grief and to assist in appropriate mourning will be one of the key tasks for health workers in these contexts and essential to understanding other psychological reactions and setting them in context.

Most children will be seen in the company of their surviving adult relatives whose own mental state will have a profound effect on the child. A child and family doctor must therefore be responsive to, and able to assess and support the whole family. These notes look briefly at grief in adults as well as children. They include:

- An introduction to basic concepts
- A brief discussion on recognising what is abnormal grief or other psychiatric disturbance in different cultural contexts
- Grief in children
- The interplay of trauma and grief in children
- Principles for working with families exposed to massive loss.

This chapter outlines a general approach to families, but does not give detailed management advice on the wide variety of symptomatic problems that can occur in grieving children. This is because there are a number of excellent manuals available on this topic, listed in the further reading.

Table 3.14.1 Some of the losses experienced by those exposed to political violence or life as refugees

Internal	External
Control	Family members
Autonomy	Friends
Security	Home
Identity	Community/country
Self-respect	Work/school
Belief in future	Money and other material possessions
Sense of belonging	
Trust	Physical health
Past	Religion
Meaning of life	Language
	Familiar diet

Why do we grieve?

The ability to form strong relationships with others is necessary for survival as human beings. We call this ability **attachment**. The sense of loss we feel when a loved one is absent, leads to searching them out and helps to keep families and groups together. Human beings could not have survived in previous eras if they did not live in groups which would enable them to feed and shelter themselves.

Loss is the sense of sadness, fear and insecurity we feel when a loved person is absent. It can also be felt for things and places.

Attachment/separation behaviour is most visible in children of 6 months to 3 years old. John Bowlby defined a cycle of behaviours that can be observed in an infant separated from its mother and then reunited with her. These are:

- Protest
- Despair
- Withdrawal
- Detachment
- Anger
- Re-engagement.

However, these behaviours can reappear in any of us throughout the life cycle when faced with separation from someone we love.

> Attachment behaviour is any form of behaviour that results in a person attaining or maintaining proximity to some clearly identified individual who is conceived as better able to cope with the world. It is most obvious whenever a person is frightened, fatigued or sick, and is assuaged by comforting and caregiving. At other times the behaviour is less in evidence. Nevertheless for a person to know that an attachment figure is available and responsive gives him a strong and pervasive feeling of security and so encourages him to value and continue the relationship. Whilst attachment behaviour is at its most obvious early in childhood, it can be observed throughout the lifecycle, especially in emergencies. Since it is seen in virtually all human beings (though in varying patterns), it is regarded as an integral part of human nature and one we share (to a varying extent) with members

of other species. The biological function attributed to it is protection. To remain within easy access of a familiar individual known to be ready and willing to come to our aid in an emergency is clearly a good insurance policy – whatever our age.
John Bowlby, *The Origins of Attachment Theory (1988)* in *A Secure Base. Clinical Applications of Attachment Theory.* London: Routledge, 1988.

Behaviour and emotions experienced after a bereavement

- Disbelief/numbing
- Sadness/despair/yearning
- Anger
- Acceptance

Note the similarity between a child's behaviour after separation from a parent, and our reactions to the loss of a loved person who has died. Thus death reactivates attachment behaviour. These feelings have sometimes been said to occur in stages. In fact their expression depends on culture, family values and individual personality. All the feelings may reoccur at different times. An individual may feel anger and sadness at the same time. An anniversary, or some other trigger in the memory, which reactivates the feelings of grief again, years after the event, may interrupt a long period of acceptance. Some have described grief as a "relapsing illness".

When discussing grief feelings with children I use the image of a wave. I ask them to imagine they are standing at the edge of the sea and a big wave comes along and knocks them over. They feel terrible but manage to struggle to their feet, then there is a period of calm water before the next wave. This time they are more prepared so that when the next wave comes it does not knock them over. What will happen over time is that, although the waves never go away completely, the periods of calm sea will grow longer, the waves get smaller and the child grow stronger (see Figure 3.14.1).

We can grieve for the loss of loved ones, for the loss of possessions, home, parts of ourselves (for example a limb after amputation), our whole selves when faced with life changing or terminal illness. Grief has mortality. It is possible to die of a broken heart. Suicide risk and the mortality of some illnesses increases after bereavement, particularly of a partner.

Mourning

Mourning is the name for the culturally appropriate processes which help people to pass through grief. All cultures mourn but in different ways. Mourning processes usually include acknowledgement and acceptance of the death, saying farewell, time periods for grieving, processes to continue attention towards the dead and to move beyond it and make new attachments. Different societies have different time periods set aside for grief, and different ideas about what is appropriate behaviour for different family members. They may also have different views on the appropriate role of children in these rituals. Sometimes families may be in conflict over what is the appropriate way to mourn. This is particularly the case in societies in a state of upheaval. See Appendix, Case example: A.

Political violence may disrupt the possibility of appropriate mourning. There may be uncertainty over missing relatives. The body may have been treated inappropriately, the normal rituals impossible to carry out during flight. Massive losses that affect whole communities also have the effect of depriving each individual of the normal support that they would receive from their community if their loss was a singular occurrence. The experience of loss is amplified by the fact that the family's loss is one of many in a community, so that the pain is amplified by this knowledge. But at the same time the family itself, not wishing to burden neighbours in a similar position to themselves, may become more reticent than usual about what is happening, and their own feelings. Because everyone is affected few are in the position to play the role of visitor, and comforter. Thus outsiders may have a significant role to play simply by encouraging and partaking as a visitor in the normal processes of mourning.

Abnormal grief

- "Too intense"
- "Too long"

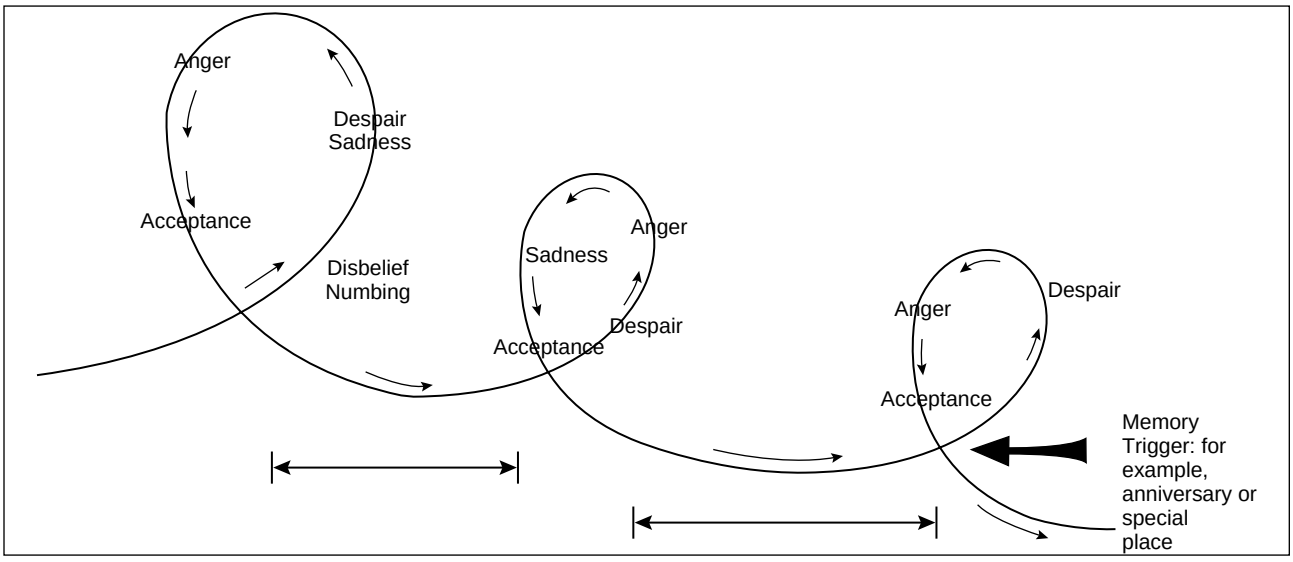


Figure 3.14.1 Waves of grief: the time intervals between waves get longer and the waves get smaller.

- "Delayed"
- "Culturally inappropriate"

The decision as to what is abnormal and inappropriate will depend on an understanding of both the family and the culture from which they come. You cannot decide what is abnormal without this cultural and personal knowledge

In Table 3.14.2 below the first column lists the feelings, thoughts perceptions and behaviours that can occur in normal grief. Some of these symptoms might also be the basis for a pathological diagnosis as indicated by the + signs in the other columns. However Western doctors need to be cautious in diagnosing psychopathology when the behaviour might either be the culturally appropriate expression of grief, or a direct result of lack of access to culturally appropriate methods of mourning.

Table 3.14.2 The symptoms of normal grief that can also occur in various psychiatric disorders

All the following are possible in normal grief	Depression	Post-traumatic stress disorder	Psychosis
Feelings			
Sadness	++	+	
Anger	+		+
Numbness	++	++	+
Fear	+	++	++
Guilt	++	+	
Nostalgia, yearning			
Anxiety	+	+	+
Thoughts			
Ruminations	++		
Intrusive thoughts	+	++	+
Unusual ideas	+		++
Suicidal thoughts	+	+	+
Perceptions			
Flashbacks		++	
Hallucinations	+		++
Behaviour			
Withdrawal	++	++	+
Aggression		+	+
Non-acceptance			+
Identification			+
Motor			
Agitation/ restlessness	+		++
Lethargy/apathy	++		++
Cognitive			
Poor memory	++	+	++
Poor attention and concentration	++	+	++
Disorientation	++	+	++
Biological			
Somatic symptoms of all kinds	+	+	+
Loss of appetite	++	+	
Sleep disturbance and nightmares	++	++	++

Which feelings and behaviours occur and which are regarded as normal for both children and adults will depend very much on how grief is expressed in that culture, and by that family as well as the religious values, temperament and personality of the individual. For example in Bosnia, it is regarded as appropriate for Serbian women to attend the funeral and to display their emotions visibly, keening and weeping. Muslim culture values a more stoical approach and sees the vivid display of emotions as inappropriate. In some cultures, for example parts of South East Asia, vivid dreams may be regarded as appropriate messages from the dead, in others (Western) dreams may be seen as an upsetting form of sleep disturbance. In Kosovar families, for example, there is often one individual (usually an older adolescent girl), who may cry a great deal, hyperventilate and faint, while the rest of the family remain stoical. The fainting girl may cause concern but she also seems to play a role in expressing grief for the rest of the family, whose concern for her also acts as a form of distraction from the loss. (See Appendix, Case example: Family B.)

Cultural bereavement

The Australian anthropologist and child psychiatrist Maurice Eisenbruch uses the term "cultural bereavement" to describe the massive losses experienced by refugees and all those displaced by war. (Eisenbruch 1991)

> Cultural bereavement is the experience of the uprooted person – or group – resulting from loss of social structures, cultural values and self identity: the person – or group – continues to live in the past, is visited by supernatural forces from the past while asleep or awake, suffers feelings of guilt over abandoning culture or homeland, feels pain if memories of the past begin to fade, but finds constant images of the past (including traumatic images) intruding into daily life, yearns to complete obligations to the dead and feels stricken by anxieties, morbid thoughts and anger that mar the ability to get on with daily life. It is not in itself a disease but an understandable response to catastrophic loss of social structure and culture.

In his work with Cambodian adolescents he found that those refugee children who had been encouraged to assimilate rapidly into a new culture, suffered more cultural bereavement than those encouraged to participate in traditional ceremonies and cultural practices.

He believes that the concept allows for a more integrated and culturally sensitive approach to the experience of loss, than attempting to classify any disabling symptoms only in terms of pathological categories according to Western diagnostic criteria. Disabling symptoms may be best addressed by a combination of restoring appropriate cultural practices and, if necessary, symptomatic relief.

Grief in childhood

Frequently asked questions:

- Do children grieve?
- Are they too young to understand?

- Should we protect them from unpleasantness and distress?
- Will loss in childhood cause later mental illness?

Children's reactions to death

The following notes are based on Western experience and should be taken as a guide. Working with victims of political violence in the Balkans has revealed that children, particularly from rural areas, have an earlier understanding of death but that in other respects the categorisation below holds true.

Under five years

There is little understanding that death is final. For example a 4-year old child in England, having helped to formally bury his dead pet rabbit in the garden, then asked if he could now dig it up so that he could have the rabbit back. Magical thinking results in misconceptions about causes and effects. An egocentric view of the world can lead to feelings of responsibility. "Mummy won't come back because I was naughty."

Reactions are similar to those following any separation. The longer the absence the greater the distress. It may be followed by detachment, so that the surviving family may think the child does not care.

Regressive behaviour, soiling, wetting, clingy behaviour, sleeplessness and minor illnesses can all occur.

Over five years

They can understand that death is irreversible, that certain physical changes occur, and that there is permanent separation. They may still not regard it as something that can affect them. They may continue to have some magical, concrete and egocentric thinking. At this age children more commonly use concepts of good and bad, they are curious about cause and effect, and able to articulate concern for others.

There is a desire to stay connected to the dead parent. Many children dream about and talk with the dead parent frequently; feel the dead parent is watching them and keep physical objects associated with them. The Harvard study found that 43% of children in a large community sample thought about the dead parent on a daily basis one year after death.

The reactions are variable. Boys are already learning to suppress feelings. 91% of the children in the Harvard study cried on first day; 50% had transient emotional and behavioural problems. Concentration and school work are affected. Repetitive play is very common.

Ten to adolescence

There is a growing understanding of abstract concepts: for example that death is universal and inevitable and can affect them personally. There is a growing concern with justice and injustice, and an awareness of inconsistencies.

The conflict between the desire for autonomy and need for closeness can be resolved by "indifference and detachment", or by identification and nostalgia. In a group for adolescent refugee boys ethnically cleansed from Northern Bosnia (all had lost their homes, some had lost their family) all spoke passionately and with great longing about their home-towns, describing them as the "most beautiful place to live". Revenge fantasies are not unusual.

There are less somatic and behavioural problems, and more depression is common. Poor concentration and lack of interest occur at school. The oldest child who has lost a same sex parent is at greatest risk.

Case example

G is a 13-year-old boy. During a long and brutal war his elder brother was killed on the front line. G was always very close to his brother. Three years later he continues to think about him on a daily basis. He visits the grave frequently and watches the video of the funeral once a week. He does not like to sleep alone and feels sad much of the time, although he is doing well at school. He talks about his brother a great deal. He would like to be as much like his brother as possible, who he believes was one of the bravest and most incorruptible people. He is angry about the peace agreement that he feels is unjust and makes a mockery of the aims for which his brother fought.

The most common immediate reactions

- Shock and disbelief
- Dismay and protest
- Apathy and feeling stunned
- Continuation of usual activities

Any of the following can occur:

- Anxiety
- Vivid memories
- Sleep problems
- Sadness and longing
- Anger and acting out behaviour
- Guilt, self reproach and shame
- School problems
- Physical complaints
- Regressive behaviour
- Social isolation
- Fantasies
- Personality changes
- Pessimism about the future
- Rapid maturing.

Age, personality, culture and family values, especially the way the parents or surviving caretakers react will all affect the expression of grief. The lack of synchronicity and different experiences within families can create problems. (See Appendix, Case examples A and Family B)

The traumatic experience can interfere with mourning. Avoidance that may be protective in coping with the trauma may prevent the acceptance that is necessary for coming to terms with the loss. Most agree that one must deal with traumatic symptoms first before grief work can be done.

Table 3.14.3 Distinguishing between the effects of loss and experiencing a traumatic event (adapted from Hendricks et al 1993)

Loss	Traumatic event
Image and memory of the lost person very important	Intrusive memories of means of death, no memory of lost person
Yearning and preoccupation with loss	Anxious and preoccupied with traumatic event
Separation anxiety changes to deep sadness	Anxiety about security can persist
Anger at loss	Irritable, diffuse unfocussed anger and rage
Guilt at not doing enough	Guilt at surviving
Seek out reminders of loved one	Hypervigilant, scanning environment for threat
Mourning as a tribute to the dead	Avoidance of thinking of event and reminders
Dream of dead person is comforting	Nightmares of event are terrifying

Long-term effects

Is grief an illness or part of normal life? If what is required is adaptation rather than cure, what is the role of the doctor or psychiatrist? Research evidence suggests that:

- Children who suffer an early bereavement do have a higher incidence of psychiatric disorder in later childhood
- Adults bereaved of a parent in childhood are more vulnerable to psychiatric disorder than the general population, particularly to depression and anxiety, precipitated by further losses.

Life events research shows that the following events are the most likely to be associated with later mental illness:

- Those that require people to undertake a major revision of assumptions about the world
- Those that are lasting in their implications
- Those that take place over a short period of time without preparation.

But when faced with a child who has experienced a traumatic loss, note the importance of the following:

- The response of the surviving parent
- Availability of other support
- Subsequent life circumstances
- Degree of continuity in the child's life
- How the loss is viewed by others
- What resources are available.

Case example: Telling the truth
The father was a member of a "Liberation Army" and killed in the fighting. His 32-year-old wife had two surviving children of eight and nine and continued to live with her husband's relatives. She told the children their father was working in another country. The children would frequently ask her why he did not phone and if he would bring them presents. They were confused because other children in the village told them their father was dead. When they questioned their mother she would start to cry, so they became nervous of asking her. Mother and brother-in-law asked for advice as to what to do and accepted my suggestion of sitting with the children and explaining in simple terms what had happened, answering all the children's questions as they came up, and sharing the experience of grief. Mother told me that the relief of not having to lie to the children had slightly eased her own distress and made it easier to respond to them. Moreover rather than being bewildered by their father's silent absence the children now talked about him in the village with pride.

Not all grieving families require a health worker's intervention. But in situations of political violence many families present to health workers because they have concerns about the long-term impact of events on the child, and want advice on how to talk about such abnormal events with their children. The health worker's role should be to facilitate the process of normal grieving, help to sustain and support the protective aspects listed above, and while treating pathology where it is evident, to avoid pathologising where it is not.

Some points on supporting grieving families

Many families are concerned that telling the child what happened will cause unnecessary distress and that as the child is "too young to understand", it is better to lie or

Case example: Telling the story
The family consisted of three surviving children (two girls and a boy) who had witnessed the death of their mother and aunt and fifteen other members of their extended family. They had been physically injured in the attack and spent some time in hospital. At our first meeting four months after the event, two of the children did not believe their mother was dead. They hoped she had survived as they had. We sat together with the children's father who told them gently that he thought that in all probability she was dead. The children cried and we did not discuss it further that day. The following week the bodies had been identified, funeral notices posted and the funeral arranged. The family had returned to the house where the massacre had occurred and appeared to be functioning well. The boy had no symptoms, although the younger girl was sad and quiet. When asked if they wanted to talk about what happened, she said no. Her brother said that he had already talked with journalists and did not feel a need to go over it again. The elder girl (14) had some intrusive thoughts and memories, and poor sleep and appetite. She wanted to walk me around the site and retell the events in detail.

The other children did not wish to join in. During the walk the elder girl told me that she now knew her mother was dead. All the children then wanted to show me all their old photographs, and all the children participated in identifying their dead relatives and telling me stories about their life before the war. After the funeral (which only the elder girl attended at her own request) the children appeared more cheerful and all were looking forward to school. The surviving family provided an extremely loving and supportive network and although the father was extremely sad, he allowed the children to talk about their mother whenever they wanted. Later the whole family was sent to another country for medical treatment for the children. They attended local schools where they learnt English, and appeared to be adjusting well. A year after the events the International Criminal Tribunal wished to interview them regarding the massacre, and the three children insisted that they would like to give their accounts. All the children made statements recorded on video and gave similar detailed stories about the events surrounding the massacre. Although they found it distressing each obviously regarded it as significant and important, and were pleased to have had the chance. This example illustrates that children in the same family will not all deal with their grief in the same way. If given the opportunity they will find the most appropriate time to tell their own stories in the way that will give it significance and meaning for themselves.

avoid the subject when it comes up. Children are very protective of surviving parents and quick to sense when a question causes distress. They may avoid asking for information because the questions make the parent cry. False information leads to confusion and a lack of trust.

- Children need clear, honest, consistent explanations available to their level of development.
- They need to accept the reality of the loss, not be protected from it.
- Magical thinking should be explored and corrected. What is imagined may be worse than reality and children may be blaming themselves for events beyond their control.
- However the insistence on getting a child to "debrief" or tell the story of their loss may not be therapeutic or appropriate. Not all cultures put a high value on the ventilation of individual feelings, as Western culture does. The therapist's goal should be to encourage a supportive atmosphere for the children, where open communication is possible, difficult questions answered, and distressing feelings tolerated. This means that the child will be free to express their grief in the manner they find appropriate to the person they most trust, and at a time of their own choosing.
- Help the family to cope with traumatic symptoms if they exist. Give the parents information as to what to expect and straightforward management advice.
- Help the child maintain connection with the lost parent. Encourage the parents to allow the child to choose a memento to keep, access to photographs and to answer the child's questions about the dead relative.
- Support the surviving family. Help them to access the appropriate agencies to solve the practical problems they will encounter. Attending to basic needs is essential. Engaging in the process of rebuilding their lives helps families to come to terms with their losses (see Appendix, Case example A).
- In situations of political violence the question of justice will be important for families. Many will state that they cannot come to terms with their losses while the fate of loved ones is unknown, bodies unidentified, or perpetrators at large. These issues will affect the children, and older children may bring them up spontaneously and wish to discuss them. Health workers may be asked their own views. Stating a willingness to learn and understand, along with an awareness of one's own biases and subjectivity is the most helpful position. Political and cultural literacy are essential. The family should be put in touch with the appropriate human rights or justice agencies if they wish to give formal evidence, so that the therapeutic and confidential nature of your own work remains clear and the family are not confused as to the purpose of the interview. Giving testimony to such agencies should always be at their own request. In this case it may prove therapeutic (see Case example: Telling the story).

Appendix

Case example: A – Complex needs and conflict in a grieving family

A is a high school student of 18, living in a rural area in the heart of a conflict region, the second eldest of seven children (four girls and three boys). She wanted to study medicine. Her life and health were normal, until the shelling began and her family fled to the forest, where they spent three months. The local police of a different ethnic origin found them and separated men from women and elderly men and sent the latter home. They got home to find their village full of army and police and themselves under siege at their home, where they were harassed and sometimes beaten. Meanwhile their invalid, pensioner father was shot in a massacre of 10 men from the village. He was buried while they were under siege. A was referred to me one month after this by a local doctor concerned at her mental state. When I first saw her she was extremely sad and frightened. She was crying all the time, ruminating about her father being captured. She found everywhere frightening, and was too frightened to go to sleep, but when she did fall asleep woke early. She had no appetite, and a diurnal mood swing.

I first assessed her at the doctor's home where we had a long talk at her instigation, about all that had happened to her. I felt the severity of her depressive symptoms might necessitate using an

antidepressant, but delayed making a decision until I was able to assess her at home with her family. I visited them a week later and found all seven of them living in one restored room of their fire damaged home. To my surprise, A was a great deal better, her sleep and appetite having returned to normal over the week. She informed me that she felt this was because of feeling she had someone to talk to, and who "wanted to come and visit". However all the female members of the family were preoccupied with father's death, tearful in discussion of it and in conflict over how to manage the grief. The mother and one sister no longer wanted to wear the symbolic mourning clothes and to move on. The other three sisters were wearing black mourning bands in their hair and wanted to do so for the appropriate period of a year. One of these sisters complained she was having some panic attacks. They also all felt angry and concerned about their material circumstances. They had no access to father's pension as this would have meant going to a police station run by the ethnic group in power to get new identity papers (all burnt) and identifying themselves as from a conflict area and as members of a family that had suffered a massacre victim. Anxiety made sleep difficult. Interestingly the boys in the family (14, 8, 7) appeared cheerful, busy and well, insisting they were symptom free, although they missed their father. All the boys attended school regularly. The girls did not go, as there was no money for books. They therefore sat around at home with little to do.

We agreed to have family meetings to help them resolve their conflict about how to grieve; and relaxation therapy to provide some symptomatic relief. We did this as a group and they practised themselves on a daily basis, mother running the group. Over the weeks there was a marked improvement in the whole family. The three girls continued to wear their mourning bands and the mother was more tolerant of this. A began to press me to help her get an ID card so that she could go to a nearby town, get a job and earn some money. However the security situation deteriorated too much for this to be possible. My last visit before evacuation was distressing, as there was fighting on the nearest main road and the sound of shelling of nearby villages. We all knew that they might have to flee again in the near future.

I returned to the family three months later. They had spent these months internally displaced pushed from one village to another, with very little to eat. During this time the 14-year-old son, who had separated himself from the family believing he endangered them, had been killed along with another male relative. The family had returned to their home to find it completely burnt to the ground except for an outhouse. They had nothing left and were using an ammunition box as a table, and sleeping under a small piece of plastic in the garden, because the outhouse attracted snakes. As previously, the healthiest members of the family appeared to be the smallest boys, who denied any symptoms except some tearfulness now and then. They appeared active and cheerful except when witnessing their mother's distress. The mother was devastated, and could not stop crying. She could not sleep or eat or function and expressed suicidal ideas. A had moved to an aunt in a nearby town and had a number of somatic symptoms. We provided clothes, basic material equipment for the house. Her mother was started on antidepressant therapy.

The family then lost contact with our service for six months. They had been provided with materials to build a warm room but the aid agency had failed to realise that with no adult males left there was no one to build it. The family therefore moved into a grim damp refugee flat in town. The mother had found the antidepressants helpful but had run out of medication. Two daughters had escaped the situation by marriage. The boys were well and attending school. The other daughters remain trapped within the prison of their mother's unremitting grief. They spent all day in the flat with their mother talking and crying. She did not wish to be left alone. They wanted to show her how much they cared for her and insisted on doing every household task, which added to her feeling of being a useless burden. We began "family work" again: encouraging the girls to join the free local youth club and to allow the mother to re-establish her maternal role in the family, supporting her by restarting the antidepressant medication at her request, and getting in touch with the aid agency about their house.

Some reflections on this case. For most families of this particular ethnic group, the immediate and respectful burial of the dead is crucial. This is followed by seven days of visiting by friends and family, who sit all day with the bereaved and discuss the dead. These normal mourning processes had not been possible either for the father or son. It seems likely that the surprisingly sudden symptomatic relief A gained from my initial intervention, was through my contributing to some of this normal mourning by being an outsider who visited and listened. A family approach meant that differences could be brought out in the open in a respectful way. The family also formed a natural group that could encourage and support each other in doing relaxation work. Attending to human rights concerns such as identity papers and security was also important. However all this was undone by the second round of conflict and loss. There is something particularly devastating about loss coming again immediately after having begun to work one's way to recovery. Being made homeless and not being given support to rebuild their house has contributed to their sense of bereavement and powerlessness, and prolonged the period of grief. The mother told me repeatedly that if she could start rebuilding her house she would feel better.

Some families are strongly patriarchal. There are different coping strategies available to boys and girls. All the women in this family came across as strong and capable, but all felt that the loss, first of an invalid father, and then of the eldest boy, had destroyed the family's capacity to function at

all. Much of the work with grieving female survivors has to address their insecurity and lack of confidence in their own self worth. This family required a complex approach: participation in normal mourning; attention to basic needs; help with family communication; symptomatic relief; help in re-establishing normal family roles and adapting to new ones in the absence of male support.

Case example: Family B – Supporting the whole family

Family B had lost more than 20 members – mostly female and children – in a massacre. I was asked to visit because of concerns for the mental health of the surviving children who had witnessed the attack and were all under six. At the first session most of the remaining extended family, including the children, had gathered to meet me in the only intact room in the house. I already knew the outline of what had happened and used this first meeting to draw a genogram. I have found that in situations of mass violence, in a culture where the extended family is of central importance, this simple technique has a number of useful functions.

- It is a collective act, everyone joins in, introducing themselves and explaining their connection to others.
- It is interesting for the children, who join in the actual drawing on a large piece of paper in the centre of the room.
- By asking them to include those who have died, it allows for a collective naming of the dead. In this family my symbolically putting a black simple line through these names took on a ritual significance and the children were quick to point out when I missed someone out.
- The naming allows the person to be identified, but how much is said about that person or what happened, is up to the family, thus it opens the possibility for story telling without forcing the issue.
- What is said about the dead is said in front of the whole family so that there is collective narrative from which the children are not excluded.

Once the genogram was done the family told me their concerns about the children and their own fears about letting the children talk as it seemed to upset them. At this meeting I gave the simple advice about communication outlined above and arranged to meet the family regularly and to have play therapy with the children. At the next meeting, the family informed me they were concerned about the eldest teenage girl who fainted regularly at the same time every afternoon, and was the most nervous and sensitive member of the family. Her sister was one of the dead and her mother was particularly concerned about her health, but never cried herself. They wanted reassurance that the girl was not seriously ill. Having provided this I wondered aloud if the teenage daughter was in some way grieving for the whole family and that this exhausting work might be causing her to faint. It also meant that mother did not have time to think about her own sadness. The daughter said she wished her mother would cry a little and not worry about her so much in which case she could look after her.

By the next meeting the daughter was no longer fainting and mother was now actively grieving. I continued with family meetings and play therapy over the next six months.

During this time the oldest child (5 years) of the section of the family in which the mother had died began to tell his father fragments of what he had seen and to ask questions about his mother. The father had taken out photographs of her to show to all the children. At no point did the children tell the story to me, nor did I insist upon it, seeing my role as facilitating and supporting communication within the family. Over the following year the children changed from tearful and withdrawn, to outgoing, cheerful, communicative and energetic. They all attended the formal reburial of their family. Their father remarried and their new step-mother was well accepted. He began on the process of rebuilding his house. They remained well at our last contact and the eldest child had begun school without problems.

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3.15 Vitamin and mineral deficiencies

Rob Moy

Minimum standards requirements

- Adequate diets
- Vitamins A, B, C, D and K
- Folic acid
- Zinc
- Copper
- Iodine

Vitamin A deficiency

Importance

- Single most important cause of childhood blindness in disadvantaged countries.
- Significant contribution to morbidity and mortality from common childhood infections even at subclinical levels of deficiency.

Prevalence

- Endemic in at least 60 countries worldwide especially in Africa, South and South-East Asia, some areas of Latin America and the Western Pacific.
- 254 million pre-school children are at risk.
- 5–10 million pre-school children clinically affected with eye disease.
- 250 000 cases of blindness per year.

Aetiological factors

- Persistent inadequate intake of vitamin A exacerbated by insufficient consumption of dietary fat leading to ineffective absorption.
- Frequent infections, especially measles, depressing appetite and increasing the body's utilisation of vitamin A and depleting liver stores.
- In an epidemiological context of poverty, social underdevelopment, hostile living environments, water shortage and food scarcity, and individual factors such as lack of breastfeeding, inappropriate weaning practices and increased physiological needs during periods of rapid growth.

Clinical effects

- Night blindness (decreased ability to generate rhodopsin in retinal rod photoreceptors essential for vision in dim light).
- Compromised integrity of epithelial surfaces leading to "dry eye" (xerophthalmia), that is conjunctival xerosis, Bitot spots, corneal xerosis, corneal ulceration, irreversible damage to the eye (keratomalacia).
- Depressed immunity.
- Increased susceptibility, duration and severity of common infections, for example acute respiratory infection, diarrhoea, measles.

Assessment of vitamin A status

- ***Serum retinol < 0.7 micromol/litre.***
- ***Conjunctival impression cytology (detecting early loss of vitamin A-dependent goblet cells and metaplasia of epithelium).***
- Vitamin A deficiency becomes a public health problem when the following are prevalent:
  - Night blindness > 1%
  - Bitot's spots > 0.5%
  - Corneal xerosis with or without ulceration > 0.01%
  - Corneal scarring > 0.05%

Treatment

Table 3.15.1 Treatment of vitamin A deficiency: WHO schedule (Oral)

	Day 1	Day 2	2 weeks later
< 6 kg	50 000 IU	50 000 IU	50 000 IU
6–10 kg	100 000 IU	100 000 IU	100 000 IU
> 10 kg	200 000 IU	200 000 IU	200 000 IU

Deep IM injection of vitamin A (retinyl palmitate) 50 000 IU for < 2 years and 100 000 IU at > 2 years if severe stomatitis, persistent vomiting or malabsorption is present

- Regular vitamin A supplements are advised for all young children in poor countries. It has been shown to reduce overall mortality and morbidity from diarrhoea.
- Vitamin A 200 000 IU orally should be given to all children admitted with persistent diarrhoea, dysentery, acute respiratory infection, severe malnutrition and measles.

- Dietary education about vitamin A rich foods (for example dark green vegetable leaves, carrots, mango, papaya, eggs, orange fruits, liver, red palm oil, fatty fish).
- Treat siblings and mother.

Beri-beri Vitamin B_1 deficiency

- May occur in areas of severe nutritional deprivation where little more than polished rice is consumed.
- Affects adults, children and breastfed infants of thiamine-deficient mothers.
- Often mistaken for oedematous malnutrition (kwashiorkor), nephritis, cerebral malaria, encephalopathy or septicaemia.
- Causes cardiovascular and neurological signs:
  - cardiac failure with breathlessness, oedema, tachycardia
  - peripheral neuritis
  - acute encephalopathy
  - aphonic form: noiseless cry due to laryngeal nerve paralysis.
- ✔ **Beri-beri is rapidly fatal.**
- Initial dose 50–100 mg thiamine hydrochloride. IM or orally, particularly effective in heart failure (have facilities for treating anaphylaxis available)
- Continue with 10 mg/day for <2 years and 25 mg/day for 2–12 years and 50 mg/day for >12 years

Pellagra Nicotinic acid (niacin) deficiency

- Common where maize is the staple diet.
- Clinical features:
  - dermatosis of parts of the skin exposed to sunlight
  - diarrhoea and malabsorption
  - rare encephalopathy.
- Treatment: 10 mg nicotinic acid three times daily for 7 days in children <2 years and 25 mg three times daily for 7 days in children >2 years. Treat other vitamin B deficiencies at the same time (thiamin and riboflavin).

Scurvy Vitamin C deficiency

- Vitamin C is needed for collagen formation (in bones, cartilage, teeth, and capillary walls)
- Is important in the healing of wounds
- Increases iron absorption
- Found in citrus fruits, vegetables and breast milk. Very little in cow's milk especially if heated
- Found in severe malnutrition and in children fed on very poor diets in institutions
- Causes spontaneous haemorrhages, especially from gums, and defective bone, cartilage and dentine formation.

Clinical features

- Local tenderness and swelling of the legs (due to subperiosteal haemorrhages)
- Pseudo-paralysis of limbs
- Haemorrhagic and spongy changes in the gums
- Petechiae and ecchymoses around the eyes
- Microscopic haematuria may be present
- Anterior ends of the ribs swell
- Mild anaemia
- Characteristic *X* ray appearance: loss of trabeculae in long bones gives ground glass appearance, dense lines of calcification in the epiphysis next to the epiphyseal plate and calcification of sub-periosteal haemorrhages.

Treatment

- Vitamin C orally 200 mg daily for 7 days
- Subsequent improvement in diet

Vitamin D deficiency: Primarily vitamin D_3 (cholecalciferol) deficiency

Vitamin D deficiency causes the following:
- Rickets (failure of mineralisation of growing bone)
- Hypocalcaemic tetany in infancy
- Osteomalacia in adults.

Nutritional rickets is most prevalent in North Africa, Middle East and Pakistan. Asian and Afro-Caribbean children are also at risk in the UK and other countries where limited sunshine is present.

Aetiology

- Vitamin D increases Ca^{++} absorption from gut, reabsorption of Ca^{++} from kidney, and a phosphate diuresis
- Vitamin D deficiency reduces Ca^{++} in ECF and increases parathyroid hormone (which increases phosphate loss by the kidney). Result is a low Ca^{++} and low phosphate. Subsequently a rise in alkaline phosphatase and then *X* ray features of rickets.
- An infant's diet especially if not breastfed contains only small amounts of vitamin D and so fortification of foods and vitamin D supplementation is recommended.
- Lack of vitamin D-containing foods such as oily fish, eggs, butter, margarine, meat and milk is important.
- Lack of sunlight exposure (UVlight) (black and brown skinned children are particularly at risk).
- If a child presents with rickets and has a normal exposure to sunlight, consider a hypocalcaemic diet (reported in South Africa and Nigeria). Cereals can bind calcium and prevent its absorption.
- Rarely, a metabolic disorder such as familial hypophosphataemic rickets. Where consanguinity is common, renal tubular disorders can produce this.
- Exclude chronic renal failure.

Clinical features

- Disturbance of the normal growth of the epiphyseal plate leads to the formation of inadequately calcified

new bone at the diaphysis edge of the plate (so called osteoid tissue). The proliferating zone on the epiphyseal side of the plate enlarges excessively producing a swelling of the plate. Osteoid tissue may also form sub-periostally. There is also demineralisation of the skeleton. The following features result from these abnormalities:

- epiphyseal swelling (especially distal radii at the wrists, also ankles and knees)
- craniotabes (soft areas of the skull bones, especially of the occiput which when pressed gently are easily depressed)
- rickety rosary (enlarged costochondral junctions)
- delayed fontanelle closure
- curvature of the shafts of the tibia and femur may occur in severe cases
- bossing of the frontal and parietal skull bones due to osteoid formation
- pidgeon chest (pectus carinatum)
- Harrison's sulci
- deformities of the thoracic and lumbar spine can produce kypho-scoliosis and lumbar lordosis
- pelvic bone deformities in the female child can lead to subsequent birthing difficulties due to damage to the inlet and outlet of the birth canal
- delayed dentition
- delayed gross motor development with generalised muscle weakness and hypotonia
- growth retardation.

Diagnosis

- Very elevated plasma alkaline phosphatase
- Usually normal, possibly slightly low plasma calcium
- Very low plasma phosphate (normally 1.6 to 2.3 mmol/litre but in this situation 0.6 to 1.0 mmol/litre)
- Lowered plasma levels of 25 hydroxy-vitamin D_3
- Typical *X* ray appearance: cupping and fraying of distal ends of the long bones such as the ulna and radius. There is widening of the metaphyseal plate due to osteoid formation. The periosteum may be raised. There may be abnormal curvature of bones and generalised under-calcification.

Prevention

Exposure to sunlight and foods such as egg yolk, milk and fortified margarine

Treatment:

25–125 micrograms vitamin D_3 (1000–5000 IU) daily for 4 weeks.

Vitamin K deficiency

- Cofactor for the hepatic synthesis of clotting factors (prothrombin, VII, IX and X)
- Sources are green leafy vegetables and synthesis by gut flora
- Deficiency may occur as a result of the lack of bile salts, the malabsorption of fats, after the use of broad spectrum antibiotics or in the breastfed newborn (see Chapter 3.48)
- Treat with Vitamin K 5–10 mg orally, IM or IV (1 mg in the newborn) (see Chapter 3.48)
- Prevent haemorrhagic disease of the newborn by giving vitamin K to all newborn infants either orally or IM.

Folic acid deficiency

- The most important issue here is the relative deficiency that predisposes to neural tube defects when present during the early period postconception in the pregnant mother
- Relative deficiency occurs in haemolytic anaemias and in preterm infants (see Chapter 3.23)
- Deficiency occurs in malabsorption syndromes such as coeliac disease and blind loop syndromes
- Anticonvulsants such as phenytoin may interfere with its metabolism.

Consequences

- Foetal abnormalities
- Megaloblastic anaemia, neutropenia and thrombocytopenia

Treatment

- All females anticipating pregnancy should be taking an additional 400 micrograms of folic acid per day before and throughout pregnancy
- To treat deficiency: infants 500 micrograms/kg once daily and children >1 year 5 mg once daily. Treat for up to 4 months and exclude concomitant vitamin B_{12} deficiency, which if untreated could result in neuropathy.
- For haemolytic anaemia treat with 2.5–5 mg orally once per day from 1 month to 12 years and 10 mg once per day after 12 years of age.
- In preterm infants 100–200 micrograms orally per day.

Iodine deficiency

- Iodine deficiency in pregnancy causes maternal hypothyroidism and cretinism in the newborn.
- Is one of the commonest causes of disability worldwide.
- Clinical features range from mild neuromuscular incoordination and cognitive deficit to severe mental retardation, spasticity and deafness, and severe stunting of growth.
- Endemic in mountainous regions far from the sea (for example Andes, Himalayas, Central Africa, Papua New Guinea) and areas where iodine is eluted from the soil by repeated flooding, for example Bangladesh.
- Prognosis is poor even after early recognition and treatment with thyroid hormone.
- Prevention by salt iodination or single oral dose of iodine in pregnancy.

Iron deficiency

See Chapter 3.21

Zinc deficiency

- Dietary sources are meat, poultry and fish. Staples will decrease bioavailability and absorption.
- Zinc deficiency impairs growth, decreases immune function and increases susceptibility to infection.
- A congenital absence (rare) of zinc absorption causes acrodermatitis enteropathica (severe mucocutaneous ulceration, failure to thrive and excess infection).
- Zinn supplementation has been shown to decrease morbidity in acute and persistent diarrhoea and respiratory infection, and to improve linear growth and weight gain.
- Zinn supplements of 2 mg/kg/day should be an essential component of mineral mix for the management of severe malnutrition. Excess copper in the diet may interfere with zinc absorption.

Copper deficiency

- Rare except in severe malnutrition and very low birth weight infants.
- In infants 200 micrograms to 1 mg/day. In children >1 year 1–3 mg/day.
- Severe deficiency causes osteoporosis.

3.16 Severe malnutrition

Michael H N Golden

Minimum standards requirements

- Scales, charts, low-reading thermometer
- ReSoMal
- Electrolyte and mineral mixtures
- Antibiotics and metronidazole
- IV 10% glucose
- Vitamins
- Blood transfusion
- Mebendazole
- F75 and F100 feeds
- Barrier skin cream

Severe malnutrition (multinutrient undernutrition) is characterised by oedema or wasting, usually with anorexia and infection. The main immediate causes of death are infections, septic shock, hypoglycaemia, electrolyte imbalance, dehydration, hypothermia, cardiac failure and severe anaemia. Every physiological and metabolic function is impaired so that they are extremely fragile, similar to the premature neonate. Two clinical pictures are seen with much overlap.

- **Marasmus** affects all ages but young infants are particularly at risk. It is due to a deficiency of Type II (growth) nutrients (usually non-breastfed infants). The baby is extremely thin with loss of subcutaneous fat producing skin wrinkles and folds. Weight-for-length or height is <70% of the median (see Chapter 6.1).
- **Kwashiorkor** usually occurs in children of 2–4 years of age. Kwashiorkor is an acute illness that suddenly appears over a few days. It is due to a deficit in the antioxidant nutrients. It presents with sodium retention and oedema, skin lesions that are like severe sunburn in a fair skinned person. There is fatty liver, with low circulating levels of all hepatic export proteins. The hair may be depigmented (this has no relation to prognosis and should be ignored clinically), but the hair pulls out very easily and painlessly (which is related to prognosis). Initially, there is a high level of circulating nitric oxide and vasodilatation that resembles toxic shock.

In severe malnutrition, biochemical abnormalities include:

- Low urea
- Severe hypoproteinaemia
- Hypokalaemia, hypophosphataemia
- Hypomagnesaemia, and
- Hypoglycaemia (see below).

Anaemia is frequently present.

Principles of treatment

Treatment is much more successful if standard treatment protocols are followed than if clinical judgements are made on individual patients. This is because the illness itself changes the clinical presentation, signs and symptoms of common complications.

The treatment of severe malnutrition is divided in two phases separated by a transition phase.

- **Phase I (initial treatment)**
  Specific objectives: return of normal homeostasis and treatment of complications.
  - The immediate treatment of life-threatening complications – hypoglycaemia, hypothermia, heart failure, septic shock, infections and infestations, severe dehydration and very severe anaemia.
  - The prevention of hypoglycaemia and hypothermia.
  - Nutritional treatment based on a maintenance diet (total 100 kcal/kg/day), divided into frequent meals (eight meals per 24 hours).
- **Transition phase**
  The diet is gradually increased over 4–5 days.
- **Phase II (rehabilitation or catch-up growth)**
  Specific objectives: promote rapid weight gain (10–20 g/kg/day) and the preparation for discharge.
  - A nutritional treatment based on a high-energy intake (160–200 kcal/kg/day) divided into six meals a day.
  - Emotional and physical stimulation.

The treatment in phase II can be given in special day-care units.

Admission criteria

- Weight-for-height of <70% of the median.
- Oedema
- Mid-upper-arm circumference of <110 mm if child is over 65 cm in length.

Assessing nutritional status and recovery

See Chapter 6.1 practical procedures.

Discharge criteria

Depend upon the quality of the follow up services. With good follow up services and a supplementary food available:
- Weight-for-height of >85% of median for 3 days
- And no oedema for 2 weeks
- And no medical complication for 2 weeks.

Without good follow up services:
- Weight-for-height of >95% of median for 3 days
- And no oedema for 2 weeks
- And no medical complication for 2 weeks
- Taking family food well and maintaining weight.

Medical and nutritional history and examination

The proforma history and examination sheet (Appendix 7.4) should be filled in by the admitting physician or experienced nurse.

Key points in the history are:
- Recent intake of foods and fluids
- Usual diet before current illness
- Breastfeeding?
- Duration and frequency of diarrhoea and vomiting
- Type of diarrhoea (watery/bloody)
- Appetite
- Family circumstances
- Chronic cough
- Contact with TB?
- Contact with measles?
- Known or suspected HIV infection

Key points on examination are:
- Oedema
- Dehydration? (very difficult to diagnose: NOT possible in the oedematous child)
- Shock? (often gives appearance of dehydration in child with oedema)
- Severe palmar pallor?
- Eye signs for vitamin A deficiency? Dry eyes, Bitot's spots, corneal ulceration, keratomalacia (see Chapters 3.15 and 3.34)
- Signs of local infection (ear, throat, skin, pneumonia)
- Signs of HIV
- Fever
- Hypothermia (rectal or mouth temperature <35.5 °C)
- Mouth ulcers or other oral problem
- Skin changes of kwashiorkor (hypo- or hyperpigmentation, desquamation, ulceration, exudative lesions resembling burns often with secondary infection such as *Candida*.

✔ **Note: Children with vitamin A deficiency are likely to be photophobic and will keep their eyes tightly closed. Examine carefully to prevent corneal rupture.**

Laboratory tests

Laboratory tests are not needed to guide or monitor treatment. Electrolytes and haemoglobin are difficult to interpret and can easily misguide. If haemoglobin is measured this should be done on admission only, and a transfusion given at this time if essential. A patient should not be given a blood transfusion after the first 48 hours. The haemoglobin nearly always falls after admission due to haemodilution with expansion of the circulation during mobilisation of oedema and export of sodium from inside the cells in marasmus – at this time, with expansion of the circulation, there is such a grave danger of precipitating heart failure that a transfusion should not be given, even for very severe anaemia.

A malaria smear is useful if malaria treatment is not given as part of the routine management of all severely malnourished children.

Details of treatment

In Phase I (initial phase) restore nutritional imbalances, metabolic function and treat complications. Phase II (catch-up growth) is a period of rapid weight gain. There is a "transition phase" between these phases.

Table 3.16.1 The phases of treatment

Phase I (1–7 days)	Transition (3–4 days)	Phase II (usually 14–21 days)
Treat dehydration, hypoglycaemia, hypothermia Treat infection and do not give iron Correct electrolyte imbalance Diet: maintenance intake Stimulate child	Moderate intake Stimulate child Treat helminths Do not give iron	Correct nutrient deficiencies and iron deficiency High intake Stimulate child Provide physical activities Prepare for discharge

- There are **routine measures** that are systematically given to all malnourished children, and additional routine treatments that are often added.
- **Specific treatments:** including emergency management of life-threatening complications and of specific diseases.
- **General points:** On admission severely malnourished children should be separated from those with infections and kept in a warm room (25–30 °C) without draughts. Washing should be minimal and when possible with warm water and the child immediately dried. The mother should be encouraged to stay with her child.

Intravenous infusion and blood transfusion

Intravenous infusions are to be avoided in all severely malnourished children: the risk of precipitating heart

failure is very high because of their atrophic heart muscle, high intracellular sodium and electrolyte imbalance.

- The only indication for IV infusion in severely malnourished children is unconsciousness due to circulatory collapse.
- The only indication for blood transfusion is when the anaemia is present on admission and is life-threatening.
- Cannulae should not be kept open but removed immediately after the prescribed treatment is given.

Nasogastric tube feeding is recommended in cases of:

- Anorexia with an intake of less than 70 kcal/kg despite patient coaxing
- Severe dehydration with inability to drink
- Child cannot drink and eat because of weakness or clouded consciousness
- Painful or severe mouth lesions (herpes, cancrum oris)
- Repeated, very frequent vomiting.

Try to not tube-feed for more than 3–4 days. Always explain the reason to the mother.

Try to breastfeed or feed by mouth every time.

Dehydration with severe malnutrition

Dehydration from diarrhoea is common in severely wasted children (marasmus) on admission. The treatment of dehydration is not the same as in the non-malnourished child (with the exception of cholera).

This section does not apply to mild diarrhoea occurring during transition from one phase to another.

Signs of dehydration in malnutrition

✔ The normal signs used to assess dehydration are all unreliable in severe malnutrition. **Assume all children with acute watery diarrhoea have some dehydration**. The interpretation of the signs relies on the history.

The specific signs are:

- History and observation of frequent WATERY diarrhoea
- History of recent sinking of the eyes: the eyes appear "staring"
- History of not passing urine for 12 hours
- History and observation of thirst.

Reduced skin turgor and sunken eyes (that are longstanding) are features of malnutrition itself. It is not possible to adequately judge the degree of dehydration in the severely malnourished child.

The appearance of dehydration in children without watery diarrhoea or in children with oedema can be caused by a toxic shock with dilatation of the blood vessels – these patients should not be treated as if they have dehydration.

✔ **Note: Low blood volume can occur with oedema.**

Routine treatment

Standard WHO-ORS solutions have too high sodium and too low potassium for children with severe malnutrition. ReSoMal (rehydration solution for malnutrition: see Appendix 7.5) is a special solution for this situation.

- For the children with watery diarrhoea in an adequate clinical state:
  At admission, give one dose of ReSoMal orally or by nasogastric tube and start to feed the child with the phase I diet. Further ReSoMal can be given after each stool or vomit.
  - 50 ml for children less than 85 cm in length (approximately <2 years)
  - 100 ml for children over 85 cm in length (>2 years)
- **For children with watery diarrhoea in a poor clinical state:**
  Start rehydration with ReSoMal immediately (10 ml/kg per hour for the first 2 hours and then 5 ml/kg per hour until rehydration is complete).

This rate is slower than for normally nourished and dehydrated children.

Completed rehydration

The rehydration is completed when the child is alert, no longer thirsty and has passed urine. There should be less sunken eyes and fontanelle and improved skin turgor. (Care: loss of sunken eyes in a severely wasted patient or the worsening of oedema can be a sign of overhydration)

The diet should now be given.

Monitoring

ReSoMal at 70 ml/kg weight per day is usually enough to restore hydration. But take care, rehydration can quickly lead to fluid overloading giving cardiac failure or sudden death. Malnourished children cannot excrete excess sodium. The clinical state of the child should be reassessed every 30 minutes during the first 2 hours then every hour. The best way to monitor the child is by regularly measuring his/her weight – this gives "fluid balance" directly and accurately, without having to measure any stool or vomit.

The ReSoMal should be stopped immediately if:

- the body weight increases by 10% or more
- the respiratory or pulse rate increase
- the jugular veins becomes engorged
- oedema appears or the eyelids become puffy
- The liver increases by ≥ 2 cm (mark its position on skin with marker pen at onset of rehydration).

Note: It is common for malnourished children to pass many small unformed stools. These must not be confused with profuse watery stools and does not require fluid replacement.

Feeding and rehydration

- Breastfeeding should not be interrupted during rehydration.
- Phase I diet should start immediately when the child is alert.
- If the child has had severe dehydration, feeding should start as soon as the child is alert and the severe dehydration has been treated (2–3 hours).

Rehydration solutions
When no commercial ReSoMal is available, a solution can be made:

To 2 litres of boiled, filtered water add:
1 bag of ORS
50 g of sugar
1 dose of mineral/vitamin mix (6.5 g)

(**Note**: This is double the quantity of water normally used – 2 litres so solution is effectively half strength.)

Emergency treatment of severe dehydration by IV infusion in severe malnutrition
Infusion should be administered only in case of circulatory collapse severe enough to cloud consciousness. **Alert children should never get an infusion.**

The main signs are:
- cold hands and feet.
- weak or absent radial pulse.
- diminished consciousness.

Severe dehydration and septic shock are difficult to differentiate in children with severe malnutrition. They both present with signs of hypovolaemic shock. Points that help to differentiate them are:
- Eyelid retraction associated with a history of diarrhoea is a sign of severe dehydration. The child with septic shock has eyelids that droop
- If the child is unconscious (or asleep) without having the eyelids together, either dehydration or hypoglycaemia (a sign of excess epinephrine) is present
- Superficial veins may be dilated in septic shock. They are always constricted in severe dehydration.

Treatment protocol for life-threatening dehydration
Immediate treatment should be given as follows:
- Give 15 ml/kg IV over 1 hour. The recommended solution is Ringer's lactate solution or 0.9% saline, each with 5% glucose.
- At the same time, insert a nasogastric tube and give ReSoMal 10 ml/kg per hour.
- Monitor carefully for signs of overhydration: check respiratory rate every 15 minutes .

If after 1 hour: the child is improving but still severely dehydrated, continue nasogastric ReSoMal 10 ml/kg/hour for up to 5 hours.

Or if after 1 hour: the child has not improved (radial pulse still weak) assume that it is a septic shock and treat it accordingly; see treatment of septic shock below.

Electrolyte problems

All severely malnourished children have deficiencies of potassium and magnesium that may take two weeks or more to correct. Oedema is partly a result of these deficiencies. **Do not treat oedema with a diuretic.** Excess body sodium exists even though the plasma sodium may be low. **Giving high sodium loads could kill the child**.

Treatment

- Give extra potassium (3–4 mmol/kg daily)
- Give extra magnesium (0.4–0.6 mmol/kg daily)

The extra potassium and magnesium should be added to the feeds during their preparation. See Appendix 7.5 for a recipe to make a combined electrolyte/mineral solution. Add 20 ml of this solution to 1 litre of feed to supply the extra potassium and magnesium required. Alternatively, use commercially available premixed sachets (specially formulated for the malnourished child with minerals already added – do NOT add to commercial preparation).
- When rehydrating, give low sodium rehydration fluid (ReSoMal) (see Appendix 7.5).
- Prepare food without adding salt.

Infections: treatment and prevention

All malnourished children must be presumed to have an infection. Because of the lack of an inflammatory response, clinical signs of infection may be entirely absent in a malnourished child with severe systemic infection.

Routine treatment

All children admitted with severe malnutrition should be given broad spectrum antibiotics.

Protocol of treatment

Specific infections
Children with specific infections should receive the appropriate antibiotic.

No specific infection and no suspected septic shock
The principle is to have a first-line treatment and a second line treatment:
- **First-line treatment** is routinely given at the admission to all severely malnourished children without septic shock, hypothermia, hypoglycaemia or specific infection (skin, eyes).
- **Second-line treatment** is given after 48 hours to children who do not respond to the first treatment line; and to all children with complications.

The choice of the antibiotics used in first line and second-line is based on local resistance patterns, the availability and cost of the drugs. It should be a broad-spectrum antimicrobial, such as amoxicillin/ampicillin (50 mg/kg IV 6 hourly for 2 days then orally 50 mg/kg 6 hourly for 5 days) plus gentamicin 5 mg/kg IV once daily for 7 days. If the child fails to improve after 48 hours add chloramphenicol or ceftriaxone/cefotaxime. Co-trimoxazole is not usually effective in severely malnourished children.

Septic shock: emergency treatment
Septic shock is a very common cause of deaths in these patients. The signs are:
- Clouding of consciousness
- Rapid respiratory rate:
  ≥ 50 breaths/minute for children from 2 to 12 months
  ≥ 40 breaths/minute for children from 12 months to 5 years

- Rapid pulse rate
- Cold hands and feet with visible subcutaneous veins
- Signs of dehydration but without a history of watery diarrhoea
- Hypothermia or hypoglycaemia
- Poor or absent bowel sounds
- An abdominal splash when the child is shaken.

It can be very difficult to distinguish between severe dehydration and septic shock.

Suspected septic shock: treatment
- A broad-spectrum antibiotic treatment (ampicillin + gentamicin) is started immediately.
- Warming the child to prevent or treat hypothermia, (see hypothermia below).
- Feeding and fluid maintenance by nasogastric tube or orally.
- Close monitoring of the vital signs (pulse, respiration, consciousness).

Circulatory collapse
Give IV infusion as described in the case of circulatory collapse due to severe dehydration. But as soon as the radial pulse becomes strong and the child regains consciousness, discontinue the infusion and start the diet orally or by nasogastric tube.

Table 3.16.2 Antibiotic treatment protocol in cases of septic shock

Drugs	Dosage	Route	Days
Amoxycillin	15 mg/kg, Four times daily	IV but IM if IV not possible	1, 2
Amoxycillin	15 mg/kg, Three times daily	oral	3, 4, 5, 6, 7
Gentamicin	5 mg/kg Once daily	IV but IM if IV not possible	1, 2, 3, 4, 5, 6, 7

Ceftriaxone can be also used at 75 mg/kg once daily IV or IM if IV is not possible. Alternatively, cefotaxime 50–100 mg/kg 12 hourly IV or IM if IV is not possible. Some units routinely give metronidazole 7.5 mg/kg orally 8 hourly for 7 days in addition to the above.

Hypothermia: prevention and treatment

Malnourished children have a low metabolic rate. The thermoneutral temperature is 28–32 °C. At 24 °C they can become hypothermic. Those with infection or extensive skin lesions are at particular risk. A hypothermic malnourished child should always be assumed to have septicaemia.

Signs

The signs of hypothermia are a temperature: rectal or oral <35.5 °C (with low reading thermometer). If axillary temperature is <35 °C or does not register, assume hypothermia.

Routine prevention

- Cover all children with clothes and blankets. Put on a warm hat (most heat is lost from the head).
- Ensure that mothers sleep with their children. **Do not leave a child alone in bed at night.** ✔
- Keep the ward doors closed during the night.
- Avoid wet nappies, clothes or bedding.
- Do not wash very ill children. Others can be washed quickly ideally with warm water and dried immediately.
- Make sure that the child is fed so that metabolic heat can be produced. Ensure feeds occur during the night. **Avoid medical examinations which leave the child cold.**

Emergency treatment of hypothermia

Immediately place the child on the caretaker's bare chest or abdomen (skin to skin) and cover both of them. Give the mother a hot drink to increase her skin blood flow.

If no adult is available clothe the child very well (including the head) and put near a lamp.

Immediately treat for hypoglycaemia (see below) and then start normal feeds.

Give broad spectrum antibiotics.

Monitor the rectal temperature every 60 minutes until the temperature is normal (>36.5°C).

Hypoglycaemia: prevention and treatment

Severely malnourished children easily develop hypoglycaemia. It is associated with serious infection. If available blood glucose is <3.0 mmol/litre. If not measurable, assume hypoglycaemia is present.

Signs

The main signs of hypoglycaemia are:
- Lethargy, limpness, loss of consciousness or convulsions
- Drowsiness/unconsciousness with the eyelids partly open, or retraction of the eyelids
- Low body temperature (<36.5 °C).

Sweating and pallor do not usually occur. ✔

Routine prevention

- Frequent small feeds, (day and night).
- Feeding should start while the child is being admitted.
- Treatment of infections.

Emergency treatment

If hypoglycaemia is suspected:
- **If the child can drink** give therapeutic milk or 50 ml of glucose 10%, or 50 ml of drinking water plus 10 g of

sugar (1 teaspoon of sugar in 3.5 tablespoons of clean water). Follow this with the first feed as soon as possible. If achievable, divide first feed into 4 and give half-hourly. If not, give whole feeds every 2 hours during the day and night for at least the first day.

- **If the child is unconscious or has convulsion** give 5 ml/kg of body weight of glucose 10 % IV and/or if IV is not possible give 5 ml/kg of glucose 10 % by nasogastric tube.

Continue frequent feeding to avoid a recurrence.

Give broad-spectrum antibiotics.

If there are convulsions other causes must be excluded – cerebral malaria, meningitis/encephalitis, thiamine deficiency, hypernatraemic/hyponatraemic dehydration (especially in hot dry climates).

If blood glucose available and is low, repeat finger or heel prick after 60 minutes.

Congestive heart failure

This is a common and dangerous complication that usually occurs several days after admission. The heart muscle is atrophic (effectively there is a cardiomyopathy). During early recovery from severe malnutrition, sodium can be mobilised from the tissues before the kidney recovers sufficiently to excrete the excess. All blood transfusions must be done as soon as possible (in the first two days after admission).

Heart failure is usually caused by inappropriate treatment:

- Misdiagnosis of dehydration with consequent inappropriate "rehydration"
- Very severe anaemia
- Overload due to blood transfusion (consider an exchange transfusion, see Chapter 6.9)
- A high sodium diet, using conventional ORS, or excess ReSoMal
- Inappropriate treatment of "refeeding diarrhoea" with rehydration solutions.

Signs

Excess weight gain is the most reliable sign – daily weights should be taken on all malnourished children. Differentiate pneumonia and heart failure by weighing the child: if weight has risen, particularly if by more than 5% then consider heart failure; if weight is lost, then consider pneumonia.

- **First sign:** fast breathing
  ≥50 breaths/minute for children from 2 to 12 months
  ≥40 breaths/minute for children from 12 months to 5 years
- ***Later signs***
  Lung crepitations
  Respiratory distress
  Rapid pulse rate
  Engorgement of the jugular vein
  Cold hands and feet
  Cyanosis of fingertips or hypoxaemia diagnosed by pulse oximetry if available (SaO_2 < 94% in air at sea level)
  Enlarged liver >2 cm from baseline (see above)

Emergency treatment of congestive cardiac failure

- Stop all oral intake and IV fluid.
- The treatment of heart failure takes precedence over feeding of the child.
- No fluid at all should be given until the cardiac function improves, even if it takes 24–48 hours.
- Give a diuretic IV, usually furosemide (1 mg/kg). This is the only situation in which diuretics should be used: **diuretics should never be given to reduce oedema in malnourished children.** ✔
- If potassium intake has been assured (F100 has adequate potassium) then use half the conventional doses of digoxin ORALLY for 24 hours only. This slows the sodium pump and allows the sodium to re-enter the cells (give 20 micrograms/kg single loading dose).

Measles: prevention and treatment

Measles is especially dangerous in severe malnutrition. ✔

Routine prevention

All children >6 months admitted with malnutrition should be vaccinated against measles on admission. A second dose of vaccine in a previously immunised child is not harmful. A second dose should be given at discharge where the prior vaccination state is uncertain or the child was not vaccinated before admission.

Treatment of measles

If a case of measles is admitted:

- Isolate the individual and any suspected cases
- Review the vaccination status of all patients in the ward
- Give two doses of vitamin A separated by one day
- Treat for measles (see Chapter 4.19) as well as malnutrition.

Micronutrient deficiencies

All will have these. Commercial F100 contains all micronutrients in the correct amounts. Give daily multivitamin supplement: zinc 2 mg/kg/day; copper 0.3 mg/kg/day. Zinc and copper can be combined with potassium and magnesium to make an electrolyte/mineral solution which is added to ReSoMal and to feeds. Premixed sachets are available but AVOID iron during the first 2 weeks until the ✔

child is gaining weight. In goitrous regions, potassium iodide should be added to the mineral mixture (12 mg/2500 ml) or the child given Lugol's iodine 5–10 drops per day.

Vitamin A: prevention and treatment

Routine preventive treatment

Oral vitamin A is particularly important for the severely malnourished child and one dose should be given routinely to each child admitted with malnutrition.

Table 3.16.3 Vitamin A dosage – preventive treatment

Weight	Dose at admission
<6 kg	50 000 IU once
6–10 kg	100 000 IU once
>10 kg	200 000 IU once

Treatment of xerophthalmia

If a child shows signs of vitamin A deficiency (xerophthalmia) or has measles, three doses of vitamin A treatment should be given.

Table 3.16.4 Vitamin A dosage – xerophthalmia

Weight	Dosage		
	Day 1	Day 2	Day 14
<6 kg	50 000 IU	50 000 IU	50 000 IU
6–10 kg	100 000 IU	100 000 IU	100 000 IU
>10 kg	200 000 IU	200 000 IU	200 000 IU

If the eyes show signs of inflammation or ulceration, give the following additional care to the affected eye(s) to prevent corneal rupture and extrusion of the lens:

- Instil chloramphenicol or tetracycline eye drops, 2–3 hourly as required for 7–10 days
- Instil atropine eye drops, 1 drop 3 times daily for 3–5 days
- Cover with sterile saline-soaked eye pads
- Bandage eye(s).

✔ **Note:** Children with vitamin A deficiency are likely to be photophobic and have their eyes closed. It is important to examine their eyes **very gently** to prevent corneal rupture.

Iron deficiency and anaemia treatment

The majority of malnourished children have anaemia. This is due to the many deficiencies they have (folic acid, riboflavin, pyridoxine, ascorbic acid, vitamin E, copper) and their inability to metabolise iron. **Iron should not be given for about 2 weeks after the start of treatment.** ✔

Routine treatment

Folic acid

Give 5 mg of folic acid on the day of admission, then 1 mg/day thereafter (in F100 already).

Iron

Iron should never been given during the Phase I or the transition phase. In malnourished patients, iron is not properly metabolised and used and is dangerous. The free iron enhances the production of free radicals that can damage cell walls. Excess free iron encourages systemic infection.

Oral iron supplement should start 14 days after admission. This is best added to the diet at a dose of one crushed tablet of ferrous sulphate (200 mg) to 2 litres of therapeutic milk or ferrous sulphate 3 mg/kg/day.

Emergency treatment of very severe anaemia

Blood transfusion in malnourished children is potentially dangerous because it can precipitate heart failure. The only two indications to consider blood transfusion are:

- A child with a Hb <4 g/100 ml.
- A child with signs of heart failure due to anaemia (at immediate risk of death).

10 ml per kg body weight of packed cells (or whole blood) are given slowly by partial EXCHANGE transfusion; ideally, and if this can be achieved, by the use of a carefully and continuously observed cannula in an artery or central vein; possible also in a vein in the antecubital fossa. 2.5 ml/kg of anaemic blood is first removed and then when 5 ml/kg of appropriately screened and cross-matched blood has been transfused, 2.5 ml/kg is again taken and the cycle repeated. The child is closely monitored for signs of congestive heart failure.

If partial exchange is not possible and heart failure is present, give 10 mg/kg ideally as packed cells otherwise as whole blood. Transfuse over 4 hours and give IV furosemide 1 mg/kg at the start of the transfusion. Monitor carefully for worsening heart failure.

Try not to transfuse again until at least 4 days have passed.

Intestinal parasites

Routine treatment

Routine deworming is given to all children over 1 year but only in Phase II or the transition phase.

For children over 1 year of age, mebendazole (1 tablet = 100 mg): single dose, 500 mg; or if dose given over 3 days, 100 mg twice daily.

In areas with widespread hookworm the 3-day scheme is preferable, otherwise the single-dose treatment of 500 mg is much easier.

Dermatosis of kwashiorkor

Shedding of the skin in scales or sheets, desquamation, exfoliation, cracking of the skin surface, ulceration of genital or peri-anal areas are all common.

There can be widespread weeping skin lesions like burns.

Zinc deficiency is usual in this situation and oral zinc supplements improve the skin (2 mg/kg/day of elemental zinc).

Treatment

- Leave the exposed area open to dry during the day.
- Apply barrier cream (zinc and castor oil ointment) or petroleum jelly or tulle gras to the raw areas and gentian violet or nystatin cream to the skin sores two times per day.
- These children should be on broad-spectrum antibiotics.
- Do not use plastic pants or disposable nappies for these children.

Continuing diarrhoea

Diarrhoea should subside during the first week of treatment. In the rehabilitation phase, loose or poorly formed stools are normal and do not need treatment providing that weight is increasing.

Treatment

Giardiasis

Giardiasis and mucosal damage are common causes of continuing diarrhoea. Where possible, examine the stools by microscopy.

If cysts or trophozoites of *Giardia lamblia* are found, give metronidazole (5 mg/kg 8 hourly for 7 days).

Lactose intolerance

Diarrhoea is only rarely due to lactose intolerance. Only treat for lactose intolerance if the continuing diarrhoea is preventing general improvement. Starter F75 is a low-lactose feed. In exceptional cases:

- Substitute milk feeds with yoghurt or a lactose-free infant formula
- Reintroduce milk feeds gradually in the rehabilitation phase.

Osmotic diarrhoea

This may be suspected if the diarrhoea worsens substantially with hyperosmolar F75 and ceases when the sugar content and osmolarity are reduced. In these cases:

- Use a lower osmolar cereal-based starter F75 (see Appendix 7.5 for recipe) or, if available, use a commercially prepared isotonic starter F75
- Introduce catch-up F100 gradually.

Malaria: treatment and prevention

In endemic areas, all malnourished children should have a rapid malaria smear on admission. Where this is not possible, all malnourished children should receive anti-malarial treatment routine for the area. The parasitaemia is usually much lower than in normal children. In initially negative children, there can be a recrudescence during nutritional replacement treatment.

Treatment

See Chapter 4.27.

If practical and in endemic areas, children should sleep under impregnated nets in the wards.

Tuberculosis

In patients treated for malnutrition, tuberculosis can be a cause of failure to gain weight. In malnourished children the diagnostic of tuberculosis is particularly difficult and misdiagnosis is common (see Chapter 4.10).

How to diagnose pulmonary TB

The signs of TB in malnourished children are often not specific (anorexia, failure-to-thrive). Asymmetric chest signs or asymmetric lymph nodes are usually TB. Pneumonia in malnourished children affects both lungs and HIV gives symmetrical lymphadenopathy (see Chapter 4.18).

Sputum is rarely available. The Mantoux test can be negative in malnutrition. Undertake a chest *X* ray if possible.

Treatment

See Chapter 4.10.

Children with TB children should not be isolated, for the following reasons:

- Children are generally not a source of transmission (no expectoration)
- Treatment quickly eliminates the risk of transmission
- An isolated child is stigmatised and neglected in resource-poor settings.

Usually paediatric TB is acquired from a sputum-positive adult in the household, so the caretaker is a much higher infection risk. He/she should have a chest *X* ray.

Malnutrition and AIDS

Basically, nutritional treatment of suspected HIV-positive patients, **is the same as for any other severely malnourished patient**. They require the same dietary and medical treatment. HIV-positive patients usually respond well to the nutritional treatment and gain weight. Testing of suspected cases is not useful and not recommended. Patients can be stigmatised and rejected if the staff or other patients know their status.

Dietary treatment of severe malnutrition

Dietary treatment in Phase I

Objectives
Progressive restoration of the electrolyte, metabolic and physiological balance by the frequent feeding of a special formula milk.

Principles
Severely malnourished children are usually anorexic, have thin bowel walls, damaged metabolism, and too much salt in their bodies. Initially they require a low salt and low protein diet and are unable to tolerate large amounts of food because their **capacity is** reduced. Initially a diet high in carbohydrate with low levels of sodium and iron and very modest protein is given. The diet leads to restoration of the metabolic and physiological function, but is insufficient for weight gain. Feeding should:

- Start quickly after admission
- Be divided into many small meals to stay within the absorptive and metabolic capacity and to prevent hypoglycaemia and hypothermia
- The child should be encouraged to eat but not be forced. Feeding a malnourished child requires time and patience. Use a cup or a bowl or a spoon or syringe to feed very weak children. If the child takes less than 70% of the prescribed diet, he or she should be fed by a nasogastric tube
- Always continue breastfeeding, and encourage the mother to breastfeed. After the breast feed give the scheduled amounts of starter formula first (see below).

Also:

- Frequent small feeds of low osmolality and low in lactose
- Night feeds are essential
- Oral or nasogastric feeds (never parenteral preparations)
- 100 kcal/kg/day
- Protein: 1–1.5 g/kg/day
- Liquid: 130 ml/kg/day to all children, no matter whether or not oedema is present.

Table 3.16.5 A recommended schedule

Days	Frequency	Volume/kg/feed	Volume/kg/day
1–2	2-hourly	11 ml	130 ml
3–5	3-hourly	16 ml	130 ml
6 onwards	4-hourly	22 ml	130 ml

What food to give
There is a special milk for Phase I called F75: if it is in short supply it can be kept for specially ill children. If it is not available then F100 should be diluted to the same strength as F75 and given in its place.

F75 has:

- 75 kcal/100 ml
- 0.9 g of protein/100 ml (around 5% kcal provided by protein)
- 2 g of fat/100 ml (around 32% of kcal provided by fat)
- 13 g of carbohydrate/100 ml (around 62% of kcal provided by carbohydrates)

Which quantity of food to give
100 kcal/kg/day. The daily amount of kcal should be divided by the number of meals given during the day, usually eight meals per day.

F75: 133 ml = 100 kcal.

Example

One child of 6 kg should receive a diet of 100 kcal/kg/day. The child will get eight meals of F75.

Number of kcal/day : 100 kcal × 6 kg = 600 kcal
Quantity of F75 per day: 800 ml (798 exactly)
Quantity per meal: 800/8 = 100 ml

Do not exceed 100 kcal/kg/day in this initial phase. Diarrhoea should gradually decrease and oedematous children should **lose weight** as the oedema goes. If diarrhoea continues see above.

Table 3.16.6 Home-made Phase I diet

Food item	Quantity
Dried skimmed milk (DSM)	25 g
OR boiled full-cream milk	300 ml
Vegetable oil	27 g (30 ml)
Sugar	105 g
Water (boiled)	Add water to make 1 litre of preparation
CMV (minerals and vitamin mix)	20 ml (should be added after the water)

Note: F75 is much better since it contains maltodextrin instead of sugar and therefore a lower osmolality and is less likely to cause osmotic diarrhoea. Alternatively 35 g/litre of starch can be added and the sugar reduced to 70 g/litre.

Dietary treatment in the transition phase

What food to give
In the transition phase full strength F100 is given in the same volume that was calculated for F75 in Phase I. There is no other change made in the transition phase.

F100 contains:

- 100 kcal/100 ml
- around 2.6 g of protein/100 ml (10% of kcal provided by protein)
- around 5.6 g of fat/100 ml (50% of kcal provided by fat)
- around 9.8 g of carbohydrate (40% of kcal provided by carbohydrate).

There are two forms of F100:

- **Commercial F100**

This therapeutic milk is prepared in a sachet. All that the nurse has to do is open the packet and dilute the contents into 2 litres of potable (boiled) water. The commercial F100 has a low osmolarity to reduce "refeeding" diarrhoea in the severely malnourished children.

Preparation of therapeutic milk
1 bag of 456 g of therapeutic milk is diluted in 2 litres of boiled water. This gives 2.4 litres of therapeutic milk.

- **Home-made F100**
  When commercial milk is not available, it can be prepared from the recipe in Table 3.16.7.

Dietary treatment in Phase II

Objectives
Catch-up growth with a rapid weight gain of the child (10–20 g/kg/day).

Usually the appetite has returned.

Principles
The child has re-established his/her physiological balance and should get enough food to gain weight as quickly as possible. The child is given a high energy diet with a normal protein content.

- The amount of intake is increased in quantity (to about 200 kcal/kg/day)
- Reduce meal frequency from eight to six meals per day.
- No limit in the quantity of food given. The child is allowed to eat as much as he/she wants but never forced to eat.
- Breastfeeding continues. Breast milk must always be offered BEFORE the high energy food is given.
- Aim for weight gain >10 g/kg/day
- Remain alert for heart failure

Table 3.16.7 Recipe to prepare 1 litre of home-made F100

Food item	Quantity
Dried skimmed milk (DSM)	80 g
Vegetable oil	60 g
Sugar	50 g
Water (boiled)	Add water to make 1 litre of preparation
CMV (minerals and vitamin mix)**	20ml (should be added after the water)

** The CMV should be added when the preparation of milk is ready.
Whisk to prevent oil from separating.
This keeps for 6 hours.

What food to give
The basic diet is composed of F100 meals. However, when the child is gaining weight quickly other foods can be introduced, for example:

- Enriched porridges (1 ml contains 1 kcal/g)
- Enriched biscuits (useful for overnight feeding if Phase II is conducted in a day-care centre)
- Local meal: composed of the usual food eaten in the area – this should be enriched in the pot with the addition of oil and CMV and sometimes DSM.

Quantity of food to give
200 kcal/kg of F100/kg of body weight per day is dispensed and offered.
Example of calculation
A child of 9 kg should receive per day: 200 kcal × 9 = 1800 kcal. The child will receive six meals per day: each meal should provide: 1800 kcal/6 = 300 kcal per meal.

The diet is composed of six meals of F100. The enriched porridge or family meal is given in ADDITION if the patient wishes to take it.

- F100 (1 ml of F100 = 1 kcal): the child should receive 300 ml of F100 per meal.

Older children and adolescents, when they are gaining weight rapidly often do not want the milk and demand "solid food". This usually slows the rate of recovery. The solid food should always be enriched.

In developing local recipies the weight gain should be compared with children taking F100 alone. If the weight gain is similar, then the recipe for the porridge is adequate.

There are several commercial preparations of solid ready-to-eat F100 under development.

Individual child monitoring

In Phase I

A daily medical and nutritional round of all the children in Phase I should be done. The children should be carefully monitored, every day, for:

- Oedema
- Weight
- Appetite: how the child is eating and the quantity eaten

Table 3.16.8 Alternative formulae where dried skim milk is not available

		F100 DSM	F100 DWM	F100 COW	F100 BUFFALO	F100 EvapMilk	F100 GOAT	F100 EWE's milk
Milk	G/ml	80	110	900	750	350	900	550
Sugar	G	50	50	50	60	50	50	50
OIL	G	60	30	25	10	30	30	30
CMV	ml	20	0	0	0	0	0	0

DSM = dried skim milk
DWM = dried whole milk
Cow = fresh cow's milk (full cream)
Buffalo/goat/ewe = fresh milk from buffalo, goat or sheep
EvapMilk = tinned evaporated milk (sugar free).

- Clinical state: consciousness, diarrhoea, vomiting, skin, etc.
- Behaviour: apathetic, alert, crying, etc.
- Temperature
- Liver size, heart rate, heart sounds.

This information should be recorded every day on an individual chart.

When to pass to transition phase
Children stay usually from 1 to 7 days in Phase I. The child can pass to the transition phase when:
- He/she regains appetite
- He/she is lively and interested
- Serious medical complications are under control
- Oedema is decreasing (but may be still present).

If after 5–7 days the child is not ready for transition phase then the child should be completely re-examined and investigated.

After 4 days in transition phase without experiencing any problem, the child is ready to pass to Phase II. Oedema should be completely clear before progressing to Phase II.

In Phase II

The monitoring in Phase II includes:
- A daily round by the nurse who checks the general state of the child. Is there oedema? How is the child eating? Is there nausea or vomiting?
- A physician round one or two times a week
- Measurement of weight every 2 days, or two times a week
- Measurement of height taken once a month.

The information should be recorded on the individual chart.

✔ **If a child develops a complication in Phase II such as refeeding diarrhoea or vomiting that requires passage of a nasogastric tube, rehydration solutions, transfusion, etc. the child is returned to Phase I and subsequently the transition phase again – the above treatments must never be given to children while in Phase II and taking very large amounts of F100 diet.**

When the child can be discharged
Children stay in Phase II until they reach the criteria of recovery. The average total length of stay is usually around 4 weeks; longer with very severe complicated malnutrition.

When the child has reached his/her target weight and is in good clinical state, he/she will be either referred to a supplementary feeding programme or sent directly home with arrangements made for follow-up.

Failure to gain weight
If the child fails to gain weight he/she should be investigated. Weight gain is defined as: poor if <5 g/kg/day; moderate if 5–10 g/kg/day and good if >10 g/kg/day. The reasons are usually:

- Food prescription or food preparation (kitchen) are wrong and the child has not received the right quantity of the right food
- The child does not eat the amount of food prescribed (dislikes the food, food is being eaten by other persons)
- Suspect hidden acute infections (urinary tract infection, acute respiratory infection, otitis media, mouth candidiasis, giardiasis, etc.)
- There are chronic hidden infections (tuberculosis, HIV)
- Re-examine, do stool and urine microscopy and chest *X* ray
- Look for poor feeding techniques, check that night feeds are occurring.

Emotional and physical stimulation

The severely malnourished child is nearly always psychosocially deprived. The illness itself makes the child unresponsive. He/she does not cry or complain. Because mothers use a cry as the signal to give attention, these children do not get the attention they need to stimulate them. The neglect is not wilful on the part of the mother at all; rather it is a failure of the two-way communication between the mother and her child.

Because they do not cry or complain, nurses and staff often neglect these children as well – this greatly compounds the problems associated with being in a strange environment. It is essential to stimulate these children – particularly the unresponsive children. The ward should be made as much like home as possible and children should sleep with their mothers. This is usually on the floor. This is much better than in a small barred cage (called a cot) where the child is isolated and cold.

- **In Phase I**
  It is essential that mother (or other caretaker) is present, feeds the child, comforts him, holds him, plays with him, talks and sings to him.
- **In Phase II**
  It is important to stimulate the child to move, to play with other children. A play area should always be present.

The daily organisation of the activities

To organise the treatment of malnourished children a schedule of activities, (care, distribution of meals), must be established. An example is given below.

Table 13.6.9 Daily organisation of activities

Hour	For children in Phase I and transition	For children in Phase II (day care)
2	Milk distribution	
5	Milk distribution	
7	Team changeover (day shift)	

7.30	Temperature	Arrival of children
8	Milk + drugs distribution	Milk distribution + drugs
9	Weight, oedema	Weight, oedema
9.30	Mother's meal	Medical round
10	Medical round	Milk distribution
11	Milk distribution	Mother's meal
12		Milk distribution + drugs
13	Dressing care	Dressing care
14	Milk distribution + drugs	
15		Porridge distribution
16	Mother's meal	Mother's meal
17	Milk distribution	Milk distribution
18	Quick medical round	Departure to home with one porridge and enriched biscuits for the night
19	Team changeover (night shift)	
20	Milk distribution + drugs	
21	Closing of windows and wrapping of child	
23	Milk distribution	

Inappropriate practices

- Too much sodium, energy and protein given during the Phase I of treatment
- No distinction made between Phase I and II
- Failure to monitor food intake
- Lack of feeding at night
- Lack of blankets and hats
- No daily schedule organised
- Diuretic given to treat oedema
- Anaemia treated from time of admission with iron supplements
- Intravenous fluids given for other indications than circulatory collapse
- Use of high sodium diet and standard oral rehydration solution
- Routine antibiotics not given
- No vitamin A or measles vaccine given.

Problems with malnutrition

A high level of care is needed

The treatment of a severely malnourished child, requires intensive care, like a premature neonate, with close monitoring, complex medical care (severe or chronic infections), a diet well enriched in nutrients (F100 etc.), and emotionally stimulating rich and physically warm environment.

Limited resources

The resources are almost always limited. The limited financial resources lead to difficulty in obtaining therapeutic milks and other fortified food, drugs and materials. However, analysis shows that these are not the main reasons for death. Inappropriate ORS, and blood transfusion (expensive) are given – the cost of these treatments would more than pay for the diets. The major cause of death and relapse is premature discharge, before the child has recovered and with no follow up or community service. Inappropriate diets may be cheap, but they prolong the stay of the patient and time to recovery. The main cost is being in hospital itself: if a child can be discharged two weeks earlier with F100 than with family meals, then this is a much cheaper option. The recording charts, weight charts and proforma are tools that help in the management of these children in a way that is not possible without such aids. However, perhaps the greatest problem is the limited human resource on the malnutrition ward, with an insufficient number of skilled personnel and constant movement of staff as soon as they are trained. The presence of demotivated personnel (often very underpaid and undervalued), who have undergone insufficient training in the very special needs of the severely malnourished and how such children are quite different from a normal child that becomes ill represent the greatest challenge.

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3.17 Acute upper airway obstruction

Christiane Ronald and David Southall

Diagnosis

Obstruction of the upper airway (larynx and trachea) is potentially life threatening. The cardinal feature is stridor, heard mainly during inspiration. **Like the wheeze in asthma, the loudness of the stridor does not indicate the severity of the obstruction.** There may also be hoarseness and a barking or seal-like cough. The severity of the obstruction is best assessed by the degree of sternal and subcostal recession, and respiratory and heart rate. Increasing agitation or drowsiness, or central cyanosis indicates severe hypoxaemia and hypercapnia and the need for urgent intervention.

Differential diagnosis

Most cases of upper airway obstruction in children are the result of infection.

Croup

Minimum standards requirements

- Oxygen
- Dexamethasone orally
- Nebulised budesonide and epinephrine
- Antibiotics: chloramphenicol/cefuroxime
- Intubation/tracheostomy

Croup is defined as an acute clinical syndrome with inspiratory stridor, barking cough, hoarseness and variable degrees of respiratory distress. This definition embraces

Table 3.17.1 Differential diagnosis of upper airway obstruction

Collapse of airway due to loss of muscle tone and build up of secretions due to poor cough reflex	Airway swellings	Space occupying lesion or structural abnormality
Depressed conscious level, drug intoxication, bulbar palsy and myopathy	Infective ● Upper respiratory tract infection in an infant ● Viral croup (laryngotracheobronchitis) ● Bacterial tracheitis ● Epiglottitis ● Severe tonsillitis for example in infectious mononucleosis ● Diphtheria Non-infective ● Recurrent croup ● Anaphylaxis ● Adenoidal hypertrophy	Can be intranasal, pharyngeal, laryngeal or in upper trachea ● Foreign body ● Retropharyngeal abscesses ● Tumour ● Extrinsic haematoma causing airway compression for example post-thyroidectomy ● Congenital pharyngeal, laryngeal or upper tracheal abnormalities for example: choanal atresia, laryngomalacia, subglottic stenosis, laryngeal web

several distinct disorders:

- **Acute viral laryngotracheobronchitis (viral croup)** is the commonest (>95%) of laryngotracheal infections. Peak incidence is the second year; most hospital admissions are between 6 months and 5 years. The stridor is usually preceded by fever (<38.5 °C) and coryza and symptoms tend to be worse at night. If narrowing is minor, the stridor will be present only when the child hyperventilates or is upset. As the narrowing progresses, the stridor becomes both inspiratory and expiratory and is present even with the child at rest. Particularly children under 3 years may develop features of increasing obstruction and hypoxaemia with marked sternal and subcostal recession, tachycardia and agitation. If the infection extends distally to the bronchi, wheeze may also be audible.
- **Recurrent or spasmodic croup:** Some children have repeated episodes of croup without preceding fever and coryza. The symptoms are of sudden onset at night, and often persist for only a few hours. It is associated with atopic disease (asthma, eczema, hay fever). The episodes can be severe, but are more commonly self-limiting.
- **Bacterial tracheitis or pseudomembranous croup,** is dangerous. It is one of the important complications of measles. Infection of the tracheal mucosa with *Streptococcus pneumoniae*, *Staphyloccocus aureus* or *Haemophilus influenzae B* results in copious, purulent secretions and mucosal necrosis. The child appears toxic with a high fever, with marked signs of respiratory obstruction. In the UK over 80% of these children need intubation and ventilatory support to maintain an adequate airway. The croupy cough and the absence of drooling help distinguish this condition from epiglottitis. Clinical and radiological signs of segmental collapse and consolidation related to bronchial occlusion are usual. The cough is often persistent and ineffective in clearing the secretions, the illness has a prolonged course, the restoration of normal mucosa usually takes several weeks.

Emergency treatment of croup

- These children (and often their mothers) can be very frightened. **Don't frighten them more by sticking things in their throat or painful injections**. Crying increases their oxygen demand and may increase laryngeal obstruction. Keep them on their parent's lap and **explain** the condition and the treatment. Tell the mother to alert nurses or doctors if the child breathes more quickly or has marked sternal recession. **These are danger signs for hypoxaemia**. ✔
- Ensure adequate oral fluid intake.
- Many children admitted to hospital have hypoxaemia. Humidified oxygen should be given through nasal

Table 3.17.2 Severity of upper airway obstruction

Severity	Mild	Moderate	Severe
Upper airway noise	Hoarse voice, barking cough, mild stridor	More severe stridor	Stridor may reduce as exhaustion occurs Stridor associated with epiglottitis may be soft as the obstruction is above rather than in the larynx. Drooling can be marked due to inability to swallow
Work of breathing (effort)	Mild increase in effort with some intercostal recession	Moderate increase in effort with nasal flare, tracheal tug and accessory muscle usage	Huge increase in work of breathing to the point of exhaustion where breathing efforts will decrease to respiratory arrest
Efficacy of breathing – conscious level and colour	Child alert, not distressed by increase in effort required No cyanosis. SaO_2 may be normal	Child distressed by breathing efforts. Cyanosis may not be visible but SaO_2 reduced	Conscious level reduced to the point of respiratory collapse due to exhaustion
Cardiovascular effects	Mild increase in heart rate	Continuing rising heart rate	Severe tachycardia to the point of collapse due to exhaustion, respiratory arrest and subsequent hypoxic cardiac arrest

- Mild croup does not need any specific treatment.
- Moderate croup may require hospital admission.
- ✔ **Severe croup should be admitted and intensive care may be necessary.**

cannulae or a facemask held just in front of the child's face by the parent. **Do not use nasopharyngeal catheters that can precipitate dangerous paroxysms of coughing**. Milder cases of croup should not routinely get oxygen as it can frighten the child or may even mask cyanosis.

- Croup can be a very painful condition, even if the child does not have a high temperature, prescribe regular paracetamol.
- There is good evidence that steroids help. Children with mild, moderate or severe croup all benefit from steroids. Give 0.6 mg/kg dexamethasone once or twice a day, this should be given orally, as it works as well as injected. If the child vomits, repeat it or give the same dose intramuscularly. An expensive and equally effective treatment is nebulised budesonide 2 mg in 2 ml; it may be repeated 30–60 minutes later.
- **Children with signs of severe obstruction should be given nebulised epinephrine (5 ml of 1 in 1000) ideally with oxygen through a facemask**. This produces a transient improvement for 30–60 minutes but rarely alters the course of the illness. Arrange for the child to be seen **quickly** by an anaesthetist. Monitor the oxygen saturation with a pulse oximeter. If effective, repeat every 2 hours as required.
- A few children need intubation. This should be done under general anaesthetic. **If there is doubt about the diagnosis or difficulty in intubation is anticipated, an ENT surgeon capable of performing a tracheostomy should be present**. In intubated children, 1 mg/kg prednisolone every 12 hours reduces the duration of intubation.
- Severely ill or toxic children and those with measles should receive an antibiotic effective against *Strep. pneumoniae*, *H. influenzae* and *Staph. aureus*. If available, cefuroxime 150 mg/kg/day in 4 doses IV or cephalexin 25 mg/kg 6 hourly orally is a good choice. An alternative is chloramphenicol 2.5 mg/kg IV or orally 6 hourly.

Inhaled foreign body
(see Chapter 3.1)

Suspect the diagnosis if sudden onset of cough and stridor in a well child. Direct question to parents and child, regarding peanuts or other possibly inhaled object.

Assessment

- Usually non-progressive stridor.
- Look for mediastinal shift, asymmetrical breathsounds.

Investigations

- Lateral *X* ray neck.
- Inspiratory/expiratory chest *X* ray or bilateral lateral decubitus chest *X* ray for infants and small children.
- 80% inhaled objects are non-radiopaque.

Management

Call ENT surgeon for laryngoscopy. Laryngeal foreign body may present very acutely with cyanosis or loss of consciousness. Therefore urgent direct laryngoscopy and/or tracheostomy or cricothyroidotomy (see Chapter 6.14) may be necessary.

Acute epiglottitis

Minimum standards requirements

- Immunisation against *H. influenzae*
- Antibiotics: chloramphenicol/cefotaxime/ceftriaxone/cefuroxime
- Intubation/tracheostomy

This is a severe infection caused by *H. influenzae*. Peak incidence is 2–3 years. It is less common than croup but important as **the diagnosis needs to be made fast as rapid progression of stridor in the ill, toxic child may be fatal within hours if not promptly treated**. Cough is not a prominent feature and the stridor has a soft quality with often an expiratory component. The child tends to drool and assume an upright posture.

Do not
- Examine the throat.
- Lie the child down.
- *X* ray neck.
- Perform invasive procedures.
- Use a nasopharyngeal tube for oxygen.

Do
- Use reassurance to calm the child.
- Attach pulse oximeter and give warm humidified oxygen if $SaO_2 < 95\%$ (use mask held, close to but not on face, by the mother)
- Call anaesthetist and ENT surgeon.
- Arrange examination under anaesthesia.
- ***Refer to intensive care unit accompanied by anaesthetist.***

Management

- Elective intubation under general anaesthetic is the treatment of choice. The diagnosis is confirmed by laryngoscopy under general anaesthetic just prior to intubation ("cherry-red epiglottis"). An ENT surgeon must be present.
- Whilst anaesthetised, the following procedures should be done:
  Blood cultures, throat swab, intravenous line.
- Recommended antibiotic therapy:
  Chloramphenicol 50 mg/kg immediately then 25 mg/kg 6 hourly. If available, cefuroxime, or cefotaxime 50 mg/kg 6 hourly or ceftriaxone 50 mg/kg once daily should be effective.

- Following intubation the child will be able to breathe humidified air spontaneously ideally with CPAP (see Chapter 1.26). Sedation (discuss with anaesthetist) may be required in order to prevent self extubation but the child will then usually require assisted ventilation. Most children will be ready for extubation after 48 hours.

An alternative is to fix the child's arms to their thorax using a bandage.

Table 3.17.3 Contrasting features of croup and epiglottitis

Feature	Croup	Epiglottitis
Onset	Over days	Over hours
Preceding coryza	Yes	No
Cough	Severe, barking	Absent or slight
Able to drink	Yes	No
Drooling saliva	No	Yes
Appearance	Unwell	Toxic, very ill
Fever	$<38.5°C$	$>38.5°C$
Stridor	Harsh, rasping	Soft
Voice muffled	Hoarse	Reluctant to speak,
Need for intubation	1%	80%

Angioneurotic oedema (see Chapter 1.23)

There are usually areas of painless swelling obvious in other parts of the body. Eyes, lips and tongue are particularly likely to be affected. Stridor is caused by laryngeal oedema.

Management

- Give epinephrine 10 micrograms/kg IM
- Epinephrine 5 ml of 1 in 1000 nebulised with 100% oxygen
- Give 100% oxygen
- Hydrocortisone 4 mg/kg IV over 15 minutes or IM
- Chlorpheniramine 250 micrograms/kg IV or oral
- 0.9% saline or 4.5% albumin (if available) 10–20 ml/kg, if shocked.
- Intubation or even tracheostomy may be required (contact ENT team).

Laryngeal burns (see Chapter 5.10)

✔ Due to inhalation of hot gases or toxic vapours. May be associated with extensive skin burns. **Be aware that airway obstruction may develop even if not obvious on first assessment.**

Management

- ***Admit to ICU*** (if available).
- Hydrocortisone 4 mg/kg IV 6 hourly.
- 4.5% albumin for shock as required.
- Intubation/tracheostomy if indicated by assessment.

Diphtheria (see Chapter 4.3)

Gradual onset of stridor in a child (usually 2–3 years old) with neck oedema and ulcerating lesions of the tonsillar bed forming a membrane. Bleeding may occur at the site. Diagnosis may be confirmed by throat swab and **urgent Gram stain** (see Chapter 4.3).

Glandular fever/infectious mononucleosis

Caused by the Ebstein-Barr virus and may be similar in presentation to diphtheria.

Diagnosis

Atypical lymphocytes on blood, monospot and Paul–Bunnell tests (usually but not always positive).

Management

- **Do not give ampicillin, amoxyl or Augmentin** for throat infection until glandular fever excluded (risk of severe skin reaction).
- IV maintenance fluids.
- IV hydrocortisone 4 mg/kg/6 hourly if signs of airway obstruction occur.
- Intubation/tracheostomy is rarely indicated.

Tonsillitis and quinsy (see Chapter 3.18)

Mediastinal tumours

Slow onset of stridor in a child with other symptoms and signs, for example pallor, lethargy. ***May be precipitated or aggravated by mediastinal radiotherapy.***

Management

- *X* ray chest and mediastinal inlet.
- Intubation may be required as a temporary measure.
- Treat primary cause.

3.18 Tonsillitis, otitis media and mastoiditis

Andrew Freeland

Minimum standards requirements

- Antibiotics: penicillin/amoxil/erythromycin
- Ephedrine nose drops
- Wicking

Tonsillitis

Tonsillitis is a common childhood disorder. The usual bacteria are beta-haemolytic streptococci, *Streptococcus pneumoniae* and *Haemophilus influenzae;* 50% of attacks are viral.

Classic symptoms include pyrexia and sore throat. Swallowing solid food is difficult and fluids must be encouraged. Painful cervical lymphadenopathy is the rule and referred earache from the IXth cranial nerve is common. Febrile convulsions may occur in younger children who may also present with acute abdominal pain without any throat symptoms due to mesenteric lymphadenitis.

Examination

- Tender lymphadenopathy behind the angle of the mandible.
- Red enlarged tonsils with or without purulent exudate.

Differential diagnosis

Diphtheria and infectious mononucleosis (see Chapter 3.17 on upper airway obstruction).

Treatment

- Paracetamol (20 mg/kg 4–6 hourly) for pain relief. Keep in mind attacks are often viral and often antibiotics are not needed.
- Penicillin is still an effective antibiotic and in **serious cases** in hospital, penicillin 12.5 mg/kg four times daily orally. If the child is sensitive to penicillin, erythromycin may be used.
- Rarely there is airway obstruction due to tonsillar enlargement, use IV benzylpenicillin 25 mg/kg 6 hourly and IV hydrocortisone 4–8 mg/kg initially and then a further dose 4 hours later if needed at 4 mg/kg.

Recurrent tonsillitis

- If the number of attacks increases with age rather than decreases, tonsillectomy is appropriate.
- As a rule of thumb, six attacks per year for two years over the age of 5 would seem a yardstick on which to base the case for tonsillectomy.
- It is often said that peritonsillar abscess (quinsy) is an indication, but one attack of quinsy is not enough to warrant the operation.

Indications for tonsillectomy

In the past tonsillectomy was done all too often. Sleep-related upper airway obstruction (see Chapter 3.19) is a good reason for tonsillectomy and about 10% of tonsil operations are currently done for this reason.

Peritonsillar abscess (Quinsy)

This is a complication of tonsillitis and presents with a unilateral swelling of the soft palate deflecting the uvula to the opposite side with associated trismus. Drainage is often necessary as well as intravenous penicilin as above.

Acute suppurative otitis media (ASOM)

Acute suppurative otitis media is a mucosal infection of the middle ear and mastoid aircells, arising via the eustachian tube. *Strep. pneumoniae* and *Haemophilus influenzae* are the commonest bacteria and about 50% are due to viruses.

The symptoms are hearing loss, earache and fever. Pain is due to the bulging tympanic membrane from accumulated pus. Rupture leads to otorrhoea with rapid symptom improvement. Localising signs may be absent in infants, who may present with fever and systemic illness. On examination the tympanic membrane is red and bulging.

Treatment

- **Many cases of otitis media are incorrectly diagnosed**; any child who is crying or has a fever will tend to have pink eardrums.

- Earache often presents at night. This is usually due to eustachian tube obstruction occurring, when the child is sleeping, from accumulated mucus in the postnasal space resulting in a negative pressure in the ear, waking the child up with discomfort. Paracetamol, sitting up and drinking, open the eustachian tube relieving symptoms. Antibiotics are unnecessary.
- In true otitis media with bulging eardrums amoxycillin <1 year = 62.5 mg, 1–4 years 125 mg, 5–12 years 250 mg, >12 years 500 mg all at three times daily should deal with the two commonest bacteria mentioned above.
- Paracetamol relieves pain and reduces fever.
- Ephedrine nose drops (0.25 % in 0.9% saline) 8 hourly for a maximum of 5 days may help open the eustachian tube and speed resolution.
- If the eardrum is perforated antibiotics may no longer be necessary but the ear must be kept dry until the resulting perforation has healed. This is achieved by teaching the parent to undertake wicking. Instructions as follows: roll a clean, soft absorbent cotton cloth or strong tissue paper into a wick. **Never use a cotton tipped applicator or flimsy paper that will fall apart in the ear. Do not use a stick of any kind**. Place the wick in the ear and remove it when wet after a few seconds. Repeat until the ear is dry. Wicking should occur at least three times daily usually for 1–2 weeks until pus is no longer present. The parent must not leave anything in the ear, must not put oil or any other fluid in the ear, and should prevent the child going swimming or putting their head under water until the ear has been dry for at least 2 weeks.

Secretory otitis media

This may lead to recurrent attacks of ASOM because the exudate acts as a culture medium for repeated infections. ***Occasionally myringotomy with grommet insertion is necessary.*** The alternative treatment is long term low dose antibiotics (trimethoprim 2 mg/kg once daily for three months). Eustachian tube function may also be improved by adenoidectomy.

Acute mastoiditis

This is a complication of ASOM. The mucosa of the mastoid system is always inflamed in ASOM. Mastoiditis occurs when the mucosal inflammation spreads to the adjacent bone, causing osteitis and eventually the outer cortex of the mastoid is breached leading to a subperiosteal abscess behind the ear. The symptoms are as with ASOM but the signs include a forward displaced pinna with a tender, fluctuant swelling in the post auricular sulcus.

Complications

Not only is the outer cortex of the mastoid involved, but so can the bone adjacent to both the middle and the posterior cranial fossa, occasionally leading to **extradural abscess, meningitis** and **brain abscess. Facial nerve paralysis** may occur from the pressure of pus on an exposed facial nerve.

Treatment

- Intravenous benzylpenicillin 50 mg/kg IV 6 hourly for 5 days and then orally 25 mg/kg 4 times daily for another 5 days and if there is no improvement within 12 hours, surgical drainage is necessary.
- The key is to provide drainage for the mastoid system. ***If it is not possible to do a full scale mastoidectomy (because of lack of equipment or expertise)*** dramatic improvement, in conjunction with intravenous antibiotics, may be obtained by incising the abscess (avoiding the mastoid tip in the small child where the facial nerve may be exposed) and opening into mastoid air cells.

Mastoidectomy

The surgical landmarks are as follows: after elevation of the periosteum it is possible to find the posterior edge of the external auditory meatus. An anterior superior landmark is the zygomatic process. A horizontal line drawn backwards from the zygomatic process connecting with a vertical line from the posterior ear canal wall will be the site of the mastoid antrum. Opening the mastoid here with a drill, or a hammer and gouge, is safe. Widening this opening without damaging the dura covering the temporal lobe superiorly, or the posterior fossa covering the lateral venus sinus posteriorly will provide good drainage. Intracranial complications will, of course, need neurosurgical expertise (see Chapter 3.44).

3.19 Sleep related upper airway obstruction

Martin Samuels

Minimum standards requirements

- Surgery
- Nasal CPAP
- Sleep video-oximetry

Introduction

The incidence of sleep-related upper airway obstruction depends on the method of diagnosis (1–3% of pre-school children). It is associated with

- enlargement of tonsils/adenoids
- reduced tone or diameter in upper airway.

Groups at risk are children with:

- Sickle cell disease
- Cerebral palsy
- Down's syndrome
- Congenital palatal problems
- Pierre–Robin sequence
- Craniofacial deformities
- Prader–Willi syndrome
- Central hypoventilation (rare).

Presenting features

- Sleep disturbance
- Snoring (occurs in >10 % of 4–5 year olds)
- Mouth breathing + halitosis
- Restlessness
- Nocturnal enuresis
- Daytime sleepiness
- Subcostal and sternal recession during sleep
- Odd sleep positions
- Nocturnal sweating
- Pulmonary hypertension
- Heart failure

✔ **These features may be associated with impaired cognitive function, daytime sleepiness and behavioral disorders. It is insidious** – the child may appear completely normal when awake and the problem shows most or only during REM sleep.

Investigations

- Sleep observation/study. Either direct observation of the child during sleep with chest and face exposed or by video recording at home during sleep (by parents)
  - Chest wall recession
  - Snoring
  - Sleep position
  - Nocturnal restlessness
- Measurement of arterial oxygen saturation using pulse oximeter during sleep
- Possibly barium swallow
- ***Possibly fibre-optic upper airway endoscopy***
- ***Possibly lateral X ray postnasal space to show size of the adenoids.***

Hypoxaemia

- Continuous lowering of baseline arterial oxygen saturation (SpO_2) during sleep
- Recurrent dips in SpO_2 associated with partially or completely obstructed breaths

Normal SpO_2 values

Methodological issues:

- Instruments used
- Exclusion of motion artefact
- Averaging
- Altitude
- Inclusion of apnoeic pauses

Normative data

Nellcor N-200: beat-to-beat mode

- (Preterms) J Pediatr 1993;123:963–8
  J Pediatr 1992;120:447–54
- (Full terms) Arch Dis Child 1991;66:569–73
- (First year) J Dev Physiol 1991;15:341–45
- (2–16 years) Pediatrics 1993;92:686–90

Table 3.19.1 Baseline SpO_2 in infants

N	Median age in days	Range	Median SpO_2	5th percentile
55	1	1–7	99.4	95.7
160	20	3–165	99.6	95.7
110	62	30–176	100.0	97.9
60	4	1–7	97.6	93.2
60	17	8–28	98.0	91.9
66	39	29–54	99.8	97.5
16	102	83–146	99.9	99.2

Summary of desaturations (pulse oximeter in non-averaged mode)

- Majority of premature and full-term infants have short-lived falls in $SpO_2 \leq 80\%$ for ≥ 4 s
- Six weeks later these are rare
- Prolonged desaturations were less frequent in the first week of life than later

Adverse effects of hypoxaemia

- Poor weight gain
- Pulmonary hypertension
- Rise in airway resistance
- Cyanotic–apnoeic episodes

Treatment

- Time – the airway enlarges with growth
- Topical steroids/decongestants
- Tonsillo-adenoidectomy
- Nasal CPAP (see Chapter 1.26)
- Nasopharyngeal tube
- Tracheostomy

Nasal CPAP (see Chapter 1.26)

- Effective – non-invasive
- Potential problems
  - – Compliance
  - – Side effects
    - Pressure sores
    - Nose bleeds
    - Drooling
    - Aerophagy

Table 3.19.2 SpO_2 levels at different altitudes

Altitude (m)	Location	N studied	Age	SpO_2 (%)		Author	Year
Sea level	UK	70	2–16 years, mean 8 years	range median	95.8−100 99.5	Poets et al	1993
Sea level	Peru	189	2 months–5 years	range mean	96−100 98.7±1.1	Reuland et al	1991
1610	Colorado	150	<48 hours	95% CI mean	88−97 93.0±0.2	Thilo et al	1991
			3 months	95% CI mean	86−97 92.2±0.4		
1670	Nairobi	87	7 days–36 months	range mean	89.3−99.3 95.7±1.6	Onyango et al	1993
2640	Bogota	189	5 days–24 months	range mean	84−100 93.3±2.1	Lozano et al	1992
2800	Colorado	72	3–670 days	range mean	88−97 91.7±0.2	Nicholas et al	1993
3100	Colorado	14	6 hours–4 months 1 week 4 months	range mean mean	81−91 80.6±5.3 86.1±4.6	Niermyer et al	1993
3658	Tibet	15	6 hours–4 months	*immigrants** *indigenous*	76−90 86−94	Niermyer et al	1995
3750	Peru	153	2–60 months	range mean	81−97 88.9±2.9	Reuland et al	1991

Values given are those in (quiet) sleep.
* *Ranges for babies born to immigrant Chinese mothers, and indigenous babies whose families have lived at this altitude for innumerable generations*.

3.20 Anaemia

Christiane Ronald and Simon Parke

Minimum standards requirements

- Iron
- Folic acid
- Antihelminthic drugs: mebendazole/albendazole/pyrantel
- Blood transfusion
- Antimalarial drugs

Definition

Table 3.20.1 Haemoglobin concentrations below which anaemia is present (at sea level) according to WHO

Children	6 months to 4 years old	11 g/dl	HcT 33%
	5 years to 11 years old	11.5 g/dl	HcT 34%
	12–14 years old	12 g/dl	HcT 36%

The problem

- Widespread in disadvantaged countries.
- Common in young children under 5 years of age.
- More than one cause of anaemia is usually found in the anaemic child.
- Treatment with iron injections may increase mortality (meningitis) and morbidity (respiratory infections, malaria) in infants.
- Genetic causes of anaemia are common.

Causes of childhood anaemia in disadvantaged situations

- Low birthweight
  Due to low iron stores (0–2 years age group) (see Chapter 3.21)
- Dietary
  Diets tend to be low in iron
- Infections
  Parasites
  Malaria (haemolysis)
  Hookworm, *Trichuris* (blood loss) and others
  Bacteria and viruses
- Genetic
  Haemoglobinopathies (HbSS and others) (chronic haemolysis), glucose-6-phosphate dehydrogenase deficiency. (see Chapters 3.22 and 3.23)

Clinical features

- May be asymptomatic until Hb < 8 g/dl (however poor appetite, weakness and tiredness may be present)
  Breathless on exertion when Hb < 6 g/dl
- Pallor
  nail beds (the best site)
  mucous membranes
- Suboptimal growth
- Physical signs of chronic haemolytic anaemia (for example bossing, prominent jaw, child may be jaundiced)
- Congestive heart failure

Investigations

The tests in bold should *always* be done before a transfusion!

- **Haemoglobin concentration** (cyanmethaemoglobin method)
- **Bloodfilm** **Malarial parasites**
  RBC: hypochromia, microcytosis, anisocytosis (iron deficiency, thalassaemia)
  sickle cells
  target cells (thalassaemia, iron deficiency)
  WBC: hypersegmented neutrophils (folate, vitamin B12 deficiency)
- Mean corpuscular volume (MCV) and reticulocyte count as the two principal criteria for the initial classification of anaemia.
- **Haemoglobin electrophoresis:** sickle cell, thalassaemia (blood can be stored for this in a sequestrene bottle).
- Red cell volume distribution (RDW) index is a useful criterion of classification alongside MCV. This can be measured with automated red cell analysers (Coulter counter). RDW index reflects the heterogenicity of the

red blood cell size and thus provides a quantitative measure of anisocytosis.
- Stool test: parasitic ova, blood.
- Haematocrit or PCV (microcentrifuge).

Management

- Establish diagnosis and severity of anaemia
- Treat malaria (oral route) – see malaria guidelines
- Haematinics (oral route)

Folic acid

- Up to 5 years: 2.5 mg once daily
- >5 years: 5 mg once daily

Iron

Table 3.20.2 Iron therapy

Age or weight (6 mg/kg elemental iron)	Ferrous sulphate 200 mg (60 mg elemental iron)	Ferrous fumarate 60 mg per 5 ml (12 mg elemental iron/ml)
2–4 months (4–6 kg)	–	2ml
4–12 months (6–10 kg)	–	2.5ml
1–3 years (10–14 kg)	$\frac{1}{2}$ tablet	4ml
3–5 years (14–19 kg)	$\frac{1}{2}$ tablet	5.5 ml
>5 years	1 tablet	–

Iron or folic acid should be given once daily for two months.

Antihelminthics

- **Albendazole**
  Drug of choice if available:
  400 mg as a single dose (200 mg if child – 2 years)
- **Mebendazole**
  Most effective against hookworm and *Trichuris trichiuria* (whipworm).
  For children over 1 year: 250 mg as single dose (or 500 mg when the child is above 2 years).
  May be repeated after 2 or 3 weeks.
- **Pyrantel**
  Most effective against hookworm: 10 mg/kg (max. 1 g) as a single dose.

Blood transfusion (see Chapter 1.14)

Only when essential
- Warm blood first under mother's dress in contact with skin especially in infants.
- Do not use blood stored for >35 days at 2–6 °C or out of fridge for >2 hours or visibly spoiled (plasma must not be pink, redcells not purple or black) or bag open or leaking.
- Check blood is correct group and patient's name and number are identical on both label and form.
- Needle/catheter 22 gauge or larger to prevent clotting.
- **If signs of heart failure give 1 mg/kg of furosemide IV at start of transfusion unless hypovolaemic shock is also present.** ✔
- Record baseline temperature and pulse rate.
- Do not allow a single unit of blood to go in over longer than 4 hours.
- Ideally in infants or those with heart failure control flow with in-line burette.
- Record observations every 30 minutes looking for heart failure and transfusion reactions.
- Record quantities given.

Indications
- Severe anaemia (Hb < 4g/dl)
- Impending or overt cardiac failure if Hb < 6 g/dl
- Hyperparasitaemia in malaria if Hb < 6 g/dl
- In sickle cell disease
  if Hb < 5 g/dl or severe infection present
  Stroke (regardless of Hb)
  Priapism (regardless of Hb)
- Children in congestive cardiac failure due to severe anaemia (consider partial exchange)
- Severe chronic haemolytic anaemia such as thalassaemia major
- Following acute severe blood loss when 20–30 % of the total blood volume (80 ml/kg) is lost and bleeding is continuing – **remember that the Hb can initially be normal!**

Volume of transfusion
- Give whole blood: 20 ml/kg or
  Required volume (ml) = weight (kg) × 4 × desired rise in Hb (g/dl)
  or
- Packed red cells:
  10–15 ml/kg or
  Required volume (ml) = weight (kg) × 3 × desired rise in Hb (g/dl)

In both cases, rate = 5 ml/kg/hour (usually over 3–4 hours unless shocked).

Add furosemide 1 mg/kg IV immediately in advance of transfusion to avoid precipitating cardiac failure (unless hypovolaemic shock) in cases of very severe anaemia.

Transfusion reactions
(see Chapter 1.14)

Control of anaemia

- Improve iron intake in infants
  - Breastfeeding for at least 6 months
  - Give breastfeeding mothers iron
  - Vitamin C-rich foods (citrus fruit juices) and/or meat, fish, beans and leafy vegetables by 6 months with cereals
  - Low-birthweight babies to receive oral iron 2 mg/kg daily from the age of 4 weeks for 6 months
- Prevent infections
  - Diarrhoea (breast milk)
  - Measles (vaccination)
  - Prevention and prompt treatment of malaria

- Routine deworming of children under 5 years every 3–6 months
- Pneumococcal pneumonia in sickle cell patients (reduce by vaccination and oral penicillin prophylaxis: this is usually begun at diagnosis and continued for at least the first 5 years of life: 125 mg twice daily for first 5 years, or 12.5 mg/kg twice daily, 250 mg twice daily 6–12 years).
- Malaria prophylaxis in sickle cell patients.

Further reading

Integrated Management of Childhood Illness, WHO, MOH, UNICEF, 1997.

O'Callaghan C, Stephenson T. *Pocket Paediatrics*, p. 241. Edinburgh: Churchill Livingstone, 1992.

Long SS, Pickering LK, Prober CC. *Pediatric Infectious Diseases*, p. 731. Churchill Livingstone, New York 1997.

World Health Organization. *Management of a Child with a Serious Infection or Severe Malnutrition*. Geneva: WHO, 2001.

3.21 Iron deficiency anaemia

Rob Moy

Definition

Nutritional iron deficiency of sufficient severity to cause anaemia, i.e. reduction in the blood haemoglobin concentration below a level which is normal for that individual (see Table 3.21.1). Lesser degrees of iron deficiency cause depletion of iron stores (indicated by low plasma ferritin, <12 micrograms/litre) and iron deficient erythropoiesis (indicated by microcytic hypochromic red blood cells).

Table 3.21.1 Cut-off points to define anaemia

Group	Subgroup	Hb (g/dl)
Children	6 months to 6 years	<11
	6–14 years	<12
Adults	Males	<13
	Females (non-pregnant)	<12
	Females (pregnant)	<11

Approaches to diagnosis

- Child coming from a population at risk (see causes)
- Symptoms, for example tiredness, lethargy, irritability, poor concentration
- Clinical signs: pallor of conjunctiva, nail beds, oral mucosa, palmar creases, absence of nail-bed blanching
- Near patient blood testing: optical haemoglobinometer, blood spot colour matching (WHO Colour Scale) microhaematocrit determination, HemoCue haemoglobinometer (photometric assay)
- Laboratory blood testing: automated Coulter counter (Hb, red cell indices), serum ferritin (reflects iron stores), other biochemical measures of iron deficiency

Adequate capillary blood specimens can be obtained from a warm finger or toe or heel of an infant with a lancet. Wipe away the first three drops of blood to encourage free flow and then sample the fourth. Avoid excessive squeezing to minimise contamination of the blood specimen with tissue fluid.

Differential diagnosis of hypochromic anaemia

- Thalassaemia
- Anaemia of chronic disease and infection
- Rare hereditary disorders of haemoglobin synthesis

Causes of iron deficiency anaemia

Iron deficiency is the commonest nutritional deficiency worldwide and the commonest cause of anaemia.

- Inadequate accumulated iron stores in preterm, low birthweight and twin babies, those whose mothers were severely anaemic in pregnancy, after early clamping of the umbilical cord or severe postnatal blood loss.
- Inadequate iron in the diet after early cessation of breastfeeding, early introduction of whole cows' milk in the first year, prolonged breastfeeding as the main food source, or weaning onto low iron-containing solids.
- Poor bioavailability of iron from high fibre cereal or root vegetable diets without meat or adequate vitamin C containing foods or juices.
- Reduced absorption of iron because of the consumption of excessive amounts of tea (tannins) or unleaven breads (phytates).
- Iron loss occurring in menstruating girls or bleeding caused by hookworm infection, *Helicobacter pylori* gastritis or schistosomiasis, often worsening the severe haemolytic anaemia due to malaria in tropical areas.
- Rapid growth creating high demands for iron in adolescents.

Effects of iron deficiency

- Tiredness and irritability
- Impaired psychomotor development in young children
- Impaired scholastic achievement in school children
- Impaired immune function
- Increased susceptibility to infection
- Reduced weight gain and growth

Prevention of iron deficiency

Primary prevention

- Encourage breastfeeding or use iron-fortified infant formula (containing around 7 mg/litre of iron) iron fortified or meat-containing weaning foods
- Vitamin C-containing citrus fruits and juices (enhances iron absorption)
- Prophylactic supplementation with iron drops (5 mg of elemental iron/kg/day) especially for low-birthweight babies (after 28 days of age)
- Deworming and community programmes to reduce reinfestation.

Secondary prevention

Screening for anaemia in at-risk population (see causes of iron-deficiency anaemia, above)

Iron therapy

- Treatment of underlying cause, for example nutritional deficit, hookworm, etc.
- Oral iron supplementation (ferrous sulphate, fumarate or gluconate);
  3 mg elemental iron/kg body weight per day of iron-containing syrup once daily dosage at least half an hour before meals to maximise absorption (once or twice weekly might be more effective then daily doses). Treatment to be continued for 3 months to replenish iron stores. An adequate response is indicated by a rise in Hb of 1 g/dl or more per month. Gastrointestinal side effects and non-compliance are common.
  - Despite theoretical risks of iron supplements increasing the risk of infection or malaria, present evidence suggests that iron supplementation for prophylactic or therapeutic uses is safe in malaria areas.
  - Parenteral iron should be avoided in children especially in the newborn (increased risk of Gram-negative sepsis) or in geographical areas with endemic malaria (increased risk of malaria in anaemic children given parenteral iron).
  - Iron therapy must be avoided in the initial stage of the management of severe malnutrition because of the risk of precipitating infection and free radical damage (see Chapter 3.16).
  - Iron therapy may make severe infections worse since there is evidence during septicaemia that bacteria feed on iron. It may be better to await recovery from severe bacterial infection before starting treatment.

Severely anaemic malnourished children with Hb $<$ 4 g/dl should receive careful blood transfusion with diuretic cover (see Chapter 3.16).

3.22 Sickle cell disease

Sally C Davies

Minimum standards requirements

- Analgesia: paracetamol/NSAIDs/opiates
- Blood transfusion
- Oxygen
- Penicillin
- Pneumococcal, hepatitis B and *Haemophilus influenzae* vaccines
- ORS
- Antimalarial drugs
- Desferrioxamine

When one sickle gene is co-inherited with a second sickle gene (Hb SS, sickle cell anaemia) or with another haemoglobin (Hb) beta-chain genetic abnormality (for example beta-thalassaemia (Sβ+ or $S\beta^0$) and Hb C (Hb SC disease)), clinical problems generally ensue.

βs is widely spread throughout Africa, the Middle East, Mediterranean countries, and India and has been carried by population movement to the Caribbean, North America and North Europe. The mutation has occurred on at least four separate occasions and the frequency of sickle cell carriers (Hb AS), who are healthy is up to 1 in 4 in West Africans and 1 in 10 in Afro-Caribbeans.

The clinical problems in sickle cell disease (SCD) relate to vaso-occlusion, caused by polymerisation of deoxygenated Hb S. This results in erythrocytes becoming sickle (or crescent/banana) shaped, stiff and poorly deformable. They adhere to the vascular endothelium leading to blood vessel blockage and tissue oxygen deprivation and damage.

SCD has marked clinical variability with a median survival in the fifth decade in advantaged countries. However, in poor countries, most people with SCD will die of one of its complications, with a high infant and childhood mortality. In addition, large blood vessel occlusion also occurs, resulting in thrombotic cerebrovascular accidents, the acute sickle chest syndrome, and placental infarction.

Diagnosis of SCD

- A positive sickle solubility test denotes the presence of sickle haemoglobin but does not define whether the person is a carrier (Hb AS) or Hb SS.
- Haemoglobin electrophoresis demonstrates the absence of Hb A and either Hb S (SS) or Hb S and another haemoglobin such as Hb C (SC).
- A full blood count shows severe (SS) to mild anaemia (SC).
- Examination of the peripheral blood shows sickled erythrocytes.

Painful vaso-occlusive crisis

- The most common clinical problem resulting from blockage of small vessels.
- The spectrum of presentation alters with age. In particular, the distribution of bony crisis shifts from peripherally to a more trunk distribution with the recession of the red bone marrow and its replacement by fatty bone marrow.
- The first presentation in a majority of infants is with the "hand-foot syndrome" or dactylitis, in which infarction of the metacarpals or metatarsals leads to an associated overlying soft-tissue reaction with swelling, redness, tenderness and, often, a marked light reflex. This may affect single or multiple hands or feet.
- In later infancy and childhood the long bones, elbows, and knees begin to be involved
- In adolescence, the pain is more likely to affect the thighs, hips, spine, ribs, shoulders, upper humerus, as well as the bones of the cranium joints and muscles.

Management of the painful vaso-occlusive crisis, as other complications of SCD, requires treatment of any precipitating causes (see Table 3.22.1) with maintenance of adequate hydration and good analgesia (see Table 3.22.2 and Chapter 1.27).

Table 3.22.1 Precipitating causes of vaso-occlusion

Problems	Treatment
Cold	Warmth and rest
Pyrexia	Antipyretics (e.g. paracetamol). Treat malaria or bacterial infection
Infection	Appropriate antibiotic/chemotherapy
Dehydration	Rehydration
Hypoxia	Inhalation of oxygen

Table 3.22.2 Management of sickle crisis

- **Antibiotics** (if bacterial infection suspected)
  First line penicillin and/or amoxycillin, antimalarial chemotherapy if appropriate
- **Rehydration**
  25–50% > maintenance (orally or IV) (Chapter 3.25)
- **Analgesia**
  Use "ladder" (see Chapter 1.28); if inadequate then use opiates (see Chapter 1.27)
- **Blood transfusion**
  Only if severely anaemic and clinically compromised (e.g. Hb < 5 g/dl in SS)
- **Oxygen**
  When hypoxaemic (either clinically or Sao_2 < 92%)

Pain and its response to treatment is generally best measured using a visual analogue scale of 10 cm:

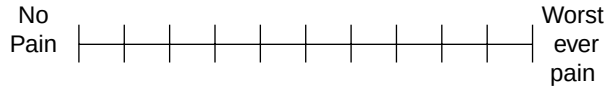

Figure 3.22.1 Visual analogue scale

or by asking a child to point at a face that best describes their pain (Figure 3.22.2).

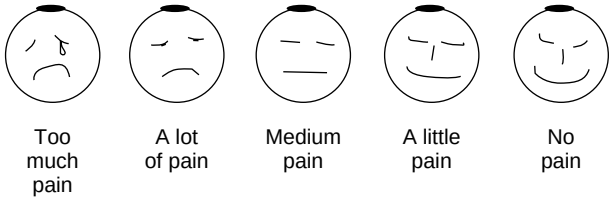

Figure 3.22.2 Pain scale for children

Infection

Sickling in the spleen and consequent autosplenectomy occurs in most SS and SB^0 patients. A degree of splenic damage also leads to immunological impairment in other types of SCD. As a result, patients are prone to overwhelming infection by encapsulated bacteria, in particular *Pneumococcus*, *Salmonella* species and *Haemophilus*.

The initial distribution of SCD was coterminous with falciparum malaria and, while the carrier state offers protection, malaria is frequently fatal for those with SCD. It is, therefore, essential to educate the parents and families as to the importance of using impregnated mosquito nets with chemoprophylaxis where available and affordable. Any acute episode of malaria infection should be treated promptly and appropriately (see Chapter 4.27).

Severe dehydration (for example from gastroenteritis) and, consequently, vaso-occlusion is a common cause of death in infants and young children (see Chapter 3.25). Severe dehydration may also result in sickling in the pulmonary vasculature (known as the "acute sickle chest syndrome") or in the major cerebral blood vessels leading to a cerebrovascular accident. Parents should be educated as to the importance of bacteriologically clean drinking water, hand-washing, and the need to start treatment with rehydration solutions early in these children (see Chapter 3.25). In addition, in hot climates, extra fluids may be needed to compensate for the increased insensible losses.

The human parvovirus B19 can infect developing erythroblasts preventing red cell production for 1–2 weeks. As the mean red cell life span in SS is only around 10 days, the haemoglobin concentration falls rapidly, known as the aplastic crisis. Urgent additive red cell transfusions can be life-saving.

Osteomyelitis, in particular caused by *Salmonella* species and *Escherichia coli*, can occur at any age. The management is no different from the general population: surgical drainage may be required with a minimum of six weeks of the appropriate antibiotics (see Chapter 3.45).

Chest infections are particularly common and, because of the risk of deaths associated with pneumococcal infection linked to hyposplenism or auto-splenectomy, any lobar pattern of infection should be treated urgently with IV (or IM if this is not possible) benzylpenicillin 50 mg/kg every 6 hours. Penicillin prophylaxis (single oral dose daily: 62.5 mg < 12 months of age, 125 mg 12–36 months of age, 250 mg > 3 years) and immunisation against the *Pneumococcus* reduce the risk of overwhelming infection and death from this complication. Immunisation against *Haemophilus influenzae* provides additional protection. Immunisation against hepatitis B can reduce transfusion risks.

Sequestration events

In sequestration there is pooling of red cells in an organ, a fall ≥2 g/dl of circulating haemoglobin, loss of function of the organ, and, in the case of the spleen and liver, enlargement. Infection may play a precipitating role.

Splenic sequestration is often underrecognised in poor countries either because it occurs so acutely that the infant dies before a diagnosis is made or because cases are sufficiently mild not to present to medical services and either resolve spontaneously or result in chronic hypersplenism (see later). Other organs that can be involved in sequestration include the liver, the lungs and the bowel.

Neurological involvement

- Meningitis is more common than in normal children and clinicians should maintain a high index of suspicion for this complication.
- Stroke is a common problem as a result of stenosis or occlusion of the cerebral vasculature, leading to hemiplegia (7%) or related complications (see Table 3.22.3).
- Most children make a good motor recovery from an initial stroke but are left with intellectual defects. Untreated a half to two-thirds of children will suffer a second cerebrovascular accident, usually within 2–3 years of the first episode, as a result of which many will die and most will be seriously impaired. Transient ischaemic attacks may presage a more major event.

Table 3.22.3 Neurological complications of sickle cell disease

- Meningitis
- Cerebrovascular accident
- Transient ischaemic attack
- Altered state of consciousness/coma
- Convulsion
- Dementia (multi-infarct or cerebral atrophy)
- Reduction in "intelligence quotient"

Treatment of stroke in SCD

- The only effective treatment is regular blood transfusions (to suppress sickle haemoglobin production and maintain oxygen delivery).
- Transfusions are required every 3–4 weeks to maintain the haemoglobin between 10.5 and 14.5 g/dl. Such hypertransfusion regimes are onerous for the patient, their family, and the medical services with the risks of transmission of infection, alloimmunisation to foreign red cell antigens and, inevitably, in the absence of treatment, the development of iron overload.
- Parenteral desferrioxamine (20–50 mg/kg SC infusion over 8–12 hours 5–7 nights weekly) is effective but expensive, can be painful, has side effects, including high tone deafness, retinopathy, osteoporosis and *Yersinia enterocolitica* infection. Desferrioxamine up to 2 g/500 ml of blood given at the time of transfusion as an IM injection is helpful but is insufficient on its own.
- Patients with altered levels of consciousness or coma should be treated as for crisis and an exchange transfusion performed.
- Convulsions should be treated (see Chapters 3.37 and 3.38).

Chronic problems of sickle cell disease in childhood

- Failure to thrive is common particularly when hypersplenism is present (sometimes from malaria infection or from chronic splenic sequestration).
- If haemoglobin is 2 g/dl or more lower than that patient's stable state (generally ≤ 5 g/dl in SS) then splenectomy can make a dramatic difference.
- Puberty may be delayed because of hypersplenism or malnutrition because of the hypermetabolic state and inadequate nutrition. Dietary advice, treatment of any chronic infections and splenectomy (if hypersplenism is present) may help.
- 30% of SS children suffer from sleep-related upper airways obstruction with consequent hypoxaemia. This should be treated as for other children with upper airways obstruction (see Chapter 3.19).
- Chronic pain resulting from damage caused by acute vaso-occlusive crises and other pains secondary to the haemolytic process occurs as shown in Figure 3.22.3.
- Avascular necrosis of the hip or shoulder can occur as young as 6 years although it is uncommon before adolescence. The initial presentation may be with the acute vaso-occlusive crisis but once disintegration of the femoral head occurs, the pain is of a chronic osteoarthritic type and should be managed as such.
- Leg ulcers, that can become seriously infected, are common and their prevalence rises with age. Appropriate antibiotics such as erythromycin and flucoxacillin, wound cleaning and protection together with rest and elevation of the leg help. Compression stockings may also assist.
- Children develop a renal tubular concentrating defect by the age of 2 years. During adolescence proteinuria, the nephrotic syndrome or chronic renal failure may develop.

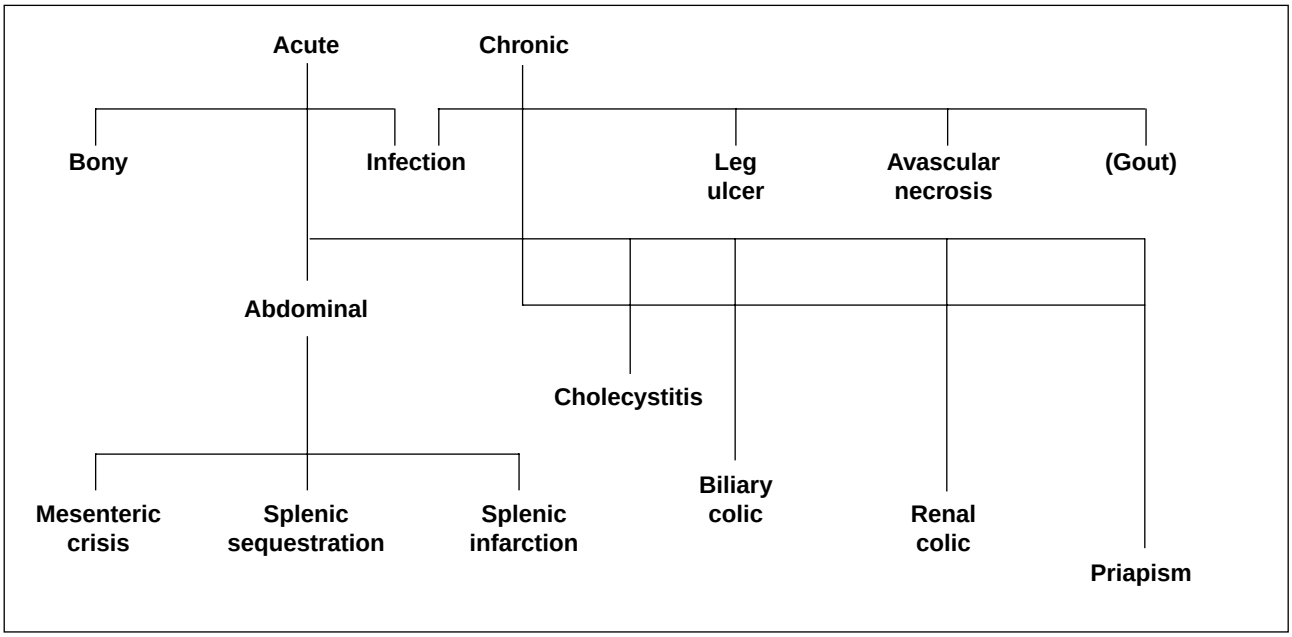

Figure 3.22.3 Pain related to sickle cell disease.

- Renal papillary necrosis may produce haematuria, urinary tract infection and renal colic. Rarely the haematuria is severe and blood transfusion is required. Renal colic is treated with copious fluids and adequate analgesia.
- Many patients are chronically jaundiced with exacerbations. There is no treatment and reassurance should be given that this rarely represents liver failure.
- A number of SS boys will experience priapism which can be either stuttering, that is lasting up to a few hours on a daily basis, or fulminant where the episode lasts for 6 hours or longer. Protracted fulminant priapism can lead to fibrosis of the corpora cavernosum and subsequent erectile impotence if it occurs in adolescence. This risk appears somewhat lower in the prepubertal boys. Many cases of stuttering priapism respond to either exercise or warm baths. It can be prevented by the ***use of the alpha-adrenergic stimulating drug etilephrin orally 0.5 mg/kg at night or 0.25 mg/kg twice daily if priapism occurs both day and night.*** Fulminant cases are usually treated by rehydration and gentle sedation of the patient with ***intracorporeal injection of an alpha-adrenergic stimulator or a surgical washout and drainage procedure.*** Exchange transfusion may be needed.

3.23 Haemolytic anaemias

Beatrix Wonke

Minimum standard requirements

- Folic acid
- Screened blood for transfusion
- Splenectomy
- Desferrioxamine
- Pneumococcal vaccine/penicillin

A variety of disorders characterised by a reduction in the life span of red blood cells.

Table 3.23.1 Commonest causes of haemolytic anaemias

With minor splenomegaly	With marked splenomegaly
G6PD deficiency	Sickle cell disease (see Chapter 3.22)
Autoimmune haemolytic anaemia	Beta-thalassaemia major
Haemolytic uraemic syndrome	Hb-E beta-thalassaemia
	Hereditary spherocytosis
Beta-thalassaemia minor (heterozygous)	Malaria in tropical splenomegaly
Hb H alpha thalassaemia syndrome	Visceral leishmaniasis (kala-azar)

Hereditary haemolytic anaemias

Red cell membrane defects (dominant inheritance)

Spherocytosis

This is the most common haemolytic anaemia due to a membrane defect. It may present at any time from birth to old age and varies in severity; from patients with Hb concentrations of 4–5 g/dl to asymptomatic ones with normal Hb.

Diagnosis

- Along with a positive family history, the clinical features are mild jaundice, pallor and splenomegaly.
- Laboratory features: blood film shows spherocytes, **increased osmotic fragility** of red cells, increased reticulocytes, **negative antiglobulin test**.

Treatment

- Folic acid (from birth to 12 months 250 micrograms/kg once daily, from 1–3 years 2.5 mg once daily and >3 years 5 mg once daily).
- Severely anaemic and symptomatic moderately anaemic children may benefit from splenectomy if the facilities available make this a low-risk procedure.
- Delay splenectomy until after the age of 5–10 years if possible.
- Administration of **pneumococcal vaccine** prior to splenectomy, prophylactic **penicillin** thereafter (under 12 months 62.5 mg twice daily, 1–3 years 125 mg twice daily, >3 years 250 mg twice daily.)

Elliptocytosis

Diagnosis

- Blood film shows 25–90% of oval, elliptical or rod-shaped red cells.
- Homozygotes tend to have severe haemolytic anaemia from infancy.

Treatment

As for spherocytosis.

Stomatocytosis

Hereditary stomatocytosis is rare, it can be acquired in several conditions, especially liver disease.

Diagnosis

Blood film shows erythrocytes with central mouth-like slit (stomatocytes).

Treatment

As for spherocytosis. Splenectomy may not be effective.

Metabolic defects

Glucose-6-phosphate dehydrogenase deficiency (G6PD) (X-linked)

There are two types of normal G6PD enzymes (type A and B). Worldwide there may be 100 million people with diminished red cell G6PD activity. G6PD A deficiency is

common in black children, their G6PD function is reduced to about 10% of normal. G6PD B deficiency (G6PD Mediterranean) is less common; the enzyme activity is reduced to 1–3%; this and the Chinese variant of G6PD deficiency are the more severe forms of the disease.

Clinical features
- Severe enzyme deficiency: chronic haemolytic anaemia, jaundice.
- <10% enzyme activity: severe episodes of haemolysis occur with oxidant stress (exposure to oxidant drugs such as antimalarials, sulphonamides, high dose aspirin, phenacetin and vitamin K analogues), fava beans (due to ingestion of the fava broad bean or inhalation of its pollen), infection (for example parvovirus). Haemoglobinuria may occur.
- 10–60% enzyme activity: acute episodes of anaemia and jaundice resulting from oxidant stress, drugs, infection.

Diagnosis
- Blood film shows "blister" and "bite" cells
- ***Enzyme assay if available is needed to make the diagnosis (but this may be normal if reticulocytes are raised).***

Treatment
- Avoid drugs causing oxidant stress (chloroquine, primaquine, sulphonamides, nitrofurantoin, quinolones, dapsone, high dose aspirin, phenacetin) or fava beans. If primaquine is necessary, this can be given weekly for 8 weeks.
- Transfusion may be necessary.

Pyruvate kinase deficiency (autosomal recessive)
This is the second commonest enzyme defect of the glycolytic pathway and affects mainly northern Europeans.

Clinical features
- Neonates may present with early jaundice (within 48 hours) and anaemia.
- In older children, haemolysis is variable and may be asymptomatic or lead to poor growth, delayed puberty and the skeletal changes associated with chronic haemolysis, such as maxillary prominence and frontal bossing.

Diagnosis
Enzyme assay for pyruvate kinase.

Treatment
Folic acid (250 micrograms/kg once daily) and splenectomy (if the facilities available make this a low-risk procedure).

Haemoglobin defects

- **Abnormal variants:** sickle (see Chapter 3.22), Hb C, Hb E, Hb D, etc.
- **Defective synthesis:** thalassaemias
- **Beta-thalassaemia major (autosomal recessive)**

Beta-thalassaemia major
In this condition there is complete or almost complete absence of the beta-globin chain synthesis. There is a high incidence of the beta-thalassaemia gene (1–15%) in southern Europe, the Middle East, India, Pakistan and South-East Asia.

Clinical features
- Anaemia becomes obvious by 3 months
- Failure to thrive, intermittent fever, poor feeding
- Infections, splenomegaly
- Stunted growth with skeletal changes, for example frontal bossing, maxillary hyperplasia
- Increased skin pigmentation
- Delayed puberty

Diagnosis
- Blood film shows microcytosis, anisocytosis, hypochromic and nucleated red cells
- Haemoglobin electrophoresis: Hb F increased (10–90%), Hb A absent, Hb A_2 can be reduced, normal or occasionally elevated.
- Serum iron and ferritin are increased

Management of beta-thalassaemia major

Blood transfusion
- Monitor Hb, growth and development and **transfuse when child stops developing or when Hb < 7 g/dl in absence of infection.** ✔
- **Blood should be ABO, rhesus (Dd, Cc, Ee), Kell matched and filtered to avoid alloimmunisation and transfusion reactions.** ✔
- **Immunise** against hepatitis B prior to transfusion.
- Transfuse 20 ml/kg of filtered red cell concentrate over 2–3 hours.
- To monitor, calculate transfused red cell concentrate in ml/kg yearly. If blood consumption is >300 ml/kg, investigate cause.
- Increased blood consumption may be due to: large spleen, large liver, autoimmune haemolytic anaemia or multiple alloantibodies.
- To prevent bone deformities, osteoporosis, extramedullary haemopoiesis **aim for pretransfusion level not less than 9 g/dl.** ✔
- Pretransfusion Hb is mandatory, post-transfusion Hb is optional.
- As a rule, the Hb drops by 1 g/week in splenectomised children, in nonsplenectomised patients by 1.5 g/week.

Iron chelation
- To avoid damage to the endocrine glands, liver and heart this should be started when the **serum ferritin is around 1000 micrograms/litre.** ✔
- **Give no more than 40 mg/kg desferrioxamine** in 10 mls of water for injection, infused subcutaneously over 10–12 hours via an infusion pump on five nights a week. Too much desferrioxamine can cause growth, hearing and eyesight problems. ✔
- Give 100–200 mg vitamin C orally at the same time as desferrioxamine. This enhances iron excretion in the urine.

- Oral chelation may be used when desferrioxamine is not available or not tolerated. Ferriprox is licensed in the EU and Kelfer is licensed in India.
- Monitor neutrophil count every two weeks
- If neutrophils $< 1.0 \times 10^9$/litre stop iron chelation and monitor recovery
- If infection is present and neutrophil count is $< 0.5 \times 10^9$/litre and there are symptoms, take blood cultures and treat with broad-spectrum antibiotic to avoid septicaemia.
- Other side effects are joint pain, nausea, fluctuating liver enzymes and zinc deficiency.

Monitoring treatment

- Measure height and weight, plot height velocity and watch for delayed puberty.
- To avoid psychological trauma and ensure the development of secondary sexual characteristics, treat if no signs of sexual development have occurred by 16 years of age (see Chapter 3.11).
- Check at least twice yearly: serum ferritin (iron overload), liver function tests, calcium, phosphate, alkaline phosphatase (hypoparathyroidism, tetany).
- ***Yearly screening for HCV and HIV infection.***
- ***If HCV is positive assess viraemia (serotype) if possible, do liver biopsy and give interferon with or without ribavarin to avoid cirrhosis and hepatoma.***
- ***If HIV-positive, continue transfusions and give latest available antiviral treatment.***
- **All blood donors should be tested for HCV and HIV.** ✔

Acquired haemolytic anaemia

- **Iso-immune:**
  Haemolytic transfusion reaction
  Haemolytic disease of the newborn (see Chapter 3.48)
  Hypersplenism
- **Malaria**
- **Secondary:**
  Renal failure
  Liver disease, etc.
- **Miscellaneous:**
  Chemicals, toxins and drugs
  Infections

3.24 Blood clotting disorders

Frank Hill

Minimum standards requirements

- Regional/national centre
- Prednisolone
- Immunisation; hepatitis B and C
- Blood clotting products

Factor deficiencies

The incidence of haemophilia is similar worldwide, 1 in 5–10 000 male births. Major advances have been made in separating haemophilia A (factor VIII deficiency) and haemophilia B (factor IX deficiency) and safe therapeutic intervention with replacement therapy. However, this is only available to the 20% of haemophiliacs who live in advantaged countries. For patients in poor countries, severe haemophilia continues to be a personal and social disaster with affected boys becoming progressively crippled during childhood from spontaneous painful intractable haemorrhages into muscles and joints. These boys die in childhood or early adult life. Severe deficiencies of the other coagulation factors (X, XI, VII, V, XIII, fibrinogen and von Willebrand's disease) are also associated with severe and at times fatal life-threatening haemorrhage.

- The largest barrier to providing replacement therapy is its high cost.
- There are also non-financial barriers including insufficient knowledge even among the medical community, lack of proper healthcare structure and low literacy.
- In the last decade WHO and WFH (World Federation of Haemophilia) have made considerable progress in setting up programmes in disadvantaged countries.
- WHO has identified as core components:
  - Training of care providers and establishing care centres
  - Identification and registration of people with haemophilia
  - Improving social awareness of haemophilia
  - Prevention of haemophilia
  - Providing safe therapeutic products
  - Developing a programme of comprehensive care.

How can delivery of haemophilia care be implemented in disadvantaged countries?

- National haemophilia societies are crucial. In addition to supporting affected families, they can lobby for support from the healthcare budget.
- WHO and WFH have visiting teams that have contributed to education and improvement through these national groups. They include international haemophilia training fellowships, workshops and twinning programmes, in order to transfer knowledge and diagnostic expertise to these embryo services.
- It is important that those planning healthcare fully appreciate that provision of laboratory diagnostic services for haemophilia and the development of safe blood transfusion services to provide safe replacement therapy will benefit a wide range of medical services.

How should the service be built and structured?

- At least one national centre should be created where the laboratory scientific and medical expertise exists to make an accurate diagnosis, which will then allow the appropriate counselling, including genetic counselling, of the patient's family (similar to cancer therapy; see Chapter 3.33). With advances in molecular biology, carriers of haemophilia can currently be identified and antenatal diagnosis provided so that a choice can be made to prevent the birth of haemophilic boys, particularly if treatment is not available.
- National registers should be set up for service planning.
- A clinical service involving paediatricians, dentists, orthopaedic surgeons and adult physicians, needs to be set up. Safe replacement therapy, probably initially derived from donated plasma, should be developed.
- Donor screening and product treatment to remove the risk of at least HIV and hepatitis B and C must be provided.
- Haemophiliacs should be vaccinated at an early age against hepatitis B and C.

What treatment should be given in the absence of replacement therapy?

Spontaneous haemorrhages into muscles and joints can be extremely painful and will lead to progressive crippling deformities. The acute episode must be managed with bed rest, for bleeds like knees splinting with a back slab to restrict movement may help. Analgesia for the pain is also required (see Chapter 1.27). Opiates may need to be used to obtain adequate pain relief. Bleeding with loss of first dentition may be severe enough to warrant blood transfusion.

Platelet deficiencies: idiopathic thrombocytopenic purpura (ITP)

- Isolated thrombocytopenia usually following a viral infection 1–3 weeks previously.
- Boys and girls equally affected and peak incidence aged 2–4 years.
- 90% chance of complete remission, but those presenting over the age of 10 years are more likely to have chronic ITP.
- Bleeding manifestations include petechiae, purpura, epistaxes, haematuria, gastrointestinal haemorrhage and rarely intracerebral haemorrhage. The child has no hepatosplenomegaly and is usually well.
- Other causes of thrombocytopenia must be excluded. If there is doubt, a bone marrow aspirate will show normal haemopoiesis with increased numbers of megakaryocytes.

Management

- Treatment is based on symptoms **not** platelet count and many patients require none.
- Petechiae on the head/neck, gastrointestinal and oral bleeding, are indicators for prednisolone (1–2 mg/kg/day after food in two divided doses for six weeks and reduce and stop irrespective of the platelet count if asymptomatic). Prednisolone does not alter the course of the disease. The time to remission is very variable.
- Chronic ITP with serious bleeding into the gastrointestinal tract or brain may require splenectomy. However, in disadvantaged countries there is a high risk of infection following splenectomy and long-term penicillin prophylaxis and pneumococcal vaccination are required.

3.25 Acute diarrhoea

Peter Sullivan

Minimum standards requirements

- ORS and ReSoMal
- IV fluids: 0.9% saline and Hartmanns
- Potassium: oral and IV
- Antibiotics: co-trimoxazole, amoxycillin, nalidixic acid, ciprofloxacin, cefotaxime, chloramphenicol, erythromycin, metronidazole, tetracycline, vancomycin, doxycycline

Introduction

Diarrhoeal diseases are a leading cause of childhood morbidity and mortality in disadvantaged countries. In 1999, 3 million children died of dehydration caused by diarrhoea, 80% of them in the first two years of their life. Around half of these deaths are due to watery diarrhoea and occur either because of lack of access to oral rehydration solution (ORS) or because of incorrect case management. About a third of deaths are caused by persistent diarrhoea and the remainder (approximately 15%) by dysentery.

Definition

Diarrhoea is the passage of loose or watery stools, usually at least three times in a 24-hour period. However, it is the consistency of the stools rather than the number that is most important. Mothers usually know when their children have diarrhoea and may provide useful working definitions in local situations. The volume of fluid lost through the stools in 24 hours can vary from 5 ml/kg (near normal) to 200 ml/kg, or more. Dehydration occurs when these losses are not replaced adequately and a deficit of water and electrolytes develops. The concentrations and amounts of electrolytes lost also vary. The total body sodium deficit in young children with severe dehydration due to diarrhoea is usually about 70–110 millimoles per litre of water deficit. Potassium and chloride losses are in a similar range.

The most common causes of dehydration are rotavirus, enterotoxigenic *Escherichia coli* (ETEC) and, during epidemics, *Vibrio cholerae* O1 or O139.

Most important issues

- Rehydration therapy
- Continued feeding
- Antibiotics **not** given routinely. Indicated in bloody diarrhoea (probable *Shigella*) and suspected cholera
- Antidiarrhoeal drugs and antiemetics should never be given and can be dangerous

Classification of diarrhoea

- **Acute watery diarrhoea** (including cholera), which lasts several hours or days: the main danger is dehydration; weight loss also occurs if feeding is not continued.
- **Acute bloody diarrhoea**, or dysentery: the main dangers are intestinal damage, sepsis and malnutrition; other complications, including dehydration, may also occur.
- **Persistent diarrhoea**, which lasts 14 days or longer: the main danger is malnutrition and serious non-intestinal infection; dehydration may also occur (see Chapter 3.26).
- **Diarrhoea with severe malnutrition** (marasmus or kwashiorkor). The main dangers are: severe systemic infection, dehydration, heart failure and vitamin and mineral deficiency (see Chapter 3.16).

Assessment of the child with diarrhoea

- Fever, vomiting, and loose stools are the common symptoms of acute gastroenteritis.
- RULE OUT OTHER SERIOUS ILLNESS, e.g. meningitis, malaria, bacterial sepsis, pneumonia, otitis media, and urinary tract infection.
- Assess for degree of dehydration, bloody diarrhoea, persistent diarrhoea, malnutrition and serious non-intestinal infections.

History

Specific points to enquire about in the history include:

- Duration of diarrhoea
- Presence of blood in the stool

- Local knowledge or reports of cholera epidemic
- Recent use of antibiotics
- Presence of fever, cough, or other important problems (e.g. convulsions, measles)
- Usual feeding practices
- Type and amount of fluids (including breast milk) and food taken during the illness
- Drugs or other remedies taken
- Immunisation history

Physical examination

The degree of dehydration is graded according to signs and symptoms that reflect the amount of fluid lost (Table 3.25.1). Infants with acute diarrhoea are more apt to dehydrate than are older children because they have a higher body surface-to-weight ratio, have a higher metabolic rate, and are dependent on others for fluid. Although the most accurate assessment of fluid status is acute weight change, the patient's premorbid weight often is not known.

In severe dehydration, prolonged skin retraction time, and decreased perfusion are more reliably predictive of dehydration than sunken fontanelle or absence of tears. A good correlation has been reported between time of capillary refill and fluid deficit. However, fever, ambient temperature and age can affect capillary refill time as well. **In severe dehydration, death follows soon if rehydration is not started quickly.** ✔

Children with some dehydration or severe dehydration should be weighed without clothing, when estimating their fluid requirements. If weighing is not possible, a child's age may be used to estimate the weight. **Treatment should never be delayed because a scale is not readily available. Also:** ✔

- Look for abdominal mass or abdominal distension.
- In an infant <1 week, diarrhoea is sometimes a sign of neonatal sepsis (see Chapter 3.48). In an infant blood in the stool may be an intussusception (see Chapter 3.49) or in the first week of life haemorrhagic disease of the newborn (see Chapter 3.48).
- **Remember:** Typhoid, surgical conditions such as intussusception, antibiotic-associated colitis, and rarely imflammatory bowel disease.

Investigations

Laboratory investigations are rarely needed. Serum electrolytes, especially sodium or potassium concentrations may be useful in severe dehydration. Stool cultures should be undertaken if at all possible in dysentery (bloody diarrhoea) but are not needed to initiate treatment in the usual case of acute watery diarrhoea. Stool microscopy is useful for diagnosing *Giardia lamblia*, *Cryptosporidium* and amoebic dysentery.

Principles of case management

Management includes two phases:

- rehydration and
- maintenance.

In the rehydration phase, the fluid deficit should be replaced and clinical hydration attained. In the maintenance phase, adequate dietary and fluid intake should be maintained. In both phases, excess fluid losses must be replaced continuously.

A child's fluid deficit can be estimated as follows:

- No signs of dehydration $<5\%$ fluid deficit: <50 ml/kg
- Some dehydration 5–10% fluid deficit: 50–100 ml/kg
- Severe dehydration $>10\%$ fluid deficit: >100 ml/kg

Rehydration therapy is based on degree of dehydration.

Note: use ReSoMal instead of standard ORS in children with severe malnutrition. ✔

- Mild dehydration ($3\% - 5\%$ fluid deficit)
  - Commence oral rehydration with 50 ml/kg over 2–4 hours.
  - The parent gives small amounts of fluid (for example one teaspoon) containing 50–90 mEq/litre of sodium (for example ORS) frequently.
  - Gradually increase the amount, as tolerated using teaspoon, syringe, medicine dropper, cup or glass.

Table 3.25.1 Clinical signs of dehydration

No dehydration ($<3\%$ weight loss)	Mild dehydration (3–5% weight loss)	Moderate dehydration (6–9% weight loss)	Severe dehydration (10+% weight loss)
NO SIGNS	Increased thirst	Loss of skin turgor, tenting when pinched	More pronounced effects than seen in moderate dehydration
	Slightly dry mucous membranes	Sunken eyes Sunken fontanelle in infants	Lack of urine output
		Restless or irritable behaviour	Hypovolaemic shock, including: a rapid and feeble pulse (the radial pulse may be undetectable), low or undetectable blood pressure, cool and poorly perfused extremities (decreased capillary refill >2 seconds) and peripheral cyanosis
		Dry mucous membranes	Rapid, deep breathing (a sign of acidosis)
			Altered consciousness or coma

- Reassess hydration after 2–4 hours, then progress to the maintenance phase or continue rehydration.

- Moderate dehydration (6–9% fluid deficit)
  - ORS should be administered as above.
  - The initial amount of fluid administered for rehydration is 100 ml/kg, given over 2–4 hours.
- Severe dehydration (≥10% fluid deficit, shock)
  - Start intravenous rehydration **immediately** (two intravenous lines if possible) or even cut down, femoral venous line, or intraosseous needle.
  - Boluses (see Table 3.25.3 or 10–20 ml/kg) of Ringer's lactate (Hartmann's) solution (the best if available) or 0.9% saline, should be administered until pulse, perfusion (**check capillary refill**), and mental status return to normal. **Do not use 5% glucose or 0.18% saline/4% glucose which can be dangerous if given quickly (hyponatraemia and cerebral oedema**).
  - When the child's level of consciousness returns to normal, he or she can take the remaining estimated deficit by mouth.
  - **Assess hydration status frequently.**

In acute diarrhoea **without dehydration**, omit the rehydration phase of therapy and start maintenance therapy immediately.

Practicalities of case management by WHO

Examine the child and select the appropriate WHO Treatment Plan.

- No signs of dehydration: WHO Treatment Plan A at home to prevent dehydration and malnutrition (see Appendix 7.3).
- Mild to moderate dehydration: WHO Treatment Plan B to treat dehydration (see below).
- Severe dehydration: WHO Treatment Plan C to treat severe dehydration urgently (see below).

Electrolyte disturbances

Knowing the levels of serum electrolytes rarely changes the management of children with diarrhoea. Indeed, these values are often misinterpreted, leading to inappropriate treatment. It is usually not helpful to measure serum electrolytes. The disorders described below are all adequately treated by oral rehydration therapy (ORT) with ORS solution.

Hypernatraemia

Some children with diarrhoea develop hypernatraemic dehydration, especially when given drinks that are hypertonic owing to their content of sugar (e.g. soft drinks, commercial fruit drinks) or salt. These draw water from the child's tissues and blood into the bowel, causing the concentration of sodium in extracellular fluid to rise. If the solute in the drink is not fully absorbed, the water remains in the bowel, causing osmotic diarrhoea.

Children with hypernatraemic dehydration (serum $Na^+ > 150$ mmol/litre) have thirst that is out of proportion to other signs of dehydration. Their most serious problem is convulsions, which usually occur when the serum sodium concentration exceeds 165 mmol/litre, and especially when intravenous therapy is given. Seizures are much less likely when hypernatraemia is treated with ORS solution, which usually causes the serum Na^+ concentration to become normal within 24 hours.

It is absolutely essential that intravenous rehydration does not lower the serum Na^+ too rapidly. Intravenous glucose solutions (5% glucose or 0.18% saline/4% glucose) are particularly dangerous and can result in cerebral oedema, usually fatal or permanently disabling.

Hyponatraemia

Children with diarrhoea who drink mostly water, or watery drinks that contain little salt, may develop hyponatraemia (serum $Na^+ < 130$ mmol/litre). Hyponatraemia is especially common in children with shigellosis and in severely malnourished children with oedema. Hyponatraemia is occasionally associated with lethargy and, less often, seizures. ORS solution is safe and effective therapy for nearly all children with hyponatraemia. An exception is children with oedema, for whom ORS solution may provide too much sodium. ReSoMal (see Chapter 3.16) may be helpful here.

Hypokalaemia

Inadequate replacement of potassium losses during diarrhoea can lead to potassium depletion and hypokalaemia (serum $K^+ < 3$ mmol/litre), especially in children with malnutrition. This can cause muscle weakness, paralytic ileus, impaired kidney function and cardiac arrhythmias. Hypokalaemia is worsened when base (bicarbonate or lactate) is given to treat acidosis without simultaneously providing potassium. Hypokalaemia can be prevented, and the potassium deficit corrected, by using ORS solution for rehydration therapy and by giving foods rich in potassium during diarrhoea and after it has stopped (bananas, coconut water, dark green leafy vegetables).

If it is necessary to give potassium intravenously (for instance serum $K^+ < 2.0$ mmol/litre or ECG signs of hypokalaemia: ST depression, T wave reduction and prominent U waves) then great care must be taken. In acute depletion, an infusion at the rate of 0.2 mmol/kg/hour can be used and the serum K^+ checked after 3 hours. The potassium for injection MUST be diluted before use and thoroughly mixed before being given. **The maximum concentration of potassium that can be given through a peripheral vein is 40 mmol/litre. The maximum infusion rate of potassium is 0.5 mmol/kg/hour.**

Note: The injectable form of KCl usually contains 1.5 g that is 20 mmol of potassium in 10 ml and can be given orally. The daily requirement of K^+ is 2.5−3.5 mmol/kg.

Replacement of ongoing fluid losses

During both rehydration and maintenance therapy, ongoing stool and vomit fluid losses must be replaced. If such losses can be measured accurately, 1 ml of ORS should be administered for each gram of diarrhoeal stool.

- Give 10 ml/kg or in older children a cup or small glass for each watery or loose stool passed, and 2 ml/kg of fluid for each vomit.
- Use either low-sodium ORS (containing 40–60 mEq/litre of sodium) or ORS containing 75–90 mEq/litre of sodium with an additional source of low-sodium fluid (for example breast milk, formula or water).

Vomiting

- Administer small, frequent volumes (for example 5 ml every minute) at first via spoon or syringe
- Gradually increase amount given
- Closely supervise

Dietary therapy

- **Breastfed infants:** Continue feeding on demand.
- **Bottle-fed infants:** Administer full-strength formulas immediately after rehydration (no longer than 4 hours). Lactose intolerance may develop and cause an exacerbation of diarrhoea with a lactose-containing formula, temporarily reduce or remove lactose from the diet.
- **Older children:** Continue their usual diet during diarrhoea. Recommended foods include starches, cereals, yoghurt, fruits and vegetables. Foods high in simple sugars and fats should be avoided. Excess fluid losses via vomiting or diarrhoea must be replaced with ORS.

Drug therapy: use of antimicrobials and "antidiarrhoeal" drugs

Antimicrobials should not be used routinely. This is because, except as noted below, it is not possible to distinguish clinically episodes that might respond, such as diarrhoea caused by enterotoxigenic E. coli, from those caused by agents unresponsive to antimicrobials, such as rotavirus or *Cryptosporidium*. Moreover, even for potentially responsive infections, selecting an effective antimicrobial requires knowledge of the likely sensitivity of the causative agent, information that is usually unavailable. In addition, use of antimicrobials adds to the cost of treatment, risks adverse reactions and enhances the development of resistant bacteria.

✔ **Antimicrobials are reliably particularly helpful only for children with bloody diarrhoea (probable shigellosis), suspected cholera with severe dehydration, and serious non-intestinal infections such as pneumonia. Antiprotozoal drugs are rarely indicated except as indicated below when a definite diagnosis is available.**

Antimicrobials for acute diarrhoea

Neonates

Diarrhoea and vomiting may be a symptom of septicaemia. If septicaemia is suspected parenteral antibotics are required (see Chapter 3.48).

Bloody diarrhoea

- **Bacterial causes:** *Campylobacter jejuni*, *Shigella sonnei*, *Sh. flexneri* and *Sh. dysenteriae*, and less commonly *salmonella*, *E. coli* O157:117 and *Aeromonas*.
- May be accompanied by abdominal pain and rectal prolapse.
- As culture facilities may not be available, sick, toxic children with bloody diarrhoea should be treated for shigella dysentery. Mild infections due to *Sh. sonnei* are usually self-limiting. *Shigella* in disadvantaged countries are commonly resistant to co-trimoxazole and ampicillin. Nalidixic acid, ciprofloxacin, ceftriaxone **or antibiotic of choice for the area**, should be used for a 5-day course. In infants with bloody diarrhoea due to infection, give ceftriaxone 75 mg/kg IV/IM once daily for 5 days.
- In infants and young children, exclude surgical causes (e.g. intussusception; see Chapter 3.49).

Salmonella

If non-typhoidal *Salmonella* is suspected in infants under 1 year of age or in immunocompromised children, blood cultures should be undertaken. If positive or the infant is toxic, an appropriate parenteral antibiotic should be given for example chloramphenicol or ceftriaxone or ciprofloxacin for 7–10 days. Look out for pneumonia or metastatic abscesses in bone, brain or elsewhere. Otherwise *Salmonella* gastroenteritis is not treated with antibiotics.

Note: Systemic *Salmonella* infection is common in malnutrition, HIV infection, sickle cell disease and schistosomiasis.

Campylobacter jejuni (also *Shigella* and *Salmonella*) may cause severe abdominal pain, mimicking a surgical emergency. Otherwise the disease is self-limiting and does not require antibiotics. If treatment is considered appropriate erythromycin (12.5 mg/kg 4 times daily) for 5 days is the antibiotic of choice.

Other causes of diarrhoea warranting antimicrobial treatment

- **Amoebic dysentery:** diagnosed by microscopy of fresh, warm stool. Treatment is metronidazole 50 mg/kg once daily (maximum dose = 2 g) for 5–7 days.
- **Cholera:** is usually only diagnosed during epidemics. If child has severe watery diarrhoea, suspect cholera or enterotoxigenic *E. coli* (only diagnosed by specialist laboratories). Treatment for cholera: tetracycline 50 mg/kg for 3 days in children >8 years. Alternative in young children is chloramphenicol 25 mg/kg 8 hourly for 3 days. In addition to rehydration, give antibiotic to which local strains of *Vibrio cholerae* are sensitive. These include tetracycline, doxycycline, co-trimoxazole, erythromycin and chloramphenicol.
- **Giardiasis:** diagnosed by microscopy of stool is usually self-limiting or asymptomatic. If symptomatic in a malnourished child or the disease is prolonged, it is justified to treat with metronidazole for 5 days (as for amoebic dysentery). Tinidazole is an alternative (50 mg/kg for 5 days).

- *Clostridium difficile* usually occurs after a course of antibiotics for some other illness and is associated with antibiotic-associated pseudomembranous colitis. Antibiotics, especially clindamycin, may alter the flora of the gastrointestinal tract and allow overgrowth of *C. difficile*. *C. difficile* produces a toxin which causes damage to gut mucosa resulting in pseudomembranous colitis. Confirmation is by culture of *C. difficile* in the faeces. Treatment is with oral vancomycin for 7–10 days which clears *C. difficile* from the gut. Dose: 10 mg/kg four times daily for older children (maximum dose in a day = 2 g).

✔ **"Antidiarrhoeal" drugs and antiemetics have no practical benefits for children with acute or persistent diarrhoea. They do not prevent dehydration or improve nutritional status, which should be the main objectives of treatment. Some, like loperamide, have dangerous, and sometimes fatal, side effects. These drugs should never be given to children below 5 years.**

Treatment of rectal prolapse

Gently push back using a surgical glove or wet cloth or if oedematous and cannot be reduced, warm compresses of magnesium sulphate may reduce the oedema.

Haemolytic uraemic syndrome

If lab tests are not available, suspect when purpura, pallor, altered consciousness and low or absent urine output are present. If lab tests are available, blood smear shows fragmented red cells and decreased or absent platelets. There will be an increase in blood urea and creatinine. (see Chapter 3.7).

Adapted WHO Treatment Plan B: oral rehydration therapy for children with some dehydration

Children with some dehydration should receive oral rehydration therapy with ORS solution in a health facility following the treatment plan described below.

How much ORS solution is needed?

Use Table 3.25.2 to estimate the amount of ORS solution needed for rehydration in the first four hours. If the child's weight is known, this should be used to determine the approximate amount of solution needed. The amount (in ml) may also be estimated by multiplying the child's weight in kg times 75. If the child's weight is not known, select the approximate amount according to the child's age.

The exact amount of solution required will depend on the child's dehydration status. Children with more marked signs of dehydration, or who continue to pass frequent watery stools, will require more solution than those with less marked signs or who are not passing frequent stools. If a child wants more than the estimated amount of ORS solution, and there are no signs of over hydration, give more.

Oedematous (puffy) eyelids are, in this context, usually a sign of overhydration. They may be a sign of chronic malnutrition. If this occurs, stop giving ORS solution, but give breast milk or plain water, and food. Do not give a diuretic. When the oedema has gone, resume giving ORS solution or home fluids according to Treatment Plan A (see Appendix 7.3).

How to give ORS solution

A family member should be taught to prepare and give ORS solution. The solution should be given to infants and young children using a clean spoon or cup. Feeding bottles should not be used. For babies, a dropper or syringe (without the needle) can be used to put small amounts of solution into the mouth.

Children under 2 years of age should be offered a teaspoonful every 1 to 2 minutes; older children may take frequent sips directly from the cup.

Vomiting often occurs during the first hour or two of treatment, especially when children drink the solution too quickly, but this rarely prevents successful oral rehydration since most of the fluid is absorbed. After this time vomiting usually stops. If the child vomits, wait 5–10 minutes and then start giving ORS solution again, but more slowly (for example a spoonful every 2–3 minutes).

Monitoring the progress of ORT

Check the child from time to time during rehydration to ensure that ORS solution is being taken satisfactorily and

Table 3.25.2 Guidelines for treating children with some dehydration: approximate amount of ORS solution to give in the *first 4 hours*

Age:	< 4 months	4–11 months	12–23 months	2–4 years	5–14 years	⩾15 years
Weight:	< 5 kg	5–7.9 kg	8–10.9 kg	11–15.9 kg	16–29.9 kg	30+ kg
Volume:	200–400 ml	400–600 ml	600–800ml	800–1200ml	1200–2200ml	2200–4000ml

Use the patient's age only when you do not know the weight. The approximate amount of ORS required (in ml) can also be calculated by multiplying the patient's weight in kg by 75.

If the patient wants more ORS than shown, give more.

Encourage the mother to continue breastfeeding her child.

For infants under 6 months who are not breastfed, also give 100–200 ml boiled/clean water during this period.

Note: During the initial stages of therapy, while still dehydrated, children can consume up to 20 ml per kg body weight per hour.

that signs of dehydration are not worsening. If at any time the child develops signs of severe dehydration, shift to Treatment Plan C (see below).

After 4 hours, reassess the child fully, following the guidelines above. Then decide what treatment to give next:

- If signs of severe dehydration have appeared, intravenous therapy should be started following Treatment Plan C. This is very unusual, however, occurring only in children who drink ORS solution poorly and pass large watery stools frequently during the rehydration period.
- If the child still has signs indicating some dehydration, continue ORT by repeating Treatment Plan B. At the same time start to offer food, milk and other fluids, as described in Treatment Plan A (see Appendix 7.3), and continue to reassess the child frequently.
- If there are no signs of dehydration, the child should be considered fully rehydrated. When rehydration is complete:
  - the skin pinch is normal
  - thirst has subsided
  - urine is passed
  - the child becomes quiet, is no longer irritable and often falls asleep.

Teach the mother how to treat her child at home with ORS solution and food following Treatment Plan A. Give her enough ORS packets for 2 days. Also teach her the signs that mean she should bring her child back.

Meeting normal fluid needs

While treatment to replace the existing water and electrolyte deficit is in progress the child's normal daily fluid requirements must also be met. This can be done as follows:

- Breastfed infants: Continue to breastfeed as often and as long as the infant wants, even during oral rehydration.
- Non-breastfed infants under 6 months of age: during the rehydration phase with ORS solution, give 100–200 ml of plain boiled water by mouth. After completing rehydration, resume full-strength milk (or formula) feeds. Give water and other fluids usually taken by the infant.
- Older children: throughout rehydration and maintenance therapy, offer as much plain boiled water to drink as they wish, in addition to ORS solution.

If oral rehydration therapy must be interrupted

If the mother and child must leave before rehydration with ORS solution is completed:

- Show the mother how much ORS solution to give to finish the 4-hour treatment at home
- Give her enough ORS packets to complete the four hour treatment and to continue oral rehydration for 2 more days, as shown in Treatment Plan A (see Appendix 7.3)
- Show her how to prepare ORS solution
- Teach her the three rules in Treatment Plan A for treating her child at home.

When oral rehydration fails or is not appropriate

In about 5% of children the signs of dehydration do not improve during ORT, or they worsen after initial improvement. The usual causes are:

- Continuing rapid stool loss (more than 15–20 ml/kg/hour), as occurs in some children with cholera
- Insufficient intake of ORS solution owing to fatigue or lethargy
- Frequent, severe vomiting.

Such children should be given ORS solution by nasogastric tube or Ringer's lactate solution (75 ml/kg IV in 4 hours), usually in hospital: **Watch carefully for fluid overload in particular pulmonary oedema**. After confirming that the signs of dehydration have improved, it is usually possible to resume ORT successfully. ✔

Rarely, ORT should not be given. This is true for children with:

- Abdominal distension with paralytic ileus, usually caused by opiate drugs (for example codeine, loperamide) and hypokalaemia
- Glucose malabsorption, indicated by a marked increase in stool output, failure of the signs of dehydration to improve and a large amount of glucose in the stool is rare and can be diagnosed using standard glucose oxidase impregnated strip (BM stick for example) applied to the stool. This will not diagnose lactose intolerance which requires a clinitest (modified Benedict's test) tablet when ORS solution is given.

In these situations, rehydration should be given IV until diarrhoea subsides; nasogastric therapy should not be used.

Giving food

Except for breast milk, food should not be given during the initial 4-hour rehydration period. However, children continued on Treatment Plan B longer than 4 hours should be given some food every 3–4 hours as described in Treatment Plan A. All children older than 4–6 months should be given some food before being sent home. This helps to emphasise to mothers the importance of continued feeding during diarrhoea.

Adapted WHO Treatment Plan C: intravenous rehydration therapy for patients with severe dehydration

The preferred treatment for children with severe dehydration is rapid intravenous rehydration. If possible, the child should be admitted to hospital. Guidelines for IV rehydration are given in Table 3.25.3.

Children who can drink, even poorly, should be given ORS solution by mouth until the intravenous drip is running. In addition, all children should start to receive some ORS solution (about 5 ml/kg/hour) when they can drink

without difficulty, which is usually within 3–4 hours (for infants) or 1–2 hours (for older patients). This provides additional base and potassium, which may not be adequately supplied by the intravenous fluid.

The most widely commercially available solution for use in intravenous rehydration is Ringer's lactate solution ($Na^+ = 131$ mmol/litre; $K^+ = 5$ mmol/litre; $HCO_3 = 29$ mmol/litre; $Ca^{2+} = 2$ mmol/litre) (also called Hartmann's Solution for Injection). It supplies an adequate concentration of sodium and sufficient lactate (which is metabolised to bicarbonate) for the correction of acidosis. The concentration of potassium is low and there is no glucose to prevent hypoglycaemia. This can be corrected by adding 100 ml of 50% glucose to 500 ml of Ringer's lactate solution giving approximately a 10% glucose solution (50 ml gives a 5% solution). It can be used in all age groups for the initial treatment of severe dehydration caused by acute diarrhoea of any aetiology.

If Ringer's lactate solution is not available, normal saline may be used but it does not contain a base to correct acidosis and does not replace potassium losses. Ringer's Lactate Solution already prepared with 5% glucose has the added advantage of providing glucose to help prevent ✔ hypoglycaemia. **Plain glucose (dextrose) solution should not be used since it does not contain electrolytes and thus does not correct the electrolyte losses or the acidosis. It does not effectively correct hypovolaemia and can produce dangerous hyponatraemia.**

Table 3.25.3 Guidelines for intravenous treatment of children with severe dehydration

Age	Ringer's lactate solution		
	First give 30 ml/kg in:		Then give 70 ml/kg in:
Infants (under 12 months)	1 hour	Repeat once if shock is still present	5 hours
Older child	30 minutes		2.5 hours

- Start IV fluids immediately. If the patient can drink, give ORS by mouth until the drip is set up. Give 100 ml/kg Ringer's lactate solution divided as indicated above
- Reassess the patient every 1–2 hours. If hydration is not improving, give the intravenous drip more rapidly.
- After 6 hours (infants) or 3 hours (older patients), re-evaluate the patient. Then choose the appropriate Treatment Plan A (see Appendix 7.3), B (above) or continue Treatment Plan C.

Monitoring the progress of intravenous rehydration

Patients should be reassessed every 15–30 minutes until a strong radial pulse is present and capillary refill is normal (< 2 seconds). Thereafter, they should be reassessed at least every hour to confirm that hydration is improving. If it is not, the intravenous drip should be given more rapidly.

When the planned amount of intravenous fluid has been given (after 3 hours for older patients, or 6 hours for infants), the child's hydration status should be reassessed fully.

Look and feel for all the signs of dehydration:

- If signs of severe dehydration are still present, **repeat** the intravenous fluid infusion as outlined in Treatment Plan C. This is very unusual, however, occurring only in children who pass large watery stools frequently during the rehydration period
- If the child is improving but still shows signs of some dehydration, **discontinue** the intravenous infusion and give ORS solution for 4 hours, as specified in Treatment Plan B (see above)
- If there are no signs of dehydration, follow Treatment Plan A (see Appendix 7.3). If possible, observe the child for at least 6 hours before discharge while the mother gives the child ORS solution, to confirm that she is able to maintain the child's hydration. Remember that the child will require therapy with ORS solution until the diarrhoea stops.

If the child cannot remain at the treatment centre, teach the mother how to give treatment at home following Treatment Plan A, give her enough ORS packets for 2 days and teach her the signs that indicate she should bring her child back.

What to do if intravenous therapy is not available

If intravenous therapy is not available at the facility, but can be given nearby (i.e. within 30 minutes), send the child immediately for intravenous treatment. If the child can drink, give the mother some ORS solution and show her how to give it to her child during the journey.

If intravenous therapy is not available nearby, health workers who have been trained can give ORS solution by nasogastric tube, at a rate of 20 ml/kg body weight per hour for 6 hours (total of 120 ml/kg body weight). If the abdomen becomes swollen, ORS solution should be given more slowly until the abdomen becomes less distended.

If nasogastric treatment is not possible but the child can drink, ORS solution should be given by mouth at a rate of 20 ml/kg body weight per hour for 6 hours (total of 120 ml/kg body weight). If this rate is too fast, the child may vomit repeatedly. In that case, give ORS solution more slowly until vomiting subsides.

Children receiving nasogastric or oral therapy should be reassessed at least every hour. If the signs of dehydration do not improve after 3 hours, the child must be taken immediately to the nearest facility where intravenous therapy is available. Otherwise, if rehydration is progressing satisfactorily, the child should be reassessed after 6 hours and a decision on further treatment made as described above for those given intravenous therapy.

If neither nasogastric nor oral therapy is possible, the child should be taken immediately to the nearest facility where intravenous or nasogastric therapy is available.

Oral rehydration solutions

The formula for oral rehydration salts (ORS) recommended by WHO and UNICEF is given in Table 3.25.4. The quantities shown are for preparation of one litre of ORS solution. The concentrations of the components of this solution are shown in Table 3.25.5.

When prepared and given correctly, ORS solution provides sufficient water and electrolytes to correct the deficits associated with acute diarrhoea. Potassium is provided to replace the large potassium losses associated with acute diarrhoea, especially in infants, thus preventing serious hypokalaemia. Citrate (or bicarbonate) is provided to prevent or correct base deficit acidosis. Glucose is essential because, when it is absorbed, it promotes the absorption of sodium and water in the small intestine. This is true irrespective of the cause of the diarrhoea. Without glucose, ORS solution would be ineffective.

Table 3.25.4 Composition by weight of WHO oral rehydration salts (ORS) to be dissolved in boiled water to produce 1 litre

Ingredient	Grams/litre
Sodium chloride	3.5
Trisodium citrate, dihydrate*	2.9 (or $NaHCO_3$ = 2.5 g/litre but is less stable)
Potassium chloride	1.5
Glucose, anhydrous#	20 (or monohydrate = 22 g/litre)

Note: The newer hypo-osmolar formulations of ORS can also be used and in some studies have been shown to be associated with a greater reduction in the frequency and volume of stools when compared with WHO ORS.

Table 3.25.5 Resulting molar concentration of components of WHO solutions

Component	ORS citrate (mmol/litre of water)	ORS bicarbonate (mmol/litre of water)
Sodium	90	90
Potassium	20	20
Chloride	80	80
Citrate	10	–
Bicarbonate	–	30
Glucose	111	111

Acknowledgements

This chapter was adapted from *Management of the patient with diarrhoea (revised 1992) third revision of WHO/CDD/SER/80. 2* and *The Treatment of Diarrhoea. A manual for physicians and other senior health workers WHO/CDR/95.3 10/95*

Further reading

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3.26 Post-infectious persistent diarrhoea

Zulfiqar A Bhutta

Minimum standards requirements

- ORS and ReSoMal
- IV fluids: 0.9% saline and Hartmanns
- Potassium: oral and IV
- Antibiotic: amoxycillin, gentamicin both IV
- Vitamin A
- Electrolyte and mineral mix
- Folic acid and later iron

Epidemiology

- Commonly defined as diarrhoea that starts acutely, but lasts for >14 days and is associated with growth faltering.
- Most cases are thus post-infectious in origin and other disorders such as inflammatory bowel disease and coeliac disease are therefore excluded.
- 4–20% of all episodes of diarrhoea in the disadvantaged world become prolonged, with associated case-fatality rates exceeding 50%.
- In parts of sub-Saharan Africa, the association of persistent diarrhoea with HIV infection is often the terminal event.

Risk factors for persistent diarrhoea

Appropriate case management of acute diarrhoea is key to the prevention of prolonged episodes.

- **Specific pathogens:** although several studies have identified an association between persistent diarrhoea and enteroaggregative *Escherichia coli* in the small bowel, this is by no means pathognomonic, nor is there a particular pattern of small bowel microbial colonisation or overgrowth seen in most cases. In HIV endemic parts of Africa an association of persistent diarrhoea with cryptosporidiosis is well recognised, but may represent a manifestation of immunodeficiency. Evidence from Bangladesh does suggest that recurrent bouts of infection with pathogens such as *Shigella* does lead to prolongation of the duration of successive diarrhoeal episodes.
- **Malnutrition:** persistent diarrhoea is commonly seen in association with significant malnutrition and the relationship may be bidirectional. It is widely recognised that diarrhoeal episodes, especially if invasive, may become prolonged in malnourished children. The recent evidence of micronutrient deficiencies, especially of zinc and vitamin A in malnourished children with persistent diarrhoea, indicates impaired immunological mechanisms for clearing infections as well as ineffective mucosal repair mechanisms.
- **Dietary risk factors:** while many children with persistent diarrhoea are lactose-intolerant, the role of specific dietary allergies in inducing and perpetuating enteropathy of malnutrition is unclear. Several studies have highlighted the high risk of prolonged diarrhoea with lactation failure and early introduction of artificial feeds in disadvantaged countries. In particular, the administration of unmodified cows' or buffalo milk is associated with prolongation of diarrhoea, suggesting the potential underlying role of milk protein enteropathy.
- **Inappropriate management of acute diarrhoea:** the association of prolongation of diarrhoea with starvation and inappropriately prolonged administration of parenteral fluids, has been recognised for over half a century. Unnecessary food withdrawal, and replacement of luminal nutrients, especially breast milk, with non-nutritive agents is a major factor in prolonging the mucosal injury after diarrhoea. In particular, blanket administration of antibiotics and any administration of antimotility agents should be avoided.

Principles of management of persistent diarrhoea

In general the management of persistent diarrhoea in malnourished children (Figure 3.26.1) represents a blend of the principles of management of diarrhoea and malnutrition (see Chapters 3.16 and 3.25). Associated malnutrition may be quite severe in affected children necessitating rapid nutritional rehabilitation, often in hospital. Given the chronicity of the disorder, prolonged hospitalisation may be quite problematic in disadvantaged countries, and

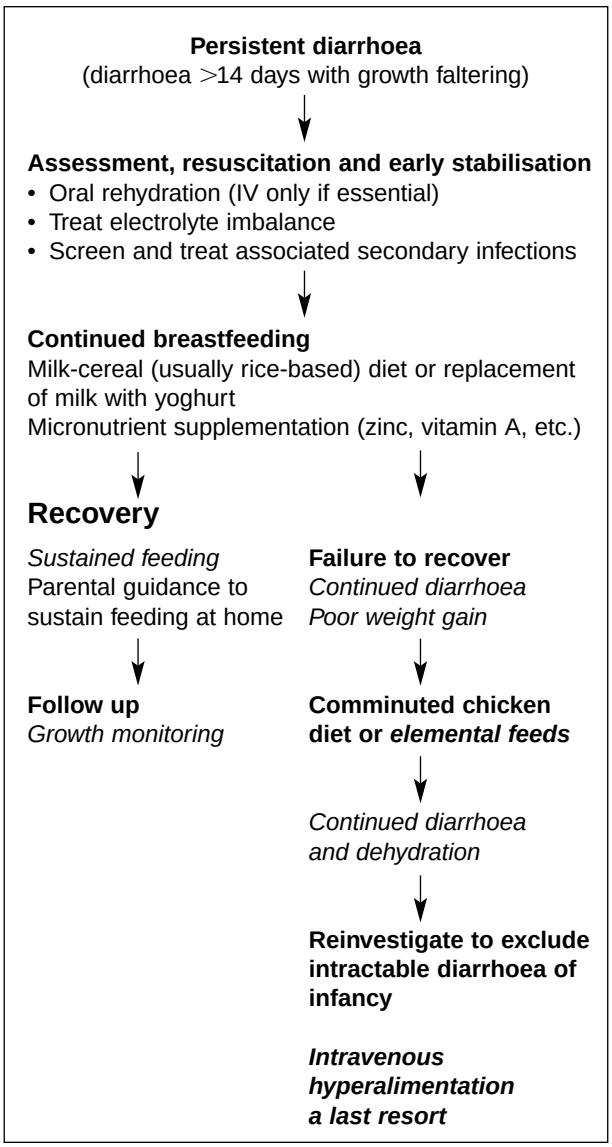

Figure 3.26.1 Outline of management.

whenever possible, ambulatory or home-based therapy must be stressed.

The following represent the basic principles of management of persistent diarrhoea and a suggested therapeutic approach is indicated in the Figure 3.26.1.

Rapid resuscitation and stabilisation

- Most children with persistent diarrhoea and associated malnutrition are not severely dehydrated and oral rehydration may be adequate.
- However, acute exacerbations and associated vomiting may require brief periods of intravenous rehydration with Ringer's lactate solution.
- Acute electrolyte imbalance such as hypokalaemia and severe acidosis may require correction (see Chapters 3.16 and 3.25).
- Associated systemic infections (bacteraemia, pneumonia and urinary tract infection) are well recognised in severely malnourished children with persistent diarrhoea and a frequent cause of early mortality. These must be screened for at admission. In severely ill children requiring hospitalisation, it may be best to cover with intravenous antibiotics at admission (usually **ampicillin** 25 mg/kg three times daily up to a maximum of 4g/day and **gentamicin** 2.5 mg/kg 8 hourly) while awaiting cultures.
- It should be emphasised that there is little role for oral antibiotics in persistent diarrhoea as in most cases the original bacterial infection triggering the prolonged diarrhoea has disappeared by the time the child presents.

Oral rehydration therapy

This is the preferred mode of rehydration and replacement of on-going losses. While in general the standard WHO oral rehydration solution (containing 90 mmol/litre of Na^+) is adequate, recent evidence indicates that hypo-osmolar rehydration fluids ReSoMal (containing 45 mmol/litre of Na^+) as well as cereal-based oral rehydration fluids may be advantageous in malnourished children. In general replacing each stool with about 50–100 ml ORS or ReSoMal is safe.

Enteral feeding and diet selection

- Most children with persistent diarrhoea are not lactose intolerant, although administration of a lactose load exceeding 5 g/kg/day is associated with higher rates of stooling and treatment failure. In general therefore withdrawal of milk and replacement with specialised (and expensive) lactose-free formulations is unnecessary.
- Alternative strategies of reducing the lactose load in malnourished children with persistent diarrhoea include addition of lactose free milk to cereals as well as replacement of milk with fermented milk products such as yoghurt. These measures have now been extensively evaluated in successive studies of the management of persistent diarrhoea in South Asia, and found to be extremely effective.
- It is rare to find persistent diarrhoea in breast-fed infants, **it must be stressed that breastfeeding must not be stopped under any circumstances.** ✔
- Rarely when dietary intolerance precludes the administration of cows' milk-based formulations or milk, it may be necessary to administer specialised milk-free diets such as a comminuted or blenderised chicken-based diet or ***an elemental formulation***. However, the latter may be almost unaffordable in most poor countries. A choice of enteral diets and formulations is given in Table 3.26.1.
- The usual energy density of any diet used for the therapy of persistent diarrhoea should be around 1 kcal/g, aiming to provide an energy intake of at least 110 kcal/kg/day and a protein intake of between 2–3 g/kg/day (in meals six times daily). Nasogastric feeding may be required during the first 2–3 days of care, particularly whilst infection is being treated.
- There should be at least three successive days of increasing weight before a response can be verified.
- Dietary failure is shown by an increase in stool frequency (>10 watery stools/day) or a failure to establish a daily weight gain within 7 days.

Table 3.26.1 Suggested composition of selected diets in children with persistent diarrhoea

Component	Khitchri (rice–lentils) (per 100 g)	Home made version of F75 diet (WHO) (per 1000 ml)	Comminuted chicken (per 100 g)	Semi-elemental diet (per 100 ml)
Protein	Mung lentils, 30 g	Dried skimmed milk, 25 g	Protein, 8 g	Protein, 2.25 g (hydrolysed)
Fat	Oil, 2 g	Vegetable oil, 27 g	Fat, 4 g	Fat, 1.65 g (medium chain triglycerides)
Minerals and micronutrients	Salt (to taste)	Vitamin mix, 140 mg Mineral mix, 20 ml	Electrolytes (sodium 0.4 mmol, potassium 1.3 mmol, calcium 0.2 mmol, phosphorus 1.5 mmol)	Electrolytes (sodium 1.9 mmol, potassium 2.3 mmol, calcium 1.8 mmol)
Carbohydrate	Rice, 60 g	Cereal flour, 35 g Sugar, 70 g		Caloreen, 5 g

- In selected circumstances when adequate intake of energy-dense food is problematic, the addition of amylase to the diet through germination techniques which increase the endogenous amylase content of foods may be helpful.

First diet: A starch-based, reduced milk concentration (low lactose) diet

The diet should contain at least 70 kcal/100 g, provide milk or yoghurt as a source of animal protein, but no more than 3.7 g lactose/kg body weight/day, and should provide at least 10% of kcalories as protein. The following example provides 83 kcal/100 g, 3.7g lactose/kg body weight/day and 11% of calories as protein:

- full-fat dried milk 11g (or whole liquid milk: 85 ml)
- rice 15 g
- vegetable oil 3.5 g
- cane sugar 3 g
- water to make 200 ml

Of the children who do not improve on this first diet, more than half will improve when given the second diet, from which the milk has been totally removed and starch (cereals) partly replaced with glucose or sucrose.

Second diet: A no-milk (lactose-free) diet with reduced cereal (starch)

The second diet should contain at least 70 kcal/100 g, and provide at least 10% of calories as protein (egg or chicken). The following example provides 75 kcal/100 g:

- whole egg 64 g
- rice 3 g
- vegetable oil 4 g
- glucose 3 g
- water to make 200 ml

Finely ground, cooked chicken (12 g) can be used in place of egg to give a diet providing 70 kcal/100 g.

Of the children who do not improve on this first diet more than half will improve when given the second diet, from which milk has been totally removed and starch (cereals) partly replaced with glucose or sucrose.

Micronutrient supplementation

Most malnourished children with persistent diarrhoea have associated deficiencies of micronutrients including zinc, iron and vitamin A. This may be a consequence of poor intake and continued enteral losses. It is therefore important to ensure that all children with persistent diarrhoea and malnutrition receive an initial dose of vitamin A orally or if that is not possible by deep intramuscular injection (<6 months 50 000 U, 6–12 months 100 000 U, >1 year 200 000 units. They should also receive a daily intake of the following for the next 2 weeks:

- A multivitamin supplement
- Folic acid (250 micrograms/kg on day 1, then 75 micrograms/kg/day)
- Zinc 3–5 mg/kg/day.
- Copper 0.3mg/kg/day
- Magnesium 0.2mmol/kg/day

While the association of significant anaemia with persistent diarrhoea is well recognised, iron replacement therapy should not be initiated until recovery from diarrhoea has started (ferrous sulphate 18 mg/kg/day or 6 mg/kg/day of elemental iron in 2 divided doses.

Follow up and nutritional rehabilitation

Given the high rates of relapse in most children with persistent diarrhoea, it is important to address the underlying risk factors and institute preventive measures. These include appropriate feeding (breastfeeding, complementary feeding) and close attention to environmental hygiene and sanitation. This poses a considerable challenge in communities deprived of basic necessities such as clean water and sewage disposal.

By the time they return home, children should be receiving a diet providing at least 110 kcal/kg/day (including milk and fresh fruit and well-cooked vegetables).

Further reading

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3.27 Upper gastroenterological disorders

Brian Coulter

Upper gastrointestinal disorders are not prominent amongst the general attendance of poor people at health centres and hospitals in disadvantaged countries. It may be that symptoms are under reported or overlooked because of more common problems such as gastroenteritis, persistent diarrhoea, intestinal helminths and malnutrition. However, certain life threatening conditions do occur including obstruction of oesophagus due to foreign body or strictures due to caustic poisoning, haematemesis due to peptic ulcer or portal hypertension and volvulus due to malrotation.

In better off communities, and particularly where ***facilities for upper gastrointestinal paediatric endoscopy*** are available, similar symptoms to those occurring in rich countries present, e.g. recurrent abdominal pain, epigastric and substernal pain, recurrent/persistent vomiting, dyspepsia and waterbrash/heartburn.

For causes of vomiting see section 2 (Chapter 2.10).

Gastro-oesophageal reflux

Gastro-oesophageal reflux (GOR) is a physiological condition in infants, children and adults but if severe may be pathological.

Table 3.27.1 Gastro-oesophageal reflux

Symptoms	Complications
Vomiting (regurgitation)	**Infants**
Waterbrash/heartburn	Apnoea
Nausea	Life-threatening event
Epigastric/retrosternal pain	**All ages** Failure to thrive* Aspiration pneumonia Haematemesis Anaemia Oesophageal stricture

* Particularly in children with cerebral palsy.

Note: Sandifer–Sutcliffe syndrome: dystonic posturing associated with GOR.

Diagnosis

- ***pH monitoring: grades the frequency and duration to which the lower oesophagus is exposed to acid (pH <4.0).***
- Barium swallow: is a much less sensitive method for diagnosis than pH monitoring. However, it is usually the only diagnostic facility available in low-income countries and will detect associated or other conditions, for example oesophageal stricture, hiatus hernia, diaphragmatic hernia and malrotation.

Management

- If child is thriving, the management is reassurance.
- Infants: positioning (30° head up from horizontal in lateral position), thickening of feeds with Carobel (Cow & Gate) or Nestargel (Nestlé).
- Moderate to severe GOR: the prokinetic agent cisapride is of value in some cases but has presently been withdrawn because of cardiac side effects, viz – lengthening of Q-T interval. Alternative drugs are metoclopramide (100–150 mg/kg 8 hourly) and domperidone (200–400 micrograms/kg every 4–8 hours). Though proof of efficacy is lacking, they may be effective in children with cerebral palsy.
- Oesophagitis: diagnosed by ***endoscopy and histology*** or by barium swallow (less sensitive). H_2 antagonists for example ranitidine (2–4 mg/kg twice daily, maximum 150 mg twice daily) or the proton pump inhibitor, omeprazole (700 micrograms–3 mg/kg/once daily) should be given.
- ***Surgery: Nissen fundoplication is undertaken in severe cases with failure of medical treatment, especially if there is severe oesophagitis, failure to thrive and aspiration pneumonia. It is commonly required in children with cerebral palsy and GOR.***

Helicobacter pylori

H. pylori is an ubiquitous bacteria which commonly infects the stomach (especially antrum) of children in poor countries from an early age. Child-to-child transmission is important. In rich countries up to 40–60% of adults are infected probably mainly during childhood. Conditions associated with *H. pylori* include:

- Chronic gastritis: often asymptomatic. Not a major cause of abdominal pain in children.

- Duodenal ulcer: *H. pylori* has a strong association with duodenal ulcer and must be eradicated to ensure healing.

Diagnosis

H. pylori can only be diagnosed by tests outlined below which are rarely available as a routine in low-income countries.

- ***Serology: good for epidemiological studies but reduced sensitivity in children under 7 years.***
- ***Urea breath test (^{13}C-UBT): sensitive and specific especially in children >6 years.***
- ***Endoscopy: histological demonstration and culture of H. pylori.***

Management

Selection of optimal antibacterial agents is difficult because of development of resistance.

Suggested regimen

Omeprazole (700 micrograms–3 mg/kg once daily up to a maximum of 40mg) plus metronidazole (7.5 mg/kg three times daily) **or** clarithromycin (7.5 mg/kg twice daily) plus amoxycillin (<1 year, 62.5 mg; 1–4 years, 125 mg; 5–12 years, 250 mg; >12 years, 250–500 mg; all three times daily).

Duodenal ulcer

Duodenal ulcers are uncommon in children but can be life threatening due to haematemesis, melaena and perforation. There is commonly a family history. Common symptoms include **nocturnal waking by pain** and epigastric pain relieved by food or antacids.

Diagnosis

- ***Endoscopy including biopsy for H. pylori is the optimal method.***
- Barium swallow: less sensitive in diagnosing acute ulceration and better at detecting scarring.

Management

- Unless facilities to diagnose *H. pylori* are available all children should be treated for eradication of presumed *H. pylori*.
- H_2 antagonists for 6–8 weeks:
  ranitidine 2–4 mg/kg twice daily (maximum 150 mg twice daily)

Non-specific upper gastrointestinal pain

Where duodenal ulcer is not considered to be the cause of upper gastrointestinal pain or dyspepsia, Gaviscon or other antacids may be used on a regular or as-required basis.

3.28 Inflammatory bowel disease

Brian Coulter

Minimum standards requirements

- Aminosalicylates
- Prednisolone
- Hydrocortisone
- Steroid enemas
- Blood transfusion
- Elemental diet
- Metronidazole

Inflammatory bowel disease (IBD) is uncommon in children in disadvantaged countries. Abdominal tuberculosis is more common. However, in the UK about 18% of children with IBD are non-white of whom most are either of Indian or Caribbean origin. Although IBD may present in infancy the mean age in the UK is approximately 12 years. Crohn's disease is more than twice as common as ulcerative colitis. A family history is common.

Diagnosis

- Clinical diagnosis of ulcerative colitis is usually straightforward. Crohn's disease may have a wide variety of symptoms, especially extraintestinal. Iron-deficiency anaemia is common in both.
- Interval between onset of symptoms and diagnosis is often over 6 months in Crohn's disease and may be 2–3 months in ulcerative colitis. Denial of symptoms is common especially in adolescents.

Investigations

- Growth parameters and investigations are a guide to the severity and duration of disease and nutritional state of the child.
- Examination of mouth and anus is essential.
- Stool examination is essential to exclude bacterial and parasitological causes of diarrhoea.
- Normal investigations, especially acute phase reactants (erythrocyte-sedimentation rate or C-reactive protein), do not exclude IBD.
- Children with Crohn's disease and severe involvement of the colon may present similarly to ulcerative colitis.

Table 3.28.1 Comparison between Crohn's disease and ulcerative colitis

Feature	Ulcerative colitis	Crohn's disease
Pathology	Mucosal disease	Transmural disease, skin lesions, strictures, fistulae
Site	Rectocolonic (rectum always involved)	Ileocolic: 50–60% Small intestine: 30–35% Colon: 10–15%
Common presenting symptoms	Diarrhoea mixed with blood/mucus Pain – lower abdominal, weight loss	Pain – right iliac fossa Diarrhoea with or without blood Growth failure Perianal disease
Extraintestinal features*	Uncommon	Common

* Oral ulcers, finger clubbing, arthritis, skin disorders, fever.

General investigations

- **Stool**
  Blood, mucus
  Microscopy for *E. histolytica*, *Schistosoma*, *Trichuris trichuria*, *G. lamblia*
  Culture for bacteria (see Chapter 3.25)
- **Full blood count**
  Hb ↓
  WBC ↑
  Platelets ↑
- **Acute phase reactants**
  Erythrocyte-sedimentation rate
  C-reactive protein
  α_1 ***glycoprotein***
- **Chemical pathology**
  Electrolytes (if diarrhoea severe)
  Ferritin (may be spuriously raised – acute phase reactant)
  Albumin

Specific investigations

- Specific investigations depend on availability of paediatric gastrointestinal facilities. Sigmoidoscopy is essential. ***Flexible endoscopy of lower and upper gastrointestinal tract is ideal.*** Barium enema (double contrast) is required in colitis **only** if colonoscopy is not available.
- Normal macroscopic appearance of lower or upper gut **does not exclude IBD. Histology is essential.**
- "Indeterminate colitis" is a term to describe patients whose histology is not typical of ulcerative colitis or Crohn's Disease. They are usually treated initially as ulcerative colitis.

Table 3.28.2 Specific investigations: Crohn's disease and ulcerate colitis

Feature	Ulcerative colitis	Crohn's disease
Endoscopy*	Proctoscopy Sigmoidoscopy Colonoscopy	Lower gut ***Upper gut****
Radiological studies	Barium enema† (double contrast)	Barium meal and follow through
White cell scan‡ (technetium labelled)	Screening	Screening

* Depending on availability.
† Only required if colonoscopy unavailable.
‡ Only available in rich countries.

Management

Management of ulcerative colitis

- Initial management depends on severity.
- Follow up: parents and older children should be taught and understand how to recognise and treat any relapse promptly.

Management of active colitis

- **Mild disease**: less than four motions/day, intermittent blood, normal acute-phase reactants, no toxicity
  - Aminosalicylates
  - Corticosteroid or mesalazine enema until bleeding stops and then alternate nights for 1 week
  - Corticosteroids orally if no response within 2 weeks
- **Moderate disease**: four to six motions/day, moderate blood, slight toxicity, anaemia and raised acute-phase reactants
  - As above. Oral corticosteroids are given in the higher dose. If poor response, treat as for severe disease.
- **Severe disease**: more than six blood motions/day, toxicity, fever, anaemia and hypoalbuminaemia
  - Intravenous pulse methylprednisolone or hydrocortisone for 3 days
  - Rectal hydrocortisone or prednisolone enemas twice daily
  - Antibiotics, for example metronidazole (benefit not proven)
  - Intravenous fluids, correction of electrolyte deficits
  - Blood transfusion
  - ***Intravenous nutrition if there is nutritional deficiency and slow response***
  - Toxic dilation: if no response to intensive therapy by 12–24 hours undertake colectomy
  - ***Intravenous cyclosporin may be of value if no response to intravenous corticosteroids***

Relapse

Prompt commencement of rectal corticosteroids or mesalazine enema. If no response give course of oral corticosteroids.

Maintenance

- Aminosalicylic acid preparations are generally given life-long.
- If relapses occur when steroids are reduced, ***give azathioprine for up to 3–5 years***. Regular monitoring of blood count every 1–2 months is important.

Indicators for colectomy

- Toxic megacolon (see above), intractable disease, growth failure.
- Risk of cancer relates to extent of disease and duration. Good maintenance therapy is important in prevention. Two-yearly colonoscopy should be considered in those with pancolitis for 10 years after commencement of disease.
- Proctocolectomy with ileoanal pouch anastomosis is curative and successful in 90% of cases if **performed by an experienced surgeon**.

Management of Crohn's disease

- Key to management is to maintain growth and nutrition and control symptoms.
- Most children will have recurrent relapses.
- Many will require surgery at some stage.
- Nutritional treatment and support.

Elemental diet

An elemental diet is effective in producing remission especially in small bowel disease. Advantages over corticosteroids are the positive effect on growth. Because of unpalatability it usually has to be given by nasogastric tube. The diet is given for 6 weeks, during which time no other food is given and then gradually reduced over 6 weeks while a hypoallergenic, then normal diet is re-introduced.

Elemental or polymeric diet may be given intermittently by nightly nasogastric or gastrostomy tube to maintain growth.

Drug therapy

See Table 3.28.3 for drug dosage in ulcerative colitis.

- Prednisolone 1–1.5 mg/kg/day (usually around 40 mg/day) is effective in small and large bowel

Table 3.28.3 Drug dosages for ulcerative colitis

Agent	Dose
Corticosteroids	
Prednisolone	1–1.5 mg/kg/day (approx 40 mg) for 2 weeks or until pain and rectal bleeding settle, then reduce to 30 mg/day for 1 week, then by 5 mg/day
Methylprednisolone	IV 1–1.5 mg/kg/day
Hydrocortisone	IV 4 mg/kg 6 hourly
Prednisolone enema or foam (20 mg in 100 ml)	50–100 ml at night
Aminosalicylates	
Sulphasalazine (tablets 10 mg and 50 mg)	10 mg/kg 4–6 hourly for acute episodes. Decrease dose by half for maintenance as soon as possible. Urine and tears will go orange. Report sore throat
Mesalazine	10–15 mg/kg three times daily
Metronidazole	10 mg/kg three times daily
Azathioprine	1.5–3 mg/kg/once daily (or 60 mg/m^2 once daily)

disease. Continue until remission occurs, then reduce slowly by 5 mg/kg/week and **slowly** tail off. If required to maintain remission, alternate-day therapy may have fewer side effects.

- Budesonide (9 mg/day in those >12 years) may be as effective as prednisolone for ileocolic disease and have less side effects.
- Mesalazine but not sulphasalazine appears to be effective for treatment and maintenance in ileal as well as colonic disease (dose 10–15 mg/kg/three times daily).
- ***Azathioprine is effective in long-term maintenance and has steroid-sparing effects. It may be useful for healing fistulae. It takes many months to act. It should be continued for at least 4 years. Blood counts should be undertaken every 1–2 months.***
- Metronidazole may be effective in controlling perianal disease and fistulae. It may also reduce small bowel overgrowth.

Surgery

Indications for surgery include: failure of medical therapy, obstruction and growth failure. Strictureplasty may be an effective method to avoid excision of bowel when structures are present.

Follow up and support for IBD

Patients and their families require long-term understanding and support. Psychological therapy may be helpful in some cases.

3.29 Gastrointestinal bleeding

Brian Coulter

Causes of bleeding from the gastrointestinal tract are many and relate to the age of the child. A good history and clinical examination are essential and will indicate specific investigations.

- In haematemesis it is important to exclude swallowed blood due to disorders of the nose and mouth.
- In infants the commonest cause of rectal bleeding is anal fissure. "Rectal bleeding" may amount to only appearance of a small amount of blood on the lavatory paper.
- Melaena has to be differentiated from dark stools associated with medication, for example iron preparations, and colouring from foods. A heavy infestation of *Enterobius vermicularis* may be seen on lower bowel endoscopy. Whether this is coincidental or a cause of rectal bleeding may not be known.

Table 3.29.1 Causes of gastrointestinal haemorrhage

Site	Clinical features
*Upper gut**	
Poisoning or treatment with salicylates	"Coffee ground" vomit
Mallory–Weiss syndrome	
Oesophagitis/gastro-oesophageal reflux	See Chapter 3.27
Duodenal or gastric ulcers	
Portal hypertension/varices	Hepatosplenomegaly (Chapter 3.10)
	Schistosomiasis (Chapter 4.30)
Midgut	
Intussusception, volvulus	Infants, "redcurrant jelly" stools (Chapter 3.49)
Meckel's diverticulum*	Often painless
Colorectal	
Infection, for example shigellosis, amoebiasis, schistosomiasis	Diarrhoea – blood/mucus mixed with stool.
Inflammatory bowel disease (IBD)	Abdominal pain
Milk protein intolerance	Eosinophils elevated in blood ***and on histology***
Polyps (single, multiple, familial, sometimes in jejunum – Peutz–Jeghers syndrome)	Blood separate or surrounding normal stools
Anus	
Fissure	Infants, constipation, local pain, "holding stool", and tags
Crohn's disease	***See*** Chapter 3.28
Miscellaneous	
Necrotising enterocolitis, duplication cyst, Henoch–Schönlein purpura, AIDS, thrombocytopenia, malignancy, connective tissue disorders	

* Large bleed from upper gastrointestinal tract may present as "red blood" in stool as well as melaena.

Table 3.29.2 Investigations for gastrointestinal bleeding

History/examination	Looking for
History	
Acute/chronic, amount of blood	Severity
Endemic area	Schistosomiasis
Haematemesis	Upper gastrointestinal disorder
Nose, mouth disorders	Swallowed blood
Pain – site	Upper or lower gastrointestinal tract
Stool	
hard, loose	Constipation/diarrhoea
blood mixed	Inflammation/infection
blood surrounding	Anal fissure, polyp
colour	Fresh blood/melaena
Inflammatory bowel disease – patient/family	See Chapter on IBD (3.28)
Bleeding tendency	Thrombocytopenia, malignancy
Examination	
Nose, mouth	Swallowed blood
Pallor, capillary refill, blood pressure	Anaemia, shock
Petechiae, telangiectasia	Thrombocytopenia, hereditary telangiectasia
Abdomen	Tenderness, hepatosplenomegaly
Anus	Fissures, tags, infection

Investigations

Depends on suspected site of bleeding and clinical features. See appropriate chapters.

- Essential to include:
  - FBC, group/cross-match (if necessary)
  - Serum ferritin/iron
  - Stool
    - direct examination – blood, mucus
    - microscopy – RBC, WBC, ova/parasites and culture
    - occult blood
- ***Isotope scan: required for diagnosis of Meckel's diverticulum***
- Barium studies
- ***Endoscopy***

3.30 Malabsorption and coeliac disease

Brian Coulter

Malabsorption

Common causes of malabsorption in low-income countries include severe malnutrition, persistent diarrhoea and HIV infection. None of these require bowel investigation. The main emphasis is on nutritional rehabilitation which regenerates the small bowel atrophy and the immune system (see management of persistent diarrhoea, and severe malnutrition (Chapters 3.16 and 3.26)). Only a limited response to nutritional support is expected in HIV infection depending generally on stage of disease.

Other causes are outlined in Chapters 2.3 and 2.11.

Coeliac disease

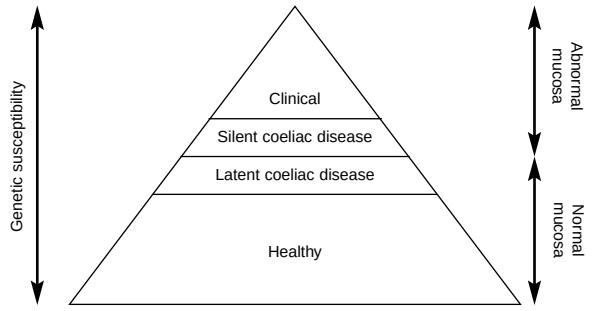
Coeliac disease is due to intolerance to wheat gliadin. It occurs in Caucasians whose diet contains substantial amounts of wheat. It mainly affects those of European origin, and also occurs in North Africa and the Middle East, and is described in the Punjab of the Indian subcontinent where wheat is a staple.

Frequency

The spectrum of coeliac disease is outlined below.

Although the gene may be present in up to 1:300 of some populations, clinical disease is manifest in approximately 1:1000 (range 1:250–1:4000).

The classical symptoms of steatorrhoea are more common in young children. In older children and adolescents coeliac disease may manifest in a wide variety of ways.



Latent coeliac disease: ***positive antigliadin antibodies***, normal small bowel but risk of developing disease

Figure 3.30.1 The spectrum of coeliac disease

Clinical Features

Table 3.30.1 Clinical features of coeliac disease

Under 2 years	Over 2 years
Steatorrhoea	Short stature
Vomiting	Delayed puberty
Abdominal distension	Iron-resistant anaemia
Irritability	Rickets/osteomalacia
Anorexia	Behaviour problems
Growth failure	With or without the gut disorders that occur in younger children

Investigations of malabsorption

Specific investigations in the diagnosis of coeliac disease are IgA antigliadin and IgA endomysial antibodies and jejunal biopsy using a Crosby or Waton capsule, ***or duodenal (second part) biopsy by endoscopy. Measure serum IgA as low levels (occurs in 1:700 population) will invalidate the above IgA antibody tests.***

Table 3.30.2 Investigations for malabsorption

General	Specific
FBC+film	***Immunoglobulins***
Ferritin	***IgA antigliadin antibodies****
Folate (RBC)	***IgA endomysial antibodies****
Vitamin B_{12} (especially in Crohn's disease)	***Small bowel biopsy****
Albumin	Villus atrophy
Hydrogen breath test	Hyperplasia of crypts
	Increased inflammatory cells
Permeability tests (for example mannitol + lactulose, xylose)	

* Child must be on a gluten-containing diet for these investigations to be valid.

Diagnosis

- ***Histology compatible*** with coeliac disease
- Clear response to gluten-free diet, that is symptoms resolve and weight and/or height increase.

- No further biopsy required.
- Diet is for life. This prevents small intestine lymphoma.
- Wheat, barley and rye should be excluded. Moderate amounts of oats, also maize, sorghum and rice are tolerated.

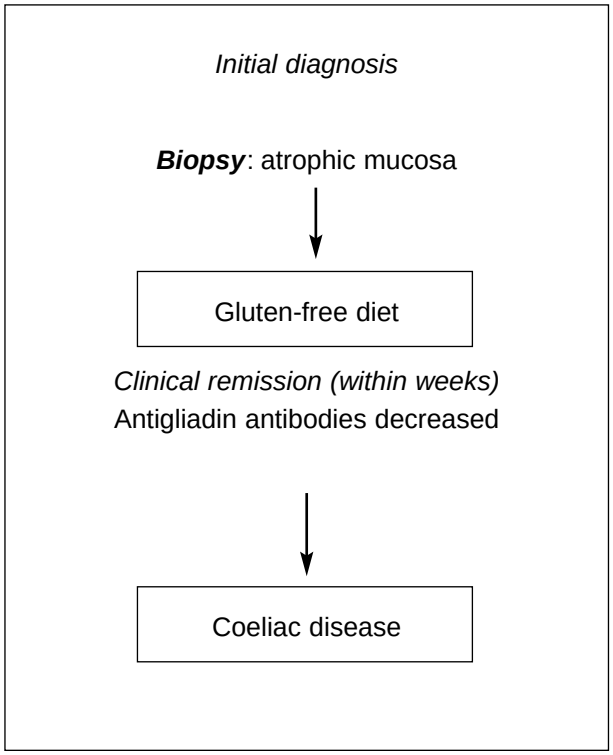


Figure 3.30.2 Diagnosis of coeliac disease

Gluten challenge

Do a gluten challenge if:

- Diagnosis is in doubt. Other diagnoses are possible, especially in children <2 years, for example milk protein intolerance, persistent diarrhoea, giardiasis
- Response to gluten-free diet is not clear-cut
- Asymptomatic subjects, for example, first-degree relatives.

Note: Gluten challenge should preferably be undertaken when child is over 6 years and before puberty to reduce effects on dentition and growth.

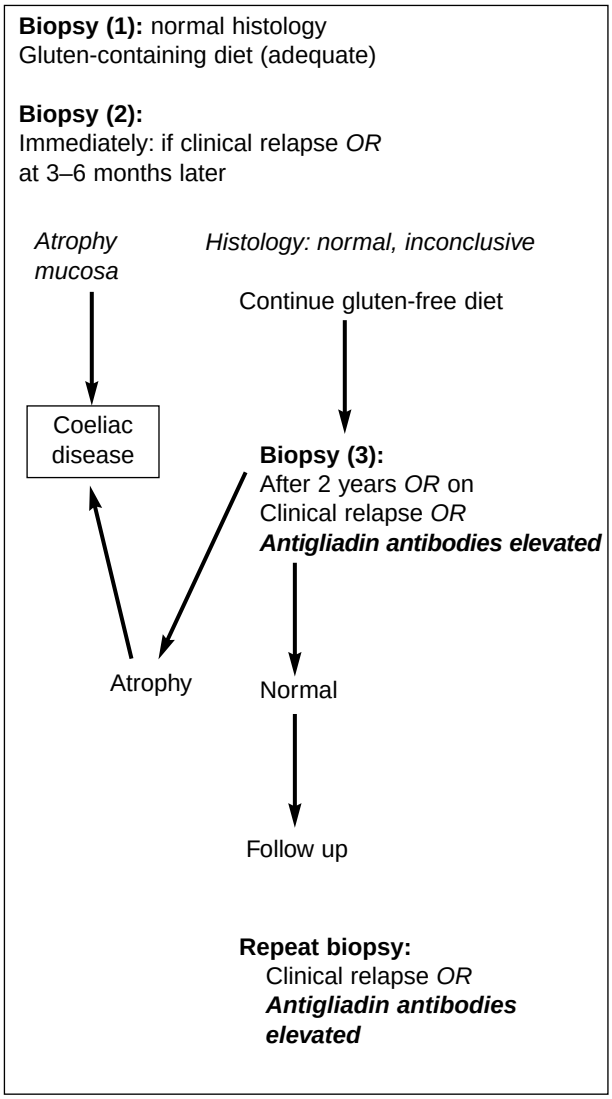


Figure 3.30.3 Gluten challenge

3.31 Constipation

Susan K Bunn

Minimum standards requirements

- Docusate
- Lactulose
- Senna or sodium picosulphate
- Glycerine suppositories
- Small volume citrate enemas

Introduction

Definition

Difficulty, delay or pain on defaecation.

This is one of the conditions referred to hospitals in rich countries. It is also common in the better-off families in poor countries. It is associated with low-roughage diets.

Normal defaecation patterns

- Breast-fed babies average three and formula-fed two stools per day.
- Children average one stool per day after 3 years but the normal range is once on alternate days to three times daily.

Diagnosis

- Faecal masses palpable abdominally; often in left and right iliac fossae, but sometimes suprapubically.
- Anal tags and fissures.
- Evidence of sexual abuse.
- Hard impacted faeces on rectal examination although only really necessary if there is a suspicion of Hirschsprung's disease (rectal examination should be done with topical lignocaine jelly (1%) if anal fissures are present, and aborted if too painful)
- Faecal masses on abdominal *X* ray (abdominal *X* ray should rarely be necessary).

Aetiology

The vast majority of constipation in childhood is "idiopathic" (see below).

Common causes of constipation

These include inadequate food or fluid intake and in young children, excessive cows' milk.

Uncommon causes

These include:

- **Neurogenic constipation**: due to spinal cord lesions – excluded by a normal neurological examination
- **Anal lesions giving pain or creating obstruction**: anal fissures most commonly, perianal skin infections, rarely congenital anterior anus and anal stenosis, rarely but must not be missed sexual abuse – excluded by superficial perineal examination
- **Hirschsprung's disease**: suspect when there is infancy onset constipation, delay (>48 hours) in passing meconium, failure to thrive, vomiting, abdominal distension, alternating constipation and diarrhoea, surprisingly little soiling for degree of constipation and an explosive gush of faeces when the examining finger is withdrawn at rectal examination
- **Endocrine conditions**: hypothyroidism, renal tubular acidosis, diabetes insipidus, hypercalcaemia – suspect if combination of failure to thrive and constipation especially with pellet stools
- **Family problems**: Coercive potty training.

Pathophysiology

When the rectum becomes chronically obstructed or incompletely emptied it can enlarge to give a megarectum. This leads to diminished urgency to defaecate leading to soiling (which is involuntary and not the deliberate act by the child, in contrast to encopresis where the child voluntarily passes normal stools in unacceptable places).

Management of idiopathic constipation

Understanding the aetiology and sequence of events in developing chronic constipation is crucial in successful physical and psychological management (Figure 3.31.1).

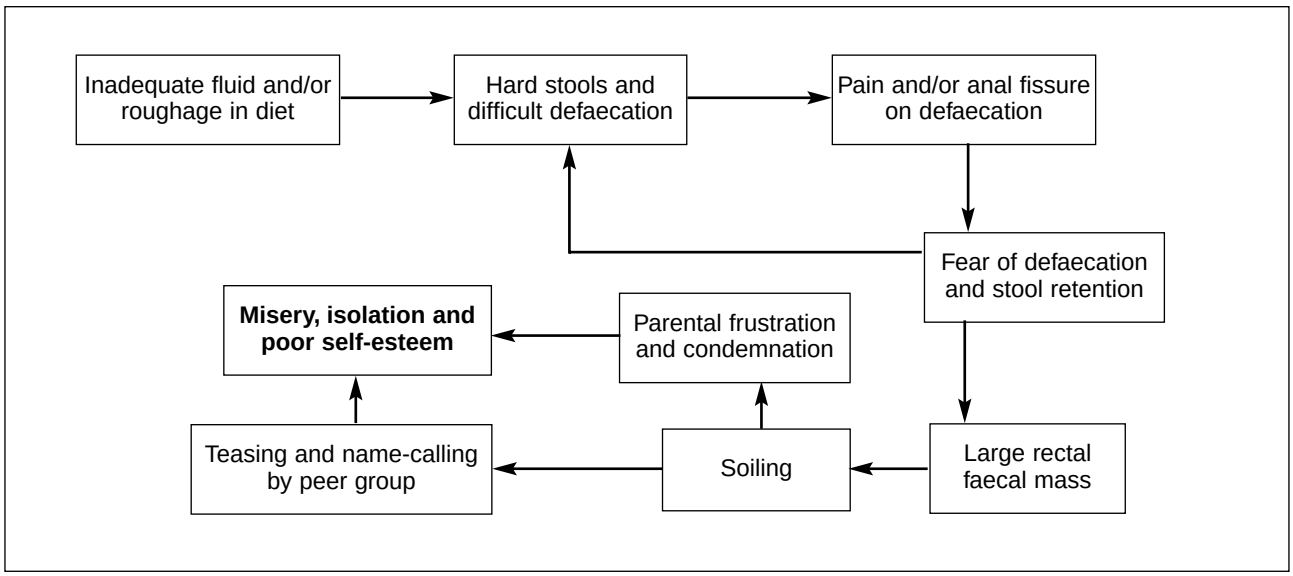


Figure 3.31.1 Flow diagram demonstrating the usual sequence of events in the development of idiopathic constipation and soiling. Each and every element of this flow diagram should be addressed and treated if management is to be fully successful.

1. **Explanation**
   Careful and thorough explanation of the problem to the parents and child. Emphasise that soiling is not purposeful and the child needs support not condemnation. Assess the need for psychological as well as physical treatment.
2. **Evacuation of retained faeces**
   To soften and lubricate retained mass give docusate sodium according to age at a dose of: <5 years 30 mg twice daily, 5–10 years 60 mg twice daily, >10 years 60 mg in the morning, 120 mg at night for 48 hours (this can be lengthened in children with very hard or large faecal masses). Following this and in order to expel the retained mass, give a stimulant laxative for example sodium picosulphate (<5 years 5 mg at night, 5–10 years 10 mg at night, >10 years 10 mg twice daily). This should be maintained for 48 hours or until the faecal mass has been cleared clinically. If picosulphate is not available a large dose of senna can be tried, but may need to be used for longer. For example, senna: <5 years 15 mg at night, 5–10 years 30 mg at night, >10 years 30 mg twice daily. Senna is available as tablets (7.5 mg sennoside) or liquid (7.5 mg sennoside in 5 ml).

   Only if the above fails give suppositories (glycerine) once daily (infant 1 g, child <12 years 2 g, and child >12 years 4 g).

   If the oral and suppository methods are unsuccessful, excessive abdominal pain develops and/or there is vomiting then stimulant enemas will be required. Phosphate enemas should not be used <2 years. For children 2–10 years give 60 ml (1/2 phosphate enema) and >10 years 120 ml (full enema). If not available a small-volume sodium citrate enema (micro-enema) can be used. However, the use of enemas can add to the child's fear of defaecation. ***The child should never be forcibly held down to receive an enema***. Give enemas **once**/day in the morning. Most children need 2–3 enemas to clear faecal mass.

   If these measures fail, the child should have manual evacuation of faecal mass under general anaesthetic (ONLY IF THIS IS SAFE AND AVAILABLE) with an anal stretch performed at the same time.
3. **Establish a regular and effective pattern of defaecation to prevent recurrence**

- Bulking agents for example lactulose (<1 year 2.5 ml twice daily, 1–5 years 5 ml twice daily, 5–10 years 10 ml twice daily and >10 years 15 ml twice daily)
- Stimulant laxatives usually senna (<2 years 0.25–0.5 ml/kg once daily; 2–6 years 5 ml daily; >6 years 10–20 ml or 1–4 tablets daily) with the aim of causing initial loose stools and then reducing dose to induce at least one soft stool per day. Often large doses will be required initially. Senna is available as tablets 7.5 mg and liquid 7.5 mg/5 ml.

Encourage increased fluid intake and high roughage diet (fruit, vegetables and cereals). If possible apply behavioural treatment with positive reinforcement of toileting, the passage of stool in the toilet and the absence of soiling.

Patients are likely to require at least one year of stimulant laxatives, and often several years of continuous or intermittent bulking laxatives.

3.32 Connective tissue disorders

Taunton R Southwood

Overview

Making a diagnosis of a rheumatic disease in a child relies primarily on clinical skill and experience, as there are few diagnostic laboratory tests. Although these diseases are rare in children, symptoms that raise the possibility of rheumatic disease are common. Rheumatic symptoms may be relatively specific, such as joint swelling, or relatively non-specific, such as fever, lethargy, pallor, anorexia, failure to thrive, muscle weakness, musculoskeletal pain, skin rash, headache and abdominal pain. The interpretation of these clinical features requires a meticulous approach to characterising the nature of each feature and considering the overall pattern of all the clinical features in the individual patient. The aims of this chapter are to assist in the recognition of rheumatic patterns of clinical features and to provide guidance for appropriate treatment and monitoring of rheumatic disease in children.

Rheumatic fever

Minimum standards requirements: rheumatic fever

- Penicillin
- Aspirin
- Prednisolone
- Haloperidol/diazepam/lorazepam
- Anti-endocarditis measures

Rheumatic fever usually presents with joint pain but may have an insidious onset especially if carditis is the predominant feature. The diagnosis of rheumatic fever is made on the following criteria and clinical features:

- Evidence of streptococcal infection (usually pharyngitis secondary to group A beta-haemolytic streptococcus) with positive throat swab culture or positive serology for recent streptococcal infection. This is usually accompanied by a prolonged fever and followed by other clinical features after a 2–3 week period. (see Chapter 3.5)
- Arthritis of the large joints. This is a reactive arthritis (rather than a septic arthritis) often affecting many joints and is migratory in nature. It is usually responds dramatically to aspirin (up to 120 mg/kg/day in 4–6 divided doses by mouth after food) or other NSAIDs (see below). The presence of joint pain without swelling may still indicate rheumatic fever in the presence of the other clinical features.
- Skin rash and subcutaneous nodules: erythema marginatum is an uncommon feature. It has a "snake-like" appearance, usually over the trunk, and occurs early in the disease but may persist for months. Subcutaneous nodules are also rare, occurring over bony prominences such as elbows and knees.
- Carditis: this may range from a tachycardia with a prolonged PR interval seen on the ECG through to myocarditis with a systolic apical mitral murmur, pericarditis or cardiac failure.
- Chorea is an involuntary movement disorder, often of the face, tongue and upper limbs. It may appear as dysarthria or clumsiness and is associated with emotional lability.

Treatment

Supportive care with bed rest is needed during the acute attack.

For arthralgia, aspirin as above or other NSAID (for example ibuprofen up to 60 mg/kg per day in 3–4 divided doses after food) is given.

Eradication of streptococcal infection with IM benzylpenicillin (1.2 million U as a single injection) or a ten day course of oral penicillin at high dose (12.5 mg/kg 4 times a day). Penicillin prophylaxis (up to 1 year 62.5 mg, 1–5 years 125 mg, 6–12 years 250 mg, >12 years 500 mg; all twice daily) should be given until the adult years. For patients allergic to penicillin, erythromycin in the same doses can be used.

Endocarditis prophylaxis requires high dose antibiotics just before and after dental and other surgical procedure (see Chapter 3.5).

For acute carditis, prednisolone orally (2 mg/kg/day) for 2–3 weeks or by intravenous infusion (see below) is effective.

Chorea may respond to haloperidol of 12.5 to 25 micrograms/kg twice daily (max 10 mg a day). Extra-pyramidal side effects may occur.

Vasculitis in children

Minimum standards requirements: Vasculitis

- Prednisolone
- Aspirin
- IV gammaglobulin (if available)

Vasculitis in childhood may be primary, including Henoch–Schönlein purpura, Kawasaki disease and the rare vasculitides, or secondary to multisystem connective tissue diseases including juvenile dermatomyositis or systemic lupus erythematosus (SLE). In all of these diseases, skin manifestations are usually prominent but the combination with other clinical features helps to ascertain the diagnosis.

Henoch–Schönlein purpura

- **Purpuric rash**: a palpable purpuric rash is most commonly seen over the buttocks and around the ankles and legs. The purpura occur in crops and may range from small petechiae-like lesions to large, ulcerating ecchymoses. Oedema and urticaria may precede the purpura, particularly at the ankles, scrotum and face.
- **Gastrointestinal pain**: abdominal pain is a prominent feature early in the disease and often accompanied by vomiting. Occasionally, frank gastrointestinal haemorrhage may occur.
- **Arthritis**: typically affects large joints of the lower limb, especially the ankles. Ankle swelling may be difficult to interpret in the presence of tissue oedema. The joint pain is usually transient.
- **Renal disease**: haematuria and proteinuria are common manifestations of the disease but are usually only detected on dip-stick urine analysis. A small proportion of children (1–3%) may develop renal failure secondary to severe glomerulonephritis.

Treatment

Henoch–Schönlein purpura is usually a self-limiting disease, requiring supportive care and symptomatic treatment with simple analgesia only. If the abdominal pain is severe, prednisolone (1–2 mg/kg/day) for a week may be helpful.

Kawasaki disease

Kawasaki disease is characterised by the combination of most of the following features in a young child (<5 years) who is extremely miserable.

- **Fever**: An irregular spiking fever which persists for 1–3 weeks despite antibiotics is characteristic during onset.
- **Skin involvement**: Skin rash is variable and polymorphic, ranging from diffuse erythema of the trunk and face to minimal macular lesions on the limbs. Tissue oedema of the dorsal surfaces of the hands, feet and perineum is characteristic. These changes are followed within days to weeks by desquamation, usually of the finger and toe tips but occasionally more widespread.
- **Mucositis and conjunctivitis**: Inflammation of the mucous membranes of the mouth and eyes results in a characteristic appearance of red eyes and red, swollen, cracked lips.
- **Lymphadenopathy**: Usually affects cervical lymph nodes, often unilaterally.
- **Cardiac disease**: Myocarditis with heart failure or pericarditis are rare but serious complications of Kawasaki disease. Coronary artery aneurysms may be present from early in the disease process. Clinical manifestations are relatively non-specific, ***but the two-dimensional echocardiographic appearances are diagnostic of the condition***.

Treatment

- Hospitalisation and monitoring of cardiac status.
- Aspirin, initially 100 mg/kg/day in 3–4 divided doses after food until the acute inflammatory phase of the disease settles, then 1–10 mg/kg/day (antiplatelet doses).
- ***Intravenous gammaglobulin 2.0 g/kg immediately on diagnosis, if available***. This treatment reduces the likelihood of coronary artery aneurysms if given once during the first 10 days of the illness.
- Corticosteroids (prednisolone 1–2 mg/kg/day) may have a role in controlling the acute inflammation of Kawasaki disease, but are generally not recommended.

Juvenile idiopathic arthritis

Juvenile idiopathic arthritis (JIA) is one of the most common physically disabling chronic diseases of children. The most prominent clinical features include joint swelling, restriction of joint movement, joint pain and tenderness at the joint margins, muscle wasting and any of the non-specific features mentioned below (Table 3.32.1). The most common mistake is to diagnose arthritis in the absence of objective evidence of persistent joint swelling.

Table 3.32.1. Diagnosis of juvenile idiopathic arthritis: all four criteria required

Presence of arthritis, defined by swelling of a peripheral joint. Loss of joint range of movement and pain on movement are sufficient for the definition of arthritis involving the hip or spine (in the absence of other causes for the pain).
Persistence of arthritis for more than 6 weeks
Onset of arthritis before the 16th birthday
Absence of any known cause for the arthritis

Classification and differential Diagnosis (Table 3.32.2)

There are a variety of different forms of JIA that are important in order to advise on the prognosis and most appropriate treatment of the illness.

- Arthritis affecting a few joints only: oligoarthritis carries the best prognosis; 30% may have arthritis affecting their adult years.
- Arthritis affecting many joints: polyarthritis is likely to persist into adult life in 40%.
- Arthritis affecting few or many joints with prominent extra-articular features:
  - Systemic arthritis: with fever, rash, enlargement of liver, spleen, lymph nodes, pericarditis is a life-threatening complication. The persistence of arthritis with this illness carries the worst prognosis: over 50% have arthritis as adults.
  - Psoriatic arthritis: often associated with a psoriatic rash and family history of psoriasis. This has a similar outcome to polyarthritis.
  - Enthesitis-related arthritis: the clinical manifestations of enthesitis include pain, tenderness and occasionally swelling localised to the exact site of tendon insertion. Other features include back pain, red, painful eyes and urethritis. It carries a 60% risk of developing ankylosing spondylitis.

Table 3.32.2. Differential diagnosis of JIA

- Transient arthritis: irritable hip, reactive arthritis
- Septic arthritis* and osteomyelitis, including immunodeficiency
- Acute lymphoblastic leukaemia*, neuroblastoma*, lymphoma*, local neoplasia*
- Bleeding diatheses: haemophilia
- Haemoglobinopathies: thalassaemia, sickle cell crisis
- Epiphyseal disorders: dysplasia, avascular necrosis, osteonecrosis, slipped upper femoral epiphysis
- Metabolic and endocrine disorders
- Traumatic joint disease, including non-accidental injury*
- Hypermobility and inherited connective tissue diseases
- Systemic connective tissue diseases, including systemic lupus erythematosus, dermatomyositis, vasculitis
- Idiopathic musculoskeletal pain syndromes
- Arthritis of inflammatory bowel disease

* Rheumatological emergencies.

Initial minimal set of investigations for differential diagnosis

- Full blood count, including white cell differential and platelet counts
- Plain radiographs of affected joints
- ***Synovial fluid aspiration, microscopy and culture***
- Blood culture

Monitoring for complications and disease progress in JIA

There are several important complications of JIA including joint failure, chronic anterior uveitis and local growth disorders, as well as the general complications of chronic inflammatory disease in children such as anaemia, fatigue, delayed puberty and growth failure. Three of these complications, joint failure, chronic anterior uveitis and growth disorders will be discussed in more detail.

Joint failure (Table 3.32.3)

- Inability to walk without pain and stiffness.
- Inability to write or perform activities of self-care without pain and stiffness.
- The integrity of joint cartilage and bone density is affected from the onset of the disease.
- If the inflammation remains poorly controlled, destruction of cartilage, joint space narrowing and erosion of bone will result in permanent loss of joint function.

Table 3.32.3 Important sites of joint contracture

Joint affected	Type of contracture	Consequence of contracture
Tibio-talar gait	Plantar flexion deformity	Circumduction or high stepping
Knee	Flexion	Quads wasting, limping gait
Hip	Flexion	Limited “swing phase” gait
Wrist	Flexion	Poor writing
Neck	Flexion	Poor neck rotation

Eye disease

- Chronic anterior uveitis is typically insidious and asymptomatic: all children with JIA should have slit lamp eye examination to detect cells in the anterior chamber and protein “flare”. **Delay in the diagnosis can lead to blindness.** ✔
- Inflammation is treated with ocular topical corticosteroids (hydrocortisone 1% eye drops or ointment 0.5%) three times daily and mydriatics (3 minutes after hydrocortisone) (atropine 0.5% eye drops or ointment 1%).
- Severe chronic anterior uveitis may require systemic treatment with corticosteroids or methotrexate.

Growth disorders

- Generalised growth failure may be due to inadequate energy intake (chronic inflammatory disease increases energy demands) or the adverse effects of medication. It is usually treated with dietary energy supplements and ***occasionally recombinant human growth hormone.***
- Local growth disturbance: bony overgrowth of the knee with an increase in leg length, sometimes with a valgus knee deformity. Arthritis of the small joints of

the hands is likely to cause premature fusion of the epiphyses and reduced growth of the affected fingers.

Treatment of JIA

- The first priority is to exclude the differential diagnoses, especially the emergencies of septic arthritis, acute lymphoblastic leukaemia or other malignancies and non-accidental injury. Septic arthritis will require large doses of intravenous antibiotics (See Chapter 3.45).
- The effective treatment of JIA usually requires a team of trained health professionals including therapists and medical staff.
- Education of the patient and family is important, especially concerning the risks and benefits of all treatment and the natural history of the disease.
- Physiotherapy, hydrotherapy and occupational therapy work together to maintain joint function and muscle bulk, correct joint deformities and rehabilitate affected joints.
- Drug treatment should begin as soon as the diagnosis is made with both:
  - **Non-steroidal anti-inflammatory drugs** (NSAIDs): Ibuprofen up to 60 mg/kg/day in three or four divided doses after food. Avoid using more than one NSAID at a time.
  - ***Intra-articular corticosteroids: Strict aseptic conditions, no-touch technique, appropriate sedation, local or general anaesthetic must be given. Triamcinolone hexacetonide is the most effective steroid at a dose of 1 mg/kg/large joint (for example knee, hip or shoulder) or 0.5 mg/kg/small joint (for example ankle, wrist, elbow). This technique requires an experienced operator.***
- For children with polyarthritis or systemic arthritis, in addition to the above, the following should be considered:
  - ***Methotrexate***: Begin with 500 micrograms/kg/week (up to 15 mg/week) starting dose, orally 1 hour before food, and increase by 2.5 mg every month until 1.0 mg/kg/week (maximum 30 mg/week). The drug may be given by subcutaneous injection in severe cases. The patient should be monitored monthly for cytopenia (full blood counts) and liver function abnormalities. Administration is sometimes accompanied by nausea, a side effect that can be improved with folic acid 1 mg once daily (not on the day of methotrexate).
  - **Intravenous methylprednisolone**: May be needed for severe disease flares or complications such as pericarditis. 30 mg/kg/dose (maximum of 1 g) is given once a day for three days by slow intravenous infusion over a 2–3-hour period. Blood pressure monitoring for acute hypertension during the administration of this medication should occur every half-hour.
  - **Sulphasalazine**: Begin with 12.5 mg/kg/day for the first week, increasing by 12.5 mg/kg/day each week until the maximum dose of 50 mg/kg/day in two divided doses is reached, or until adverse drug reactions occur. These may include skin rash, nausea, abdominal pain and pancytopenia. Monitoring with 1–2 monthly full blood counts is a sensible precaution.

Minimum standards requirements

- NSAIDS
- Aspirin
- Prednisolone
- Sulphasalazine
- Ocular steroids and mydriatics
- Intra-articular steroids
- Physiotherapy and family support

3.33 The treatment of children with cancer

Barry Pizer and Tim Eden

Minimum standards requirements

- Regional/national centre chemotherapy and radiotherapy
- Antibiotics
- Blood products
- Palliative care (see Chapter 1.28)
- Indwelling long-term vascular access

Introduction

✔ ● **More than five times as many children present with cancer each year in disadvantaged countries than in rich nations**.

- With an increasing global population, principally in disadvantaged countries, the number of children will continue to increase in terms of absolute numbers and proportionately in such poorer nations.
- As malnutrition and infection decline, particularly in young children, the worldwide contribution to mortality from cancer will increase.

✔ ● **Only 10–20% of all children with cancer in poor countries receive curative therapy, and most do not even receive any form of palliative care**.

Epidemiology

The estimated incidence rate of cancer in children (aged <15 years) is approximately 120 per million per year for both rich and poor countries. Most cancers probably result from the interaction of environmental factors with a genetic predisposition. For example, African Burkitt's lymphoma is related to infection with the Epstein–Barr virus (EBV) very early in life, with persistence of induced genetic rearrangements within B lymphocytes. However, the widespread use of medicinal plants which may increase the likelihood of cell transformation by EBV, chronic malnutrition which induces immunosuppression and frequent infections which cause B-cell proliferation are all probable aetiological factors.

There are clear variations in the incidence of different childhood cancers around the world, for example an excess of retinoblastoma in India, Pakistan and sub-Saharan Africa. In many countries, most noticeably those in sub-Saharan Africa, the AIDS panepidemic has been associated with a significant increase in cancers such as Kaposi's sarcoma and retinoblastoma.

Problems of treating children with cancer in disadvantaged countries

The problems listed below are not exclusive to disadvantaged countries, and not mutually exclusive, i.e. many factors interact, compounding difficulties in treating cancer.

- **Poverty**: national, regional, local and personal
- **High cost of treatment**
  Cost of cytotoxic agents and of diagnostic imaging and pathology
  - Cost of supportive care:
    infection
    blood products
  - Cost of caring for critically ill children:
    high-dependency/intensive care
    postoperative care
- Lack of suitable treatment centres and training programmes
- Poor transportation and communication
  Difficulties and cost of travelling to treatment centres particularly for repeated visits
- Impact on family structure:
  - Loss of parental income
  - Disintegration of family
  - Change in role of the family, especially where both parents need to work to maintain the family income
- High prevalence of infections, anaemia and malnutrition
  - Confounding diagnosis
  - Decreasing tolerance to cytotoxic therapy
- Lack of education and knowledge of uncommon diseases
  - Amongst healthcare workers
  - Amongst families/communities
  - Results in delay in diagnosis, lack of understanding of need for treatment and poor compliance with treatment.
- Reliance on traditional medicines

- Unrealistic preconceptions of cancer
  - Healthcare workers
  - Families/communities

In Swahili – 'The never healing sore', i.e. zero expectation of cure, therefore don't try.

Management of children with cancer in disadvantaged countries

✔ **A decision has to be made, depending on resources, as to whether or not children with cancer are going to receive treatment aimed at curing their disease or be given palliative care to reduce their suffering. BOTH MUST BE SEEN AS ACTIVE FORMS OF THERAPY**. If curative treatments are to be undertaken, then it is our view that it should, whenever possible, be given in a specialist children's cancer centre (see below). We feel there is little benefit and the potential for increasing suffering by 'half treatment' of cancer in children. It has to be done fully and professionally **or** the child treated palliatively (see Chapter 1.28 for additional information on this). However, in countries where there is an improving infrastructure but short of a specialist centre, the following cancers may have a good or reasonable chance of cure:

- **Burkitt's lymphoma**: single agent – cyclophosphamide, cyclophosphamide, vincristine, methotrexate, prednisolone (COMP) chemotherapy
- **Wilms' tumour**
  stage I: surgery plus ten doses of vincristine (at weekly intervals)
  stage II: (possibly stage III) – surgery plus vincristine/actinomycin for six months
- **Hodgkin's disease**: Chlorambucil, vinblastine, procarbazine, prednisolone (ChlVPP) or mustine, vinblastine, procarbazine, prednisolone (MVPP) – six to eight courses
- **Neuroblastoma** (stage I and II) – surgery alone
- **Retinoblastoma** – enucleation (radiotherapy in some cases)
- **Brain tumours**
  Resectable low grade gliomas: surgery alone
  Medulloblastoma and ependymoma (resectable/non-metastatic): surgery followed by ***radiotherapy***
- **Resectable embryonal rhabdomyosarcoma** (certain sites): surgery plus vincristine/actinomycin D – four courses
- **Non-Burkitt's/non-Hodgkin's lymphoma**: early stage – surgery plus COMP or cyclophosphamide, adriamycin, vincristine, prednisolone (CHOP)
- **Standard risk acute lymphoblastic leukaemia** Children aged 2–10 years, with white cell count $<50 \times 10^9$/litre may have a reasonable chance of cure with induction chemotherapy (vincristine, prednisolone, asparaginase) followed by maintenance chemotherapy as described below without the use of intensification modules. ***CNS-directed therapy with cranial radiotherapy plus limited intrathecal methotrexate or intrathecal methotrexate throughout therapy is, however, required in all cases. Asparaginase is expensive.***

Specialist cancer centres

Establishment

- Specialist centres, and the use of standard treatment protocols (discussed below), have been fundamental to the ever-improving survival of children with cancer.
- Cancer is a relatively rare disease and the treatment is usually complex.
- Management requires a dedicated and experienced multidisciplinary team.
- **Every country should have at least one adequately equipped and funded centre**. ✔

Advantages of a specialist children's cancer centre

- Development of medical, nursing and paramedical expertise.
- Improved supportive care, including pain relief for children.
- Facilities to protect cancer patients from other children suffering from contagious diseases.
- Opportunities for training and accreditation in paediatric oncology.
- Stimulus for the development of similar units in the same part of the world.
- Improved support, education and counselling of affected children and their families.
- Improved opportunities for research, including the development of treatment protocols relevant to the particular region or country.
- Development of links with national and international oncology units and organisations.

Requirements of a specialist children's cancer centre

- Dedicated paediatric oncologist(s) and nursing staff.
- General surgeon and neurosurgeon trained in paediatric surgery.
- Blood and platelet banking facilities.
- Pathologist with experience of paediatric tumours with adequate histology and cytology facilities (immunohistochemistry desirable – can be in a centralised laboratory if more than one centre/country).
- Haematology, biochemistry and microbiology laboratories with good quality control.
- Diagnostic imaging: *X* ray, ultrasound; computed tomography imaging desirable, especially for brain tumours.
- Computer facilities with Internet facility (for e-mail to link centre, Medline searches, patient database).

Above all, a keenness of all staff to work together to learn and make the unit successful.

Unit database

All units should keep a record of treatment, including details of patient demographics, diagnosis, treatment, side effects and survival. This will help the identification of specific problems and the development of more effective treatment protocols for treatment and supportive care. The aim should be to try and create a truly population-based database.

Links with other units and organisations

Provision should be made for communication and transportation for patients from remote areas. Links with healthcare facilities in rural areas should be established to continue care where appropriate (for example district hospitals, community health care workers).

Links with units in well-resourced nations

Links with an established unit in a richer nation, can have several advantages as follows:

- Development of standardised treatment protocols including those for supportive care
- Provision of help for difficult diagnoses or therapeutic dilemmas
- Exchange educational visits
- Increased profile of the specialist unit
- Improve support from governmental and non-governmental organisations
- Research.

In addition, links with international organisations is to be encouraged, for example with the International Society of Paediatric Oncology (SIOP).

Principles of the curative treatment of children with cancer as undertaken in a specialised unit

Diagnosis

- A complete history and examination.
- Investigations to confirm histology, determine the extent of the tumour (staging) and identify any tumour-related toxicity (for example disturbance of renal/liver/bone-marrow function).

Pathology

- Good histopathology is essential for the individual and is the only way to compile true incidence figures, survival data and to identify favourable histological subgroups.
- Close involvement of the pathologist before biopsy or surgery so that the surgeon can obtain an optimal specimen in the right fixative.
- The International Society of Paediatric Oncology are proposing to provide a centralised review of diagnoses in problem cases.

Imaging

- To define the dimensions of the primary tumour and to determine the degree of tumour spread (staging).
- Good plain posterior-anterior and lateral chest *X* rays are quite adequate for chest imaging.
- Ultrasonography affords good visualisation of the abdomen and pelvis, although computed tomography (CT) may have advantages in some patients.
- Intravenous urography and cavagrams may also be useful in patients with abdominal tumours.
- For the brain, CT scanning is a necessary part of investigation and management. MRI imaging has advantages over CT, but availability of this technique is very limited.

Biochemical markers

Useful in the diagnosis of a limited number of tumours, for example urinary catecholamines in neuroblastoma and serum alpha-fetoprotein in hepatoblastoma and germ cell tumours.

Treatment protocols

- Each unit should use a standard protocol for each tumour type with necessary variations for tumour stage.
- Protocols should be based on established and effective protocols used by national and international groups.
- Protocols may require modification based on the resources, drug availability, cost and the level of supportive care that can be provided by the unit.

Chemotherapy

- Childhood malignant tumours have almost always disseminated, requiring treatment with systemic chemotherapy.
- Cytotoxic drugs prevent cell division by a variety of mechanisms.
- Although occasionally single agent therapy is given (for example for stage I Wilms' tumour), the great majority of treatment protocols employ a combination of drugs used synergistically to produce maximal cell kill with acceptable toxicity and to prevent tumour cell resistance.

Surgery

- Important in obtaining diagnostic material, and as local therapy to reduce tumour bulk. Surgeons should be specially trained and have experience in oncology.
- Operating facilities must be of high quality to reduce the risk of infection.
- There must be adequate support from blood transfusion services.

- Several treatment protocols use preoperative chemotherapy, which may reduce tumour size, and thus reduce perioperative risks.

Radiotherapy

- Radiotherapy is used to treat regional tumour extension including nodal disease and as part of local tumour control to eradicate local residual microscopic (or sometimes macroscopic) disease following surgery.
- It has a particular role to play in certain brain tumours and may also be used as curative therapy in early-stage Hodgkin's disease.
- It is also frequently used in the management of bone and soft tissue sarcomas and in the prevention of overt central nervous system disease in acute lymphoblastic leukaemia.
- Megavoltage machines have advantages over older orthovoltage therapy in giving a more controllable beam and to avoid damage to skin and overlying tissues when administered to deep tissues.
- The whole of the original tumour volume is generally irradiated plus a safety margin (usually 1–2 cm) of surrounding normal tissue.
- The combination of chemotherapy and radiotherapy can increase late local effects and should be minimised whenever possible.

Procedures

Bone-marrow aspiration

- Needed in the diagnosis of leukaemia and lymphoma but also to identify any bone-marrow infiltration with solid tumours such as neuroblastoma.
- ✔ **Is a painful procedure and must be done either under a general anaesthetic or under sedation along with infiltration with local anaesthetic of the skin and subcutaneous tissues down to periosteal level**.
- Aspiration is preferably performed from the posterior iliac crest but can also be taken from the anterior crest.

Lumbar puncture

- In the diagnosis of malignant meningitis, especially with leukaemia and lymphoma, but also in certain brain tumours, for example medulloblastoma and other solid tumours, particularly those affecting the head and neck.
- Lumbar puncture is a painful procedure and in a child should usually be performed under general anaesthetic or sedation with local anaesthetic wherever possible (especially if multiple lumbar punctures are needed).

Side effects of the disease and/or its treatment

Chemotherapy

Bone-marrow suppression is the most important and life-threatening side-effect.

Specific side effects of chemotherapy

Neurotoxicity	Vincristine (muscle weakness due to peripheral neuropathy, constipation, rarely encephalopathy)
Cardiomyopathy	Doxorubicin/daunorubicin
Respiratory	Bleomycin
Urinary Tract	Cisplatin (renal), ifosfamide (renal and bladder), cyclophosphamide (bladder)
Liver	Thioguanine, actinomycin D
Hearing	Cisplatin

Infection

- Neutropenia, at diagnosis in leukaemia and following most chemotherapy, produces a risk of significant bacterial and fungal sepsis when the neutrophil count is less than 1.0×10^9/litre and particularly when less than 0.2×10^9/litre.
- The greatest risk is from the absorption of Gram-negative bowel organisms such as *E. coli*, *Proteus*, *Klebsiella* and *Pseudomonas*.
- Gram-positive organisms from the skin and mucosal surfaces, especially staphylococci, may also cause significant morbidity.
- Life-saving measures include identification of those at risk, close observation and the empirical administration of intravenous antibiotics to patients with a neutrophil count of less than 1.0×10^9/litre who develop fever (for example more than 38°C for two hours or 38.5°C on one occasion).
- The antibiotic regimen should be determined by each centre depending on the prevailing flora and the cost and availability of antibiotics.
- First-line therapy for febrile neutropenia should generally be with a combination of a broad spectrum beta-lactam antibiotic and an aminoglycoside.
- If the temperature fails to remit, or if Gram-positive organisms are isolated, then therapy with vancomycin is recommended.
- For microbiologically proven septicaemia, 5–7 days antibiotics should be given, the choice of which depends on the antibiotic sensitivity of the isolated organism.
- Newer, very broad spectrum antibiotics, such as the carbapenems and quinolones, are best avoided as they are expensive and may promote fungal colonisation and bacterial resistance.
- Most patients should receive oral nystatin (100 000 units in 1 ml) (2 ml four times a day) to prevent fungal gut colonisation. Patients should be encouraged to retain the nystatin in the mouth for as long as possible.
- If systemic fungal infection is proven or suspected (for example if fever fails to remit after four to five days of antibiotics), then intravenous amphotericin, despite its renal toxicity, is still the drug of choice and is widely available.

- *Pneumocystis carinii* pneumonia especially in patients with leukaemia, requires prophylaxis with co-trimoxazole (calculated on a dose of 150 mg/m^2/day of trimethoprim given twice a week).
- Viral infections are generally tolerated, but chickenpox and measles cause life-threatening infections in immunosuppressed patients. Whenever possible, children must be isolated from direct contact with these infections. Immunoglobulin therapy, including zoster immune globulin, may be life-saving but is rarely available.
- High-dose aciclovir is the treatment of choice for zoster infections, but is expensive and not yet widely available throughout the world.

Bleeding/anaemia

- Adequate blood banking facilities (see Chapter 1.14) are a fundamental part of therapy. Red cell transfusion should be reserved for symptomatic anaemia, or when the haemoglobin falls to a very low level, for example less than 6 g/dl.
- Platelets should be reserved for patients with florid petechiae or overt bleeding or to cover procedures such as lumbar puncture, when a platelet count above 40×10^9/litre is essential.
- Prophylactic platelet transfusions in response to specific platelet counts are not recommended.
- In the presence of fever, bleeding may occur at higher platelet counts than would normally be expected.

Nutrition

- Maintenance of adequate nutrition is essential. "Cancer wasting" or cachexia is a well-recognised complication of paediatric tumours and is associated with a decreased tolerance to chemotherapy and its side effects and possibly an increase in cancer mortality.
- Poor nutritional status may result from:
  - Stress
  - Pain
  - Increased metabolism (tumour/infection)
  - Anorexia
  - Alterations in taste and smell
  - Chemotherapy induced nausea and mucositis (for example stomatitis/oesophagitis)
  - Radiotherapy induced mucositis and dry mouth (xerostomia)
  - Surgery-induced pain, bowel obstruction and appetite suppression
- Each child should have a nutritional assessment including measurement of height or length, weight and mid upper-arm circumference. These should be plotted on a standard percentile chart (see Chapter 6.1).
- Nutritional support should be given to children who consistently show a decrease across percentile lines.
- A high-calorie diet with adequate protein should be given to all children with cancer, supplemented when necessary with specific additives providing additional calories and protein.
- If sufficient food is unable to be taken orally, then enteral feeding via a nasogastric tube (particularly overnight) should be given.
- Total parenteral nutrition should be avoided, as it is expensive and associated with a high risk of complications including infection and metabolic disturbance.

Nausea and vomiting

- Nausea and vomiting is a very unpleasant side-effect of chemotherapy and can lead to poor compliance with therapy and additional complications such as metabolic disturbance, dehydration and oesophageal tears.
- Chemotherapeutic agents vary in their emetogenic potential from very low (for example vincristine and etoposide) to very high (for example cisplatin and actinomycin D). Antiemetic therapy should be given wherever possible, preferably prophylactically, but certainly to patients with established retching and vomiting.

Antiemetic agents

- **Metoclopramide**: effective in high dose but a greater risk of extrapyramidal side effects exists in children. Up to 150 micrograms/kg/two to three times daily orally or slowly (over 2 minutes) IV injection. **AVOID IM route**.
- **Chlorpromazine**: orally or IV (IV can cause severe hypotension) 500 micrograms/kg every 4–6 hours.
- **Prochlorperazine**: orally or IV slowly over 10 minutes 100–250 micrograms/kg every 8–12 hours (only if >10 kg or >1 year of age).

These drugs are generally available but have a high incidence of side effects including drowsiness. May be more effective when combined with steroids.

- **Benzodiazepines**: main effect is sedation and amnesia. Useful for anticipatory nausea.
- **Steroids**: main effect is in combination with other agents (prednisolone 0.5 mg/kg every 12 hours.
- ***5HT$_3$ antagonists: the most effective antiemetics especially when combined with steroids. Expensive***.

Alopecia

Inevitable with most chemotherapy but reversible.

Oral mucositis

- A common side-effect of many cytotoxic agents and also radiotherapy.
- Good, simple oral hygiene should be maintained and prophylactic nystatin (2 ml four times daily) given.

Infertility

- This mainly occurs in males and is a consequence of several cytotoxic agents, especially the alkylating agents such as cyclophosphamide or radiation to the gonads. Girls may suffer from ovarian failure causing a premature menopause after certain therapies.
- Families should receive counselling in this respect.

Second tumours

- Chemotherapy results in a small but important risk of second tumours, especially acute myeloid leukaemia.
- It is especially associated with alkylating agents such as cyclophosphamide (especially if used with radiotherapy) and topoisomerase-2 inhibitors such as etoposide.

Venous access

- Venepuncture for administration of chemotherapy and blood sampling is painful and especially difficult in the young child.
- Repeated venepuncture results in loss of venous access due to venous thrombosis and may significantly compromise therapy.
- Several agents, especially vinca alkaloids are extremely vesicant when extravasated.
- ***The placement of a long-term central venous catheter, for example Broviac or Hickman lines, should be considered in children receiving intravenous chemotherapy. Such lines are relatively cheap, but should be placed by an experienced surgeon, and are associated with an increased risk of infection, particularly from skin organisms such as staphylococci.***
- Short-term percutaneous placement of medium length or long lines under local anaesthetic may provide an alternative means of venous access.

Psychological support

Cancer and its treatment is a frightening experience for many patients and every attempt should be made to reduce the child's fears. An explanation of the diagnosis and treatment should be given in clear, understandable terms to the child's family and also to the affected child or young adult wherever appropriate. All aspects of treatment should be clearly explained, including details of supportive care, such as infection control, mouth care and care of lines and procedures, such as surgery, bone-marrow aspirate and lumbar puncture. The family must always be fully involved in the patient care.

Treatment of individual tumour types

A detailed discussion of the presentation and management of every type of tumour is outside the scope of this book.

Acute lymphoblastic leukaemia (ALL)

Presentation

- Myelosuppression:
  - anaemia, infection (can be life-threatening)
  - thrombocytopenia (bruising/bleeding, petechiae)
- Lymphadenopathy, hepatosplenomegaly
- Organ infiltration – abdominal pain (diffuse, non-specific)
- Bone pain/limp

Diagnosis

- Blood film can be diagnostic for patients with very high white cell counts
- Bone-marrow aspirates (always required):
  - morphology (for example FAB system)
  - cytochemistry and immunocytochemistry (if available)
- Lumbar puncture: CSF cell count and cytospin for lymphoblasts
- Chest *X* ray (T-cell leukaemia)

Treatment

Particularly for patients with high white cell counts, biochemical disturbance as a result of rapid, sometimes spontaneous tumour lysis is a major risk, and intravenous fluids, allopurinol and close monitoring of renal function is required at the start of treatment. The treatment is divided into a number of phases.

Induction

Four weeks of oral prednisolone or dexamethasone with weekly vincristine injections will result in a 90% remission rate, ***although the addition of a third drug, asparaginase (9–12 doses every 48 hours) is associated with improved long-term survival. If asparaginase is not available or too expensive, then anthracyclines (for example doxorubicin) can be substituted.***

CNS directed therapy

This is needed in **all** ***patients to prevent CNS relapse. Standard therapy is to give 5–6 doses of intrathecal methotrexate together with cranial irradiation (24 Gy). For standard risk (not high risk) patients, irradiation can be replaced with intrathecal methotrexate at regular intervals throughout the treatment period, although some units may find radiotherapy easier to administer.***

Intensification therapy

The administration of periods of more intensive therapy, for example with drugs such as cyclophosphamide, daunorubicin and cytosine, has been associated with increased survival, although this treatment carries the risk of severe myelosuppression.

Continuation therapy

This essential part of treatment generally lasts for two to three years. Most regimens employ daily oral mercaptopurine and weekly oral methotrexate with vincristine and a short course of steroid given every month.

Prognosis

With current therapy in specialised centres one can expect at least 50% of standard risk patients (i.e. white cell count at diagnosis $<50 \times 10^9$/litre and aged 2–10 years) to survive.

Acute myeloid leukaemia (AML)

Presentation
As with ALL with more likelihood of tissue infiltration
- Gum hypertrophy: monocytic leukaemia
- Skin involvement: myleoblastic leukaemia
- Disseminated intravascular coagulation: promyelocytic leukaemia

Diagnosis
See ALL.

Treatment
This is less successful than for ALL. Induction therapy is based on 8–10 days of intensive chemotherapy with drugs such as daunorubicin, thioguanine and cytosine. Remission rates of over 80% can be achieved, but these regimens are associated with severe and prolonged myelosuppression with a significant risk of toxic death. This risk should be carefully considered before curative therapy is attempted. Consolidation therapy is again based on intensive and life-threatening chemotherapy. The risk of CNS relapse is less than with ALL. Lumbar puncture with triple intrathecal chemotherapy (methotrexate, hydrocortisone, cytosine) should be given with each course of chemotherapy.

Prognosis
Less than 50% of children will be expected to survive long term, with a high risk of toxic death rate following intensive chemotherapy.

Non-Hodgkin's lymphoma (NHL)

Childhood NHLs are a heterogeneous group of usually diffuse lymphocytic or lymphoblastic neoplasms arising from both B and T cells. Burkitt's lymphoma, a B-lineage NHL is the most common childhood malignancy reported from tropical Africa and is also prevalent in South America and in parts of South East Asia.

Presentation
Lymphomas can arise in any area of lymphoid tissue and thus presenting features are protean. The majority of non-Burkitt's B-cell lymphomas are disseminated at diagnosis, often with diffuse abdominal disease.
- T-cell NHL presents with thymic and/or nodal involvement, often with signs of airway or superior vena cava obstruction. Both types frequently have marrow involvement and sometimes CNS disease.
- Burkitt's lymphoma is an aggressive tumour, usually affecting the head and neck, but also arising from several abdominal organs. Head tumours present usually with extensive jaw involvement with swelling of the jaw and tooth loosening, gum expansion, bleeding, ulceration or exophthalmus.

Diagnosis
The diagnosis is frequently suggested on clinical examination (for example classical features of Burkitt's or T-cell lymphoma). Diagnosis is supported by appropriate imaging (*X* ray, ultrasound). Bone-marrow aspirate and lumbar puncture should be performed. Biopsy is necessary when the diagnosis cannot be made on a marrow examination.

Treatment
Burkitt's lymphoma
This is an extremely chemosensitive tumour and a high remission rate can be achieved by a single course of cyclophosphamide. ***Repeated courses of this single agent may be successful in some early stage patients, but the success of therapy is improved, particularly for advanced disease, with the use of multi-agent chemotherapy using combinations such as COMP (cyclophosphamide, vincristine, methotrexate, prednisolone), for example given over a 6-month period.*** As with ALL, biochemical disturbance as a result of rapid tumour lysis is a major risk, and intravenous fluids, allopurinol and close monitoring of renal function is required.

Non-Burkitt's B-cell NHL
Repeated courses of multi-agent chemotherapy with COMP or CHOP (cyclophosphamide, adriamycin, vincristine, prednisolone) is often successful especially for early stage disease. For advanced disease, more intensive regimens such as the French LMB protocols may result in a high success rate, although toxicity of these regimens is potentially high.

T-cell NHL
As opposed to B-cell NHL, therapy for T-cell disease is usually based on leukaemia-type therapy (with intensification modules and continuing chemotherapy). CNS-directed therapy with cranial irradiation should be used.

Prognosis
Burkitt's lymphoma
Prognosis varies according to the stage of disease, although overall at least 50–75% of patients will be cured with modern therapy. CNS disease is, however, associated with a poor outcome.

Non-Burkitt's B-cell NHL
The prognosis is poorer than with Burkitt's and depends on stage and intensity of treatment. In low-stage disease a survival of at least 75% is expected. Prognosis is worse with extensive disease, particularly with bone-marrow or CNS involvement.

T-cell NHL
With modern leukaemia-type therapy, survival is around 50% or greater.

Hodgkin's disease

Unlike NHL, Hodgkin's lymphoma tends to be confined to the lymph nodes or spleen, although spread to other sites such as lungs, liver and bone may occur. Most children

present with a primary painless neck mass, although any nodal group may be involved. Patients are staged according to the Ann Arbor system, which incorporates an A and B designation for the absence or presence respectively of fever, night sweats and weight loss.

Diagnosis
Diagnosis is generally made by lymph node biopsy. Staging investigations include chest *X* ray and abdominal ultrasound. Bone-marrow aspirate and trephine should be performed on patients with evidence of advanced disease.

Treatment
In the past, radiotherapy was widely used, often using extensive radiation fields, for example the "Mantle" or "Inverted Y techniques" to cover all known sites of disease. Radiation is still used in localised disease, but generally chemotherapy is favoured for most patients using regimens such as ChlVPP (chlorambucil, vinblastine, procarbazine, prednisolone) or MVPP (nitrogen mustard replacing chlorambucil.). Six to eight courses given every month are used. Such chemotherapy may be given as an outpatient, and is relatively non-toxic, although the risk of infertility in boys is high.

Prognosis
Hodgkin's generally carries a good prognosis. For patients with stage I and II tumours, over 80% are expected to be cured. Even in advanced disease, over half of patients would be expected to survive.

Brain tumours

Presentation
About 60% of childhood brain tumours arise in the posterior fossa and present usually with signs and symptoms of raised intracranial pressure due to obstruction of CSF pathways. A variety of other presenting features may occur depending on the site and rate of progression of the tumour. These include irritability, behavioural disturbance, cranial nerve palsies, long tract signs particularly truncal ataxia, endocrine abnormalities, visual disturbance and seizures.

Diagnosis
Modern imaging with ***CT scanning (or preferably MRI if available)*** has revolutionised the management of brain tumours and should be performed if CNS tumours are suspected. Some tumours have characteristic appearances on imaging (for example diffuse brainstem glioma and optic nerve glioma) although most tumours require histological confirmation.

Treatment
For most tumours, modern neurosurgery (see Chapter 3.44) is vital to management. Prompt relief of raised intracranial pressure is often required and may be life-saving. This is achieved with dexamethasone, which when used perioperatively has also been shown to significantly reduce operative mortality. Surgery may also be required to relieve hydrocephalus (for example with ventricular peritoneal shunting). The aim of definitive surgery is to provide a histological diagnosis and usually to de-bulk the tumour as completely as possible. Tumour resection is required for most tumours, including all posterior fossa tumours outside the brainstem, tumours of the cerebral hemispheres and craniopharyngiomas. Some tumour types may be cured with surgery alone (for example cerebellar low-grade astrocytoma), although others, for example medulloblastoma, ***require adjuvant radiotherapy. Generally a large dose of radiotherapy is given to the tumour bed with some tumours, for example medulloblastoma, requiring whole CNS radiotherapy due to the high risk of CSF dissemination. To date chemotherapy has had relatively little impact on the treatment of brain tumours.***

The following is a brief guide for the management and prognosis of individual tumour types:

Medulloblastoma
Surgical resection (as complete as possible). ***Postoperative craniospinal radiation therapy (children aged 3 years and over); 55 Gy to the posterior fossa and 36 Gy to the whole CNS.***

Prognosis
Around 60% for children with non-metastatic disease. Children with medulloblastoma aged less than 3 years have a much worse prognosis than older children. Radiotherapy may be curative but most centres do not advocate this, as radiation therapy to the developing brain is associated with a very high incidence of severe handicap. Prolonged chemotherapy can be used to try and delay radiotherapy but even then survival is only around 20%.

Cerebellar low-grade astrocytoma
Surgical resection – postoperative radiotherapy is not required if the resection has been adequate.

Prognosis
At least 80% following total resection.

Supratentorial low-grade astrocytoma
Surgical resection for accessible lesions, although many of these tumours, for example those involving the hypothalamus and optic pathways, are not resectable. ***In these cases, focal radiotherapy should generally be given, particularly in patients with progressive disease.***

Prognosis
Variable, principally depending on the site of the tumour.

High-grade glioma
Surgical resection (as complete as possible) ***and postoperative focal radiotherapy.***

Prognosis
Overall, very poor at around 15%. Survival for patients with complete resection and Grade 3 (anaplastic astrocytoma) tumours have a much better chance than those with subtotal resection and Grade 4 tumors (glioblastoma multiforme).

Ependymoma
Surgical resection (as complete as possible) and ***post-operative focal radiotherapy***.

Prognosis
Around 30–50%; principally depending on the degree of tumour resection.

Brainstem glioma
- Focal exophytic tumours: surgery followed by focal radiotherapy.

Prognosis
Around 30–50%, principally depending on the degree of tumour resection.

Diffuse (malignant) brainstem gliomas
- ***possibly palliative radiotherapy***.

Prognosis
Fatal (<5% survival).

Craniopharyngioma
Surgical resection (high perioperative mortality rate). ***Radiotherapy sometimes used for recurrent tumours***.

Prognosis
Variable. All patients suffer panhypopituitarism requiring hormone-replacement therapy.

Germ cell tumours
Surgery ***followed by craniospinal radiotherapy*** (high risk of CSF dissemination) ***with boost to primary tumour. Chemotherapy is used in some protocols for malignant intracranial teratomas***.

Germinoma

Prognosis
At least 80% with radiotherapy.

Malignant teratoma

Prognosis
Around 70% in non-metastatic cases.

Complications
Brain tumour therapy, particularly radiotherapy to the whole brain and spine has a very high risk of sequelae particularly in young children, including neuropsychological disability, growth failure (growth hormone deficiency and poor spinal growth) and hypothyroidism.

Wilms' tumour (nephroblastoma)

This tumour occurs in nearly all parts of the world and is one of the most curable of all childhood cancers.

Presentation
Most patients present with a large, generally painless, flank mass with or without haematuria and hypertension. The diagnosis may be confused with the abdominal distension associated with malnutrition and with other flank masses, for example neuroblastoma and splenomegaly associated with malaria or haemoglobinopathy.

Diagnosis
The clinical presentation is often suggestive. The presence of a renal tumour can be confirmed by ultrasonography which should also assess the presence of inferior vena cava (IVC) involvement. Alternatively intravenous urogram (IVU) (with injection into the feet to perform a cavagram), can be used as ***may CT scan (with contrast) if available***. A chest *X* ray should look for the evidence of lung metastases.

Treatment
Primary surgery still appears to be the best approach. Adjuvant chemotherapy should be given, and has dramatically increased survival of this disease. The most active agents are vincristine, actinomycin D and adriamycin. For stage I tumours, vincristine given weekly for ten weeks is appropriate. For stage II disease, vincristine and actinomycin should be given for 6 months, a regimen which may also be used for stage III tumours, although the addition of adriamycin appears to improve survival. ***For stage IV tumours and for so-called "unfavourable (anaplastic)" histology groups, all three drugs should be given for 6–12 months. Radiotherapy to the abdomen should only be given if residual bulky disease is present after surgery. Patients with pulmonary metastases at diagnosis should receive lung irradiation (20 Gy)***.

Prognosis
For patients with stage I and II tumours (favourable histology) at least 80% should be cured. Stage III and IV tumours have survival around 60–70 and 50–60% respectively. The prognosis is, however, poor for patients with unfavourable histology.

Neuroblastoma

This biologically unusual tumour can arise from any part of the sympathetic nervous system, although around 60% originate from the adrenal gland. Localised stage I and stage II disease and the unique stage IV S disease of infancy have a good outlook, although for the 80% of patients that present with advanced tumours, the prognosis is very poor.

Presentation
A large proportion of patients present with an abdominal (adrenal) or pelvic mass often extending across the midline. Paraspinal masses extending into the spinal canal causing cord compression and thoracic primaries causing airway obstruction also occur. Most patients (65%) present with metastatic disease which often causes bone pain and limp, marrow infiltration mimicking leukaemia, skin infiltration or orbital masses causing proptosis or periorbital bruising.

Diagnosis
Ultrasound of the abdomen (***CT abdomen if available***), chest *X* ray (***CT chest for thoracic tumours if available***), abdominal *X* ray (calcification is often a feature of primary tumours), ***24-hour urine collection for catecholamine metabolites*** (secreted in 85% of cases), bone-marrow aspirate and trephine (bilateral) are all helpful. Although the diagnosis can be made without tumour biopsy for patients with classic features of stage IV disease,

histological confirmation is required for localised tumours and for advanced disease where the diagnosis is in doubt.

Treatment and prognosis
Patients with stage I and II disease should be treated with surgical excision, which, if complete is associated with an 80% or greater survival. ***For stage III patients, prognosis is around 40% with treatment including multi-agent chemotherapy with drugs such as cyclophosphamide, vincristine, adriamycin and platinum followed by surgical excision of the tumour***. Stage IV disease carries a very poor prognosis.

Soft-tissue sarcomas

These tumours arise from undifferentiated embryonic tissue. The most common of these is rhabdomyosarcoma, a tumour of striated muscle. Rhabdomyosarcomas can arise anywhere where there is such striated muscle or embryonic remnants thereof, but the most common sites include the orbit, head and neck, including the nasopharynx, genitourinary tract in both boys and girls, and the extremities. Two main histological types are recognised: the more common embryonal type and the less common alveolar type which generally carries a much poorer prognosis.

Presentation
Most rhabdomyosarcomas present as diffuse masses, but orbital lesions generally present with proptosis and diplopia and nasopharyngeal lesions often present with nasal obstruction, epistaxis and pain. At least 25% of sarcomas will have metastases at diagnosis, most commonly to the lungs and lymph nodes.

Diagnosis
Histological confirmation is required by biopsy or excision of the primary tumour. Initial radical surgery should not be performed. Primary tumours should be defined by ***CT scan*** if available (particularly important in head and neck and orbital tumours) although other techniques such as tomography and ultrasound may be useful. For head and neck lesions, lumbar puncture with careful CSF examination is required. Parameningeal tumours are those in which CSF involvement is demonstrated or possible due to the proximity of the tumour to the meninges based on ***CT scanning***. Metastatic surveillance includes chest *X* ray, abdominal ultrasound (***or CT if available***), and bilateral bone marrow aspiration.

Treatment
In view of the high rate of local and distal dissemination, chemotherapy is required for all patients. ***The VAC regimen (vincristine, actinomycin D and cyclophosphamide four to nine courses), is most commonly used. In more recently devised regimens, ifosfamide has replaced cyclophosphamide (IVA ifosfamide, actinomycin D, vincristine) although ifosfamide carries a far greater risk of side effects, including haemorrhagic cystitis and nephropathy. Unless the tumour can be completely excised, local therapy should generally be performed after cytoreductive chemotherapy (for example after three to six courses)***. Surgery is the usual local therapy for such sites such as the extremities and genitourinary systems. For head and neck tumours, surgical excision of the primary tumour is usually extremely difficult and ***radiotherapy should be considered***.

Radiotherapy is the treatment of choice following chemotherapy for orbital tumours.

For parameningeal tumours, whole CNS radiotherapy and intrathecal methotrexate is advised.

Prognosis
For completely resected tumours, the prognosis is good, with at least 70% survival. For those with regional disease the prognosis is less good with about 40–50% survival. Survival is particularly poor for patients with metastatic disease (less than 20%) and for parameningeal tumours and thus careful consideration should be given before embarking on a curative treatment for these categories. Alveolar histology confers a significantly worse prognosis for all stages and sites.

Kaposi's sarcoma

This tumour has become a major healthcare problem in areas affected by the AIDS panepidemic. Younger children tend to present with disseminated suppurative lymphadenopathy and conjunctival disease, whereas in older children, skin nodules predominate.

Treatment
Radiotherapy may control locally aggressive tumours. Kaposi's sarcoma may also respond to chemotherapy including agents such as vincristine, actinomycin D and DTIC.

Bone sarcomas

About half of all sarcomas occur in the bone, predominate types being osteosarcoma and Ewing's sarcoma.

Presentation
A bone sarcoma usually presents as a painful mass which may be hot and tender mimicking osteomyelitis. 95% of osteosarcomas arise in long bones and half occur in the upper tibia or lower femur. About half of Ewing's sarcomas occur in long bones, usually in the shaft, with the remainder occurring in the pelvis, shoulder, skull and vertebrae. About 20% of patients with Ewing's sarcoma and 10–20% of those with osteosarcoma have metastatic disease at diagnosis.

Diagnosis
The diagnosis is suggested on plain *X* ray with osteosarcoma showing bony expansion with both osteoblastic and lytic activity. Ewing's sarcoma generally appears as an ill-defined lytic lesion. Diagnosis is confirmed with biopsy, preferably using an open technique under direct vision. Chest *X* ray (or CT of chest if available) is used to detect lung metastases, the lung being the most common metastatic site for both tumours.

Treatment and prognosis

Ewing's sarcoma

Chemotherapy using vincristine, actinomycin D, adriamycin and cyclophosphamide (or ifosfamide) should be given to control both local and metastatic disease. Local therapy with wide surgical excision should be performed. ***If this is not possible, high-dose radiotherapy (for example 45–50 Gy) should be given***, although for long bone sites amputation may be appropriate in some patients.

Overall prognosis is around 40% but depends on the site and adequacy of local tumour control. The prognosis for patients with metastatic disease is very poor.

Osteosarcoma

Amputation of the long bone containing the primary tumour only gives a cure rate of about 20%. ***Chemotherapy either before or after local therapy has increased survival to around 50% for non-metastatic patients. The current European protocol uses six courses of cisplatinum and doxorubicin (three pre- and three postsurgery) which may be feasible in many low-income countries.*** Local control is either with amputation or when available, tumour resection and ***endoprosthetic bone replacement or rotation plasty.***

Retinoblastoma

Although rare in many countries, retinoblastoma is a common paediatric cancer in many areas, including sub-Saharan Africa, Pakistan and India. Two forms are identified: an autosomal dominant heritable form which may affect one or both eyes and a sporadic (non-heritable) form which is always unilateral.

Presentation

Most children present within the first few years of life with a white mass in the pupil or with a squint. In patients with a family history, routine surveillance may detect an early lesion. Delayed presentation may result in a protruding fungating orbital mass.

Treatment and prognosis

Enucleation of the involved eye is the standard therapy and is curative in about 75% of patients with localised disease. ***Very small tumours may also be effectively treated with a cobalt plaque, local irradiation, light coagulation or cryotherapy. External beam radiotherapy may be curative in early cases, but cataract formation usually results.*** Extensive spread outside the orbit is usually fatal. ***Relatively simple chemotherapy, for example with carboplatin, vincristine and etoposide, appears to be effective in reducing large tumours, facilitating preservation of vision and possibly preventing metastatic spread***.

Liver tumours

The two main types are hepatocellular carcinoma (HCC) and hepatoblastoma. Although both are rare in Europe and North America, in several parts of the world, for example East Africa and New Guinea, HCC is a relatively frequent childhood malignancy. In children with HCC, as in adults, there is a clear and possibly causative association with hepatitis B infection both in the presence and absence of coexisting cirrhosis. Hepatoblastoma generally presents in children aged less than 3 years.

Presentation

The presentation in both hepatoblastoma and HCC is similar with most patients presenting with abdominal distention and a right upper quadrant mass. Additional features, particularly for HCC, include abdominal pain, nausea, weight loss, anorexia and jaundice. Features of underlying chronic liver disease may be present with HCC.

Diagnosis

The liver mass may be seen on ultrasound of the abdomen and ***CT if available***. The diagnosis should be confirmed by biopsy. ***Alpha-fetoprotein is elevated*** in nearly all cases of hepatoblastoma and about 65% of cases of HCC. ***In these patients, the alpha-fetoprotein level may be used as a tumour marker to monitor progress***.

Treatment and prognosis

Surgical excision is the definitive treatment for both tumours. ***Hepatoblastoma is a chemosensitive tumour and preoperative chemotherapy significantly improves prognosis, facilitating surgical excision and controlling distant metastases. The most active agents are doxorubicin and cisplatin***.

Prognosis for localised and non-metastatic tumours is around 50%, although the surgery is difficult and carries significant risks.

The overall prognosis for HCC is very poor. This disease is much less responsive to chemotherapy than hepatoblastoma and unfortunately these tumours are often multicentric or extensively invasive making resection possible in less than 30% of patients. Of these cases, only one-third survive long term.

Palliative chemotherapy and radiotherapy

As stated above, if curative treatment is not possible or has failed, then the focus must be on providing palliative care, particularly symptom control including adequate pain relief (see Chapter 1.27). Occasionally palliative chemotherapy may be appropriate for example the use of steroids with or without vincristine in relapse or incurable ALL and lymphomas. Steroids are also used in controlling symptoms such as headache due to certain brain tumours (see Chapter 1.28). ***Palliative radiotherapy may be useful in treating bone pain caused by tumour infiltration, for example in neuroblastoma and bone tumours themselves, and may be helpful in controlling symptoms caused by compression of nerves (including spinal cord) or other vital organs***.

Conclusion

Although in many countries with limited resources, curative treatment of children with cancer may not be at present achievable, children will present with often distressing symptoms, which we must strive to alleviate and palliate. As infections in particular become more controllable in disadvantaged countries, cancer starts to emerge as a major cause of morbidity and mortality. Some allocation of resources becomes inevitable and since paediatric oncology requires a multidisciplinary approach, thinking and acting on the problems faced by children with cancer can lead to improvement of care for all children in hospitals.

The authors will gladly give advice about issues raised in this chapter, for example use of specific treatment protocols, development of links between centres and contact with international organisations such as SIOP.

Further reading

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3.34 Eye disorders

John Sandford-Smith

Minimum standards requirements

- Vitamin A
- Ocular antibiotics
- Fluoroscein
- Ocular steroids
- Aciclovir
- Occlusive pads
- Glasses and other visual aids

Eye diseases are common in children everywhere. Because the eye is an external organ and liable to infection and injury, eye diseases are even more common in children in disadvantaged countries. The two most important children's eye disorders in poor countries are vitamin A deficiency (xerophthalmia) and trachoma. Both these can be prevented by appropriate action in the community, which is both very cheap and very effective for both disorders.

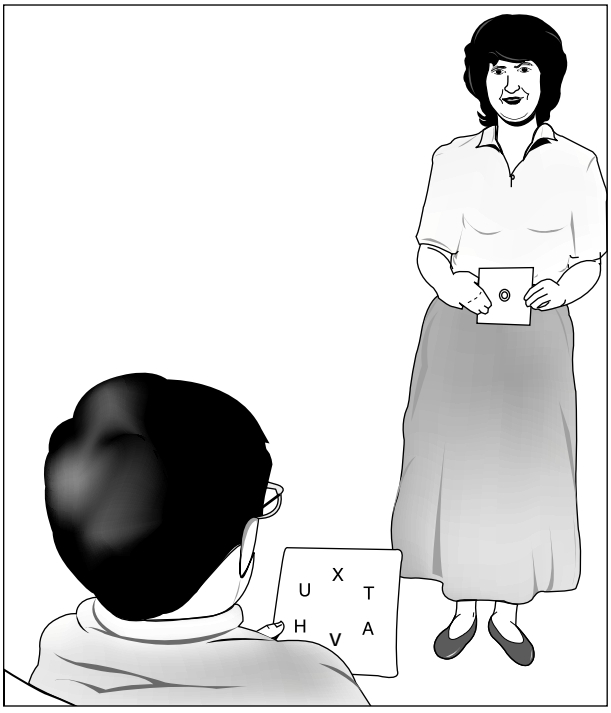


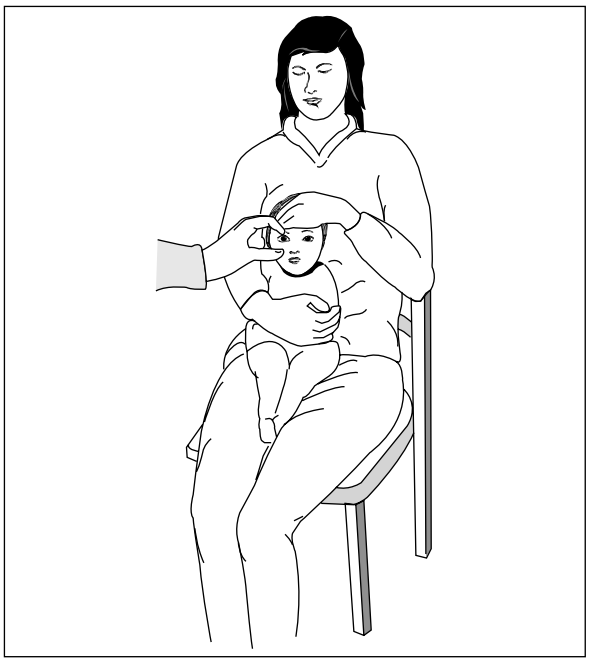
Figure 3.34.1 Vision-testing chart

Eye examination and diagnosis: basic equipment

- Vision-testing chart. Show only one letter at a time and get the child to match the letter on a chart (Figure 3.34.1).
- A bright torch light which can give a focused beam of light.
- The ophthalmoscope.
  - The primary purpose is for examination of the fundus; that is the retina, choroid and optic nerve.
  - The ophthalmoscope can also be used for examination of the ocular media, that is the cornea, lens and vitreous. Dial a small positive lens in the ophthalmoscope, about +2 or +3, and hold it about 20 cm from the patient's eye. In the healthy eye with a dilated pupil there will be a clear red glow of light reflected from the retina called the red reflex, and any opacity in the cornea, lens or vitreous will appear as a black shadow against this red reflex.
  - The ophthalmoscope can also be used to act like a magnifying lens to examine in detail the conjunctiva, the sclera, the iris, etc. To do this a very strong positive lens is dialled in the ophthalmoscope (about +20) and it is held very close to the patient's eye.
- Mydriatic drops (**cyclopentolate.HCl** 0.5% or **atropine** ointment in <3 months of age) local anaesthetic drops (**tetracaine.HCl** 0.5 and 1% or **proxymetocaine HCl**) and sterile **fluorescein** paper strips.
- ***Binocular telescopic magnifying glasses are very useful but not essential.***
- ***More sophisticated equipment like a tonometer for measuring intraocular pressure, a slit lamp and a binocular indirect ophthalmoscope are luxuries which usually are only available in a specialist clinic.***

Gaining the confidence and trust of a child is the most important step in a successful eye examination which should not be painful or unpleasant, other than possibly putting drops in the child's eye. If child is finding it hard to cooperate, examine the parents' or older siblings' eyes first to gain child's confidence. Sometimes with very small children with a serious eye problem an anaesthetic may be required.

Two ways of examining the eyes of young children. The same techniques can be used to apply drops or ointment



Examining the eyes of babies and young children is often very difficult. It requires patience and encouragement to gain the confidence of the child. If it is still difficult to get a good view, the following techniques may be helpful:

1. Seat the baby on his mother's lap, so that her hands restrain his/her arms, and steady his/her head.
2. Wrap the baby in a sheet or blanket, with his/her head on the examiner's lap, and his/her body on his mother's lap. Gently hold open his/her lids with the fingers and thumb of one hand. The other hand is then free to instil any eye drops, or hold a torch or condensing lens. This is probably the best way to get a satisfactory view of the eye, but it also provokes the greatest resentment from the baby.
3. In very difficult cases, it may be necessary to instil a drop of local anaesthetic, and use a speculum to hold open the eyelids. However, any type of speculum can seriously damage the cornea. Only an experienced person must use it and with the greatest of care.

Presenting symptoms of eye disease include:

- Red, sore, irritable or discharging eyes
- Impairment or loss of vision
- Squint.

Red, sore, irritable or discharging eyes

- If **bilateral** it is nearly always caused by conjunctivitis, and if **unilateral** the usual cause is an ulcer or injury to cornea or iritis (occasionally dacryocystitis).
- Evert the upper eyelid to inspect the upper tarsal conjunctiva.
- Apply fluorescein stain to cornea to diagnose an ulcer or identify a foreign body. The green fluorescein dye will stain the ulcer or the foreign body.

Conjunctivitis

- *Acute* **bacterial conjunctivitis** causes a mucopurulent discharge from the conjunctiva and is usually self-limiting, resolving after a few days. Give topical antibiotics as drops or ointment to speed recovery.
  - Acute bacterial conjunctivitis is **dangerous in neonates** when caused by sexually transmitted disease. The cornea in a neonate is at much greater risk and neonates produce less tears to wash away bacteria. **Treatment is urgent**. ✔
  - WHO recommended treatment for severe neonatal conjunctivitis is a single intramuscular injection of either ceftriaxone 50 mg/kg (max. 125 mg) or **kanamycin** 25 mg/kg (max. 75 mg) and hourly **tetracycline ointment** or **chloramphenicol drops or ointment**.
- **Acute viral conjunctivitis** is also a self-limiting disease and usually lasts for a week or so. Tear secretions are usually watery rather than mucopurulent. There is no specific treatment but it is customary to give antibiotic drops.
- **Allergic conjunctivitis** is common and causes recurrent severe itching of eyes. In severe cases there will be pigmentation and proliferation of conjunctiva of limbus and tarsal conjunctiva proliferates to form papillae. Non-steroidal anti-inflammatory drops relieve symptoms but in severe cases use topical steroids

(hydrocortisone 1% eye drops or betamethasone 0.1% eye drops or ointment). If prolonged treatment is necessary, patients must be checked to make sure they are not developing steroid-induced glaucoma.

Trachoma

This is described in Chapter 3.35.

Corneal ulcers

Corneal ulcers are usually **unilateral**. There is usually pain and photophobia. Dilated blood vessels are only around margin of cornea. Staining the eye with fluorescein will show the outline of the ulcer.

- **Herpes simplex** ulcers are typically branched and irregular.
  Treat by applying aciclovir ointment 3% two hourly until the epithelium has healed. If unavailable apply local anaesthetic drops (see above) to the eye and use a sterile cottonwool bud to wipe away all diseased corneal epithelial cells. Pad the eye with antibiotic ointment and mydriatics.
- **Bacterial corneal ulcers** are more serious and can rapidly progress to destroy the cornea and the eye. They must be treated as an emergency.
  - If possible first perform a Gram stain and microscopy of tissue scraped with great care from the edge of the ulcer with a scalpel **blade**. This will often give helpful information about the cause of the ulcer and so make the treatment more specific.
  - Antibiotic drops should be given hourly or two hourly for 48 hours and then four times a day.
  - The choice of antibiotic depends on the availability and also the results of the Gram stain.
  - Ofloxacin (0.3%) or ciprofloxacin (0.3%). Both have a good spectrum of activity against Gram-positive and Gram-negative bacteria. In most circumstances these would be the first choice.
  - Concentrated locally made antibiotic drops are very helpful if ofloxacin or ciprofloxacin are not available. The antibiotic drops can be made up by diluting antibiotic powder for injection in 5 ml of sterile water or 0.9% saline. They should only be used for 48 hours and then discarded. The following are the recommended strengths. Gentamicin 15 mg/ml or Amikacin 50 mg/ml should be used for Gram negative organisms, a cephalosporin either cefuroxime, ceftazidine or cephazolin should be used for Gram positive organisms, the strength being 50 mg/ml. If a Gram stain is not possible, two types of drops can be given alternately every hour.
  - Chloramphenicol (0.5% drops and 1% ointment) is a cheap and readily available alternative if none of the above is available.
- **Fungal corneal ulcers** are very common in hot, humid climates. The branching filaments of the fungus can be identified on a Gram stain. The treatment is unfortunately very difficult because topical antifungal drugs are very hard to obtain and the response to treatment is slow. Natamycin is sometimes available as an eye ointment. Econazole, clotrimazole and ketoconazole are all available as skin creams and it may be necessary to use these or use systemic antifungal agents in desperate cases.

Iritis

Iritis is a less common cause of an acute red eye. The pupil is constricted and irregular and there are often deposits known as keratic precipitates on the posterior surface of the cornea. Give intensive topical steroids hourly (prednisolone, betamethasone or dexamethasone drops) and keep the pupil dilated with mydriatics (atropine 0.5%–1% twice daily).

Vitamin A deficiency (xerophthalmia)

Xerophthalmia usually affects only malnourished children (see vitamin A deficiency Chapters 3.15 and 3.16).

- In the early stages the conjunctiva appears dry and wrinkled, but this is not easy to detect.
- As the disease progresses, the cornea also appears dry and then shows signs of corneal ulceration. Ulcers may progress very rapidly to destroy the entire cornea. Eventually the whole eye shrinks or the child may be left with a dense corneal scar.
- In communities where vitamin A deficiency is common, older children are frequently found with corneal scars dating from early childhood. In most cases malnutrition is a chronic problem, and the disease is precipitated by an acute infective illness that is nearly always measles. Xerophthalmia and measles are particularly important because these ulcers are very frequently bilateral, whereas most other causes of corneal ulceration and scarring usually only affect one eye. There are three other factors which may precipitate corneal destruction in xerophthalmia:
  - **Herpes simplex**. Severe and often bilateral herpes simplex ulcers may develop
  - **Traditional eye medicines**. Application of toxic substances may cause damage and chemical burns to the conjunctiva and cornea
  - **Exposure**. Sick and malnourished children may lie with their eyes open and exposed so the cornea is not protected by the eyelid.

Management

- Apply topical antibiotics and ensure adequate closure of eyelids. Give **local aciclovir** if *herpes simplex* is suspected. Give **topical steroids** (hydrocortisone 1% or betamethasone 0.1% eye drops or ointment) if a clear history of toxic traditional eye medication is obtained.
- Vitamin A capsules (200 000 IU >1 year or 100 000 IU <1 year per day, for 2 days). Systemic antibiotics and rehydration may also be indicated.

The child who cannot see or cannot see well

If only one eye is affected, the child and family may not be aware of the problem. However, a child with poor vision

in one eye only will often develop a squint in that eye (see below).

Cornea

Bilateral corneal scarring bad enough to cause serious visual impairment is most usually a consequence of xerophthalmia and measles (both preventable, vitamin A and immunisation). Careful refraction may improve the sight. ***An optical iridectomy or a corneal graft may also help***.

Cataract

Cataract is the most common congenital ocular abnormality. It may be present at birth, or may develop in early childhood (juvenile cataract). It may be complete, presenting as a dense white opaque mass in the pupil or be incomplete and less obvious. There will be a normal pupil light reaction so that the pupil constricts on shining a light into the eye. Other causes of a white mass inside pupil, in particular retinoblastoma, will **not** have any pupil light reaction.

Congenital cataracts require **early expert surgical treatment** or the child will develop a nystagmus which will prevent the development of good vision.

Congenital Glaucoma

Congenital glaucoma usually presents with photophobia, a hazy cornea and often enlargement of the eye called buphthalmos.

Urgent specialist surgery is required to control intraocular pressure and save what sight is available, **otherwise the child will become irreversibly blind**.

Retinal Diseases

- **Retinopathy of prematurity** is the commonest cause of acquired retinal disease. It is associated with excessive oxygen given to premature babies. It is now particularly common in middle income countries, for example Latin America, Eastern Europe, Middle East and Asia. In countries with highly developed intensive neonatal care it is uncommon and in poor countries most very premature babies do not survive.
- **Retinitis pigmentosa** is the most common congenital disorder of the retina. It affects the peripheral retina and causes night blindness.
- **Vitamin A deficiency** also causes night blindness by affecting rod photoreceptors in the peripheral retina.
- **Retinoblastoma** is important because it **is one of the few eye diseases which can kill a child if not properly treated**. The tumour can present in one eye or in both eyes as a white mass in the pupil, a squint, a painful inflamed eye or a mass in the orbit. If the eye is removed before the tumour has spread the child's life may be saved.

Optic nerve

Optic nerve hypoplasia or optic atrophy may be congenital. It may also be acquired following meningitis, or rarely an infection such as typhoid or measles.

Cortical blindness

Cortical blindness occurs following severe brain insults such as meningitis or cerebral malaria. The pupil light reflex is normal but the child cannot see. In some cases the vision gradually improves with time.

Management of blindness

- In the majority of cases treatment is rehabilitation and education rather than medical.
- Cataracts and glaucoma, in particular, must be recognised and diagnosed early to preserve and save as much sight as possible.
- Most blind children have some sight and should have an opportunity to use low-cost visual aids. Simple aids, manufactured locally, may enable children to read and so transform their whole chance for education. These may be a strongly positive lens worn as spectacles or used as a stand magnifier.

Squint

Squint is a common but complex disorder and there are many different causes and types. Basic questions include:

- **Does the child really have a squint?** To perform a corneal light reflex, ask the child to look at a light and observe the reflections from the cornea. If reflections in the two eyes are symmetrical and central there is no squint, but if one is asymmetrical then that eye is squinting.
- **Does the squint alternate?** Cover the non-squinting eye. If the squinting eye moves to look at the light or object being held and if child can use either eye to fixate then the squint alternates. This means that the vision is fairly good in each eye and the treatment of the squint is purely cosmetic.
- **If the squint does not alternate, is there any disease in the squinting eye?** Test the pupil light reflex and then dilate the pupils with mydriatics. Look for diseases such as cataract, retinal scar and in particular retinoblastoma. Refer for treatment. **Treatment for retinoblastoma is urgent enucleation**.
- **Is there a refractive error, for example hypermetropia (long sight) or myopia (short sight)?** This requires refraction tests.
- **Is the squinting eye amblyopic (that is it has poor vision)?** At first, squints cause double vision (diplopia) which the child finds confusing. As time passes the visual acuity in the squinting eye becomes permanently suppressed. The treatment for amblyopia is to force the child to use the squinting eye by wearing an occlusive patch over the healthy eye for about one hour a day for a few weeks.

✔ **Amblyopia only develops in young children and it can only be treated in young children (under 5 years)**.

Surgery may be required but should not be considered until eye disease and refractive errors have been excluded and amblyopia treated.

3.35 Trachoma

Anthony Solomon and David Mabey

Minimum standards requirements

- Ocular tetracycline
- Doxycycline/tetracycline >12 years
- Azithromycin
- Surgery

Trachoma is the most common cause of preventable blindness worldwide. It is caused by *Chlamydia trachomatis*, certain serotypes of which preferentially infect the corneal and conjunctival epithelium. The organism is transmitted from person to person by direct contact, fomites, and possibly by flies. Disease clusters in families – the greatest risk factor for infection is sharing a bedroom with an active case. Repeated episodes of active infection over many years causes an accumulation of scar tissue in the tarsal plate and tarsal conjunctiva of the upper lids. Contraction of the scar may produce trichiasis and/or entropion, and the resulting corneal abrasion by inturned lashes leads to corneal scarring. This eventually causes blindness. In paediatric practice in endemic areas, active infection is seen frequently. Blinding complications may start to appear in the late second and third decades of life.

Clinical features

These are best presented using the simplified clinical grading system as a framework. Examination for trachoma involves inspection of the lashes and cornea, followed by eversion of the upper eyelid to examine the tarsal conjunctiva. A 2.5 × magnifying loupe and torch (or daylight) should be used. These tools are sufficient to determine the presence or absence of signs that are considered in this grading scheme. Each eye is graded separately.

- Trachomatous inflammation
  - Trachomatous inflammation – follicular (TF): the presence of five or more follicles at least 0.5 mm in diameter deep to the part of the conjunctiva that overlies the upper tarsal plate. Follicles appear as yellow-grey semitransparent patches or swellings beneath the conjunctiva. Fewer than five follicles, or follicles at the nasal or temporal margin may be normal.
  - Trachomatous inflammation – intense (TI): pronounced inflammatory thickening of the tarsal conjunctiva that obscures more than half of the normal deep tarsal blood vessels.

  TF and TI are both forms of active trachoma; in other words, they represent active infection with *Chlamydia trachomatis*. Patients with active trachoma may be asymptomatic or complain of irritable, red eyes or a whitish discharge. Infection of the cornea may produce pain and sensitivity to light.
- Trachomatous scarring: the presence of scarring in the upper tarsal conjunctiva. Scars should be easily visible as white bands, lines, or sheets. This represents old, healed infection and itself is asymptomatic.
- Trachomatous trichiasis: at least one eyelash rubs on the eyeball, or evidence of recent removal of inturned eyelashes. Trichiasis is intensely irritating to the sufferer, and he or she may choose to pull out their eyelashes in an attempt to reduce discomfort. There may be discharge from superadded bacterial infection of the abraded cornea. Even in hyperendemic areas, it is unusual to observe trachomatous trichiasis in children.
- Corneal opacity: easily visible corneal opacity, at least part of which overlies the pupil. Such corneal opacities cause significant visual impairment.

It is important to remember that these grades are not mutually exclusive. A patient with active trachoma (trachomatous inflammation – follicular and/or trachomatous inflammation – intense) may also show signs of the late complications of the disease.

There are other signs of trachoma that have not been included in the simplified grading scheme:

- Papillae are often visible during active infection, but are not specific for trachoma. These are small elevations of the conjunctival surface that give the conjunctiva a velvety appearance. They are more easily seen using a slit lamp.
- A superficial punctate keratitis can be associated with invasion of the cornea by *C. trachomatis*. Slit-lamp examination (after instillation of fluorescein) is best to visualise this sign.
- Fibrovascular connective tissue may grow down from the limbus to invade the anterior layers of the superior cornea in response to infection. The ingrowth is known as a pannus. The new blood vessels may persist after resolution of the active infection.

- Sometimes follicles are found under the bulbar conjunctiva at the limbus as well as deep to the tarsal conjunctiva. Scarring of these limbal follicles may leave small depressions known as Herbert's pits.

Treatment

For active disease (trachomatous inflammation – follicular and intense) antibiotics are required. Topical tetracycline eye ointment 1% is effective when applied to both eyes twice daily for six weeks.

In addition to topical treatment, oral antibiotics are recommended for patients with trachomatous inflammation – intense (children >12 years) doxycycline 100 mg once daily for three weeks or tetracycline 250 mg four times daily for three weeks can be used. These antibiotics should not be given to children under the age of 12 years or pregnant females. A single oral dose of azithromycin 20 mg/kg (to a maximum of 1 g) has been shown to be as effective in resolving active infection as six weeks of monitored twice-daily topical tetracycline. Even when distributed on a community-wide basis, the incidence of adverse effects associated with azithromycin at this dose has been pleasingly low. The drug offers great promise for ongoing trachoma control efforts, particularly in the light of Pfizer's initiative to pilot an azithromycin donation programme in five countries in which trachoma is highly endemic.

Trichiasis or entropion requires surgical management to rotate the margin of the eyelid outwards, so that contact between the lashes and globe is interrupted. The bilamellar tarsal rotation is the procedure currently recommended by the World Health Organization; it is performed under local anaesthetic and can be undertaken at the village level by trained ophthalmic assistants.

Corneal opacity can be managed by corneal graft. Unfortunately, few endemic countries have the resources to establish a transplant programme, and because of new vessel growth from the limbus and abnormalities of the tear film in the trachomatous eye, the risk of graft rejection or failure is greatly increased.

The identification of signs of trachoma in an individual should prompt the screening of other family members.

Prevention

Blindness from trachoma is preventable. Eyelid surgery and antibiotic treatment represent tertiary and secondary prevention respectively. Primary prevention is also important if trachoma is truly to be controlled in the long term. Facial cleanliness has been shown to be associated with a decreased prevalence of active disease. Environmental changes to improve sanitation, enhance water supply and control flies provide a range of health benefits in addition to their impact on trachoma. The acronym SAFE (surgery, antibiotics, facial cleanliness, environmental change) has recently been adopted by WHO to encapsulate its approach to the fight against trachoma. WHO plans to use the "SAFE" strategy to achieve the elimination of trachoma as a blinding disease by the year 2020.

3.36 Coma

Bernhards Ogutu and Charles Newton

Minimum standards requirements

- **ABC** and intensive care
- Clinical chemistry and haematology
- Blood film for malaria
- Toxicology
- Chest *X* ray, cultures, lumbar puncture
- Neuroimaging if possible

Introduction

Coma is a state of unresponsiveness, in which the child is unable to be aroused by external stimuli, physical, verbal, sensory or inner needs. It results from a process either diffusely affecting the cerebral hemispheres or directly impairing the function of the reticular activating system in the brainstem. It may be caused by systemic disorders (metabolic encephalopathies), or intracranial diseases which are either diffuse or focal.

Coma is a medical emergency that requires immediate assessment and detection of reversible causes. Initial quick resuscitative measures are paramount, ✔

Table 3.36.1. Common causes of coma in children

Disorder	Disease entities
● Trauma	Head injury (consider child abuse)
● Seizure	Overt seizures, status epilepticus, subclinical seizures, postictal state
● Infections	Bacterial meningitis; *Streptococcus pneumoniae, Haemophilus influenzae, Nisseria meningitidis*, streptococci (group B), *Pseudomonas* species, tuberculosis Viruses: *herpes simplex*, Japanese B virus (JBV) (in Asia), *herpes zoster* Acute spirochitaemia; syphilis, Lyme disease, leptospirosis Parasitic **malaria**, ricketsial Cerebral abscess Fungal; *Cryptococcus neoformans*
● Metabolic	Hypoglycaemia (excess insulin or metabolic disorders), hyperglycaemia (diabetic ketoacidosis) Hypoxaemia secondary to cardiac/respiratory/shock Electrolyte imbalance: hyponatraemia or hypernatraemia Severe dehydration Severe malnutrition Organ failure: liver failure, renal failure, Addison's disease, respiratory failure Drugs; opiates, salicylates, organophosphates, benzodiazepines, thiazines, ***aluminium in patients undergoing dialysis,*** barbiturates, anti-depressants Other: porphyrias, Reye's syndrome
● Poisoning	Alcohol, recreational drugs, accidental/deliberate poisoning
● Tumours	Primary: meduloblastoma, astrocytoma, Secondary: leukaemias, sarcomas
● Vascular	Haemorrhage (subdural/subarachnoid), hypertension, hypotension, thrombosis, aortic stenosis, cardiac asystole Vasculitis and collagen vascular syndromes
● Systematic inflammatory response syndrome (usually with shock)	Sepsis, trauma, burns, peritonitis

before undertaking a full clinical assessment of the child.

Initial assessment and clinical examination

- Assess airway, respiration (rate, pattern and depth) and circulation by measuring: pulse or heart rate, blood pressure, capillary refill time, temperature both core and surface and oxygen saturation.
- **Stabilise the respiratory and cardiovascular systems**.
- Ascertain and treat any life-threatening conditions like airway obstruction, haemorrhage and seizures.
- Detect immediate reversible causes of coma like hypoglycaemia, hyperglycaemia and seizures (see Table 3.36.1).
- Exclude trauma.

Clinical examination is directed towards identifying signs suggesting causes, extent of injury and level of consciousness. A general examination should be undertaken; the depth guided by history and presumptive cause of coma. Look for rashes (for example purpura of meningococcaemia), tick bites, signs of trauma, evidence of ingestion of drugs or chemicals and evidence of organ failure.

History

A detailed history should be taken from the parent or guardian, preferably one who was around at the onset of the coma. It should focus on the possible cause of coma, rate of development of unconsciousness, extent of injury, signs of deterioration or recovery and past medical history.

Examination

- Pulse:
  Bradycardia may indicate raised intracranial pressure or reflect the effects of poisons or drug overdoses.
- Blood pressure:
  Hypertensive encephalopathy or signs of raised intracranial pressure (RICP).
- Temperature:
  sepsis.
- Respiratory pattern:
  Irregular due to brainstem lesion or raised intracranial pressure (RICP)
  Rapid due to acidosis or aspirin ingestion
  Slow due to opiate ingestion.
- Pupil size and reactivity:
  Small due to opiate ingestion
  Large due to amphetamine ingestion and RICP

Coma scales have been devised to measure the depth of coma, improve agreement between clinicians and avoid the use of ambiguous words such as "stuperous." The widely used ones are the Glasgow Coma Score or the paediatric modification (Adelaide Coma Score) (Table 3.36.2).

Table 3.36.2 Coma scales

	Glasgow Coma Scale (4–15 years)			Adelaide Coma Scale (<4 years)	
Activity	**Best response**	**Score**	**Activity**	**Best response**	**Score**
Eye opening	Spontaneous	4	Eye opening	Eyes open spontaneously	4
	To verbal stimuli	3		To request	3
	To pain	2		To pain	2
	None	1		No response to pain	1
Verbal	Orientated	5	Verbal	Orientated, alert	5
	Confused	4		Recognisable and relevant words but less than usual. Spontaneous cry	4
	Inappropriate words	3		Cries only to pain	3
	Incomprehensible sounds	2		Moans only to pain	2
	None to pain	1		No response to pain	1
				Obeys commands	6
Motor	Follows commands	6	Motor	Localises painful stimulus	5
	Localises pain	5		Withdrawals from pain	4
	Withdraws in response to pain	4		Abnormal flexion to pain (decorticate)	3
	Abnormal flexion in response to pain (decerebrate)	3		Abnormal extension to pain (decerebrate)	2
	Abnormal extension in response to pain (decerebrate)	2		No response to pain	1
	No response to pain	1			

Refer to Figure 3.36.1 on where and how to perform painful stimulation and Figure 3.36.2 for motor responses.

Figure 3.36.1 Areas for exerting painful stimuli.

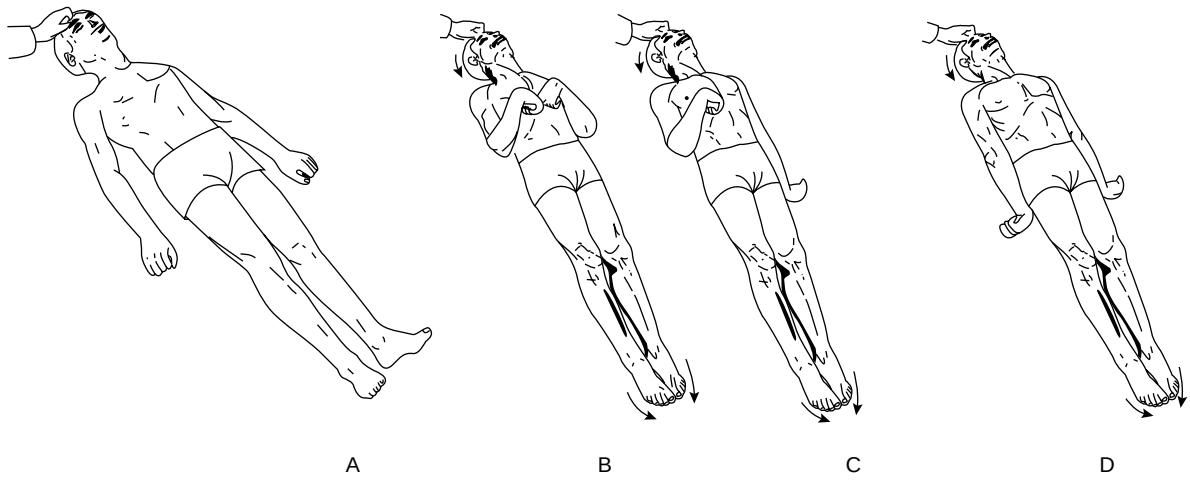
Unequal/unreactive due to raised intracranial pressure (RICP).

- Skin rashes:
  Infectious, for example meningococcal septicaemia, Dengue fever.
- Breath odour:
  Diabetic ketoacidosis, alcohol ingestion, inborn errors of metabolism.
- Hepatomegaly:
  Reye's syndrome, other metabolic disorders.
- Fundi:
  Papilloedema, retinal haemorrhages.
- Posture/oculocephalic reflexes (Figure 3.36.2):
  Abnormal in raised intracranial pressure.

The purpose of neurological examination is to establish a baseline and identify features of raised intracranial pressure (including herniation syndromes), focal deficits (for example space-occupying lesions) and lateralising signs (hemiplegic syndromes). Examination may also help in providing prognostic information.

Cranial nerves

- Pupillary reactions
  - Use a bright torch
  - Consider the effect of drugs used in treatment for example benzodiazepines for fits.
- Ocular movements
  - Eyelid response
  - Corneal response
- Oculocephalic reflexes (Figure 3.36.3)
  - Turn the head sharply to one side, the eyes move to the opposite side in the normal state.
  - If the eyes only partly deviate or remain fixed then this is abnormal.
  - **Check that there is no cervical injury before performing the manoeuvre**.



A, motionless; B, decorticate; C, decorticate/decerebrate; D, decerebrate.

Figure 3.36.2 Posture positions.

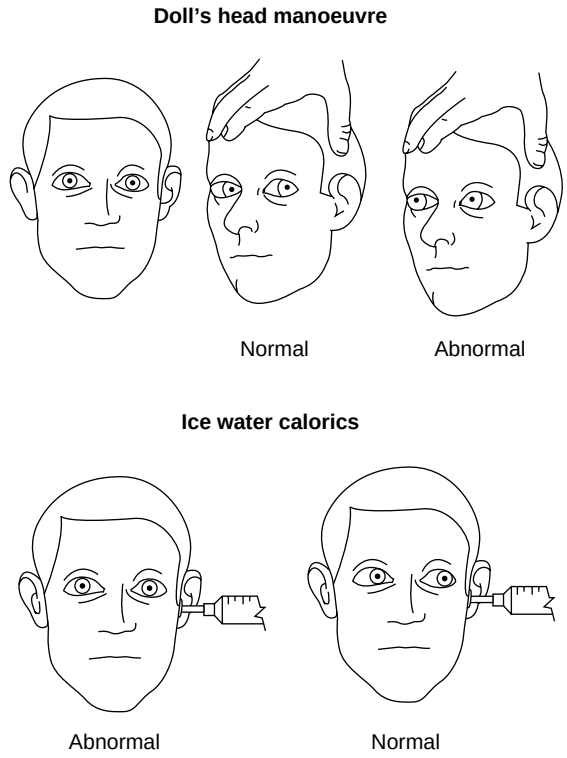


Figure 3.36.3 Oculocephalic and oculovestibular responses.

Plum F, Posner JB. "*The Diagnosis of Stupour and Coma*", ed. Davies FA Philadelphia: Saunders, 3rd edition 1980.

- Oculovestibular or caloric response (Figure 3.36.3)
  - Ascertain the tympanic membrane is intact and there is no wax in the external meatus.
  - Tilt the head forward at 30°, instill ice cold water into the ear. The eyes turn to the side of the stimulus in case of a normal brainstem.

Motor function

- Motor activity, i.e. tremor, multifocal, or none
- Motor response or postures, i.e. normal, decerebrate state, decorticate rigidity, flaccid, extension or flexion of contralateral limbs (Figure 3.36.2)

Respiratory pattern

- May be difficult to identify in children.
  - Irregular: consider seizures
  - Cheyne–Stokes: raised intracranial pressure, cardiac failure
  - Kussmaul: acidosis, central neurogenic hyperventilation, midbrain injury, tumour or stroke.
  - Apneustic (periodic) breathing: pontine damage, central herniation
- Signs and symptoms of raised intracranial pressure (RICP)
  - Preceding history of headache
  - Recurrent vomiting
  - VIth cranial nerve palsy
  - Sluggish or no pupillary reactions
  - Dilated retinal veins with reduced pulsations
  - Papilloedema
  - Subhyaloid retinal haemorrhages

Investigations

The investigations are guided by the presumptive clinical diagnosis.

Essential investigations

- Clinical chemistry: blood glucose, electrolytes, creatinine, urea, ***blood gases***, liver function tests (including clotting)
- Blood film for malarial parasites
- Haematological parameters: full blood count, peripheral blood film, blood clotting
- ***Toxicological tests, i.e. salicylates, organophosphates, opiates, alcohol, paracetamol***
- Blood cultures
- Lumbar puncture in case of high index of suspicion of central nervous system infection. **It should be delayed if there are features of raised intracranial pressure, the child is too sick to tolerate the flexed position needed to do a lumber puncture, infection at puncture site, bleeding tendency and rash of meningococcal septicaemia**. The child should be given antibiotics to cover the possibility of bacterial meningitis ✔
- Chest *X* ray: tuberculosis, severe pneumonia

If facilities are available

- ***Neuroimaging: computerised tomography or magnetic resonance imaging***
  Particularly useful in detecting space-occupying lesions, traumatic injury
  Contrast dye should be given if an infection or a tumour is suspected
- ***Plasma ammonia and plasma/CSF lactate.***
- ***Urine/plasma for organic/amino acids***

Other investigations

These will depend on the cause of the coma.

- ***Hormonal assays, i.e. thyroid hormones, cortisol, ketosteroids (adrenal insufficiency)***
- ***Neurophysiological tests***
  - ***Electroencephalography: (EEG)***
    To detect seizures
    To identify unilateral lesions
    May identify encephalitis
    May be useful in prognosis
  - ***Evoked potential responses: – to detect brainstem lesions.***
- ***Neuroimaging: CT scan, MRI scan, Magnetic resonance angiography***

Differential diagnosis

Determine whether the cause is primarily intra- or extracranial. Intracranial conditions may be subdivided into those with or without focal signs. Extracranial causes include encephalopathies arising from metabolic derangements or exogenous toxins. The common causes are as shown in Table 3.36.1.

Management

Prognosis depends on the aetiology, age of the child and the state of the patient i.e. the level of consciousness on admission and the initial response to appropriate interventions. The presumptive cause of coma guides the treatment. Consider the following interventions.

Immediate management

- Support respiration if respiratory effort is not adequate to maintain desired oxygen saturation and/or carbon dioxide excretion.
- Support circulation to maintain adequate cerebal perfusion (aim to keep systolic blood pressure at normal values for age: avoid hypotension).
- Maintain normoglycaemic state, be cautious of administering insulin to hyperglycaemic patients, since hyperglycaemia may be stress induced.
- Assess and maintain electrolyte balance (avoid hyponatraemia: use 0.9% saline plus added glucose NOT 0.18% saline, 4% glucose). If possible keep serum sodium in normal range, >135–145 mmol/litre.
- Treat seizures if present and give prophylactic anticonvulsants if the child has repeated seizures.
- Insert nasogastric tube to aspirate stomach contents. Perform gastric lavage in circumstances such as drug or chemical ingestion.
- Regulate temperature (avoid hyperthermia: that is above 37.5°C)
- Undertake appropriate medical management of raised intracranial pressure if noted:
  - ***Support ventilation (maintain a PCO_2 of 3.5–5.0 kPa)***
  - Mannitol (250–500 mg/kg; that is 1.25–2.5 ml/kg of 20% IV over 15 minutes, and repeat as required based on response and clinical signs, ***provided serum osmolality is not greater than 325 mosm/litre*** (max. total dose 2 g/kg)
  - Dexamethasone (for oedema surrounding a space-occupying lesion) 0.5 mg/kg 6 hourly
- Catheterisation for bladder care and urine-output monitoring
- Plan for continued regular clinical assessment, mainly nursing observations.

Intermediate management

- Prevent child falling out of the bed.
- Nutritional support: parenteral and/or oral to prevent malnutrition during period of unconciousness.
- Skin care: prevent bed sores by turning the patient.
- Eye padding to avoid xerophthalmia.
- Family counselling, support and consent in case of invasive procedures.
- Appropriate surgical intervention if indicated.
- Chest physiotherapy to avoid hypostatic pneumonia.
- Restrict fluids to 80% of maintainance if evidence of water retention is seen (provided blood pressure is normal).
- Prevent deep vein thrombosis by physiotherapy/stockings.
- Maintain oral and dental hygiene.
- Appropriate care for central and peripheral venous or arterial access to avoid infection by maintaining sterility when handling the sites.
- Watch out for nosocomial infection.

Long-term management

Rehabilitation, family education and support for disabilities that may arise. Recovery can occur up to two years following admission. Some deficits, (for example cortical blindness), recover faster and more completely than others (for example quadriplegia or deafness). Seizures need to be looked for, since epilepsy can be treated.

The most important issue is prioritization of the management steps of the individual child, depending on the cause of coma and the level of care required.

3.37 Epilepsy

Deb K Pal and Charles Newton

Minimum standards requirements

- Anticonvulsants: phenobarbitone, phenytoin, sodium valproate, carbamazepine, ethosuximide
- Temperature control
- Prednisolone
- ***EEG and neuroimaging if possible***

Epilepsy is a symptom caused by a CNS disorder, and is usually defined as the occurrence of two unprovoked seizures. In over 70% of cases, a cause cannot be identified (idiopathic epilepsy); although genetic causes may be important, since there is often a family history. Most children with epilepsy live in disadvantaged countries where the incidence rates are estimated to be twice that of Western countries and where more than 70% are untreated.

The impact of epilepsy on children and families is wide. To reduce disability, management is best shared with other workers who can visit the family closer to home, such as community doctors, health or disability workers.

Confirming the diagnosis of epilepsy

There is no justification for a trial of anti-epileptic drugs if the diagnosis is unclear. The diagnosis of epilepsy is purely a clinical one and rests usually on a good history or eyewitness account or ideally a home video of an event.

Features of the seizures that are important:

- Timing and duration
- Provocation factors
- The early phase of the attack
- Movements
- Sensory symptoms
- Level of responsiveness
- Nature of offset.

In early childhood, breath-holding attacks, reflex anoxic seizures and febrile syncope may be commonly mistaken for epileptic seizures. Syncope, hypoglycaemia and non-epileptic attacks also enter the differential diagnosis (see Chapter 2.6).

Defining the epilepsy syndrome

See Table 3.37.1 overleaf.

Prognostic features of epilepsy

When a syndrome cannot be identified precisely, then the following features can serve as a guide to prognosis.

Table 3.37.2 Prognosis in epilepsy

Good outcome	Adverse outcome
Single seizure type	Multiple seizure types
No additional impairment	Additional neurological impairment (especially cognitive)
Late age at onset (for the syndrome)	Early age at onset (for the syndrome)
Provoked by illness, stress, flashing lights	Unprovoked
Short seizures	Status epilepticus
Low frequency of seizures	High frequency of seizures
Good initial response to anti-epileptic drugs	Poor initial response to anti-epileptic drugs, requiring polytherapy

Once the diagnosis and prognosis have been assessed, assemble a problem list:

- What effect does the epilepsy have on the development of the child?
- Are learning or motor problems present?
- Is the child attending school and getting opportunities to play with other children?

Role of investigations

The history and sometimes the examination will usually indicate the cause. Children can be managed without the need for EEG or neuroimaging. ***EEG and computer tomography (CT) should be reserved for intractable cases*** or those with neurological signs suggesting a space-occupying lesion. **Such problems and the imaging required to identify them, will usually require the support of a specialised neurosurgical centre, at least** ✔

Table 3.37.1 Clinical classification: most common syndromes of epilepsy in children

Syndrome	Features	Treatment
Generalised		
Tonic or tonic-clonic (Grand mal)	Loss of consciousness Stiffening, convulsive movements Incontinence Post-ictal drowsiness, headache	Phenobarbitone, phenytoin sodium valproate, carbamazepine
Absences	Vacant stare, with decrease in awareness, responsiveness and memory. Precipitated by hyperventilation	***Ethosuximide****, sodium valproate. Avoid carbamazepine and phenytoin
Myoclonic	Head nodding or jerks of limbs	***Ethosuximide****, sodium valproate. Avoid carbamazepine and phenytoin
Infantile spasms	Sudden flexion of head, trunk and limbs; sometimes extensor spasms Associated with hypsarrhythmia on the EEG and developmental delay	Steroids (requires careful monitoring, avoid if this is not feasible) sodium valproate
Lennox–Gastaut syndrome	Multiple seizure types: tonic (especially night), atonic, absences, generalised tonic-clonic. Associated with slow spike and wave and developmental delay	Sodium valproate, carbamazepine, phenytoin and combinations
Secondary generalised seizures	Seizures starting as partial, developing (sometimes rapidly) into generalised tonic-clonic seizures	Carbamazepine, sodium valproate, phenytoin, phenobarbitone
Partial		
Simple	Convulsive movements involving eyes, face, parts of limbs. May have sensory symptoms	Carbamazepine, sodium valproate
Complex	Aura of abdominal discomfort, vacant stare, loss of contact with surroundings, lip smacking, chewing, swallowing, facial flush, hallucinations. Post-ictal tiredness and headaches May produce apnoea	Carbamazepine, sodium valproate phenobarbitone
Benign epilepsy of childhood with Rolandic spikes	Most common, usually starts age 3–10 years. Predominantly simple partial seizures involving oropharyngeal muscles (gurgling) face or limbs, mostly during sleep Characteristic EEG, normal intelligence	Often do not need treatment. Carbamazepine, sodium valproate, phenobarbitone

* *Treatment may not be affordable*.

one of which should exist in every country. Skull *X* ray is not useful in established epilepsy.

Selecting rational anti-epilepsy drugs

Phenobarbitone, phenytoin, carbamazepine and sodium valproate should be available.

Convulsive status epilepticus

(see Chapter 3.38)

How to start treatment?

- Monotherapy is the aim, to reduce the side effects and interactions.
- Try to avoid drugs which impair development; for example, phenobarbitone except in infancy.

- If possible, always prescribe the same brand, as there may be pharmacodynamic differences.
- Always start in low doses to minimise side effects and improve chances of compliance.
- Remember to warn about any likely side effects, especially if they are temporary, such as drowsiness.
- Increase the dose of drug gradually until the maintenance range is reached, for example two weeks or so between increments.

How to monitor treatment?

- Case notes should record the diagnosis, problem list, dates and types of seizures, indication for treatment, past treatment with response and side effects of treatment, and information that has been given to child and guardians.
- Hand out medical cards to be kept as a seizure diary, reminder of prescription and clinic dates. Graphic symbols can be used for the illiterate.
- Regularly review the child to check on progress, more often if seizure control has not been attained, or there are side effects or drug changes.

When to change treatment?

- Consider changing treatment if side effects are troublesome.
- Introduce the second anti-epileptic drug in the normal way, first checking for possible drug interactions. Once established, begin to withdraw the first anti-epileptic gradually (see Table 3.37.3). If seizure control is not attained with monotherapy, seek specialist referral.

Whom to refer?

This depends upon local facilities. One-third of patients will be intractable to treatment with first-line anti-epileptic drugs. Some of them may not have epilepsy, others may have poor prognosis syndromes. They will require specialist assessment and treatment advice. Epilepsy may also be a part of complex developmental disorders involving the CNS and these children may also benefit from specialist input.

When and how to stop treatment?

In children with a good prognosis, 12–24 months' seizure freedom is associated with a 70% chance of continuing seizure remission. Withdrawal must be a gradual and closely monitored process. If seizures recur after a decrement, they usually remit once the last decrement has been reversed. Withdrawal period depends on the drug, for example phenobarbitone over 4–6 months, carbamazepine over 2–3 months.

Social issues

Promoting social integration

Children need to participate as fully as possible in the normal activities of their peers, at school, at play, in the home and preparing for employment. Community workers should be involved in the wider management, and parents' fears and anxieties discussed.

Supporting parents

Parents often tend to overprotect their children who have epilepsy, and may lack confidence in dealing with seizures. In many societies epilepsy carries a stigma. Opportunities to discuss first aid, behaviour, and other concerns are vital and can be provided by health workers or parent groups.

First aid advice

The general theme to be emphasised is that children with epilepsy should be encouraged to live as full and normal life as possible. There are very few absolute restrictions, these include climbing trees or riding bikes or motorcycles. Children should be accompanied when swimming or around hazards such as stoves and fires. During a convulsion **place in the recovery position**, protect the person from hard or sharp objects in the vicinity and cushion the head. **Do not put anything in the mouth or try to restrain the limbs**. Let the person recover by themselves; they may need to rest or sleep. ✔

Table 3.37.3 Anti-epileptic drugs

Drug	Usual dose	Side effects and toxicity
Carbamazepine	10–20 mg/kg/day in 2 divided doses daily starting dose: 5 mg/kg/day	Ataxia, diplopia, rashes, aplastic anaemia (watch for mouth ulcers, bruising or other signs)
Phenobarbitone	3–5 mg/kg/day <5 years 2–3 mg/kg/day >5 years Give once daily at night or twice daily Starting dose: 1.5 mg/kg/day	Drowsiness, agitation, rashes Developmental impairment
Phenytoin	8–10 mg/kg/day <3 years 4–7 mg/kg/day >3 years Give once daily or twice daily starting dose: 3 mg/kg/day	Gum hypertrophy, hirsutism, acne, ataxia, diplopia, nystagmus, neuropathy, choreoathetosis, encephalopathy, lymphoma, megaloblastic anaemia
Sodium valproate	15–40 mg/kg/day, twice daily Starting dose: 10 mg/kg/day	Nausea, epigastric pain, alopecia, weight gain, tremor, hepatitis, pancreatitis, encephalopathy

Febrile seizures

A febrile seizure is a seizure that occurs in children aged between 3 months and 7 years with febrile illness not caused by an intracranial disease. The commonest age of onset is 14–18 months. Febrile seizures are common, occur in 2–5% of all the children and account for about one-third of all childhood seizures.

Clinical presentation

Febrile seizures are usually brief, generalised, clonic or tonic-clonic convulsions lasting less than 10 minutes with minimal post-ictal confusion or weakness. About 20% of febrile seizures are complex, i.e focal, or last longer than 15 minutes or occur more often than once in 24 hours. Complex febrile seizures may suggest an underlying central nervous system cause and are associated with a poorer outcome.

Febrile seizures occur while the child has a recognisable infection, most commonly upper airway infections and viral illnesses such as gastroenteritis. Other causes include pneumonia, urinary tract infections and after vaccinations. Shigellosis, roseola infantum and malaria have an unusually high incidence of seizures. Most children have a core temperature of 38–41°C but it may occur at the onset of the febrile illness and the child may have a normal temperature at the time of seizure.

An increased frequency of febrile seizures occurs in children of parents and siblings who have had febrile seizures and siblings with epilepsy.

Identify the cause

Check blood glucose (fingerprick if available), film for malaria parasites, urea, creatinine, calcium, electrolytes and full blood count. Consider urinalysis, lumbar puncture, cultures of blood, urine, pharyngeal swab and cerebrospinal fluid, relevant *X* rays. A lumbar puncture is mandatory when meningitis is thought to be a possibility, particularly in children less than 2 years of age. (Unless evidence of raised intracranial pressure when IV antibiotics should be given anyway).

Differential diagnosis

Exclude:
- Encephalitis
- Acute encephalopathies of metabolic or toxic origin
- Cerebral malaria
- Electrolyte disorders
- Hypoglycaemia
- Anoxia
- Trauma
- Haemorhage
- Tumour.

Other entities that can be confused with febrile seizures are:
- Febrile delirium
- Febrile rigors.

Treatment (see Chapter 3.38)

The choice of the anticonvulsant depends on the availability.
- Lorazepam is a new drug, with a half-life longer than diazepam.
- Fosphenytoin is a prodrug of phenytoin but unlike phenytoin can be given intramuscularly. It can be given as a bolus intravenously.
- Children with prolonged seizures need to have their vital signs monitored. Anticonvulsants can cause hypotension and respiratory depression.
- If seizures are not controlled by the long-acting anticonvulsant; there is a role for general anaesthesia and muscle relaxants with respiratory support in the centres that have the facility.
- A second single febrile seizure lasting less than 5 minutes does not warrant treatment, but if the child has multiple seizures then a prophylactic anticonvulsant should be started.
- Parents/guardians are often frightened by seizures in their children. They should be REASSURED and told that febrile seizures are common in children and this does not mean that the child will have brain damage. They should also be educated on prevention and management of seizures at home.

Sequelae

- About one-third of children with febrile seizures will have another febrile seizure.
- 3% will have at least one afebrile seizure.
- 2% may develop epilepsy (recurrent afebrile seizures).
- Approximately 65% of children with simple febrile seizures will have had no further seizures by 7 years of age.
- Recurrent seizures tend to re-occur, particularly in children aged less than 1 year or those with a positive family history.

Epilepsy

The risk factors for the development of epilepsy are:
- Complex febrile seizures
- Prior abnormal neurological function
- Multiple febrile seizures
- Family history of epilepsy
- Age of less than 1 year at the first seizure.

Long-term care: home treatment

- Rectal diazepam for parents to administer in case of prolonged seizures (2.5 mg for <1 year, 5 mg for 1–3 years, 10 mg for >3 years).
- Oral or rectal paracetamol to prevent/treat febrile seizures.
- Advice to parents.

3.38 Management of prolonged seizures

Bernhards Ogutu and Charles Newton

Minimum standards requirements: Status epilepticus

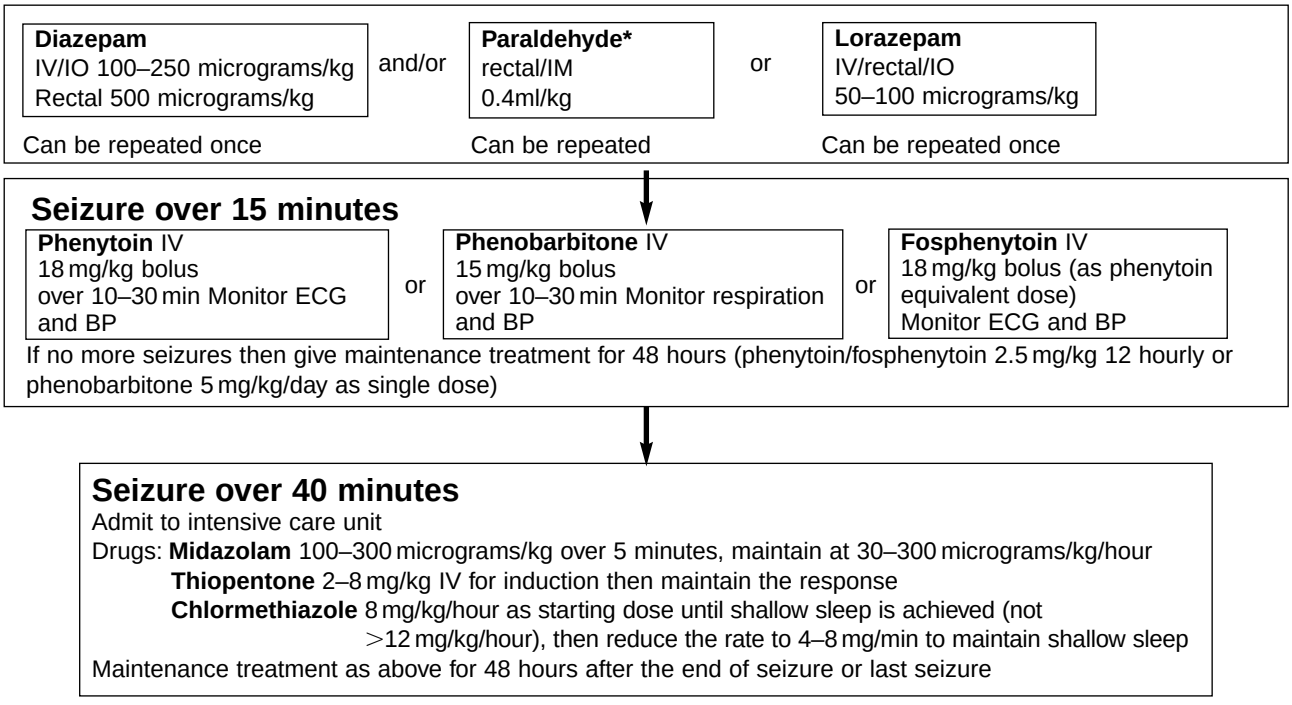
- **ABC** and intensive care
- Anticonvulsants: diazepam, paraldehyde, lorazepam, midazolam, chlormethiazole, thiopentone
- Temperature control
- Mannitol
- Dexamethasone

Evaluation and immediate management of a major fit or status epilepticus

During a seizure

- Put the child onto a left lateral decubitus position.
- Ensure the airway is patent and there is adequate respiratory effort and adequate circulatory volume.
- If the seizure lasts more than 5 minutes (or the duration is not known) anticonvulsant treatment should be given. Short recurrent seizures lasting less than 5 minutes should also be treated (see flow chart in Figure 3.38.1).
- Treat the fever if present by exposure, tepid sponging and rectal paracetamol (40 mg/kg loading dose, 20 mg/kg if <3 months).

In case of excessive respiratory depression from benzodiazepines have available flumazenil (see Chapter 5.12).



*Plastic syringe can be used if dose given immediately. Maximum 5 ml IM at one site.

Figure 3.38.1 Treatment of major seizures

3.39 Neuropathies

Allie Moosa

Minimum standards requirements

- Muscle biopsy
- Prevention of scoliosis
- Prevention of muscle contractures

Neuropathies are diseases affecting:
- The anterior horn cell
- The peripheral nerves.

Anterior horn cell disease

The most common are:
- Poliomyelitis (see Chapter 4.21)
- Spinal muscular atrophy.

Spinal muscular atrophy

This is a motor neurone disease of the spinal cord and brain stem, inherited as an autosomal recessive disorder and associated with deletions of the survival motor neurone (SMN) and neuronal apoptosis inhibitory protein (NAIP) genes. It is the second commonest autosomal recessive disorder after cystic fibrosis.

Clinical features

These children have delayed motor development but normal social, language and intellectual development. They are floppy and weak. The weakness is proximal more than distal and affects lower more than upper limbs. They are areflexic and fasciculation of the tongue is diagnostic (observed with the tongue at rest in the mouth).

There are three clinical subtypes depending on severity.
- **Severe infantile type**: These infants never sit, crawl or walk. The onset is before or soon after birth. They have severe intercostal and bulbar weakness but the diaphragm is spared. Most die from respiratory failure before their second birthday.
- **Intermediate type**: These infants can sit but are unable to walk. They may or may not have respiratory and bulbar weakness and this factor determines their prognosis. If absent these children can survive into adulthood.
- **Mild type (also known as Kugelberg–Welander type)**. The onset is later and the children can walk but do so late and with difficulty. Respiratory and bulbar weakness are usually not present. A coarse tremor of the hands is frequently seen in this and the intermediate form. This is a useful sign to distinguish this type from muscular dystrophy with which it is often confused (see Table 3.39.1).

Table 3.39.1 Duchenne muscular dystrophy versus spinal musclar atrophy (mild type)

Features	Spinal muscular atrophy	Duchenne muscular dystrophy
Motor milestones	Delayed	Normal or delayed
Hypotonia	+++	+/−
Pseudohypertrophy	+/−	+++
Tremor of hands	+++	−
Fasciculation of the tongue	+	−
IQ	Normal	Normal or low
ECG	Baseline tremor	R and Q waves changes
Creatine kinase	Normal or slightly increased (×2)	Very high (×100)
EMG	Chronic denervation	Myopathic
Muscle biopsy	Denervation	Dystrophic

+ mild, +++ marked, − absent

Diagnosis (see Table 3.39.1)
Since the discovery of the gene defect, muscle biopsy is rarely needed. Deletion of the *SMN* gene is found in almost all cases of spinal muscular atrophy of all three types. It can be detected rapidly by the polymerase chain reaction. Blood (2–5 ml in EDTA tube) or DNA extracted from it, can be sent ***by post to a laboratory doing the test and diagnosis confirmed within a few days***.

Management
Management is supportive. The most important complication in the intermediate form is the development of scoliosis. This can be delayed by getting the child to stand with support of lightweight calipers for as long as possible. If the child is confined to the wheelchair, a brace may be required to control the scoliosis. ***Surgery (fusion of spine) may be necessary***.

Peripheral neuropathy

The two commonest causes of peripheral neuropathy in children are Guillain–Barré syndrome (see Chapter 3.40) and hereditary motor and sensory neuropathy.

Hereditary motor and sensory neuropathy

This is the commonest chronic peripheral neuropathy in children. It is progressive. There are several types but the commonest is Type I (peroneal muscular atrophy). It is dominantly inherited and most children are asymptomatic until late childhood, when unsteady clumsy gait with frequent falls occur. There is weakness and wasting of the muscles of the anterior compartment of the leg. The parents are often asymptomatic. The diagnosis is confirmed by ***finding very low motor conduction velocities in both the patient and one of the parents indicating demyelination. Type II is similar but rare and shows axonal rather than demyelinating changes on nerve conduction studies***. There is no treatment for these diseases other than special boots and ankle orthosis to stabilise the ankle.

Other peripheral neuropathies

These include leucodystrophies (where peripheral nerve demyelination occurs as part of CNS demyelination), **toxic neuropathy (glue or benzene sniffing, lead, drugs) and diphtheria**. ✔

3.40 Guillain–Barré syndrome

Allie Moosa and Charles Newton

Minimum standards requirements

- ABC and intensive care
- Lumbar puncture

This is the commonest peripheral neuropathy seen in childhood. It is a demyelinating neuropathy induced by an autoimmune process precipitated by a preceding viral or other infection. It has a peak incidence around 8–9 years in rich countries and 3–4 years in disadvantaged ones, possibly due to overcrowding. Rarely, an acute axonal form occurs especially in some countries like China.

Clinical features

The onset is usually acute. There is often a history of a preceding upper respiratory tract infection and insidious sensory symptoms, for example muscle tenderness, occasionally an unsteady gait and frequent falls. The weakness starts in the lower limbs and ascends to affect the trunk, upper limbs, respiratory (intercostal and diaphragm), bulbar and facial muscles. It is usually symmetrical and affects both proximal and distal muscles and may take 10–30 days to reach its maximum. Cranial nerve involvement often precedes respiratory difficulties. Reflexes are absent. Sensory loss is minimal and of the "glove and stocking" distribution. Ophthalmoplegia, papilloedema and bladder involvement rarely occurs. Autonomic dysfunction occurs in many children resulting in hypertension, hypotension and cardiac arrhythmias. In some patients the paralysis occurs rapidly with quadriplegia and respiratory paralysis within 2–5 days.

Chronic inflammatory demyelinating polyradiculoneuropathy

The disease may evolve into chronic inflammatory demyelinating polyradiculoneuropathy. This disease is similar to Guillain–Barré syndrome, consists of a progressive or relapsing motor and sensory dysfunction, lasts at least two months, with hyporeflexia of all four limbs. The importance of identifying this condition is that it responds to steroids (prednisolone 2 mg/kg/day).

Diagnosis

The diagnosis is confirmed by finding a high CSF protein and low CSF lymphocyte count. These findings may not be present in the first week after onset. Other causes of acute flaccid paralysis need to be considered (Table 3.40.1).

Management

- Admit the child to monitor for impending respiratory and bulbar paralysis and autonomic dysfunction.
- Measure respiratory and heart rate, blood pressure, ***vital capacity*** (or peak flow) and airway protection frequently. ***Blood gas analysis may be required and helpful.***
- ***If the vital capacity is <50% normal for age and/or there is significant respiratory failure with hypoxaemia and hypercapnia, ventilate the child if possible.***
- If bulbar and respiratory paralysis ensues, airway protection, tube feeding and ventilatory support will be necessary. Airway protection can be achieved by intubation or tracheostomy.
- Children who require ventilation, can be given ***high-dose human immune globulin if available, 2 mg/kg IV over 6 hours. This can be repeated once or twice over the next few days***.

Prognosis

Recovery is usually complete within 4–6 months in most children but may take up to 2 years. About 5% of children will have minor motor sequelae. About 2–3% will die from respiratory failure or autonomic dysfunction. Poor prognostic factors include onset of weakness within 8 days of preceding infection, rapid progression, cranial nerve involvement and a CSF protein of >800 mg/litre in the first week of the disease. The prognosis is generally better in children than adults.

Table 3.40.1 Acute flaccid paralysis

Cause	Features
Spinal cord	
Poliomyelitis	Preceding fever, headache and meningeal irritation Asymmetrical weakness, CSF pleocytosis
Enterovirus: Japanese B encephalitis	Similar to poliomyelitis
Trauma	History and evidence of trauma
Myelitis	Paraplegia, segmental sensory loss Bladder and bowel sphincter disturbance
Epidural abscess	Fever, vertebral tenderness
Neuropathies	
Guillain–Barré virus	Symmetrical, ascending weakness
Diphtheria	Preceding history of diphtheric pharyngitis, cardiac involvement, deep sensation impaired
Botulism	Bulbar symptoms before onset of weakness, ophthalmoplegia
Tick paralysis	Rapid progressive paralysis, no sensory loss Normal CSF protein Resolves quickly once tick removed
Metabolic	
Acute intermittent porphyria Hereditary tyrosinaemia	Family history, other symptoms
Muscle	
Myasthenia gravis (rare but treatable)	Fluctuating weakness worse with activity better after rest Tensilon test positive
Acute viral myositis	Tender muscles, high ***creatine kinase level***
Other causes	
Organophosphate poisoning	History of exposure, excessive salivation, twitching of muscles, meiosis, tachycardia

3.41 Muscular dystrophies

Allie Moosa

Minimum standards requirements

- Creatine phosphokinase measurement
- Prevention of scoliosis and contractures
- Prednisolone

A group of inherited disorders that cause progressive muscle weakness and share a common pathological process of muscle fibre degeneration and fibrosis.

Duchenne muscular dystrophy

This is the most common muscular dystrophy, caused by deficiency of dystrophin, a structural protein found on the inner side of the sarcolemmal membrane. The deficiency is caused by deletions or point mutations of the dystrophin gene which is located on the short arm of chromosome Xp21.

Clinical features

Duchenne muscular dystrophy is X-linked; it affects boys and is transmitted by females. Affected infants are normal in the first two years but will have very high serum creatine kinase levels. They usually present between 2–5 years with delayed walking, frequent falls, difficulty in climbing stairs and in getting up from the floor. The weakness affects proximal more than distal muscles and the pelvic more than the shoulder girdle. Facial muscles are unaffected. Prominent calves and thighs are characteristic. With time, they walk on their toes with marked lumbar lordosis and a waddling gait. The arm reflexes are lost early but ankle jerks are preserved. Once confined to a wheelchair, they rapidly develop contractures of the knees, hips, ankles and scoliosis. Intellectual impairment may occur or develop in some patients, often related to the onset of respiratory failure. The ECG shows dominant R waves in right-sided and deep Q waves in left-sided leads and inverted T waves in most patients.

Diagnosis

The serum creatine kinase is very high (100 times normal). ***The EMG is myopathic*** and muscle biopsy shows dystrophic changes and absent dystrophin. ***Deletions in the dystrophin gene can be identified by the polymerase chain reaction in DNA extracted from blood in 60–80% of patients***.

Management

There is no effective drug treatment. A course of oral prednisolone (0.75 mg/kg/day) for 3–6 months can produce a small but significant improvement in muscle strength, but has many side effects and these must be weighed against the slight benefit. Night splints to keep the ankles at 90° may delay shortening of the tendoachilles. When walking is becoming difficult, the fitting of lightweight calipers and intensive physiotherapy may keep the child ambulant a few more years. Once wheelchair-bound, a rigid seat and adequate postural support of the spine, may prevent scoliosis.

Prognosis

The weakness is progressive and by 10–12 years, a wheelchair is needed. Later as respiratory muscle weakness develops, nocturnal hypoventilation may cause disturbed sleep and morning headaches (due to hypoventilation and carbon dioxide retention). Assisted non-invasive ventilation (using a nasal mask) at night will improve the quality of life (see Chapter 1.26). Death occurs in the twenties from respiratory or cardiac failure.

Genetic counselling

An elevated creatine kinase (on three separate occasions) in female relatives, indicates carrier status. Some will also have an abnormal muscle biopsy with some fibres showing normal and others absent dystrophin. Normal creatine kinase and muscle biopsy does not exclude the carrier state. ***Prenatal diagnosis is possible in some but not all families but requires specialised molecular genetic techniques***.

Becker muscular dystrophy

This is a milder variant of Duchenne muscular dystrophy and is rare. Onset is between 5–10 years and ambulation is maintained beyond 15 years and often into adulthood. It is caused by partial deficiency of dystrophin.

Severe autosomal recessive muscular dystrophy of childhood

This is rare but prevalent in North Africa. It has a clinical picture similar to Duchenne muscular dystrophy but affects boys and girls. It is associated with deficiency of one of the sarcoglycans, a group of sarcolemmal proteins intimately linked to dystrophin. This can be demonstrated in muscle biopsy specimens. The intelligence remains normal and the heart is unaffected.

Congenital muscular dystrophy

Infants with congenital muscular dystrophy are born floppy and weak and have contractures. They have delayed motor milestones and most are unable to walk. They develop a characteristic long, thin, expressionless face with open mouth. There is no ophthalmoplegia. There are several subgroups, some with eye or brain abnormalities and demyelination. Merosin is deficient in one subgroup. The creatine kinase may be elevated or normal and the diagnosis is made by muscle biopsy. Management is supportive. Congenital muscular dystrophy must be distinguished from other causes of hypotonia (other congenital myopathies, infantile spinal muscular atrophy, organic acidaemias).

3.42 Breath-holding spells

Allie Moosa

Minimum standards requirements

- Haemoglobin measurement
- Oxygenation measurement (see Chapter 3.19)

(See also Chapter 2.6)
Breath-holding spells occur in about 4% of children under the age of 5 years. They typically start between the age of 6–18 months and usually cease before the age of 5 years. They are infrequent (less than one per month) but occasionally occur more often. There are two types of spells differentiated by the presence of **cyanosis** or **pallor**. The mechanism of the two types are different.

Type of spells

- **Cyanotic breath-holding spells**
  These are provoked by anger, frustration, fright and pain. The infant cries vigorously, holds his/her breath in expiration, goes blue, loses consciousness and becomes limp. Rarely this is followed by brief stiffening of the body. The infant then starts breathing again and the attack ends. The attacks may be due to cerebral ischaemia from sudden rise in intrathoracic pressure that impedes venous return; intrapulmonary right–left shunting also plays a part.
- **Pallid breath-holding spells**
  These are less common than the cyanotic type (about one-fifth of all cases). The attack is provoked by pain usually from a mild bump on the head. The child cries, loses consciousness, develops marked pallor and goes stiff. Occasionally the child loses consciousness immediately after the bump without crying. A few clonic jerks may occur (reflex anoxic seizures). These pallid spells are caused by vagal asystole and can be induced by pressure on the eyeballs (oculocardiac reflex), ***although it is not necessary to elicit this reflex, and if thought to be important, should only be done under controlled conditions with EEG and ECG monitoring***.

Diagnosis

The diagnosis is made from a careful history of sequence of events. **These attacks are frequently confused with epilepsy**. In epilepsy the cyanosis occurs after the tonic-clonic phase of the seizure. In breath-holding spells they occur before but more importantly the diagnosis rests on the fact that the attacks are always precipitated by an appropriate stimulus. An EEG is not necessary except where the diagnosis is in doubt and epilepsy is suspected. **Always exclude anaemia which is a well documented cause of breath-holding episodes**. **Also exclude hypoxaemia which is also a cause**. ✔

Prognosis

These attacks are frightening for the parents but are harmless. They cease with time and do not have any long-term effects. There is no risk of subsequent epilepsy. Some infants with pallid type go on to develop faints in later childhood.

Management

The parents need to be given an explanation of these attacks and reassured about their harmless nature. There is no effective drug and no need for drug treatment. **Treat anaemia and hypoxaemia if present**.

3.43 Migraine

Allie Moosa

Minimum standards requirements

- Paracetamol and ibuprofen
- Antiemetics
- Propranolol, clonidine and pizotifen

Introduction

Migraine is a common cause of recurrent headaches in children. Its prevalence increases with age. It may be preceded by a history of recurrent abdominal pain and vomiting at a younger age (abdominal migraine, cyclical vomiting). The headache is typically throbbing in nature, temporal or frontal in location, more often bilateral than unilateral (compared to adults) and commonly associated with nausea, vomiting, pallor and sometimes photophobia. It usually lasts 1–3 hours but sometimes persists for 24 hours. It is relieved by sleep. It is precipitated by stress (school examinations, family pressure, undue expectations) and sometimes by hunger, fatigue, lack of sleep, exposure to sun and some foods (chocolates, Coca-cola, caffeine drinks, nuts, cheese). A positive family history of migraine, especially on the maternal side, is found in over 90% of patients and the diagnosis of migraine must be questioned in the absence of such a history. Between the attacks the child is well.

Classification of migraine

Migraine is classified into three types.

- **Common migraine**: There is no aura in this type. It is the commonest form in children accounting for over 80% of children with migraine.
- **Classical migraine**: An aura precedes the headache which is rare in children (about 10%). Visual aura include hemianopia (loss of half of visual field), scotoma (small areas of visual loss), fortification spectra (brilliant white zig-zag lines), blurred vision and flashes of lights. Occasionally sensory auras occur consisting of paraesthesia round the mouth and numbness of hands and feet.
- **Complicated migraine**: Rarely neurological signs occur during the headache and persist for varying periods after the headache. Ophthalmoplegic migraine (IIIrd cranial nerve palsy) is rare and must be distinguished from a berry aneurysm or other space occupying lesion compressing the IIIrd cranial nerve.

Hemiplegic migraine is the occurrence of hemisyndrome (weakness or numbness down one side of body) with the headache. Recurrent attacks of hemiplegic migraine are rare in children but occasionally, starting in infancy, a child may have alternating hemiplegia as a manifestation of migraine.

Basilar migraine results from vasoconstriction of the basilar and posterior cerebral arteries. Symptoms include vertigo, tinnitus, diplopia, blurred vision, ataxia and occipital headaches. There is complete recovery after the attack. Minor head trauma may precipitate basilar migraine.

Management

A careful history and examination is essential to confirm the diagnosis of migraine. Investigations are rarely needed. Explanation of the attacks and the relatively benign nature and good prognosis will reassure the parents and the child and by itself will lead to a reduction in frequency and severity of the headaches in over half of the children. Where possible, precipitating factors need to be identified and eliminated or reduced. In particular dietary factors such as chocolate, Coca-cola, caffeine drinks and cheese should be avoided.

Acute attack

Rest and sleep in a quiet, darkened room is usually preferred by patients. A simple analgesic alone or in combination with an non-steroidal anti-inflammatory agent is often all that is required and if given at the onset will abort or reduce the severity of the headaches. Paracetamol (dose 20 mg/kg repeatable every 6 hours as necessary) and ibuprofen (5 mg/kg/dose) are useful agents. If nausea and vomiting are troublesome, then an antiemetic may be prescribed: metoclopramide at 100 micrograms/kg/dose orally, or prochlorperazine: 200 micrograms/kg (max. 12.5 mg) orally IM or IV immediately then 100 micrograms/kg /dose 6–8 hourly, orally or rectally. **Prochlorperazine not licensed for children <10 kg**. Extrapyramidal side effects may occur.

Sumatriptan, a serotonin receptor agonist is highly effective for the acute attack but its use in children <12 years awaits further evaluation. Dose for adults is 50–100 mg orally as soon as possible after onset.

Prophylactic treatment

If the headaches are frequent (three to five per month) and troublesome then continuous prophylaxis is required usually for a period of one year. If headaches recur, the course of treatment is repeated. The drug of choice for children is propranolol but pizotifen and clonidine have all been tried in children with varying degrees of success.

- Propranolol is a beta-blocker and given in children above 7 years of age. (dose 7–12 years 10–20 mg 2–3 times daily orally, >12 years 20–40 mg 2–3 times daily) **It must not be given to children with asthma or diabetes, and may cause depression**.

✔

- Clonidine can be given at a dose of 2 micrograms/kg every 8 hours. Maximum dose 200 micrograms/day.
- Pizotifen given at a dose of initially 500 micrograms and building up to 1 mg at night or 500 micrograms 8 hourly. It may cause weight gain.

Prognosis of migraine

The prognosis is generally good. About half of children with migraine will undergo spontaneous prolonged remission after the age of 10 years. In most children the headaches are infrequent and rarely interfere with schooling or daily activities. In some the headaches are frequent and troublesome, these will require prophylactic treatment.

3.44 Neurosurgical disorders

Jonathan Punt

Minimum standards requirements

- Maternal folic acid before and during pregnancy
- Regional/national centre
- Dexamethasone and mannitol
- Intensive care
- Emergency burr holes
- Antibiotics
- Anticonvulsants
- Shunts

Introduction

Every country needs at least one hospital equipped to manage children with neurosurgical problems. The central and essential component of accurate assessment of the most frequently encountered intracranial neurosurgical emergencies is the prompt identification of the presence of raised intracranial pressure. Once this is recognised and controlled, the precise diagnosis of the site and cause can await ***sophisticated neuro-imaging by ultrasound scan, computed tomography (CT) or magnetic resonance imaging (MRI)***.

Raised intracranial pressure

The signs and symptoms are different for the pre-speech and younger infant than for the older child.

Babies and children < 2 years

- Abnormally rapid head growth
- Separation of cranial sutures
- Bulging of anterior fontanelle (note the anterior fontanelle usually closes by 18 months)
- Dilatation of scalp veins
- Irritability
- Vomiting
- Loss of truncal tone
- Fluctuating level of responsiveness
- Irregular rate and rhythm of breathing, usually with slowing of respiratory rate
- Irregular heart rate, usually with bradycardia but occasionally with tachycardia
- Decerebrate attacks

It is important to note that features may be non-specific, as in irritability and vomiting; that there may be marked fluctuations in the younger child's condition from minute to minute and from hour to hour; and that frank unconsciousness occurs relatively late, often being preceded by apnoea. Decerebrate attacks can be mistaken for epileptic seizures; in the former the child extends all four limbs and trunk, where in the latter flexion of the upper limbs is more usual and there are clear tonic-clonic phases.

Older children

- Headaches
- Vomiting
- Loss of postural control of the trunk
- **Failing vision**
- Diplopia
- **Neck pain and extension** ✔
- **Decerebrate attacks**
- Irregular rate and rhythm of breathing, usually with **slowing of respiratory rate**
- Irregular heart rate, usually with bradycardia but occasionally with tachycardia, and mounting hypertension with widening pulse pressure
- **Diminishing level of consciousness**

The most urgent features are failing vision, neck pain and extension, decerebrate attacks, diminishing level of consciousness and cardiorespiratory failure, as they all indicate incipient terminal events. Failing visual acuity is also urgent as it indicates severe papilloedema and a danger of permanent visual loss. The absence of papilloedema does not exclude raised intracranial pressure; its presence does indicate that there is a risk of permanent visual loss.

Accurate cerebral localisation on a clinical basis is difficult in children and virtually impossible in babies and young children, but the following can be fairly dependable features of a supratentorial mass lesion:

- Dysphasia
- Visual field defects
- Epileptic seizures

- Unilateral pupillary dilatation indicates a mass ipsilateral to the dilated pupil, or on the side of the pupil that dilated first in the case of bilateral pupillary dilatation.

Management

Whereas the definitive solution is removal of the causative lesion, this will often have to await the availability of ***imaging by computed tomography and transfer to a neurosurgical facility***. The emergency relief of raised intracranial pressure can be achieved by one or more of the following medical measures:

- Dexamethasone by slow intravenous injection (500 micrograms/kg immediately and then 100 micrograms/ kg every 6 hours)
- Mannitol 20% by intravenous infusion (250–500 mg/kg and repeat as required based on response and clinical signs (max. total dose 2 gm/kg) infusion over 20 minutes)
- ***Intubation and artificial ventilation to $PaCO_2$ of about 4 kPa***

In a dire emergency, and faced with a rapidly deteriorating child with no immediate prospects of evacuation for neuro-imaging and specialist neurosurgical care, then the following measures can be employed if there is no history of head injury.

Babies

Transfontanelle needle tapping of the subdural space, and if there is no subdural effusion, then the needle is advanced into the cerebral ventricle in the hope of finding and relieving hydrocephalus.

Infants and children

Right frontal burr-hole and ventricular drainage (see below).

If there is a history of head injury, then the procedure for "blind" burr-holes is followed (see below).

Head injuries (See Chapter 5.5)

Intracranial sepsis

Spontaneous extradural, subdural or intracerebral abscesses most commonly arise in children as a complication of an acute, or very occasionally chronic, episode of infection in the paranasal sinuses or middle ear. The cardinal clinical features are:

- Raised intracranial pressure
- Signs of focal neurological disturbance, including epileptic seizures
- Systemic signs of sepsis. These may be absent.

The diagnosis can be confirmed by ***CT scan with intravenous contrast enhancement***.

- Evacuation of pus is important to relieve raised intracranial pressure and mass effect, and to provide material for accurate microbiological diagnosis. Intracerebral abscesses can often be drained satisfactorily by burr-hole aspiration, which may have to be repeated. Extradural and subdural collections will usually require a major craniotomy.
- Raised intracranial pressure will usually be very severe and may require use of mannitol.
- Pending microbiological diagnosis, or in the absence of such support, the most useful antibiotics are a combination of cefuroxime (IV 60 mg/kg 6 hourly for 3 days, then 25 mg/kg 6 hourly) and metronidazole (7.5 mg/kg by intravenous infusion over 20 minutes every 8 hours) for a minimum of three weeks. A further six weeks of an appropriate oral antibiotic, such as amoxycillin, is usually necessary.
- Amoxycillin

Route:	Oral	
Dose:	1 month–12 years, 8 mg/kg	12–18 years, 500 mg
Frequency:	every 8 hours	

- An ENT surgeon may need to drain fronto-ethmoid sinuses or mastoids to prevent recurrence.

Hydrocephalus

Hydrocephalus can be diagnosed by transfontanelle ultrasound in those with an open anterior fontanelle, and by CT in older infants and children. CT will also demonstrate the likely cause.

The emergency relief of hydrocephalus, or suspected hydrocephalus, is by transfontanelle needle drainage in babies, or by burr-hole drainage and insertion of an external ventricular drain in older children. The best site for burr-hole drainage is on the right coronal suture in the midorbital line. The external guides are a triangulation comprising the root of the nose (medially) and the external auditory meatus (posteriorly) and the midorbital line. If the ventricles are enlarged, they will be encountered at a depth of no more than 6 cm from the scalp. The landmarks for transfontanelle puncture are the same as for subdural puncture (see above), but the needle is angled more steeply following the points for burr-hole drainage. CSF is allowed to drain spontaneously. Most babies will tolerate venting of up to 50 ml of CSF. Following withdrawal of the needle, the skin puncture is closed with a suture to prevent external leakage of CSF. It is important to have the CSF examined by a microbiology laboratory, remembering that subacute, partially treated, "neglected" pyogenic meningitis and tuberculous meningitis can present with hydrocephalus.

Definitive treatment may be by removal of the obstructing lesion in the case of a tumour or other mass; or by establishment of a permanent CSF diversion by way of insertion of an implanted ventriculo-peritoneal shunt or neuroendoscopic third ventriculostomy. Hydrocephalus shunts have a high incidence of malfunction: of those inserted in childhood, 40% require further surgery within two years of insertion, and 80% within twenty years. The shunt hardware is obscenely and unjustifiably expensive due to the

marketing processes involved. The more recently reintroduced alternative treatment of neuroendoscopic third ventriculostomy therefore holds considerable appeal, as when successful the patient is free of the tyranny of shunt complications.

The symptoms of shunt blockage are essentially those of raised intracranial pressure and require urgent attention if death or disability is to be avoided. The most reliable eye sign is loss of upward gaze. Blockages usually affect the ventricular end of the catheter rather than the peritoneal end. Revision should ideally be preceded by plain radiographic shunt series to exclude any fracture or disconnection, and ***CT or MRI to demonstrate ventricular size and to exclude any concomitant pathology***.

In the event of rapid deterioration precluding transfer to a fully equipped facility, the shunt reservoir, if present, can be tapped and CSF aspirated; this will not always be successful, as it is usually the ventricular catheter that is at fault. The choice then lies between insertion of an external ventricular drain (see above) or "blind" shunt revision. If the latter is employed, then exploration should commence at the cranial end.

If the shunt is found to be obviously infected then it should be removed and replaced by an external ventricular drain, pending appropriate antibiotic therapy and shunt reinsertion. Neuroendoscopic third ventriculostomy is an excellent alternative to shunt revision.

Shunt infections can present acutely with features of cerebral irritation, fever and seizures if there is major ventriculitis occurring within a few days of insertion; however this is relatively rare. More often shunt infections present with symptoms of malfunction as mentioned above. If plain radiographs do not show any obvious malposition, disconnection or fracture then infection should be suspected. Most shunt infections occur within a few months of insertion and so early shunt failure should raise concerns regarding infection. A few patients with chronic colonisation present with recurrent abdominal pains and low-grade pyrexia. In general, illnesses associated with high fever in patients with shunts are usually due to some other infection. In patients with myelomeningocele urinary infection must always be considered. If shunt infection is a possibility it is sensible to aspirate a small volume of CSF from the shunt reservoir, if present, prior to revision. It is important to remember that the common infecting organisms tend to adhere to the shunt tubing and may not be cultured from the CSF; **any tubing removed at revisional operations should therefore be sent for culture along with CSF samples**. ✔

The only method that is guaranteed to eliminate shunt infection is removal of all components, including any loose or retained fragments from earlier procedures, interval external drainage, appropriate antibiotics and shunt reinsertion through fresh incisions. As with all serious infections success is dependent upon accurate microbiological diagnosis. The most frequently encountered organisms are *Staphylococcus epidermidis* and *Staphylococcus aureus*. The most useful antibiotics are vancomycin by intravenous injection (loading dose is 15 mg/kg then 10 mg/kg every 6 hours) coupled with vancomycin by once daily intraventricular injection (5–20 mg) via the external ventricular drain. This can be supplemented by rifampicin orally (10–20 mg/kg once daily maximum 600 mg). The duration of treatment depends upon how rapidly the CSF becomes sterile but seven days minimum is recommended.

Myelomeningocele

Myelomeningocele is the commonest major congenital malformation compatible with survival. Incidence has been progressively falling for twenty years. Although there are regional variations the overall frequency is 0.7–0.8 per 1000 live births. The objective of management in the immediate postnatal period is the prevention of infection of the central nervous system. This is achieved by early closure of the lesion.

The level of the open lesion is noted and an assessment made of the sensorimotor level; the state of the sphincters; of any orthopaedic deformity; and the presence of major hydrocephalus, as evidenced by signs of raised intracranial pressure (see above). The prospects for independent ambulation as an older child or adult relate to the sensorimotor level: good extension at the knees is required, and so paralysis at or above L3–L4 is generally incompatible with independent ambulation without sophisticated appliances.

The ideal is to achieve closure within 24 hours of birth. **The majority of lesions have adequate skin in the wall of the sac as long as this is not unnecessarily sacrificed by a wide incision**. **The technique employed involves mobilisation of the neural placode, watertight dural repair and closure of the skin**. Whilst awaiting closure, the lesion should be protected with a dressing of moist sterile 0.9% saline, which is replaced every few hours to prevent desiccation. ✔

Most babies will require surgical treatment for hydrocephalus in the first few weeks of postnatal life. This will usually be ***by insertion of a ventriculoperitoneal shunt,*** although some babies are amenable to ***neuroendoscopic third ventriculostomy***.

The prospects for the child thereafter depend upon skilled multidisciplinary orthopaedic and urological care. Further neurosurgical interventions may be required to deal with complications of hydrocephalus, secondary tethering of the spinal cord and development of hydromyelia and hindbrain herniation (Chiari malformation). Spinal corrective surgery may be required in adolescence.

It is clear that the commitment is a lifelong one and this is a challenge to families and healthcare systems.

The aim should be to prevent as many as possible of these anomalies by adequate maternal nutrition prior to conception and during pregnancy. **FOLIC ACID TAKEN PRIOR TO CONCEPTION AND FOR THE FIRST TRIMESTER OF PREGNANCY ABOLISHES 75% OF CASES OF MYELOMENINGOCELE AND ANENCEPHALY**. (See Chapter 3.15.) ✔

Minimal requirements for paediatric neurosurgery

Staff

- ***Neurosurgeon with at least six months' training in paediatric neurosurgery gained in a centre that is nationally recognised as suitable for providing vocational higher surgical training in paediatric neurosurgery***.
- ***Paediatric anaesthetist***.
- ***Neuroradiologist with training in paediatric neuroradiology is preferable to general paediatric radiologist. The alternative is an image-link facility (Internet link) with a centre that has a paediatric neuroradiologist. Recent advances in image transfer make international consultation viable***.
- ***Neuropathologist. Again recent advances in image transfer make international consultation viable***.
- ***Microbiologist***.
- ***Resident medical staff in disciplines of neurosurgery and paediatrics. Children with neurosurgical conditions should neither be disadvantaged medically by being in a neurosurgical environment, nor surgically by being in a paediatric environment***.
- ***Paediatric trained nurses to care for children at all times***.

Plant and equipment

- ***Children should be nursed in a dedicated child's area of a neurosurgical ward, or in a ward in a children's department***.
- ***Access to a children's intensive care unit and to a neonatal intensive care unit***.
- ***Ultrasound scan and CT accessible and available at all times***.
- ***Operating room with appropriate anaesthetic equipment for children of all ages and sizes, and environmental control or other device to maintain body temperature***.
- ***Children-only operating lists***.

3.45 Orthopaedic problems other than injuries

Steve Mannion

Minimum standards requirements

- Antibiotics
- *X* rays
- Erythrocyte-sedimentation rate and C-reactive protein
- Antituberculous drugs
- Orthopaedic procedures

Injuries are by no means the only paediatric orthopaedic problems in the disadvantaged world. There is a great burden of orthopaedic infective conditions which, if treated suboptimally, can lead to considerable handicap. Furthermore, there is the same spectrum of non-infective conditions as is seen in rich countries which, due to the limited resources available in underdeveloped healthcare systems, represent a considerable diagnostic and therapeutic challenge.

Infections

Paediatric musculoskeletal infections are a common presentation in resource-poor countries. Morbidity and mortality can be prevented by prompt diagnosis, antibiotics and surgery where indicated. Infection should be suspected in any child presenting with pain or swelling in the limbs, spine or pelvis.

Pyomyositis

- Pus within muscle.
- Due to bacterial infection of muscle, almost always due to *Staphylococcus aureus*.
- Common in the tropics, exceedingly rare in the advantaged world.
- May be a history of previous injection/trauma to site.
- Signs: general malaise, swinging fever, decreased range of motion, fluctuant swelling, tenderness.

Treatment

- If diagnosed early (unusual) may respond to antibiotic therapy (flucloxacillin).
- Most cases will require incision and drainage of abscess under general anaesthesia.
- At operation:
  Incise along the long axis of the tender/swollen area
  Mark this area prior to anaesthesia ✔
  Drain all pus
  Irrigate thoroughly
  Insert wick to maintain drainage and prevent recurrence of abscess.
- Post-operatively:
  Analgesia
  5 days of antibiotics
  Change dressings daily
  Evaluate for signs of recurrence/other foci of infection.

Osteomyelitis

Infection within bone. Common in poor countries with several different manifestations:

- Acute haematogenous osteomyelitis
- Neonatal osteomyelitis
- Subacute haematogenous osteomyelitis
- Chronic osteomyelitis.

Acute haematogenous osteomyelitis

Pathogenesis

- Causes unknown
- Infection starts in metaphyseal venous sinusoids
- Vessels thrombose
- Pus develops in the medullary cavity leading to a build up of pressure
- Untreated, pus bursts through cortex and spreads under periosteum, rendering bone ischaemic (see **chronic** osteomyelitis below)
- In infants and children pathogen is almost always *S. aureus*, (for neonates see below); exception in sickle cell disease where *Salmonella paratyphi* is common.

Diagnosis

- Any child with **fever and unexplained bone pain** – high index of suspicion. ✔
  50% will have history of recent infection.
  Refuse to move affected limb.

Investigations

- In poor countries **clinical examination is the mainstay of diagnosis**. ✔

- White blood cell count – unreliable.
- Erythrocyte-sedimentation rate: raised in 90% cases.
- Blood culture positive in 40–50%.
- Plain *X* rays: bony changes take 7–14 days.
- Aspiration, Gram stain, look for acid fast bacilli.
- ***Bone scan, ultrasound – if available***.

Treatment
- Prior to the formation of pus in the medullary cavity antibiotics alone may suffice.
- Due to the predominance of *S. aureus* as the causative organism, initial antibiotic should be (flu)cloxacillin whilst culture results awaited (25 mg/kg IV or orally 6 hourly for 3 weeks).
- Once an abscess has formed this should be drained surgically.

Operative treatment
- Incision, drilling and drainage of osteomyelitic abscess.
- Mark area of maximal swelling/tenderness prior to anaesthesia.
- Longitudinal incision.
- Dissect onto and incise periosteum.
- Drill cortex of bone; if no pus at one site, further drill holes proximally and/or distally until pus obtained.
- Copious irrigation.
- Leave wound open, apply dry or antiseptic dressing.
- Monitor postoperatively for recurrence/other foci of infection, leave wound to granulate.

Neonatal osteomyelitis

There are several features unique to neonatal osteomyelitis:
- In the neonate metaphyseal vessels communicate with epiphyseal, thus permitting the spread of infection into the epiphysis and ultimately into the joint. Thus acute haematogenous osteomyelitis and septic arthritis may occur together. This can lead to complete lysis of areas such as the femoral head and neck and the proximal humerus, or premature physeal arrest.
- As the immune system of the neonate is immature there may be a less marked inflammatory response to infection with an absence of fever, raised white blood cells or erythrocyte-sedimentation rate.
- Multiple foci of infection are more common.
- A wider spectrum of infecting organisms is found; not only *S. aureus* but also group B streptococci and Gram-negative coliforms.
- Antibiotic treatment consists of gentamicin plus (flu) cloxacillin (see Chaper 3.48 for doses).

Subacute haematogenous osteomyelitis

This differs in presentation from that of acute haematogenous osteomyelitis as it:
- Often has an insidious onset
- Clinical signs are less marked
- Investigations may be inconclusive or equivocal
- Location is usually metaphyseal with plain *X* rays showing a solitary lytic lesion (abscess) with a sclerotic margin
- The differential diagnosis includes a neoplasm

The usual causative organism is, as for acute haematogenous osteomyelitis, *S. aureus*.

Treatment is surgical curettage of the lesion followed by antibiotic therapy.

Chronic osteomyelitis

If acute osteomyelitis goes untreated the pressure due to the intramedullary pus eventually increases until it bursts through the cortical bone into the subperiosteal space. If still undecompressed the pus spreads proximally and distally stripping the periosteum and thus rendering this cortical bone ischaemic (having been deprived of both intramedullary and periosteal blood supply).

The avascular cortical bone thus dies and becomes a focus of chronic infection called a "sequestrum". Simultaneously a periosteal reaction occurs under the stripped periosteum resulting in the laying down of new bone, "involucrum".

Appearance on plain *X* ray is characteristic; sclerotic sequestrum is separated (by the abscess cavity) from an irregular and enveloping involucrum.

Chronic osteomyelitis is difficult to treat even with optimal resources. Some guidelines as to its management are:
- If an osteomyelitic abscess is beginning to point, or there are signs of an underlying abscess, then this should be incised and drained
- In weight-bearing bones there should be no attempt at removal of sequestrum until the overlying involucrum is mature. This maintains the potential for weight bearing and ambulation
- Periods of immobilisation should be minimised in order to retain ranges of motion and function of nearby joints
- Sequestrum that begins to point through the skin can be removed/excised
- In many cases the clinical picture which results is one of intermittent flare ups of infection which can be treated by incision and drainage of abscesses, excision of sequestrae and antibiotic (flucloxacillin) suppression of infection as required
- Curative treatment is often elusive even in specialised centres and a degree of morbidity is unfortunately inevitable.

Septic arthritis

Septic arthritis is infection of a synovial joint.

Features
- More common in males than females
- Peak incidence around 2 years old
- Swollen, tender, warm joint with restricted range of motion
- Usually fever, 38–40 °C
- The patient is usually **systemically unwell**.

Diagnosis
- Mainstay of diagnosis is **clinical examination**.
- White blood cells raised in 30–60%.
- Elevation of the erythrocyte-sedimentation rate is more sensitive (except in the neonate).

- Plain *X* rays often normal until evidence of bone destruction at 7–14 days.
- ✔ **Aspiration of the joint is the definitive test**.
- Common pathogens include *S. aureus*, *H. influenzae*, group A and B streptococci, pneumococci, Gram-negative coliforms (in neonates).

Treatment
- Antibiotic therapy should not begin until after joint aspiration and blood cultures have been taken.
- Start with flucloxacillin (infants and children) or flucloxacillin and gentamicin (neonates).
- Some studies have shown that a combination of aspiration and antibiotic therapy is sufficient treatment but this must be followed by close monitoring to ensure improvement.
- If the child fails to improve then surgical washout and drainage is required either via open arthrotomy or ***(if available) by arthroscopic means***.

Postoperatively
- Continue antibiotic therapy, monitor for recurrence.
- Early mobilisation of affected joint to prevent stiffness.
- If treated early, prognosis for functional recovery is good. However, if presenting late there may already have been destruction of the articular surface.
- Beware coexisting osteomyelitis, present in around 15% of cases of septic arthritis.

Tuberculosis

Tuberculosis as an entity is covered in detail in Chapter 4.10, but it is important to remember the potential orthopaedic manifestations.
- It can cause both osteomyelitis and septic arthritis.
- In both cases the signs are less marked than in their non-mycobacterial forms, and the history usually more chronic.
- May be associated with systemic manifestations of tuberculous disease (respiratory, renal).
- Spinal tuberculosis (Pott's disease) can be the cause of both paraplegia and scoliotic deformity.
- Treatment consists of surgical drainage/curettage of abscess cavities combined with anti-tuberculous chemotherapy. For chronic disease and joint destruction spinal stabilisation/joint arthrodeses may be indicated.

Non-infective conditions

The non-infective paediatric orthopaedic conditions described below can be exceedingly difficult to treat in the disadvantaged world context. Firstly, without any form of population screening procedure in place or comprehensive primary healthcare provision, many cases will present late. Secondly advanced diagnostic modalities (ultrasound, arthrography) needed to direct treatment may not be available. Finally, where surgery is indicated, the operative techniques often need highly specialist training and/or ***specialised resources such as internal fixation and perioperative fluoroscopy***.

Thankfully, the conditions described are rare, typically occurring at a rate of less than 0.1%. They thus present far less commonly than the orthopaedic paediatric infections and cause a lesser burden of handicap overall.

Developmental dysplasia of the hip

Formerly known as "congenital dislocation of the hip", this complex condition has now been renamed "developmental dysplasia" in recognition of its variable characteristics such as the fact that it is not always present at birth nor does it always feature hip dislocation.
- Reported initial (neonatal) rates vary between 3 and 17 per 1000 live births but the rate of established dislocation is much lower; around 1 per 1000.
- Aetiology is multifactorial; increased rates are seen in female children, first borns, breech position, oligohydramnios and there is undoubtedly a genetic influence (increased rates with positive family history, affected siblings).
- Early detection depends upon neonatal screening, often not available in poor countries.
- If screening is to be carried out it should involve Barlow/Ortolani tests for newborns followed by subsequent re-examination/ultrasonography of suspected cases at 1 month of age.
- Plain *X* rays are of limited use before 6 months.

Treatment

As alluded to above, treatment of this condition in the disadvantaged world context is very difficult.
- Up to 6 months of age gentle closed reduction can be undertaken, then maintained in a Pavlik harness.
- If a Pavlik harness is unavailable then a plaster spica, maintaining the hips in flexion and abduction will achieve the same.
- In children over 6 months of age, closed reduction can still be attempted but is less and less likely to be successful due to the interposed joint capsule preventing stable, concentric reduction.
- If closed reduction fails, then open reduction can be attempted if surgical skills allow and infection avoided.
- Later presentation with proximal femoral/acetabular abnormalities may require ***complex secondary reconstructive procedures which are really within the realm of the specialist surgeon only***.

The reality of developmental dysplasia of the hip in the disadvantaged world is that cases will often not present until after the age of 18 months when the child has failed to walk or has an obviously abnormal gait. By this time bony changes may have occurred, the only treatment then being complex secondary procedures the skills and resources for which are usually unavailable in a disadvantaged world setting.

Congenital talipes equino-varus

There are three classes of talipes equino-varus (clubfoot):
- **Postural**: arises from intrauterine positioning and resolves fully with passive stretching within a few weeks of birth. Parents can be trained to do this.

- **Congenital**: arises in an otherwise normal child, varying degrees of severity; 1 in every 1000 live births, bilateral in 30–40%.
- **Syndromic**: associated with other syndromes such as arthrogryphosis, often severe and refractive to treatment.

Treatment

- The goal of talipes treatment is to obtain a functional, plantigrade, stable foot by the time the child begins to walk (i.e. before one year of age).
- If recognised in the neonatal period then gentle daily parental manipulation may be successful or alternatively manipulation and taping by qualified personnel (for example physiotherapist)
- For cases that fail to resolve in the first 6–12 weeks serial manipulation and plaster casting is indicated with cast changes every 2–4 weeks.
- If the deformity still fails to resolve, then there may be a place for limited percutaneous soft tissue releases (Achilles tendon or plantar fascia) at an age of 3–9 months. These techniques are relatively easily learned, have low morbidity and are user friendly in the disadvantaged world setting. They should be combined with manipulation and casting.
- For the case which still fails to resolve then more extensive surgery, such as a postero-medial release, is required. The timing of this surgery is usually between 6 months and one year of age. Although specialist training is required to learn this operation it can be relatively easily assimilated by the non-orthopaedic surgeon and, being only a soft tissue release, does not require any "high tech" surgical resources.
- Unfortunately, similarly to developmental dysplasia of the hip, the reality of this condition in poor countries is for the child to present late (over 18 months of age) when the deformity is fixed and secondary bony changes have occurred. Correction at this stage requires a combination of bony and soft tissue surgery which is really only in the realm of the orthopaedic specialist surgeon.
- In the adolescent child with fixed, chronic deformity the procedure of choice may be an arthrodesis (fusion) combined with correction of deformity performed at skeletal maturity.

Perthes disease (Legg–Calvé–Perthes disease)

Perthes disease is a disease of uncertain aetiology involving a process of fragmentation and repair of the femoral head possibly due to underlying idiopathic osteonecrosis.

- Usually occurs in a susceptible child between 4 and 8 years old, but can occur as young as 2 or as old as 12 years.
- Five times more common in boys, 10% bilateral, associated with hyperactivity.
- Presents with limping or waddling gait with groin, thigh or knee pain.
- *X* rays show varying degrees and stages of fragmentation and repair of the femoral head.
- Prognosis relates to the degree of fragmentation and the potential for repair and remodelling prior to epiphyseal closure. Good prognosis, therefore is associated with early onset and male sex (epiphyses close later).

Treatment

- In the majority of cases no specific treatment is indicated. The femoral head will repair and remodel satisfactorily and eventual outcome will be good. Bed rest, activity restriction and abduction braces have no proven impact on the natural history of the disease.
- In the small proportion of cases which may benefit from surgery the issue is **containment**. A very deformed femoral head may not seat or move properly in the acetabulum and thus lead to secondary arthrosis. A proportion of these cases may benefit from varus osteotomies of the proximal femur or pelvic osteotomies. ***Assessment for these procedures requires at the very least arthrography and the procedures themselves are very much the preserve of the orthopaedic specialist surgeon.***

Slipped upper femoral epiphysis

Slipped upper femoral epiphysis (SUFE, also known as "slipped capital femoral epiphysis", SCFE) is a disease in which the epiphysis becomes posteriorly displaced on the femoral neck.

- Prevalence 1–10 per 100 000, higher in black populations.
- Twice as common in boys as girls, at risk age for boys 10–17, girls 8–15 years. Most affected children obese, 40% bilateral hip involvement.
- Aetiology unknown; possibly endocrine.
- Onset may be abrupt or gradual. Sudden slips present with severe pain and inability to walk, chronic slips with pain often referred to the knees, a slight limp and limited internal rotation of the hip.
- Plain anteroposterior and lateral *X* rays are the most important diagnostic studies. Severity can be classified according to the degree of epiphyseal displacement. Greater than 30% displacement can result in premature osteoarthrosis.

Treatment

- The goal of treatment for SUFE is to stabilise the slippage and to promote premature fusion of the epiphysis if possible.
- Ideal treatment is fixation *in situ* with a single cannulated screw. Given the posterior position of slippage the point of entry of the screw needs to be anterior on the femoral neck. This procedure needs to be done ***under fluoroscopic or X ray control.***
- Where internal fixation or peroperative imaging is not available an alternative, would be spica cast immobilisation. However this is often logistically difficult and the physis may still be open even after cast removal.
- For the most severe degrees of slippage, in the hands of a specialist surgeon, reduction and fixation of the slip or femoral neck realignment osteotomies may be indicated.
- The commonest complications of operative treatment for SUFE are those of chondrolysis and osteonecrosis of the femoral head due to vascular compromise.

Genu varum/valgum

Varying degrees of knock- and bowed knees are common in the paediatric population. Most of these are merely variants of the normal physiologic knee-angle development appropriate to the child's age. Very few will require any form of intervention.

- Normal development: Babies are born with a varus knee angle which reduces with growth to become neutral at 18 months to 2 years of age. Thereafter the knee becomes increasingly valgus to a peak at 5–7 years, following which the angle gradually declines to the 5–9° of valgus seen in most adults.
- **Blount's disease** is a developmental condition affecting the proximal tibial physis and resulting in progressive varus deformity.
- Treatment of degrees of genu valgum/varum depends upon the age of the child and the severity of the condition. Bracing is of no proven benefit. Various corrective osteotomies are possible but these should be restricted to only those cases with functional handicap and are certainly not indicated merely on cosmetic grounds.

Scoliosis

Scoliosis is deformity of the spine characterised by lateral curvature and rotation.

- The commonest cause of paediatric scoliosis in the disadvantaged world is probably tuberculosis (**Pott's disease**). *X* ray appearances can be strongly suggestive of this diagnosis and then antituberculous chemotherapy commenced.
- The scoliotic deformity is descibed as idiopathic where there is no known aetiology. Contrary to popular belief most idiopathic scoliosis is only of cosmetic significance; only the most severe cases will have any degree of cardiorespiratory compromise.
- Scoliotic bracing is expensive, has compliance problems and is unlikely to be available in poor countries. If available it may have a role in slowing the progression of curves which are between 20° and 40°.
- Curves that are under 40° at the time of skeletal maturity are unlikely to progress further.

3.46 Developmental disorders and learning difficulties in children attending hospital

Prudence Hamadé, Mohammed Arzomand, David Cundall and Allie Moosa

Minimum standards requirements

- Good antenatal care including folic acid, iodinisation, iron, antimalarials, antitetanus immunisation
- Effective neonatal resuscitation
- Accident prevention
- Management of acute respiratory infection
- Management of diarrhoea and dehydration
- Multidisciplinary care
- Aids for disabled children

Children with special needs (disabled children)

Definitions

- "Special needs" is a term used to describe a child with an impairment that results in disability.
- "Impairment" the functional limitation within the individual caused by physical, mental or sensory problems.
- "Disability" the loss or limitation of opportunities to take part in the normal life of the community on an equal level with others that results from the impairment.

Children's rights

Article 23 of the UN Convention defines the right of disabled children to special care, education and training designed to help them achieve the greatest possible self-reliance and to lead a full and active life in society. It also encourages states to develop free and accessible services where possible and to share information with other countries regarding the latest outcomes of research into the management of disabling conditions.

The main care of children takes place in the community. Children with special needs and chronic illness and their parents are entitled to receive the same standard of care as any other family when their child is in need of acute care for any other condition described in this book. The attitude of healthcare professionals should reflect this important principle. Many cultures in poor countries have a greater degree of acceptance of disabled people. However, some cultures regard disability as a punishment or a reason for shame. The behaviour of hospital staff will go some way to dispelling these prejudices.

Planning of services

Ministries of Health and hospitals should consider establishing a register of disabled children but only after careful consideration of the aims of registration, the likely benefits and costs and the resources available.

Aims of a service for disabled children with special needs

- To provide health services which ensure that children reach their maximum potential optimising their independence and ability to lead a high-quality life.
- To promote active involvement of disabled children and their families in all aspects of healthcare, working in partnership with health professionals.
- To promote access to the hospital for families with disabled children.
- To provide comprehensive integrated and coordinated services.
- To enable health services to work with other key agencies, such as social services and education and training services.

Objectives

- To ensure that impairment and disability are promptly identified and treated where possible.
- To ensure services are developed to take into account ethnic, cultural, religious and linguistic needs of children and their families.
- To ensure that plans are developed in partnership with the child and family and that they are properly co-ordinated in partnership with other agencies both statutory and voluntary.

The prevention of impairment and disability in children

This is the main issue in poor countries where facilities to support such children are very limited. Of most importance are the quality of antenatal care, the quality of neonatal resuscitation (see Chapters 1.22 and 3.48), the prevention

of cerebral oedema due to inappropriate fluid management (see Chapters 3.25 and 3.36), the prevention of hypoxic ischaemic cerebral injury (see Chapters 1.20 and 3.6) and the avoidance of accidents and protection from abuse (see Chapters 5.1 and 5.2).

Antenatal and early infant care

- Every attempt should be made to provide good nutrition to women who are potentially to become pregnant. Folic acid at the time of conception is vital in preventing spina bifida and other congenital abnormalities.
- Doctors and nurses working in hospital maternity services should work closely with local leaders, women's groups, and the Ministry of Health to improve pregnancy outcomes.
- Iodinisation of salt is inexpensive and should be universal.
- Iron-deficiency anaemia during pregnancy is associated with low birthweight babies and should be screened for and prevented.
- Malaria in pregnant women is another cause of low birth weight and prematurity and should be prevented and if contracted be treated vigorously (see Chapter 4.27). Ministers of Health should be persuaded of the value of providing malaria prophylaxis for all pregnant women in endemic areas or in the most dangerous seasons.
- Immunisation against tetanus (see Chapters 1.29 and 4.9).
- Effective neonatal resuscitation should be available 24 hours a day in every maternity hospital and for all home deliveries. Staff must be trained and should have the basic equipment (see Chapters 1.22, 1.24 and 3.48) to prevent those causes of birth asphyxia which arise after the delivery of the baby.
- Breastfeeding must be supported (see Chapter 1.3) and special support given to help mothers provide breast milk to babies with developmental impairments which make sucking or attachment difficult.
- Obstetric care within hospitals should aim to prevent impairments due to complications of labour and delivery.
- Adequate training and facilities for the correct management of dehydration in gastroenteritis (Chapter 3.25), cerebral oedema, for example in malaria (Chapter 4.27), hypoxic ischaemic injury, for example in acute lower respiratory infections (Chapter 3.1) or severe anaemia from malaria (Chapter 4.27) all reduce preventable brain damage.
- Paediatricians in hospitals should advocate for programmes of accident prevention (see Chapter 5.1) and the prevention of injuries to children resulting from conflict, displacement or other social factors.

Management of disabled children with special needs

Identification and primary diagnosis

- A diagnosis that there is an impairment and if possible its cause should be established as soon as clinically possible and be communicated in a culturally sensitive manner to parents in accordance with locally developed guidelines.
- This communication must include information about the local availability of services and social support.

Comprehensive interdisciplinary assessment

- Assessment should always include the child's strengths as well as weaknesses and an assessment of home circumstances.
- This should result in decisions about management, including any immediate surgical or medical treatments available to alleviate the condition.
- Should include an assessment of sensory, motor, behavioural and intellectual capabilities as outlined below.
- Educational needs will also need to be assessed at appropriate ages.

Convening a team to plan long-term management

- The team will include those people whose skills and training are relevant to the needs of the child. The team is often led by a named paediatrician.
- Representatives from outside agencies such as education and social services must be included.
- A care manager or key worker should be appointed who will act as a liaison between professionals and parents to ensure that the child fully benefits from the available resources.

Development of local guidelines for clinicians

- Hospital staff should aim for an early diagnosis and identification of treatable causes of disability.
- Resources to support the child and family should be sought.
- In the absence of social support, hospitals must develop sensitive policies to inform parents of the diagnosis and expected prognosis in a way which is compatible with the best outcome for the child.
- Such policies should be decided by each hospital and all personnel should be informed of the policy.
- Culturally sensitive disclosure of information about diagnosis and expected prognosis should be given by a senior clinician with experience in this area and who is aware of local attitudes and beliefs regarding disability and the services available to the child and family.
- Services should be developed as resources allow.
- Policies with regard to the intensity of resuscitative treatment given to children with various impairments should be developed by doctors, other health professionals, representatives of the local community including disabled people and politicians. These policies must take into account ethnic and cultural issues and local support available for the care of severely disabled

children. Such policies must be reviewed frequently. (see Chapter 1.5).

- Development of services for and the rights of disabled people should be promoted wherever possible. Front line staff should feel confident that they know and can work within the framework of the policy.
- ***Metabolic disorders such as phenylketonurea, hypothyroidism, and disorders of glucose metabolism require long-term management and expensive treatment regimes. Lack of early treatment leads to severe mental handicap, which may in the long-term prove more expensive. Some families may not be able to cope with the time and expense required for adequate treatment***. **Ethical dilemmas of this sort should be part of the policy guidelines developed by hospital services at the highest level in collaboration with the local Ministry of Health and community leaders**.

Diagnosis

All newborn babies should be examined before leaving hospital by a member of staff (usually a nurse/midwife, or a paediatrician) if available who has been trained to carry out a competent neonatal examination. Any possible impairment must be reviewed by an experienced paediatrician.

The neonatal examination

- **General**: Signs of dysmorphism should be looked for. The baby should be examined for tone and observed to have normal limb movements. Disordered tone, feeding difficulties, irritability and seizures should be noted.
- **Hips**: The hips should be checked for dislocation remembering the three major risk factors of family history, female sex and breech presentation. Dislocated hips should be referred to an orthopaedic specialist.
- **Jaundice**: Any jaundice in the first 24 hours should be considered seriously and monitored appropriately. Causes of jaundice such as blood group incompatibilities diagnosed and treated. Severe jaundice can lead to mental retardation (see Chapter 3.48).
- **Cardiovascular system**: Should be examined (particularly oxygenation, pulse volumes and heart sounds) and if abnormalities are detected the baby should be referred to a paediatrician (see Chapter 3.5).
- **Hearing**: Behaviour should be noticed, although hearing defects are difficult to detect in the neonatal period without ***special equipment***.
- **Vision**: The child's eyes should be examined for infection, which must be treated with suitable medication. The absence of cataracts should be ascertained by the presence of a good red reflex in each eye (see Chapter 3.34).

Comprehensive assessment of children with impairments

Most children in disadvantaged countries are born at home and therefore, children with disabilities are more likely to present at the hospital in later life.

History (see Tables 3.46.1 and 3.46.2)

A complete paediatric history, including antenatal, perinatal, postnatal and family history should always be taken. Many countries have found that the "Ten Questions" are helpful in establishing the prevalence and distribution of various impairments:

1. Compared with other children, did he/she have any serious delay in sitting, standing or walking?
2. Compared with other children does he/she have difficulty seeing, either in the daytime or at night?
3. Does he/she appear to have difficulty hearing?
4. When you tell him/her to do something, does he/she seem to understand what you are saying?
5. Does he/she have difficulty in walking or moving his/her arms or does he/she have any weakness and/or stiffness in the arms or legs?
6. Does he/she sometimes have fits, become rigid or lose consciousness?
7. Does he/she learn things like other children his/her age?
8. Does he/she speak at all (can he/she make himself/herself understood in words. Can he/she say any recognisable words)?
9. For 3–9–year olds ask: is his/her speech in any way different from normal (not clear enough to be understood by people other than his/her immediate family)?

   For 2 year olds ask: can he/she name at least one object (for example an animal, a toy, a cup, a spoon)
10. Compared with other children of his/her age, does he/she appear in any way to have difficulties in learning?

Examination

A full clinical examination of all physical, sensory and psychological systems should be undertaken.

Additional issues

- Determine how the child and family have adapted their lives in response to the child's difficulties.
- Determine extent to which available treatment, training and management will improve the situation.
- Evaluate the emotional adjustment of the child and family to the disability.
- Investigate the educational facilities available to the child and how they may be adapted to his/her needs.
- Determine child's and family's strengths, abilities and positive personality traits which can be encouraged to help them cope with the disability.

Protocols for particular conditions

These should be developed to ensure that the child is thoroughly investigated initially and reviewed at regular intervals to ensure he/she can reach his/her maximum potential.

For example, a protocol for a child with Down's syndrome could include:

- Full medical examination
- ***Chromosome studies (if facilities are available)***
- ECG and chest *X* ray with ***echocardiography (if available)***

- Development of care plan with parents/carers as partners
- Audiological assessment
- Visual assessment
- Assessment by speech therapist (where available) to promote communication skills
- Assessment by the occupational therapist (where available) to determine any aids or equipment which may be of help
- Thyroid function test at appropriate intervals.

Sensory impairments

Liaison between health services and local education facilities is particularly important for the support and understanding of children with sensory impairments.

Visual Impairment

Evaluation

Newborn babies can follow a large object and focus on the mother's face. Impaired vision can therefore be detected soon after birth. It is normally the mother who will suspect this because the baby is not looking at her when she is breastfeeding. There may be roving eye movements.

- Use appropriate objects to confirm visual impairment, for example human face in neonates, toys in older infants and pictures (whose dimensions correspond to Snellen letters) in older children.
- Determine if visual impairment is an isolated problem or associated with other developmental defects (for example cerebral palsy, mental retardation) by detailed history and physical examination.
- Check for the red reflex: shine a light on the pupil from an arm's length: normally, it will appear red because of light reflected from the retina. If it appears white, then consider dense cataract, severe retinopathy of prematurity and retinoblastoma. If red reflex is normal, check the pupillary response to light: if normal it makes a local cause, i.e. optic nerve or retinal degeneration unlikely and is then most likely due to occipital lobe damage.
- Check the retina and optic nerve to exclude optic atrophy and retinal degeneration. If in doubt refer to an ophthalmologist.

Causes

Common causes of blindness are optic atrophy, congenital cataracts, and retinal degeneration and in poor countries, vitamin A deficiency and onchocerciasis.

Close liaison between paediatricians and ophthalmologists is required to develop policies to detect and treat visual defects as early as possible.

Management

- Treatable causes
  - Cataract: Congenital cataract should be referred to an ophthalmologist as soon as possible for early treatment. If no treatment is available, parents should be shown ways of stimulating residual vision by playing with bright lights and presenting visual stimuli to the child as much as possible.
  - Xerophthalmia: Treat with vitamin A (see Chapter 3.15).
  - Eye infections: (See Chapter 3.34).

Community health workers should have training sessions on eye care emphasising simple hygiene measures and sources of food rich in vitamin A to be found in local diets.

- Non-treatable causes
  - Visual assessment and provision of suitable visual aids.
  - Surgical correction of squints (when possible).
  - Mobility training for blind children and their carers.

The family will need support and advice about appropriate schooling, changes to the home and mobility training.

Hearing Impairment

Hearing loss is a hidden defect and may easily be missed if health workers are not vigilant. Because hearing defects often lead to the lack of development of speech and language, the child is sometimes assumed to be mentally retarded and may be further isolated from his or her family and society because of this. All children presenting with failure to develop language should have a good quality hearing assessment.

Hearing is essential for language development, therefore early detection of hearing impairment is essential. A newborn responds to sudden noise by the startle response. A normal baby will listen to the mother's voice. Formal hearing assessments in the newborn are possible ***using the acoustic cradle***. The distraction test is used at 4–8 months of age and audiometry in older children.

There are two types of hearing loss:

- Conductive: The commonest cause is recurrent/chronic infective otitis media (see Chapter 3.18).
- Sensorineural deafness: The commonest causes are meningitis, cerebral malaria, genetic defects and congenital infections. A hearing aid is needed and the child may have to be taught a sign language.

The following children are at risk:

- Family history of sensorineural hearing loss
- Dysmorphic features
- Abnormalities of the pinnae
- Severe birth asphyxia
- Severe neonatal jaundice
- Neurological abnormalities
- Postnatal infections such as meningitis or measles.

Most important is to identify and treat causes of conductive deafness (see Chapter 3.18).

- Treatable causes of sensorineural hearing impairment are very rare.
- Hospitals should seek to develop services for children with irreparable hearing problems such as simple audiological assessment and the provision of hearing aids.

Neurological problems

General advice

- Parents and carers should be given information and training so that they can modify daily activities to promote the development of the child and prevent the

development of contractures. Lifting, carrying, seating, playing and bathing will all need to be discussed and demonstrated.
- Physiotherapy should be commenced as early as possible to prevent the development of contractures in hypertonic children.
- Good positioning and movement are helped by appropriate aids and appliances (see Chapter 1.31).
- Local people are often resourceful in developing appropriate equipment for their own children out of locally available materials at cost. Advice from occupational and physiotherapists is very useful when available.
- Communication aids may also be required and the advice of speech and language therapists is very useful.
- Children with motor difficulties often have feeding difficulties and may not have the same access to food sources as children without impairments. Community health workers should receive training on feeding techniques in order to improve the nutritional state of these children as well as receiving instructions on how to show parents simple physiotherapies.
- Feeding may require the placement of and management of a nasogastric tube and parents should be shown how to undertake this.
- A care plan should be developed and a key worker appointed to monitor long-term plans to support the parents and keep them informed and involved in the long-term planning of services for their child.
- Aids to enable the child to have mobility, an effective means of communication and access to education should be developed in the community.
- All hospitals should seek to develop specialist therapy services to help such children.

Neural tube defects

(See section on paraparesis and incontinence below.)
- Should where possible be prevented by adequate maternal nutrition and vitamins at the time of conception.
- Children born with neural tube defects should be treated urgently to prevent worsening of their condition.(see Chapter 3.44).
- Parental wishes in terms of surgical treatment are very important.
- Later complications involve the urinary tract and bowel function. Poor blood flow to the lower limbs associated with a lack of sensation and mobility may result in pressure sores.
- Many children with spina bifida require alternative means of mobility.
- Spina bifida occulta may result in clumsiness and continence problems. Some of the associated problems may be improved by surgical intervention.

Delayed development (see Tables 3.46.1 and 3.46.2)

Delayed development presupposes knowledge of normal development. Development proceeds in an orderly fashion

Table 3.46.1 Normal milestones in development

Age	Milestone
Birth	Focuses with eyes and responds to sound
4–6 weeks	Social smile
6–7 months	Sits without support, transfers objects
9–10 months	Gets to sitting position, pulls to stand, pincer grasp, waves bye-bye
12 months	Stands, walks with one hand held, two or three words, stranger anxiety
15 months	Walks, drinks from cup
18 months	Walks upstairs, ten words, feeds with spoon
2 years	Runs, draws straight line, two word sentences
3 years	Draws circle, draws cross, dresses in simple clothes without assistance
4 years	Hops on one leg, fluent speech

Table 3.46.2 Warning signs in development

Age	Sign
10 weeks	Not smiling
3 months	Not responding to noises or voice, not focusing on face, not vocalising, not lifting up head when lying on stomach
6 months	Not interested in people, noises, toys, does not laugh or smile, squint, hand preference, primitive reflexes still present
9–12 months	Not sitting, not saying “baba”, “mama,” not imitating speech sounds, no pincer grasp
18 months	Not walking, no words, no eye contact, not naming familiar objects, not interested in animals, cars and other objects, passive – not moving about exploring, running, climbing; excessive periods of rocking and head banging
3 years	Unaware of surroundings, not imitating adult activities, little or no speech, long periods of repetitive behaviour, unable to follow simple command
4 years	Unintelligible speech
At any age	Parental concern, regression of acquired skills

but there is considerable variation in the age at which milestones are achieved.

Developmental assessment

The purpose of developmental assessment is threefold:

- To confirm normal or delayed development.
- To identify possible causes of delayed development.
- To plan strategy for intervention.

To achieve these aims, a detailed history and physical examination are essential. Particular emphasis is placed on perinatal and developmental history. Allowance must be made for prematurity. Microcephaly, dysmorphic features, signs of neglect must be excluded and a detailed neurological examination including primitive reflexes undertaken.

Answer the following questions:

- Does the child have global retardation (all areas of development delayed)? If this is the case the most likely disorder is diffuse damage to the central nervous system in disadvantaged countries and genetic disorders in rich countries.
- Is the delay confined to one area of development? If motor area this suggests a possible neuro-muscular disorder. Delayed speech development with normal motor and social skills could suggest hearing disorder.
- Has the child lost previously acquired skills and if so, is it progressive? This suggests a neurodegenerative disorder.

Delayed walking (not walking by 18 months)

- Family history of late walking and otherwise normal: give reassurance
- Global delay (especially in language/social skills): the child probably has mental impairment.
- Severe neglect.
- Cerebral palsy with upper motor neurone signs (spasticity, clonus, brisk reflexes) or dystonia, ataxia and involuntary movements.
- Neuromuscular disorders (see Chapters 3.39 and 3.41) with flaccid weakness, wasting or fasciculation of muscle, absent or diminished reflexes.
- Congenital dislocation of the hips and rickets can cause delayed walking.

Delayed language development

For meaningful speech to develop, the infant must be able to hear, have intact language pathways and normal oropharyngeal structures.

The following approach in evaluating a child with language delay is useful:

- Is there a hearing defect?
- What is the problem in language delay: is it in understanding or in expressing thoughts or both?
- Is the delay confined to language or is it part of global delay (consider severe learning difficulty)?
- Is there any dysfunction/defect of the mouth and pharynx (obvious on physical examination, comprehension will be normal)?
- Does the child have a problem with social interaction (consider autism: loss of social interaction, little or no non-verbal communication, no eye contact and repetitive ritualistic behaviour)?

Cerebral palsy (see Table 3.46.3)

Cerebral palsy refers to the disturbance of movement and/or posture that results from a non-progressive lesion of the developing brain. Commonest causes: hypoxic-ischaemic insult to the brain occurring prenatally, rarely postnatally (for example meningitis, head injury).

- Spastic diplegia (common with prematurity).
- Spastic quadraplegia and spastic hemiplegia.
- Choreoathetoid type (abnormal non-purposeful, writhing movements induced by voluntary activity).
- Ataxic type involves mainly the cerebellum and is rare.

- Diagnosis

The child normally presents with delayed development and is found to have abnormalities of tone, delay in motor development, abnormal posture or movements and persistence of primitive reflexes. The diagnosis is made on clinical grounds and investigations are not required.

- Evaluation

Assess the functional status of the child with regard to the motor system, (this is best performed by a physiotherapist) and identify associated problems.

Table 3.46.3 Associated problems in children with cerebral palsy

Problems	Action
Visual and hearing impairment	Refer to appropriate specialist
Seizure disorder	Anticonvulsants
Contractures	Physiotherapy and rarely surgery
Feeding difficulties, failure to thrive	Monitor intake Correct positioning for feeding Increase calories Consider gastro-oesophageal reflux
Constipation	Diet and stool softners
Learning difficulties	Additional help with schooling
Dislocation of hips	***Surgery to relieve pain***
Recurrent aspirations and respiratory infections	Antibiotics
Gastro-oesophageal reflux	Feed thickener (starch), if available H_2 receptor antagonist (for example ranitidine) ***and surgery***. May lead to oesophageal stricture and consequent malnutrition as well as inhalational pneumonia

- Management

The child with cerebral palsy has multiple problems and invariably will require care from a multidisciplinary team. The doctor and the physiotherapist play a prominent role. Physiotherapy advice enables parents to move and handle children in their daily activities to improve mobility and aim to prevent contractures. Parents need support in ensuring that the educational needs of the child are met and that the child is integrated as fully as possible into society.

- Deterioration in a child with cerebral palsy

Children with cerebral palsy usually remain stable. If a child shows apparent deterioration consider the following:

- Pain from dislocation of hips
- Dyspepsia from gastro-oesophageal reflux
- Non-convulsive status epilepticus
- Deterioration in mobility during growth spurt
- Wrong diagnosis – the child may have a progressive neurodegenerative disease.

Paraparesis and incontinence

Paraparesis, paralysis of both legs, is usually due to a spinal cord problem. This may be congenital, as in spina bifida or acquired, for example following trauma, infection or malignancy. Some causes are treatable if diagnosed early, for example TB of the spine or Burkitt's lymphoma. Thorough clinical assessment to establish the level of the lesion, and reassessment to look for changes, is essential. Any suspicion of a space-occupying lesion needs surgical advice.

Many children with paraparesis **will** suffer preventable complications **unless** carers and staff are aware of the risks of:

- **Poor nutrition**: many children with paraparesis find it difficult to eat and drink. They need good food to withstand infection, keep their muscles from wasting, prevent constipation and maintain good skin.
- **Contractures**: all joints need to be moved through their full range of movement to prevent contractures developing. If the child has presented late and contractures are already established, a programme of gradual passive stretching may help to improve the range of movement.
- **Pressure sores**: are prevented by ensuring that the child is moved regularly. The child can often learn to do this by using arms and upper body strength to pull on a suspended strap or ring to move their own position. The child can use a mirror to inspect their own skin to look for sore patches. Established pressure sores take a long time to heal, must be kept clean and free from pressure.

Rehabilitation should start immediately but will depend on whether the child's spine is stable. A creative approach to mobility, using locally available materials, (see *Disabled Village Children* by David Werner) is more likely to succeed than waiting for sophisticated rehabilitation equipment to be purchased.

Incontinence is usually associated with paraparesis, and can be both socially and medically disastrous. Some children have neuropathic bladders which are usually full and may lead to hydronephrosis and renal damage. These children need intermittent clean catheterisation to prevent back pressure and infection. Clean catheterisation may be required up to every 3–4 hours. This technique can be easily learnt by a carer or the older child. Other children have bladders which are not full and empty themselves frequently. These children are at less medical risk of kidney problems but it is much more difficult to enable them to be socially dry without complex surgery to enhance the size of the bladder.

Most children with bowel continence problems associated with paraparesis will be constipated due to relative immobility. A good diet and plenty of fluids will prevent constipation. Bowel evacuation in young children is often managed by abdominal massage. Older children can learn to use a Shandling catheter which is a plastic tube passed up the rectum for a washout of bowel contents with saline. ***Surgical approaches include using the appendix as a catheterisable stoma, or a caecostomy, for washing out the bowel.***

Learning difficulties and developmental delay

- Children who do not meet their expected developmental milestones should be assessed for possible causes.
- Some children have specific learning difficulties and may be assumed to have general learning difficulties unless they are carefully assessed. Full psychological assessment is helpful if available.
- Treatable causes, for example hypothyroidism, abuse/neglect, malnutrition, anaemia etc. should be ascertained. Problems such as autism and attention deficit disorder, with or without hyperactivity, should be documented.
- In planning services for these children, social and educational involvement is essential.

Severe learning difficulties

Severe learning difficulties (formerly called mental retardation) are suspected when there is global developmental delay especially in language, social and fine motor skills. Gross motor milestones may be normal. Some causes are fetal alcohol syndrome, hypoxic ischaemic injury to the brain, Down's syndrome, Fragile X syndrome and neurocutaneous syndrome. In most cases, severe learning difficulty is idiopathic. **Treatable causes should be excluded** (hypothyroidism and phenylketonuria).

The parents will need considerable support in coming to terms with the diagnosis and its implications. They may refuse to accept the diagnosis and should be taught and encouraged to stimulate the cognitive, language and motor development. Provide advice on appropriate play activities, suitable toys and reading material. Some children will be able to attend mainstream schooling but will need additional help, others will be better off in special schools. Their progress must be continuously monitored and associated problems dealt with. They deserve the same care as normal children.

Autism and communication disorders
- Usually presents in the second or third year of life.
- Is primarily a communication disorder associated with an absence of or deviant speech and language development.
- Is often associated with obsessional behaviours or interests.
- May or may not be associated with mental retardation.
- Often associated with learning difficulties because of inability to understand social situations.
- May result from abuse or neglect.

Treatment is largely educational and supportive.

Dyspraxia and dyslexia

Attention deficit disorder
A major problem associated with the following:
- Difficulty with concentration
- Impulsivity
- Difficulty in predicting the outcome of actions and therefore child does not learn from mistakes
- Strong association with hyperkinesis
- Poor social interaction because of difficulty with turn taking
- Poor listening skills
- Improves with maturity.

Treatment is difficult. The most important issue is to recognise the disorder, explain it to the parents and provide them with family/other support to cope with it. ***Sedatives, such as methyl-phenidate, may be very helpful***.

Behaviour disorders
- Exclude attention deficit disorders and other developmental impairments.
- Try to exclude abuse (see Chapter 5.2).

Psychiatric illness (see Chapter 3.13)
- These are rare in young children.
- Severe malnutrition, deprivation and abuse can lead to depression and the signs of frozen awareness/watchfulness (see Chapter 5.2).

Child-friendly and child-safe environments

- All buildings used as health facilities and playgrounds for children should be surveyed with the needs of their disabled users in mind.
- When new buildings are planned, it should be remembered that wheelchairs need wider doors and that where steps are used ramps should be provided.
- If the building has several floors lifts should when possible be in place. If this is not possible clinics serving people with disabilities should be on the ground floor.
- Areas used by visually impaired people should be well lit with steps and drops highlighted. Written notices should be as large and clear as possible.
- Special facilities may need to be provided for deaf and blind children to access information.

Transition to adult life for children with disabilities

A human rights perspective

Transition from childhood to adulthood takes time and the process of adolescence is experienced and managed in very different ways in different cultures. This transition is much more challenging for disabled children whose abilities to achieve independence may be constrained by their condition. Disabled children are more at risk of abuse and exploitation and are likely to be more vulnerable as they go through adolescence into adulthood. It is best to view this transition from a **human rights perspective**. Thus the disabled child has rights as stated within the UN Declaration of the Rights of the Child and the same perspective informs any consideration of the transition to adult life.

Conditions, cultures and economies

Different cultures and economies make it relatively easy or difficult for young people with different types of conditions to integrate and find a role. For example, a young person with a severe physical disability, such as spastic quadriplegia but of normal intelligence may find it relatively easy to find a fulfilling role as an adult in a technologically advanced urban environment where there are relatively few physical barriers for wheelchair access. However, a young person with learning difficulties and good mobility may find it difficult to find a fulfilling role in such a society. By contrast, a less technologically advanced society can be much more accepting of the young person with learning difficulties for whom there are many welcome roles in the rural economy, and the intellectually competent but physically impaired young person may find it much more difficult to find fulfilment in such an environment.

There may be very different cultural expectations for young men and young women and deep-seated prejudices and cultural taboos which cause further disablement and devaluing of young disabled people unless the human rights perspective is paramount.

The challenges of transition

Independence
Good practice includes involving children in decision-making about their own lives well before they enter adolescence. Learning from failures as well as successes is part of normal development. Many children in economically poor communities are expected to work on the land or in industry, look after livestock or take responsibility for child care of their younger siblings at ages when they are not developmentally equipped to do so. Many children who have been involved in civil war and other armed conflict have been deprived of an ordinary childhood and may have had "independence" forced upon them at an early age (see Chapter 3.14). Disabled children may have similar experiences or worse, for example being used as beggars which deprives them of their human right to a childhood. If "independence" means the insecurity of street children

progressing to prostitution or a life of petty crime then this is not the sort of independence that young people need.

At the other extreme, disabled children worldwide are often overprotected by their families. Their families may feel ashamed or there may be cultural taboos and beliefs about the origins of particular conditions. Parents may wish to do everything for their disabled child but this can result in the child not learning from experience. The end result may be that disabled young people do not get the opportunities for education and training which will enhance their self-esteem and ability to at least make some contribution to society rather than be seen merely as an object of pity and charity.

To enable the disabled child to become an integrated member of adult society is a challenge which requires:

- Imagination and flexibility on the part of the health, education and social services
- Active engagement with the young person, his or her family and community
- A real commitment to working with the strengths of the young person and minimising his/her weaknesses by reducing the barriers to his/her participation in society
- Anticipating difficulties in advance and balancing the risk of failure against the benefits of increasing independence.

Information

Disabled young people often do not have access to information about their own condition, necessary health education to prevent secondary problems developing, training and employment opportunities, self-help groups and their rights.

Sexuality

The challenge of emerging sexuality is often more difficult for the disabled young person. Young people often have inaccurate information about the basic facts of sexual development and disabled young people often miss out on the opportunity to learn these facts in a straightforward way. Many young people may be unaware of any genetic implications of their own condition although it is more common to assume that there are genetic risks to their offspring when this is **not** the case.

Families, and indeed some health professionals, may make inaccurate assumptions about the ability of disabled young people to have normal sexual experiences. These young people may have their own inaccurate beliefs which may cause much unnecessary suffering unless they have the opportunity to understand the facts about their own bodies. Even when there are some physical problems which will affect sexual experience, for example lack of genital sensation for some young people with paraplegia, this does not preclude an active and fulfilling sexual relationship.

Services for the transition to adult life

Health facilities providing services for children with disabilities should develop expertise in enabling children to make the transition to adult life. This expertise is likely to be achieved by developing shared knowledge amongst a group of relevant professionals working in partnership with young people. The service should be able to offer:

- Information which is relevant and up to date
- Individual counselling
- Opportunities to meet other young people with similar difficulties
- Careers advice
- A service to loan out equipment to increase independence
- Close links with education facilities and any social and housing services.

Further reading

Werner, D. *Disabled Village Children*. A guide for community health workers, rehabilitation workers and families. USA. The Hesperian Foundation 1999.

3.47 Skin diseases

Rod J Hay

Minimum standards requirements

- Antiscabies treatment
- Antibacterial treatment
- Antifungal treatment
- Topical steroids
- Emollients
- Antiviral treatment

In poor countries, skin disease is dominated by bacterial infections such as impetigo and parasitic conditions including scabies and pediculosis. It is often poorly managed and may present a real economic cost to families through use of ineffective remedies. It is important to recognise whether cases reflect individual or community problems; treatment of single cases of scabies makes little impact if there is widespread infection in the community.

Scabies

- Scabies is a parasitic infection caused by the mite, *Sarcoptes scabiei*, which spreads from human to human, usually by direct contact.
- The adult female burrows a tunnel into the stratum corneum or outer skin layer producing eggs which hatch into larvae in 3–4 days.
- "Outbreaks" in communities may follow a cyclical pattern with peaks of incidence occurring every 4–7 years.
- Infection in adults usually reflects overcrowding in households and transmission through contact with infected individuals including infants.

Clinical presentation

Main sites for infection include fingers, wrists, elbows, external genitalia, ankles, buttocks; the face and head may be affected in babies but these sites are seldom involved in older children.

Important clues are:

- Itching in several members of same household
- Lesions in characteristic sites particularly lateral borders of fingers
- Papules, pustules and sinuous tracks or burrows (5–10 mm).

In onchocerciasis itching is also common but lesions are seldom found on fingers.

Diagnosis

Remove mites from their burrows with a sterile needle and examine under low power of the microscope.

Complications

- Secondary bacterial (streptococcal) infection is common (see below).
- In severely immunocompromised individuals (for example AIDS) a crusted form of scabies, without severe itching but with large numbers of mites, may occur.

Table 3.47.1 Topical treatment of scabies

Antiscabetic	Treatment	Side-effects
Sulphur	Given as a 5–10% application in white soft paraffin or as soap. Treat for 1–2 weeks	Local irritation
25% Benzyl benzoate emulsion	One application followed by another 2–3 days later	Local itching, eczema
5% Permethrin cream	One application (another is often necessary)	Minimal itching
0.5% Malathion lotion	One or two applications	Itching
1% Gamma-benzene hexachloride lotion	One to four applications	Caution in children Seizures have been recorded
1% Crotamiton cream	1–2 weeks of treatment. Not very effective although can reduce itching	

Treatment

- Cheapest options are sulphur based. However they are slow and require daily applications for 7–14 days.
- Permethrin is the most rapidly active but also the most expensive.
- All potentially affected areas are treated including soles of feet and, in babies, the scalp.
- ✔ **Failure of antiscabetics often occurs because there is no place where individuals can apply treatments in privacy**.
- Treat all members of household, including those without itching.
- Clean or change clothes after first treatment.
- Resistance to gamma benzene hexachloride occurs.
- Ivermectin (oral) is highly effective for crusted scabies but is not suitable for young children (single dose of 150 micrograms/kg). (No food to be taken for 2 hours before or after dose.)
- Community-based treatments, although ideal, are seldom practised as they are difficult and, although individually cheap, comparatively costly to apply to large numbers.

Impetigo

- The term, "pyoderma", is used to describe a range of superficial bacterial infections which includes impetigo, folliculitis, abscesses (furunculosis) or secondary bacterial infection, for example of scabies.
- Impetigo is a form of pyogenic infection, which involves the epidermis, caused by Group A streptococci or *Staphylococcus aureus*. It is not possible to separate the two infections clinically.
- Ecthyma occurs where impetigo penetrates deeper to affect the dermis with ulceration.

Clinical presentation

- Impetigo presents with oozing and yellowish crusted plaques, often on exposed sites such as the face.
- May be multiple, and form blisters – in which case *Staph. aureus* is the usual cause.
- May be transmitted to other parts of the body and to other children.
- Secondary infection of scabies may occur – papules become pustular and there may be surrounding impetiginised crusts on scabetic burrows.
- Boils (furuncules) are also common and are always caused by *Staph. aureus*. Lesions are large tender fluctuant masses with surrounding inflammation. May occur in other members of same household.

Complications

A serious complication of streptococcal impetigo or pyoderma is glomerulonephritis which follows infection by nephritogenic strains. In tropical environments poststreptococcal glomerulonephritis more often follows skin rather than throat infection.

Management

- Impetigo is transmissible and treatment should include other contacts with lesions.
- Should cover both *Staph. aureus* and streptococci – unless laboratory facilities for culture are available.
- A topical agent may be used but for widespread lesions oral treatment is usual (see Table 3.47.2). Choice is influenced by cost, extent of disease and type of lesions.
- Most *Staph. aureus* strains, even in remote communities, are resistant to both penicillin and tetracycline.
- Most topical azole antifungals (for example clotrimazole, miconazole), apart from ketoconazole, have activity against Gram-positive bacteria.
- Boils are best managed by incision and drainage.

Tropical ulcer (tropical phagadenic ulcer)

- Tropical ulcer mainly occurs in children and teenagers but is seldom seen in the rich countries.
- It is patchily distributed in endemic foci throughout Africa, India and the West Pacific.
- Associated with humid areas or areas subject to local flooding.
- Considered to result from synergistic bacterial infection of which one anaerobic organism is usually *Fusobacterium ulcerans*. Other bacteria present in lesions include spiral bacteria and Gram-negative bacteria. *F. ulcerans* has also been isolated from mud or stagnant water in vicinity of cases.
- Initial lesion is a soft papule with surrounding hyperpigmentation overlying an area of skin necrosis. This

Table 3.47.2 Treatment of impetigo

Agent	Route	Use	Cost
Cloxacillin, flucloxacillin	Oral: 12.5–25 mg/kg four times a day	For widespread and severe impetigo. Rapid – clearance in 3–5 days	Expensive
Mupirocin	Topical	For localised infections. Rapid – 3–7 days	Moderate
Fucidin	Topical	As for mupirocin	Moderate
Clioquinol	Topical	Slow 7–14 days, may stain skin. Irritant	Cheap
Potassium permanganate (alternatives chlorhexidine, povidone iodine)	Topical	Simple to use, stains skin. Slow 7–14 days	Cheap

develops over at least one week and when the overlying skin sloughs a regular and deep ulcer, 3–10 cm in diameter, is revealed.

Complications

With proper care and regular irrigation or cleansing of lesions the area will heal. About 5–10% may progress to chronic ulceration and, in some cases, secondary squamous carcinoma or more serious infection, for example underlying osteomyelitis may develop.

Management

Objective of treatment is to allow rapid healing without secondary infection.

Regimen:

- Dilute antiseptic, for example potassium permanganate solution, or 0.9% saline for cleansing ulcer and surrounding skin
- Daily dressings.

A single IM dose of benzyl penicillin (50 mg/kg) or oral metronidazole (7.5 mg/kg every 8 hours). The former is particularly important in areas where yaws is also endemic as it will cover both conditions. If healing is delayed local pinch grafting may be necessary.

Cutaneous leishmaniasis
(see section on leishmaniasis, Chapter 4.26)

Superficial fungal infections

- Common childhood fungal infections are scalp ringworm or tinea capitis and oropharyngeal candidosis (see Chapter 4.18).
- Tinea infections are caused by dermatophyte fungi, which are adapted to survive on the outer layer of the skin, the stratum corneum, or structures such as hair or nails derived from it. Dermatophyte infections are caused by one of three genera of fungi, *Trichophyton*, *Microsporum* and *Epidermophyton*, which are acquired by spread from soil, animal or human sources, geo-, zoo-, or anthropophilic infections, respectively. By convention they are known by the term "tinea" followed by appropriate Latin word for site affected for example tinea pedis (feet), tinea corporis (body) or tinea capitis (scalp).
- Tinea capitis is often endemic in rural or urban areas of disadvantaged countries and inner-city areas of industrialised countries. Prevalence rates may reach over 20% in some communities.
- Main signs of infections are:
  - Scaling
  - Hair loss – diffuse or in localised patches. Scalp hairs in affected areas may break at scalp level or a few millimetres above skin
  - Itching – variable
- Key clue to diagnosis is presence of broken hairs. Confirmation is by culture of scrapings taken from scalp surface with a sterilised scalpel or sterile scalp brushes. Presence of infection can also be verified by microscopy of hair samples.

Complications

- Kerion is a severe pustular reaction on the scalp, which accompanies a strong immune response to ringworm infection.
- Favus is a widespread crusting form.
- Secondary infection with bacteria may occur, usually where there are crusts overlying surface of inflamed lesions.

Management

- Culture of fungus can distinguish whether infection is from a human or an animal source, viz. zoophilic, (*Microsporum canis* – cats and dogs, *Trichophyton verrucosum* – cattle) and the anthropophilic species (*T. violaceum, T. tonsurans, T. soudanense, M. audouinii*).
- Presence of infections in close contacts, for example schoolmates or family may signal child to child spread and alert schools to other infected children. In disadvantaged countries mass treatments have a low priority because of health resources and because cases usually self-heal.
- Children with severe symptoms, for example kerions, favus or widespread hair loss, should be treated.
- Whitfield's ointment or imidazole antifungals, for example clotrimazole, are generally ineffective in scalp ringworm.
- Treatment of choice is griseofulvin, which is available in oral tablet or solution form, 10 mg/kg once daily (after food) and up to 20 mg/kg in refractory infections. Single-dose treatments with 1.0 g immediate dose, sometimes repeated after one month, have been successful for mass treatments of infected classes in school. If possible a topical treatment such as an imidazole cream (clotrimazole) two or three times daily, ketoconazole shampoo or selenium sulphide shampoo should be given to prevent spread to others. Occasionally kerions may require topical or oral steroids but these are not part of their initial treatment.

Eczema (atopic dermatitis)

- Eczema is a specific inflammatory disease involving epidermis and dermis.
- In childhood the commonest form of eczema is atopic dermatitis. Atopic dermatitis is uncommon in rural areas of disadvantaged countries and appears to be associated with urban environments and increased affluence.

Clinical presentation

- Severe itching and a scaling rash affecting the skin flexures, for example elbows, behind knees, neck.

- Scratching may be very severe and sufficient to disturb sleep.

Management principles

- Moisturise skin with emollients. Thicker more greasy preparations such as white soft paraffin or a 50:50 mixture of white soft paraffin and liquid paraffin are preferred to creams as they provide longer-lasting effects.
- Treat inflammatory lesions with topical corticosteroids (once or twice daily). Weaker strength preparations (1% hydrocortisone) are best although it may be necessary to use medium to strong topical steroids in some cases (never use the latter on the face). Use corticosteroids intermittently relying for long-term management on emollients.
- Treat complications. These are secondary bacterial infections, usually *Staph. aureus* and acute *herpes simplex* (eczema herpeticum): aciclovir cream five times daily for 5–10 days (until healed) or aciclovir orally if severe 20 mg/kg four times daily for 5–7 days and contact dermatitis which may include allergy to topical medicaments such as lanolin and corticosteroids. An oral antibiotic (for example cloxacillin or flucloxacillin 12.5–25 mg/kg four times a day) in acute flare up of eczema may produce a good response.

Atopic eczema ranges from a mild skin rash to a severe condition which can dominate family life and may cause major family stress. Food allergy is a rare cause and skin testing for precipitating factors is usually not helpful. In industrialised countries there are patient organisations (for example National Eczema Society, UK) which provide support and advice to patients and families.

Hypopigmentation and hyperpigmentation disorders

Often secondary to other inflammatory processes which should be treated. There are no effective, cheap or easily administered treatments for the pigmentary changes themselves. The common fungal disease, pityriasis versicolor, may present with hypopigmented patches on the trunk which coalesce; however these are scaly. Treatment with topical antifungal azole creams (for example clotrimazole or miconazole) are effective.

3.48 Neonatal medicine

Anthony Williams, Silvia Patrizi, James I Hagadorn and Nicholas Guerina

Preterm labour

Labour commencing before 37 completed weeks (259 days) from the first day of the last menstrual period (if known) is considered "preterm", placing the baby at increased risk of complications (see below). In the absence of special facilities mortality increases substantially under 32 weeks of gestation and survival at under 28 weeks is unlikely. When preterm delivery is anticipated, realistic expectations should be discussed with the parents and any limitations on resuscitative efforts should be agreed upon. Paediatric outcome at gestations less than 32 weeks can be improved by attention to the following points before delivery:

- Administration of antenatal corticosteroids to the mother (dexamethasone 6 mg twice daily for four doses).
- Tocolytic therapy to defer delivery, "buying time" for steroids to take effect.
- Treating any ascending infection with antibiotics. If infection is likely, delivery of the baby should be expedited. The most suitable antibiotic for the mother is a combination of a beta-lactam and aminoglycoside (for example ampicillin and gentamicin), or equivalent broad-spectrum coverage in standard adult doses. Intrapartum risk factors for fetal/newborn infection at all gestational ages include rupture of membranes for 18 hours or more, maternal fever, and chorioamnionitis; the incidence of infection and adverse outcome is further increased if there is preterm labour in addition to any of these risks.

Delivery

Preparation for delivery

- Equipment listed in box must be present
- The delivery room should be warm (22°–25°C).
- A trained resuscitator needs to be present at all deliveries (ideally following a Neonatal Life Support course – see Chapter 1.22, 1.24 and Appendix 7.11).

At delivery

- Once delivered the baby should be held below the mother until the cord stops pulsating.
- If the baby is active and establishes breathing he or she should be placed on the mother's chest, dried and covered with a warm, dry towel or blanket to prevent hypothermia.
- If the baby does not establish breathing, commence resuscitation.

Essential equipment needed for all deliveries

- Warm surface and towels to dry and wrap the baby
- Disposable gloves
- A suction device connected to a suction catheter (8 or 10FG) manual and (ideally) electrically powered unit
- Face masks of suitable size and a pressure-limited ($<$30–40 cm H_2O) inflation device
- An oxygen source
- Nasogastric tubes, sizes 6 and 8FG
- Sterile umbilical cord clamps
- Adhesive tape (for example *Micropore*)
- Sterile scissors
- Stethoscope
- Clock
- Namebands

For the use of attendants trained in neonatal resuscitation (to be immediately available for all deliveries)

- Two neonatal laryngoscopes (with spare bulbs)
- Endotracheal tubes, sizes 2.5, 3.0 and 3.5, with adapters to fit the inflation device
- Umbilical venous catheter (or use sterile feeding tube)
- 1 ml and 5 ml syringes
- Emergency drugs:
  - 1 in 10 000 epinephrine
  - 4.2% sodium bicarbonate
  - Vitamin K for injection (1 mg)
  - 0.9% saline for infusion
  - 10% glucose for infusion
  - Naloxone where opiates used in labour

Resuscitation (see Chapter 1.22)

This section contains a brief outline of newborn resuscitation and should not be used as a substitute for formal training (see Self-instructional Educational Program in Neonatal Resuscitation and Perinatal Care, Chapter 1.24).

- **The following signs indicate that a baby needs resuscitation**: apnoea or gasping respirations, hypotonia, pallor or cyanosis, or bradycardia (heart rate under 100 beats/minute). Heart rate may be assessed by gently squeezing the base of the umbilical cord between finger and thumb to determine umbilical artery pulsations, but as pulsations are not always felt, cardiac auscultation is better. The infant's status may be more formally assessed by the Apgar score (see Chapter 1.22), but this procedure should not be used to direct resuscitation efforts or delay resuscitation.
- Dry the baby and place on a flat, dry surface under an overhead heater (if available).
- Using low-pressure suction, clear secretions from the nose and pharynx.
- Apply a facemask of appropriate size over the mouth and nose and ventilate with air or oxygen: a self-inflating bag or flow device with thumb occlusion can be used – whatever is available there **must be a blow-off valve or simple water manometer which prevents pressure exceeding 30–40 cm H_2O**. Breaths should have an inspiratory time of 2–3 seconds (see Appendix 7.10).
- Watch for chest wall movements, auscultate to ensure breath sounds are symmetrical and count the heart rate – is it increasing?
- If the pulse rate does not rapidly respond to mask ventilation, check the chest is moving. If not the baby will require **endotracheal intubation** by somebody trained to undertake this procedure.
- **If the heart rate is under 60 beats/minute cardiac massage should be given**: place two fingers on the sternum between the nipples and depress about 100 times per minute to 1/3 of the anteroposterior diameter of the chest.
- **If the heart rate remains below 60 beats/minute** once adequate ventilation via an endotracheal tube has been established, give 0.1–0.3 ml/kg of 1 in 10 000 epinephrine down the endotracheal tube.
- Obtain venous access – the quickest way is to pass an umbilical venous catheter (UVC).
- Repeat epinephrine a second time through the UVC, giving 0.1–0.3 ml/kg of 1 in 10 000 solution.
- Sodium bicarbonate (4.2%) 4 ml/kg and volume support (10 ml/kg of 0.9% sodium chloride) may be given through the UVC.
- Epinephrine, glucose and sodium bicarbonate can be given intracardiac (see Chapter 6.16).
- Epinephrine BUT NO OTHER DRUGS can be given into the trachea.
- ✔ **If no pulse rate is present after 15 minutes and at least two or more doses of epinephrine have been given the senior person present may decide whether to abandon further resuscitation efforts**.

Documentation

The following data should be recorded in the notes of every newborn baby.

Mother's data

- Name, address, date of birth, and any identifying number
- Parity and previous obstetric history
- Height
- Blood group
- First day of last menstrual period
- ***Results of any antenatal serology (for example rubella, syphilis, Rh titres, HIV status)***
- Illness during the pregnancy
- Drugs taken during the pregnancy
- Family history of any illnesses

Father's data

- Full name, address and date of birth
- Family history of any illnesses

Labour and delivery data

- Time of onset: whether induction of labour or spontaneous
- Time membranes ruptured and any other known risk factors for infection (see below)
- Duration of first and second stage of labour
- Drugs given to the mother in labour
- Presentation and mode of delivery
- Apgar score (see below)
- Full details of any resuscitation
- Time, dose, route of administration and full generic name of any drugs given to the mother
- ***Cord pH (if measured)***

Baby data

- Temperature shortly after delivery to document adequate thermoregulation
- Birthweight
- Head circumference (best measured after 24 hours when moulding subsided)
- Length (ideally)
- Full physical examination, noting any abnormalities or evidence of birth trauma detected
- Details of dose, preparation and route of administration of any drugs given at delivery (for example vitamin K)
- **If not already given, ensure vitamin K 1 mg intramuscularly** ✔

Terminology

- **A low birthweight baby** is one weighing <2.5 kg at birth. Low birthweight may be attributable to preterm delivery or intrauterine growth retardation (see below).
- **A preterm baby** is one born before 37 completed weeks have elapsed since the first day of the last menstrual period (259 days). All but about 1% of preterm babies are born after 32 weeks of gestation.

- **A small-for-gestational age (SGA) baby** is one whose birth weight falls below the 10th percentile on a birthweight chart. Note: probably at least a quarter of SGA babies have not suffered intrauterine growth retardation but are just constitutionally small by virtue of maternal size. The mean birthweight of babies born to mothers 4 feet 10 inch tall is about 500 g less than that of babies born to mothers 6 foot 0 inch tall. This discrepancy increases to about 1 kg if extremes of mid-pregnancy weight are also taken into account.
- **Intrauterine growth retardation (IUGR)** also known as **intrauterine growth constraint** is a term which refers to slowing of fetal growth velocity. It is possible for a large fetus to suffer IUGR and still have a birthweight >10th percentile.
- A large for gestational age (LGA) baby >90th percentile on a birth weight chart.

For most clinical purposes it is sufficient to classify babies as "low birthweight", "preterm" or "small-for-gestational age".

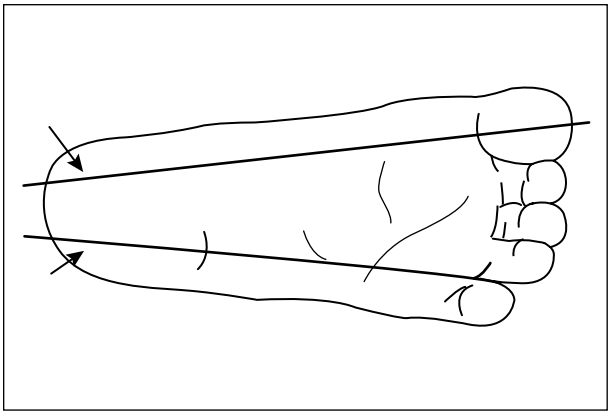


Figure 3.48.1 Heel prick zones. For blood sampling a baby's heel should be pricked *outside* the lines shown and in the arrowed areas.

General problems of preterm babies

Assessing gestation

Sometimes a mother is unaware of her last menstrual period. The baby's gestational age can then be assessed to within ±2 weeks on morphological criteria related to skin, nipples, ears and foot creases (Table 3.48.1).

Common complications

The more immature the baby, the more likely are the following:

- Poor temperature control, hypothermia
- Jaundice
- Respiratory distress
- Difficulty establishing feeds
- Cerebral haemorrhage
- Infection – early or late onset
- Apnoeic attacks
- Hypoglycaemia

Temperature

The development of incubators earlier in the 20th century significantly reduced the mortality of preterm babies. Nursing babies in incubators is covered by standard texts but Table 3.48.2 gives settings from which to start, adjusting incubator temperature up or down to maintain the baby's axillary temperature at 36.5°C.

If an incubator is not available a small baby's temperature can be maintained by attending to the following

Table 3.48.1 Gestational assessment

Score	1	2	3	4
Breast size	<5 mm	5–10 mm	10 mm	
Nipple formation	No areola nipple visible	Areola present nipple well formed	Areola raised. Nipple well formed	
Skin opacity	Numerous veins and venules present	Veins and tributaries seen	Large blood vessels seen	Few blood vessels seen or none at all
Scalp hair	Fine hair	Coarse and silky individual strands		
Ear cartilage	No cartilage in antitragus	Cartilage in antitragus	Cartilage present in antihelix	Cartilage in helix
Fingernails	Do not reach finger tips	Reach finger tips	Nails pass finger tips	
Plantar skin creases	No skin creases	Anterior transverse crease only	Two-thirds anterior sole creases	Whole sole covered

Score:	7	8	9	10	11	12	13	14	15	16	17	18	19	20	21
Week:	27	28	29	30	31	32	33	34	35	36	37	38	38	39	40

Notes:
Test fingernails by scratching them along your hand.
Skin creases are the deep creases not the fine lines.

Table 3.48.2 Incubator temperatures

Weight	Day 1	Day 2	Day 3	Day 4
<1200 g	35°C	34°C	34°C	33.5°C
1200–1500 g	34°C	34°C	33.5°C	33.5°C
1500–2500 g	33.5°C	33°C	32°C	32°C
>2500 g	33°C	32.5°C	31°C	30.5°C

Figure 3.48.2 Skin–skin nursing.

points:
- Dry the baby as soon as possible at birth.
- Put the baby against the mother's chest, inside her clothes.
- Put a thick hat on the baby's exposed head (it forms a significant part of the surface area from which heat loss will occur).

Such skin–skin nursing is often known as the "kangaroo mother method". Randomised trials in both industrialised and resource poor countries have shown that it is associated with increased prevalence of breastfeeding, reduced incidence of apnoea and reduced risk of infection.

Feeding

Babies born after 34 weeks are generally mature enough to suck and swallow but may be less demanding of feeds than term babies. Attention to the following points can help a preterm baby to establish breastfeeding:
- Encourage early and prolonged skin contact (see above)
- Encourage small, frequent feeds by waking the baby every three to four hours and putting to the breast
- If the baby will not latch on and suck, encourage the mother to express her breast milk after attempting a feed; the expressed milk can be given to the baby by gavage (orogastric/nasogastric tube) or cup
- Avoid giving formula or breast milk by bottle – a small feeding cup (about the size of a medicine measuring cup with a smooth rim), or a syringe can be used. Give 20 ml/kg of expressed breast milk every 3 hours
- If too unwell to suck or drink from a cup, give expressed breast milk via nasogastric tube
- As the baby becomes more vigorous encourage a transition to demand breastfeeding.

Other problems

Problems such as respiratory distress, jaundice, infection and apnoeic attacks are covered in the relevant general sections below. **Give all unwell neonates 1 mg IM of only vitamin K**. ✔

Birth trauma

Swellings around the head

- The commonest is a **caput succedaneum**, oedematous tissue over the occiput present after vaginal delivery. This usually resolves within 24-hours and is of no consequence.
- A **cephalhaematoma** is a lateral (sometimes bilateral) fluctuant swelling well circumscribed by the sutures. It does not cross the midline and anatomically represents a subperiosteal haemorrhage. There may be an associated skull fracture but neither this nor the swelling itself usually need treatment. The only important complication can be worsening of physiological jaundice (see below) as the blood is broken down and resorbed.
- A **subgaleal haemorrhage** (bleeding between the skull periosteum and the scalp aponeurosis) is the least common but most dangerous scalp swelling – it represents haemorrhage beneath the aponeurosis of the scalp. Onset and progression is often insidious, often leading to significant haemorrhage. The boggy swelling of the head, extending from above the eyes to the occiput may only be noticed after the baby has developed hypovolaemic shock (see below for treatment).

Nerve palsies

- **Facial nerve palsies** are usually associated with forceps delivery. They usually resolve within 2–3 days.
- **Brachial plexus** usually follows shoulder dystocia and reflects traction injury to the upper roots of the brachial plexus. The arm is flaccid and the wrist flexed – this can most clearly be demonstrated by eliciting a Moro reflex. Look for signs of respiratory distress as the ipsilateral phrenic nerve is sometimes affected. An *X* ray

should be obtained to exclude a pseudoparesis associated with clavicular fracture. The humerus should also be included in the *X* ray since humeral fractures may also occur. Most brachial plexus palsies resolve within 3–4 weeks of delivery but rarely are permanent. Once fractures have been ruled out the mother can be shown how to perform passive movements to reduce the possibility of joint contractures developing.

Fractures

The most common are skull and clavicular fractures. Neither usually require specific treatment.

Common congenital anomalies

Talipes equinovarus

Talipes equinovarus is inversion and flexion of the foot at the ankle. It is usually "positional", i.e. the foot can easily be brought into the normal position. This, and talipes calcaneovalgus (in which the foot is everted and dorsiflexed) do not require treatment. If talipes is fixed, and the foot cannot easily be put in a normal position, it needs treatment (it is helpful to note that this form of fixed talipes is usually associated with the presence of a groove on the medial aspect of the foot). The foot should be splinted and strapped in the position closest to normal and an orthopaedic surgeon's advice sought. ✔ **Whenever talipes is present, be sure to examine the hips carefully for evidence of developmental dysplasia (also known as "congenital dislocation of the hip")**.

Extra digits

It is important to distinguish "preaxial" extra digits (radial aspect of the hand) from "post axial" (ulnar). The former may be associated with other congenital anomalies, particularly of the heart, spine, kidney or gut (tracheal or anorectal). Postaxial polydactyly is, however, of cosmetic significance only. Digits are often held by only a thin pedicle of tissue which can be excised after ligating the base.

Supernumerary nipples and preauricular skin tags

These are often found and are of cosmetic concern only. They should be left alone.

Fluid and electrolyte balance

✔ **When giving fluid or blood intravenously, you must use an in line infusion chamber/burette to avoid fluid overload**.

Water requirements

- Body water content is high at birth. Giving large volumes of fluid will rapidly make a baby oedematous and worsen any respiratory disease (respiratory distress is normally associated with fluid retention which will exacerbate the problem). A **simple general rule** is to start an ill newborn baby who cannot take enteral fluids (breast milk) on 60 ml/kg/day IV 10% glucose, increasing in daily steps of 20–30 ml/kg/day to a maximum of 140–180 ml/kg/day. In a small-for-gestational age baby it may be necessary to begin with an intake of 90 ml/kg/day in order to meet the glucose requirements (see glucose requirements, below).
- Insensible water loss (mainly through the skin) is high in some circumstances, particularly at under 29 weeks gestation, ***or when an overhead heater (radiant warmer) rather than an incubator is used***. Helpful measures to reduce insensible water loss are:
  - Maintaining a high level of humidity in the incubator. Aim at producing between 50–80% relative humidity – at 70–80% condensation is often just beginning to appear on the cooler incubator walls. Maintaining humidity also helps to keep very immature babies warm because evaporative heat loss is reduced.
  - Clothing the baby, or wrapping the body below the head with bubble wrap or aluminium kitchen foil.
- When an overhead heater is used, the infant should not be covered, and the heater output must be adjusted in direct response to the infant's skin temperature ***(typically achieved by a continuous temperature probe servo system)***.
- In the first week of life, high insensible water loss will be reflected by high weight loss (>10% of birthweight) and often an increase in the plasma sodium concentration to 150 mmol/litre or higher. If either occurs the fluid intake should be increased by 30 ml/kg/day. When nursing a low birthweight infant under an overhead heater it is advisable to pre-empt this situation by adding in an extra allowance of 30 ml/kg/day right from the start, i.e. start at 90 ml/kg/day rather than 60 ml/kg/day. Note however that even 30 ml/kg/day might not be enough to meet the insensible losses of a very preterm baby (under 29 weeks) under a radiant heater. Such babies are much better nursed in humidified incubators.
- In very low birthweight infants enteral feeds should be advanced slowly with 20–30 ml/kg/day increments. Babies who are being enterally fed but are unable to breastfeed can be given expressed breast milk by orogastric tube in the amounts referred to above, i.e.:

Day 1	60 ml/kg/day
Day 2	80–90 ml/kg/day
Day 3	100–120 ml/kg/day
Day 4	120–150 ml/kg/day
Day 5	140–180 ml/kg/day

- Monitor the fluid intake by weighing daily and recording frequency of urine output. Infants under 29 weeks should be weighed more than once each day; fluid intake may need to be adjusted frequently to maintain fluid balance. Urine output can be monitored by measuring the difference between wet nappies (diapers)

and a dry one using kitchen scales. Look for signs of fluid overload (oedema) or dehydration. If possible measure the plasma electrolytes, but always remember these cannot be interpreted without information on body weight and urine output.

Electrolyte requirements

Electrolyte solutions added to daily maintenance fluids are shown in Table 3.48.3.

- Sodium 2.5 mmol/kg/day will meet the needs of most term babies. This can be provided by supplementing the daily intravenous 10% glucose allowance with small amounts of 30% sodium chloride (contains 5 mmol Na^+ per ml) or 23% solution (contains 4 mmol of Na^+ per ml). The sodium requirements of preterm babies may be much higher as urinary sodium losses may approximate 10 mmol/kg/day in those of 29 weeks' gestation or less. In general, 10% glucose with 0.225% sodium chloride will provide average daily requirements (infusion rate is adjusted for day post delivery and weight changes as above).
- Sodium supplements can be commenced on the second day of life in babies receiving intravenous fluids but if there is respiratory distress it is wise to wait until the diuresis associated with recovery begins (often delayed until the third or fourth day of life).
- Potassium supplements. 1–2 mmol/kg/day will meet requirements and can be provided by adding small amounts of concentrated (15%) potassium chloride to 10% glucose in amounts shown in Table 3.48.3. If potassium supplements are given the plasma potassium concentration should be monitored daily. It is preferable to administer KCl by continuous IV infusion over 12–24 hours. Supplemental KCl should always be given slowly (over 1–6 hours) at a maximum IV infusion rate of 0.5–1 mmol/kg/hour. The concentration of KCl in peripheral IV solutions should not exceed 60 mmol/litre.
- Potassium and calcium supplements are best given orally and unless low serum values are identified are not usually needed until babies begin to grow.

Glucose requirements

Infusing glucose at the following rates will match the normal hepatic glucose output and therefore maintain blood glucose concentration at an acceptable level:

Term infant	3–5 mg/kg/min
Preterm, appropriate weight for gestation	4–6 mg/kg/min
Small-for-gestational age	6–8 mg/kg/min

These infusion rates provide minimal glucose requirements to maintain euglycaemia but higher rates may be required for growth. If much higher rates of infusion are needed consider hyperinsulinism as a cause of the problem. Always use 10% glucose for peripheral IV infusions; a central venous line or umbilical venous catheter will be needed if high glucose requirements or limits on fluid volume necessitate a more concentrated solution (see section on hypoglycaemia below).

Bleeding and hypovolaemic shock

Causes

A baby's blood volume approximates 80 ml/kg of body weight. Intrapartum fetal haemorrhage of relatively small amounts of blood can therefore result in hypovolaemic shock at delivery. Common causes are abruptio placentae, fetomaternal haemorrhage (this can be diagnosed retrospectively by ordering the Kleihauer–Betke test on maternal blood) or sub-galeal haemorrhage (see above). Other causes are vasa praevia and accidental incision of the placenta during Caeserean section. Bleeding in the first week of life is uncommon but may signify haemorrhagic disease of the newborn or clotting factor deficiency.

Table 3.48.3 Quantities of electrolyte solutions required for daily maintenance

	Sodium chloride 30% (5 mmol/ml)		Potassium chloride 15% (2 mmol/ml)		Calcium gluconate 10%* (0.225 mmol/ml)	
Dose	At 3 mmol/kg/day: ml/100 ml	At 5 mmol/kg/day: ml/100 ml	At 2 mmol/kg/day: ml/100 ml	At 3 mmol/kg/day: ml/100 ml	At 0.8 mmol/kg/day: ml/100 ml	At 1.0 mmol/kg/day: ml/100 ml
40 ml/kg/day	1.50	2.50	2.50	3.75	8.90	11.10
60 ml/kg/day	1.00	1.65	1.70	2.50	5.80	8.00
75 ml/kg/day	0.80	1.35	1.30	2.00	4.70	6.30
90 ml/kg/day	0.65	1.10	1.10	1.70	3.90	5.20
105 ml/kg/day	0.55	0.95	0.95	1.40	3.30	4.20
120 ml/kg/day	0.50	0.85	0.81	1.25	2.90	4.00
135 ml/kg/day	0.45	0.75	0.74	1.10	2.60	3.50
150 ml/kg/day	0.40	0.65	0.67	1.00	2.30	3.00
165 ml/kg/day	0.35	0.60	0.60	0.90	2.10	2.80
180 ml/kg/day	0.30	0.55	0.54	0.80	1.90	2.50
200 ml/kg/day	0.30	0.50	0.45	0.75	1.80	2.20

To replace nasogastric drainage add 5 ml of 15% KCl (10 mmol) to 500 ml of 0.9% NaCl and give IV an amount equivalent to gastric losses.

* IV calcium if extravasated causes severe skin burns.

Presenting features

- The baby will look pale, have weak peripheral pulses, tachypnoea and a tachycardia that may exceed 200 beats/minute. Blood pressure may be low or undetectable, even in a term baby.
- ✔ **In the acute stage packed cell volume and haemoglobin concentration are unreliable indicators of the amount of blood lost – they may be normal in an infant with acute hypovolaemic shock**.
- Common sites of blood loss include the umbilical stump and gastrointestinal tract. In the latter case there may be doubt as to whether blood is of infant or maternal origin (blood swallowed at delivery or from a bleeding nipple). In some cases this can be resolved by the Apt's test.

Apt's test

- Mix one part of the blood containing fluid (vomitus, gastric aspirate or liquid stool) with five parts distilled water. Centrifuge then mix 1 ml of supernatant with 0.25 ml of 0.25% NaOH. A yellow-brown colour signifies maternal blood whereas fetal haemoglobin remains pink.

  Revealed blood loss rarely results in hypovolaemic shock.

Treatment

- In emergency in a shocked baby give O Rh-negative blood (20 ml/kg) at a rate depending on the degree of shock (usually the first 10 ml/kg can be safely given over 15 minutes), monitoring the response and reducing the rate of infusion as improvement occurs. Sometimes a further 10 ml/kg or 20 ml/kg may be necessary.
- If blood is not immediately available use 10–20 ml/kg of 4.5% albumin or 0.9% saline.
- If there is overt bleeding take a blood sample for platelet count, film and clotting studies. Then give 1 mg vitamin K (phytomenadione or phytonadione) IV. If bleeding continues give 20 ml/kg of fresh frozen plasma, if available. Reserve platelets unless the count is $<60\times10^9$ per litre. Bleeding due to haemorrhagic disease of the newborn usually stops within 30 minutes of vitamin K administration.

Enteral feeding

Type of milk

Breast milk will supply the nutrient requirements of almost all babies. For more preterm babies (<1500 g) the following supplements may be needed:

- Multivitamin supplement. A supply of vitamin D 400 IU/day is particularly important for bone mineralisation.
- Iron supplement from about 4 weeks of age. Preterm babies have reduced iron stores compared with term babies. The daily dietary supplementation is 2 to 4 mg/kg of elemental iron, up to a maximum of 16 mg/day.
- Phosphorus supplements. Sometimes very small babies (<1.25 kg) become hypophosphataemic (plasma phosphorus <1.5 mmol/litre) as a result of phosphorus deficiency. If untreated this may result in metabolic bone disease. Addition of a concentrated phosphorus salt to feedings to provide an additional 50 mg/kg/day of phosphorus will prevent this (adding 0.05 ml/kg of a 4 mmol/ml phosphorus solution to each of eight feeds per day will give 50 mg/kg/day supplemental dietary phosphorus).
- Babies with renal sodium wasting may also need sodium supplements. If the plasma sodium concentration falls below 130 mmol/litre, add an additional 2.5 mmol/kg/day sodium to the milk until it rises into the acceptable range (134–144 mmol/litre).

Method of feeding

See feeding under the general problems of preterm babies above.

Hypoglycaemia

Hypoglycaemia is a common problem in the nursery; it can occur in babies who appear well and in babies who are sick and can present with or without symptoms. For these reasons it is important to identify an infant at risk for adequate prevention and to diagnose it promptly. ✔ **Untreated hypoglycaemia can result in brain damage**.

The overall incidence is 1 to 5 per 1000 live births. However, in high-risk infants the incidence can be up to 30% (for example 8% in large-for-gestational age and 15% in preterm and small-for-gestational age infants).

Definition

The definition of hypoglycaemia is controversial and no studies have determined an absolute value at which organ dysfunction will occur. It is known, however, that prolonged low levels are associated with brain injury. The neonatologists, at this time, like to maintain blood sugar levels at least above 2.2 mmol/litre (40 mg/dl).

Causes of hypoglycaemia

Increased utilisation of glucose: hyperinsulinism

- Infants of diabetic mothers
- Erythroblastosis fetalis
- Islet cell hyperplasia
- Beckwith–Wiedemann syndrome
- Insulin-producing tumours
- Maternal beta-agonist tocolytic therapy
- Abrupt interruption of high glucose infusion
- Malpositioned umbilical arterial catheter infusing high concentration of glucose into coeliac and mesenteric arteries (T11–T12) stimulating insulin release

Decreased production/stores

- Prematurity
- Small-for-gestational age
- Inadequate caloric intake

Increased utilisation and/or decreased production or other causes

- Perinatal stress (asphyxia, sepsis, shock, hypothermia)
- Polycythaemia
- Defects in carbohydrate metabolism (galactosaemia, fructose intolerance, glycogen storage disease)

- Endocrine deficiency (adrenal insufficiency, hypothalamic insufficiency, glucagon deficiency)
- Defects in amino acid metabolism (maple syrup urine disease, propionic acidemia, methylmalonic acidaemia, tyrosinaemia)
- Exchange transfusion

Infants at risk for hypoglycaemia

- Infants of diabetic mothers
- Preterm babies
- Small-for-gestational age babies
- Large-for-gestational age babies
- Post-term babies
- Sick babies with infections and respiratory failure
- Fasted babies

Diagnosis of hypoglycaemia

There are few normal data on blood glucose concentration in the first week of life, particularly for healthy breast-fed term babies. Moreover **there is little evidence that a transient low blood glucose concentration in term babies who show no physical signs is harmful**. Indications to measure the blood glucose concentration of a term baby include seizures (see below), pronounced hypotonia or diminished consciousness. Association between such signs and low blood glucose concentration is described as "symptomatic hypoglycaemia". There is no place for measuring blood glucose concentration just because a baby has not fed.

- Beware of blaming signs in the baby on "hypoglycaemia". Remember a baby who seems drowsy may be infected (see below) and low blood glucose concentration may merely be an associated finding, not the root cause of the problem.
- Blood glucose concentration in the first 6 hours of life is very often low (<1.5–2.0 mmol/litre). There is no evidence this is harmful for otherwise healthy babies who adapt by mobilising other fuels. Consequently early testing (<6 hours of age) is pointless – unless neurological signs are present.

When to test

- **Symptomatic infants** (lethargy, poor feeding, temperature instability, respiratory distress, new-onset apnoea/bradycardia, jitteriness, seizures): immediately.
- **Infants at risk**: soon after birth (within 6 hours), then 3-hourly until stable at 2.5 mmol/litre (45 mg/dl) or higher. Continue to monitor until feeds are well established.
- **Infants with hypoglycaemia**: check blood glucose 20–30 min from beginning of treatment, then hourly until stable at 2.5 mmol/litre (45 mg/dl) or higher. Continue to monitor frequently (every 4–8 hours) during treatment and while decreasing supplemental intravenous glucose infusions.

Laboratory diagnosis

- **Reagent strips** are useful and rapid but in general are less reliable than laboratory plasma glucose measurements. Reagent strips may show a glucose level as much as 15% lower than plasma levels since whole blood is used. When ever possible, it is preferable to use a calibrated glucometer when using reagent strips.
- **Laboratory plasma glucose determinations**, if available, are useful for confirming hypoglycaemia detected by reagent strips, but blood samples must be processed promptly for accurate values since glycolysis occurs in standing whole blood samples. **Do not wait for laboratory confirmation, before initiating therapy**. ✔

Management of hypoglycaemia

Infants at risk for hypoglycaemia, appearing to be well

- Initiate early feeding within 1 hour after birth with breast milk or formula (only if breast milk is not available), repeated every 2–3 hours.
- Feeding with 5% glucose is not recommended in infants with hyperinsulinism because milk provides more energy.
- Infants of diabetic mothers are unlikely to develop hypoglycaemia on the second day of life if tests in the first 24 hours are satisfactory.

Infants with symptomatic hypoglycaemia, or unable to feed or who failed correction of glucose levels with enteral feeding

- Start intravenous glucose bolus 200 mg/kg over 5 min (2 ml/kg of 10% glucose in water). **Remember that giving high concentration of glucose solutions by bolus injections can itself be dangerous, particularly from effects on the brain**. ✔
- Follow with maintenance infusion of 10% glucose at a rate of 5–8 mg/kg/min.
- If further episodes of hypoglycaemia occur, bolus should be repeated and the infusion rate increased by 10–15%.
- Exclude infection. A baby who seems drowsy may be infected and a low blood glucose may be an associated finding not the main cause of the problem.
- When administering boluses, never use high concentrations of glucose (>10%) because of risk of intraventricular haemorrhage and/or cerebral oedema.
- The concentration of glucose in the maintenance fluids can be increased in accordance with the total daily fluids requirements.
- If using concentrations greater than 12.5% a central venous line or umbilical venous catheter needs to be inserted because of risk of tissue damage in case of fluid extravasation.
- Most infants will correct hypoglycaemia with infusion of 5–8 mg/kg/min; it is not infrequent though that infants with severe intrauterine growth retardation and those with hyperinsulinism may require infusion rates up to 12–15 mg/kg/min.
- When normal blood glucose levels have been stable for 12–24 hours and the infant is tolerating enteral feeding, decrease the intravenous glucose infusion by

10–20% each time levels are greater than 2.5 mmol/L (45 mg/dl).
- Always decrease intravenous infusion gradually because of the risk of precipitating hypoglycaemia.
- If unable to gain intravenous access, Hypostop gel an oral glucose mixture containing 500 micrograms of glucose per ml can be helpful. Give 1–2 ml to the oral mucosa.

If hypoglycaemia persists beyond the first week of life and requires large infusions of glucose (greater than 8 mg/kg/min), evaluation for endocrine or metabolic disorders should be considered (if possible).

Jaundice (see algorithms for management, Chapter 2.16)

Physiological jaundice

"Physiological jaundice" is common, affecting at least a third of normal term babies. Jaundice can be considered physiological and does not require treatment or investigation if the following criteria are met:
- Jaundice is not present in the first 24 hours of life
- The baby is well, free of signs of infection (see below) without enlargement of liver or spleen
- The bilirubin concentration does not exceed 300 micromoles/litre (approximately 17 mg/dl) at any stage (term babies only – a much lower acceptable level is set for preterm babies, see below)
- The bilirubin concentration reaches a peak on the fourth or fifth day of life
- The jaundice has fully resolved by the end of the second week of life.

The risk of exaggerated physiological jaundice can be reduced by encouraging early, unrestricted demand breastfeeding. **Note**: There is no evidence whatsoever to support the widely held belief that giving extra water either reduces the risk of jaundice or is helpful in treatment. In fact the converse has been shown: giving water is likely to reduce the frequency of breastfeeds and **increase** risk of jaundice. Dehydration should be avoided by encouraging frequent feeds.

Measuring bilirubin concentration

- Bilirubin concentration can be most simply and accurately measured by simple spectrophotometry of serum obtained by centrifuging blood in a capillary tube. Several easily operated machines are available. If used, staff should be trained, and the machine calibrated daily and checked with control specimens of known bilirubin content. Using dirty tubes (or cuvettes), haemolysed or lipaemic samples can produce significant errors.
- If laboratory equipment is not available a rough estimate of bilirubin concentration can be obtained by:
  - Using the "icterometer" (Gosset's icterometer). This is a perspex strip shaded with yellow pigment. It can be applied to the skin on the tip of the nose (in dark-skinned babies the gums are more reliable) and the bilirubin read off by colour matching. It is only accurate to about ±20% and can underread at high levels. Therefore an accurate serum bilirubin measurement is needed before providing treatment such as exchange transfusion.
  - Another rough method is visual inspection (Kramer):

Any jaundice detectable	>90 micromoles/litre
Head and neck only	70–130 micromoles/litre
Trunk, elbows and knees	190–310 micromoles/litre
Hands and feet jaundiced	>300 micromoles/litre

Again, a laboratory (spectrophotometer) reading is needed before initiating an exchange transfusion.

Prolonged jaundice

In prolonged jaundice (jaundice present after 14 days of age) it is important not just to know the total bilirubin concentration but the proportion of **conjugated bilirubin**. Conjugated bilirubin is not neurotoxic but its presence signifies presence of biliary obstruction attributable to potentially serious conditions such as neonatal hepatitis or biliary atresia (see Chapters 3.9 and 3.10). If laboratory investigation is not available, the history may be informative – are the stools unpigmented or the urine dark? Also the urine can be tested with a reagent strip – if positive for bilirubin the diagnosis of biliary obstruction is supported.

Pathological jaundice

Causes

Jaundice which does not satisfy the above criteria is pathological and may place the baby at risk of **kernicterus** and/or **sensorineural deafness**. In the first week of life the following factors may lead to jaundice sufficiently severe to require treatment:
- **Preterm delivery**. Even moderate prematurity (for example birth at 36 weeks) significantly increases the risk of early or severe jaundice occurring and the risk of associated sequelae. Consequently the bilirubin treatment charts (see below) give lower treatment thresholds for babies 31–34 weeks gestation. At under 31 weeks, treatment is started at even lower bilirubin levels.
- **Haemolytic disease**. This may be isoimmune (for example Rh or ABO incompatibility) or due to red cell disorders, for example hereditary spherocytosis or glucose–6 phosphate dehydrogenase deficiency (G6PD).
- **Infection**. Haemolysis and impaired elimination of bilirubin may be associated with septicaemia. Congenital infection (for example rubella, cytomegalovirus infection) may also be associated with jaundice, but other features such as rash, hepatosplenomegaly, thrombocytopenia will be present, and there is usually a significant conjugated bilirubin level (up to 50% of total bilirubin).
- **Rarer causes**. These include inborn errors of metabolism (galactosaemia) and congenital hypothyroidism.
- **Obstructive jaundice**. Rarely presents in the first week of life but is important in the differential diagnosis of prolonged jaundice (see above).

Investigation

In a baby who develops jaundice in the first 24 hours the most likely causes are infection and haemolytic disease. History and examination may be helpful. Has the mother had previously affected babies or is she known to have a hereditary haemolytic disorder? Are there risk factors for infection or clinical signs of sepsis (see neonatal infections below)? Is there hepatosplenomegaly suggesting congenital infection or haemolytic disease? Useful laboratory tests are:

- Mother's and baby's ABO and Rh blood groups. Save serum to cross-match if exchange transfusion needed
- Direct Coombs test (if positive indicates an isoimmune haemolytic anaemia)
- Complete blood count and reticulocyte count (anaemia and reticulocytosis indicating haemolysis and/or abnormal white blood cells indicating possible infection)
- Peripheral blood smear (abnormal red cell morphology and/or fragmented red cell forms suggesting a specific red cell disorder and/or haemolysis)
- Thyroid function tests and urine test for non-glucose reducing substance (possible galactosaemia) are useful tests for high levels of bilirubin that are prolonged.

Treatment

The bilirubin treatment charts (Figure 3.48.3) show intervention lines for the two principal treatments – phototherapy and exchange transfusion. In general the smaller the baby and the sicker the baby, the greater the urgency to intervene. Bilirubin in plasma is normally bound to albumin, but in a sick, acidotic baby less binding occurs, and more "free" bilirubin will be available to enter the central nervous system. Therefore intervene about 40 micromoles/litre below the indicated line in such circumstances.

The specific bilirubin levels for which phototherapy and exchange transfusions need to be considered in infants under 31 weeks gestation are less certain. A frequently used guideline is to initiate phototherapy when the bilirubin level approaches 85 micromoles/litre per kg birthweight (approximately 5 mg/dl per kg birthweight), and to consider an exchange transfusion for levels above 170 micromoles/l per kg birthweight.

- **Phototherapy**. This uses light in the blue-green end of the spectrum (**not ultraviolet**) to convert bilirubin to its water-soluble isomer – (lumirubin) which can be excreted in urine and stools. The baby should be nursed naked in an incubator to allow maximum skin exposure but can be removed for breastfeeds as necessary (intermittent treatment has been shown to be as effective as continuous). A mask should be used to shield the eyes. Troublesome side effects of phototherapy include rashes and profuse watery stools but these do not require treatment. The total daily fluid intake may need to be increased, especially in preterm babies, to minimise additional water losses from evaporation and convection.
- **Exchange transfusion**. Bilirubin levels rising above certain threshold values place an infant at risk for developing kernicterus. In these cases, the bilirubin level should be immediately lowered with a double volume exchange transfusion; a volume of the infant's blood equal to the bodyweight in kg × 2 × 80 ml/kg is exchanged in small aliquots with packed red cells O Rh-negative blood, or blood cross-matched against maternal antibodies (see exchange transfusion on Chapter 6.9). Double-volume exchange transfusion is recommended in **term babies** if:
  - Haemolysing or ill and bilirubin = 300–400 micromoles/litre
  - Well, not haemolysing but bilirubin = 425–500 micromoles/litre

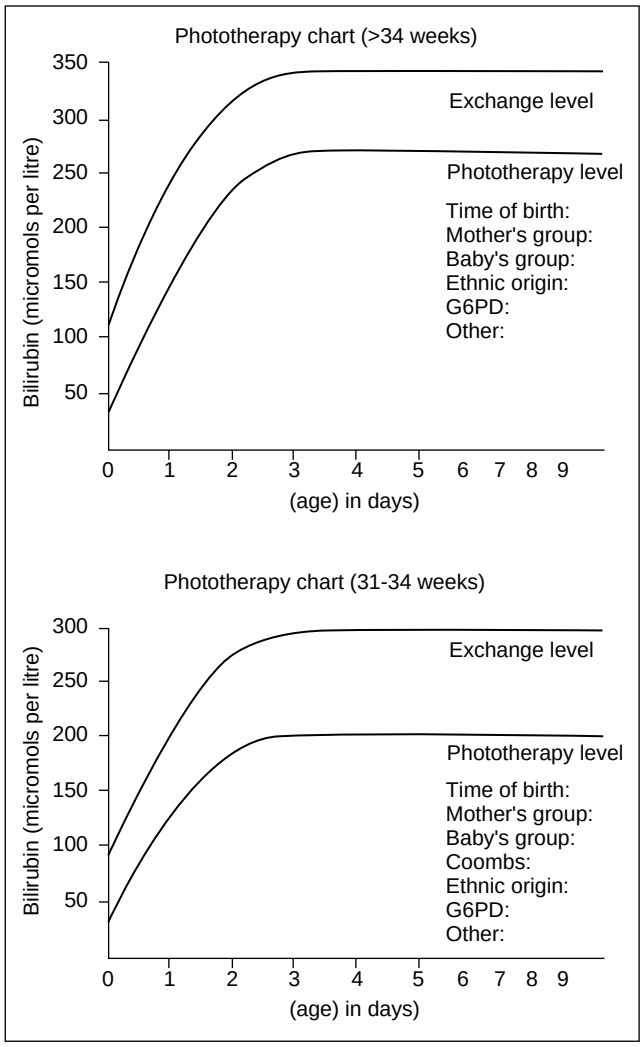


Figure 3.48.3 Bilirubin treatment charts.

For details of doing an exchange transfusion see Chapter 6.9.

Respiratory distress

Definition

Three cardinal signs characterise respiratory distress:

- Tachypnoea: respiratory rate >60 per minute.
- Retractions (recessions): tugging of the soft tissues between the ribs or at the edges of the rib cage.
- Grunting: a prolonged expiratory effort usually with an audible noise.

Two of these signs are sufficient to make the diagnosis. Note that cyanosis is not a necessary diagnostic feature

but is often present if oxygenation is compromised by the underlying lung condition.

If pulse oximetry is available, SaO_2 in distressed infants will be <94% in air at sea level, usually <90% (normal values for SaO_2 for healthy infants should be determined if the hospital is based at high altitude: see Chapter 3.19).

Causes of early respiratory distress

"Early" respiratory distress – presenting in the first 12 hours of life – has a number of causes:

- "Transient tachypnoea" associated with a delay in clearing of fetal lung fluid
- Congenital pneumonia or sepsis
- Surfactant deficiency (also known as "respiratory distress syndrome")
- Pneumothorax
- Meconium aspiration
- Congenital abnormalities of the lung or airways (including diaphragmatic hernia).

Maternal fever during labour and prolonged rupture of the fetal membranes (>18 hours) particularly point to pneumonia or sepsis. Pneumothorax should be considered if the baby has been resuscitated using positive-pressure ventilation (although this has been described as occurring spontaneously in about 1% of normal term babies). Transient tachypnoea is more common among babies delivered prematurely, or by elective caesarean section (in the absence of spontaneous labour). Surfactant deficiency and infection are the most likely causes in preterm babies.

Note that congenital heart disease does not usually cause early respiratory distress. **Cyanosis is the more likely presentation**; respiratory distress associated with heart failure normally occurs after the first week of life in association with tachycardia, pallor, sweating, hepatomegaly and excessive weight gain.

Principles of treatment

- Assess the baby's oxygenation, clinically or using pulse oximetry if available. If cyanotic give oxygen until pink. If pulse oximetry is available keep SaO_2 94–98% (to avoid retinopathy do not give too much oxygen and therefore do not permit SaO_2 values of >98% in preterm infants).
- ***Where there are facilities an arterial sample should be obtained for blood gas analysis.***
- Once the baby is adequately oxygenated blood should be taken for culture, an intravenous line established and antibiotics given. **Antibiotic therapy is always indicated** until an infectious process has been ruled out; neonatal sepsis or pneumonia, if present, progress rapidly and cannot be excluded on clinical grounds. Ampicillin/penicillin and an aminoglycoside (or a third-generation cephalosporin) are the most appropriate empiric antibiotic choices. ✔
- Infants with mild, steadily resolving respiratory symptoms may be observed without further diagnostic procedures, but if there is severe distress, or the pulmonary symptoms are not rapidly improving, a diagnostic chest *X* ray should be obtained if available.
- Oral feeding should be avoided if possible: intravenous infusion of 10% glucose (60 ml/kg/day) is safest, ideally through a peripheral vein but if not possible through an umbilical venous catheter. If there are no facilities for intravenous infusion, breast milk or 10% glucose may be given in limited quantities (up to 60 ml/kg/day) by **orogastric tube**. Note: **nasogastric tubes** may contribute to upper airway resistance so an orogastric tube is preferred in infants with pulmonary compromise.

Assisted ventilation (see Chapters 1.26 and 6.18)

Continuous positive airway pressure (CPAP)

Indications for CPAP and continuous negative extrathoracic pressure (CNEP) are:

- Respiratory distress syndrome
  CPAP or CNEP are non-invasive and nurse controlled therapies which can provide powerful support to the breathing of infants with respiratory failure resulting from a number of causes (see Chapters 1.26 and 6.18)
- Apnoea of prematurity (see below)
  Preterm infants with normal lungs who develop late-onset (greater than 48 hours old) apnoeic attacks failing to respond to simple measures.
- Post-ventilation
  A period of nasal CPAP or CNEP following ventilation is useful to prevent atelectasis.

Pneumothorax

It may be possible to diagnose a pneumothorax clinically. The abdomen is often distended by downwards displacement of liver and spleen. The breath sounds may be reduced on the affected side. Transillumination can be useful if a ***"cold light" (fibreoptic light source)*** is available: the affected side may glow brightly. A chest *X* ray is the most reliable, if available – mediastinal shift indicates tension and a chest drain will need to be inserted. **If there is rapid clinical deterioration, however, with severe hypoxaemia and/or cardiovascular compromise (bradycardia, hypotension), and a pneumothorax is strongly suspected, immediate needle thoraco-centesis (see Chapter 6.11) prior to *X* ray confirmation may be required**. A pneumothorax that does not result in severe respiratory distress, and is not under tension, may spontaneously resolve without mechanical removal of the pleural air, but careful monitoring is required. ✔

Apnoea

Apnoea is a cessation of respiration or a hypoxaemic event associated with signs of cardiorespiratory decompensation (bradycardia, cyanosis, pallor). Apnoeic episodes are common in preterm babies <32-weeks gestation ("apnoea of prematurity"). In term babies, apnoea usually signifies an underlying pathologic condition.

Causes of apnoea include:

Apnoea of prematurity
These are often characterised by a brief cessation of respiration that responds to gentle tactile stimulation, but they may vary significantly in duration and severity, especially in very low birthweight infants. Sometimes isolated bradycardia and/or brief oxygen desaturation events are identified without clinically apparent apnoea. The aetiology of apnoea of prematurity is often a mixture of impaired central nervous system respiratory control ("central apnoea"), intrapulmonary shunting and upper airway obstruction. The frequency and severity of apnoea events, nonetheless, may be reduced or even eliminated by treatment with a methylxanthine (aminophylline, theophylline, caffeine) which acts on the respiratory centre. Caffeine has become the preferred methylxanthine by some neonatologists because it has a long half-life (daily dosing) and serum levels do not have to be monitored. Continuous positive airway pressure (CPAP) is also effective. ***Rarely mechanical ventilation is necessary***.

Apnoea of immaturity
The diagnosis of "apnoea of immaturity" is one of exclusion. Other processes may cause or exacerbate apnoea. In the case of a preterm baby these include:

- Respiratory distress (surfactant deficiency, pneumonia, pulmonary oedema due to a persistent ductus arteriosus)
- Intraventricular haemorrhage
- Hypoglycaemia
- Temperature instability
- Sepsis.

Severe anaemia may also contribute to apnoea.

Pulmonary parenchymal disease
Any condition causing decreased lung compliance or impaired gas exchange can contribute to apnoea. Appropriate pulmonary support should be provided for adequate gas exchange and the underlying pulmonary condition should be treated.

Airway obstruction
This may result from simple malpositioning of the head (for example hyperflexion or hyperextension of the neck), especially in premature infants. Congenital airway anomalies may also present as apnoea (e.g tracheo-oesophageal fistula or aberrant thoracic blood vessel compressing the trachea – "vascular sling"). Maintaining proper head positioning or surgical correction of the underlying anomaly should be provided.

Infection (see below)
This should always be excluded; antibiotics should be given until infection has been ruled out by subsequent clinical findings and laboratory results (complete blood counts, chest *X* ray, bacterial cultures).

Seizures
See discussion of seizures below. Seizures may present primarily as apnoea. Consider this possibility, especially in term or near-term infants with no other identified cause for apnoea. There may be a poor response to positive pressure ventilation. A history of an operative vaginal delivery (for example forceps) or other birth trauma may signal possible intracranial haemorrhage when this condition occurs in the first one to three postnatal days.

Maternal medication
The most common cause of apnoea in this setting is intrapartum maternal narcotic administration for maternal pain/sedation. The condition can be reversed by the administration of naloxone hydrochloride (100 micrograms/kg, usually given IM). Naloxone should not be administered if there is a history of chronic narcotic use in pregnancy since acute neonatal narcotic withdrawal may be precipitated. Exposure to high magnesium sulphate levels have also been associated with apnoea in the immediate postnatal period, but this is usually a self-limiting process that rarely requires mechanical ventilation.

Neonatal infections

Bacterial sepsis in the neonate may initially present with any number of subtle, non-specific changes in activity or physical findings. There may be a change in feeding pattern, emesis, irritability, pallor, diminished tone, and/or decreased skin perfusion. Other presenting symptoms and physical findings include lethargy, apnoea, tachypnoea, cyanosis, petechiae and early jaundice. There may be fever but this is not common, especially with bacterial infections occurring in the first week. Temperature instability with hypothermia, however, may be seen. Abnormal glucose homeostasis (hypoglycaemia or hyperglycaemia) and/or metabolic acidosis are commonly associated findings.

Early-onset sepsis (first week)

Early-onset sepsis usually occurs as a result of bacteria acquired by vertical transmission from mother to infant intrapartum. The most frequently observed organisms vary around the world. Gram-negative enterics (especially *Escherichia coli* K1) predominate in many regions. Gram-positive cocci are also common and include group B

Table 3.48.4 Caffeine in apnoea of prematurity

Drug and preparations	Each dose	Dose frequency	Administration notes
Caffeine citrate	20 mg/kg 5–8 mg/kg maintenance	Loading dose once daily	If oral dose too large, divide into two and give 1 hour apart
Caffeine base	10 mg/kg 2.5–4 mg/kg maintenance	Loading dose once daily	Give IV loading dose over 30–60 minutes diluted as much as possible

β-haemolytic *Streptococcus*, other streptococcal species, *Staphylococcus*, and *Enterococcus*. Less commonly *Listeria monocytogenes* is isolated from newborn infants with sepsis, especially when there are foodborne epidemics.

Maternal risk factors for early onset sepsis
These include:
- Maternal chorioamnionitis
- Intrapartum maternal fever (especially 38°C or greater)
- Premature rupture of membranes
- Prolonged rupture of membranes (18 hours or greater)
- Preterm labour
- Maternal bacteruria during pregnancy (especially with group B β-haemolytic *Streptococcus*)
- Prior infected infant (group B β-haemolytic *Streptococcus*).

Although early-onset sepsis in the newborn usually results from bacteria acquired from the mother at, or shortly before delivery, vaginal cultures cannot be used to determine the choice of antibiotics when treating the symptomatic newborn suspected of being septic. Lower vaginal and rectal swabs taken within 5–6 weeks prior to delivery can help identify women colonised with group B β-haemolytic *Streptococcus*, and intrapartum prophylaxis with penicillin or ampicillin can be used to help prevent neonatal infection. This approach, however, has not been proven to be useful for any other bacterial pathogen commonly implicated in early onset sepsis.

Laboratory evaluation of the infant
In a baby who is generally unwell with no clinically obvious infective focus the following investigations should be performed:
- Blood culture (about 1 ml venous blood). This should be obtained from a peripheral vein after preparing the skin with an antibacterial wash such as povidone-iodine and/or 70% ethanol or isopropyl alcohol. The blood culture is the gold standard for neonatal sepsis, but it is not 100% sensitive. The sensitivity may be further reduced if intrapartum antibiotics were administered to the mother.
- White blood cell count (WBC) with differential cell count. The WBC in newborn infants shortly after birth typically ranges from 10×10^9/litre to 30×10^9/litre. Lower and higher counts may be seen in the absence of infection. There is usually a predominance of mature neutrophils plus band forms. A total WBC less than 5×10^9/litre or an elevated ratio of forms to total neutrophils (mature neutrophils plus bands) (0.3 or greater) are concerning for infection. **The predictive value of the WBC and differential count when obtained shortly after birth is, however, very poor, and it should not be used to decide on the use of antibiotics in symptomatic infants**. A follow-up WBC and differential at 12–24 hours may be more predictive of infection in infants suspected of having infection, and this may aid in the decision to extend antibiotic therapy in the absence of an identified focus for infection or a positive blood culture.
- Chest *X* ray. This should be obtained if there are any respiratory symptoms.
- Lumbar puncture (cytology and culture).
- Suprapubic aspirate of urine for culture. This procedure is of little value in the infant suspected of having sepsis shortly after birth, but it may have a greater yield in infants with new onset symptoms later in the first week. A urinary tract infection should always be considered in neonates with late onset sepsis (see below).
- Blood glucose concentration.
- Serum bilirubin concentration if baby appears jaundiced.
- Surface cultures (ear canal, umbilical stump) and gastric aspirate cultures do not correlate with either the likelihood of sepsis or the causative agent in septic infants; these cultures should **not** be obtained.

In infants with suspected sepsis, priority should be given to stabilising cardiovascular and respiratory problems while simultaneously obtaining a blood culture followed by prompt administration of antibiotics. Other tests can be obtained once the infant is stable and antibiotics have been given.

Antibiotic treatment (beware of increasing multi-drug resistant organisms)
- Beta-lactam antibiotic plus aminoglycoside. The most frequently used agents are ampicillin and gentamicin, but alternative broad-spectrum coverage may be used. Penicillin may be used if ampicillin is not available, but it has a narrower spectrum limited to Gram-positive bacteria; ampicillin may also provide better coverage for certain Gram-positive pathogens as well. Beta-lactams and aminoglycosides may be synergistic in treating some of the most frequently encountered neonatal pathogens.
- Third-generation cephalosporins such as cefotaxime, ceftazidime or ceftriaxone may be used, but some Gram-positive bacteria may not be covered if a penicillin derivative is not included (for example *Enterococcus* and *Listeria*). These antibiotics may be given intramuscularly if intravenous access cannot be obtained, but frequent use of these drugs may contribute to the development of strains of multiple drug resistant bacteria in nurseries. Ceftriaxone has a longer half-life and can be dosed once daily; however, it should be avoided in infants with hyperbilirubinaemia. In infants with suspected Gram-negative meningitis accompanying early-onset sepsis, the inclusion of a third-generation cephalosporin may be preferred because of a theoretical greater killing power for enteric bacteria in the cerebrospinal fluid.
- Flucloxacillin (intravenous or oral) is preferable for *paronychia* or *septic spots* as these are usually caused by coagulase-positive staphylococci.
- Ciprofloxacin may be helpful in infections of resistant organisms.

Late-onset sepsis

Organisms are less likely to reflect those of the maternal genital tract, although the same pathogens may be identified in infants presenting from home. The most common infections

are infected umbilicus and infected skin or nails. A circumcision wound can also be the site of serious infection.

In the hospital setting, infection more commonly occurs from nosocomial pathogens including coagulase negative staphylococci, Gram-negative enterics (for example *Klebsiella oxytoca*, *K. pneumoniae, Enterobacter cloacae*), *Staphylococcus aureus*, *Pseudomonas* species, streptococcal species and *Enterococcus*. Fungal sepsis must also be considered. Investigate as for early-onset sepsis, with the inclusion of a suprapubic sample of urine, and treat with parenteral broad-spectrum antibiotic therapy directed towards the most commonly encountered pathogens for the particular nursery.

Neonatal tetanus (see Chapter 4.9)

Meningitis (see Chapter 4.1)

Bacterial meningitis in the newborn is usually caused by the organisms named above (early- and late-onset sepsis sections). Mortality and morbidity vary with the pathogen, but on average approximately one-third of affected babies die and up to one-half of the survivors have long-term disability.

Presenting features

May include lethargy, irritability, hypotonia, seizures, generalised signs of accompanying sepsis and a bulging or tense anterior fontanelle. **Always measure and note carefully the head circumference**. ✔

Investigations

Lumbar puncture is essential if meningitis is suspected, and should be undertaken in all newborns with non-localising features of sepsis.

- Cerebrospinal fluid (CSF) cell counts, chemistries and Gram stain. An elevated CSF leucocyte count with a pleocytosis is characteristic. White cell counts of up to 25 cells/cubic millimetre may be seen in term newborn infant, but the cells should be predominately monocytes and macrophages. Rarely, the cell count

Table 3.48.5 Antibiotics in early-onset sepsis

Antibiotic	Each dose IV or IM	Dose frequency	Postnatal age (days)	Gestational age at birth (weeks)	Administration notes
Ampicillin	50–100 mg/kg	12 hourly	<7	Any	Reduce dose frequency in severe renal impairment and birth asphyxia
	50–100 mg/kg	8 hourly	>7	Any	
Gentamicin 20 mg in 2 ml ampoules	4 mg/kg	36 hourly	<7	<28	Measure trough and peak levels: <32 weeks before and after 3rd dose
	4 mg/kg then 24 hours later	Loading dose	>7	<28	
	2.5 mg/kg	24 hourly	>7	<28	>32 weeks before and after 4th dose
	3 mg/kg	24 hourly	<7	28–32	Aim for trough <2 mg/litre peak 6–10 mg/litre
	4 mg/kg then 18 hours later	Loading dose	>7	28–32	
	2.5 mg/kg	18 hourly		28–32	
	2.5 mg/kg	18 hourly	<7	32–38	
	2.5 mg/kg	12 hourly	>7	32–38	
	2.5 mg/kg	12 hourly	<7	>38	
	2.5 mg/kg	8 hourly	>7	>38	
Benzyl Penicillin	50 mg/kg	12 hourly	<7	Any	Reduce dose frequency in severe renal impairment and birth asphyxia
	50 mg/kg	8 hourly	>7	Any	
Cefotaxime	50 mg/kg	12 hourly	<7	Any	Reduce dose by 50% in severe renal impairment
	50 mg/kg	8–12 hourly	>7	Any	
Ceftriaxone	50 mg/kg	24 hourly	Any	Any	Avoid in infants <36 weeks gestation or if jaundiced. Follow special IM preparation instructions
Ceftazidime	50 mg/kg	12 hourly	<7	Any	Increase dose interval to 24 hours in severe renal impairment
	50 mg/kg	8 hourly	>7	Any	
(Flu)cloxacillin	50 mg/kg	12 hourly	<7	Any	Increase dose interval to 24 hours in severe renal impairment. Double the dose in severe infection and if CNS is involved
	50 mg/kg	8 hourly	>7	Any	

may be normal early in bacterial meningitis, but an evolving pleocytosis essentially always develops so a follow-up lumbar puncture at 24 hours may be helpful. The CSF protein in meningitis may be high (more than 2.0 g/litre in a term baby) and the CSF glucose is typically low (less than 1.0 mmol/litre or <30% of blood glucose value). The Gram stain may reveal bacteria, but antibiotic therapy should not be directed on the basis of this result, because rapidly growing bacilli may be mistaken for cocci, or the state of the organism may result in variable staining.

- The CSF picture in preterm babies who have sustained an intraventricular haemorrhage can be confusing: sometimes there is a mild reactive pleocytosis present for the first few weeks of life. If there is clinical suspicion this should be treated as bacterial meningitis until cultures are known to be negative.
- If a "bloody tap" is obtained it is best to treat the baby as though infected and repeat the lumbar puncture after 24 hours. The ratio of red blood cells to white blood cells has been used in older infants and children, but the validity of this ratio has not been demonstrated in neonates and should probably not be used.
- If a CSF pleocytosis is present but no organism is identified, consider ***intracranial imaging (for example head computed tomography with contrast if available)*** to rule out a parameningeal focus (for example brain abscess). This may be particularly important if seizures occur or there are focal neurologic findings. Meningitis caused by *Citrobacter diversus* is frequently accompanied by brain abscess, so, when possible, a ***contrast head computed tomography is always appropriate with this organism if available***.

Treatment

Empiric therapy is the same as for neonatal sepsis (beta-lactam antibiotic and aminoglycoside), but it may be preferable to add a third-generation cephalosporin given the theoretical improved CSF killing power for sensitive Gram-negative bacteria. Therapy can be adjusted once the bacteria has been identified and antibiotic sensitivities are determined. The duration of treatment is at least 14 days for uncomplicated Gram-positive bacteria and 21 days for Gram-negative bacteria. A repeat lumbar puncture may be useful at the end of therapy, especially with Gram-negative meningitis; extending therapy may be appropriate if there is a persistant elevation of the CSF protein. If a parameningeal focus is identified a much longer treatment course is indicated (months). There is no evidence that intrathecal or intraventricular antibiotic therapy improves outcome.

Be vigilant for the presence of seizures and treat as appropriate.

Skin, eye and mucous membrane infections

Conjunctivitis

Most conjunctivitis presents as "sticky eyes", but this may not always be of bacterial origin, especially if it occurs in the first few days (i.e. chemical conjunctivitis from ophthalmologic prophylaxis with topical antibiotic). A bacterial process must be considered in all cases. Infants with a crusting serous discharge without significant conjunctival inflammation may simply have blocked nasolacrimal tear ducts. This usually responds to gentle pressure applied in a downward motion along the nose immediately adjacent to the eyes. The discharge may be cleaned from the eye with sterile 0.9% saline drops. If the condition worsens, or there is conjunctival inflamation, topical therapy with erythromycin, neomycin or chloramphenicol ophthalmic ointments or drops may be applied after obtaining a swab culture.

Rapidly progressive purulent conjunctivitis occurring within the first few days must in countries with high rates of sexually transmitted diseases always be assumed to be due to *Neisseria gonorrhoeae* which must be promptly identified and aggressively treated with parenteral antibiotics and irrigation. Most strains are now resistant to penicillin. Swab for microscopy (Gram-negative intracellular diplococci) and culture (special medium is required such as Thayer–Martin agar with incubation under increased carbon dioxide). Treatment should be initiated promptly before culture confirmation. Intravenous penicillin (see page Table 3.48.5) for 7 days has been used successfully, but because of increased worldwide resistance (penicillinase-producing *Gonococcus*), a third-generation cephalosporin is often selected as the first-line therapy (ceftriaxone 50 mg/kg per day IV or IM for 2 days. If ceftriaxone is not available, kanamycin 25 mg/kg in a single IM dose (maximum 75 mg) or spectinomycin 25 mg/kg IM in a single dose (maximum 75 mg) should be given. ✔

The eyes should be frequently irrigated with sterile normal saline or boiled and cooled clean water. The eyes should be wiped from the inside to outside edge using a clean cotton wool swab for each eye. Hands should always be washed before and after the procedure. Topical antibiotics are not required, but consideration should be given to possible concomitant chlamydial infection.

If presumed or diagnosed gonococcal or chlamydial infection, the mother and partner should also be treated.

In countries with a low rate of sexually transmitted diseases staphylococcal and Gram-negative organisms are more likely. Staphylococcal infections can be treated with cloxacillin or flucloxacillin 30 mg/kg every 6–8 hours for 5 days.

Chlamydial conjunctivitis

Chlamydia trachomatis is the most common cause of infectious conjunctivitis in the neonate. It typically presents between 5 and 14 days, and symptoms vary from mild conjunctival inflammation to severe inflammation with purulent discharge. ***Confirmation of Chlamydia can be made by culture or rapid antigen detection***, but these are highly specialised procedures that may not be readily available. A ***Giemsa stain of a conjunctival scraping (not the purulent exudate) may show basophilic intracytoplasmic inclusion bodies***. Care must be taken when obtaining the scraping so as not to cause ocular trauma. Treatment with erythromycin ethylsuccinate for three weeks effectively treats this infection and also may eradicate

Table 3.48.6 Erythromycin for *Chlamydia* infection

Drug	Each dose	Dose frequency	Age	Weight	Indication/ administration notes
Erythromycin	IV or oral				
	10 mg/kg	12 hourly	<7 days	Any weight	Infuse over 30–60 minutes. Caution in hyperbilirubinaemia as displaces bilirubin from albumin.
		8 hourly	>7 days	<2 kg	
	15 mg/kg	8 hourly	>7 days	>2 kg	Drug interactions include increased serum levels of digoxin, theophylline and potentially caffiene.

upper respiratory tract colonisation. Topical therapy with sulphonamide drops (10%) or tetracycline ointment (1%) or neomycin drops (two drops) every 6 hours for three weeks may also be effective. Ensure the mother is appropriately referred for treatment.

Pustules

Pustules are most commonly caused by *Staphylococcus aureus*. Most often these occur in small clusters in an otherwise healthy infant. Topical therapy may be all that is needed. Oral therapy with a penicillinase-resistant penicillin (for example flucloxacillin, see Table 3.48.5) or first-generation cephalosporin (for example cephalexin 12.5 mg/kg 6 hourly for 7 days) may also be used. Sometimes staphylococcal pustules can be difficult to distinguish from erythema toxicum, a benign, non-infectious newborn rash. Careful examination of the contents of a pustule by Wright stain reveals numerous eosinophils in erythema toxicum compared to neutrophils and cocci bacteria with *Staphylococcus*.

Umbilical infection

Clean the area with soap and warm water and remove/drain pus and crusts. Dry and paint the area with antiseptic such as gentian violet, povidone-iodine or chlorhexidine.

Cellulitis

This is most commonly caused by streptococci, but *Staphylococcus aureus*, as well as Gram-negative enterics and anaerobes should be considered when infection occurs at sites where there have been breaks in the skin. Parenteral antibiotic therapy with intravenous flucloxacillin (see Table 3.48.5) and intravenous penicillin (see Table 3.48.5) should be given. Omphalitis is characterised by periumbilical erythema and induration, often with purulent discharge from the umbilical stump. Treatment should be directed against both Gram-negative and Gram-positive bacteria, and parenteral antibiotics are required (for example a penicillinase-resistant penicillin and aminoglycoside). Omphalitis may become rapidly progressive with spread to deep tissues and the peritoneum, or along umbilical blood vessels. Clostridia should be considered in the setting of poor maternal immunity or poor aseptic umbilical cord care and if possible give metronidazole.

Scalded skin syndrome

These are rare infections due to staphyloccocal organisms with allergic reaction producing the effect of both serious infection as well as burns.

Superficial candidiasis ("thrush" and "monilial" rash)

Superficial candidiasis of the oral mucosa ("thrush") commonly manifests as white patches which do not easily scrape with a spatula. The nappy area may be affected as well ("monilial" rash): **unlike irritant dermatitis** the erythema extends into skin folds and there may be small raised erythematous lesions. Treat with oral nystatin suspension, 1 ml after feeds (divide it between each cheek with a small syringe). Topical nystatin ointment may be used to treat the skin rash but only in combination with oral nystatin.

Necrotising enterocolitis

This is a very serious condition with a mortality of approximately 20–40%. It usually occurs in premature babies, generally after the first week of life. Prevalence is inversely related to birthweight and gestation, and is more common in ill infants. About 15–20% of affected babies have never been fed. It appears to be less common in babies fed exclusively on human milk.

Presenting features

These include peripheral circulatory failure, abdominal distension (sometimes sufficient to embarrass respiration), bile-stained emesis or gastric aspirates, and blood and mucus in the stools. Features of multisystem failure such as coagulopathy, petechial haemorrhage, oliguria and haematuria may be associated.

Investigations

These must include abdominal *X* rays: look for abnormal gas pattern, free intraperitoneal air – best seen with a left side down (lateral decubitus *X* ray) where free air may be easily seen overlying the dense hepatic tissue – intramural gas (pneumatosis intestinalis) or gas in the portal tracts of the liver. A complete blood count with differential cell count, platelet count, blood culture, serum electrolytes and regular weighing should be obtained. Continuous heart and respiratory rate monitoring should be provided,

Table 3.48.7 Metronidazole in necrotising enterocolitis

Drug	Loading dose	Regular dose	Administration notes
Metronidazole	IV or oral 15 mg/kg	IV or oral 7.5 mg/kg every 12 hours	Infuse over 30 minutes Injection solutions can be given rectally

if available, and frequent blood pressure measurements should be made.

Treatment

- **Stop all enteral feeds** and provide intravenous fluids, typically at 120 ml/kg/day of 10% glucose with added electrolytes. Adjust fluids as indicated based on weight change, urine output and serum electrolyte determinations.
- Place an orogastric tube and place on low-pressure continuous suction, if available, or leave the tube open with intermittent gastric aspiration (every 4 hours). The goal here is to keep the intestines decompressed. The quantity of gastric fluid aspirated is usually of relatively small volume so replacement fluid is seldom required (see Table 3.48.3).
- Start parenteral broad spectrum antibiotics, usually with ampicillin and gentamicin. (see Table 3.48.5) Anaerobic coverage with metronidazole (see Table 3.48.7) may be added, especially if pneumatosis, perforation or evidence of peritonitis are present.
- Treat shock with 0.9% saline or colloid such as 4.5% albumin, 20 ml/kg over 1 hour. Repeat if necessary.
- If bleeding give 1 mg vitamin K IV and fresh frozen plasma 20 ml/kg (if available).
- The principal goal of therapy is bowel rest and antibiotic treatment of any contributing or evolving bacterial infection. The length of this therapy is usually 10–21 days depending on the severity of the process. Serial abdominal *X* ray studies are indicated early in this disease to monitor for pneumatosis intestinalis or perforation. ***Ideally parenteral nutrition should be given at this time***, in place of simple 10% glucose and electrolytes. Enteral feeds (breast milk) are re-introduced slowly at the end of antibiotic therapy (20–30 ml/kg/day) with careful monitoring for abdominal distension or other signs of obstruction.
- **Note**: Even in hospitals with good surgical support, perforation of the bowel is not necessarily an indication for a laparotomy. The conventional surgical approach has been laparotomy with resection of the perforated and adjacent necrotic bowel. A stroma and mucus fistula may be created with later anastomosis. An alternative surgical approach is to place a peritoneal drain with laparotomy reserved for later complications if they develop (for example bowel obstruction from adhesions or bowel wall strictures). Although there is some controversy about which approach is best, studies suggest that the overall mortality may be similar with either approach.

Seizures

Seizures (fits, convulsions) have been reported to affect about 0.1% of term babies and 10% of those <1500 g at birth.

Presenting features

Clinical findings may be subtle (apnoea staring, lip smacking/grimacing, deviation of the eyes, cycling movements of limbs), or more obvious tonic (extensor) posturing or clonic movements may occur. Involvement of a limb or one side of the body only does not necessarily imply a focal cause in the neonate. A bulging anterior fontanelle may suggest intracranial haemorrhage or infection. **Always measure and note the head circumference**. ✔ Sometimes involuntary movements (for example extreme jitteriness) or benign myoclonic jerks can be hard to distinguish from seizures; the presence of associated autonomic instability and/or lateral eye deviations may signal seizure activity whereas the absence of these findings and elimination of the movements when the limbs are restrained indicate the absence of seizures.

Differential diagnosis

- Hypoxic ischaemic encephalopathy (see below). This is the most common cause of seizures in a term baby. Onset is usually within the first 24 hours and almost never commences after the third day.
- Intracranial haemorrhage, subarachnoid haemorrhage or cerebral infarction are also common causes of neonatal seizures. With subarachnoid haemorrhage seizures may or may not be focal, but unilateral tonic-clonic seizures are often observed with cerebral infarction. Although intraventricular haemorrhage occurs most frequently in low-birthweight infants or at gestational ages under 32 weeks, this may also be found in term or near-term infants with neonatal seizures. Always give 1 mg vitamin K IV.
- Infection. Although meningitis is not a common cause of neonatal convulsions it must always be excluded by lumbar puncture and antibiotics commenced pending results of culture (see above and Chapter 4.1).
- Metabolic causes:
  - Hypoglycaemia: check blood glucose.
  - Hypocalcaemia: check plasma calcium and magnesium concentration.

- Hyponatraemia/hypernatraemia. Seizures are uncommon unless plasma sodium is <120 mmol/litre or >160 mmol/litre. Seizures in infants with hypernatraemia may result from cavernous sinus thrombosis. A rapid fall or rise in serum sodium, as may occur with too rapid therapeutic correction, may be more injurious than the absolute value.
- Pyridoxine dependency is an excessively rare metabolic disorder that can present with severe seizures.
- Kernicterus (see jaundice, above).
- ***Other rare inborn errors of metabolism (for example urea cycle defects, non-ketotic hyperglycinaemia) – require measurement of serum amino acids, urine fatty acids, serum lactate and pyruvate, and blood ammonium***.
- Maternal substance abuse, particularly opiate withdrawal.

Investigations

These should include:

- Lumbar puncture and blood culture.
- Blood glucose, calcium, urea and electrolytes. Blood ammonia if available (arterial).
- ***Arterial blood gas to help further assess acid–base status***.
- Cranial ultrasound.
- ***Intracranial imaging (head computed tomography if available)***.
- Baseline and follow-up electroencephalograms may aid in the diagnosis and treatment.
- Save urine, plasma and CSF for metabolic studies if seizures are protracted.

Treatment

- Stop feeds and place an intravenous line.
- Start antibiotic therapy (see Infections above)

Table 3.48.8 Treatment of seizures: anticonvulsants

Drug and preparations	Each dose	Dose and frequency	Administration notes	*Blood levels*/cautions
Phenobarbitone Injection Oral solution	IV, IM or oral	Loading dose 20 mg/kg followed by maintenance 12–24 hours later of 5 mg/kg Once daily generally but with time may need to be given 12 hourly	Slow IV over 30 minutes Loading dose may be repeated but at 10 mg/kg if clinically indicated	***Monitor plasma levels. Therapeutic range 15–30 mg/litre although increasing up to 40 mg/litre should be considered in resistant seizures***
Phenytoin Injection Oral solution	Slow IV over 30 minutes or oral IV or oral	Loading dose of 15–20 mg/kg Single dose – may be repeated if clinically indicated *Then* 12 hours later 1.5 mg/kg 12 hourly increasing to usually 2–4 mg/kg 12 hourly	Give IV over 20–30 minutes Orally poorly absorbed, particularly in premature infants Usual maximum 7.5 mg/kg 12 hourly	***Therapeutic range 5–17 mg/litre. Wide variation in levels so monitor closely and adjust dose and interval accordingly. Measure trough level. May not reach steady state for up to 14 days and dose adjustment can have marked effect on levels so monitor closely***
Paraldehyde Injection rectal solution (olive or arachis oil)	Rectal, IV or IM	0.2 to 0.3 ml/kg loading dose and repeat once at 4–6 hours later	All doses as paraldehyde undiluted. Use injection rectally or ready-diluted rectal solution. Dilution with an equal volume of olive oil. If using a plastic syringe administer immediately.	IM injections may cause sterile abscesses (maximum 1 ml at one site)
Clonazepam Injection	Slow IV	100 micrograms/kg loading dose then 10–30 micrograms/kg/hour as a continuous infusion. Not for >3 days	Up to 200 micrograms/kg/24 hours may be required in first 48 hours Use slow IV injection over 20 minutes	Caution: respiratory depression and increased pulmonary secretions particularly if accumulation occurs. **If not ventilated use lower dose because of respiratory depression** ✔

- Treat hypoglycaemia if present (see Hypoglycaemia, above).
- Monitor heart and respiratory rate, oxygenation (ideally with pulse oximetry) and blood pressure. Treat SaO_2 or cyanosis with oxygen.
- Consider anticonvulsant therapy (Table 3.48.8): the earlier fits appear, the more frequent they are (more than two or three per hour), and the longer they last (more than 3 minutes) the more likely this will be required. Fits which interfere with respiration will need to be treated. Anticonvulsants can be given as follows:
  - **Phenobarbitone (first line)**. Give 20 mg/kg intravenously – seizures have been reported to respond to this dose 40% of the time; an additional 10 mg/kg may be required if seizures persist or recur (70% response rate)
  - **Phenytoin (second line)**. Give 20 mg/kg loading dose by slow infusion and monitor for hypotension and cardiac arrhythmia
  - **Paraldehyde** (see Table 3.48.8)
  - **Clonazepam IV infusion**. 100 micrograms/kg loading dose then 10–30 micrograms/kg/hour as an infusion (see Table 3.48.8) Not for >3 days.
  - **Sodium valproate**: 20 mg/kg orally then 10 mg/kg 12 hourly
  - **Carbamazepine**: 2.5 mg/kg 12 hourly orally. Increase by 2.5 mg/kg every 3–7 days until correct dose achieved (usually 5 mg/kg 8–12 hourly).

✔ **Note: anticonvulsants may precipitate a need for respiratory support**.

Once seizures are controlled, maintenance therapy with a single agent is often possible (usually phenobarbitone). Discontinuation of treatment depends on the underlying aetiology but aim to withdraw anticonvulsants as soon as possible. If available, blood phenytoin and phenobarbitone levels are useful for monitoring therapy.

Hypoxaemic ischaemic encephalopathy

The term "hypoxaemic ischaemic encephalopathy" describes the abnormal neurological state of babies who have suffered significant perinatal asphyxia signified by:

- ***cord blood pH* <*7.0***; low Apgar score (three or less at 5 minutes) despite appropriate resuscitation measures
- Neonatal neurological abnormalities
- Evidence of multiorgan dysfunction such as oliguria, haematuria (signifying acute tubular necrosis), increased transaminase levels (hepatic necrosis), myocardial dysfunction.

Presenting features

Sarnat's clinical grading system (Table 3.48.9) may be used to help guide treatment and give some indication of prognosis.

Treatment

- Treatment is generally supportive, giving close attention to monitoring of gas exchange, blood pressure and fluid balance. Avoid hyponatraemia which may result from inappropriate antidiuretic hormone secretion and excessive intravenous hypotonic solutions. Note that acute renal failure is often present; if so restrict fluids to measured unine output and gut losses plus 15 ml/kg/24 hours for full term and 24 ml/kg/24 hours for preterm infants (to reflect insensible losses) and avoid giving potassium supplements.
- Seizures are treated as described above. Note however that increasing doses of anticonvulsants may precipitate a need for ***mechanical ventilation*** and confound the clinical staging criteria above which apply only to **unsedated** babies.

Prognosis

Good in stage 1, guarded in stage 2 and very poor in stage 3.

About 50% of stage 2 babies will recover without sequelae. Babies in stage 3 will either die or be left severely disabled. A judgement must therefore be made with the family about the implementation or continuation of intensive care in such cases.

Special problems for young infants

Neonates are susceptible to infection and display less specific clinical signs when they are unwell.

Diarrhoea

See Chapter 3.25.

Table 3.48.9 Sarnat's grading of hypoxaemic ischaemic encephalopathy

Sarnat stage:	Mild (stage 1)	Moderate (stage 2)	Severe (stage 3)
Conscious level:	Hyperalert	Lethargic	Stuporose
Muscle tone:	Normal	Hypotonic	Flaccid
Seizures:	Rare	Common	Severe
Feeding:	Sucks weakly	Needs tube feeds	Needs tube feeds ***or IV feeds***
Respiration:	Spontaneous	Spontaneous	Absent

Special points

- Frequent breastfeeding is essential. Encourage more frequent feeds.
- If dehydration give oral rehydration solution **in addition to breast milk** (see Chapter 3.25 for details).
- If bloody diarrhoea, assume dysentery but avoid cotrimoxazole in infants who are jaundiced or premature.
- In the septic and unwell infant, give IV antibiotics as outlined in Table 3.48.5.
- **Beware intussusception** although it is rare under 1 month of age (see Chapter 3.49).

Hypothermia

Can be due to a cold environment, malnutrition or serious infection.

- Use a low-reading thermometer. If core (rectal) is <32°C then severe; between 32 and 35.9°C is moderate. Alternatively axillary temperature <35°C. If temperature does not register on normal thermometer, assume hypothermia.
- Warm the infant by thermostatically controlled heated mattress (37–38°C) or air heated incubator 35–36°C.
- If not available, a cot heated with a **hot-water bottle removed before infant is placed in it** can be effective.
- Cover the head with a warm bonnet and dress the baby in warm DRY clothes. Keep nappy dry.
- Use "kangaroo care": skin-to-skin contact with the mother, between her breasts and covered with a blanket (see above) is probably the most effective method.
- Take care when examining the infant not to allow temperature to fall (ideally room temperature should be >25°C.
- Avoid overheating by regular axillary temperatures.
- Feed 2 hourly. Also feed during the night (4 hourly).
- Avoid washing baby and draughts.
- Baby to sleep with the mother during the night.

Acknowledgements

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3.49 Surgical problems

Devendra Kumar Gupta, Anupam Lall, Manoj Shenoy and Leela Kapila

Indirect inguinal hernia

The protrusion of the abdominal viscous into a peritoneal sac (the processus vaginalis) in the inguinal canal. The contents of the sac are usually intestine but may be omentum, Meckel's diverticulum or ovary and fallopian tube in females.

- 50% seen in the first year of life and mostly before 6 months of age.
- Patent processus vaginalis (not a hernia) is present in 80% of boys at birth, in 40% at 2 years and in 20% of adult men.
- A bulge in the groin, which sometimes extends to the scrotum, appearing when the child cries or strains, but disappears when he relaxes is certainly a hernia. Hernias are seldom symptomatic except when they are very large or are incarcerated or strangulated. On physical examination, cough impulse is the most important sign. A soft bulge that is reducible on digital pressure is also a diagnostic feature. Hernia in neonates may be transilluminant so it is not a very reliable test to differentiate with hydrocoele.

✔ **Needle aspiration is contraindicated** in any inguinal swelling for fear of perforating the intestines.

Differential diagnosis

Should include:

- Lymphadenopathy: firm, immobile, nonreducible, and no cough impulse
- Hydrocoele: can reach the upper pole of the swelling, transilluminant, no cough impulse is present
- Hydrocoele of the cord: separate from testes, nonreducible, no cough impulse, upper limit is reachable
- Undescended testis: scrotum empty and hypoplastic, cough impulse, may be reducible
- Femoral and direct inguinal hernias: rare but should be kept in mind.

Treatment

All inguinal hernias should be promptly repaired unless there is another medical condition that makes the anaesthetic risks prohibitive. Premature infants with hernia should not be discharged without a repair of the hernia, as the chances of incarceration are high. ***An expert paediatric anaesthetist is required, as anaesthesia risks are higher. Postoperative apnoea is common in premature babies and at times may require ventilatory support. If these facilities are not available the baby should be referred to higher centres or the surgery is deferred till the risk of anaesthesia is low.***

- There are reasons to avoid delay, especially in infants.
  - Spontaneous disappearance of inguinal hernia does not occur.
  - Risk of incarceration is greater in infants.
  - Operation is technically more difficult and the risk of injury to the vas and testicular vessels are greater in long-standing and incarcerated hernia.
  - Increasing age does not decrease risk of anaesthesia provided experienced anaesthetist is available.

A herniotomy is performed through an incision in the lowermost transverse inguinal skin crease. The sac is identified and transfixed. Herniorraphy is contraindicated as it disturbs the normal anatomy of the inguinal canal. Bilateral exploration and repair is indicated in patients with bilateral hernias and in infants under the age of 1 year as a contralateral sac is present in 50% of cases. Direct inguinal hernia and femoral hernia requires Cooper's ligament repair.

Incarcerated hernia

Occurs when the intestine becomes stuck at the internal inguinal ring. If prolonged the blood supply may also become compromised causing strangulation.There is a sudden increase in the size of the hernia with severe pain and symptoms of bowel obstruction (vomiting and abdominal distension). On examination a hard, tender, fixed mass in the groin is palpable with increased bowel sounds on auscultation. It may be confused with the torsion of testis, acute inguinal lymphadenitis and tense infected hydrocoele.

Treatment

The treatment includes:

- Adequate sedation and administration of analgesics to calm the baby
- Cold fomentation (reduces the oedema)

- Gentle pressure is applied to reduce the hernial contents (signs of peritonitis are a contraindication)

After reduction of the hernia the child should be admitted to the hospital and checked hourly to be certain that there is no damage to the intestine or testis and to reduce a recurrent incarceration promptly if it occurs.

Herniotomy is performed **after 48 hours** to allow tissue oedema to subside.

Hydrocoele

Accumulation of fluid in the scrotum (communicate via a patent processus vaginalis with the peritoneal cavity). Rarely secondary to epididymo-orchitis, tumor and torsion of testis.

- Usually asymptomatic.
- Testis is not palpable separately, and upper pole of the swelling is reachable, reduces on lying down and is transilluminant (hernia in a neonate may also be transilluminant).
- No cough impulse is present.

Differential diagnosis

Should be differentiated from **inguinal hernia** and underlying pathology like **tumours and torsion of testis** should not be missed. **Spermatocoele** and **varicocoele** are non-transilluminant, have worm-like feeling on palpation and are separate from the testes.

Surgical treatment

This is rarely indicated. More than 90% will spontaneously disappear. **Surgery is indicated when it has not disappeared by the age of 2 years** and those that are larger and symptomatic. Herniotomy, as done for inguinal hernia, is the procedure of choice (high ligation of the sac).

Undescended testis (cryptorchidism)

An undescended testis is one which cannot be made to reach the bottom of the scrotum. It represents the second most common problem in paediatric surgery after indirect inguinal hernia and should be distinguished from the more common **retractile testis**.

The incidence of undescended testis is 2.7–3% at birth in full-term infants decreasing to 1.5% after 1 year of age and thereafter remains the same. It is more common in premature infants approaching 100% at gestational age of 32 weeks or less.

- An **ectopic testis** is one that has strayed from the inguinal canal, usually to the thigh, perineum, and base of penis, femoral and abdominal region.
- An **ascending testis** is one that is in the scrotum at birth, but the spermatic cord fails to elongate at the same rate as body growth and the testis progressively becomes higher in the inguinal canal in childhood.
- An **impalpable testis** is quite uncommon (less than 10%) and **agenesis** is rare (20% of all impalpable testes). A fully descended but grossly hypoplastic testis may be impalpable and only identified by exploration. Normal descent of testes occurs at about the seventh month of fetal life when the gubernaculum swells and shortens, drawing the testis through the inguinal canal into the scrotum. Failure of descent may occur because of hormonal failure (inadequate gonadotrophins and testosterone), a dysgenetic testis or an anatomic abnormality such as abnormal or malplaced gubernaculum, obstruction of inguinal canal or scrotum or short vas or vessels.

Sequelae of non-descent

- The higher temperature of the extrascrotal testis causes testicular dysplasia with interstitial fibrosis and poor development of seminiferous tubules thus hampering spermatogenesis. Testosterone production is unaffected by position, thus a male with bilateral undescended testes will develop secondary sexual characteristics but will be sterile.
- Due to dysplasia, higher chances of malignancy are present (10–20 times). The risk of malignant degeneration is not altered by orchidopexy. Malignancy usually develops in the second or third decade of life.
- A testis in the inguinal region is more prone to direct trauma and torsion.

Examination

On examination, always look for a hernia. The position and size of the testis should be noted. If impalpable, ectopic locations of the testis should be examined.

Treatment

The histological changes in the testes occur as early as 6 months of postnatal life and therefore a child who has an undescended testis should be operated at the earliest time possible to prevent them.

The best time for orchidopexy is about 1 year of age, but where surgical expertise is available, it should be done before 9 months of age. The hernial sac should be dissected from the cord structures and a high ligation done.

- The testis is placed in an extra-dartos pouch in the scrotum; an adequate dissection should be done to avoid tension on the pedicle while placing the testis in the scrotum. Any torsion of the pedicle should be avoided. Retroperitoneal dissection and careful snipping off of lateral peritoneal bands will give an adequate length to the cord.
- In cases of impalpable testis, in about 50% a useful testis can be brought down and in the other 50% there is either no testis present (testicular agenesis or intrauterine torsion-vanishing testis) or a useless and potentially neoplastic testis is removed.
- For an abdominal testis, laparoscopy is useful to identify and confirm the position of the testis and

simultaneously permit the ligation of spermatic vessels (Fowler-Stephen's stage I operation).
- Later the testis can be brought into the scrotum by inguinal exploration, testicular blood supply being supported by artery to the vas. ***Microvascular transfer of testis is the best procedure to avoid atrophy of the testis, but it needs great surgical expertise and equipment***.
- For psychological reasons, if orchidectomy has been done, prosthetic replacement should be performed.
- In bilateral undescended testes, especially with hypospadias, an intersex disorder should be suspected and the child should be further investigated.

Prognosis

There is 2% recurrence, 2–5% incidence of atrophy, 70–80% fertility after unilateral orchidopexy and 40% fertility after bilateral orchidopexy.

Phimosis

Phimosis is defined as the excessive tightness of the foreskin, preventing retraction behind the glans. It occurs in 1–2% males. The foreskin normally cannot be retracted in infants and non-retractability of the foreskin is not pathologic until the age of 3 years. Forced retraction may cause phimosis by producing tears in the foreskin, which heals with scarring and contraction. If there is pooling of urine and repeated attacks of balanoposthitis, then simple dilatation of the foreskin can be done and the mother advised regarding local hygiene.

After the age of 3 years, the foreskin becomes naturally retractile. The mother has to be explained that daily retraction and cleansing of the glans will prevent recurrence of the phimosis. At the same time it is of utmost importance to emphasise the importance of retracting the prepuce from the glans to avoid paraphimosis, which is an inability to bring the foreskin in its natural position because it is trapped in the sulcus at the base of the glans.

Circumcision is only indicated where the prepucial skin is scarred and fibrotic due to balanitis xerotica obliterans.

Hypospadias

A condition where the urethra opens on the ventral aspect of the penis at a point proximal to the normal site. When opening on the dorsal aspect, termed "epispadias", there is usually associated extrophy of the bladder.
- Hypospadias is one of the commonest congenital anomalies of the male genitalia occurring in 1 in 300 male births. There are various degrees of severity depending on how far back the urethral meatus lies. It may be associated with undescended testes and in severe cases there is a possibility of an intersex problem.
- Ventral curvature of the shaft of penis is called a "chordee"; it is due to fibrosis of the urethral plate, shortened skin or fibrosis of the corpora cavernosa. The prepuce is deficient ventrally and an unsightly dorsal hood of redundant skin is present.
- Congenital short urethra is a deformity where there is ventral curvature of the shaft of penis without hypospadias.

The disabilities of hypospadias are cosmesis of the penis, a stream that is deflected downwards or splashes and in severe hypospadias, children have to void in a sitting position (as in females). Uncorrected chordee interferes with intercourse and there is infertility in severe hypospadias (penoscrotal and perineal) as semen is not directed into the vagina.

Treatment

The dictum is that every boy has a birthright to void standing and this should be corrected before school-going age so that the child does not feel ostracised in society.

In severe cases of hypospadias, intersex disorders and associated urological abnormalities such as pelvic-ureteric-junction obstruction or renal agenesis should be ruled out.

Principles of surgery

- Correction of chordee to straighten the penis (orthoplasty).
- Movement of the urinary meatus to its normal position on the tip of the penis (urethroplasty).
- Correction of the deformity of the glans to give it a conical shape (glansplasty).

No infant with hypospadias should be circumcised, as the prepuce is essential for the repair. It can be undertaken as a one-time or staged procedure. It depends on the degree of chordee and the severity of the hypospadias. ✔

Bladder stones

In disadvantaged countries, bladder stones are quite common due to the prevalence of malnutrition. The stones are composed of ammonium acid urate and oxalate and are seen in lower socio-economic groups. Such stones are usually related to a high dietary intake of rice or wheat and low intake of milk and animal protein (see Chapter 3.7).
- Children present with increased frequency of urine and strangury or haematuria (the child usually holds the penis and rubs it with the finger and cries during micturition).
- Children may present with an episode of retention of urine if the bladder stone becomes impacted at the bladder neck or in the urethra.
- On per-rectal examination, a stone may be palpable on bimanual palpation.
- A plain abdominal *X* ray may reveal calcified stones. Abdominal ultrasonography will detect non-calcified stones.

Treatment

Open stone surgery is the modality of choice ***if expensive equipment for cystolithopaxy is not available***.

If there is no infection, two-layered closure of the bladder is sufficient, requiring no catheter or suprapubic drainage.

Once the stones are removed, recurrence is rare.

Cervical swellings

The neck is one of the commonest sites of cystic and solid swellings during childhood. Lesions are either developmental anomalies arising from the remnants of branchial arches, thyroglossal tract, the jugular lymphatics or the skin or acquired as in disease of the salivary gland, lymph nodes or the thyroid gland.

- Lymphangiomas (cystic hygroma)
- Branchial cysts/fistulae
- Thyroglossal cyst
- Ectopic thyroid/ thyroid swellings
- Epidermal cyst
- Swelling of salivary glands
- Haemangiomas
- Lymph-node swellings

Lymphadenopathy

Enlargement of lymph nodes may result from acute or chronic infection and from primary or secondary neoplasia.

- Infection is the commonest cause of lymph node enlargement in childhood.
- Tuberculosis is the most important pathogen in poor countries.
- In many cases lymph nodes are reacting to an upper respiratory tract or an ear infection. This is known as "non-specific reactive hyperplasia".
- Primary tumours of the lymph nodes include lymphoma and leukaemia.

  A lymph node enlargement of more than 1 cm in size is significant and a persistent node >3 cm in size requires a surgical biopsy.

A careful history regarding repeated attacks of upper respiratory tract infection, boils on the scalp or drainage area, and ear discharge, should be taken. A positive family history of tuberculosis is an important feature of tubercular lymphadenitis. A history of the pattern of fever, loss of weight and appetite, and the presence of night sweats are important features in making a differential diagnosis.

On careful physical examination, all sites of lymph nodes (cervical, axillary, inguinal and abdominal) should be examined. The size, number, consistency, tenderness, and presence or absence of fluctuations should be noted. On abdominal examination liver, spleen and mesenteric lymph nodes should be palpated. The drainage area of the lymph nodes should be examined for boils, furuncles, injury or neoplastic swelling. Tonsils should be inspected for enlargement and suppuration.

- In **acute lymphadenitis**, the affected nodes are enlarged, painful and tender, restricting movement of local areas of the body. Fever and leukocytosis are common. Untreated infections may resolve spontaneously, progress to suppuration and abscess formation, or become chronic.
- In **tubercular lymphadenitis**, lymph nodes are enlarged painlessly, become matted together and fixed to adjacent structures. Caseation leads to the formation of "cold" abscesses, which lack the local and systemic signs of acute inflammation (fever, tenderness and erythema). When a cold abscess ruptures through the deep fascia (a "collar stud abscess") the skin becomes red and thin, takes on a blue tinge and then gives way to establish an indolent tubercular sinus. On aspiration, straw-coloured fluid is present, compared to the thick pus that is usually present in an acute abscess. Confirmation depends on culture of the organisms or visualisation of acid-fast bacilli on microscopy.
- In **primary neoplasia**, the nodes are painless, rubbery in consistency and discrete. Liver and spleen enlargement may or may not be present. Systemic features of low-grade fever, night sweats, or loss of weight and appetite points towards the diagnosis.
- **Secondary enlargement of the lymph nodes** due to neoplasia is rare in childhood.
- **Primary cancers** are soft-tissue sarcomas, mainly rhabdomyosarcoma, germ cell tumours and neuroblastomas. The nodes are large, firm to hard in consistency and fixed to underlying structures.

Investigations

- Blood tests
  The erythrocyte-sedimentation rate is usually raised in chronic infection and neoplasms. Leukocytosis is seen in acute lymphadenitis and abscess formation.
- Mantoux test
  To diagnose tuberculosis (start with 1 in 10 000 and then 1 in 1000). A strongly positive test is a pointer towards the diagnosis; if the test is negative, it does not rule out the disease (especially in the presence of HIV infection).
- *X* ray chest
  To identify there is the pulmonary lesion of primary complex or the hilar lymphadenopathy seen in cases of tuberculosis. Mediastinal widening is seen in patients with lymphomas.
- FNAC (fine-needle aspiration cytology) of lymph nodes is helpful in cases of persistent lymph nodes, which do not reduce in size after a week's course of antibiotics. Lymphomas cannot be diagnosed on FNAC and a surgical biopsy is mandatory.

Treatment

Acute lymphadenitis

- Antibiotics are prescribed. Penicillin is usually appropriate as most infections occur outside the hospital settings. Oral or intravenous preparations may be used. If improvement has not occurred within 48 hours, a broad-spectrum antibiotic such as an oral/intravenous cephalosporin may be started.
- Anti-inflammatory medication (to reduce the pain and swelling).

- Hot fomentation (relieves pain and reduces the swelling).

Fluctuation, or other local signs of abscess formation, indicate the need for **incision and drainage of pus**, best performed under general anaesthesia. All the loculi are broken and necrotic material is curetted out. A sample should be sent for microscopy (including Zeihl–Neelsen staining), culture and sensitivity, and appropriate antibiotics prescribed. The precipitating cause of acute lymphadenitis should also be treated.

Tubercular lymphadenitis

- Anti-tubercular treatment leads to resolution (full course of 9 months should be undertaken with four drugs for two months and two drugs for the next 7 months; see Chapter 4.10).
- Cold abscesses require drainage or aspiration which, even if successful, will require a drain otherwise a sinus will form. Aspiration in a non-dependent position will help to avoid formation of a sinus.

Lymphomas

After diagnosis, further investigations will be required to stage the disease and its treatment (see Chapter 3.33).

Cystic hygroma

A hamartoma of the jugular lymph sac which presents in infancy and is more common in boys than girls. It produces a major neck swelling and is diagnosed by inspection. The swelling is usually found as a unilateral, fluctuant, transilluminant swelling centred on the carotid triangle. The cysts are of varying sizes and contain clear fluid. A haemangiomatous element may be present in the swelling, giving it a reddish tinge instead of light blue colour. Cysts may enlarge suddenly due to viral or bacterial infection or haemorrhage. If the cyst compresses airways and vessels, it may cause stridor, respiratory distress and superior vena caval syndrome. Surgical excision is difficult and complete removal is mandatory to prevent recurrence. Non-surgical procedures include aspiration of the swelling and injection of agents such as hypertonic saline.

Branchial cysts, sinuses and fistulae

These are uncommon in childhood. They usually arise from the second cleft and emerge from beneath the anterior border of the sternomastoid in the upper third of the neck. The fluid contents are milky and contain cholesterol crystals. They may become infected, therefore require excision as soon as they are diagnosed.

Sinuses and fistulae most commonly arise from the second branchial cleft, occasionally from the first and third. They present as a small discharging sinus on the skin overlying the lower end of the sternomastoid muscle. They usually present in early childhood and sinuses may present at any time during childhood and sometimes may be complicated by infection and abscess formation. The treatment of both is surgical excision of the whole tract up to the pyriform fossa to prevent recurrence. Methylene blue is injected or a nylon thread guided in the fistula to delineate it during surgical dissection for appropriate excision.

Thyroglossal cyst

The descent of the thyroid gland from the floor of the fetal mouth leaves a tract from the foramen caecum of the tongue to the thyroid isthmus. A cyst lined by respiratory epithelium may arise anywhere along the tract, but is usually subhyoid (75%). The swelling is in the midline and moves with swallowing and also with protrusion of the tongue. An infected cyst may be mistaken for acute bacterial lymphadenitis, or an ectopic thyroid may cause a similar swelling.

The thyroglossal cyst and the entire tract along with the central portion of hyoid bone should be excised to minimise the risk of recurrence (Sistrunk's operation).

Ectopic thyroid

An ectopic thyroid should be suspected on surgical dissection if the cyst appears to be soft and solid. ***A preoperative isotope scan can identify this if available***. If an ectopic thyroid is encountered, the thyroid is divided in the midline rotated on its vascular pedicles and placed behind the strap muscles.

Epidermoid cyst

Inclusion dermoid cysts arise from ectodermal cells detached during fetal growth. They are often in the midline or along lines of embryonic fusion. They contain sebaceous cheesy material surrounded by squamous epithelium. They enlarge slowly and should be removed completely; the capsule should not be breached to prevent recurrence.

Haemangiomas

They are the most common tumours of infancy and the most common congenital anomalies. They are present in about 1–3% of all newborn infants. This increases to 10% by 1 year of age. Haemangiomas can be capillary or cavernous.

- The natural history of **capillary haemangiomas** is that they initially present shortly after birth as a pale, pink or bright red spot or patch on the skin. There is subsequently a rapid growth in infancy for 3–6 months, followed by a static phase. At 18–24 months

the lesion starts involuting. 50% will involute by 5 years and 90% by 7 years. Rarely the lesion persists and requires excision.
- **A cavernous haemangioma** has a deeper component in subcutaneous tissues or muscles and is less likely to regress completely.

Management

Management of these lesions consists of an accurate diagnosis and careful observation. Parents need reassurance when the lesion is growing rapidly. Problems of ulceration, bleeding and rarely infection occur secondary to minor trauma. These are best treated non-operatively.
- Surgical excision is indicated when there is functional or gross cosmetic disability (for example a haemangioma on the eyelid).
- Steroids may be used to induce involution in large haemangiomas (prednisolone 1–2 mg/kg/day for 2–4 weeks; the dose is tapered off before stopping the therapy). Intra-lesional steroids can be used to induce regression in the size of haemangiomas in and around the eye. ***Lasers have also been used to regress the lesion***.

Obstructive jaundice in infancy

This is most commonly caused by extrahepatic biliary atresia, choledochal cyst or inspissated bile syndrome.
- The most difficult differential diagnosis is neonatal hepatitis.
- If jaundice in the newborn persists, the stools are never yellow, and the urine brown, then a conjugated bilirubin level should be measured and urobilinogen and bilirubin looked for in the urine.

Empyema thoracis (see Chapter 3.1)

This is defined as an accumulation of pus in the pleural space. In most children, this results from an infected pleural effusion associated with ongoing uncontrolled pulmonary sepsis or pneumonia. An infection of the pleural space is unlikely when there is a healthy underlying lung that is completely expanded. Empyemas/effusions may be diffuse and involve the entire pleural space or they may be intralobar, diaphragmatic or paramediastinal.

Before the advent of antibiotic therapy, *Pneumococcous* and *Streptococcus* were the organisms most frequently associated. Presently *Staphylococcus aureus* is the most common organism. In poor countries *Mycobacterium tuberculosis* is an important cause.

Other reasons for empyema include extension of lung abscess, trauma and extension of subphrenic abscess.

An empyema usually presents with pleuritic chest pain and a heavy sensation on the involved side.

Children are febrile, tachypnoeic, tachycardic and may have a cough that is productive (purulent sputum).

Examination

On examination there is reduced respiratory excursion, pain and dullness on percussion. A friction rub or distant to absent breath sounds may be heard on auscultation of the involved side.
- Chest *X* rays in the anteroposterior and lateral views are necessary for the accurate localisation of the empyema. The underlying lung may show consolidation or evidence of infection by tubercular organisms. There may be evidence of mediastinal shift to the opposite side. The presence of pneumatocoeles indicates staphylococcal infection.
- An ultrasound scan may help distinguish fluid from consolidation in a patient with complete opacification. It is also helpful in localising a loculated empyema that may be drained. It is essential to evaluate the condition of the underlying lung to decide for decortication or pneumonectomy.
- ***A computed tomography scan of the chest is ideal but often unavailable***.

Management

Treatment depends on the cause, whether it is acute or chronic, the state of the underlying lung, the presence of a bronchopleural fistula, an ability to obliterate the space and the patient's clinical condition and nutritional status.
- Fluid from the effusion should be aspirated (thoracocentesis) under ultrasound control after local anaesthesia (see Chapter 3.1).
- If the fluid is serosanguinous, thoracocentesis and appropriate intravenous antibiotics (benzylpenicillin and flucloxacillin) till the temperature settles (change according to sensitivity) and then orally for a total period of 6 weeks, can be a definitive treatment.
- If the fluid is purulent and thick, then a tube thoracostomy is indicated. An intercostal tube should be placed in a dependent position to encourage the pleural space to drain completely. Simultaneously, physiotherapy should be instituted to expand the lung and obliterate the space (see Chapter 6.12 for placement of chest drains).
- If loculated and undrained pockets are present and the lung is not expanding on tube thoracostomy, an open surgical procedure (decortication) will be required.
- If the underlying lung is badly damaged, and will not expand on vigorous physiotherapy, then pneumonectomy is indicated and de-cortication may not be effective. In pneumonectomy, the space can be obliterated by paralysis of the diaphragm (removing a segment of phrenic nerve) or muscle transposition and thoracoplasty may be tried.
- In tubercular empyema, a 6–8-week course of antitubercular treatment (see Chapter 4.10) is essential for optimum results. Surgical therapy should be withheld, except for emergency drainage, until the tubercular disease in the lung has regressed or stabilised, as shown on the serial chest *X* rays.

Urinary tract infection (UTI) due to surgical causes

Recurrent UTI needs investigation to exclude a number of structural and functional abnormalities:

- Vesicoureteric reflux (see Chapter 3.8)
- Posterior urethral valves
- Neurogenic bladder (see Chapter 3.46)
- Urethral strictures
- Bladder stones (see Chapter 3.7)
- Diverticulum of the bladder and urethra
- Voiding dysfunction.

Umbilical pathology

Umbilical hernia

- A defect in the umbilical ring, which generally closes at birth leading to protrusion of a loop of bowel or omentum through it. Some degree of herniation is seen in 20% of newborn babies, still more in premature babies or when there is any increase in intra-abdominal pressure (ascites, etc.).
- Swelling appears on crying and straining, reduces when child is calm.
- Can be reduced with an audible gurgle.

Most close spontaneously in the first 12 months of life but may take up to 3 years. Strangulation and incarceration are virtually unknown, therefore it is safe to wait. **Strapping with coin application is contradicated**: it leads to maceration of skin and infection without any real advantage of inducing closure so is contraindicated.

Surgical indications are a large hernia not closed by 3 years of age or an incarceration.

Umbilical discharge

- **Purulent discharge** is seen in umbilical sepsis. **Neonatal tetanus** is a serious condition in which mortality is very high (see Chapter 4.9) (cow dung application, as practised in rural India, is one cause). Portal thrombosis may occur secondary to it and manifest later as portal hypertension. Appropriate antibiotics should be instituted at the earliest possible stage (benzylpenicillin) and local hygiene maintained.
- **Mucus/serous discharge** is seen in umbilical polyps and **granulomas**. Silver nitrate application will enable these to epithelialise. If these persist, excision will be required. Umbilical fistula may be present and require exploration and excision.
- **Urinary discharge** is seen with a patent urachus in association with a lower urinary tract obstruction. Surgical treatment involves excision of the urachal remnant after investigation and relief of any underlying outlet obstruction.
- **Faecal discharge** is seen with a patent vitello-intestinal duct. This is a persistence of the connection between the yolk sac and the midgut which normally disappears at about the sixth week of gestation. All remnants are to be excised, which may necessitate a laparotomy to search for any discontinuous segments of the tract.

Appendicitis

Appendicitis is the most common abdominal surgical emergency. Although diagnosis and treatment have improved, **appendicitis continues to cause significant morbidity and still remains, although rarely, a cause of death**. ✔

Appendicitis results from luminal obstruction following infection or impaction by a faecolith. Inflammation of the appendix does not inevitably lead to perforation as spontaneous resolution may occur.

Clinical presentation

- Appendicitis presentation is very variable.
- Pain is invariably present and nearly always the first symptom. Early visceral pain is non-specific in the epigastric or umbilical region and **only later does pain** becomes localised over the appendix, most typically at McBurney's point. Pain with a pelvic appendix is often delayed in onset because the inflammed appendix does not contact the peritoneum till rupture occurs and infection spreads. Pain of a retrocaecal appendix may be in the flank or back.
- Anorexia, nausea and vomiting typically follow the onset of pain within a few hours.
- Diarrhoea occurs more frequently in children than adults and can result in misdiagnosis. It may indicate a pelvic abscess.
- The child with acute appendicitis lies in bed with minimal movement. There may be fever and tachycardia.
- Patients may be asymptomatic before perforation and symptoms may be present for longer than 48 hours without perforation. In general, however the longer the duration of symptoms, the greater the risk of perforation.

Examination

Auscultation of the chest to rule out a lower respiratory tract infection is essential.

The single most important aspect of evaluation is serial examination done by the same person. This decreases the number of unnecessary operations.

Investigation

There may be an increase in white blood cell count but this is unreliable.

Ultrasonography is an effective diagnostic aid with a sensitivity of about 85% and a specificity of about 90%. Demonstration of a non-compressible appendix that is 7 mm or larger in anteroposterior diameter is the primary criterion.

Management

- The initial management involves intravenous fluids and adequate analgesia.
- Adequate resuscitation (see Chapter 3.6) must be performed in a patient who presents with peritonitis before surgery is undertaken.
- For early non-ruptured appendicitis, perioperative antibiotics (cefuroxime and metronidazole) are given.
- For perforated appendicitis after appendicectomy, saline irrigation of the peritoneal cavity with the patient in the head-high position is advisable in an attempt to remove as much infected material as possible. Intravenous antibiotics should be given for at least 5 days.
- Cefuroxime (50 mg/kg 8–12 hourly) plus metronidazole (7.5 mg/kg 8 hourly IV over 20 minutes) or
- Ampicillin IV (25–50 mg/kg 8 hourly: max. 4 g/day), gentamicin (2.5 mg/kg 8 hourly) and metronidazole (7.5 mg/kg 8 hourly). (See page 394 for gentamicin doses.)
- If the initial presentation is with an appendicular mass, conservative treatment with IV antibiotics is carried out till symptoms subside with a plan for an interval appendicectomy.

Complications

Complications following appendicectomy are wound infection, abscess formation (local, subphrenic or pelvic) and paralytic ileus. A late complication may be an adhesive bowel obstruction.

Pyloric stenosis

A classical cause of gastric outlet obstruction in infants with a prevalence rate of about 1.5 to 4 in 1000 live births among whites but is less common in Africans and Asians. It is more common in males than females with a ratio of between 2:1 and 5:1. There appears to be an increased risk to first-born infants with a positive family history and certain ABO types.

Cause

No definite cause has been elucidated. Pathologically there is marked muscle hypertrophy primarily involving the circular layer, which produces partial or complete luminal obstruction.

Presentation

Typically presents between 2–8 weeks of age with a peak occurrence at 3–5 weeks. The **vomiting is projectile and non-bilious**. Occasionally, there is coffee-ground vomiting due to gastritis or oesophagitis. The child remains hungry after vomiting and is otherwise not ill looking or febrile. 2–5% of infants have jaundice associated with indirect hyperbilirubinaemia. Non-bilious projectile vomiting, visible gastric peristalsis in the left upper abdomen, and in those presenting late a hypochloraemic hypokalaemic metabolic alkalosis are the cardinal features of pyloric stenosis.

Diagnosis

A definite diagnosis can be made in 75% of infants with pyloric stenosis by careful physical examination of the upper abdomen: **an essential prerequisite is a calm and cooperative child, warm environment, good light and patience**. With the patient in the supine position, in the mother's left arm and sucking on the left breast, and the surgeon sitting on the left side of the patient, the left hand is used to feel the classically described "olive" to the right of the rectus muscle. Visible gastric peristalsis is often noticed.

Investigations

- Ultrasonography is the most commonly used imaging technique for diagnosis. A positive study is a pyloric canal length of 16 mm or more and a pyloric muscle thickness of 4 mm or more. A diameter of more than 14 mm is also considered abnormal.
- Blood investigations in an advanced situation may show the typical hypochloraemic, hypokalaemic metabolic alkalosis.

Management

- It is most important to prepare the patient appropriately and adequately for anaesthesia and surgery.
- Intravenous fluid resuscitation with 5% glucose in 0.45% saline with 20–40 mEq/litre of potassium chloride is the optimal fluid.
- Urine output and serum electrolytes should be monitored.
- The stomach should be aspirated before the operation.
- Ramsted's pyloromyotomy performed through a right upper quadrant or supraumbilical incision is curative and associated with a low morbidity.
- The majority of the patients can be started on feeds about 6 hours after surgery.
- Those who present with haematemesis from gastritis may benefit by delayed feedings for an additional 6–12 hours after surgery.
- Vomiting in the early postoperative period is thought to be secondary to discordant gastric peristalsis or atony.

Intussusception

The telescoping of a portion of the intestine into the lumen of an immediately adjoining part. Typically it occurs in a well-nourished child 4–12 months of age. Male to female ratio is 3:2 and it is more common in Caucasians.

The pathogenesis of intussusception is unclear. It usually originates in the ileum close to the ileocaecal junction and proceeds into the ascending colon. In 2–8% there is a specific lead point such as a Meckel's diverticulum, polyp

or a duplication cyst. Adenoviral infection resulting in lymphoid hyperplasia may act as a lead point.

Clinical presentation

- The infant is suddenly disturbed by what seems to be violent abdominal pain. The pain is colicky, intermittent and severe; with spasms the infant draws up the knees to the abdomen, screams, becomes pale and may sweat and vomiting occurs soon after. The infant may pass a normal stool, seems to recover immediately and may resume normal eating habits, until stricken by another bout of colicky abdominal pain. The vomiting is initially reflex but with a delayed diagnosis becomes secondary to intestinal obstruction and is often bile-stained.
- Classically, infants pass stool that resembles redcurrant jelly.
- **The triad of pain, vomiting and blood per rectum is present in only one-third of patients**: 1 in 10 infants with intussusception will have diarrhoea before signs and symptoms attributable to intussusception become obvious; this is often a cause for delay in diagnosis.
- Pallor, persistent apathy and dehydration are common signs.
- Abdominal examination shows emptiness in the right lower quadrant and a **sausage-shaped mass in the right hypochondrium** extending along the line of the transverse colon. **The mass is not always easy to palpate and its absence does not rule out an intussusception**.
- Fever and leukocytosis are common and tachycardia results from episodes of colic and hypovolaemia from dehydration.

Investigation

- Abdominal *X* ray may show a soft-tissue mass across the central abdomen with dilated loops of bowel.
- **Ultrasonography has become the standard non-invasive diagnostic test and is very reliable. Doughnut (target or concentric ring) and pseudokidney sign suggest a diagnosis of intussusception**.

Management

The most important aspect of treatment is adequate resuscitation prior to intervention. This involves establishing reliable intravenous access, collecting blood for baseline investigations and for cross-match, passing a nasogastric tube for decompression, intravenous fluids and analgesia. Some patients may require one or more boluses of 10–20 ml/kg of albumin/normal saline when first seen.

Broad-spectrum intravenous antibiotics such as a combination of cefuroxime (25–50 mg/kg 8 hourly, depending on the degree of infection) and metronidazole (7.5 mg/kg 8 hourly IV over 20 minutes) are started and the urine output monitored.

Management is initially non-surgical, i.e. with the use of air enema. Sedation should be used for the procedure.

- **A surgeon should be present in the *X* ray department when the radiologist attempts reduction and if perforation occurs surgery should be performed immediately**. ✔
- **An absolute contraindication to rectal reduction is evidence of peritonitis, indicating the presence of a gangrenous intestine**.

If hydrostatic reduction fails and if the patient is stable, a repeat reduction may be attempted. Once the intussusception reduces, the child should be observed overnight with careful monitoring of the fluid and electrolytes.

If reduction fails, the child is taken for surgery where by gentle manipulation (pushing and not pulling) the intussusception can be reduced. The appendix may be removed, recorded and parents informed. If a pathological lead point is found, a resection anastomosis is performed. If the bowel is not viable, it is resected and a primary anastomosis performed. Feeds are started the day after the operation and increased gradually.

Intravenous antibiotics should be given for at least 48 hours and longer (7 days) if peritonitis is present.

The interval between the onset of symptoms and institution of treatment is of paramount importance and mortality can be reduced if the condition is recognised and treated early. ✔

Intestinal obstruction

This is the most common condition requiring emergency surgery in infants and children. Most causes result from complications of congenital anomalies or from inflammatory conditions that affect the bowel.

Causes

- Extrinsic causes: incarcerated hernia and vascular bands, intussusception, anomalies of rotation (volvulus and Ladd's bands, paraduodenal and paracaecal hernias), postoperative adhesions.
- Intrinsic causes: inspissation of bowel contents (meconium ileus, distal intestinal obstruction syndrome in patients with cystic fibrosis), roundworm obstruction.
- Peristaltic dysfunction: Hirschsprung's disease.
- Inflammatory lesions: tuberculosis, Crohn's disease.

Symptoms and signs

Patients present with cramping abdominal pain with anorexia, nausea and vomiting which progresses to become bile-stained. Abdominal distension occurs with the degree being directly related to the site of obstruction in the gastrointestinal tract; the distension being greater the more distal the obstruction.

On examination, the patient may have tachycardia and signs of dehydration. Tenderness and hyperactive bowel sounds are present on abdominal examination.

Chest and abdominal films are taken to confirm the diagnosis of obstruction and rule out the presence of free air.

Treatment

- The goal of treatment is to relieve obstruction before ischaemic bowel injury occurs.
- Intravenous access is established and blood collected for baseline investigations including a full blood count, urea, creatinine and electrolytes and cross-match.
- Intravenous fluids (0.9% saline with 10% glucose) are started using the guidelines of 4 ml/kg/hour for the first 10 kg, 2 ml/kg/hour for the next 10 kg and 1 ml/kg/hour for the next 10 kg. Potassium is added to the fluids, once a good urine output is established (2–3 mmol/kg/24 hours).

For example:

A child weighing 22 kg would need 40+20+2= 62 ml/hour.

- Some patients may need one or more intravenous boluses (10–20 ml/kg) with 0.9% saline/4.5% albumin at the start of resuscitation.
- A nasogastric tube is passed for decompression.
- Broad-spectrum intravenous antibiotics such as:
  - Cefuroxime 50 mg/kg 8 hourly or 12 hourly in the neonate and metronidazole 7.5 mg/kg 8 hourly IV
  - Benzylpenicillin 50 mg/kg 6 hourly, gentamicin 2.5 mg/kg 8 hourly and metronidazole 7.5 mg/kg 8 hourly. (See page 394 for gentamicin doses.)
- Once the patient is adequately resuscitated and fluid and electrolyte imbalances corrected, laparotomy is performed and the cause treated.
- At all times adequate analgesia should be given (see Chapter 1.27).

Hirschsprung's disease

This is characterised by an absence of ganglion cells in the affected intestine. The incidence is about 1 in 4400–7000 live births; the male to female ratio is about 4:1 and in long segment disease it approaches 1:1. The longer the segment of aganglionosis, the higher the familial incidence.

Associated conditions

The associated conditions include Down's syndrome (4–16%), Waardenburg syndrome, multiple endocrine neoplasia 2A and Von Recklinghausen's disease. A higher incidence of enterocolitis has been noted in patients with Hirschsprung's disease and Down's syndrome.

Presentation

The usual presentation is with a delayed passage of meconium beyond 48 hours after birth; 95% of full-term infants pass meconium within 24 hours after birth and the remainder within 48 hours. The child then has episodes of constipation, abdominal distension, vomiting and poor feeding and fails to thrive. He/she may also present with a history of constipation with explosive diarrhoea, the latter indicating the development of enterocolitis.

Differential diagnosis

Hirschsprung's disease should be considered in the differential diagnosis of any child who has constipation dating back to the newborn period.

Examination

On examination the child has a distended abdomen and after a rectal examination there is often the explosive passage of flatus and faeces.

- A plain *X* ray of the abdomen may show dilated bowel loops with paucity of air in the location of the rectum. Barium enema may show the characteristic coning.
- Rectal biopsy remains the gold standard for diagnosis. It should be performed at least 2 cm above the anal valves as the normal anus has a paucity/absence of ganglion cells at the level of the internal sphincter. Though suction rectal biopsy with acetylcholinesterase staining has become the accepted standard for diagnosis in most centres, a full-thickness rectal biopsy under general anaesthesia is equally useful where such facilities are not available.

Treatment

Enterocolitis remains the major cause of morbidity and has a mortality rate of about 6–30%. It manifests clinically as explosive diarrhoea, abdominal distension and fever. The pathophysiology is not fully understood. The diagnosis is made on clinical grounds and treatment is conservative with intravenous fluids and rectal washouts to decompress the colon.

Surgery

The surgical treatment of Hirschsprung's disease has evolved from a three-stage procedure (initial colostomy with multiple seromuscular biopsies, pull-through of the ganglionic colon as the second stage and closure of colostomy as the third stage) through a two-stage procedure (colostomy at the transition zone initially and pull-through at a second stage) to a one-stage procedure without a colostomy. The essential prerequisite of a primary pull-through is adequate preparation with colonic washouts.

Perforative peritonitis

The causes of perforation include amoebiasis, typhoid, tuberculosis, roundworm perforation, and Hirschsprung's disease (see relevant Chapters: 4.33, 4.10, 4.11, 4.28).

Management starts with an adequate history, clinical examination followed by chest and abdominal *X* rays. Adequate resuscitation should be carried out as outlined in the section on intestinal obstruction. After this a laparotomy is performed and the cause treated.

Further reading

O'Neill JA, Rowe MI, Grosfeld JL, Fonkalsrud EW, Coran AG. *Paediatric Surgery*, 5th edn. St. Louis, Missouri: Mosby Year-Book, 1998.

Hutson JM, Beasley SW, Woodward AA. *Jones' Clinical Paediatric Surgery. Diagnosis and Management*, 4th ed. Carlton: Blackwell Scientific Publications, 1999.

Spitz L, Coran AG. *Rob & Smith's Operative Surgery. Pediatric Surgery*, 5th edn. London: Chapman & Hall, 1995.

Section 4

Infectious diseases and individual infections

How to use this book

This is a comprehensive text for all paediatricians caring for children in hospital. It can be used by those with limited resources and also where greater resources are available. We have identified the different levels of care in the following ways:

- **Minimum standards requirements** are given in a highlighted box at the the beginning of each clinical chapter.
- ***A standard of care*** where resources are not limited appears as bold, italicised text.
- **Key points** of particular importance in management of children are identified by a tick in the margin and bold text.

In this way we hope the book will act as a user-friendly, speedy reference on any paediatric ward.

Reducing suffering in the treatment of infectious disease in children

A recent controlled study in children and a series of meta-analyses have suggested that once daily gentamicin is as effective as and no more toxic than three times a day regimes. In an attempt to reduce discomfort and pain, we therefore recommend the following regime for gentamicin when given outside the neonatal period:

Age 1 month to 12 years: 6 mg/kg once daily
Age 12–18 years: 4–5 mg/kg once daily

Intramuscular injections of antibiotics are painful. Their use has to be balanced against the insertion of a venous cannula which is also painful, especially if multiple attempts are required to insert a cannula.

Whenever possible we recommend that if parenteral antibiotics have to be given, that in the relevant infections, a once daily antibiotic such as ceftriaxone or gentamicin (see above), are administered intramuscularly when intravenous access is difficult.

Of course, whenever it is clinically as effective, antibiotics should be given orally.

41 Bacterial meningitis

Elizabeth Molyneux and Sarah Morley

The incidence of bacterial meningitis is about ten times greater in disadvantaged than in well resourced countries and outcome is worse. Mortality is reported as 12%–44% in resource-poor countries and <5% in advantaged countries. Sequelae are underreported and frequent, including significant neurological impairment and hearing loss.

Causes

- Worldwide the commonest causes are *Streptococcus pneumoniae*, *Haemophilus influenzae* and *Neisseria meningitidis*.
- Neonatal meningitis is commonly caused by *Streptococcus faecalis*, *Klebsiella* species and *Salmonella* species and carries a poor prognosis. *Listeria monocytogenes* and Group B *streptococci* cause both early and late neonatal infections and have a better prognosis than infections caused by anaerobic bacteria.

Diagnosis

- Older child: fever, neck stiffness, vomiting, headache, altered consciousness and possibly convulsions.
- Neonates: signs are more subtle and non-specific and include poor feeding, hyper- or hypothermia, convulsions, apnoea, irritability and a bulging fontanelle.
- Contraindications to lumbar puncture include raised intracranial pressure, child too sick to tolerate a flexed position, infection at puncture site, bleeding tendency (blood clotting platelet disorder), or a widespread petechial rash suggesting meningococcal disease. In these situations antibiotics should be started and lumbar puncture delayed until it is safe to undertake.
- Gram stain of CSF may identify bacteria in about two-thirds of cases and provides a guide to choice of antibiotic therapy in the absence of culture facilities.
- Other laboratory tests of help include blood culture and ***latex agglutination tests*** of CSF, and for general management, full blood count, serum electrolytes and glucose, and urine specific gravity. In malarial areas undertake blood smear and treat appropriately.

Other conditions

- Consider tuberculous meningitis in children who do not respond to the initial antibiotics and particularly if two or more of the following are present: history >7 days, HIV

Table 4.1.1 Bacterial meningitis, typical findings in cerebrospinal fluid

Condition	White cell count ($\times 10^9$/litre)	Cell differential	Protein (g/litre)	Glucose (mmol/litre)
Normal	0.5 <22 in full term, <30 in premature neonates	PMN ⩽2 but <15 neonate	<0.5	Two-thirds blood glucose
Acute bacterial meningitis*	100 to >300 000	Mostly PMN. Monocytes in *Listeria* infection	>1.0	<2.5
Tuberculous meningitis	50–500 sometimes higher	Lymphocytes early but also PMN	>>1.0	<2.5, Usually 0
Herpes encephalitis	usually <500	Mostly lymphocytes PMN early in the disease	>0.5	Normal
Cerebral abscess	10–200	PMN or lymphocytes	>1.0	Normal
Traumatic tap	WBC and RBC	RBC/WBC=500/1	Increases by 0.001 g/litre per 1000 RBC	

* Bacterial meningitis can occur without a pleocytosis. Partial treatment will alter these findings.
PMN=polymorphonuclear granulocytosis; WBC=white blood cells; RBC=red blood cells.

known or suspected, patient remains unconscious, CSF has high white blood cell count (typically >500/ml) mostly lymphocytes, elevated protein (0.8–4 g/litre) and low glucose (<1.5 mmol/litre), Chest *X* ray suggesting tuberculosis, optic atrophy, focal neurological deficit or extrapyramidal movements (see Chapter 4.10).

- Children with HIV are more prone to meningitis and septicaemia from *S. pneumoniae* and *Salmonella* species. Relapse is more frequent and treatment may have to be lengthened.
- *Listeria monocytogenes* may present with headache and little neck stiffness.
- Fungal infections, mostly in children with HIV, often cause severe headache without neck stiffness. Lumbar puncture alone may improve symptoms.

Therapy

Antibiotic choices depend upon activity against the infecting organism, CSF penetration, cost and availability of the

Table 4.1.2 Antibiotic therapy in bacterial meningitis: based on age and when organism is unknown or antibiotic sensitivity cannot be performed

Age	Probable pathogens	Antibiotics of choice	Alternative antibiotics	Comments
Neonates	Gram-negative bacteria Group B *streptococci* *Listeria* *Neiserria meningitidis* *Streptococcus pneumoniae* *Haemophilus influenzae*	Ampicillin + third-generation cephalosporin (cefotaxime or ceftriaxone)	Benzylpenicillin plus gentamicin or ceftazidime (both regimes will need to add in ampicillin if *Listeria* is to be treated)	See Chapter 3.48 No improvement after third day, exclude obvious cause, for example subdural effusions or an abscess by looking for persistent fever plus focal neurological signs or a reduced level of consciousness. An ultrasound scan and neurosurgical input may be required, if available. Repeat lumbar puncture and look for evidence of an improvement (for example a fall in white blood cell count or increase in CSF glucose). Consider a third antibiotic. Look for other signs of infection such as cellulitis at injection sites, arthritis or osteomyelitis.
Infants over 1 month and children 1 month to 5 years	*Neisseria meningitidis* *Streptococcus pneumoniae* *Haemophilus influenzae*	Third-generation cephalosporin (cefotaxime or ceftriaxone IV (add vancomycin or rifampicin if pneumococcal resistance suspected)	Chloramphenicol plus ampicillin (add vancomycin or rifampicin if pneumococcal resistance)	
Children over 5 years	*Neisseria meningitidis* *Streptococcus pneumoniae*	Third-generation cephalosporin (cefotaxime or ceftriaxone IV (add vancomycin or rifampicin if pneumococcal resistance suspected)	Chloramphenicol plus ampicillin or benzylpenicillin (add vancomycin or rifampicin if pneumococcal resistance)	

Notes:

1. Give all antibiotics parenterally for at least 3 days or until temperature has settled and condition is improving; chloramphenicol can then be given orally.
2. Oral chloramphenicol is not reliably absorbed by malnourished children and should be prescribed with caution.
3. The intravenous route is recommended. In the absence of intravenous access, chloramphenicol, cefotaxime and ceftriaxone may be administered intramuscularly. If there is no alternative, high oral doses of chloramphenicol have also been used, although it is not recommended in infants below 3 months of age.
4. The antibiotic regimen should be rationalised once and if culture and sensitivity results for the infecting organism become available.

Table 4.1.3 Antibiotic therapy in bacterial meningitis

Organism	Antibiotics of choice	Alternative antibiotics	Duration
Haemophilus influenzae	Ceftriaxone/cefotaxime	Ampicillin plus chloramphenicol*	10–14 days
*Streptococcus pneumoniae***	Ceftriaxone/cefotaxime	Ampicillin/ benzylpenicillin plus chloramphenicol*	10–14 days
Neisseria meningitidis	Ceftriaxone/cefotaxime	Benzylpenicillin plus chloramphenicol*	7 days
Gram negative bacilli (including *E. coli*)	Ceftriaxone/cefotaxime	Ampicillin plus gentamicin or chloramphenicol*	At least 21 days***
Salmonella enteritidis	Ceftriaxone/cefotaxime plus IV ciprofloxacin (if available)	Meropenem or chloramphenicol* plus ampicillin (may be incomplete cover and excess mortality compared to cephalosporins	At least 21 days***
Listeria monocytogenes	Ampicillin plus gentamicin (see page 394)		10–14 days
Group B *Streptococcus*	Benzylpenicillin plus gentamicin or ceftriaxone/cefotaxime		10–14 days
Staphylococcus spp.	Flucloxacillin plus gentamicin	Flucloxacillin plus chloramphenicol*	10–14 days

* Chloramphenicol should be used with caution in children under 3 months of age. Monitoring of serum levels is advisable in this group.
** *S. pneumoniae* resistant to penicillins and cephalosporins are increasingly prevalent. If resistance is suspected add either rifampicin or vancomycin (see doses below).
*** Gram-negative infections are difficult to treat and have a high rate of sequelae. Repeat lumbar pucture to ensure response to antibiotics may be indicated if the clinical picture is not improving.

Notes
- Choice depends on local antibiotic resistance patterns, national guidelines and drug availability
- Give all antibiotics parenterally for at least 3 days
- Once culture and sensitivity results are available empirical antibiotics should be changed accordingly
- Do not delay antibiotic therapy if cephalosporins unavailable, use the next most appropriate antibiotic combination

Table 4.1.4 Bacterial meningitis: antibiotic doses

Antibiotic	Route	Dose
Ampicillin	IV	100mg/kg/6 hourly (max. single dose 3 g)
Benzylpenicillin	IV	50 mg/kg/4 hourly
Cefotaxime	IV	50 mg/kg/6 hourly (max. single dose 4 g)
Ceftriaxone	IV or IM	80 mg/kg/24 hours once daily* (max. single dose 4 g)
Chloramphenicol	IV	25 mg/kg 6 hourly **(after loading dose of 50 mg/kg)
	Oral	25 mg/kg 6 hourly**
	IM	An oily preparation of chloramphenicol is available and is usually used in a single dose of 50–100 mg/kg with a maximum dose of 3 g. The dose may be repeated after 24 hours. It is recommended only if more suitable alternatives are unavailable
Flucloxacillin or cloxacillin	IV	50 mg/kg 6 hourly (max. dose 8 g/day)
Gentamicin	IV or IM	1 month–12 years 2.5 mg/kg 8 hourly*** (see Chapter 3.48 for neonatal doses and page 394)
Ciprofloxacin	IV	10 mg/kg 12 hourly (5 mg/kg/12 hourly in the neonate)
Meropenem	IV	40 mg/kg 8 hourly (maximum single dose 2 g) by slow IV injection over 5 minutes
Vancomycin	IV	15 mg/kg loading dose and then 10 mg/kg 6 hourly*** (total daily dose should not exceed 2 g)

* Ideally 80 mg/kg 12 hourly should be given for the first three doses followed by 80 mg/kg per 24 hours.
** Although not recommended in children less than 3 months old or in malnourished children, the evidence for this is slight.
***Monitoring of drug levels strongly advised if at all possible.
See page 394 for gentamicin doses.

antibiotic, route of administration and local patterns of antibiotic resistance (Tables 4.1.2–4.1.4). The degree of diagnostic certainty is also important, especially in the case of meningitis with minimal rash, as treatment should be given for all the common causes of bacterial meningitis according to the child's age group.

- Antimicrobial resistance has emerged among the three major bacterial pathogens causing meningitis outside the neonatal period. In the *Meningococcus* intermediate penicillin resistance is increasingly common in many countries and chloramphenicol resistance is emerging. *Haemophilus influenzae* are also frequently beta-lactamase resistant and chloramphenicol resistance is described. Third-generation cephalosporins are therefore the drug of choice for both organisms although, if they are precluded on the basis of cost, chloramphenicol is still an acceptable alternative. Pneumococci resistant to penicillin and to chloramphenicol are widespread, and third-generation cephalosporins are the drugs of choice. However, pneumococcal resistance to third-generation cephalosporins is found in many parts of the world. Treatment of these strains requires the addition of vancomycin or rifampicin to therapy with third-generation cephalosporins.
- Third-generation cephalosporins, cefotaxime, ceftriaxone or ceftazidime, may be necessary first-choice antibiotics in some areas. In neonates ceftazidime which is active against *Pseudomonas* infections may be the most suitable. In epidemic meningococcal meningitis areas, oily chloramphenicol is useful.
- The antibiotic regimen should be rationalised once culture and sensitivity results for the infecting organism become available.
- During confirmed epidemics of meningococcal meningitis and where there are other signs such as petechial rash, lumbar punctures are unnecessary. If resources are very limited, oily chloramphenical (100 mg/kg IM) as a single dose up to 3 g can be curative. If the oily dose is too large for one buttock, divide into two doses.

Duration

- Neonates require 14–21 days treatment. In children, a 10-day course is usually adequate for pneumococcal and *Haemophilus*, and 7 days for meningococcal infections. Seven days of ceftriaxone is usually sufficient (Table 4.1.3).

Corticosteroids

- There is **no** clear evidence that corticosteroids are helpful in bacterial meningitis where there is delay in presentation and antibiotics have already been given some hours earlier. Steroids are generally not indicated in meningococcal disease.
- Dexamethasone has been proven to reduce the incidence of neurological sequelae in meningitis due to *Haemophilus influenzae*. As similar mechanisms operate in meningococcal and pneumococcal meningitis, some authorities recommend the administration of dexamethasone 150 micrograms/kg four times daily for 2 days in bacterial meningitis. The first dose should be given concurrently with, or a maximum of 4 hours after first antibiotic administration.
- Steroids are used in tuberculous meningitis and may prevent hearing loss in some children with *H. influenzae* meningitis, if the child presents during the early part of the disease. In tuberculous meningitis complicated by decreased level of consciousness and/or focal neurological signs, dexamethasone 150 micrograms/kg four times daily for 2–3 weeks, tailing down the dose over a further 2–3 weeks can be helpful.
- **Do not use steroids in** the newborn, in suspected cerebral malaria or in viral encephalitis, or in areas where there is a high incidence of penicillin-resistant pneumococcal disease.

Supportive care

- **Fluids**: maintenance fluids should be given once shock or dehydration has been corrected, initially by the intravenous route but later by nasogastric tube or orally. The degree of dehydration may be underestimated, and deep breathing may be a sign of acidosis. Recent studies suggest that inappropriate secretion of antidiuretic hormone (ADH) is **not common** and that the low serum sodium levels, increased ADH secretion and mild hypertension as found in meningitis are a normal response to raised intracranial pressure. **Avoid overhydration by careful fluid balance and in particular avoid intravenous fluids with low sodium levels such as 5% glucose**. Use 0.9% N saline with added glucose to make a 10% glucose in N saline infusion. If electrolytes are being measured, maintain serum Na^+ in the high normal range and above 135 mmol/litre. ✔
- A **nasogastric tube** may be helpful in the unconscious child or those who are vomiting, in order to protect the airway. A small amount of milk (1 ml/kg/hour) down this nasogastric tube may prevent gastric erosions and improve bowel function.
- **Urine output** should be carefully monitored, particularly in the unconscious child.
- **Seizures**: must be controlled with anticonvulsants, but there is no data to support routine use of prophylactic anticonvulsants (see Chapters 3.36–3.37 on seizures and coma).
- If there is a high **fever** (>39°C) apply temperature-reduction methods including paracetamol.
- **Blood glucose** must be regularly monitored particularly in the infant and young child. Hypoglycaemia must be considered in any child with seizures or altered consciousness and corrected as follows: 5 ml/kg of 10% glucose IV and recheck blood glucose 30 minutes later. If it remains low (<2.5 mmol/litre) repeat the intravenous glucose dose (5 ml/kg).
- **Nutritional support**: a nasogastric tube should be inserted if the child is unable to feed himself or herself after 48 hours. Continue expressed breast milk if breastfed or give milk feeds 15 ml/kg every 3 hours.

Monitoring

- Careful observation is essential.
- Raised ICP and shock are most severe complications. Early recognition and treatment essential.
- Daily weights and urine specific gravity help assess fluid requirement.
- Temperature, pulse, blood pressure, capillary refill time (normal <2 seconds), respiratory rate and effort, conscious level and pupillary responses should be monitored frequently after admission (4–6 hourly), particularly in patients with meningococcal disease (see Chapter 4.6 on meningococcal disease). Pulse oximetry is valuable, if available, for monitoring oxygenation and for identifying early on evidence of respiratory compromise.
- Regular (4 hourly) blood glucose monitoring (hypoglycaemia is common).
- Seizures should be treated immediately.
- A critical care pathway is an ideal way of incorporating observations, treatment, laboratory findings on one chart. Doses and treatments can be standardised and incorporated on the chart.
- If available it is ideal to monitor electrolytes (sodium, potassium, calcium and magnesium, urea and/or creatinine) and replacement of deficits (hyponatraemia due to excessive intravenous administration of hypo-osmolar solutions is common and can predispose to seizures). Monitoring of full blood count and coagulation screen should be carried out regularly if initially abnormal.

Nursing care

- Turn an unconscious child 2 hourly, keeping the child dry, and prevent overheating. Insert nasogastric tube if there is persistent vomiting.

Minimum standards requirements

Meningitis

- Lumbar puncture
- Early parenteral antibiotics
- Antituberculous drugs (if indicated)
- Dexamethasone
- Anticonvulsants
- Monitoring of vital signs, fluid balance, blood glucose and electrolytes
- Immunisation and prophylaxis for contacts
- Follow up for neurological sequelae

- Cooperation of the mother will increase if she is included in progress reports and made part of the caring team.

Complications

- Seizures with or without hypoglycaemia.
- If fever does not settle quickly and if the child's condition deteriorates or is not improving, repeat lumbar puncture.
- If fontanelle is patent, ***do a head ultrasound scan to look for ventriculitis, ventricular dilatation, subdural effusion or brain abscess. In older children, computed tomography or magnetic resonance imaging may be required.***
- Aspiration pneumonia may occur in the unconscious child.
- Hydrocephalus, deafness, visual loss, epilepsy and neurological deficits may develop and be evident either early in disease or at follow up.

Follow up

- Undertake hearing tests in all children, neurological assessments and head circumference (in infants) in the first 6 months after recovery.
- New sequelae are unlikely to develop after discharge but may have been missed.
- Physiotherapy may be required.

Immunisations

Meningococcal capsular polysaccharide vaccine is available for A, C and WY strains. Conjugated C vaccine is in regular use for all ages in some countries (see Chapter 4.6 on meningococcal disease). Hib vaccine is incorporated into many childhood immunisation programmes in rich countries but its cost has so far precluded its use in most resource-poor countries.

Bacterial meningitis: prophylaxis for contacts

Neisseria meningitidis
Rifampicin for two days for all household contacts*:
adults 600 mg twice daily
1 month–12 years 10 mg/kg twice daily
neonates 5 mg/kg twice daily

Haemophilus influenzae
Rifampicin or an oral cephalosporin for four days for all non-vaccinated household contacts under 4 years old

* See Chapter 4.6 on meningococcal disease for alternative antibiotics and vaccination.

4.2 Sexually transmitted diseases

Dankwart F Wittenberg

Minimum standards requirements

- Health education programmes
- Child protection if abused
- Antibacterial drugs
- Antiviral drugs
- Podophylline/trichloroacetic acid

There are more than 20 different infections that may be spread by the sexual route. These extend from the classic sexually transmitted diseases like syphilis or gonorrhoea, through conditions like genital herpes or human papillomavirus that are mainly sexually transmitted, to those infections like hepatitis B and C that can also be transmitted by sexual means.

Anogenital infections in childhood are most commonly acquired through sexual contact or abuse, but may arise as a result of close personal contact within the family or on the playground; and some systemic infections may be transmitted by sexual means without being considered venereal illnesses.

The diagnosis of sexually transmitted disease is considered in the following circumstances:

- A history of recent sexual abuse
- The isolation of sexually transmitted organisms in cases without obvious trauma leading to a diagnosis of chronic sexual abuse
- Specific syndromes and diseases usually transmitted by the sexual route in adults
- Congenital syphilis or perinatally acquired chlamydia or gonorrhoea transmitted from mother in utero or postnatally (see Chapter 3.48).

Sexual abuse

A child known to have been abused recently

Sexually abused children are at risk of acquiring an infection from the perpetrator. In relation to the high frequency of sexual abuse, the typical sexually acquired infections are fairly rare, but the risk depends on a number of epidemiological factors.

The diagnosis of potential infection of a child presenting with sexual abuse includes an active microbiological search by culture of vulval, perineal or anal swabs. Bacterial infections like gonorrhoea, syphilis or chlamydia usually become manifest soon after the assault by the development of local ulcers and infected vaginal or vulval discharge.

The sexually transmitted viral diseases such as herpesvirus type 2 can also become evident soon after the incident, but diseases with a longer latency period such as human papillomavirus are more difficult to link directly to the episode of sexual abuse.

The management of the child potentially infected after sexual abuse consists of the following:

- Management of the sexual abuse – discussed elsewhere (Chapter 5.2)
- Local management of injuries including tetanus toxoid if applicable
- Bacteriological swabs
- Serological tests for syphilis, hepatitis B and HIV, repeated 6 weeks later
- Prophylactic broad-spectrum antibiotic: ceftriaxone 50 mg/kg IM as a single dose (maximum dose 4 g) plus erythromycin 20–40 mg/kg/day in three divided doses for 7 days
- Follow up and appropriate treatment of identified infection (see below).

The presence of a sexually transmissible infection in a child calling attention to the possibility of sexual abuse

This group of children presents with symptoms and signs suggestive of genital, urinary or lower intestinal infection. In children between 2 and 10 years old, the finding of genital, anal or pharyngeal infection with *Neisseria gonorrhoeae*, *Treponema pallidum* or *Chlamydia trachomatis* should prompt a search for evidence of sexual abuse. On the other hand, herpesvirus type 2, *trichomonas vaginalis*, *Mycoplasma* spp. and bacterial vaginosis are not so commonly acquired as a result of sexually transmitted infection in this age group. While human papillomavirus types 6, 11, 16 and 18 are also usually transmitted by sexual means and may present with condylomata, a long latency in the onset of clinical signs means that these may have been transmitted from mother to child during birth,

and close domestic contact other than sexual abuse has also been shown in such cases.

Specific syndromes or diseases usually associated with sexual transmission in adults

These conditions occur particularly in sexually active adolescents. In view of the rampant spread of HIV infection, the approach to the management of sexually transmitted diseases in children and adolescents must include the following aspects:

- Treatment of the symptoms and causes in a typical syndromic approach to STDs, as described below
- Identification of those without symptoms. There is a recognised risk of co-infection, and as both syphilis and HIV may be asymptomatic, serological tests for syphilis (VDRL) and HIV (ELISA) should be offered with appropriate counselling in all patients
- Prevention of new infection by education around safe sex practices and condom use
- Motivation of health-seeking behaviour.

Genital ulcers and lymphadenitis

The infections presenting with genital ulcers with or without inguinal adenopathy and bubos are most often acquired as a result of voluntary or involuntary sexual activity, but may occur as a result of non-sexual inoculation through close domestic or play contact or indirect transmission. The patient should be carefully examined to determine the site, number, size and appearance of the ulcers, the type of exudate, the presence of associated pain, erythema and swelling, or of draining lymphadenopathy.

Regional epidemiological factors determine the relative frequency and likelihood of genital herpes (herpesvirus type 2), syphilis, chancroid, granuloma inguinale or lymphogranuloma venereum.

Genital herpes

This causes painful vesicular or shallow ulcerative lesions on the genitalia. Grouped or single lesions occur on a thin erythematous base but with generally uninflamed intervening epithelium. These regress spontaneously but may recur. Oral aciclovir 200 mg five times daily does not prevent future recurrences, but if started early, will reduce the intensity and duration of symptoms. Locally, anaesthetic and antiseptic creams help to relieve symptoms.

Chancre of primary syphilis

This is a painless ulcer with a serous exudate which is highly infectious. The diagnosis can be made by direct dark field examination or immunofluorescent antibody stains. At this stage, serological tests for syphilis are usually still negative. The treatment in children >12 years consists of benzathine penicillin, 50 000 U/kg IM as a single dose (50 000 U = 30 mg).

Chancroid

This is caused by *Haemophilus ducreyi*. Painful papulovesicles or ulcers on the genitalia are associated with suppurative inguinal adenopathy. In the absence of adenopathy, the condition has to be differentiated from herpes or syphilis, the latter of which is usually painless. In treatment of children >12 years, the following are satisfactory: azithromycin 1 g orally as a single dose, ceftriaxone 125–250 mg IM as a single dose, or erythromycin base 500 mg orally four times daily for 7 days.

Lymphogranuloma venereum and granuloma inguinale

The conditions associated with prominent inguinal lymphadenopathy and less obvious genital ulcers are lymphogranuloma venereum, caused by *Chlamydia trachomatis*, and granuloma inguinale, in which ulcers, subcutaneous nodules and granulomas occur in the genital or anal regions. In these cases, treatment for children >12 years with doxycycline 100 mg orally twice daily or erythromycin 500 mg orally four times daily should be continued for 21 days.

Urethritis and vulvovaginitis

These patients present typically with a discharge from urethra or vagina. The character of the discharge may be non-specific or it may have typical features allowing a presumptive diagnosis concerning its aetiology. Together with the discharge, there may be other features such as itching, discomfort or dysuria. There may be inflammatory erythema and swelling of the tissues. Where pruritus is a major symptom, *Trichomonas* or *Candida albicans* should be suspected. The appearance of the discharge may be typically white cheesy in *Candida* or creamy-purulent and frothy in *Trichomonas* infection, but often is fairly non-specific.

The organisms responsible for this mode of presentation include *Neisseria gonorrhoeae*, *Chlamydia trachomatis*, *Trichomonas*, *Candida* spp., *Gardnerella vaginalis* and *Ureaplasma* spp. In the syndromic approach to the management of patients with surface epithelial infection, broad-spectrum treatment aimed at gonorrhoea, *Chlamydia* and *Trichomonas* or *Candida* is given at the same time as bacterial swabs are taken for culture. Where laboratory resources are scarce, bacteriological investigations may be reserved for those not responding appropriately to the first course of therapy.

The treatment suggested for children >12 years involves ceftriaxone 125–250 mg IM as a single dose against *Neisseria gonorrhoeae* together with azithromycin 1 g orally single dose *or* doxycycline 100 mg orally twice daily for 7 days (alternative erythromycin 40–50 mg/kg/day for 14 days in pre-adolescents) for *Chlamydia*. Where *Trichomonas* or bacterial vaginosis due to *Gardnerella vaginalis* is identified or strongly suspected, metronidazole is added as 15–30 mg/kg/day in three divided doses for 7 days. *Candida* infection is treated with local nystatin (100 000 U/ml 3–4 times daily).

Acute balanoposthitis

Inflammation of the glans and prepuce can have a large number of infectious and also non-infectious causes. In the usual case, there is erythema and swelling of the glans and prepuce together with local exudate. Most such cases are not due to sexually transmitted infection, but are caused by beta-haemolytic streptococci, *Staphylococcus aureus* or *Candida albicans*. These arise secondary to local trauma including ritual circumcision. Allergic contact dermatitis and rarer causes such as psoriasis or pemphigus should also be considered. Sexually transmitted organisms include *Chlamydia*, *Gardnerella vaginalis*, *Trichomonas*, *Candida albicans*, syphilis, herpes virus and papillomavirus. If "milking" along the length of the urethra produces a purulent discharge also, STDs are more likely.

Accordingly, the evaluation of a boy presenting with balanoposthitis includes examination for the presence of urethral discharge and a urine dipstick. A swab should be sent for microbiological confirmation. A suggested treatment for children >12 years is azithromycin 1 g orally in one dose, or erythromycin 40–50 mg/kg per day in four divided doses for 14 days, plus metronidazole 15–30 mg/kg per day in three divided doses for 7 days. In the presence of urethral discharge, treatment should also include antibiotic cover for gonorrhoea.

Genital warts

Condylomata acuminata are fleshy, soft, pedunculated or flat warty lesions that may sometimes have quite a narrow base. They occur singly or in clusters. In sexually active adolescent boys, they may occur on the shaft or corona of the penis, and in girls on the genital mucosal surface both inside and outside the vagina. Perineal cutaneous condylomata are not always acquired sexually. Human papillomavirus types 6 and 11 cause these warts. Apart from the visible wart, the infection may be quite asymptomatic, particularly where lesions occur intravaginally. They must be differentiated from the flat papular condylomata lata of syphilis, skin tags and molluscum contagiosum. A diagnostic test is to apply 5% acetic acid to the lesion: condylomata acuminata turn white, but false-positive results are common.

Local treatment is satisfactory in most instances, although recurrences occur. Trichloroacetic acid or 10–25% podophyllin may be applied to external lesions, taking care not to involve normal skin. This is repeated in 7 days. ***Laser vaporisation under anaesthesia*** or excision biopsy may be used for lesions which are less accessible or intravaginal.

The association with genital dysplasia and carcinoma should be remembered and therefore Pap smears and regular follow up are indicated in girls with human papillomavirus infection.

Pelvic inflammatory disease (PID) and epididymitis

The deep infections of the upper female genital tract present with features of infection, such as fever and leucocytosis, together with lower abdominal pain and a vaginal discharge. There may be signs of pelvic peritonitis or a tender mass on vaginal or rectal examination. Epididymitis in males presents typically with unilateral pain, swelling and tenderness of the testis, together with urethral discharge. This can be distinguished from testicular torsion by means of an ultrasound examination. In sexually active adolescents, these infections are most often caused by *Neisseria gonorrhoeae* or *Chlamydia trachomatis*. Such patients may be very ill and require hospitalisation including possible surgical drainage. General supportive therapy is given as required. The antibiotic therapy aims at the above two organisms and should therefore include a third-generation cephalosporin like ceftriaxone, or a quinolone such as ciprofloxacin, plus doxycycline or erythromycin. In severe cases, other intravenous broad-spectrum antibiotics including aminoglycosides and clindamycin may be considered.

4.3 Diphtheria

Brian Coulter

Minimum standards requirements

- ABCD (especially airway protection)
- Immunisation (Chapter 1.29) and prophylaxis of contacts
- Early parenteral antibiotics
- Dexamethasone
- Early antitoxin
- Bed rest, close observation and ECG monitoring
- Intubation/tracheostomy (Chapters 3.17 and 6.14)

In countries with adequate coverage of immunisation (>70%) diphtheria is now uncommon. Epidemics still occur associated with a fall in level of immunisation as has happened in the mid 1980s, and early 1990s in Russia and Ukraine and other republics of the former USSR. It affects all ages.

Epidemiology

- When levels of immunisation are low, children are the major group affected. Young infants are protected by maternal antibody.
- With improvement in expanded programme of immunisation rates, affected age groups shift to older children and adults. Boosters at school entry and school leaving are essential to provide adequate herd immunity.
- Mass movement of people, for example refugees or army personnel, are important sources of spread in epidemics.
- Commoner in autumn and winter.
- In tropical countries, skin infection by *C. diphtheriae* provides a reservoir that results in natural immunity of the carrier and subclinical spread within the community.

Pathogenesis

- *C. diphtheriae* invades upper respiratory tract.
- Incubation period is 2–4 days.

Table 4.3.1 Clinical features of diphtheria

Site	Comments
Pharynx +++	Affected in over 90% of cases
Tonsil ±	Yellow/white to grey/black (if haemorrhagic) thick membrane which extends beyond the tonsils and covers the adjacent pharyngeal wall. Bleeds when separated from underlying tissue. Pharyngeal membrane may extend to nares, palate or larynx. There may be distortion of soft palate, tonsils etc. If confined to tonsils, little toxaemia
Nasal ±	Serosanguinous discharge Sore nose and lip Little toxaemia Highly infectious
Neck	Enlarged, tender cervical nodes. Surrounding oedema: “Bull neck”
Skin 0−+	Any type of lesion, for example bites, impetigo, may be infected. May progress to ulcer with punched-out sharp edges. Important reservoir for transmission and natural immunisation May result in respiratory colonisation
Other sites	Conjunctiva, ear and vulva
Levels of toxaemia 0, ±,++,+++	Low-grade fever (rarely >38.9°C) Weak, rapid pulse, limp, apathetic, restless Rarely haemorrhagic diathesis

Table 4.3.2 Complications of diphtheria

Complication	Weeks	Comments
Toxaemia	1	Related to extent of membrane and amount of toxin absorbed. May result in fatal cardiovascular collapse in first 10 days. Disseminated intravascular coagulation. Survivors of severe toxaemia usually have further cardiovascular or neurological complications
Myocarditis	2–3 (range 1–6)	Onset related to severity of toxaemia Soft first heart sound, apical systolic murmur ECG: conduction abnormalities, ST-T wave changes. ***Echocardiogram:*** left ventricular dilation, reduced contractility, hypertrophied left ventricle, sometimes pericardial fluid Biochemistry: ***blood myoglobin levels elevated, elevated lactate dehydrogenase, elevated creatine phosphokinase*** Mortality: high in early onset, severe carditis
Palatal paralysis	3	Probably due to local absorption of toxin: "fluids come down nose" Resolves in a few days
Visual accommodation	4–5	Blurring of vision, sometimes strabismus
Bulbar, heart, respiratory and limb nerves	6–8	Bilateral, resolve completely if patient survives

- Diphtheria toxin causes necrosis and exudation in local tissue that results in formation of the "membrane". An attempt at removal of the membrane causes bleeding.
- Toxin is distributed by blood and lymphatic system which results in toxaemia, and causes cardiac and neurological complications.
- Non-toxin producing *C. diphtheriae* may cause focal disease but not cardiac and neurological complications. Vaccination does not protect against this organism.

Clinical features

Symptoms are initially due to disease of upper respiratory tract and associated toxaemia. Later symptoms relate to the level of toxin absorbed into the circulation. Cases with small membranes and low toxaemia recover spontaneously and most remain subclinical.

Diagnosis

- Unless all children with upper respiratory symptoms, including croup, have an appropriate examination, diphtheria will be missed.
- A portion of membrane or a swab taken from beneath it should be sent for Gram stain and culture. Laboratory should be informed of suspected diagnosis so that appropriate culture medium is used.

Management (see also Chapter 3.17)

Aim is to neutralise toxin released into blood by the bacillus and to kill the bacteria.

- Admit to isolation (***on ICU if possible***) cared for by staff that are fully immunised.
- Be prepared for intubation/tracheostomy especially if laryngeal diphtheria is suspected.
- Dexamethasone (0.6 mg/kg once or twice daily IV or orally if possible) should be given in cases of moderate to severe airway obstruction and when there is swelling of the neck.
- Take great care when examining the throat or taking a sample of the membrane as it may precipitate complete airway obstruction.
- Intravenous or nasogastric maintenance fluids if child cannot drink
- Benzylpenicillin 50 mg/kg given 4 hourly IV. Change to procaine penicillin 25 000–50 000 units/kg IM once daily when toxic symptoms have subsided or where toxicity is slight or, if the child can drink, to penicillin V 12.5 mg/kg 6 hourly. Erythromycin 40–50 mg/kg per day in four divided doses (max. 2 g/day) IV, and orally when child can swallow, is an alternative. Antibiotics should be given for 7–10 days.
- **Antitoxin must be given as soon as possible** (after the test dose). The dose is dependent on the severity of the disease rather than the site of the membrane; although the two usually coincide:

  Nasal and tonsillar (mild disease) 20 000 units IM

  Laryngeal with symptoms (moderately severe): 40 000 units IM/IV

  Nasopharyngeal (moderately severe): 60 000–100 000 units IV depending on severity

  Combined sites/delayed diagnosis (malignant disease): 60 000–100 000 units IV

In practice, give 60 000 units to all cases with visible membrane and neck swelling.

Commercially available antitoxin is extremely expensive but highly purified. Some countries, for example Vietnam, make their own antitoxin but it is much **less purified** than the Aventis Pasteur vaccine for example, and **cannot be given intravenously**.

Test dose and desensitisation

See Chapter 1.23

Test dose

- As antitoxin is horse serum a test dose with 0.1 ml of 1 in 1000 dilution in saline is given intradermally.
  - Positive reaction is 10 mm erythema occurring within 20 min.
  - If no reaction, give full dose IV/IM as appropriate.
  - Have epinephrine 1 in 1000 and syringe available to give IM if anaphylaxis occurs (10 micrograms/kg)
- Desensitisation: (if test dose positive)
  Give graduated doses of increased strength every 20 minutes commencing with:
  - 0.1 ml of 1 in 20 dilution in saline SC followed by 1 in 10 dilution
  - 0.1 ml of undiluted SC then 0.3 ml and 0.5 ml IM
  - Then 0.1 ml undiluted IV

Additional treatment

- Give oxygen if cyanosed or SaO_2 <90%. Use nasal cannulae or facemask held close to child's face by the mother. **DO NOT use nasal or nasopharyngeal catheters** as these can precipitate complete airway obstruction. Be aware that giving oxygen does NOT compensate for hypoventilation which if severe will require intubation/cricothyroidotomy/tracheostomy (see Chapter 6.14). Note that intubation may dislodge the membrane producing complete airway obstruction.
- Bed rest and observation for 2–3 weeks at least, depending on severity.
- Regular monitoring of cardiac function. Serial ECGs two or three times per week through the critical period from admission until towards the end of the second week of illness. Rhythm disturbances, particularly atrioventricular block sometimes going on to complete heart block are not uncommon, and are often the earliest evidence of cardiac involvement. With severe cardiac involvement (which often follows from severe local disease) the children develop a low output state and die from cardiac failure or arrhythmias. Poor urine output and rising creatinine are early indicators of poor prognosis and should be monitored, together with serum potassium which should be kept in the normal range (see Chapter 3.7). Strict bed rest is essential for all children until the critical period for cardiac problems has passed (minimum of two weeks from onset). Captopril at the earliest sign of any cardiac involvement, may be helpful (100 micrograms/kg once daily; as test dose–supine and monitor blood pressure carefully followed by 100–200 micrograms/kg 8 hourly). Prednisolone 1.5 mg/kg/day for two weeks may be of value in reducing incidence of myocarditis.
- Nasogastric feeds if palatal or bulbar paralysis occurs. Bulbar problems rarely become evident until several weeks later, so even if children come through the phase of upper airway obstruction and survive the cardiac problems, they should remain in close contact with the hospital for at least six weeks.
- Immunise on discharge.

Prevention

- Maintaining immunity at all age levels in the community is important. Additional immunisation at school entry and leaving (see Chapter 1.29).
- Give immunised household contacts a booster of toxoid.
- Give all unimmunised contacts one dose of IM benzathine penicillin (600 000 units for <5 years and 1.2 million units for >5 years). Immunise and check daily for signs of diphtheria.

4.4 Leprosy

Terence J Ryan

Minimum standards requirements

- Clinical awareness
- Rifampicin, doxycycline, chloramphenicol
- Public health measures

Leprosy is caused by the *Mycobacterium leprae*. The organism invokes an immunological response in the skin and especially focuses on superficial cutaneous nerves, resulting in anaesthesia and paralysis of hand, foot and facial muscles.

In countries where the prevalence is low, leprosy may be forgotten. As a consequence, integration of leprosy into general health services requires that there is flow from the anxious and fearful family or from the traditional healers (whom 85% of such families consult) to health centres where common skin diseases due to bacteria or fungus or parasites are well managed. Conditions not diagnosed or not responding are referred onwards to a centre with greater expertise and adequate supplies of essential drugs.

The clinical spectrum of leprosy ranges from the early single lesion through multiple, increasingly symmetrical, lesions to widespread infiltration of the skin with a tendency to coalesce into nodules. The single or few lesions occur in persons with high resistance and may resolve even without treatment. More generalised forms of the disease occur over several years of incubation. It is these which are the public health problem because they are teeming with bacilli which are released into the environment from the nose or skin wounds.

Diagnosis

Early diagnosis requires an examination of the skin and the recognition of the early hypopigmented lesion which is insensitive to light touch, pin pricks or hot and cold sensation. Also requiring early diagnosis are the highly inflammatory and destructive immunological responses known as "reaction".

Differential diagnosis

- The **early single lesion** is hypopigmented, not totally depigmented and, if there are two or three lesions, they are usually asymmetrical. The lesions persist for months and are unresponsive to moisturising creams and antifungals.
- **Vitiligo** is totally depigmented and usually symmetrical. Reaction to light touch, pin-prick and hot and cold is not impaired.
- **Pityriasis alba** is a very common dry patch of eczema, usually symmetrical, especially on the cheeks and extensor surface of the limbs. It responds to moisturising creams and hydrocortisone and it is not fixed. The pigment is lost by surface exfoliation rather than being unformed. There is no impairment of sensation. The lesion may itch.
- **Pityriasis versicolor** is a common infection of the skin producing depigmentation especially of the upper trunk and a fine scale. It is mildly inflammatory and on a white skin can appear to be a dull reddish brown colour. The infection responds to selenium sulphide shampoo, Whitfield's ointment or ketoconazole, although the depigmentation requires sunlight exposure for rapid repigmentation.
- **Postinflammatory depigmentation** is preceded by an undisputed injury such as a burn, chickenpox or fungal infection or plaque of psoriasis. There may be mild impairment of sensation and it is clearly scarred.

Reaction

"Reactions" are of two types.

- Erythema nodosum-like, with multiple, tender, symmetrical red lumps in the skin due to immune complexes and, hence, associated with general ill health and fever. It often responds to rest and non-steroidal anti-inflammatory drugs. There is a background or story of other features of leprosy over a prolonged period.
- The other type of reaction is a single red plaque-like swelling. It is often focused over a palpable cutaneous nerve which may be destroyed in the process. It requires the prescription of high-dosage oral steroids (prednisolone 1 mg/kg/day for 7 days) and can be confused with an acute, very painful staphylococcal or streptococcal erysipelas swelling requiring antibiotics. A previous history of leprosy is usual.

The leprosy process infiltrates cutaneous nerves which become palpable and are a helpful confirmation of the

disease: radial, ulnar, posterior cervical, lateral popliteal and muscular cutaneous on the dorsum of the foot. Early signs of damage to the ulnar nerves include flexion of the fourth and fifth fingers or claw hand, a failure to be able to flex all the fingers to meet together with the thumb. A failure to blink and a tendency to foot drop are other early signs of nerve damage.

Treatment of leprosy

The recommended standard regimen for multibacillary leprosy is:

- Rifampicin: 20 mg/kg (maximum 600 mg) once a month (supervised)
- Dapsone: 1 mg/kg daily
- Clofazimine: 3.5 mg/kg once a month (supervised to aid compliance) and 1 mg/kg daily

Duration: At least 2 years

The recommended standard regimen for paucibacillary leprosy is:

- Rifampicin: 20 mg/kg (maximum 600 mg) once a month (supervised)
- Dapsone: 1 mg/kg daily

Duration: At least 6 months

Treatment

Multidrug therapy cures leprosy. Multidrug therapy should be given under supervision by experts able to provide full advice on the preventive management of disability and who may confirm the diseases by skin smears or biopsies and can manage reactions. Standard drug therapy is available free from government programmes for the elimination of leprosy. WHO guidelines for multidrug therapy include a single dose for a single lesion, or two drugs for lesions which contain more than one bacteria. A daily regimen for one year of three drugs is necessary for more widespread multibacillary disease. Lepromatous leprosy is subject to reaction even after one year of therapy and patients must be educated to return for diagnosis and appropriate therapy promptly. Relapse after completion of therapy is uncommon but well documented. There is still a fear of the stigma of leprosy. The emphasis of therapy is that it is a cure and rapidly renders the patient non-infectious.

Children may be more troubled by the haemolytic side effect of dapsone, and are less tolerant to rifampicin. New drug regimens include ofloxacin (fluoroquinolone), minocycline (tetracycline) and clarithromycin (macrolide). Several experimental and clinical studies have demonstrated that these drugs either alone or in combination with other antileprosy drugs have significant bactericidal activity. Patients presenting with single skin lesion paucibacillary leprosy can be treated with only one dose containing rifampicin 20 mg/kg, ofloxacin 15 mg/kg and minocycline 100 mg (only for children >12 years). Multibacillary leprosy patients who do not accept clofazimine can be treated with this combination given monthly for 24 months.

4.5 Leptospirosis

Roberto Jiminez

Minimum standards requirements

- Recognition and treatment of shock
- Antibiotics: amoxycillin, penicillin, doxycycline (parenteral for severe disease)
- High-dependency intensive care (see Chapter 1.25)
- Public health measures

Leptospirosis is a zoonotic disease caused by Leptospira species with a worldwide distribution. Transmission to humans is from infected animal urine. The onset is usually abrupt. The clinical course is usually biphasic and with multisystemic involvement. The initial (septicaemic) phase lasts 4–7 days, the second (immune) phase 4–30 days. It can be lethal in the acute period and is similar to diseases such as dengue, malaria, hepatitis and viral illnesses.

Clinical manifestations

- General symptoms: Headache, myalgia, vomiting and anorexia, arthralgia, macular rash.
- Central nervous system: CSF pleocytosis and elevated protein, meningism, neurological symptoms.
- Renal system: Pyuria, haematuria, proteinuria, oligouria/anuria, dysuria, back pain.
- Gastro-intestinal: Abdominal pain, diarrhoea, constipation, abnormal liver function tests, hepatomegaly, jaundice, gastrointestinal bleeding
- Respiratory system: Cough , pharyngitis, otitis media, chest pain, pneumonitis, pulmonary oedema and haemoptysis.
- Cardiac system: Arrhythmias, conduction and other ECG abnormalities.
- Haematology: Blood clotting disorder, petechiae, bruises, epistaxis, thrombocytopenia, lymphadenopathy, splenomegaly.
- Eyes: Conjunctival bleeding, photophobia, retro-orbital pain, uveitis, papilloedema.

History and examination

- ENQUIRE about headache, fever, abdominal pain, breathing difficulties and cough, diuresis, bleeding, diarrhoea or vomiting.
- ASSESS: vital signs (blood pressure, pulse, respiratory rate), "alarm signs", blood film for malaria parasite. Consider Dengue fever.
- WATCH OUT for "alarm signs" of leptospirosis: abdominal pain, respiratory distress, jaundice, bleeding and oliguria.

Classification

- Mild disease: Headache, fever, myalgia, no evidence of bleeding.
- Moderate disease: Headache, fever, myalgia, abdominal pain and jaundice.
- Severe disease: Weil's disease or ictohaemorrhagic fever: shock, abdominal pain, respiratory failure, pulmonary haemorrhage, acute renal failure, altered consciousness and bleeding.

Diagnosis

- Blood culture in initial phase and urine in the second phase.
- ***Fourfold or greater rise in agglutination titre.***
- ***Rapid diagnosis with specific IgM (ELISA).***

Management

- Mild disease
  - Discharge home with advice about hydration and "alarm signs"
  - Antibiotics
    - <10 years: amoxycillin 15mg/kg three times daily for 7 days or if allergic: erythromycin 10–15 mg/kg/day three times daily for 7 days.
    - >10 years: doxycycline 100 mg twice daily for 7 days.

- Moderate disease:
  - Investigations: amylase, liver and renal function, full blood count and blood clotting, chest *X* ray.
  - Observe for 48 hours, monitor vital signs 4 hourly.
  - If abdominal pain/respiratory distress settle, discharge.
  - Antibiotics: benzylpenicillin 25–50 mg/kg IV 6 hourly for 3 days then change to oral penicillin.
- Severe disease
  - Investigations: as above.
  - Give oxygen as required, intravenous fluids and pass nasogastric tube.
  - Keep accurate fluid balance chart.
  - Pulmonary haemorrhage may require assisted ventilation with PEEP.
  - Pulmonary oedema: fluid restrict, oxygen, diuretics.
  - Management of disseminated intravascular coagulation, renal failure, myocarditis.
  - Antibiotics: benzylpenicillin 50 mg/kg IV 6 hourly for 3 days and then change to oral.

4.6 Meningococcal disease

Sarah Morley and Michael Levin

Minimum standards requirements

- Early parenteral antibiotics
- Treatment of shock
- Neurological assessment and cerebral protection
- Frequent reassessment of clinical status
- Electrolyte monitoring and replacement
- Replacement of platelets, clotting factors and red cells
- Follow public health procedures

Introduction

Meningococcal disease is caused by *Neisseria meningitidis*, a Gram-negative diplococcus which is a commensal of the human nasopharynx. Endemic meningococcal disease, primarily affects children under 5 years old. Some areas, in particular, the meningitis belt in sub-Saharan Africa, suffer from epidemics of meningococcal disease. Temperate climates usually experience an increase in disease during winter months whereas in sub-Saharan Africa, conditions during the dry season cause a sharp rise in incidence.

Predominant disease-causing organisms are serogroups A, B and C and W135 with other serogroups generally only

Table 4.6.1 Common presenting symptoms and signs of meningitis and meningococcal septicaemia

Meningitis	Meningococcal septicaemia
Symptoms Fever Headache Nausea and vomiting Rash Drowsiness or irritability Neck and back pain, and stiffness Convulsions	*Symptoms* Fever Petechial/purpuric rash Shivering/rigors Malaise and lethargy/confusion Headache Nausea and vomiting Limb and joint pain Absence of neck stiffness Collapse
Signs Fever Non-blanching rash Neck stiffness/positive Kernig's sign/opisthotonus Decreased conscious level	*Signs* Fever Petechial/purpuric rash Shock: tachycardia low pulse volume cool peripheries capillary refill time >2 seconds hypotension (late sign) urine output reduced (<1 ml/kg/hour) tachypnoea hypoxaemia decreased conscious level Cardiac insufficiency: pulmonary oedema, hepatomegaly
Infants Signs of meningitis may be non-specific with neck stiffness frequently absent. Bulging fontanelle may be present. Suspect meningitis in any febrile infant especially where there is marked irritability, vomiting, and poor feeding	

causing infection in specific patient groups (for example complement deficiency and the immunocompromised). Serogroup A is associated with epidemic disease in the meningitis belt of Africa, Middle East and southern Mediterranean regions and less commonly in other developing countries. Serogroups B and C are largely responsible for endemic disease in temperate countries.

Clinical features

In general, meningococcal disease presents either as **meningitis** or as **meningococcal septicaemia**, although many patients present with a mixed picture. In developed countries, the majority of cases may present with septicaemia and frequently with shock, whereas in African serogroup A epidemics, meningitis is the commonest presentation.

Meningococcal disease should be suspected in any patient who presents with a non-blanching (petechial or purpuric) rash. However 13% of cases may present with a maculopapular rash and 7% may have no rash. Severity of rash does not correlate with severity of disease.

Life-threatening features of meningococcal disease

Shock: particularly uncompensated shock (hypotension and tachycardia)
Shock causes the majority of deaths due to meningococcal disease and is a medical emergency

Raised intracranial pressure
Decreased conscious level (Glasgow Coma Score*/or Modified Children's' Coma Score <8 or deteriorating)
Focal neurological abnormalities especially false localising signs, for example pupillary dilatation
Abnormal postures (decorticate or decerebrate)
Convulsions
Rising blood pressure with falling pulse rate
* Glasgow Coma Score (GCS) (see Chapter 3.36)

Table 4.6.2 Investigations in meningococcal disease

Investigations		Comment
Microbiology	Lumbar puncture**	For Gram stain and culture* Remember contraindications
	Skin scrapings from a purpuric lesion	For Gram stain and culture*
	Throat swab	Culture*
	Blood culture	Gold standard diagnostic test for septicaemia Positive in 30% or more of previously untreated cases
Special microbiology ***Advanced methods for case ascertainment in specialist laboratories***	***Meningococcal serology*** ***Rapid antigen screen on serum or CSF Meningococcal polymerase chain reaction***	Acute and convalescent samples required
Haematology	Full blood count	Low haemoglobin In milder disease or meningitis usually high neutrophil count In severe septicaemia low total white cell count with neutropenia is common. Low platelet count in disseminated intravascular coagulation
	Coagulation screen	Prolonged PT, KCTT and TT. Raised ***fibrin degradation products***
Biochemistry	Full urea, creatinine and electrolyte screen including ***calcium, phosphate and magnesium***.	Hypokalaemia Hypocalcaemia Hypomagnesaemia Hypophosphataemia Metabolic acidosis Raised urea and creatinine (if severe, suspect prerenal failure)

* Meningococci should be cultured on appropriate media (Mueller-Hinton or chocolate agar) to allow identification and ***serogrouping with antibiotic sensitivity analysis***.
** Where laboratory facilities are scarce, diagnosis of uncomplicated meningitis can be made by lumbar puncture alone – colour, cell count, glucose sticks, Albustix.

CSF features consistent with meningococcal meningitis
Turbid or purulent (may be clear or blood stained)
White blood cells >500 cells/mm^3 (<3 cells/mm^3 in normal CSF)
Protein usually >0.8 g/litre (<0.6 g/litre in normal CSF)
Glucose reduced compared to blood glucose concentration
Gram-negative diplococci (intra- or extracellular) in 72% of previously untreated cases

When not to perform a lumbar puncture
Lumbar puncture may precipitate coning if there is significantly raised intracranial pressure. In septicaemia, lumbar puncture is unlikely to be helpful and may cause rapid deterioration in an unstable child

Contraindications
- **Suspected critically raised intracranial pressure**

Glasgow Coma Score/Modified Children's Coma Score <8 (or if child is unresponsive to pain)
Focal neurological signs including pupillary abnormalities
Unexplained hypertension/bradycardia
- **Shock**

Not to be performed when significant clotting disorder or low platelet count (<100 × 10^9/litre) is present

Management of meningococcal disease (see Chapter 4.1 for uncomplicated meningococcal meningitis)

Principles

- In suspected cases give injection of benzylpenicillin before transfer of child to hospital.
  Recommended doses of benzylpenicillin are:
  <1 year: 300 mg
  1–10 years: 600 mg
  >10 years: 1.2 g
- On admission, early antimicrobial therapy, for example benzylpenicillin (for dose and alternatives see Table 4.6.3) which should ideally be given intravenously but, if this is not possible, intramuscularly.
- Close monitoring and aggressive supportive therapy if features of shock or raised intracranial pressure develop.
- **Never delay antimicrobial therapy if facilities are not available for immediate lumbar puncture or blood culture**.
- **The most appropriate available antibiotic should be used**. In general, intravenous benzylpenicillin (or intravenous ampicillin or intravenous chloramphenicol) is the drug of choice where meningococcal disease is the most likely diagnosis. Where the diagnosis is uncertain, or where there is a high prevalence of penicillin resistant meningococci, broadspectrum antibiotics should be used (see Table 4.6.3), ideally including a third generation cephalosporin. Do not delay administration if cefotaxime or ceftriaxone are unavailable (use benzylpenicillin, ampicillin or chloramphenicol instead for the initial dose).
- Risk of transmission disappears after 24–48 hours of antibiotic therapy. Isolation is not essential but staff should maintain good hygienic practice and wear masks and gloves during invasive procedures such as intubation, airway and mouth care and line insertion.
- Parenteral antibiotic treatment should be given for 7 days if the diagnosis of meningococcal disease is certain. Once culture and sensitivity results are available treatment should be modified appropriately (see Table 4.6.3).
- Notification to local public health authority.
- Prophylactic antibiotics and possibly vaccination for close contacts.

Table 4.6.3 Antibiotic doses

Antibiotic	Route	Dose
Ampicillin	IV	400 mg/kg/24 hours in four divided doses (max. single dose 3 g)
Benzylpenicillin	IV	300 mg/kg/24 hours in six divided doses (maximum single dose 2.4 g)
Cefotaxime	IV	200 mg/kg/24 hours in four divided doses (max. single dose 4 g)
Ceftriaxone	IV/IM	80 mg/kg/24 hours once daily* (max. single dose 4 g)
Chloramphenicol	IV	100 mg/kg/24 hours in four divided doses**
	Oral	100 mg/kg/24 hours in four divided doses***
	IM	An intramuscular preparation of oily chloramphenicol is available in some areas and is usually used in a single dose of 50–100 mg/kg with a maximum dose of 3 g. It is recommended only if more suitable alternatives are unavailable. Dose may be repeated at 24–48 hours.

* Ideally 80 mg/kg should be given 12 hourly for the first 3 doses followed by 80 mg/kg 24 hourly.
** Chloramphenicol should be used with caution in infants less than 3 months of age. Monitoring of serum levels is recommended and lower doses with wider dosage intervals may be required.
*** Oral chloramphenicol is usually used only following 3–4 days of parenteral antibiotics. Although not recommended for children less than 3 months of age or in malnourished children the evidence for this is slight.

Other points

- Early recognition of life-threatening disease (shock and raised intracranial pressure) is vitally important. There is a very high risk of death if patients are not resuscitated aggressively at presentation.

- Assess airway patency, breathing and circulation (ABC) and examine for signs of shock and raised intracranial pressure (see above). Management regimens differ for different presentations: shock; raised intracranial pressure; meningitis uncomplicated by either shock or raised intracranial pressure.
- Many children present with a mixed picture and may require treatment of shock as well as management of neurological complications.
- Meningococcal disease is often progressive and patients may continue to deteriorate after antibiotic and supportive therapy have been initiated. All suspected cases should be closely monitored for cardiovascular and neurological deterioration for at least 24 hours.
- Management of children with severe shock or raised intracranial pressure who do not respond fully to initial resuscitation is complex. Every effort should be made to admit these patients to an appropriate intensive care facility.

Shock (see Chapter 3.6)

- **THIS IS A MEDICAL EMERGENCY**
  - Assess ABC and give high flow oxygen. Check blood glucose (for example BM stix).
  - Obtain intravenous or intraosseous access.
  - Take blood for culture, full blood count, coagulation screen and urea and electrolytes and commence appropriate intravenous antibiotics.
- **DO NOT PERFORM LUMBAR PUNCTURE**
  - Commence fluid resuscitation immediately using 20 ml/kg of crystalloid or colloid given as fast as possible. Reassess and use repeated fluid boluses of 20 ml/kg if signs of shock persist. **Note**: very large volumes of fluid resuscitation may be required early in disease. Use either 0.9% sodium chloride or other non-glucose containing crystalloid or ideally a colloid such as 4.5% human serum albumin. Blood products such as packed cells, fresh frozen plasma and platelets may be required.
  - Patients who remain shocked after 40 ml/kg colloid/crystalloid will probably benefit from inotropic support, for example dopamine 10–20 micrograms/ kg/min IV by peripheral intravenous cannula (see Chapter 1.25).
  - Shocked patients are at significant risk of developing pulmonary oedema as fluid therapy increases. Ideal therapy is ***mechanical ventilation*** for patients who require more than 40 ml/kg fluids. In disadvantaged countries, where facilities for mechanical ventilation are unavailable, further fluid boluses should be undertaken cautiously with repeated boluses of 5–10 ml/kg of crystalloid, colloid or blood products as appropriate. If pulmonary oedema develops (with tachypnoea, hypoxia, cough and fine crackles, raised jugular venous pressure and hepatomegaly) further fluid administration should be withheld until the patient stabilises. Inotropic support, as described above, may be of benefit.
  - Full neurological and cardiovascular assessment with regular (at least hourly) assessment of: pupillary responses, conscious level, pulse, blood pressure, capillary refill time, respiratory rate and effort (pulse oximetry if available) and temperature.
  - Regular (ideally 4 hourly initially) monitoring of electrolytes (sodium, potassium, ***calcium and magnesium, phosphate***, urea and/or creatinine) and glucose and replacement of deficits. ***Blood gases should be undertaken to detect metabolic acidosis from shock or respiratory acidosis due to ventilatory insufficiency***. Severe metabolic acidosis, which does not respond to fluid therapy, may require sodium bicarbonate correction. Regular blood gas monitoring is essential for ventilated patients.
  - Monitor full blood count and coagulation regularly if initially abnormal. Replacement of red cells should aim to maintain haemoglobin levels around 10–12 g/dl. Platelets and coagulation factors (usually fresh frozen plasma and cryoprecipitate) should be replaced as required in order to prevent bleeding.
  - Hydration will usually be via intravenous route but nasogastric feeding is appropriate if tolerated. Urine output should be monitored (by indwelling catheter if conscious level depressed). Insert nasogastric tube for gastric drainage if there is persistent vomiting or where conscious level is decreased.

Suspected raised intracranial pressure

- **THIS IS A MEDICAL EMERGENCY**
  - Assess ABCD, give high flow oxygen (10 litres/min), and obtain intravenous/intraosseous access.
  - Treat shock (see above), if present, but exercise caution with fluid therapy.
- **DO NOT PERFORM LUMBAR PUNCTURE**
  - Give mannitol 250–500 mg/kg IV (this should be repeated if signs of raised intracranial pressure persist up to a maximum total dose of 2 gm/kg or if available a serum osmolality upto 325 mosm/l). If mannitol is, unavailable give furosemide 1 mg/kg IV.
- **IF SIGNS OF RAISED INTRACRANIAL PRESSURE PERSIST**

  Where signs persist despite the above therapy, ideal management would include:
  - Rapid sequence induction of anaesthesia and intubation for both airway protection (if Glasgow Coma Score <8 and/or child is unresponsive to painful stimuli) and stabilisation of P_{CO_2}
  - ***Mechanical ventilation with optimal sedation and maintenance of P_{CO_2} within the normal range (ideally between 4 and 4.5 kPa)***

  Other useful techniques include:
  - Placing patient supine with a 30° head-up position
  - Avoidance of central venous catheters in internal jugular veins
  - Antipyretics to maintain normal temperature
  - Full neurological and cardiovascular assessment with regular (at least hourly) assessment of: pupillary

responses, conscious level, pulse, blood pressure, capillary refill time, respiratory rate and effort (pulse oximetry if available) and temperature
- Monitoring electrolytes, gases, clotting and full blood count as recommended for shock.

Prognosis

- Even with optimal intensive care, around 5–10% of patients with meningococcal septicaemia will die. Where intensive care is unavailable this may rise to more than 40%.
- Mortality of meningitis is generally much lower (around 2%).
- Most frequent complication of meningitis is hearing impairment or deafness which may affect up to 10% of survivors.
- Survivors of septicaemia may require skin grafting of necrotic lesions and amputation of necrotic digits or limbs.
- In general, most survivors make a virtually complete recovery although subtle neurological abnormalities (for example behavioural and developmental problems, mild motor abnormalities) are not uncommon.

Prevention of meningococcal disease

Education

Increasing awareness of primary healthcare workers and general public to the presenting symptoms of meningococcal disease and emphasising need for early presentation and treatment may have a major impact on mortality and morbidity.

Prophylaxis of contacts

- Transmission is via droplet spread to close contacts. Around 4–25% of people are colonised at any one time but outbreaks of disease are not generally related to colonisation rate. Household contacts of a case may be at 800 times increased risk of disease than the general population.
- Chemoprophylaxis is used to prevent secondary cases by elimination of nasal carriage. Administer as soon as possible (within 48 hours after presentation of index case) (see Table 4.6.4).
- Follow local public health guidelines when determining who should receive antibiotic prophylaxis. In general only immediate family (or those sharing accommodation) and kissing contacts should be treated. Health care workers should receive prophylaxis only where they have experienced extensive contact with patient's respiratory secretions, for example during intubation. Doses are shown in Table 4.6.4.

Table 4.6.4 Antibiotic prophylaxis in meningococcal disease

Antibiotic	Route	Dose
Rifampicin	Oral	<1 month 5 mg/kg twice daily for 2 days 1 month–12 years 10 mg/kg twice daily for 2 days >12 years 600 mg twice daily for 2 days
Ceftriaxone	IV	<12 years 125 mg ⩾12 years 250 mg (suitable for pregnant women)
Ciprofloxacin	Oral	500 mg single dose (adults only)

- Where index case has proven serogroup A or C disease, consideration should be given to vaccinating close contacts with appropriate polysaccharide or polysaccharide conjugate vaccine.
- During larger outbreaks or epidemics, wider scale prophylaxis is occasionally used but should only be carried out under guidance of local/national public health authorities. Public education regarding presenting symptoms of meningococcal disease and emphasising need for early presentation may be more beneficial than wide-scale distribution of antibiotics.

Vaccination

Vaccines based on the capsular polysaccharide of serogroups A and C (±Y and W135) have been available for several years and have been used for vaccination of contacts (as above) and for protection of travellers to endemic areas. They are unable to reliably induce immunity in infants and are not generally used for population vaccination campaigns except in epidemic situations. Where widespread epidemics of meningococcal disease occur (for example in meningitis belt in sub-Saharan Africa) mass vaccination campaigns have proved useful in reducing attack rate; such campaigns are administered by local public health authorities.

New conjugated polysaccharide vaccines for serogroups A and C are now becoming available and offer the possibility of inducing immunity in all age groups. C-conjugate vaccine has been recently included in the routine immunisation schedule for infants in the UK. A broadly effective vaccine for serogroup B disease remains elusive.

4.7 Pertussis

Alan Smyth

Minimum standards requirements

- Immunisation (see Chapter 1.29)
- Erythromycin
- Oxygen (see Chapter 3.1) plus close observation
- Anticonvulsants (see Chapter 3.38)
- Prophylaxis and immunisation for unimmunised siblings

Infection with the organism *Bordetella pertussis* (a Gram-negative bacillus) causes a clinical syndrome commonly referred to as "whooping cough". The illness classically has three stages:

Stage 1: **Catarrhal stage** (1–2 weeks). The symptoms are those of an upper respiratory infection.

Stage 2: **Paroxysmal stage** (2–4 weeks). The child has severe episodes of coughing – usually up to 10 coughs without drawing breath – and then a sharp inspiration or "whoop". The prolonged coughing (often with vomiting) may lead to poor feeding, with weight loss and sometimes rectal prolapse. Other complications such as subconjunctival haemorrhages and ulceration of the frenulum may develop.

Stage 3: **Convalescent stage** (1–2 weeks). The episodes of coughing subside. Occasionally the child may continue to cough for months.

✔ **Pertussis should be prevented by universal infant immunisation**.

Effects on the young infant

Infants may become infected with pertussis before they have been immunised or if immunisation is not available (or the parents have refused). Young infants with pertussis have a different clinical picture:

- Apnoea with hypoxaemia
- Bradycardia
- Seizures
- Cough and poor feeding.

Diagnosis

The laboratory facilities needed to diagnose pertussis are not available in many hospitals. ***Culture from a pernasal swab should be undertaken on Bordet–Gengou medium***.

An absolute lymphocytosis (with a typical clinical picture) is highly suggestive (the total lymphocyte count may be over 30×10^9/litre).

Treatment

The following groups of children should be admitted:

- Infants <6 months
- Complications such as: pneumonia, convulsions, dehydration or severe under-nutrition
- Apnoea or cyanosis.

Supportive treatment

- Maintain nutrition and hydration.
- Give oxygen according to the criteria for ALRI in Chapter 3.1.
- Give gentle suction of secretions (avoid triggering coughing).
- Low dose continuous oxygen (0.5–1.0 litres/minute) via nasal cannulae may reduce apnoeic episodes in infants. Do not use nasopharyngeal cannulae which can provoke coughing spasms.
- Do not give cough suppressants, sedatives or antihistamines.
- Encourage breastfeeding. If the infant cannot drink, pass a nasogastric tube.
- If there is severe respiratory distress, consider intravenous maintenance fluids to avoid aspiration but avoid malnutrition.

Specific treatment

- Treat pneumonia, complicating pertussis, according to the ALRI protocol (see Chapter 3.1)
- Give DPT vaccine to any unimmunised siblings.
- Treat convulsions as in Chapter 3.38.
- Erythromycin will shorten the illness, if given during the catarrhal stage and may prevent spread to siblings or other patients. The oral dose is 10 mg/kg 6 hourly. Also give erythromycin to any infant in the family <1 year who has respiratory symptoms.

4.8 Streptococcal disease

James Tumwine

Minimum standards requirements

- Antibiotics (penicillin/erythromycin)
- For resistant pneumococci (cefotaxime/ceftriaxone, vancomycin)
- Pneumococcal vaccine postsplenectomy and in sickle cell disease

Streptococci are Gram-positive bacteria and are classified into I, J, or nonhaemolytic groups.

Other groups include E, F, G, H, K, L, M, N, O and V.

Group A streptococci (GAS)

- Important bacteriological cause of upper respiratory tract infection (URTI) and skin infections, for example impetigo. Complications of URTI include cervical lymphadenitis and retropharyngeal abscess and of skin infection, erysipelas.
- Rheumatic fever follows throat infections and is associated with streptococcal serotypes such as 1, 3, 5, 6, 18, 19 and 24.
- Acute glomerulonephritis may follow throat or skin (more commonly in disadvantaged countries) infections and is associated with serotypes such as 12, 49, 55 and 57.
- Scarlet fever presents with tonsillitis and a characteristic rash, circumoral pallor and strawberry tongue. Rash starts with generalised erythema which is punctate, i.e. like sand paper and palpable, followed by desquamation.
- Risk factors for severe GAS infection such as necrotising fasciitis include varicella, HIV, diabetes mellitus, chronic heart or respiratory disease.
- Diagnosis is by culture, for example throat, blood, skin and other tissues. ***Rapid antigen tests are available. Antistreptolysin (ASOT) titres are mainly raised following throat infections and anti-DNase following skin infections.***

Treatment

Resistance to penicillin is rare. Erythromycin is used if there is allergy to penicillin. Treatment of carriers is not recommended except during outbreaks of acute glomerulonephritis, rheumatic fever and recurrent GAS infections, despite adequate therapy.

Group B streptococci (GBS)

- Causes chorioamnionitis, urinary tract infections and endometritis in women. Pneumonia, septic arthritis, osteomyelitis, septicaemia and meningitis in newborn and older infants (range 1 day to 3 months).
- Neonates may be infected perinatally or in the nursery (nosocomial). Risk factors for GBS disease include prematurity (<37 weeks of gestation), premature rupture of membranes, maternal intrapartum fever, chorioamnionitis and urinary tract infection of mother (see Chapter 3.48 on neonatal infections).

Table 4.8.1 Streptococci and associated conditions

Streptococci	Group (Lancefield)	Reaction (Haemolytic)	Disease caused
S. pyogenes (GAS)	A	J	Tonsillitis, pyoderma, impetigo, scarlet fever (subsequent rheumatic fever, acute glomerulonephritis) Necrotising fasciitis, toxic shock syndrome
S. agalactiae (GBS)	B	J	Neonatal sepsis/meningitis
S. equisimillis (GCS)	C	J	Endocarditis, pneumonia, cellulitis, septicaemia
S. faecalis (GDS)	D	J or none	Normal gut flora. May cause peritonitis, urinary tract infection, endocarditis and septicaemia
S. viridans	–	I	Mouth commensal. May cause endocarditis, dental caries
S. pneumoniae	–	–	Pneumonia, meningitis, otitis media, sinusitis

Streptococcus pneumoniae

Gram-positive diplococcus (lancet shaped). At least 85 pathogenic serotypes are known. Types 1, 3, 4, 6, 7, 8, 9, 12, 14, 18, 19 and 23 are the most virulent.

- Common infections include pneumonia, meningitis, peritonitis, otitis media, sinusitis, arthritis and conjunctivitis.
- Pneumococcal infections are more common in children with defective splenic function, for example sickle cell anaemia, splenectomy; also nephrotic syndrome, chronic renal failure, diabetes mellitus, malabsorption, heart failure, skull fracture, neurosugery and those with congenital or acquired immunodeficiency such as agammaglobulinaemia, and HIV infection.
- Patients with white blood cell counts of more than 15×10^9/litre are likely to have bacteraemia.
- Culture of *Str. pneumoniae* from the respiratory tract is not useful because of asymptomatic carriers. ***Rapid tests for detecting capsular antigen in CSF, serum, urine and joint/pleural aspirate are available, and are useful in children who have commenced antibiotics.***

Treatment

- In last two decades resistance of *Str. pneumoniae* to antibiotics such as penicillin and chloramphenicol has emerged.
- In many countries up to 5–40% of isolates may be resistant to penicillin G.
- If resistance to chloramphenicol or penicillin is suspected, give either cefotaxime or ceftriaxone. If resistance to these two drugs is considered, add vancomycin to ceftriaxone or cefotaxime. If results of sensitivity confirm susceptibility to penicillin G, ceftriaxone or cefotaxime, then vancomycin may be stopped.

Pneumococcal vaccine

- A polyvalent vaccine (23 serotypes) is available to protect children over 2 years of age susceptible to invasive pneumococcal disease, for example sickle cell disease, splenectomy.
- Dose is 0.5 ml subcutaneously or intramuscularly. Side-effects include fever, myalgia and pain at injection site.
- Revaccination is recommended 3–5 years after initial dose in children younger than 10 years who are at risk. Value of revaccination is uncertain.
- A pneumococcal conjugated vaccine has been developed which is effective in children <2 years and is more effective than the above.

Chemoprophylaxis

- Daily oral penicillin V (125 mg twice daily for <5 years, 250 mg twice daily for older children) is recommended for children at risk, for example sickle cell disease, splenectomy. Emergence of penicillin-resistant pneumococci limits its value. Chemoprophylaxis to prevent endocarditis is discussed in Chapter 3.5.

Other groups of streptococci (C, D, E, F, G, H, K, L, M, N, O and V)

Cause diseases such as infective endocarditis, urinary tract infection and pneumonia. Susceptibility to penicillin is variable and treatment with an aminoglycoside (such as gentamicin) and penicillin G or ampicillin is recommended.

Table 4.8.2 Drugs for the treatment of pneumococcal disease

Disease	Antibiotic	Dose (/kg) and route	Dose interval	Duration/comments
Otitis media	Amoxicillin (oral)	12.5 mg/kg oral	8 hours	5–7 days
	Amoxicillin – clavulanic acid	12.5 mg/kg oral	8 hours	
	Cefaclor	12.5 mg/kg oral	8 hours	
	Erythromycin	12.5 mg/kg oral	6 hours	5–10 days
Sinusitis	As for otitis media	As for otitis media	As for otitis media	As for otitis media
Meningitis	Penicillin G	50 mg/kg IV	4–6 hours	10–14 days for all antibiotics below
*	Chloramphenicol	Load 50 mg/kg IV then 25 mg/kg	6 hours	
	Cefotaxime	50 mg/IV	6–8 hours	Maximum single dose 4 g
	Ceftriaxone	100 mg IV	24 hours	Maximum single dose 4 g/day
	Vancomycin	Load 15 mg/kg IV then 10 mg/kg IV	6 hours	Total daily dose not >2 g Drug levels needed
	Meropenem	20–40 mg/kg slow IV injection over 5 minutes	8 hours	Maximum single dose 2 g

4.9 Tetanus

Christiane Ronald and David Southall

Minimum standards requirements

- Immunisation and prevention (see Chapter 1.29)
- ABCD (see Chapter 1.19) especially airway protection
- Antitetanus immunoglobulin
- Diazepam, chlorpromazine and midazolam for acute spasms
- Morphine (see Chapter 1.27)
- Early IV penicillin
- Intensive/close observational care including ECG monitoring/tracheostomy IPPV and paralysis (if available)

Causes 800 000–1 million deaths/year worldwide (about 400 000 in the neonate). For infection to occur, two conditions must be met:

- A wound with a degree of necrosis
- A wound contaminated with material containing *Clostridium tetani* (a Gram-positive obligate anaerobe widely distributed in the environment).

The umbilical stump is a common site of entry for neonatal tetanus which carries up to 60–80% mortality. Ear piercing in neonates is also a common cause (for example in Vietnam). In up to 30% of infected children no wound can be found.

Cases of tetanus in older children follow small puncture wounds, accidents and trauma in the partial or unvaccinated child.

Once the *C. tetani* spore is inoculated into necrotic tissue with a low oxygen concentration it changes into a vegetative form, which elaborates tetanospasmin which ascends peripheral nerves to the spinal cord where it binds to cerebral gangliosides and impairs inhibitory synapses. This causes muscle rigidity, spasm and autonomic overactivity.

The clinical presentation depends upon the distance the injury is from the spinal cord, the longer the incubation period which varies from 3–21 days. The shorter the incubation period and the time from onset of symptoms to the first spasm, the worse the outcome.

More than 90% of patients develop trismus ("locked jaw"). As the disease progresses, spasm of other muscle groups occurs. The **spasms are extremely painful**, may be prolonged, giving rise to opisthotonus. The sympathetic system can be affected causing lability of temperature, blood pressure and cardiac function.

The approach to treatment given in this chapter is appropriate for both neonatal and childhood tetanus.

Clinical presentation

A previously well neonate, presents at 3–20 days with irritability, decreased sucking, trismus, muscle spasms or convulsions.

An older child presents following a minor injury or bite. Some infections follow chronic otitis media.

Management of established tetanus

Management is targeted at:

- Neutralising existing toxin and preventing its further production
- Control of spasms
- Prevention of complications.

On admission

- Secure and maintain the airway, ensure adequacy of ventilation.
- If the child is in **acute spasm**, this should be terminated by giving **diazepam by bolus IV infusion over 15 minutes (dose 200 micrograms/kg) or rectally (400 micrograms/kg)**. Ensure that for intravenous infusion, diazepam is diluted to 100 micrograms/ml and that extravasation does not occur (very irritant).
- An intravenous line should be secured immediately.
- Pass a nasogastric tube in order to give fluids, food and drugs with minimal disturbance. Feeds need to be given frequently (ideally hourly) and in small amounts due to reduced gut motility.
- Any obvious wound should be debrided and cleaned and previously ill-advised sutures should be removed (see Chapter 5.3).
- Finally the disease itself does not induce immunity, so after recovery tetanus vaccine should be given for future prevention.

Antibiotics

Penicillin should be given to eradicate toxin-producing *C. tetani*. **Benzylpenicillin 50 mg/kg every 6 hours IV or, if not possible, IM, should be given for** 48 hours and then, if enteral antibiotics are possible, oral penicillin 12.5 mg/kg 6 hourly for 7 days. If not possible continue benzylpenicillin. Metronidazole is used in some centres for older children.

Associated septicaemia is not uncommon in the neonate and additional broader spectrum antibiotics will often be required (see Chapter 3.48 for treatment). Nosocomial infections are also common, especially pneumonia.

Neutralisation of toxin

Antitetanus human immunoglobulin is the preparation of choice for neutralising unbound tetanospasmin. It is given by intravenous infusion over 30 minutes at a dose of **5000–10 000 units immediately on admission**. Adverse reactions are rare. **Local instillation is of no benefit and neither is intrathecal administration**.

For neutralisation of the toxin, HTIG is not available in most countries where it is needed. An equine immunoglobulin may be available and is used, 500–1000 units/kg IM (maximum dose 20 000 units). There is a risk of anaphylaxis (see Chapter 1.23 for management). Epinephrine must be immediately available.

Management of spasms and hypertonicity

- Spasms can usually be controlled by slow IV injection of diazepam 200 micrograms/kg every 3–6 hours or by continuous infusion of midazolam (30–100 micrograms/kg/hour).
- If this is not possible, nasogastric diazepam 250–500 micrograms/kg 6 hourly alternating with chlorpromazine 500 micrograms/kg 6 hourly can be given. The first dose of chlorpromazine can be given as a bolus IM if spasms are severe.
- Paracetamol 25 mg/kg 6 hourly for pain (20 mg/kg in the neonate). If this is insufficient the WHO pain ladder approach should be adopted. Oral or IV morphine may be needed (see Chapter 1.27).

Alternative antispasmodic or sedative drugs

- ***Baclofen for children >1 year (start at 750 micrograms/kg/24 hours and increase to 2 mg/kg/24 hours in three divided doses)***
- Phenobarbitone (15 mg/kg in one or two divided doses) as loading dose then 5 mg/kg/day
- Paraldehyde (0.4 ml/kg rectally in arachis oil or 0.9% saline repeated 4–6 hourly) in older child but repeated once in the neonate.

Ventilation and prevention of complications

- Many patients have major problems with pharyngeal spasms/upper airway obstruction and are best managed with a tracheostomy and pharmacological control of the spasms (sometimes the tracheostomy may need to be undertaken as an emergency procedure). Up to a third of those needing a tracheostomy do not require ventilation.
- Intubation can be very difficult because of pharyngeal/laryngeal spasm and often a mini-tracheostomy without prior intubation may be appropriate, providing experts on the procedure and on anaesthesia are present.
- Infusions of morphine and midazolam, alongside muscle relaxants (atracurium/pancuronium), are essential to minimise suffering. (Under no circumstances should paralysis be given without infusions of morphine +/− midazolam).

Neonates rarely receive ventilation because few places where tetanus occurs have appropriate ventilators or staff who know how to intubate or ventilate a child. An alternative is bag and mask ventilation as often as necessary for the apnoeas that occur secondary to bouts of spasms.

- **Good nursing and frequent monitoring with particular attention to suction of secretions from the airway, maintenance of adequate hydration, mouth hygiene, turning of the patient to avoid orthostatic pneumonia and bed sores, will reduce complications**. The child should be nursed in a quiet environment with low level lighting. Sudden loud noises should be avoided. Invasive procedures should be kept to a minimum and preceded by appropriate analgesia/sedation. There must be **continuous** observation by experienced personnel. ✔
- In the intensive care unit, cardiac function should ideally be monitored by ECG to detect toxin-induced arrhythmias and autonomic instability. If arrhythmias occur, sedation with morphine may be helpful (see Chapter 1.27).
- Intensive care of severe cases of tetanus may be necessary for up to 3–4 weeks.

It is important to realise that the child/baby has unimpaired consciousness and is often aware of what is taking place unless heavily sedated.

Prescribe appropriate (regular and frequent) analgesia, as sedatives and antispasmodics alone do not prevent the suffering resulting from painful spasms or painful procedures.

The spasms are also very frightening and distressing for the parents.

- Associated septicaemia is not uncommon in the neonate and additional broad spectrum antibiotics will often be required (see Chapter 3.48). Nosocomial infections are also common especially pneumonia.

Monitoring

- Glucose, urea and electrolytes **(as few blood tests as possible as likely to precipitate spasms)** ✔
- Cardiac monitoring
- Pulse oximetry
- Fluid input/output
- Calorific intake

Prognosis

Prognosis for neonatal tetanus is poor, especially with a short incubation period (<7 days) or with rapid evolution of symptoms. Pyrexia, tachycardia and frequent spasms (>20 in 24 hours) also indicate a poor prognosis. Quality of nursing care and the ***availability of intensive care facilities greatly affect the outcome***.

Prevention

✔ **Every child should receive tetanus vaccine according to the expanded programme of immunisation (EPI). All pregnant women should receive two doses antenatally** and then tetanus toxoid should be given combined with diphtheria and pertussis to infants according to national schedules. Note that both HIV infection and placental malaria reduce the transplacental transfer of antitetanus antibodies in utero. A booster should be given whenever a dirty wound is sustained. Sterile handling of the umbilical cords by midwives or appropriately trained traditional birth attendants should also reduce the incidence of neonatal tetanus.

Further reading

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410 Tuberculosis

Brian Coulter

Minimum standards requirements

- Mantoux, Chest *X* ray, HIV test
- Microscopy of samples for acid-fast bacilli
- Prolonged, observed multidrug therapy
- Dexamethasone
- Tracing, screening and prophylaxis of contacts

Major factors in the global increase in tuberculosis since the mid-1980s include the HIV pandemic, migration of people from countries with a high prevalence of tuberculosis to industrialised countries (particularly refugees), poverty, overcrowding and failure of investment in tuberculosis control programmes. Multidrug resistance is a major concern.

Epidemiology

- In low-income countries risk of developing infection is up to 2.5% per annum
- Age periods of highest risk of developing disease are 0–5 years (especially under 1 year) and at puberty.
- Spread is by untreated smear positive adults who may infect up to 10–15 people per year.
- Children are generally uninfectious, except for adolescents with cavitatory disease.
- Children with untreated tuberculosis contribute to the pool of adults with reactivated disease.

Tuberculosis in adolescence

May result from reactivation of a primary infection, exogenous infection or both. There is a strong hypersensitivity reaction in the lungs with local infiltration and often cavity formation. Pulmonary lymph node enlargement and extrathoracic dissemination is uncommon.

Factors which predispose tuberculosis infected children to develop systemic disease

- Tuberculosis infection in previous two years
- Age <5, especially <1 year
- Close contact
- Intercurrent infection, for example measles, pertussis
- Malnutrition
- Immunosuppression

Tuberculin sensitivity

- This is tested using the Mantoux test.
- Multiple puncture techniques, for example Heaf test, are used for screening populations of children.
- For the Mantoux test, use either 5 or 10 units (depending on the manufacturer and the country's policy) and 2 units for surveys.
- For the Mantoux test inject tuberculin (PPD-S) **intradermally** into the upper third of flexor surface of the forearm with 1 ml syringe and a short bevel gauge 25–27 needle producing a wheal of at least 5 mm. Read transverse diameter of induration at 48–72 hours.

Interpretation

- Regard induration of 0–5 mm as negative, 6–9 mm indeterminate, likely to be associated with environmental mycobacteria, and 10 mm or more indicative of infection with *Mycobacteria tuberculosis* except in the child who has had BCG in the previous few years when induration of 15 mm is required.
- In disadvantaged countries where BCG is given at birth, many infants may have a negative tuberculin test by 3–4 years of age; and thus an induration of 10 mm in children this age and older may be regarded as supportive of *M. tuberculosis* infection.
- Negative or reduced response to tuberculin occurs in malnutrition, immunosuppression associated with HIV

or other immunodeficiency states, recent viral or some bacterial diseases such as pertussis, overwhelming tuberculosis and non-respiratory tuberculosis; thus an induration of 6–9 mm may be indicative of tuberculosis.

Pathogenesis

- Inhalation of the tubercle bacillus into an alveolus establishes the primary (*Ghon*) focus. In 4–8 weeks before the cell-mediated immune response occurs, there is spread to regional lymph nodes and small numbers of bacilli disseminate throughout the body in the lymphohaematogenous system.
- Certain organs favour survival of tubercle bacilli, for example regional nodes, epiphyseal lines of bones, cerebral cortex, renal parenchyma and apical regions of the lungs (Simon focus).
- Establishment of an adequate cell-mediated immune response (which coincides with appearance of sensitisation to tuberculin) in most cases results in control or eradication of proliferating tubercle bacilli at these sites.
- Primary focus is seldom detected on chest *X* ray; enlarged hilar/paratracheal nodes or parenchymal complications are the usual evidence of the primary complex.
- **Primary tuberculosis of the lung is** usually a manifestation of lymphobronchial disease, with local compression or erosion of the bronchi. **Extrathoracic disease** is due to local spread of disease at metastatic sites, for example lymph nodes, brain, bone, kidney and abdomen.
- Dissemination of large numbers of tubercle bacilli may result in acute miliary disease or less commonly a chronic disseminated (cryptic) tuberculosis.
- Erythema nodosum and phlyctenular conjunctivitis are hypersensitivity reactions which may occur during primary tuberculosis.
- Risk of developing symptomatic disease following primary tuberculosis is highest in the first 1–2 years after infection.

Clinical features

In rich countries a majority of children with respiratory tuberculosis are **asymptomatic** and are picked up through contact tracing and will generally have early primary disease. In poor countries, only children with **symptomatic** disease present and are thus only the tip of the iceberg.

Clinical features of children presenting with tuberculosis

Symptoms	Physical signs
Cough	
Wheeze (young children)	Lymphadenopathy*
Fever	Hepatosplenomegaly*
Weight loss	Malnutrition*
Anorexia	
Diarrhoea*	
Lethargy	

* Common in HIV-infected children

HIV and tuberculosis

- Features of tuberculosis in children with perinatally acquired HIV infection are not well defined.
- Many HIV-infected infants probably succumb to bacterial infections and *Pneumocystis carinii* pneumonia before contracting tuberculous infection.
- In older children there is difficulty in diagnosis due to the following: there is often a negative tuberculin response, confusion with HIV-related chest disorders including lymphocytic interstitial pneumonitis (LIP), superimposed viral/bacterial infections, chronic interstitial pneumonitis, Kaposi sarcoma and bronchiectasis; and lack of facilities to culture *M. tuberculosis.*
- HIV/tuberculosis co-infected children are more likely to develop disseminated tuberculosis and meningitis, and may have a poor response to treatment and a higher mortality.
- Because of the difficulty in confirming tuberculosis in symptomatic HIV-infected children, many children probably receive unnecessary tuberculous chemotherapy.
- Finger clubbing may be seen in chronic tuberculosis and is common in HIV-related pulmonary disorders.

Respiratory tuberculosis

- Most respiratory tuberculosis results from complications of lymphobronchial disease and includes segmental lesions, consolidation, collapse and obstructive emphysema.
- In young children small cavities may develop during the course of primary (especially progressive) tuberculosis but they are classically seen in the adolescent period.
- Large pleural effusions usually occur in older children and adolescents.
- Radiological features of pulmonary tuberculosis may be atypical in HIV infection and malnutrition.

Pericarditis

Tuberculosis should be considered in all cases of pericarditis. *M. tuberculosis* may be cultured from an aspirate in over half the cases.

Lymph node disease

- May result from a focus in the upper lung fields or from haematogenous spread.
- Diagnosis may be made by biopsy or needle aspiration.
- Swelling and softening of nodes may continue for months after treatment has been completed.
- In rich countries, environmental mycobacteria are now a far commoner cause of chronic granulomatous disease of cervical lymph nodes than tuberculosis in indigenous young children.

Miliary tuberculosis

- Is commonest in young children and in those immunosuppressed usually occurring within 3–12 months of primary infection.

- Chest *X* ray (except in early stages) will demonstrate a "snow storm" appearance.
- Meningitis is a common complication.

Meningitis

- Commonest in children under 5 years and often occurs within 6 months of infection.
- Onset is usually insidious and diagnosis is often delayed. Late diagnosis is invariably complicated by neurological dysfunction or death.
- CSF: cell count is usually less than 500 per mm^3 and mainly lymphocytic but polymorphoneutrophils may be prominent early on which may cause confusion with partially treated bacterial meningitis. Protein is usually raised (0.8–4 g/litre) and glucose low.
- ***Brain imaging, for example CT or MRI*** should be undertaken at diagnosis and 3–4 months, and at any time there is neurological deterioration to detect complications such as hydrocephalus and tuberculomata.
- Management: isoniazid (15–20 mg/kg once daily orally, IM or slow IV injection maximum 500 mg daily), rifampicin (20 mg/kg once daily orally or IV infusion over 2–3 hours maximum 600 mg daily) and pyrazinamide (40 mg/kg once daily orally maximum 2 gm daily) are given for at least 6 months; including pyrazinamide for 6 months in severe cases. If drug resistance is suspected ethionamide (20 mg/kg) may be added as fourth drug. Ethambutol 15–20 mg/kg/day (maximum 1.5 g daily) orally and streptomycin (15 mg/kg by deep IM injection) once daily maximum dose 1 g are alternative fourth drugs but their value in meningitis is probably limited. Except in very mild cases (stage I), corticosteroids should be given. Dexamethasone 0.6 mg/kg/day in two divided doses or prednisolone 3–4 mg/kg/day is given for 2–3 weeks and tailed off over 2–3 months. ***A ventriculoperitoneal shunt may be required for obstructive hydrocephalus***.

Bone and joints

- Frequently missed at the early stages because of a low index of suspicion.
- Spine is affected in half the cases followed by knee, hip and ankle. The most serious complication is spinal compression.
- Diagnosis is made by histology. Ziehl–Neelsen stain and culture of tissue may be positive and ***if in doubt specimens should be sent for polymerase chain reaction***.

Abdominal tuberculosis

- May present with ascites, abdominal nodes or masses, or diarrhoea with or without abdominal pain.
- Diagnosis is usually made on bacteriological examination of ascitic fluid or a biopsy.
- Ultrasound and ***CT/MRI are helpful***.

Perinatal tuberculosis

- Congenital tuberculosis is rare but should always be considered in sick neonates/infants especially in areas where HIV/tuberculosis co-infection is common.
- If a mother has completed tuberculosis chemotherapy during pregnancy or has inactive disease her infant should be given BCG at birth. If she has active disease or is still requiring treatment, the infant should be given isoniazid 6 mg/kg once daily for 3–6 months. A tuberculin test and chest *X* ray is then performed. If negative, BCG is given, if positive, full investigations for tuberculosis are undertaken. If no evidence of infection is detected isoniazid and rifampicin are given for 3–4 months. If tuberculosis is suspected, full treatment is given (see Tables 4.10.1 and 4.10.2 on management).

Diagnosis

Standard methods for diagnosis are the tuberculin test and a chest *X* ray. Gastric aspiration or sputum induction (see below) and bronchoscopy are reserved for difficult and complex cases, and when drug resistance is suspected. In poor countries the tuberculin test is often negative (or unavailable) and the chest *X* ray may not be easy to interpret due to poor quality films. Many children are given a "therapeutic trial" of tuberculosis chemotherapy and thus tuberculosis is often overdiagnosed, especially in areas with high HIV prevalence.

Diagnosis of infection (see above)

Mantoux induration of $\geqslant$10 mm in children who have not received BCG vaccination or $\geqslant$15 mm in children who have received BCG recently.

Diagnosis of disease

Tuberculosis is demonstrable, for example by chest *X* ray and/or child is symptomatic.

- Gastric aspiration should be undertaken in the early morning while child is recumbent. Ziehl–Neelsen smear of gastric aspirate is seldom positive and culture is positive, under optimal conditions, in only 30–50% of cases.
- Alternative methods are laryngeal swabs, and sputum induction using nebulized 3% hypertonic saline. Neither has a higher sensitivity than gastric aspiration.
- **The polymerase chain reaction on histological specimens may be useful** but does not appear to be any more sensitive than culture of gastric aspirates (at least at present)
- Young children, especially those who are sick, malnourished or deteriorating, or where tuberculous meningitis is suspected, should be considered for treatment even though investigations are inconclusive. In other cases with pulmonary disease where the diagnosis is not clear, a course of appropriate antibiotics should be given and the chest *X* ray repeated after a

month. If there is no improvement or deterioration a full course of anti-tuberculosis chemotherapy is given and progress carefully monitored to document response.

- Increase in weight (measured daily) and loss of fever (measured twice daily) indicate response to treatment. If treatment is given for suspected rather than proven tuberculosis, no response to symptoms within 4 weeks suggests tuberculosis is unlikely.

Diagnosis of tuberculosis in young children unable to expectorate sputum

History/examination

- Contact
- Failure to recover from illness
- Weight loss/malnutrition
- Persistent cough (>2 weeks)
- Peripheral lymphadenopathy
- HIV infection

Investigations

- Tuberculin test >10 mm or >5 mm in malnutrition or HIV
- Chest *X* ray: lymphadenopathy collapse/consolidation with or without persistent opacity
- Histology: lymph node or other tissue biopsy
- Smear/culture: gastric aspiration, sputum induction
- Ultrasound: chest/abdomen/lymph nodes/pericardium/brain
- ***CT/MRI***
- HIV antibody tests (if relevant)

Management

- Except in adolescents with cavitatory disease, most tuberculosis in children is paucibacillary (low number of mycobacteria)
- With the exception of meningitis (see above) both pulmonary and extrapulmonary tuberculosis may be treated with standard six-month chemotherapy, viz. isoniazid, rifampicin and pyrazinamide for two months and isoniazid and rifampicin for the remaining four months. In countries unable to afford rifampicin for six months' therapy, ethambutol is substituted for rifampicin for the continuation phase and the total period of treatment is eight months. Ethambutol should not be given in a dose higher than 15 mg/kg/day to children under 5 years, as they may be unable to report visual disturbance associated with optic neuritis. **Thiacetazone must be avoided in children who might possibly have HIV infection**.
- Presently DOTS (directly observed therapy short course) is not generally practised for children as it is presumed that parents will supervise treatment but where DOTS is practised in the community it may be appropriate to include children.

Table 4.10.1 Regimens for treatment of tuberculosis

Regimens	Duration
Standard daily	
Isoniazid, rifampicin, pyrazinamide (ethambutol):* 2 months, then isoniazid, rifampicin: 4 months	6 months
Isoniazid, rifampicin: 9 months*	9 months
Isoniazid, rifampicin, pyrazinamide 2 months, then isoniazid, rifampicin	4 months**
Intermittent thrice weekly	
Isoniazid, rifampicin, pyrazinamide 2 months, then isoniazid, rifampicin thrice weekly: 4 months	6 months
Alternative less potent daily	
Isoniazid, rifampicin, pyrazinamide (ethambutol):* 2 months, then isoniazid, ethambutol: 6 months	8 months
Isoniazid, rifampicin, pyrazinamide (ethambutol):* 2 months, then isoniazid, thiacetazone: 6 months	8 months

* Add ethambutol in smear-positive cases or if drug resistance is suspected. Streptomycin is an alternative.

** For hilar lymphadenopathy alone.

Reproduced and adapted from *Forfar and Arneil's Textbook of Paediatrics*, Churchill Livingstone, 5th edn, 1998.

- Most children are treated with daily chemotherapy. Doses for thrice weekly intermittent chemotherapy are outlined in Table 4.10.2. If pyrazinamide is not given, isoniazid and rifampicin should be given for 9 months. If sputum is smear-positive or resistance is suspected, ethambutol (or streptomycin) should be given as a fourth drug during the initial phase, at least.
- Adverse reactions to tuberculosis chemotherapy are uncommon and if they occur it is usually within 6–8 weeks of starting treatment. Liver transaminases may increase two- to threefold during treatment with isoniazid and rifampicin, but drug therapy may be continued if there is no jaundice or symptoms of liver toxicity, for example nausea, vomiting, malaise or liver tenderness. Viral hepatitis should be considered if jaundice occurs.
- Adjunct treatment with corticosteroids is of value in meningitis (see above) and may enhance resolution of disease in lymphobronchial disease, pericarditis, pleural effusion and severe miliary disease with alveolar capillary block. Prednisolone 1.5 mg/kg/day is given for 2–3 weeks and then tailed off over 6–8 weeks.
- Avoid streptomycin where possible: injections painful, irreversible auditory nerve damage may occur and increased risk for HIV from mishandling of syringes and needles.
- Avoid ethambutol in young children who cannot report a deterioration in sight or colour perception. Maximum dose of ethambutol is 15 mg/kg/day.
- **Avoid thiacetazone in HIV** or strongly suspected HIV infection where there is a high risk of severe (**sometimes fatal**) skin reactions.

Table 4.10.2 Recommended drugs in the treatment of tuberculosis

Drug	Daily dose			Thrice-weekly dose			Side-effects
	Children	Adolescents		Children	Adolescents		
		<50 kg	>50 kg		<50 kg	>50 kg	
Isoniazid	6 mg/kg orally, 1M, IV 15–20 mg/kg (meningitis)	300 mg	300 mg	15 mg/kg (max. 900 mg)	15 mg/kg	max. 900 mg	Hepatic enzyme elevation, hepatitis, peripheral neuropathy, hypersensitivity
Rifampicin	10 mg/kg orally, IV 20 mg (meningitis)	450 mg	600 mg	15 mg/kg (max. 600 mg)	15 mg/kg	max. 900 mg	Orange discoloration of secretions and urine (also contact lens), nausea, vomiting, hepatitis, febrile reactions, thrombocytopenia
Pyrazinamide	30–35 mg/kg orally 40 mg/kg (meningitis)	1.5 g	2.0 g	50 mg/kg	2.0 g	2.5 g	Hepatotoxicity, hyperuricaemia, arthralgia, gastrointestinal upset, skin rash
Ethambutol	15–20 mg/kg orally*	15 mg/kg (max. 2.5 g)	15 mg/kg (max. 2.5 g)	30 mg/kg	30 mg/kg	max. 2.5 g	Optic neuritis, skin rash
Streptomycin	15–20 mg/kg IM	750 mg	1.0 g	15–20 mg/kg	750 mg	1.0 g	Ototoxicity, nephrotoxicity
Thiacetazone	4 mg/kg orally	150 mg	150 mg	Not recommended	Not recommended	Not recommended	Gastrointestinal disturbance, vertigo, visual disturbance, hepatitis, agranulocytosis, exfoliative dermatitis in HIV infection
Ethionamide	15–20 mg/kg orally (divided doses). In meningitis 20 mg/kg	750 mg	1.0 g	Not recommended	Not recommended	Not recommended	Gastrointestinal disturbance, hepatoxicity, allergic reactions

* In young children give 15 mg/kg throughout.
Reproduced and adapted from Forfar and Arneil's *Textbook of Paediatrics*, Churchill Livingstone, 5th edn, 1998.

Prevention

- Diagnosis and treatment of "smear-positive" tuberculosis in adults combined with contact tracing is the key to prevention of childhood tuberculosis.
- Tuberculin-positive children with normal chest *X* rays should be given prophylaxis, either isoniazid and rifampicin for 3 months or isoniazid alone for 6 months. The age limit for prophylactic therapy depends on national policy, for example under 5 years in low-income countries.
- Neonatal BCG may reduce the risk of tuberculosis meningitis and disseminated disease by 60–80% but has a limited effect on prevention of pulmonary disease.

4.11 Typhoid and paratyphoid

Zulfiqar A Bhutta

Minimum standards requirements

- Blood culture and full blood count
- Antibiotics: chloramphenicol, amoxil, cefotaxime/ceftriaxone, ciprofloxacin
- Dexamethasone
- Public health measures: sanitation and immunisation

Typhoid

Epidemiology

Despite vast advances in public health and hygiene in much of the advantaged world, typhoid fever continues to plague many poor countries. Although accurate community-based figures are unavailable, it is estimated that over 16 million cases occur annually with the vast majority of cases in Asia with over 0.6 million deaths. Recent economic and political upheavals in parts of Asia have led to an upsurge in this disorder with several large-scale outbreaks. Population-based incidence rates are estimated at 500–1000 cases per 100 000 population in endemic areas.

In recent years typhoid fever has been notable for the emergence of drug resistance. The first cases of chloramphenicol-resistant typhoid emerged in early 1970s, followed by the emergence of multidrug-resistant (MDR) typhoid (resistant to ampicillin, chloramphenicol and trimethoprim-sulphamethoxazole (co-trimoxazole)) in the mid-1980s. Over the last two years however, the development of quinolone resistance in *Salmonella typhi* from

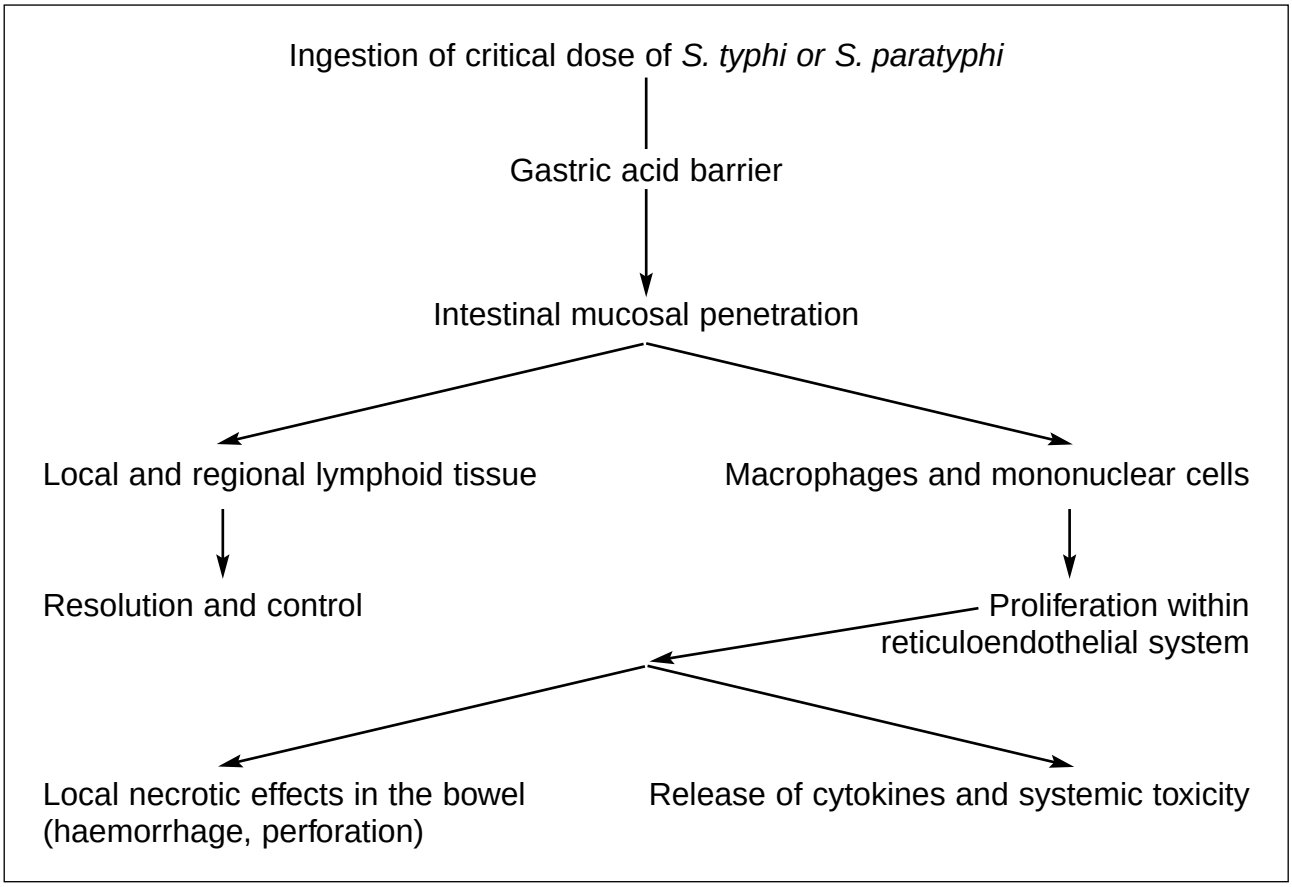

Figure 4.11.1 The pathogenesis of typhoid.

various parts of Asia has raised the extremely worrying prospect of a "super-resistant" variant of typhoid.

In contrast to classic descriptions of milder disease, because of increasing drug resistance in *Salmonella paratyphi*, paratyphoid fever is now of comparable severity and virulence to typhoid. Both types of illness will therefore be described.

Pathogenesis

The disease occurs by the ingestion of a Gram-negative flagellar organism *Salmonella enterica* serovar Typhi (*S. typhi*). A larger infecting dose leads to a shorter incubation period and more severe infection.

The organism crosses the intestinal mucosal barrier after attachment to the microvilli by an intricate mechanism involving membrane ruffling, actin rearrangement and internalisation in an intracellular vacuole. Once inside the intestinal cells, *S. typhi* find their way into the circulation and reside within the macrophages of the reticuloendothelial system (Figure 4.11.1).

The clinical syndrome is produced by a release of proinflammatory cytokines (the interleukins IL-6 and IL-1β; and tumour necrosis factor-α, TNF-a) from the infected cells leading to fever, rigors, inanition, anorexia etc.

The local effects such as intestinal haemorrhage and perforation are comparatively rare in childhood as there is relative lymphoid hyperplasia of the intestinal wall. However, malnourished children, especially adolescents, may be at greater risk of these complications.

Clinical features

The classic step-ladder rise of fever is relatively rare in childhood. Much of the presentation of typhoid fever in various geographic locations and populations is tampered by coexisting morbidities and early administration of antibiotics. In malaria-endemic areas and in parts of the world where schistosomiasis is common, the presentation of typhoid may also be atypical. Data in the box from 2000 cases show the common clinical features of typhoid in endemic areas.

Common clinical features of typhoid fever in childhood (Karachi, Pakistan)

High-grade fever	95%
Coated tongue	76%
Anorexia	70%
Vomiting	39%
Hepatomegaly	37%
Diarrhoea	36%
Toxicity	29%
Abdominal pain	21%
Pallor	20%
Splenomegaly	17%
Constipation	7%
Headache	4%
Jaundice	2%
Obtundation	2%
Ileus	1%
Intestinal perforation	0.5%

Although data from South America and other parts of Africa suggest that typhoid may present as a mild illness in young children, this may vary in different parts of the world. There is emerging evidence from South Asia that the presentation of typhoid may be more dramatic in children under 5 years of age, with comparatively higher rates of complications and hospitalisation. Diarrhoea, toxicity and complications such as disseminated intravascular complications are also more common in infancy, with higher case fatality rates. However, some of the other features of typhoid fever seen in adults, such as relative bradycardia, are rare and rose spots may only be visible at an early stage of the illness in fair-skinned children.

It must also be recognised that MDR typhoid appears to be a more severe clinical illness with higher rates of toxicity, complications and case fatality rates. This appears to be a consistent finding and potentially related to the increased virulence of MDR *S. typhi* as well as higher rates of bacteraemia. In endemic areas, therefore, it may be prudent to treat all severely ill, toxic children, especially those requiring hospitalisation, with second-line antibiotics.

Acute perforation of the intestine with haemorrhage and peritonitis can occur. This presents with severe abdominal pain, vomiting, abdominal tenderness, severe pallor and shock. An abscess may form together with enlargement of the liver and spleen. Manage peritonitis as in Chapter 3.49.

Diagnosis of typhoid

The sensitivity of blood cultures in diagnosing typhoid fever in many parts of the developing world is limited, as microbiological facilities may be basic and widespread antibiotic prescribing may render bacteriological confirmation difficult. Although bone marrow and ***duodenal fluid cultures*** may increase the likelihood of bacteriological confirmation of typhoid, these are difficult to obtain and relatively invasive.

The serological diagnosis of typhoid is also fraught with problems as a single Widal test may be positive in only 50% of cases in endemic areas, and serial tests may be required in cases presenting in the first week of illness. ***Newer serological tests such as a dot-ELISA, co-agglutination and the Tubex® are promising***, but are comparatively expensive and have yet to find widespread acceptability.

The mainstay of diagnosis of typhoid in endemic areas therefore remains clinical. **Thus any high-grade fever of >72 hours' duration associated with any of the aforementioned features, especially with no localizing upper respiratory signs or meningitis or malaria, must be suspected as typhoid and managed accordingly**. While leucopenia (white cell count $<4\times10^9$/litre) with a left shift in neutrophils, may be seen in a third of children, young infants may also commonly present with a leucocytosis. ✔

Therapy of typhoid

Making an early diagnosis of typhoid fever and instituting appropriate supportive measures and specific antibiotic

therapy is the key to the appropriate management of typhoid fever. The following are the important principles of management:

- Adequate rest, hydration and attention to correction of fluid-electrolyte imbalance
- Antipyretic therapy (paracetamol) as required if fever >39°C
- Soft, easily digestible diet should be continued unless the child has abdominal distension or ileus
- Regular monitoring for clinical recovery and potential complications
- Antibiotic therapy: the right choice, dosage and duration are critical to curing typhoid with minimal complications. Traditional therapy with either chloramphenicol or amoxycillin is associated with relapse rates of 5–15% and 4–8% respectively.
- **If drug resistance is not locally a problem**, start with oral chloramphenicol and/or oral amoxicillin/ampicillin (initially intravenous if vomiting). **If drug resistance is prevalent**, use cefixime or ceftriaxone or ciprofloxacin (associated with higher cure rates).

While epidemics are usually associated with a single dominant clone of *S. typhi*, in endemic situations there may be several coexistent strains of *S. typhi* and a clinical judgment may need to be made when instituting antibiotic therapy before culture results become available. This is particularly important as delay in the institution of appropriate second-line antibiotic therapy in resistant cases of typhoid leads to a significant increase in the morbidity and mortality. Despite the availability of newer orally administrable drugs such as quinolones and third-generation cephalosporins, blanket administration of these agents to all cases of suspected typhoid is expensive and will only lead to the rapid development of further resistance.

Given the recent evidence that MDR typhoid is a more severe clinical illness from the outset, the algorithm in Figure 4.11.2 may be acceptable for selection of antibiotics and management of typhoid.

Table 4.11.1 shows the main antibiotics that can be employed for the treatment of both sensitive and MDR infections with *S. typhi.*

Corticosteroids

In severely ill and toxic children with typhoid requiring hospitalisation, **dexamethasone** IV (0.5–1 mg/kg/day 8 hourly for up to six doses) **may be life-saving**. However, **avoid using steroids in ambulatory settings** as they mask abdominal complications and peritonitis.

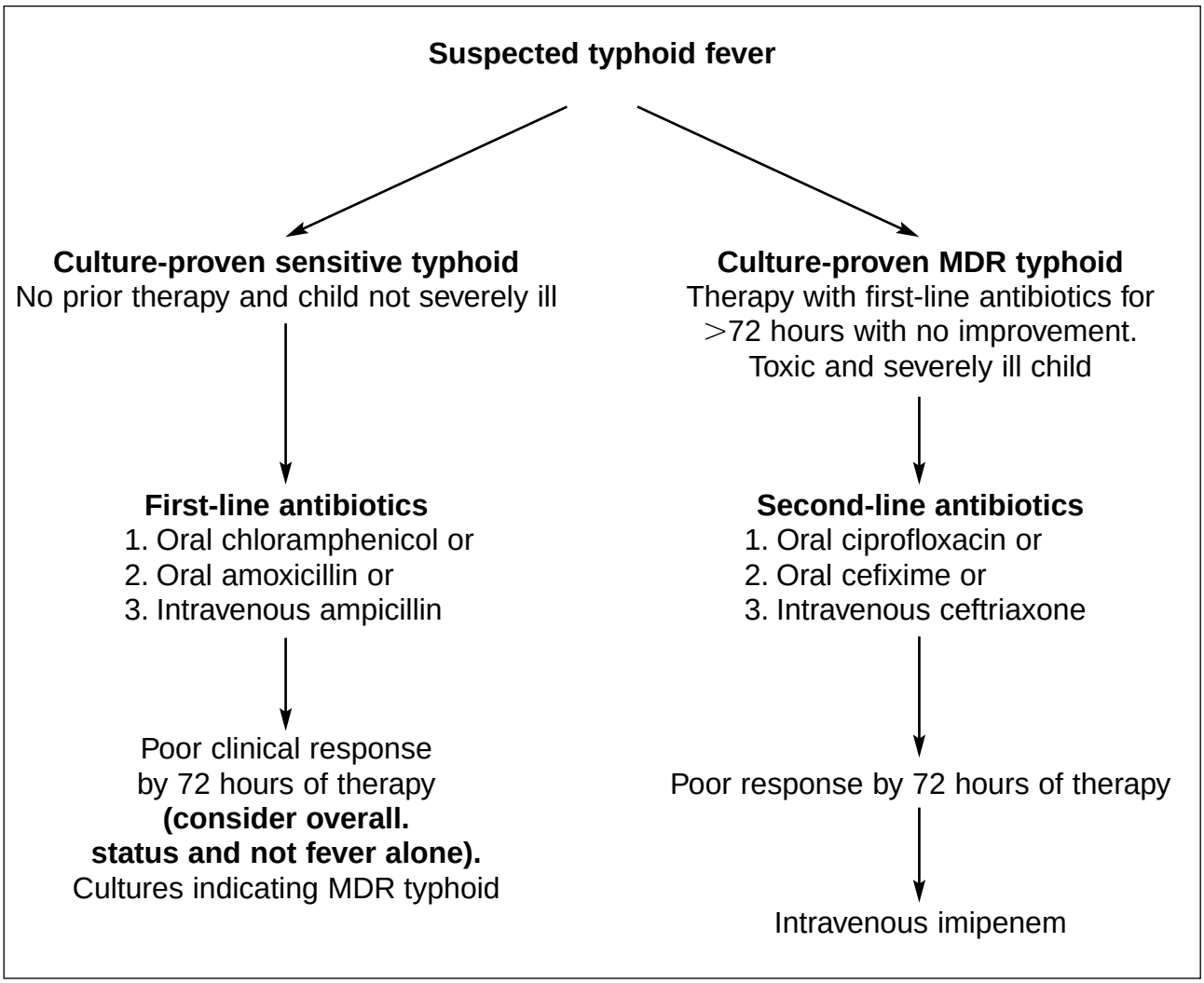

Figure 4.11.2 Management of typhoid.

Table 4.11.1 Antibiotics in *S. typhi* infections

Drug	Route	Dose (frequency)	Duration (days)
Chloramphenicol	oral	60–75 mg/kg/day (6 hourly)	14 days
Ampicillin/amoxicillin	IV/oral	100 mg/kg/day (6–8 hourly)	14 days
Ciprofloxacin	oral/IV	20–30 mg/kg/day (12 hourly)	7–10 days
Ceftriaxone	IV/IM	65–100 mg/kg/day (once daily)	7–14 days
Cefixime	oral	8 mg/kg/day (12 hourly)	14 days
Imipenem	IV	60 mg/kg/day (8 hourly)	10–14 days

Preventive measures for typhoid

The continued presence of typhoid in much of the disadvantaged world is an indication of the poor state of public health and sanitation. It is important therefore to be aware of the important risk factors for developing typhoid, in order to institute preventive measures during outbreaks.

There is recent epidemiological evidence that prior usage of antibiotics is associated with an increased risk of subsequent development of typhoid. The precise reasons for this are unclear but may be related to alterations in intestinal flora, increasing the predisposition to colonisation and infection with pathogenic strains of *S. typhi.* Thus controlling indiscriminate use of antibiotics may not only reduce the emergence of drug-resistant strains, but also reduce the risk of development of typhoid.

Of the major risk factors for outbreaks of typhoid, contamination of water supplies with sewage is the most important. During outbreaks therefore, a combination of central chlorination as well as domestic water purification are important. In endemic situations consumption of street foods, especially ice cream and cut-fruit has been recognised as an important risk factor. The human-to-human spread by chronic carriers is also important and attempts should therefore be made to target foodhandlers and high-risk groups for *S. typhi* carriage screening.

Of the available vaccines against typhoid, the classic heat-inactivated whole-cell vaccine is associated with an unacceptably high rate of side effects. There are two newer vaccines which offer protection in older school-age children, but are not recommended for use in children under 3 years of age:

- The Vi polysaccharide vaccine can be administered in two doses at any stage and has a 70–80% protective efficacy for at least five years. Recent data from the Tajikistan outbreak also suggests that large scale administration of the Vi vaccine also contributed to the arrest of the epidemic.
- The oral attenuated ty-21a vaccine has also been evaluated and found to be comparably effective. However, it is generally available in capsule form and therefore difficult to administer to young, especially pre-school children.

Given the high rates and morbidity of typhoid in young children, there is a clear need for the development of a Vi-conjugate vaccine, which could be potentially employed within the Extended Programme of Immunisation vaccination schedule.

Non-typhoidal salmonella infections

These infections usually give rise to a self-limiting gastroenteritis. This is manifest as diarrhoea with abdominal cramping pains, nausea and vomiting. There is usually a fever and there may be blood and mucus in the stools. See Chapter 3.25 for treatment of this level of infection. A reactive polyarticular arthritis may develop two weeks after the diarrhoea.

Occasionally, particularly in the neonate and in the immunosuppressed, the malnourished, or children with sickle cell disease, these infections can become very serious by spreading to the following sites: meninges (meningitis), bones (osteomyelitis) and joints (septic arthritis), lungs (pneumonia and empyema) and soft tissues (giving abscesses). This is a particular problem in children with HIV infection.

Treatment for metastatic infections should be urgently given by intravenous or intramuscular injection. Initial treatment should ideally be with the broad-spectrum antibiotics cefotaxime or ceftriaxone and if later sensitivity tests become available the organisms may be sensitive to amoxicillin (usually resistant now), co-trimoxazole and ciprofloxacin. Chloramphenicol may be effective in the absence of the above.

- Cefotaxime

Neonates <7 days	50 mg/kg every 12 hours
Neonates >7 days	50 mg/kg every 8 hours
Infants and children	50 mg/kg every 6 hours

- Ceftriaxone: All ages 50 mg/kg once daily. In very severe infections 80–100 mg/kg once daily may be given (maximum dose 4 g/day).
- Co-trimoxazole: 18 mg/kg by IV infusion 12 hourly. In very severe infections, 27 mg co-trimoxazole IV 12 hourly.
- Ciprofloxacin 10–15 mg/kg twice daily by IV infusion.

412 Other bacterial infections and diseases

Brian Coulter, David Southall and Charles Newton

Anthrax

- An infection from animals caused by *Bacillus anthracis*.
- Produces a rapidly swelling boil which ruptures and discharges pus and develops a black centre.
- Local lymphadenopathy develops.
- Systemic illness with fever develops and may include pneumonia which is very serious.
- Treatment is with benzylpenicillin IV (or IM if not possible) 25 mg/kg 6 hourly up to 50 mg/kg 6 hourly if severely ill for at least 5 days then oral penicillin V for a further 10 days.
- It is important to notify authorities if such an infection is identified.

Botulism

Botulism is a toxin produced by *Clostridium botulinum*.

- Toxin blocks release of acetylcholine, producing a myasthenic syndrome.
- Botulism most commonly follows ingestion of foods (especially canned foods) that have not been sterilised properly.
- Occurs in infants after weaning (infantile botulism) and rarely from contaminated wounds.

Foodborne type

- Within hours of ingestion of contaminated foods, vision becomes blurred (pupils dilated, unreactive to light), bulbar signs develop (respiratory difficulties), followed by diffuse paralysis (with areflexia). Dryness of mouth and bladder distension are characteristic.

Infantile botulism

- Occurs in infants usually between 3 and 6 months old, particularly after introduction of weaning food (honey in particular has been implicated). Breast fed babies may be at higher risk.
- Constipation is an early feature, followed by poor sucking and ptosis (with other cranial nerve involvement), hypotonia (with areflexia) and paralysis.
- Respiratory difficulties and aspiration are common, often requiring respiratory support. Aminoglycosides may exacerbate symptoms.

Diagnosis

- *C. botulinum* may be isolated or the toxin detected in stool, or rarely in blood for some months after infection.
- ***Electromyogram is characteristic, with an incremental response to 25–50 Hz repetitive nerve stimulation***.

Management

This is supportive. ***Human botulism immunoglobulin may be effective but is rarely available in poor countries***. Respiratory and nutritional support are major aspects of management which determine outcome.

Brucellosis

- An infection from animals by Brucella species usually through infected milk.
- Causes a chronic illness with fever, pain and swelling of joints and anaemia.
- Treatment is with co-trimoxazole for four weeks: 18–24 mg co-trimoxazole/kg twice daily.

Or paediatric liquid 240 mg/5 ml (200 mg sulphamethoxazole plus 40 mg trimethoprim):

6 weeks to 6 months:	2.5 ml twice daily
6 months to 6 years:	5 ml twice daily
6–12 years:	10 ml twice daily

Campylobacter infection

Acute gastroenteritis with **considerable abdominal pain**, fever and bloody diarrhoea (see Chapter 3.25). Most children recover without treatment with antibiotics although erythromycin or ciprofloxacin are both effective.

Chlamydia infections

Chlamydia trachomatis causes trachoma (see Chapter 3.35), infections of the genital tract (see Chapter 4.2), conjunctivitis in the newborn which is less severe than that due to the *Gonoccocus* (see Chapter 3.48).

C. pneumonia produces a chronic pneumonitis in the infant. It is important not to forget this cause of acute respiratory infection which responds well to erythromycin (see Chapter 3.1).

Haemophilus influenzae infections

- Haemophilus influenzae causes serious infections in infants and young children including:
  - pneumonia (see Chapter 3.1).
  - middle ear infections (see Chapter 3.18).
  - acute epiglottitis (see Chapter 3.17).
  - meningitis (see Chapter 4.1).
- ✔ Infections can be prevented by an extremely effective conjugate vaccine. **Every country should attempt to immunize their infants against this cause of many serious illnesses, deaths and handicap**.

Plague

- *Yersinia pestis* is carried to children by the fleas of infected rats.
- It occurs in epidemics.
- It presents with an acute fever and painful, tender large swollen lymph nodes (buboes).
- It can produce pneumonia and septicaemia.
- Prompt treatment on suspicion is essential.
- Streptomycin is the treatment of choice for severe cases (15–20 mg/kg 1 M daily, maximum dose 1 g) for 7 days. Tetracyline (children >8 years 250–500 mg 6 hourly) and chloramphenicol 15–25 mg/kg 6 hourly are alternative drugs for 7 days.

Shigellosis

- This causes an acute gastroenteritis particularly affecting the large bowel.
- There is blood and mucus in the diarrhoea.
- There is often a high fever.
- There may be tenesmus (a continuous feeling of wanting to defaecate).
- Septicaemia may occur.
- See Chapter 3.25 for advice on treatment.

Staphylococcal infections

- Most common presentation is with a pus forming skin infection (impetigo): see Chapter 3.47.
- However, this bacterium can be bloodborne to other parts of the body where it produces serious infections:
  - Pneumonia is particularly dangerous (see Chapter 3.1).
  - Osteomyelitis is also dangerous and difficult to diagnose (see Chapter 3.45).
  - Pyomyositis can occur.
  - Occasionally they cause mastoiditis (see Chapter 3.18) and laryngotracheitis (see Chapter 3.17).

The two groups of antibiotics most effective against this organism are: flucloxacillin or cloxacillin and sodium fusidate (fucidin)

Treatment with sodium fucidate

Use in combination with an other antistaphylococcal agent if possible, to avoid resistance.

Oral

Absorption is not as good as IV but the oral route should be used when possible: 250 mg fusidic acid in 5 ml. (175 mg sodium fusidate in 5 ml)

Under 1 year	0.3 ml/kg three times daily
1–5 years	5 ml three times daily
5–12 years	10 ml three times daily
>12 years	15 ml or 500 mg as tablets (250 mg sodium fusidate per tablet) three times daily

Intravenous infusion

6–7 mg/kg of sodium fusidate 8 hourly (children >50 kg in weight give 500 mg IV 8 hourly.

Dose may be doubled in severe infections.

Dilute in 0.9% saline to concentration of 1 mg/1 ml and give slowly over at least 6 hours (over 2 hours by central line).

413 Acute encephalitis

Bridget Wills

Minimum standards requirements

- ABCD and intensive care (Chapters 1.19 and 1.25)
- Lumbar puncture, clinical chemistry
- Temperature control
- Anticonvulsants (Chapter 3.38)
- Antibiotics

Encephalitis is an inflammatory process involving primarily the brain, but sometimes also the meninges (meningoencephalitis) or spinal cord (encephalomyelitis). Primary encephalitis refers to cases in which the causative agent invades and replicates within the nervous system, whilst in postinfectious encephalitis the clinical manifestations appear to be caused by an immunological response to the agent. In practice it can be difficult to differentiate between the two entities.

Aetiology

- In many instances no specific aetiological agent can be identified.
- Geographic location and seasonal variation, amongst other epidemiological factors, influence the frequency of infection with specific agents.
- Viruses are the responsible pathogen in the vast majority of cases.
- Arboviruses are the most important worldwide cause of encephalitis but the major contributor within the arbovirus group, the Japanese encephalitis virus (JEV), is limited to Asia and the Pacific Rim. Enteroviruses are a common, seasonal cause of encephalitis in Europe and the United States, whilst herpes simplex type 1 (HSV-1) causes sporadic encephalitis worldwide.
- The common childhood viral infections such as measles, mumps, rubella, and chickenpox (varicella zoster virus (VZV)) may all involve the nervous system.
- Spirochaetal infections including syphilis, leptospirosis and Lyme disease are well recognised to cause meningoencephalitis; other organisms such as *Brucella* are occasionally implicated. *Mycoplasma pneumoniae* is an important and treatable cause. Neurological involvement may occur in chlamydial and rickettsial infections, and both fungi (*Cryptoccoccus*) and parasites (*Naegleria*, *Acanthamoeba*) may cause meningoencephalitis.
- Encephalitis has been noted to occur following a wide variety of immunisations. Fortunately, improvements in vaccine technology in recent years have meant that such complications are now rare.

Clinical features

The following manifestations commonly occur, whatever the aetiological agent.

- An acute systemic illness with fever, headache, nausea and vomiting.
- Generalised seizures; less commonly focal.
- Behavioural or personality changes.
- Deteriorating conscious level; confusion and drowsiness, lapsing into coma.
- Neck stiffness; common but not invariable.
- Manifestations of involvement of any part of the nervous system, for example hemiparesis, ataxia, myelitis, movement disorder, brainstem abnormalities.
- A rash may point to a specific diagnosis, for example measles, VZV, enteroviruses.
- Signs of raised intracranial pressure (ICP) may be present. The possible contribution of raised ICP to the clinical picture should always be considered, since this may be amenable to treatment.

The severity varies from a mild illness with fever, a single brief seizure and confusion lasting for two or three days to a more prolonged illness with fluctuating level of consciousness and evolution of neurological signs over several weeks. Occasionally the course may be fulminating with death within a few days.

Diagnosis

The following investigations (Table 4.13.1) should be considered in all cases but may be constrained by lack of resources. Efforts should be directed towards identifying those diseases that are treatable, common locally, or indicated by specific details in the history.

CSF examination and culture provide valuable diagnostic information, but if the child shows evidence of raised ICP or has signs suggestive of a space-occupying lesion, lumbar puncture may precipitate herniation. Lumbar puncture should be deferred until considered safe, and antimicrobial therapy should be prescribed empirically, directed towards the common pathogens and antibiotic sensitivity patterns in the region.

Typical findings in the CSF in viral encephalitis are documented in Table 4.13.2, together with typical features on ***EEG and neuroimaging***. In general it is possible to differentiate viral from bacterial CNS infections from the CSF picture. If there is doubt, empirical antibiotic therapy may be given pending CSF culture results, or alternatively if the child is stable, the lumbar puncture should be repeated after 24–48 hours whilst observing the clinical condition closely.

Table 4.13.1 Suggested investigations in children with acute encephalitis, with reference to differential diagnosis

Investigation	Relevance
Blood glucose	Hypoglycaemia (common in infants and children with severe infections and poor oral intake/vomiting) Hyperglycaemia (diabetes) Metabolic encephalopathies, inborn errors of metabolism
Full blood count, blood film	Cerebral malaria (in endemic regions, returning travellers, etc.)
Urea and electrolytes	Hyponatraemia, syndrome of inappropriate secretion of antidiuretic hormone
Liver function tests	Reye's syndrome, metabolic encephalopathies Liver failure
Arterial blood gas	Metabolic encephalopathies To assess severity, particularly in those with brainstem compromise
Ammonia	Reye's syndrome, metabolic encephalopathies
Blood culture Widal test	Typhoid/other septicaemias may have encephalopathic features
Acute/convalescent serology	To include locally relevant pathogens (for example Japanese encephalitis serology in Asia) and those suggested by history and examination (for example measles, mumps, varicella, HSV, *Mycoplasma*, *Legionella*)
Toxicology	Heavy metals, pesticides
Erythrocyte-sedimentation rate ***and autoantibodies***	Collagen vascular disorders
Cerebrospinal fluid (CSF) examination and culture	Bacterial meningitis Tuberculous meningitis Intracranial haemorrhage
Electroencephalography (EEG)	Status epilepticus
Neuroimaging ***(with contrast enhancement)***	Space-occupying lesion (malignancy, brain abscess) Tuberculous meningitis Intracranial haemorrhage

Investigations in bold are mandatory, but CSF examination may be deferred if there are features suggestive of raised ICP or space-occupying lesion. Empirical antibiotics should be given until CNS bacterial infection can be definitively excluded. The remaining investigations will depend on the clinical presentation and local availability/resources.

Table 4.13.2. Typical findings in viral encephalitis

Investigation	Findings
Cerebrospinal fluid: microscopy and biochemistry	• Rarely may be normal • Usually lymphocyte predominant pleocytosis (a few to several thousand white blood cells/mm^3). • In early disease polymorphonuclear cells may predominate. • Mildly elevated or normal protein • Normal CSF/plasma glucose ratio • Absence of microorganisms on Gram stain
Electroencephalography (EEG)	• Virtually always abnormal • Diffuse slow waves, occasionally unilateral • Patterns may suggest particular causative agents, for example HSV (see below) subacute sclerosing panencephalitis
Neuroimaging	• May be normal • Cerebral oedema is common • Features may suggest particular causative agents, for example HSV, Japanese encephalitis virus

Management

In the majority of cases no specific treatment is available and management is primarily supportive.

- Bed rest, analgesia for headaches. Care with sedation, since a deterioration in conscious level may be obscured and/or respiratory depression may occur.
- Temperature control (avoid fever >38°C).
- Adequate oxygenation (SaO_2 >94%).
- Regular monitoring of electrolytes and review of fluid balance. **Fluid restriction may not be appropriate if the cardiac output is low**. Aim to keep the serum sodium (and other electrolytes) within the normal range. Consider the possible causes of hyponatraemia (for example vomiting/gastrointestinal losses, excessive hypotonic intravenous fluids, overhydration, syndrome of inappropriate secretion of antidiuretic hormone) and act accordingly.
- ✔ **Critically ill children, particularly those with evidence of brainstem involvement or raised ICP, should be managed in an intensive care unit if possible**. ***Assisted ventilation and cardiovascular support should be instituted early if there is evidence of compromise***.
- Control of seizures. Diazepam 200 micrograms/kg rectally or IV, initially. For repeated seizures, in infants consider phenobarbitone (loading dose 20 mg/kg IM or IV over 30 minutes followed by maintenance of 5 mg/kg/day IV, IM or oral) and in older children phenytoin (loading dose 15–20 mg/kg IV over 1 hour followed by maintenance of 4 mg/kg twice daily IV or oral). Caution is required in the use of anticonvulsant drugs with the potential for respiratory depression. ***EEG monitoring*** may reveal subclinical seizure activity.
- Consider the use of mannitol (250–500 mg/kg/dose which may be repeated afer 1 hour) if there is evidence of raised ICP but remember that electrolyte and fluid balance monitoring are critical (hypernatraemia is as dangerous as hyponatraemia). ***ICP monitoring should only be undertaken in centres experienced in this technique***.
- If there are pointers in the history/examination/preliminary investigations to suggest a specific diagnosis such as HSV, *Mycoplasma*, Lyme disease, the relevant treatment should be given (see below).
- If bacterial or tuberculous meningitis cannot be excluded, and the child is severely ill, consider cover with appropriate drugs until these diagnoses can be definitively ruled out (see Chapters 4.1 and 4.10).
- ✔ A second lumbar puncture 24–48 hours after admission may help with this decision **(provided raised ICP is not present)**.

The illness may be prolonged and survivors may be left with significant neurological sequelae. Physiotherapy and rehabilitation should be commenced once the acute stage of the illness is over, and the child is stable. Some children remain in hospital for many months and relatives require considerable support to cope with the often devastating effects on the family. A number of children without overt neurological sequelae are left with subtle problems including visual and hearing impairments, learning difficulties, and behavioural problems. Long-term follow-up is needed to detect and manage these problems.

Specific features of encephalitic infections

Japanese encephalitis virus

- Most common cause of encephalitis worldwide. Estimated 50 000 cases and 15 000 deaths each year.
- Currently limited to Asia and the Pacific Rim.
- Transmitted by *Culex* mosquitoes, with enzootic cycle (pigs and birds).
- Most infections are asymptomatic: ratio 200–300 : 1 encephalitis.
- Extrapyramidal and brainstem involvement is common in cases with encephalitis. Patients may have Parkinsonian features acutely, some later developing choreoathetoid movement disorders. Gradual improvement over several months is usual in survivors.
- Myelitis may occur, usually accompanied by some encephalitic features. Prognosis for recovery from myelitis is poor.
- Diagnosis rests on ***IgM/IgG capture ELISA in serum and CSF***. Viral isolation is difficult, as the viraemia is short-lived.
- Thalamic, basal ganglia and brainstem lesions are often apparent on ***imaging (CT/MRI)***.
- There are no specific features on ***EEG***.
- Treatment is supportive only, but effective vaccines are available.
- Prognosis is poor. Up to 30% with encephalitis die in the acute stage. Neurological sequelae are common in survivors but do tend to improve with time.

Herpes simplex virus

- Causes sporadic encephalitis worldwide (usually HSV-1).
- Encephalitis is more frequently a manifestation of recurrent than primary infection. There is no correlation between the presence of herpetic skin lesions and the diagnosis of HSV encephalitis.
- Seizures, focal and generalised, are a prominent feature.
- Personality changes, temporal lobe phenomena, and dysphasia are also common.
- CSF findings:
  - Lymphocytic pleocytosis: <50 to 2000/mm^3
  - Red blood cells are present in CSF in >80% of cases, reflecting haemorrhagic necrosis
  - Protein levels are usually moderately elevated, but may reach very high levels as the disease progresses (3–5 g/litre)
  - Up to 25% may have a relatively low CSF glucose
  - Occasionally the CSF is entirely normal in early disease.
- Diagnosis is by ***polymearase chain reaction or serology on CSF***. Viral isolation is difficult.

- ***EEG*** may show a typical pattern of multifocal periodic lateralising episodic discharges (PLEDs) on a slow background, often with a temporal lobe focus.
- ***CT/MRI*** may show lesions (often haemorrhagic) in the temporal lobes. In early disease scans may be normal.
- Treatment is with high dose intravenous aciclovir for at least 14 days (20 mg/kg 8 hourly IV, given as an infusion over 1 hour).
- Early diagnosis and treatment improve outcome significantly. HSV is a possibility in all patients with encephalitis although in areas of the world where Japanese encephalitis is endemic, HSV is responsible for only a very small minority of the total number of cases. **If resources permit, start aciclovir (dosage as above) in all cases without a definitive diagnosis and continue until HSV is excluded, or an alternative diagnosis is reached**.
- Mortality can still be up to 20%, with around 15% left with severe sequelae. Relapses occur occasionally.

Varicella zoster virus

- Usually only results in mild encephalitis with acute cerebellar ataxia as the main feature. Seizures and coma are rare.
- Prognosis is good.

Measles

- Acute encephalitis may occur 6–8 days after onset of the rash and may be severe with up to 10% mortality, and frequent sequelae.
- Delayed chronic encephalitis may also occur in the form of subacute sclerosing pan encephalitis (SSPE), presenting with cognitive deterioration and myoclonic jerks. In such cases the EEG shows stereotypic polyphasic complexes on a background of excess slow activity.

Rabies (see Chapter 4.22)

- Saliva (plus virus) from an infected mammal enters via a bite, skin abrasion, or rarely through intact skin/mucous membranes.
- The incubation period varies from a few days to many months. A history of animal bite may not be elicited at the time of presentation.
- There is an initial prodrome of fever and malaise lasting a few days, followed by second phase of excitement, hyperacusis, hydrophobia and pharyngeal spasms.
- Lastly, a paralytic phase occurs (rarely this may be the only manifestation).
- Death is inevitable once neurological signs are apparent.
- Effective prevention is available with the human diploid cell vaccine and should be combined with passive immunisation if exposure has occurred.

Mycoplasma

- Neurological involvement occurs in up to 7% of infections.
- Both direct invasion of CNS and immune-mediated disease occur.
- Aseptic meningitis, transverse myelitis and Guillain–Barré syndrome are the common manifestations.
- Diagnosis is by ***complement fixation titres***.
- If the diagnosis is suspected, treat intravenously or enterally if tolerated: erythromycin (10 mg/kg/dose 6 hourly for 10 days), although this is not effective for the immune-mediated disease.

Lyme disease

- Caused by the spirochaete, *Borrelia burgdorferi*.
- Transmitted by *Ixodes* ticks from animal reservoirs. Seasonal incidence (spring to autumn) reflects exposure to ticks in rural areas of Europe and America.
- About 70% of patients have a characteristic rash spreading from the site of the tick bite – erythema chronicum migrans. (Tick bites may not be noticed however.)
- A second stage of systemic symptoms occurs over the next few weeks to months.
- Up to 15% have neurological involvement, commonly aseptic meningitis with a facial palsy (sometimes bilateral). Rarely transverse myelitis, Guillain–Barré syndrome or cerebellar ataxia may occur.
- Cardiac and joint manifestations may be evident late in the second stage.
- Finally, a third stage of progressive encephalomyelitis, sometimes with chronic arthritis may occur.
- Confirmation of the clinical diagnosis can be difficult. The ***polymerase chain reaction is useful if positive***. Serological diagnosis relies on ***IgM antibody detection,*** since IgG responses are often positive in endemic areas and may not be related to current neurological disease.
- Treatment of those with neurological involvement is benzyl penicillin 50mg/kg IV 4 hourly for 10 days) or a third-generation cephalosporin (for example ceftriaxone 75 mg/kg once daily IV or IM for 14 days).

414 Chickenpox

James Tumwine

Minimum standards requirements

- Antipyretics and antipruritus treatment, for example chlorpheniramine/promethazine
- Antibiotics for secondary infection
- Aciclovir in immunosuppressed and neonates

- Caused by varicella zoster virus (a herpesvirus). Spread by direct contact, droplet or airborne transmission and is very contagious.
- Commonest between 5 and 9 years of age. In immunocompetent children it is a mild disease and one attack virtually assures life-long immunity.
- Groups at increased risk: immunodeficiency, for example HIV infection, those on chemotherapy, long-term steroids (defined as those within the previous three months who have received prednisolone (or its equivalent) at a daily dose of 2 mg/kg/day or >40 mg/day for at least one week or 1 mg/kg/day for one month). Patients on lower doses of steroids plus another immunosuppressant drug or medical problem, for example nephrotic syndrome, should be included or those on salicylate therapy or with chronic lung or skin problems.
- Children with chickenpox are at increased risk of developing Reye's syndrome if given aspirin and other non-steroidal anti-inflammatory drugs.

Clinical presentation

- Incubation period is 14–21 days. There is low-grade fever and headache, followed by the rash mostly on the trunk and face. Rash develops into successive small single oval vesicles with an erythematous base which break within two days to develop into scabs and heal. It is very itchy, and scratching may result in secondary bacterial infection and scar formation.
- Course of disease is about one week. Children are infectious one or two days before rash appears, until one or two days after all lesions have formed scabs.
- Complications include: septicaemia, bronchopneumonia, hepatitis, thrombocytopenia, purpura, pericarditis, myocarditis, endocarditis, arthritis, myositis, glomerulonephritis, postinfectious encephalitis especially with cerebellar involvement.
- Guillain–Barré syndrome, facial nerve palsy, transverse myelitis, hypothalamic involvement, optic neuritis and transient loss of vision have been reported.
- Intrauterine infection especially in first two trimesters may result in a congenital varicella syndrome, that is intrauterine growth retardation, scarred skin, limb atrophy, mental retardation, CNS and eye complications. Only 1–2% of infants with intrauterine exposure develop complications.
- In mothers, chickenpox but not shingles, occurring five days before or two days after delivery may result in a severe infection in the neonate. This is probably the consequence of impaired immunity. The infant should be treated as soon as possible with ***varicella-zoster immunoglobulin*** if available (see below).

Management

- Keep child clean, cut and clean under nails to discourage scratching and secondary infection. Baking soda baths or calamine lotion may relieve itching. Antihistamines such as promethazine 125 micrograms/kg three times a day and 500 micrograms/kg at night or chlorpheniramine 1 mg twice daily (up to 1 year), 1–2 mg three times a day (1–5 years), 2–4 mg three to four times a day (6–12 years), 4 mg three to six times a day (>12 years) may reduce scratching. Give paracetamol for fever. Appropriate antibiotics are given for secondary bacterial infection with *Staphylococcus aureus* or *Streptococcus pyogenes*.
- Aciclovir 10 mg/kg/8 hourly or 250 mg/m^2 8 hourly (IV) for 7–10 days is recommended for immunocompromised children who develop chickenpox. Oral aciclovir (20 mg/kg four times a day) is given for HIV-infected patients whose ***CD4$^+$ counts*** are relatively normal.
- If available, IM varicella-zoster immunoglobulin (VZIG) 0–5 years 250 mg (1 vial); 6–10 years 500 mg (2 vials); >10 years 750 mg (3 vials) may modify the disease if given shortly (not more than four days) after exposure. Indications include: immunocompromised children including HIV-infected pregnant women and premature infants <28 weeks gestation, who have had an intimate contact (face-to-face) with chickenpox

or herpes zoster. Neonates whose mothers develop varicella from 7 days before to 28 days after delivery are offered VZIG 250 mg as a single IM injection.
- If VZIG is not available, oral aciclovir (as above doses) may be given.

Prevention

- Live attenuated varicella vaccine given as two subcutaneous or intramuscular injections one month apart, confers over 95% protection against severe disease.
- Contraindicated in immunocompromised children and those receiving aspirin.
- Patients who are under immunosuppressive treatment (including steroid therapy) are generally immunised when in complete remission. Total lymphocyte count should be $>1.2\times10^9$ litre and there should be no other evidence of a lack of cellular immune competence.
- Should not be given within three months of VZIG.

4.15 Dengue

Sirijitt Vasanawathana

Minimum standards requirements

- ABCD
- Recognition and treatment of shock
- Blood transfusion and replacement of clotting factors and platelets
- High-dependency/intensive care
- Vector control

- Dengue infection is caused by an RNA virus of Flaviviridae family.
- It first appeared in an epidemic in the Philippines in 1957, subsequently in Thailand, and then in other South-East Asian countries.
- Dengue virus is composed of four serotypes, type 1, 2, 3 and 4.
- *Aedes aegypti* and *Aedes albopictus* are the main vectors.
- Epidemics occur every year during rainy season.
- Dengue haemorrhagic fever is a serious clinical manifestation of dengue virus infection.

Differential diagnosis

Depending largely on age and immunological response, Dengue virus infection may be:

- Asymptomatic
- An undifferentiated febrile illness (viral syndrome)
- Dengue fever
- Dengue hemorrhagic fever

Undifferentiated fever

Infants and children who have been infected with Dengue virus for the first time (i.e. primary Dengue infection) develop fever indistinguishable from other viral infections. A maculopapular rash may accompany the fever or appear during convalescence.

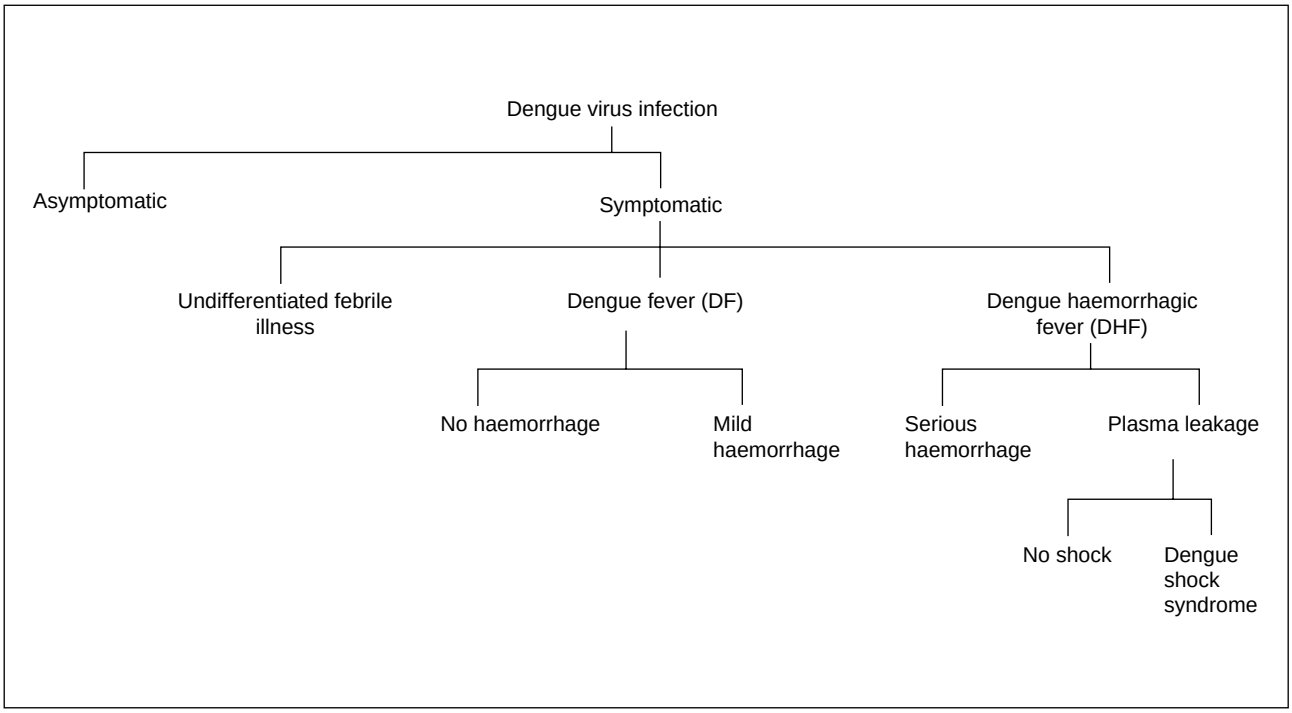

Figure 4.15.1 Manifestations of Dengue syndrome.

Dengue fever

Dengue fever is most common in young adults and older children. There is an acute biphasic fever with headache, myalgia, arthralgia, rashes and leucopenia. Although it is commonly benign, there may be an incapacitating disease with severe muscle and joint pain (break-bone fever), particularly in adults, and occasionally there is an unusual haemorrhage. Infection with one Dengue serotype gives solid immunity to that particular serotype but only partial protection against others.

Dengue hemorrhagic fever

- Peak incidence 5–9-year olds.
- Commonly in children who have experienced a previous primary Dengue infection due to a different serotype (except in infants 6–12 months old).
- Causes a significant number of deaths.
- An acute fever associated with haemorrhagic diathesis and a tendency to develop fatal shock (Dengue shock syndrome).
- Abnormal haemostasis and plasma leakage are the main pathophysiological effects, with thrombocytopenia and haemoconcentration.

Pathogenesis of Dengue haemorrhagic fever

Dengue hameorrhagic fever occurs predominantly in patients with secondary infections (previous Dengue fever).

Antibody-dependent enhancement and immune enhancement are important in development of disease. Hypotheses for Dengue haemorrhagic fever include:

- Non-neutralizing antibodies to Dengue virus enhance viral uptake and replication in target cells (monocytes).
- Enhanced viral replication in presence of monocyte dysfunction and activated T-lymphocytes.
- Secondary infection of an individual who has low or absent neutralising antibodies against Dengue virus leads to a booster antibody response and a steep rise in antibody levels.
- Antibodies against Dengue virus bind to complement system which directs a selective attack on cells expressing viral antigens on their surface.

Grading of severity of Dengue haemorrhagic fever

Severity is classified into four grades according to two pathophysiological hallmarks: **shock** and **bleeding**.

Grade 1: Fever accompanied by non-specific constitutional symptoms. Only haemorrhagic manifestation is a positive tourniquet test.

Grade 2: Spontaneous bleeding in skin and/or other haemorrhages.

Grade 3: Circulatory failure: rapid and weak pulse, narrowing of pulse pressure (20 mmHg or less) or hypotension, cold clammy skin and restlessness.

Grade 4: Profound shock with undetectable blood pressure and pulse.

Table 4.15.1 Clinical manifestations and diagnosis of Dengue fever and Dengue haemorrhagic fever

Clinical manifestations/diagnosis	Dengue fever	Dengue haemorrhagic fever
Clinical manifestations		
Fever	High continuous fever	High continuous fever
Headache*	Yes	Yes
Periorbital pain*	Yes	Yes
Myalgia*	Yes	Yes
Arthralgia/bone pain*	Yes	Only arthralgia
Rash*	Yes	Yes
Hepatomegaly with or without tenderness	Usually no	Yes
Bleeding*, for example tourniquet test, petechiae, epistaxis, melaena, etc.	Yes	Yes
Leucopenia*	Yes	Yes
Platelet count	Normal or mild decrease	$<100 \times 10^9$/litre
Rising haematocrit >20% of normal	No	Yes
Plasma leakage (ascites, pleural effusion)	No	Yes
Diagnosis		
Clinical manifestation	Fever with two WHO criteria and positive tourniquet test (petechiae >10/square inch)	Fever Hepatomegaly with or without tenderness in right upper quadrant Pleural effusion and ascites
Laboratory test	Platelets normal or mildly decreased	Platelet count $<100 \times 10^9$/litre Haematocrit >20% of normal
Confirmation	***Positive serology (ELISA, hemagglutination test)*** ***Positive viral culture***	As per Dengue fever

* WHO criteria.

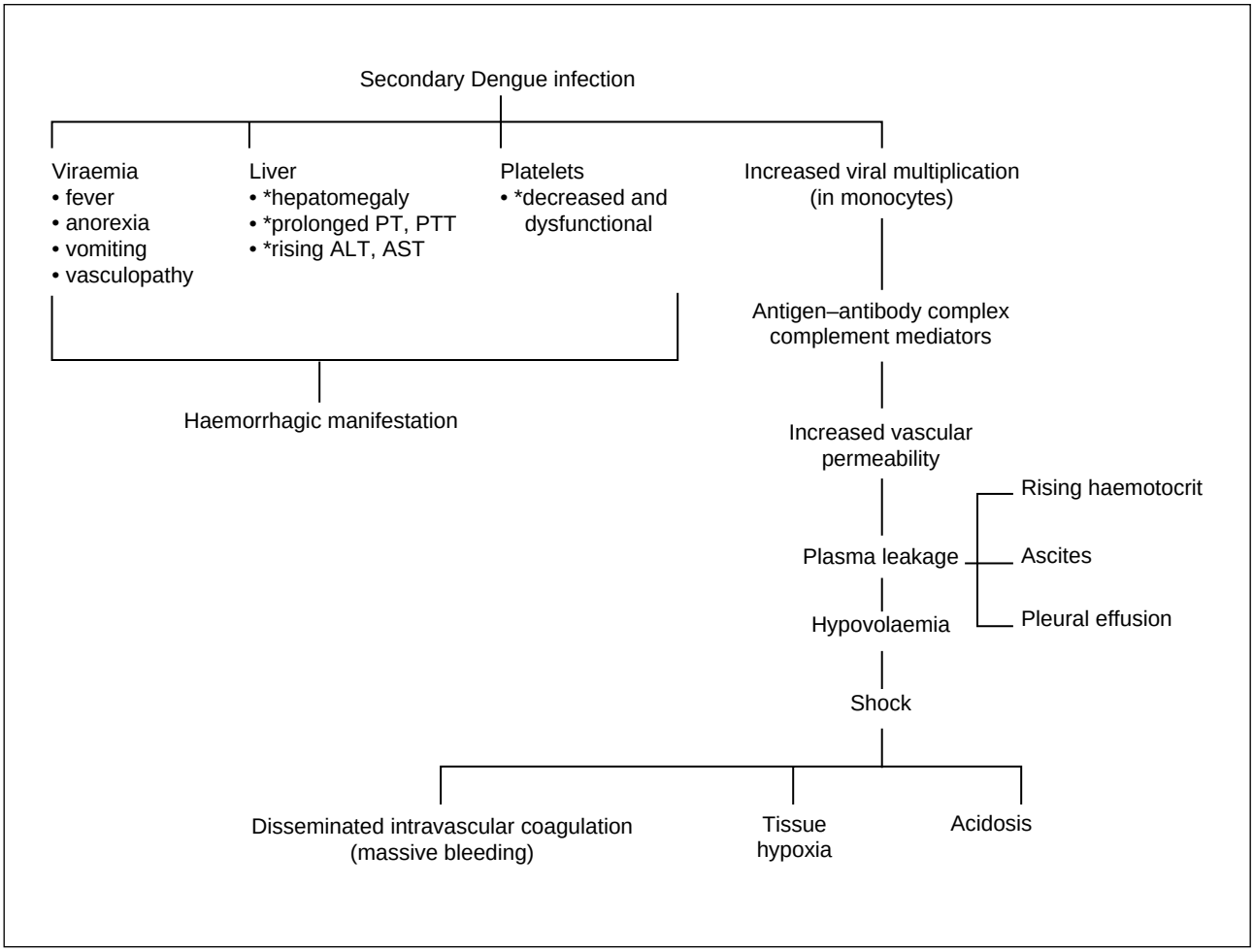

Figure 4.15.2 Pathophysiology of dengue haemorrhagic fever.
*** Important features.**

Table 4.15.2 Clinical course of Dengue haemorrhagic fever

Phase	Symptoms	Signs	Lab test
1. Febrile phase Duration 3–5 days (Days 1–5)	High continuous fever (39–40°C), headache, anorexia, nausea, vomiting, myalgia, epigastric discomfort, right upper quadrant pain, fine rubelliform maculopapular rash	Facial flushing, injected conjunctivae Tourniquet test positive Tenderness at right upper quadrant Hepatomegaly Lymphadenopathy	Not significant
2. Haemorrhagic shock or toxic phase Duration 1–2 days (Days 4–6)	Fever declines Abdominal pain especially right upper quadrant, bleeding in skin and mucosa (nose, gum, gastrointestinal tract haematemesis and melaena).	Right upper quadrant tenderness Hepatomegaly Tourniquet test positive (see figure)	Full blood count: rising, haematocrit, decreased platelet count, ($<100\times10^9$/litre) Leucopenia
	About 30% of cases will develop **shock** (sweating, irritable, restless, severe abdominal pain) Shock may progress to profound shock and child may die within 24 hours	Cold clammy skin (prolonged capillary refill time >2 seconds) Thready pulse, Tachycardia Narrow pulse pressure ($\leqslant 20$ mmHg), for example 90/70, 100/80	
3. Convalescence phase Duration 2–3 days	No fever or low-grade biphasic fever Increased appetite Diuresis	Convalescence rash on extremities with itching Confluent petechial rash with scattered round pale areas Sinus bradycardia	Stable haematocrit Rising platelet count

Management of Dengue fever and Dengue haemorrhagic fever

Dengue fever

- Management is symptomatic and supportive.
- Give antipyretics, analgesics (paracetamol) or mild sedatives for pain along with oral or intravenous fluids at maintenance levels. Control fever.
- Avoid salicylates and non-steroidal anti-inflammatory drugs, they may cause bleeding, acidosis, hepatotoxicity and Reye's syndrome.

Dengue haemorrhagic fever

- Management during the febrile phase is similar to that of Dengue fever; use antipyretics with caution.
- Rise in hematocrit value indicates loss of intravascular fluid and a need for parenteral fluid therapy.
- Volume replacement during period of leakage (24–48 hours) should be minimum, sufficient to maintain effective circulation.
  Normal maintenance fluids are: 5% glucose in 0.9% saline

Age	0–2 years	100 ml/kg/24 hours
	3–5 years	90 ml/kg/24 hours
	6–9 years	75 ml/kg/24 hours
	10–14 years	50 ml/kg/24 hours
	15–18 years	30 ml/kg/24 hours

  Aim to give full maintenance as above or even restrict figure above to 75% but treat shock episodes with additional boluses of 10 ml/kg over 10–15 minutes of plasma expanders (such as Dextran 40, hydroxyethyl starch, 4.5% albumin or plasma) or blood as appropriate. Colloid is important in patients with massive capillary leakage.
- Discontinue intravenous fluid when the haematocrit is less than 40 vol.%, vital signs are stable or the patient has regained appetite and diuresis has commenced.
- In an obese child use ideal body weight to calculate intravenous fluids. Intravenous fluids for example for a 7-year-old girl with body weight 40 kg, should be calculated by age multiplied by 2 and plus 8 ($7\times2+8=22$ kg).

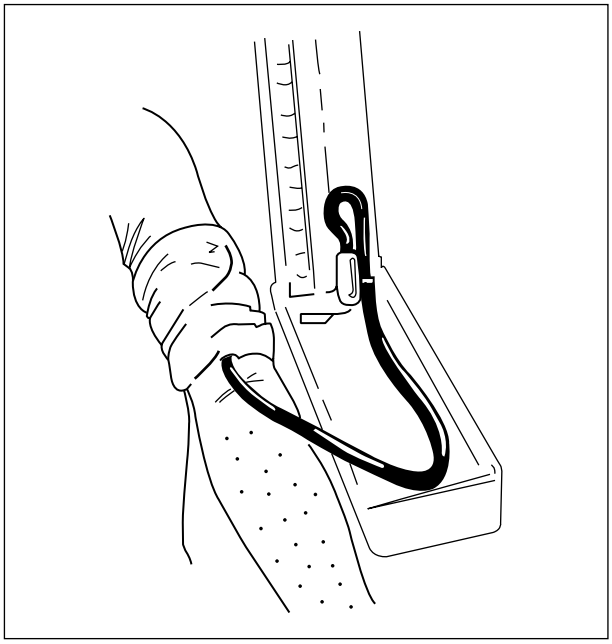

Figure 4.15.3 Tourniquet test. Apply blood pressure cuff inflated to level of mean arterial pressure (systolic + diastolic blood pressure, divided by 2) for 5 minutes. Positive test: petechiae ⩾10/square inch after removing cuff.

Table 4.15.3 Management of Dengue haemorrhagic fever (without shock)

Day of illness	Assessment	Management
Days 1–3 (febrile phase)	• Temperature, blood pressure, pulse rate, tourniquet test • Signs of dehydration • Diagnosis following WHO criteria	• Tepid sponge, paracetamol, avoid non-steroidal anti-inflammatory drugs • ORS/fruit juice/water frequently and in small amounts • Vomiting: try domperidone 200 micrograms/kg 6–8 hourly • Admit if the child has signs of dehydration, gastronitestinal bleeding or cannot drink • IV fluid: 10% glucose/0.9% saline 2–3 ml/kg/hour
Days 4–6 (toxic phase)	• Vital signs, tourniquet test • Full blood count • Signs of dehydration • Palpate liver/right upper quadrant pain • Serial haematocrit every 4 hours and platelet count	• Supportive treatment (as above) • Admit if child has signs of dehydration, gastrointestinal bleeding, rising haematocrit, signs of circulatory impairment (narrowed pulse pressure, prolonged capillary refill >2 seconds, tachycardia), unable to drink • Intravenous fluid: 10% glucose/0.9% saline 3–5 ml/kg/hour if haematocrit <45%; 5–7 ml/kg/hour if haematocrit >45%
Days 7–9 (convalescence phase)	• Vital signs • Haematocrit • Convalescence rash • Urine output	• Reduce intravenous fluids 24 hours after highest haematocrit • Discontinue intravenous fluid or reduce if the child can drink normal requirement or if there is a diuresis

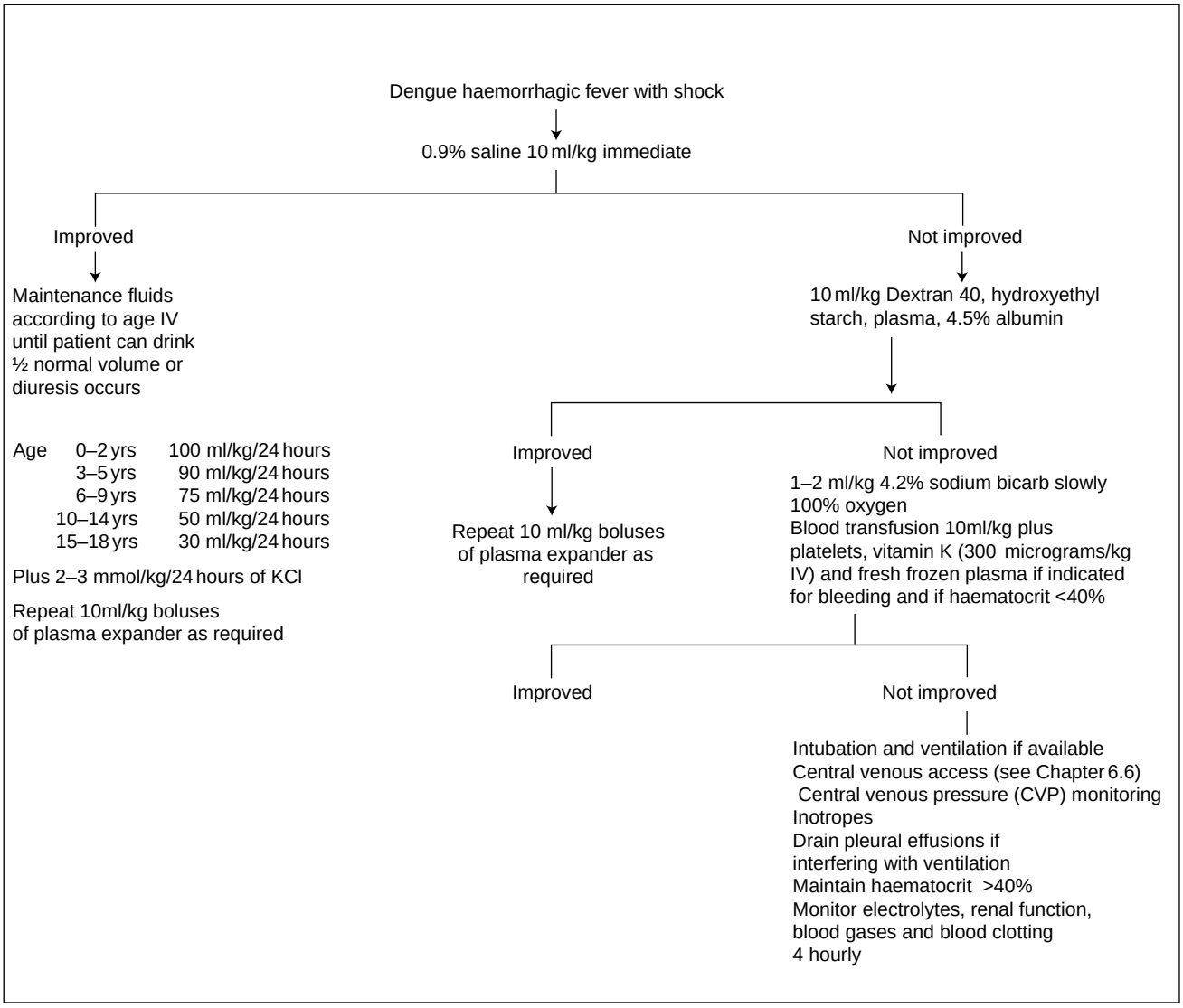

Figure 4.15.4 Management of Dengue haemorrhagic fever with shock (see Chapter 3.6).

If circulatory overload develops when large amounts of intravenous fluid are given in intractable shock, give furosemide 1 mg/kg IV, then observe vital signs. Repeat furosemide until improvement occurs. If the patient redevelops shock after furosemide, infuse plasma expander to maintain circulation and treat as intractable shock as shown in Figure 4.15.4.

Dengue infection with unusual manifestations

Vital organs are not primarily involved in Dengue haemorrhagic fever. Signs and symptoms of organ involvement are usually secondary to plasma leakage, shock and haemorrhage. Central nervous system disorder may be manifested by convulsions, spasticity and/or change in consciousness level. Hepatic dysfunction and renal failure may occur especially in cases with prolonged shock.

4.16 Acute hepatitis (AH)

Alastair Baker and Anil Dhawan

Minimum standards requirements

- Liver function tests, International Normalised Ratio, PTT, blood glucose
- Vitamin K
- Immunisation against hepatitis B

Introduction

- Acute hepatitis causes a liver dysfunction of duration less than six months.
- Transaminases are abnormal but patients are not necessarily jaundiced.
- Acute hepatitis may be cholestatic and may be complicated by acute liver failure as described in Chapter 3.9.
- Of particular importance are infectious viral hepatitides, especially hepatitis A, since it is common and other concurrent diseases may be overlooked.

Management of acute hepatitis

- ***If available, exclude hepatitis A (with HAV IgM)*** and attempt diagnosis with tests available.
- Monitor synthetic function for liver failure (International Normalised Ratio).
- Monitor for complications, including hypoglycaemia, encephalopathy, bone-marrow aplasia, secondary sepsis (see Chapter 3.9).
- Treat complications when possible.
- Give vitamin K 300 micrograms/kg.
- Give antiemetics if severe nausea and vomiting (see Chapters 1.27 and 1.28).
- Give intravenous fluids ONLY if oral or nasogastric rehydration is not possible.
- See Chapter 3.9 for the management of acute liver failure if this develops.
- If available, immunise family contacts for HAV A and B (HAV A two doses two weeks apart; HAV B three doses first immediately, second one month later and the third three to six months later).

Hepatitis A (HAV)

This is a picornavirus spread by the faecal–oral route.

- The incubation period from infection to raised transaminases is 10–20 days.
- The pre-ictal period may show anorexia, nausea, vomiting, fever and liver tenderness.
- Jaundice is related to age with >90% of children under 2 years asymptomatic but only 76% of teenagers.
- A minority of cholestatic cases have a relapsing course and 0.1–0.2% develop acute liver failure. The prognosis

Table 4.16.1. Causes of acute hepatitis

Aetiological group	Examples	Possible cholestasis
Viral	HAV, HBV, HCV, HEV, delta superinfection, cytomegalovirus, Epstein–Barr virus, herpes simplex, parvovirus, measles, mumps, varicella, rubella, adenovirus, ECHO, coxsackie. Flaviviruses (for example yellow fever, Dengue, Lassa, Ebola, Rift Valley fever and others).	HAV, HBV, HEV,
Bacterial/fungal	*Salmonella*, *Leptospira*, any septicaemia	Not usually
Protozoal + parasitic	See below	Not usually
Drugs and toxins	See Chapter 5.12	Drug cholestasis
Shock/haemodynamic	Cardiac arrest, post-cardiac surgery, heat stroke, radiation	Occurs 7–10 days after injury
Immune	Autoimmune, lupus, Kawasaki disease	Autoimmune, lupus, Kawasaki disease
Infiltrative	Leukaemias, haemophagocytic syndromes, Hodgkin's disease	Leukaemias, Hodgkin's disease
Metabolic	Urea cycle disorders, Wilson's disease	Usually not except Wilson's disease

for almost all is excellent. Chronic liver disease does not develop but occasional patients have a transient nodular regenerative phase with evidence of portal hypertension lasting up to one year. Aplastic anaemia is a rare complication.
- HAV vaccine is highly efficacious and without significant side effects.

Acute hepatitis B (see chronic liver disease, Chapter 3.10)

- This is spread by blood/body fluid products, vertical transmission from mother to baby, and by sexual contact. The risk of all such spread is much greater than for the HIV infection.
- The incubation period is 60–90 days but rarely seven months.
- The risk of acute liver failure is less than 1% and that of chronic liver disease depends on the patient's age.

Hepatitis C

- The spread of hepatitis C is the same as for Hepatitis B but vertical spread is rare (6%).
- Incubation period of 2–26 weeks followed by acute hepatitis almost always asymptomatic.
- Chronic hepatitis ensues in 30–90%.

Hepatitis E

- Spread by the faecal–oral route and is endemic in southern Europe, the Middle East and Asia.
- Rare in children, highest rate in adolescents of 3%. A relapsing course is seen.
- The prognosis is usually good but mortality is recognised in pregnant women.

Epstein–Barr virus (EBV) infection

- Accompanied by hepatosplenomegaly and hepatitis.
- The prognosis is usually good but rare cases are complicated by lymphoproliferative disease or haemophagocytic syndrome.

Cytomegalovirus infection

Spread is the same as hepatitis but symptoms are usually only seen in the newborn and immunocompromised.

Parvovirus B19 infection

Can be accompanied by acute liver failure and aplastic anaemia.

4.17 Herpes virus infections

Brian Coulter and David Southall

- These are covered in various parts of the book.
- For the management of herpes simplex infections of the mouth and oesphagus see Chapter 4.18 on HIV infection.
- For herpes encephalitis, see Chapter 4.13.
- For chickenpox, see Chapter 4.14.

418 Human immunodeficiency virus infection in children

Gareth Tudor-Williams and David Southall

Minimum standards requirements

- Preventive campaigns
- Antiretroviral drugs to prevent vertical transmission
- Correction of nutritional deficiencies
- Antibacterial drugs
- Antituberculous treatment
- Dexamethasone
- Antifungal treatment
- Antiviral treatment
- Analgesia and palliative care (see Chapters 1.27 and 1.28)
- Counselling and family support

Epidemiology

- Broadly, there are two major strains of the human immunodeficiency virus: HIV1 and HIV2. HIV1 is the more pathogenic and is responsible for the global epidemic. HIV2 is largely confined to West Africa. This chapter reflects current management of HIV1 but the principles apply to both strains.
- Infection with HIV leads to progressive destruction of the cellular immune system, ultimately resulting in an acquired immune deficiency syndrome (AIDS) in the vast majority of infected individuals.
- HIV/AIDS is now a leading cause of death in children.
- As of December 1999, an estimated 1800 children become infected with HIV each day worldwide. A cummulative total of over 5 million children have been infected; of these 3.8 million have already died.
- 95% of the world's HIV-infected children have been from disadvantaged countries. 90% have been from sub-Saharan Africa, but the prevalence is rising particularly in India, South-East Asia, and countries of the former Soviet Union.
- More than 90% of children acquire HIV perinatally (vertically) from their mothers. The rest are infected through transfusion of infected blood products or via unsterilised needles (extent unknown but probably small), or sexually amongst adolescents, or in younger children through child sexual abuse.
- In non-breastfed infants, vertical transmission occurs mainly around the time of delivery, with transmission rates in disadvantaged countries ranging from 17 to 24%. Breastfeeding roughly doubles the risk of transmission, thus in breastfed cohorts from disadvantaged countries the rates of transmission are 25–45%.
- Management ideally begins before birth, with counselling and voluntary testing of HIV-infected women during pregnancy, and instigation of measures to reduce transmission. In rich countries, antiretroviral therapy for mothers and infants, elective (prelabour) caesarean section and avoidance of breastfeeding has reduced transmission rates to less than 2%.
- Without prenatal counselling and screening, as is frequently the case in disadvantaged countries, management begins only when a child becomes symptomatic, with subsequent identification of the HIV infection in the mother.
- In all societies, even those with high prevalence, HIV is an extremely stigmatising condition and the mother or both parents may be reluctant to undergo testing. Confidentiality is essential.
- Counselling must be confidential, requires time and should be undertaken by trained staff.
- Even if a child born to an infected mother is uninfected, he or she is inevitably affected. More than 13 million children are estimated to have lost one or both parents to AIDS. These children may be abandoned by relatives, ostracised by the community, poorly educated and highly vulnerable. Many support themselves and surviving siblings by commercial sex work, and may acquire HIV infection as a result.

Natural history data

- The probability of dying by the age of 12 months or by 5 years was found to be 0.1 and 0.2 respectively in one large European study representative of advantaged countries in the era before "highly active antiretroviral therapy" (HAART).
- Data from large, long-term, prospective, perinatally-recruited cohort studies are limited in disadvantaged countries.
- Small studies from sub-Saharan Africa all suggest disease progression is more rapid (probability of death by 12 months = 0.23–0.35, and by 5 years = 0.57–0.89).

However even in disadvantaged countries, some children may be relatively symptom-free during the first decade of life.

- Such is the scale of the epidemic that all of the gains in reducing infant mortality rates over the last 20 years have been lost in urban populations in disadvantaged countries with high HIV seroprevalence.
- Growth failure, generalised lymphadenopathy, hepatosplenomegaly, persistent diarrhoea, pulmonary infections, chronic cough and recurrent fevers are the most frequent clinical manifestations.
- The most common causes of death are pneumonia, diarrhoea and malnutrition. Post-mortem studies from the Côte d'Ivoire and clinical studies in Malawi and South Africa, showed that *Pneumocystis carinii* pneumonia (PCP) is a frequent cause of death in children under 15 months.
- Malignancy is a relatively rare AIDS-defining illness in children, compared with HIV-infected adults. However substantial increases in Kaposi sarcoma in children have been reported from East and Central Africa. Coinfection with the human herpes virus (HHV8) is a crucial aetiological factor. Kaposi sarcoma typically presents with large, non-tender, firm mobile nodes in the head and neck region and there may be skin lesions and pulmonary disorders. Median survival in one series was only three months.

Diagnostic issues

Confirming a diagnosis of HIV infection in young children can be difficult in resource-limited settings.

Clinical diagnosis

- The symptoms and signs are often rather non-specific. The most recent, modified WHO clinical case definition for paediatric AIDS is a useful tool for epidemiological surveillance but lacks sensitivity and has a low positive predictive value (PPV). It is therefore not useful for confirming a diagnosis of HIV infection in an individual child.
- The presence of oral candidiasis does not distinguish HIV-infected from HIV-uninfected children. However, failure of oral candidiasis to respond to treatment or rapid relapse is a highly specific sign of HIV infection.
- Chronic parotitis or shingles are unusual signs in healthy children, and are each highly suggestive of HIV infection. Herpes zoster ophthalmicus (i.e. shingles around one eye) is said to have >95% positive predictive value for HIV infection in African children.
- Geographic variation in patterns of disease must be recognised: *Penicillium marneffei* infection, an opportunistic fungal disease presenting with nodular skin lesions, is an AIDS-defining illness that has been reported in South-East Asia. Giant molluscum contagiosum has been a presenting sign in children in Eastern Europe.
- None of these clinical features is a sensitive marker of HIV infection in childhood populations, in that a minority of HIV-infected children manifest them.

Laboratory diagnosis

- The simplest lab test is an HIV antibody test (usually done by ELISA).
- Even this may not be affordable or available in many settings.
- Infants acquire maternal HIV IgG transplacentally, which can be detected by ELISA up to 18 months of age. Thus antibody tests cannot reliably distinguish infected from uninfected children until they are 18 months old.
- Many children born to HIV-infected mothers die before this age and a diagnosis of HIV infection remains presumptive, dependent on signs and symptoms of the terminal illness.
- To confirm the diagnosis in children <18 months, it is necessary to use assays that detect the virus itself or viral components: such tests include ***antigen detection tests, virus culture, amplification techniques and HIV-specific IgA tests***.
- ***Antigen test: the HIV nucleocapsid p24 antigen can be detected by ELISA, confirming infection. The sensitivity has been enhanced by acid dissociation or heat-denaturation techniques but a negative result does not exclude infection.***
- ***The following tests are only available in sophisticated laboratories and are therefore not done routinely in disadvantaged countries:***
  - ***Culture of the virus is technically difficult and is too expensive and slow for widespread diagnostic application even in developed countries.***
  - ***A variety of amplification techniques allow viral nucleic acid (DNA or RNA) to be detected at extremely low copy numbers. These techniques include polymerase chain reaction (PCR), nucleic acid sequence-based amplification (NASBA) and branched chain DNA (bDNA) amplification.***
  - ***These tests were originally developed to detect the HIV subtype B which is prevalent in the USA and Europe. In Africa the predominant subtypes are A, C, and D. In Asia, subtype E is predominant. It is important to ensure that whichever assay is used for the infant's sample can amplify the maternal virus, to avoid false-negative results from the children.***
  - ***In the first weeks of life, not all infected infants can be detected even by these highly sensitive methods, because virus is mostly acquired peripartum and takes time to establish a high level of viraemia. Therefore if resources are scarce, a single test is best postponed until beyond 30 days of life, at which point the sensitivity is .95%.***
  - ***Fifty microlitres of blood collected on a filter paper from a heel prick may be sufficient to detect the virus by PCR. Provided the samples are fully air dried before being put into plastic ziplocked pouches they can be stored indefinitely at room temperature. Transportation from the field to a centralised laboratory is***

easy and non-hazardous, but inevitably introduces delays and many families may not return for results.

- ***Rapid salivary tests (IgG or IgA) are under investigation and avoid the risks associated with obtaining blood from children (including needlestick injury to the healthcare worker). IgA is not transferred transplacentally and is therefore only detected in infected individuals, but it is not usually secreted in detectable quantities until at least 3 months of age. Salivary tests pick up antibody from the crevicular fluid which is a transudate that is secreted at the gingival margin and diluted in the mouth by saliva. It is important to reassure health professionals and lay people that although saliva tests may be positive this does not mean that HIV can be transmitted by kissing. It is extremely difficult to obtain culturable virus from saliva.***
- ***Improved rapid screening and confirmatory serological blood tests have been developed that require only a drop of blood to be added to reagents, requiring no laboratory skills. Whilst these have remarkably good specificity and sensitivity, they are still too expensive ($3 per test) for widespread screening in many countries.***

Ancillary lab tests

- ***Hypergammaglobulinaemia*** is strongly associated with HIV infection, but may also be elevated in children with recurrent or chronic infections such as malaria. The difference between total protein estimations and albumin levels (frequently available as part of routine liver function tests) reflect the globulin fraction: a total protein >85 g/litre with a normal albumin is suggestive of hypergammaglobulinaemia.
- ***A low CD4 count or CD4 : CD8*** ratio suggests HIV infection but requires highly specialised equipment. **A low total lymphocyte count is a much cheaper though** less specific surrogate marker of HIV infection.
- HIV infection can also cause anaemia or thrombocytopaenia. It is appropriate in children presenting with idiopathic thrombocytopenic purpura to test for HIV.
- Lack of thymic shadow on chest *X* ray is a feature of advanced disease, but is clearly not specific; the thymus tends to shrink in volume in response to a variety of acute infections in childhood.

Perinatally acquired HIV infection

- HIV1 transmission occurs more frequently than HIV2.
- Occurs in late pregnancy/during delivery/through breastfeeding
- Transmission is more likely with advanced maternal HIV disease:
  - Premature labour
  - Prolonged rupture of membranes
  - Contact maternal blood
  - In the first twin
  - Maternal genital infection
- Breastfeeding versus artificial feeding
  - Infant feeding is the most immediate issue which raises complex questions. Breastfeeding may double the risk of transmission. This risk is continuous throughout the period of breastfeeding but is not linear, being relatively higher in the earlier months. The risk of transmission must be balanced against the risks associated with artificial feeding. These include not only increased morbidity and mortality due to enteric and other infections, but financial pressures on the family, potential stigmatisation of the mothers, and erosion of community breastfeeding practices. Where safe alternatives are an acceptable and affordable alternative, mothers should be given information and support to feed their infants artificially.
  - If the mother decides to breastfeed, some evidence exists in favour of EXCLUSIVE breastfeeding, which means giving nothing other than breast milk throughout the period of breastfeeding. Many cultures give babies herbal infusions or water or other animal milk, particularly as a replacement for

TABLE 4.18.1. Signs and symptoms for use in endemic areas with limited access to diagnostic laboratories

Signs or condition specific to HIV infection	Signs uncommon in HIV-negative children	Signs common in both HIV-positive as well as ill non HIV-infected children
Pneumocystis pneumonia	Molluscum contagiosum	Persistent diarrhoea (>14 days)
Oesphageal candidiasis	with multiple lesions	Failure to thrive (especially in
Herpes zoster	Oral thrush (especially after the neonatal period,	breast-fed infants)
Lymphoid interstitial pneumonia	without antibiotic, lasting >1 month or recurrent)	Persistent cough (>1 month)
Kaposi's sarcoma	Generalised pruritic dermatitis	Generalised lymphadenopathy
Chronic parotid enlargement	Recurrent severe infections (three or more in 1 year)	Hepatosplenomegaly
	Persistent and/or recurrent fever lasting >1 week	
	Neurological dysfunction (progressive neurological impairment, delayed development, intellectual impairment, hypertonia)	

colostrum. Such alternatives are potentially antigenic, causing inflammatory changes in the infant gut mucosa and thereby increasing the risk of HIV transmission when maternal milk is ingested.

- Breast abscesses or cracked nipples have been associated with higher rates of HIV transmission, and subclinical mastitis has been implicated. Breastfeeding counsellors may have a role to play in reducing the complications.

  Many studies are underway exploring abrupt and early weaning at for example 4–6 months of age. Whether this is an acceptable and practical solution remains to be seen.

- Antiviral drugs.

  At the present time, the most affordable intervention is nevirapine administered as a 200 mg single oral dose to the mother in labour, and as a 2 mg/kg single oral dose to the baby at 48–72 hours of age (cost $4 per treatment).
- Elective caesarian section reduces risk by >50%, but in disadvantaged countries this benefit must be weighed against the higher morbidity and mortality from this mode of delivery in HIV infected mothers. The lack of trained personnel and the higher service costs also mitigate against this.
- If membranes ruptured >4 hours, give chlorhexidine vaginal disinfection.
- Avoid invasive procedures but without predjudicing the care of the mother/baby – forceps, ***scalp electrodes/samples***, artificial rupture of membranes.
- Treat genital infection.
- The use of antibiotics to decrease chronic chorioamnionitis, bacterial vaginosis, preterm labour and HIV transmission is under investigation.

Management

- The aim of treatment should be to maintain the best possible quality of life for the child for as long as possible, without bankrupting the family. This is a disease affecting the whole family, and the child must be treated in the context of all of the family's needs.
- Currently there are far more questions than evidence-based answers: published data on many management issues in the context of disadvantaged countries are not available.
- Much can be achieved with compassionate supportive care, by applying existing guidelines (such as Integrated Management of Childhood Illness algorithms) with an awareness of the need for early diagnosis and intervention in the HIV-infected child.
- Diagnosis of infections such as tuberculosis, lower respiratory infections, bacteraemia – particularly with non-typhoid salmonellae, staphylococci or streptococci – and opportunistic infections can be difficult, and often relies on empiric trials of therapy.

A low threshold for antibiotic use is appropriate, but may exacerbate diarrhoea and candidiasis, and may only be effective, in the presence of diarrhoea and malabsorption, if given parenterally.

Failure to thrive

Nutrition is a long-term concern in all infected infants. Stunting frequently develops within the first 12 months, although most children maintain normal weight-for-height ratios. Close monitoring of growth, and early protein/calorie, vitamin A and other micronutrient supplementation need evaluating.

- Regular vitamin A as per WHO guidelines (see Chapter 3.15)
- Supplementary feeding if possible (aim for 150 kcal/kg/day)
- Exclude/treat *Candida*
- Exclude/treat enteric infection
- Consider zinc deficiency (see Chapter 3.15)
- Consider fever
- Consider depression
- Consider pain

Respiratory disorders

Symptoms include cough, shortness of breath, fever, sweats and cyanosis.

The aetiology of acute respiratory infections is similar to community-acquired infections in immunocompetent children. However, they may require more prolonged courses of treatment: *Mycobacterium tuberculosis, Pneumococcus*, *Haemophilus influenzae*, *Staphylococcus aureus*, *Mycoplasma pneumonia* (see Chapters 3.1 and 4.10).

Specific HIV-related causes

Pneumocystis carinii pneumonia (PCP)

- Especially <6 months of age.
- Severe generalised pneumonia with ventilatory/perfusion mismatch and severe hypoxaemia.
- High fever is uncommon compared with bacterial pneumonia.
- Chest *X* ray, showing diffuse rather than local abnormalities, with interstitial infiltrates and hyperinflation.
- Nasopharyngeal aspirate has a low sensitivity with conventional staining techniques and requires ***PCR***. Induced sputum techniques greatly increase diagnostic yield. Beware risk of infection to operators especially of, for example, multiple drug-resistant tuberculosis. ***Bronchoalveolar lavage*** may provide diagnosis if adequate resources available.
- Treat with co-trimoxazole 60 mg/kg IV 12 hourly for a minimum of 7 days followed by oral for another 2 weeks (IV if severe nausea) PLUS high-dose dexamethasone for the first 5 days (150 micrograms/kg/dose 6 hourly for 4 days).
- Prevention: Ideally prophylaxis to all infants born to HIV-positive mothers after 6 weeks of age. Continue to 12 months if HIV-positive or suspected HIV infection. Give permanent prophylaxis to all children recovered

from PCP. Co-trimoxazole once daily on 3 days of the week ($<0.25\ m^2$=120 mg; 0.25–0.5 m^2=240 mg; 0.5–0.75 m^2=360 mg; 0.75–1.0 m^2=480 mg; $>1.0\ m^2$=960 mg).

This policy is under review because of possible co-trimoxazole-resistant organisms

Lymphoid interstitial pneumonitis (LIP)

Associated with increased risk of lower respiratory infection, including bronchiectasis. Also associated with lymphadenopathy, parotid, adenoid and tonsillar enlargement and clubbing (may produce sleep-related upper airway obstruction; see Chapter 3.19).

- Can produce severe ventilatory perfusion mismatch with hypoxaemia but may be asymptomatic.
- Chest *X* ray diffuse infiltrations and hilar lymphadenopathy persisting for >2 months despite antibiotic treatment.
- *X* ray appearance often more severe than clinical features.
- May be mistaken for miliary TB but the child is systematically too well.
- Probably related to Epstein–Barr virus infection.

Tuberculosis (see Chapter 4.10)

Tuberculosis should be treated with standard regimes, the emphasis being on achieving high adherence rates. The development of multidrug-resistant tuberculosis is a very real threat if compliance is poor. Directly observed therapy (DOT) may be the best approach.

- Diagnosis extremely difficult in HIV-infected patients.
- Avoid, if practical, contact with infected person.
- BCG at birth but NOT to any infant/child with active HIV.
- Give prophylaxis if HIV-positive child is in close contact with tuberculosis, but has not had BCG and skin test >5 mm but no evidence of active tuberculosis on chest *X* ray/examination. However, even early in HIV infection there is suppression of skin reaction to tuberculin testing. Prophylaxis is as follows:

  Isoniazid 10–15 mg/kg once daily up to 300 mg daily for 6 months. If resistant to isoniazid give rifampicin 10 mg/kg once daily plus pyrazinamide 35 mg/kg/day (maximum 2 g/day) for 2–3 months.

Other lung infections

Other opportunistic lung infections that may occur include *Pseudomonas aeruginosa*, *chlamydia, Mycoplasma*, *Cryptococcus neoformans*, *Aspergillus, Cytomegalovirus*, *Histoplasma*, *Coccidioides*, *Legionella* and *Nocardia*.

Gastrointestinal disorders

Oral/oesophageal problems

- *Candida* is the most common problem.
  - White plaques on mucosa, difficult to remove.
  - Loss of taste, pain on swallowing, reluctance to eat, increased salivation.
  - Crying during feeds.

  Oesophageal candidiasis is an AIDS-defining diagnosis and occurs in association with profound immune impairment (advanced HIV disease). Oral *Candida* may not be present and the only clinical symptoms may be reluctance to feed.

Oral treatment

- Local gentian violet 0.5% aqueous solution twice daily for 7 days (one teaspoon (5 ml) crystals/1 litre of water. Filter off residue. Use in 7days)

  or
- Nystatin 100 000 IU oral suspension 4–6 times/day for 7 days

  or
- Clotrimazole 1%, miconazole 2% gel, or amphotericin B suspension/lozenges 3 times daily

Oesophageal treatment (confirmed or suspected)

Fluconazole 3 mg/kg once daily or once every 48 hours in the neonate for at least 3 months.

- **Viral oesophagitis**
  - Herpes simplex infection (HSV): treat with aciclovir 20–40 mg/kg orally or IV four times daily for 7 days (maximum single dose=800 mg).
  - Cytomegalovirus (CMV) infection: treat with ganciclovir IV 5 mg/kg every 12 hours for 14–21 days.
- Reflux oesophagitis may also be present.
  Treat with antacids and or H_2 antagonist, for instance ranitidine 2 mg/kg twice daily (see Chapter 3.27).
- Idiopathic aphthous ulcers: if possible need to be differentiated from HSV ***by viral culture***. Pay attention to oral hygiene. Thalidomide is useful if they are severe.
- Severe periodontal/gingival disease (cancrum oris)
  - Treat with penicillin 6–12.5 mg/kg four times daily
  - Provide materials and education for dental hygiene
- Rarely malignancy (Kaposi sarcoma or non-Hodgkin's lymphoma) or oral hairy leucoplakia (white lacy markings on sides of the tongue associated with Epstein–Barr virus infection; no treatment required). Visceral Kaposi's sarcoma may present with persistent diarrhoea, intestinal obstruction and abdominal pain.

Persistent diarrhoea (see Chapter 3.26)

Case management should concur with the local prevalence of treatable infections. Giardiasis, Cryptosporidiosis, microsporidiosis, *Shigella*, *Salmonella*, *Campylobacter*, enteropathogenic *E. coli* and *Yersinia* may each contribute to gastronintestinal dysfunction. HIV itself may cause an enteropathy, and in highly immunosuppressed children, atypical mycobacterial infection and protozoa such as *Blastocystis hominis* may cause diarrhoea. Even with sophisticated microbiology, no pathogen may be found, and malabsorption due to lactase deficiency and other brush-border defects should be considered.

Chronic or recurrent diarrhoea

- Normal endemic pathogens may be responsible, for example: rotavirus, *Giardia lamblia*, *Campylobacter*

jejuni (see Chapters 3.25 and 4.11), salmonellae (typhoid and non-typhoid), *E. coli*, *Shigella*, *Entamoeba histolytica*, and *Strongyloides stercoralis*.
- Vitamin A supplementation may help (see Chapter 3.15).
- Opportunistic infections may occur
  - Bacterial: atypical mycobacteria infections, for example *Mycobacterium avium* complex (MAC) (see below).
  - Protozoa and parasites: cryptosporidia/microsporidia, *Isospora belli*. Treat with azithromycin 10 mg/kg once daily
  - Viral: cytomegalovirus, herpes simplex virus
  - Fungal: histoplasmosis, coccidiomycosis and *Candida.*
    Treat if severe with fluconazole 3 mg/kg once daily
  - May be secondary to antibiotics: direct effects or through *Clostridium difficile*. Stop antibiotics as soon as possible. Give live yoghurt with or without oral vancomycin
- Diagnosis: Stool microscopy/culture – fresh. Blood culture.
- Prevention: Care with food preparation, clean water, avoid bird and animal faeces, avoid swimming in fresh water, avoid reptiles (salmonellae).

Abdominal pain

Most frequently related to infections but occasionally tumours (non-Hodgkin's lymphoma and Kaposi sarcoma).

Malabsorption

HIV itself is associated with an enteropathy. Lactase deficiency and other brush-border defects occur. Consider trial of a lactose-free diet.

Central nervous system disorders

Found in 85% at postmortem.

HIV encephalopathy

- Rapid onset or chronic and relapsing forms.
- Hypertonic (spastic) diplegia and expressive language delay.
- Acquired microcephaly with developmental regression (loss of skills).
- White matter disease predominates. Does not cause seizures and therefore seizures need fully investigating for another pathology.

Encephalitis

Focal
- *Toxoplasmosis gondii*
  - Prevention: Avoid cats and cat faeces
    Avoid raw uncooked/partially cooked food
    Can be acquired congenitally
  - Diagnosis: ***Brain imaging and serology***
  - Treat with co-trimoxazole 60 mg/kg orally (IV if severe nausea) 12 hourly for 2 weeks.
  - Then life-long prophylaxis: sulphadiazine 85–120 mg/kg/day in two doses pyrimethamine 1 mg/kg/day (max. 25 mg) folinic acid 5 mg every 3 days
- *Papovavirus–JC*: Produces progressive multifocal leucoencephalopathy which is untreatable.
- Tuberculosis
- Lymphoma
- Fungal lesion: this is rare.

Diffuse
- CMV encephalitis: Treat with ganciclovir 5 mg/kg orally or IV 12 hourly for 14–21 days
- Malaria

Meningitis

- Bacterial meninigitis is no more common than in the normal population and has the usual spectrum of pathogens, for example *Pneumococcus*, *Haemophilus influenza* and *Meningococcus*.
- Viral
- Cryptococcosis and other fungi
  - Prevention: Avoid bird faeces
  - Clinical features: Chronic onset, headache common, fever, meningism usually but not always present
  - Diagnosis: Need to stain CSF sample with Indian ink
  - Treatment
    Fluconazole 6–12 mg/kg orally or if severe nausea IV once daily for 14 days
    High relapse rate, therefore give prophylactic fluconazole 3–6 mg/kg/day
- Syphilis
  Treat with benzylpenicillin IV 50 mg/kg every 6 hours for 48 hours then oral penicillin 25 mg/kg 6 hourly for 3 weeks (20% may have a systemic febrile response to penicillin)
- Tuberculosis
  See Chapter 4.10.

Cerebral abscess

Acute bacterial or tuberculosis.

Skin disorders

Seroconversion rash

Maculopapular erythematous (very rarely observed in infants).

Viral infections

Varicella
- Chickenpox can be very severe/fatal (affecting lungs/brain)
- Herpes zoster: can involve single or multiple dermatomes and may affect the eyes.
  Treat with:
  - **IV aciclovir (oral does not work)** ✔

<3 months	10 mg/kg 8 hourly
>3 months	20 mg/kg 8 hourly

- Valaciclovir is the prodrug of aciclovir and achieves excellent blood levels orally and is an alternative to IV aciclovir if available.
- ***VZIG within 96 hours of contact (if available)***

HSV1 and 2
Affecting lips, mouth and anogenital areas (rare in children unless sexually abused).
- May be recurrent and severe.
- Treat with oral aciclovir (20 mg/kg 4 times daily for 5–7 days (maximum single dose 800 mg)).

Molluscum contagiosum
Umbilicated papular lesions.

Optimally use ***highly active anti-retroviral therapy*** (no other measures are effective). If neglected, giant lesions can result which require surgical excision.

Measles
- Prevent by immunisation (see below and Chapter 1.29).
- May not have a rash.
- Giant cell pneumonitis may occur.
- Treat with Vitamin A and human ***immunoglobulin (if available)***.

Viral warts
These can be persistent and severe. Topical treatment is ineffective (see Molluscum contagiosum above).

Bacterial infections

Impetigo and furunculitis due to *Staphylococcus aureus* are common. Treat with (flu)cloxacillin 12.5–25 mg/kg four times daily orally.

Fungal infection

Fungal infection is common and involves nails, feet, hands and groin. Treat with topical antifungal cream (miconazole 2% twice daily until healed). If severe, fluconazole 3 mg/kg once daily orally or griseofulvin (10 mg/kg once daily after food). Will need treatment for 4–6 weeks.

Seborrheic dermatitis and pityriasis versicolor
- Hypopigmented, scaly, macules are present
- Caused by the yeast *Malassezia furfur*
- Treat with aqueous cream, selenium-based shampoos; topical antifungal creams with or without 1% hydrocortisone. If severe, give oral fluconazole 3mg/kg daily.

Non-specific pruritic papular rash

- A common and severe problem for children with HIV infection.
- Treatment:
  - Bathe in a skin antiseptic wash (for example dilute chlorhexidine solution)
  - Antihistamines: chlorpheniramine

<1 year	1 mg twice daily
1–5 years	1–2 mg three times daily
6–12 years	2–4 mg three times daily
>12 years	4 mg three times daily

  - Aqueous cream and calamine lotion may help

Drug side effects

- Can be severe, for example erythema multiforme, Stevens–Johnson syndrome or toxic epidermal necrolysis.
- Commonest with co-trimoxazole, sulphadiazine, antituberculous drugs (such as thiacetazone).
- Management: withdraw drug and treat symptoms.

Infestations

Sarcoptes scabiei (see Chapter 3.47) may occur as in children withour HIV infection.

Malignancy

Kaposi's sarcoma. This affects the skin of the upper body/feet. There are single/multiple, flat/raised, purple red plaques. Lymphadenopathy is a common presentation. Diagnosis is confirmed by biopsy of a lesion or lymph node.

Eye involvement (see Chapter 3.34)

- HIV retinopathy: microangiopathy with soft exudates. Is asymptomatic and does not require treatment. Needs to be differentiated from tuberculosis.
- Cytomegalovirus retinitis: is the most common cause of visual loss in HIV. Treat with ganciclovir 5 mg/kg IV or orally 12 hourly for 14–21 days.
- Herpes zoster: may produce corneal ulceration and retinal necrosis.
- Toxoplasmosis.
- Malignancy for example non-Hodgkin's lymphoma or Kaposi's sarcoma.

Mycobacterium avium complex (MAC)

- Produces a systemic infection with fever, chronic diarrhoea, abdominal pain, chronic malabsorption, generalised lymphadenopathy, obstructive jaundice (from lymph node enlargement around porta hepatis).
- Treat with:
  clarithromycin 7.5 mg/kg twice daily IV or orally
  or azithromycin 10 mg/kg once daily
  AND ciprofloxacin and rifabutin
- In rich countries, prophylaxis with the above drugs if ***CD4 cell counts $<$ 50/mm^3***.

Fever of unknown origin

- HIV infection itself can cause fever.
- In an endemic area always treat for malaria (ideally after a blood film). Malaria has not usually been reported to be more severe in HIV-infected children in terms of parasite density or response to treatment. The main interaction between the two diseases has been the acquisition of HIV by children through transfusion for malaria-associated anaemia.

- Have a low threshold for diagnosing septicaemia and meningitis and giving powerful empiric antibiotics if severe sepsis is suspected.
- Consider tuberculosis and non-Hodgkin's lymphoma.

Immunisation

- Immunisations should generally follow the Expanded Programme on Immunisation scheme, with the ✔ **exception of BCG and yellow fever which should not be given to HIV symptomatic children**. In HIV endemic areas, BCG is routinely administered postnatally. This should be given even to infants of mothers known to be HIV infected, since the damage to the immune system generally occurs after the onset of viraemia, i.e. after the first 6 weeks of life. There is no evidence of frequent dissemination occurring after neonatal administration of BCG, although BCGosis is not an easy diagnosis to establish, and there may be unrecognised cases.
- There are theoretical risks from giving live oral polio vaccine, particularly to other immunocompromised members of the household. However, cases of vaccine-associated paralytic illness are rare and oral poliomyelitis vaccine (OPV) continues to be recommended.
- Live attenuated measles vaccine is recommended by WHO for children in disadvantaged countries, where the risks from wild-type measles virus are high. Responses to the vaccine tend to be lower in HIV-infected children with more advanced disease. WHO recommend giving an extra dose of measles vaccine at 6 months as well as standard dose at 9 months to HIV-infected children.
- Results from studies of conjugated pneumococcal vaccines are awaited with interest, and the role of Hib immunisation needs evaluating.

Antiretroviral therapy

- Highly active antiretroviral therapy (HAART) has radically altered the natural history of HIV in children, but a minimum of three drugs are needed for long-term suppression of the virus, and careful monitoring of treatment is required (cost around $10 000 per annum).
- Antiretroviral therapy will only be sustainable in countries with robust economies, although wealthy individual families may be able to afford treatment.
- The use of suboptimal single or dual combinations of antiretroviral drugs is controversial. Studies in rich countries have shown that a small proportion of children have had sustained benefit from such interventions. For the majority however, any benefits are short-lived (typically six months for monotherapy with a nucleoside analogue such as zidovudine (AZT). The limited benefits for an individual child must be carefully considered in the light of the family's resources, and unrealistic expectations must be avoided.
- A comprehensive and regularly updated guide to the current use of antiretroviral agents in children in the USA can be accessed via the web [http://www.hivatis.org] *Working Group on Antiretroviral Therapy and Medical Management of HIV Infected Children. Guidelines for the use of antiretroviral agents in paediatric HIV infection.*
- At present, the priority for the use of antiretrovirals in disadvantaged countries is in the prevention of mother–infant transmission.

Encouraging new data are emerging on the ability to restore HIV-specific immunity in adults treated during acute infection, using HAART and closely supervised treatment interruptions. Given the plasticity of the infant immune system and the fact that the timing of transmission is known, there is great current interest in early antiretroviral treatment and similar immune boosting strategies that may provide affordable long-term control of the disease. This is a fast moving field. It is not an unrealistic goal to make effective antiretroviral treatment available for infected children wherever they live.

Terminal care of the child dying from HIV Infection

See Chapters 1.27 and 1.28. Local groups for the support of families with HIV infection are essential and ideally should be funded by local government.

Summary

- The major practical focus should be on prevention of childhood HIV infection. Implementing effective strategies for reduction of mother–infant transmission, such as prenatal counselling and screening of mothers and administration of antiretroviral drugs for mother and baby. Unfortunately establishing the infrastructure to implement effective interventions is lagging far behind the scientific advances in this field.
- Limiting the use of blood transfusions and trying to ensure the blood supply is safe, and preventing sexual transmission amongst adolescents are vital public health issues.
- Identifying a child as HIV-infected should not be an excuse for therapeutic nihilism. Much can be achieved using algorithms such as those provided by the Integrated Management of Childhood Illness (IMCI), with a heightened awareness of the need for early diagnosis and intervention.
- Surmounting the sense of hopelessness amongst health professionals who are dealing with overwhelming numbers of patients without resources is a critical issue. This may come in part from research that identifies practical interventions which improve the quality of life for HIV-infected children and their families.
- It is essential that disadvantaged countries be permitted by multinational drug companies to develop low cost and effective forms of HAART, without being limited by International Patent regulations.

419 Measles

Bernard Brabin

Minimum standards requirements

- Immunisation (see Chapter 1.29)
- Vitamin A (see Chapter 3.15)
- Oral rehydration solution (Chapter 3.25) and nutritional provision (Chapter 3.16)
- Antibiotics for secondary infection
- Oxygen (Chapter 3.1)
- Nebulised epinephrine and corticosteroids for croup (Chapter 3.17)
- Eye pads (Chapter 3.34)
- Public health measures

Clinical features

- Prodromal period (3–5 days): acute coryza-like illness with fever, cough and conjunctivitis. Febrile seizures may occur. Koplik's spots appear by days 2–4 (tiny bluish white specks upon a red base on the buccal mucosa of the cheeks; they look like grains of salt).
- Maculopapular rash (day 4), commencing on face and neck, behind the ears and along the hairline and spreads to become generalised and reaching the feet after day 3. Fades after 5–6 days in order of appearance, developing brownish colour and often becomes scaly. If severe there may be petechiae and ecchymoses.
- Persistence of fever beyond day 3 of rash is usually due to complications. Rash is due to infiltration of lymphocytes into areas of virus replication in skin.

Epidemiology

- Droplet spread of virus in nasopharyngeal secretions. Is most infectious before appearance of rash and for at least 7 days after onset of first symptoms. Incubation period is 10–12 days. Quarantine can be lifted 2 days after fever subsides.
- Epidemic cycles of infection in urban areas may occur every two years. In isolated communities, all age groups are affected. In disadvantaged countries, population peak incidence is at 1–2 years, with mortality between 1% and 5%, although during epidemics it may rise to 30%. Mortality is low in the well nourished. Children who acquire infection in overcrowded conditions tend to have more severe disease, probably due to a larger infecting dose of virus.

  Pneumonia accounts for about three quarters of measles deaths. Measles is more severe in HIV-infected children.
- In disadvantaged countries, measles commonly occurs in previously vaccinated children. This is partly explained by a persistent maternal antibody at 9 months of age when vaccine is usually given.
- Rare in infants <3 months.

Complications

- Recovery following acute measles may be delayed for weeks or months due to failure to thrive, recurrent infections, persistent pneumonia and diarrhoea.
- **Pneumonia (see Chapter 3.1)**
  - Bacterial pneumonia usually occurs during convalescence and after several days of an afebrile period. Most frequent cause of death. Incidence 10–25% of hospitalised cases in developing countries.
  - Viral pneumonia occurs during acute phase of measles and may progress to giant cell pneumonia in immunosuppressed (for example leukaemia, HIV).
  - Mediastinal emphysema occurs in 1 in 300 measles cases and may lead to subcutaneous emphysema.
- **Diarrhoea**. Incidence 20–40%. May become persistent and frequently precipitates malnutrition (see Chapter 3.16).
- **Tracheobronchitis**. Presents as croup. Laryngeal tissue sometimes becomes necrotic which may lead to laryngeal obstruction (see Chapter 3.17).
- **Otitis media**. Common, especially in infants. Mastoiditis may develop. Important cause of chronic otitis media and hearing impairment (see Chapter 3.18).
- **Stomatitis**. Mucosal inflammation and ulceration with bleeding gums and secondary *Candida albicans* and herpes simplex infections. Causes difficulty in eating and worsens malnutrition. Cancrum oris (noma) may develop.

- **Xerophthalmia**. Vitamin A deficiency may combine with measles to precipitate xerophthalmia and blindness.
- **Malnutrition**. Malnutrition secondary to measles results from anorexia and poor nutrition following infection. Mortality is high (>15%) (see Chapter 3.16).
- **Tuberculosis**. Tuberculosis, including tuberculous meningitis, may first be noticed in the post measles period (see Chapter 4.10).
- **Encephalitis**
  - Acute allergic encephalitis: a demyelinating disorder and the most common CNS complication of measles. Onset often in second week as exanthem is clearing. Occurs in 1–2 per 1000 cases of measles. Virus is not found in brain.
  - Acute measles inclusion body encephalitis: results from direct invasion of brain cells by virus (virus may be isolated from CSF). More rapid onset if there is immunosuppression or malignancy.
  - SSPE (subacute sclerosing panencephalitis): long latent period (several years) between infection and onset of symptoms. Commonly, measles occurred at an early age. Characterised by lethargy, psychological changes, myoclonic jerks, and mental deterioration eventually leading to death. Virus has been isolated from brain biopsy specimens.
  - Atypical measles may have prolonged fever and present with pneumonia or rarely encephalitis. Rash may or may not appear. Prolonged fever for 2–3 weeks with diarrhoea may simulate enteric fever.

Differential diagnosis

- Other exanthema and drug reactions.
- Koplik spots are the most helpful diagnostic feature in the prodromal period.

Case assessment and classification

Cases may be classified into:
- Uncomplicated measles
- Severe measles requiring treatment or urgent referral.

Management

Mild measles

- Small frequent feeds. Infants should continue breastfeeding. Extra energy should be provided with vegetable oil or sugar to cereals (a teaspoon of each). Follow-up nutritional support.
- Paracetamol for temperature >39°C.
- Saline drops for blocked nose.
- Oral hygiene by rinsing mouth several times daily. Apply 1% gentian violet to mouth sores. Treat oral thrush (see Chapter 4.18).
- If mouth ulcers secondarily infected, use antibiotic (penicillin or metronidazole orally for 5 days.
- If mouth too sore to feed or drink, nasogastric tube may be required.
- Ocular hygiene for purulent conjunctivitis, daily washings (with sterile 0.9% saline or boiled water using cotton-wool swabs and tetracycline eye ointment three times daily. NEVER USE TOPICAL STEROIDS. Consider protective eye pads.
- Vitamin A capsule 200 000 IU (>1 year) or 100 000 IU (<1 year). **Give a second capsule the next day**.
- Oral rehydration solution (ORS) for diarrhoea.
- Oral antibiotic if clear indication of lower respiratory tract infection (co-trimoxazole, amoxicillin, ampicillin) (see Chapter 3.1).
- Admit if signs or symptoms of severe measles.

Table 4.19.1 Clinical features of severe disease

Signs	Complications
Cough, tachypnoea, or chest indrawing	Pneumonia
Stridor when quiet	Croup, necrotising tracheitis
Severe diarrhoea	Dehydration
Recent severe weight loss	Malnutrition
Corneal damage or Bitot spots	Blindness
Ear discharge	Otitis media, deafness
Lethargy, convulsions	Encephalitis
Inability to drink or eat	Dehydration, malnutrition
Blood in the stools	Dysentery, haemorrhagic measles
Severe stomatitis	Cancrum oris

Severe measles

Admit and isolate.

In addition to the care for mild measles:
- Parenteral antibiotics for pneumonia or septicaemia, for example benzylpenicillin or ceftriaxone/cefotaxime if available. (Flu)cloxacillin plus gentamicin or cefuroxime (if available) if *Staphylococcus aureus* is suspected. If stridor associated with fever is present use ceftriaxone/cefotaxime (if available) or chloramphenicol. Rapidly spreading pulmonary tuberculosis may be difficult to distinguish from a progressive pyogenic pneumonia.
- Oxygen as required.
- Croup: nebulised epinephrine, 1 ml epinephrine (1 in 1000) mixed with 1 ml of saline every 2 hours, **careful observation** (see Chapter 3.17 which also describes the use of oral steroids or nebulised budesonide which can be life saving in this situation).
- Diarrhoea: oral rehydration and appropriate antibiotic if bloody stools. Persistent diarrhoea requires nutritional support.
- Otitis media: antibiotics and regular aural hygiene. Screen for hearing impairment during follow-up.
- Xerophthalmia: protective eye pad, give vitamin A capsules (see above).

- Malnutrition: treat according to management guidelines (see Chapter 3.16).
- Encephalopathy: follow management guideline for convulsions and coma (see Chapters 3.36 and 3.38).

Prevention and follow up

- ***Gammaglobulin*** (if available) for susceptible contacts of measles cases if <1 year old.
- Improve vaccination coverage (see Chapter 1.29 on immunisation).
- Follow up vitamin A dose if malnourished or eye disorders.
- Measles control by immunisation is one of the most important public health interventions in reducing child mortality. If measles admitted, immunise all other unimmunised children >6 months in the hospital, with follow up second dose in all aged 6–9 months as soon after 9 months as possible.

4.20 Mumps

James Tumwine

Mumps is caused by a virus of the paramyxovirus family (also measles and parainfluenza). The virus is spread by airborne droplets through the respiratory tract, mouth, and possibly conjunctivae and urine, and is present in saliva, CSF, blood and urine.

Clinical presentation

- Incubation period is 14–24 days. Onset is with painful swelling of parotid glands, fever, general malaise, and occasionally headache. Parotid swelling may be unilateral at first, followed a couple of days later by swelling of the opposite parotid gland, with pain on opening the dry mouth.
- Mild meningoencephalitis is common, there may be nausea and vomiting, and abdominal pain.
- Orchitis presents with fever and tender oedematous swelling of the testis. In 10–20% of cases the second testicle may be affected. However, infertility is rare.
- Differential diagnosis of parotitis includes cervical adenitis, pyogenic parotitis, recurrent parotitis, tumours of the parotid and tooth infections.
- Mumps orchitis can mimic hernias, tumours, haematomas, epididymo-orchitis and testicular torsion.

Complications

Complications include oophoritis, mastitis, pancreatitis, nephritis, myocarditis, thyroiditis, labyrinthine disturbance, painful swelling of the lacrimal glands, optic neuritis, uveokeratitis, rapid loss of vision, arthritis, jaundice, pneumonia and thrombocytopenia. Transient or permanent unilateral nerve deafness has been reported.

Infection during pregnancy very rarely causes disease of the fetus, for example aqueductal stenosis and hydrocephalus.

Management

Symptomatic treatment includes analgesics, fluids and scrotal support for orchitis. Value of corticosteroids for orchitis is not established.

Prevention

Measles, mumps, rubella (MMR) immunisation is routine in rich countries and has reduced mumps by over 90%.

4.21 Poliomyelitis

Allie Moosa and Charles Newton

Minimum standards requirements

- Immunisation
- ABCD including airway protection if bulbar palsy
- Bed rest and physiotherapy
- Nutritional support and hydration
- Nasogastric feeding
- Intensive supportive care for bulbar palsy
- Public health measures

- Due to both vertical and mass vaccination campaigns, the number of reported cases has fallen by 90% since 1988. Wild polio virus is now mainly confined to South Central Asia, especially the Indian subcontinent, and sub-Saharan Africa.
- Poliomyelitis is caused by polioviruses type 1, 2 and 3 that are ingested and multiply in tonsils and Peyers' patches of the gut. In most cases, infection is contained at this point and the child is asymptomatic.

Severity

Minor illness

Associated with viraemia and non-specific symptoms, for example nausea, vomiting, abdominal pain and sore throat.

Major illness

- Non-paralytic poliomyelitis
  Occurs in minority of symptomatic children. Incubation period is 10–14 days and symptoms include: fever, headache and 2–5 days later, signs of meningeal irritation with severe pain and stiffness of neck, back and limbs.
- Paralytic poliomyelitis
  - Paralysis occurs within first two days of major illness.
  - Affects any muscles but particularly large ones and those of lower limbs.
  - Asymmetrical paralysis, flaccid muscles, absent tendon reflexes. Intact sensation. Paralysis is maximal within 3–5 days of onset and rarely extends once temperature is settled.
  - In **bulbar form**, involvement of cranial nerve nuclei and vital centres in the brainstem results in paralysis of facial, pharyngeal, laryngeal and tongue muscles, causing swallowing difficulties, aspiration and respiratory failure.
  - Hypertension may occur; also transient bladder paralysis.

Diagnosis

CSF initially shows neutrophil predominance and after 5–7 days is mainly lymphocytic. Protein is normal or slightly elevated. Glucose is normal. Virus can be isolated from throat and stool for up to three months after onset. Differential diagnosis includes other causes of acute flaccid paralysis (see Chapters 3.39 and 3.40).

Prognosis

- Depends on extent of paralysis and quality of care during acute phase.
- Early identification of and intervention for respiratory and bulbar paralysis will reduce mortality to 5–10%.
- With appropriate physiotherapy, improvement in function of paralysed muscles can occur for up to 18 months.
- Factors that adversely affect outcome include intramuscular injections, muscle fatigue, corticosteroid therapy and immunocompromised states. Removal of tonsils and teeth during incubation period increases risk of bulbar paralysis.

Management

Acute phase

- Absolute bed rest is mandatory, avoid intramuscular injections and exercise.
- Analgesics for severe pain. Keep paralysed muscles in neutral position to prevent contractures.

- Gentle passive exercises and warm compresses to help relieve pain. Active exercises are introduced when temperature has settled for a few days.
- Respiratory paralysis requires ventilatory support; also bulbar paralysis, nasogastric tube feeding and possibly tracheostomy.

Convalescent phase

- Aim to improve motor function, prevent deformities and generally reintegrate child into society.
- Encourage active participation by parents in rehabilitation process. Educational and emotional needs of child must not be neglected. Services of orthopaedic surgeon and orthoptist may be required.

Prevention

See Chapter 1.29 on immunisation.

4.22 Rabies

Mary Warrell

Minimum standards requirements

- Immunisation
- Wound care
- Rabies immune globulin
- Epinephrine (in case of anaphylaxis)
- Sedatives and terminal care to relieve suffering (see Chapter 1.28)

Clinical features of rabies encephalitis

Rabies virus infects some mammal species and when transmitted to man the incubation period is usually 20 to 90 days. Virus travels up the nerves and once it is in the brain the disease is invariably fatal. The illness begins insidiously with non-specific physical or behavioural signs. The site of an animal bite often becomes itchy or painful. Furious and paralytic (dumb) forms of rabies can occur. Furious rabies is characterised by agitation, hyperexcitability and hydrophobia which is due to spasms of the inspiratory and perhaps laryngopharangeal muscles. These occur on attempting to drink water or from a drought of air. Flaccid paralysis without hydrophobia occurs in some patients. Diagnosis during life depends upon ***finding virus antigen in skin biopsies, or culture of virus from, or PCR on, saliva or CSF***. A postmortem needle biopsy of the brain shows rabies antigen in neurones using an immunofluorescent test. A few patients have survived some months, although with gross CNS sequelae, after intensive management.

Management of rabies encephalitis

If the diagnosis is certain, the treatment of rabies encephalitis is palliative and terminal care should ensure adequate sedation and analgesia if necessary. Relatives and staff should wear gloves when handling the child and secretions. Although there is a theoretical risk of transmission of infection to attendants, this has not been virologically documented, so the risk of infection is very small. However if rabies vaccine is available, pre-exposure treatment should be offered to them.

Aetiology

Dog bites are the usual cause of rabies in man, but cats, wolves, foxes, jackals, mongooses, and rarely domestic animals, bats and other mammals may transmit the infection to humans.

Estimating the risk of exposure to rabies

- Is there a bite wound with broken skin? Have mucous membranes or an existing skin lesion been contaminated by virus in the animal's saliva? Intact skin is a barrier against the virus.
- How did the animal behave? An unprovoked attack by a frantic dog is a high risk, but so is contact with a paralysed animal, or an unusually tame wild mammal.
- Is the biting animal a local rabies vector species, or could it have been infected by a vector, for example a fox?
- Vaccinated animals are unlikely to be rabid, but vaccinated dogs can transmit the infection.
- If possible have the animal's brain examined for rabies. Alternatively have the animal kept under safe observation, and stop vaccine treatment of the patient if it is still healthy after 10 days.

Post-exposure treatment

This is very urgent. ✔

Wound care

This is important for all bites, irrespective of rabies risk.

- Scrub and flush the lesion repeatedly and energetically with **soap or detergent and water**. Remove foreign material. Local analgesia may be necessary (see Chapter 1.13).
- Apply povidone iodine (or 70% ethyl alcohol, **but this is painful!**).
- Do not suture wound, or at least delay suturing.
- Give tetanus immunisation if appropriate.
- Treat bacterial infection of wounds with oral antibiotic, for example amoxicillin/clavulanic acid 25–50 mg/kg/day in 3 divided doses (dose based on amoxicillin content) or tetracycline 25–50 mg/kg/day in 4 divided doses (not advised 8–10 years beause of dental staining).

Rabies vaccine

Active immunisation with vaccine should be given whenever there is a risk from contact with a suspect rabid animal. Rabies vaccines are suitable for people of all ages including pregnant women.

Vaccines recommended by the WHO are:

- Purified chick embryo cell vaccine (PCEC): rabipur
- Purified vero cell vaccine (PVRV): verorab (0.5 ml/vial).
- Purified duck embryo vaccine (PDEV): lyssavac N
- Human diploid cell vaccine (HDCV).

Three postexposure regimens

- Standard intramuscular regimen
  One ampoule (1 ml or 0.5 ml) intramuscularly (IM) into the deltoid, or anterolateral thigh in small children, on days 0, 3, 7, 14 and 28, a total of five doses. ***Do not inject into the buttock.***
- Economical eight-site intradermal regimen
  (Use vaccines containing 1 ml per ampoule. Total of less than two ampoules needed).

  Day 0: draw up 1 ml of vaccine into 1 ml (Mantoux type) syringe. Inject 0.1 ml intradermally (ID) into each of eight sites (deltoid, thigh, suprascapular and lower anterior abdominal wall) using all the vaccine (see Figure 4.22.1).

  Day 7: give 0.1 ml ID into four sites (deltoid and thighs).

  Days 28 and 90: 0.1 ml ID at one site.

This regimen is the treatment of choice when RIG (rabies immunoglobulin, see below) is not available.

- Economical two-site intradermal regimen

  Dose: 0.1 ml for PVRV and 0.2 ml for all other vaccines.

  Days 0, 3 and 7: give ID into two sites (deltoids).

  Days 28 and 90: ID dose at one site.

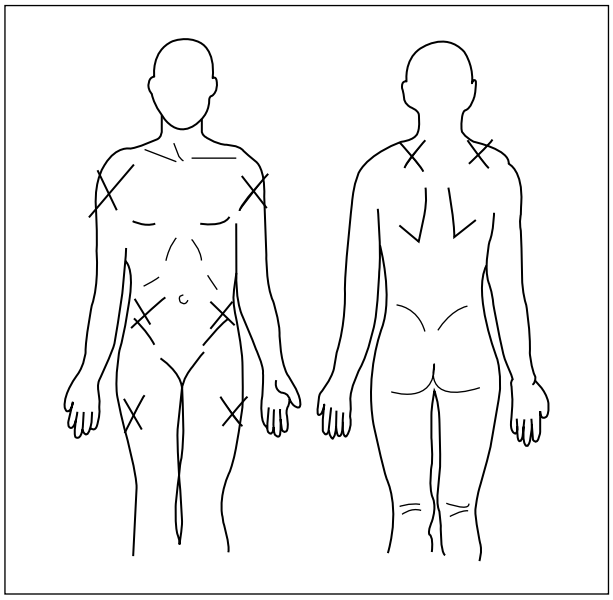

Figure 4.22.1 Distribution of intradermal injections on day 0 for eight-site postexposure rabies vaccine regimen. Injections on day 7 are over deltoids and thighs only.

In both the eight- and two-site intradermal regimens, **take care that intradermal injections raise a papule**. If vaccine goes subcutaneous, repeat the injection nearby. Ampoules may be shared between patients if stored at 4°C for one day (or longer if preservative present). **A sterile needle and syringe must be used for each patient**.

Unused vaccine may be used for pre-exposure treatment (for example of relatives or hospital staff); see below.

If treatment is delayed >48 hours after bite or immunosuppression suspected (for example severely malnourished, AIDS or corticosteroids therapy), give the first dose of an intramuscular course intradermal at eight sites (see above), or for two-site regimen (see above), double the first dose. No change in dosage for the eight-site regimen.

- **Side effects of vaccines** are mild local or non-specific generalized symptoms. Transient maculopapular or urticarial rashes are occasionally seen.
- **Other vaccines**
  Tissue culture rabies vaccines not listed above and vaccines of nervous tissue origin (for example Semple vaccine and suckling mouse brain vaccine), should be used according to manufacturers' instructions.

Rabies immunoglobulin (RIG)

- Passive immunisation with RIG is recommended to accompany vaccine following all contacts with suspected rabid animals where skin has been broken or mucous membranes have been contaminated.
- It is vital for bites on head, neck, hands or in multiple bites.
- Dosage: equine RIG (40 IU/kg) or human RIG (20 IU/kg) is infiltrated into and around wound on day 0. If not anatomically possible (for example on a finger) inject any remainder intramuscularly, at a place remote from vaccine site, but not into the buttock.
- **Skin tests are not useful in predicting anaphylactoid reactions to equine RIG. Take care that epinephrine is immediately available in case of an anaphylactoid reaction (see Chapter 1.23)**. Patients with signs such as urticaria or dyspnoea should be treated with epinephrine 10 micrograms/kg IM (0.01 ml/kg of 1 in 1000 solution [1 mg/ml]). An antihistamine, for example chlorpheniramine (200 micrograms/kg IM or IV by slow injection) should also be given and hydrocortisone 4–8 mg/kg IV or IM should also be given (maximum dose 100 mg). ✔

Postexposure treatment for previously vaccinated patients

Anyone requiring postexposure treatment who has previously had a complete pre- or postexposure course of one of the vaccines listed above, may have a short booster course:

- One dose of vaccine IM or ID on days 0 and 3. RIG is not necessary.
- **Treatment and thorough wound care is still urgent**.

Pre-exposure treatment

- Pre-exposure vaccination is the best means of rabies prevention. No one who had pre-exposure treatment and postexposure booster injections is known to have died of rabies.
- Pre-exposure regimen; three doses required: Inject one dose of a vaccine IM (1 ampoule) or ID (0.1 ml) on days 0, 7 and 28 (or 21). One or two days variation does not matter. A booster after one year enhances and prolongs antibody response.
- Patients on malaria prophylaxis with chloroquine should have IM not ID injections for pre-exposure treatment.
- **If contact with a rabid animal occurs, postexposure booster vaccine treatment is still required** (see above).

4.23 Viral haemorrhagic fevers

James Bunn

Minimum standards requirements

- ABCD (see Chapter 1.19)
- Treat shock (see Chapter 3.6)
- Blood transfusion and clotting factors (see Chapter 1.14)
- Isolation and infection control
- Intensive/supportive care (see Chapter 1.25)
- Public health measures

Overview

- Viral haemorrhagic fevers (VHFs) are a group of severe infections caused by viruses that normally affect animals.
- Human infection is characterised by high fever and, in a proportion, haemorrhage.
- Animal hosts such as rodents are usually asymptomatic and are often infected with virus since birth, excreting it in urine or body fluids throughout life.
- In primary cases, transmission to humans occurs by a variety of routes, such as food contaminated with urine (for example Lassa, Junin, Machupo and Hantaan fevers) via an arthropod vector such as ticks (Crimean–Congo and Omsk fevers), or mosquitoes (Rift Valley fever). Hosts for Ebola and Marburg haemorrhagic fevers are not yet known.
- Humans with disease are usually highly infectious.
- Most VHFs cause severe disease with a high mortality, especially following human-to-human spread (secondary cases).
- Some (for example Lassa) may also cause asymptomatic or mild illness.
- Symptomatic disease is commonly mistaken for other febrile illnesses, typically malaria, typhoid fever, or *Shigella* dysentery, which fail to respond to treatment.
- Individual VHFs are geographically restricted in distribution. As for all geographical illnesses, you only need to know of those present in the local area. VHFs are fortunately rare.

Lassa fever

- Distribution: West Africa (Nigeria, Sierra Leone, Liberia, Guinea).
- Host: Mastomys rat (habitat is rural).
- Transmission
  Primary: Mainly from contact with host (rat) urine. Food may be contaminated.
  Secondary: Patient to carer, or to hospital and laboratory staff is common, particularly from haemorrhagic cases. Maternal illness is particularly severe, with a high risk of vertical transmission to the baby (which is invariably fatal).

Prevalence

Relatively common.

- Most primary human infections are not severe, and many are subclinical.
- Childhood seroprevalence in Sierra Leone can be as high as 20% in some rural villages.
- Outbreaks may occur in displaced communities or when humans enter host habitat.

Clinical features

- High fever (>39°C), with cough and vomiting in 65% of hospital cases.
- Abdominal pain and diarrhoea are common (35% of cases).
- In children wheeze and pleural effusions are more frequent than in adults.
- Sore throat and pharyngeal ulcers occur less frequently than in adults, but are highly suggestive of Lassa.
- In children oedema, especially of the face, and overt bleeding are seen in 10% of cases, and in a febrile child from an appropriate area should suggest Lassa fever.
- At the epicentre of the transmission area, Lassa is a common cause of a febrile child with convulsions.

Diagnosis of Lassa fever

- Clinical case diagnosis
  An unexplained febrile illness compatible with Lassa, in a child from an area of known transmission, with no response in fever or illness to an antimalarial plus a broad-spectrum antibiotic (for example chloramphenicol).

  Note: Malaria parasitaemia in an area of endemic malaria transmission is not sufficient to exclude other

causes of fever (for example VHF) as the cause of a febrile illness, as many adults and older children may have coincidental "asymptomatic" malaria parasitaemia.

- Supportive indirect laboratory tests
  - Raised liver transaminases AST/SGOT (in adults this reflects poorer prognosis).
  - Low initial white blood cell counts, but often have a normal platelet count.
- Confirmation of diagnosis.
  - ***Positive specific IgM serology*** (on admission only 50% of cases are positive).
  - ***Rising IgG titres*** to Lassa on acute and convalescent serum.
  - ***Isolation of virus***. This is rarely appropriate, and ✔ **due to the high risks of laboratory infection samples should not be taken without senior expert advice**.

Samples need to be marked as high infection risk ideally with standard yellow hazard tape, and sent in two sealed plastic bags. Samples should only be taken where laboratory staff are aware of the potential risks, and have the necessary precautions to handle such specimens safely. The lab should be informed that the specimen is coming.

Management

- Appropriate symptomatic management of fever, distress and pain.
- Fluid and nutritional requirements.
- Supportive care includes oxygen (if hypoxic) and initial intravenous volume replacement if hypovolaemic (see Chapter 3.6 on shock).
- Blood transfusion may be required for a falling PCV or haemorrhage. Fresh frozen plasma (FFP) may not be of benefit, as inhibitors of clotting factors may cause bleeding.
- ***Early ribavirin can improve prognosis in severe disease but is very expensive.***

Infection control

See below and Chapter 1.6.

Ebola

- Distribution: Central Africa (Sudan, Democratic Republic of the Congo, Gabon, Cote d'Ivoire Uganda).
- Host: Main animal reservoir unknown.
- Transmission

  Primary: Infection occurs mainly in adults trekking in tropical Central African forests. Transmission from primates to humans is recorded.

  Secondary: Patients with advanced disease are viraemic and highly infectious.
  - Once in a human host, transmission to carers, hospital and laboratory staff is frequent (30% of doctors in an outbreak in Kikwit, Democratic Republic of the Congo, developed Ebola), however once effective infection control measures have been implemented, secondary cases are rare.
  - The disease is invariably severe, with a high death rate, but only 20% of cases in a Democratic Republic of the Congo outbreak were under 15 years old.
  - Children are at low risk in the community, and boys, have half the incidence of girls, possibly because they are less involved in the care of sick adults.
  - Invariably in children, there is a history of contact with a primary case and an outbreak of an illness, which could be Ebola is present in the hospital/community.
  - Postmortem transmission does occur, possibly through skin contact.

Prevalence

Low. Occurs sporadically in well-localised outbreaks.

Clinical disease (figures for adults)

- Fever is invariably present, and diarrhoea occurs in 85%. This is bloody in 20% and can be confused with *Shigella* dysentery.
- Vomiting and abdominal pain are common (75%).
- Headaches, myalgia, or arthralgias are reported in 50%.
- Sore throat occurs in 50%, and is a distinguishing feature, as is conjunctival injection (45%).
- A maculopapular rash, although poorly visible on black African skin, is common.
- Cough occurs in 10% of cases.
- Bleeding is seen in 40% of cases, and is usually either gastrointestinal, oral, at injection sites, or as skin petechiae. This is a major diagnostic sign.
- Hospital mortality of 80%. Recovery starts two weeks into the illness.

Diagnosis of Ebola

- Clinical
  - **Suspected clinical case (during epidemic)**
    Any febrile illness associated with haemorrhage. No contact history required.
  - **Probable case (during epidemic)**
    A febrile illness occurring within three weeks of contact with a case of Ebola.

  or

    A febrile illness in which three or more of the clinical features above are present.
  - **Possible clinical case (non-epidemic)**
    An unexplained severe febrile illness, particularly with haemorrhage, in an area of Ebola transmission, with no response to an antimalarial plus a broad-spectrum antibiotic (for example chloramphenicol).
- Indirect laboratory tests supportive of diagnosis.
  Raised liver transaminases AST/SGOT.
  Low/normal initial white blood cell count.

- Confirmation of diagnosis
  Early serological tests were difficult to interpret, but newer specific IgM ELISAs may allow diagnosis of acute cases on a single positive test. However IgM is not always positive at presentation.
  - ✔ ***Specific IgG (by ELISA) rises too slowly to use as a test of acute infection, but may be useful in epidemiological surveys.***
  - ***Isolation of the virus is not appropriate outside a specialised laboratory.***
  - ***A postmortem skin biopsy (in formalin at room temperature) is not infectious, and can allow a diagnosis to be made using immunohistochemistry.***

Samples need to be marked as high infection risk ideally with standard yellow hazard tape, and sent in two sealed plastic bags. Samples should only be taken where laboratory staff are aware of the potential risks, and have the necessary precautions to handle such specimens safely. The lab should be informed that the specimen is coming.

Infection control

See below and Chapter 1.6.

Notification

Consider formal identification of a possible outbreak of Ebola if there is a new illness of high mortality in adults in a recognised area of transmission, particularly if hospital-acquired secondary cases have occurred.

Management

- Apart from supportive care, particularly with respect to adequate fluid and nutritional intake, there are no specific treatments which modify the course of illness.
- Antimalarial and antibiotic therapy should be given routinely, directed at treating possible alternative diagnoses (for example shigellosis or typhoid).

Infection control of VHFs

- At increased risk are laboratory staff, midwives, and those staff and family handling body fluids and excreta.
- High-risk patient groups are those with active haemorrhage, those confused and agitated, and pregnant mothers.

Barrier nursing

- Secondary spread is usually by contact with blood, urine infected secretions, used needles, or stool, but some viruses (for example Ebola) have also been found on patients' skin.
- There is little clinical evidence of respiratory aerosol spread for the VHFs, although virus may be present in the nose and oropharynx.
- Surgical and obstetric procedures carry a particularly high risk of infection to staff.
- ✔ **Transmission is substantially reduced by strict adherence to barrier nursing, disinfection of excreta and clear labelling of "at risk" specimens.**
- **Only essential samples should be taken.**
- The laboratory should be aware and prepared to receive specimens.
- Family contact should be restricted to the minimum required for care.
- Soap and water should be available for hand washing before and after patient contact.
- ✔ **For all carers, including family, careful barrier nursing with gloves and plastic aprons is mandatory, and stocks must be easily on hand.**
- Hospital staff and carers are advised to wear double gloves, plastic aprons, gowns (boots), head covering, HEPA-type face masks and goggles/eye shields. However in a tropical setting these can only be tolerated for a few hours at a time so arrange work to account for this.
- If outer gloves are not changed between patients, gloved hands should be washed in 1:100 bleach.
- Appropriate disposal of excreta and clinical waste is essential, so incinerate burnable clinical waste daily, and flush excreta down a dedicated toilet, having added 1:10 household bleach (0.5% chlorine) first.
  - Disinfect bedpans and urine bottles with 1:10 bleach.
  - Disinfect beds and equipment with 1:100 bleach.
  - Disinfect the dead with 1:10 bleach before burying in a sealed plastic bag.
- Consider using seropositive staff to nurse these patients. The identification and use of these has been successful in some outbreaks. They must follow the ward infection control measures.
- Remember convalescent patients may continue to excrete virus for many months (in both Lassa and Ebola).
- Fear in both staff and the community needs to be addressed openly, and staff and carers educated on the role of barrier nursing measures, and the risks involved if they are not followed.
- Careful attention where local culture and customs (for example burial rites, "widow cleansing", care of the sick, etc.) cause "high-risk" activity. Education and participation of community leaders is important to ensure safe practice.

Who should one isolate?

- On admission all patients likely to have a VHF.
- A **clinical diagnostic probability** on the basis of fever, contact history, haemorrhagic and non-haemorrhagic clinical signs, initial laboratory tests, and geography. These categories are:
  - suspected clinical VHF
  - probable VHF
  - illness probably not a VHF.

Distinguishing signs (for example conjunctivitis in Ebola) are particularly helpful in categorising cases.

- Isolation for suspected and probable cases on presentation to hospital.

- Isolation ideally in single rooms, but an identified, separate communal ward for probable and confirmed cases is often all that is available.
  - This should have an adjoining toilet, for safe waste disposal.
  - There should be a separate, adjoining area for changing into and storing isolation clothing.
  - Supplies of gloves and gowns, etc. need to be easily available.
  - Hand-washing facilities are mandatory.
- **Written infection control measures should be clearly visible on the ward**.
- The area should be marked as "access restricted" to only those trained in VHF isolation precautions, and attention should be paid to screening windows.

Differential diagnosis of VHFs

- The important differential diagnoses are, depending on geography, falciparum malaria, typhoid, meningococcaemia, *Shigella* or non-specific bloody dysentery, severe sepsis, leptospirosis, plague, yellow fever and dengue.
- It is crucial to exclude other treatable disease in patients presenting with symptoms suggestive of a VHF and to initiate therapy directed at these.
- All patients should therefore receive a broad-spectrum antibiotic (for example chloramphenicol), and in some areas, an antimalarial.
- In an endemic area, or during a known outbreak, the clinical diagnosis of a VHF is relatively easy. Difficulty arises where sporadic or new cases occur.
- A history of contact with a case in the previous three weeks and a history of recent travel to a transmission area should be sought. As no VHF has an incubation longer than three weeks, travellers or contacts of known or suspected cases who are well after this period are unlikely to be infected

Further reading

The following very practical resources are for those who require additional VHF control information.

Infection Control for Viral Haemorrhagic Fevers in the African Health Care Setting. WHO and CDC, 1998 (WHOP/EMC/ESR/98.2) and online at www.cdc.gov/ncidod/publicat.htm

WHO Recommended Guidelines for Epidemic Preparedness and Response: Ebola Haemorrhagic Fever (EHF). Geneva: WHO, 1997 (WHO/EMC/DIS/ 97.7), and online at www.who.int/emc/diseases/ebola/index.html

4.24 Yellow fever

George Wyatt

Minimum standards requirements

- Immunisation
- Blood clotting factors
- Intensive supportive care
- Vector control

Yellow fever is a flavivirus infection spread by the bite of *Aedes* mosquitoes.

Epidemiology

- Currently confined to tropical Africa and parts of South America especially around the Amazon basin; there is no yellow fever in Asia.
- Reservoir of infection exists in jungle primates and mosquitoes that bite them in tree canopy.
- Humans are infected when they visit the jungle and may take infection to urban areas. If vector mosquitoes, especially *Aedes egypti*, are allowed to breed in urban environments, and yellow fever is introduced, major epidemics of yellow fever may result.

Pathophysiology

Symptoms are due to toxic effects on the liver, kidneys and sometimes other organs such as the heart and brain.

Clinical features

- Incubation period 3–6 days.
- Many patients have an initial febrile illness – chills and muscle pains, from which they recover.
- Others, after an illness of about 5 days have a brief period of apparent improvement followed by deterioration and the following symptoms:
  - Vomiting – bilious, then black (coffee ground)
  - Jaundice
  - Bleeding – gums, nose, stomach
  - Proteinuria
  - Oliguria and renal failure
- Mortality is around 50%.

Laboratory diagnosis

- Leucopenia and heavy proteinuria.
- ***Paired acute and convalescent serum specimens show a fourfold or greater rise in antibody level and sometimes an IgM response; IgG antibody levels may be due to previous immunisation.***
- Postmortem liver biopsy specimens show mid zone necrosis of hepatic lobules often with eosinophilic Councilman bodies.
- ***Confirmation: isolation of virus from blood or liver specimens by inoculation of suckling mice or tissue culture cells.***
- ***Polymerase chain reaction may detect virus in blood or liver specimens.***

Management

- No specific antiviral treatment is available. Careful nursing and symptom control.
- Use antiemetics **and paracetamol** cautiously. **Avoid aspirin** in view of bleeding tendency.
- Rehydrate.
- ***Platelet transfusion and fresh frozen plasma if available*** for bleeding disorder.
- Nurse suspected patients under permethrin-treated bed nets as blood may remain infective for mosquitoes up to 5 days after onset.
- **Suspected cases of yellow fever must be notified** to national public health authorities who in turn notify WHO.

Prevention

- Elimination of breeding sites of *Aedes egypti* mosquitoes around dwellings.
- Immunisation of local population with live attenuated 17D yellow fever vaccine. Immunisation effective after 10 days.

4.25 African trypanosomiasis

George Wyatt

Minimum standards requirements

- Hydration and nutritional support
- Confirm the diagnosis
- Pentamidine/suramin/melarsoprol
- Prednisolone
- Public health measures and vector control

Varieties

Gambian trypanosomiasis caused by *Trypanosoma brucei gambiense*, is a slowly progressive disease of West and Central Africa. Rhodesian trypanosomiasis caused by *T. b. rhodesiense* is a subacute infection found in East and Southern Africa. Trypanosomiasis of wild and domestic animals is often caused by other "subspecies" of *T. brucei* which are indistinguishable morphologically from those causing human infection.

Transmission

- By the bite of infected tsetse flies (*Glossina*).
- Riverine tsetse (*Glossina palpalis* group) are responsible for transmission of *T. b. gambiense*, chiefly from a human reservoir. Infection may be endemic or epidemic.
- Woodland tsetse flies (*Glossina morsitans* group) are mainly responsible for sporadic transmission of *T. b. rhodesiense* from animals to humans.
- Congenital transmission is also well recognised.

Clinical features

A painful bite lesion (the trypanosomal chancre) may form at the site of the infected bite and last for up to three weeks but this is much commoner in *rhodesiense* than in *T. b. gambiense* infections.

Haemolymphatic stage 1

- Symptoms of fever and malaise lasting about a week are associated with waves of parasitaemia. Lymph nodes (especially those at the back of the neck Gambian disease) become enlarged.
- There may be short-lived oedematous swellings of face or limbs and sometimes a patchy circular erythematous rash or skin itching.
- Symptoms are often minor in Gambian disease, and this stage may last for months to years.
- In Rhodesian disease, patients are usually more ill with tachycardia, high fever, hepatosplenomegaly, myocarditis, anaemia and sometimes jaundice.

Meningo-encephalitic stage 2

- Severe headache and altered behaviour are often seen.
- Patients may become apathetic, depressed or frankly psychotic.
- Sleep is disturbed so that patients are often awake during the night and sleep by day; eventually deep coma results.
- Ataxia and cerebellar signs are frequent.
- Delayed response to pain after deep pressure, the appearance of primitive reflexes and altered tendon reflexes may be seen.
- Death often results from intercurrent infection.

Diagnosis

- In *T. b. rhodesiense* infections trypanosomes can usually be found in thick blood films; these are also useful for *T. b. gambiense* infections but may be negative during periods of low parasitaemia.
- More sensitive methods of examining the blood include microhaematocrit centrifugation, use of the QBC (Quantitative Buffy Coat) technique and the mini-anion exchange column method.
- When there are enlarged lymph nodes, microscopy of aspirates from these nodes often demonstrates trypanosomes.
- ***Serological methods including the CATT (card agglutination test for trypanosomiasis) for Gambian disease. Serological tests are usually sensitive but need to be confirmed by finding parasites.***
- Treatment depends upon evaluation of the stage of infection so that a lumbar puncture is essential. The most certain evidence for CNS invasion is by finding

trypanosomes in the cerebrospinal fluid but a raised lymphocyte count and/or raised protein content are also presumptive evidence of invasion.

Treatment

The drugs used for treatment are toxic. They should only be started after a parasitological diagnosis has been confirmed and after the patient's general condition has been improved by attention to hydration and nutrition and an antimalarial drug given. Pentamidine can harm the heart, liver, kidneys, bone marrow and pancreas.

Stage 1 Gambian disease

- Pentamidine isethionate 4 mg/kg IM or via IV infusion daily for 7–10 days.
- Children must lie down for an hour after an injection and have careful checks of pulse and blood pressure (**risk of severe hypotension**). Also check for hypoglycaemia and arrhythmias which can be side effects.
- Where possible, regular blood counts, blood glucose and electrolytes should be done.

Stage 1 Rhodesian disease

- Suramin: initial dose of 5 mg/kg slowly over 5 minutes IV day 1, 10 mg/kg IV day 3, 20 mg/kg IV day 5, then 20 mg/kg IV weekly for four further doses.
- Initial low dose is to reduce the risk of idiosyncratic anaphylactic reactions to suramin. Have IM epinephrine available (see Chapter 1.23).
- Test urine for albumin before each dose and modify regime if more than a trace of protein is seen.
  This regime may also be used for Gambian trypanosomiasis.

Stage 2 Gambian or Rhodesian disease

- Suramin IV on days 1 and 3 as above. Followed by:
- Melarsoprol IV

day 5	0.36 mg/kg
day 6	0.72 mg/kg
day 7	1.10 mg/kg
day 14	1.44 mg/kg
day 15	1.80 mg/kg
day 16	2.16 mg/kg
day 23	2.52 mg/kg
day 24	2.88 mg/kg
day 25	3.24 mg/kg
day 32	3.24 mg/kg
day 33	3.24 mg/kg
day 34	3.60 mg/kg

Give slowly with patient supine and fasting. ✔ Must remain supine and fasting for 5 hours after injection.

- Prednisolone 1 mg/kg orally should be started on day 2 and tapered rapidly after day 34; this is to reduce the risk of reactive encephalopathy which can otherwise result in the death of 5% of treated patients.
- Recent evidence suggests that equivalent results in Gambian disease can be achieved with IV melarsoprol 2.2 mg/kg daily for 10 days.
- Melarsoprol is a highly toxic drug which can cause skin necrosis on extravasation, liver, kidney or bone marrow damage or exfoliative dermatitis.
- ***Eflornithine given IV 100 mg/kg once every 6 hours for 14 days is an alternative treatment for stage 2 Gambian disease but it is so expensive that it is usually reserved for recurrent disease after melarsoprol treatment.***

Follow up

- Treated patients should be seen at three-monthly intervals for the first year and six-monthly intervals for the second year and repeat lumbar puncture should be performed at the first sign of meningoencephalitis to detect relapse early.
- Cases should be reported so that effective surveillance and public health action is taken.

4.26 Leishmaniasis

Brian Coulter

Minimum standards requirements

- Public health measures and vector control

Leishmaniasis: visceral

- Bone marrow/splenic/lymph node aspirate
- Pentavalent antimonials
- Aminosidine

Leishmaniasis: cutaneous and mucocutaneous

- Topical 15% aminosidine plus 12% methyl benzathonium
- Pentavalent antimonials
- Ketoconazole/itraconazole

- Leishmaniasis is caused by *Leishmania*, a protozoa whose reservoir is in animals, for example rodents, dogs and in some areas (for example India) in humans. The vector is the female sandfly.
- There are three main clinical types of disease:
  - cutaneous (CL)
  - mucocutaneous (MCL)
  - visceral leishmaniasis (VL) or kala-azar

Parasite and life cycle

- About 21 of the 30 or more species of *Leishmania* infect humans. They are morphologically similar and can only be differentiated by isoenzyme analysis which identifies the zymodeme in the cultured parasite.
- In animals (and humans) *Leishmania* live in macrophages in the reticuloendothelial system in the form of amastigotes (Leishman–Donovan bodies). When taken up by the biting sandfly it transforms into a promastigote which has a flagellum.
- There are two main genera of sandfly responsible for transmission, *Phlebotomus* in the Old World and *Lutzomyia* in the New World (Central and South America). Sandflies breed in organic material in dark moist sites, such as cracks in masonry, termite hills or leaves on the forest floor. The female obtains its blood meal at night by feeding on animals and humans if living or working in the vicinity.

Epidemiology

- Old World CL and VL are found in the Mediterranean basin, Middle East, Ethiopia, Kenya, the Sudan, India, Bangladesh and southern regions of the former Soviet Union and China. Where AIDS and VL coexist, for example Spain, there are major problems in treatment of VL. Drug resistance in VL is a problem in India and the Sudan.
- In the New World CL and MCL are the main forms of infection, VL occurs mainly in north-east Brazil.

Immunology

- A strong cell-mediated immune (CMI) response is required for control and recovery from disease. Polyclonal stimulation of B cells results in high levels of IgG.
- Subclinical infection is common. CL usually heals spontaneously but untreated MCL will progress and VL will result in death. Development of VL indicates that the host's CMI is unable to control the infection, and if untreated progressive immunosuppression will develop.
- Death is usually due to a secondary infection, for example respiratory tract or gut.

Cutaneous and mucocutaneous leishmaniasis

Cutaneous leishmaniasis

L. tropica, *L. major*, *L. aethiopica* in Old World; *L. mexicana* and *L. amazonensis* in New World. Single or multiple nodules develop on exposed areas especially the face or

extremities and usually ulcerate. Most heal spontaneously within months to a year or so.

Mucutaneous leishmaniasis

L. braziliensis.
Starts as a nodule, as in CL, but at about the time of healing metastatic lesions occur on mucosal surfaces, such as nasal mucosa or oropharynx. Untreated, progressive destruction of local tissue occurs.

Diagnosis

- Slit skin smear or aspiration should be undertaken from the raised margin of the lesion (not the base of the ulcer). Material is spread on a slide, dried, fixed in methanol and stained with Giemsa or Leishman.
- If a biopsy is undertaken (for example in MCL) impression smears should be done before fixing.
- If available the specimen should be cultured.

Management

- Most CL are self-limiting. Specific treatment is indicated for multiple, large and disfiguring lesions and all MCL.
- Clean lesion, give antibiotics if necessary.
- Apply topical ointment containing 15% aminosidine and 12% methylbenzathonium chloride twice daily for 10–20 days.
- If unsuccessful give a pentavalent antimonial as for VL (see below) for 3 weeks; a prolonged course (6–8 weeks) may be required for MCL.
- Alternative, less effective, systemic treatments include 4 weeks of ketoconazole (3 mg/kg once daily – **danger of liver dysfunction**) or itraconazole (older child) (3–5 mg/kg once daily).
- Intralesion injection (four per ulcer) of pentavalent antimonial, alternate days for 2–3 doses for adolescents/adults, if tolerated, is effective. Approximately 1 ml is injected into the edge and the base of the lesion (four sites per ulcer) using a 1 ml syringe and fine (24 gauge) needle.

Visceral leishmaniasis

L. donovani and *L. infantum* in Old World; *L. chagasi* in New World.

- Major presenting features include the trio of prolonged fever, anaemia and moderate to marked splenomegaly. In early stages the child is often only mildly unwell and may have a reasonable appetite. In a minority of cases, the onset may be acute with a high temperature, toxaemia and mild splenomegaly.
- Pancytopenia is the main laboratory finding.

Clinical features of visceral leishmaniasis

Incubation period:	2–4 months (weeks to 2 years)
Fever:	Intermittent at first
Anaemia:	Marrow depression, hypersplenism
Splenomegaly:	Progressive enlargement
Hepatomegaly	
Weight loss	
Epistaxis:	Haemorrhage from other sites may occur in advanced disease
Diarrhoea:	Invasion of gut by amastigotes, secondary infection
Cough	
Oedema:	Hypoalbuminaemia. Hair and skin signs of malnutrition in chronic forms
Lymphadenopathy:	In some African countries

Clinical pathology of visceral leishmaniasis

Haemoglobin: low; normochromic, normocytic film
White blood cells: low, $2–3\times10^9$/litre. Eosinophils low
Platelets: low, $<100\times10^9$/litre
Reticulocytes: low
Serum albumin: low
Serum globulin: elevated
Liver transaminases and serum bilirubin: normal

Differential diagnosis

- Differential diagnosis of marked hepatosplenomegaly, anaemia and pancytopenia includes hyperreactive splenomegaly (tropical splenomegaly) syndrome and schistosomiasis; also myeloid leukaemia and myelofibrosis.
- In acute onset disease, malaria, disseminated tuberculosis, typhoid, brucellosis, African trypanosomiasis, relapsing fever and leukaemia should be considered.
- HIV infection or co-infection.

Diagnosis

- In children, diagnosis is usually confirmed by demonstrating amastigotes on bone marrow aspirate.
- Splenic aspirates have a higher sensitivity and procedure is safe in skilled hands as long as platelet count is above 40×10^9/litre and coagulation is normal.
- Repeat marrow or splenic aspiration to monitor progress if required.
- If there is lymphadenopathy diagnosis may be attempted by fine-needle aspiration.
- ***Serological antibody tests such as ELISA have a high sensitivity and are particularly helpful if a parasitological diagnosis cannot be achieved.***
- When a microscopic diagnosis cannot be made ***the polymerase chain reaction (PCR) should***

be undertaken. The value of the PCR is being evaluated.

Management

- If appropriate, consider HIV infection.
  Secondary disorders include malaria, respiratory and gut infections, and tuberculosis.
- Blood transfusion for anaemia is seldom required as child has usually adapted to low haemoglobin.
- Give haematinics and vitamin supplements during nutritional rehabilitation and convalescence.
- ***Liposomal amphotericin B (expensive) is the treatment of choice in rich countries. 3 mg/kg is given IV (over 30–60 minutes) once daily for 10 days. An initial test dose of 100 micrograms/kg (maximum 1 mg) is infused over 15 minutes. Observe for 1 hour to ensure anaphylaxis does not occur then proceed. A shorter course may be just as effective viz 3 mg/day for 5 days then 3 mg/kg on day 14 and 21 (21 mg/kg in total).*** Total doses of 18–30 mg/kg (up to 40 mg/kg in HIV-infected children) are required depending on resistance patterns in the area. Trials of variation in total dose and intervals of injections are in progress. May induce hypokalaemia.
- Standard treatment is with pentavalent antimonials, i.e. **sodium stibogluconate** (Pentostam 100 mg Sb/ml) or **meglumine antimoniate** (Glucantime, 85 mg/Sb/ml); 20 mg Sb/kg/day is given IV or IM (painful) in a single dose. If venous access is available, 10 mg/Sb/day 12 hourly may be more effective; each injection is given over 5 minutes and is stopped if coughing or substernal pain occurs, and allows a shorter course (21 days). Urinary excretion of antimony is rapid (half-life 2 hours) although slow accumulation occurs.
- Duration of antimony treatment is usually 4 weeks but prolonged treatment (up to 6 weeks) may be necessary in resistant areas.
- Serious toxicity is rare in children, but if a prolonged course or high dosage is required, or toxicity is suspected, liver function tests and an ECG looking for conduction disorders should be done (serious toxicity may require dimercaprol).
- Resistance to antimonials is increasing especially in India and alternative drugs have to be considered (see below).

Paromomycin (aminosidine)
Paromycin (an aminoglucoside) is an alternative less toxic agent and may if necessary be given with antimonials. Give IV or IM in a dose of 16–20 mg/kg once daily for 3–4 weeks.

Follow up and prognosis

Symptomatic improvement occurs usually within a few days and a haematological response within two weeks. Splenomegaly slowly regresses but may take a year or more to resolve. Prolonged follow up (at least a year) is necessary to detect relapse. Relapse is treated with a repeat, prolonged course of antimonials (up to 8 weeks). Unresponsiveness will require alternative drugs such as liposomal amphotericin B (if available), aminosidine, standard amphotericin B or pentamidine. Trials of miltefosine are in progress.

Prevention and control

Prevention is similar to malaria and includes insect repellents and use of fine mesh bed nets impregnated with permethrin. Control includes spraying of sandfly resting sites and houses, destruction of animal reservoirs and treatment of cases.

4.27 Malaria

Elizabeth Molyneux

Minimum standards requirements

- Preventive measures (impregnated bed nets and antimalarial drugs)
- Blood smear, measure blood glucose and Hb
- Antimalarials (follow local guidelines): quinine, pyrimethamine/sulfadoxine, amiodoquine, metakelfin, halfantrine, chloroquine, artemether, mefloquine
- Anticonvulsants (see Chapter 3.38)
- Intravenous glucose and hydration
- Blood transfusion (+ furosemide)
- Supportive intensive care
- Vector control

Life cycle

The infected *Anopheles* female mosquito injects sporozoites into the blood stream of an individual. Sporozoites circulate for about 30 minutes before being phagocytosed or entering liver parenchymal cells. The liver phase prior to re-entry in the circulation is called the pre-erythrocytic phase and varies depending on the species. At the end of this phase merozoites invade the red blood cells and begin the erythrocytic phase.

In two species (*Plasmodium vivax* and *Plasmodium ovale*) some hepatic stage parasites remain within the liver cells with formation of the dormant phase called hypnozoites. For various reasons (perhaps waning immunity) at a later date the dormant phases activate and reseed blood. This leads to manifestations of malaria not from a new infection but from the latent exo-erythrocytic phase. *P. falciparum* differs from the other species in that infected erythrocytes adhere to capillary epithelium, thus disappearing from the circulation and evading destruction by the spleen. This process may account for the fact that only *P. falciparum* causes life-threatening disease and is a major cause of mortality in children.

Clinical features

- Typical features include high-grade fever alternating with cold spells, rigors, chills and sweating. There are usually associated myalgias and arthralgias.
- However, features in children under 5 years may be non-specific with fever, vomiting, diarrhoea, and abdominal pain being the main symptoms.
- In older immune individuals the only symptoms may be fever with headache and joint pains.
- All fevers in children from a malaria endemic area are therefore due to malaria until proven otherwise.

Diagnosis

- Blood smear for malaria: thick slide for diagnosis, thin slide to confirm type of malarial parasite. Typically ring forms inside red blood cells are seen but there may also be gametocytes.
- Level of parasitaemia usually scored as 1–4+. (If the malarial smear is $\geqslant 3$ there is a high level parasitaemia.) In areas where parasitic density is measured the smear is reported as parasites/mm^3.

Uncomplicated malaria

Child has fever and a positive blood smear. There is no evidence of altered consciousness, hypoglycaemia, severe anaemia, jaundice or respiratory difficulties.

Management

Management for a child who has always lived in an endemic area

- No need to admit to hospital.
- Give first-line antimalarial treatment as recommended in local national guidelines.
- Ensure tablets or syrup are swallowed and not vomited.
- Encourage oral fluids.
- Measures to lower temperature may be necessary.
- If pale (probably anaemic) give haematinics (iron, but not folic acid if sulfadoxine-pyrimethamine has been used for malaria treatment).

Management of a child visiting an endemic area for the first time

Hospital admission for management of *P. falciparum* is always advisable.

Advice
Discuss preventive efforts with carers, for example bed net at night, ideally impregnated with insecticide.

Tell mother to return after two days if fever persists; earlier if child deteriorates.

Severe malaria

- Child is febrile and has a positive blood smear.
- There may also be vomiting, diarrhoea or a cough.
- Conscious state may be altered; there may be a history of convulsions.
- There may be evidence of hypoglycaemia and acidosis or severe anaemia, jaundice or generalised weakness (unable to sit up).

As temperature in malaria may fluctuate, a single reading may be normal.

Cerebral malaria

A significant majority of children with *P. falciparum* proceed to develop altered consciousness, severe anaemia, acidosis, or any combination of these. Where transmission of *P. falciparum* is endemic, malaria is the commonest cause of coma in children, especially in age range 1–5 years.

Coma develops rapidly, often within one or two days of onset of fever, sometimes within hours. Convulsions are usual and may be repeated. Clinical features suggest a metabolic encephalopathy, with raised intracranial pressure. Opisthotonos, decorticate or decerebrate posturing, hypotonia and conjugate eye movements are common. Oculovestibular reflexes and pupillary responses are usually intact. Papilloedema is found in a small minority of cases. A unique retinopathy with patchy retinal whitening and pallor of vessels has been described. In fatal cases brain swelling is commonly present at autopsy, but cerebral herniation is not usually found even in patients who had undergone lumbar puncture.

Hypoglycaemia, acidosis, hyperpyrexia and convulsions (sometimes undetectable without EEG) are common accompaniments of cerebral malaria, and require appropriate management.

No physical signs are diagnostic of coma due to malaria, and incidental parasitaemia is common in endemic areas, so other causes of coma must always be carefully sought, and if necessary treated on the basis of presumptive diagnosis.

Even with optimal treatment, the case fatality rate is 15–30%, and about 10% of survivors have residual neurological sequelae (hemiparesis, spasticity, cerebellar ataxia) that may partially or completely resolve over time.

Investigations

- Thick and thin films for malarial parasites.
- Blood glucose (for example by BM stix).
- Lumbar puncture if meningitis suspected – contraindications include: Glasgow Coma Score <8, papilloedema or suspicion of raised intracranial pressure including a tense fontanelle in infants, or respiratory difficulty. In such a situation, give intravenous antibiotics to treat meningitis.

Management

- Treat convulsions (see Chapter 3.36 on coma and Chapter 3.38 on convulsions).
- Treat hypoglycaemia.
- Treat hypovolaemic shock (if present) (see Chapter 3.6).
- Initiate antimalarial therapy.
- Treat severe anaemia.

Hypoglycaemia
Hypoglycaemia is defined as blood glucose <2.5 mmol/litre in well-nourished children; <3.5 mmol/litre in malnourished children.

Hypoglycaemia is common and is due to poor intake, increased metabolic needs of the patient and parasites and impaired hepatic gluconeogenesis.

- Prevent hypoglycaemia with a 10% glucose infusion IV (add 10 ml 50% glucose to 90 ml of 5% glucose solution).
- Treat hypoglycaemia with 5 ml/kg of 10% glucose solution IV. Recheck blood glucose after 30 minutes and repeat glucose bolus if blood glucose is still low.

  If child still unable to swallow after 48 hours, start nasogastric feeds.

  When a gag reflex is present and the child is able to swallow, introduce oral fluids.

Severe anaemia
Give blood if haemoglobin <5 g/dl or haematocrit <15% or evidence of cardiac failure or if haemoglobin >5 g/dl (haematocrit >15%) but very heavy parasitaemia and falling haemoglobin.

Give packed cells 10 ml/kg or fresh whole blood 20 ml/kg over 3–4 hours. If severely malnourished, circulatory overload is more likely and give packed cells if possible; if not give IV furosemide (1–2 mg/kg) with 10 ml/kg of whole blood.

Diuretics are not normally needed unless there is evidence of fluid overload.

Supportive care
- Nurse in recovery position and turn 2 hourly. Do not allow child to lie in a wet bed and provide special care to pressure points.
- Check blood glucose 4–6 hourly and haemoglobin/haematocrit daily.
- Watch urine output – aim at 1 ml/kg/hour. If despite rehydration urine output is <4 ml/kg/24 hours give IV furosemide 2 mg/kg. If no response double dose at hourly intervals to a maximum of 8 mg/kg.
- Monitor coma score 4 hourly.
- Treat convulsions, hypoglycaemia, hyperpyrexia (>39°C)
- Shock is unusual in malaria. If present consider septicaemia, do a blood culture and start a broad-spectrum antibiotic IV (penicillin and chloramphenicol **or** cefotaxime or ceftriaxone) in addition to antimalarials.

- As stated above in a comatosed child in whom you cannot exclude meningitis, give appropriate antibiotic intravenously.
- If there is deep or laboured breathing suggestive of acidosis, give extra intravenous fluid to correct hypovolaemia.
- During rehydration, examine frequently for fluid overload (increased liver, gallop rhythm, fine crackles at lung bases, raised jugular venous pressure).
- In infants always use an in-line infusion chamber for intravenous rehydration. If this is not available and supervision is poor, consider nasogastric rehydration.

Moderate anaemia

- If anaemia associated with malaria is not severe (defined as haemoglobin 6–9.3 g/dl) treat as follows: give iron once daily for 14 days in combination with folic acid (1 tablet contains ferrous sulphate 200 mg, equivalent to 65 mg elemental iron plus 250 micrograms folic acid). Give half tablet to child 10–20 kg and one tablet daily to child >20 kg.
- If child is taking sulfadoxine-pyrimethamine for malaria, do not give folic acid until two weeks later (it interferes with antimalarial action).
- An alternative is iron syrup (ferrous fumarate) 140 mg in 5 ml and equivalent to 45 mg of iron. Give once daily as follows:

Weight	Dose
3–6 kg	1 ml
6–10 kg	1.25 ml
10–15 kg	2.0 ml
15–20 kg	2.5 ml
20–30 kg	4 ml

Plus separate folic acid 250 micrograms/kg/day

Treat for three months where possible (one month to correct anaemia and one to three months to build iron stores).

Antimalarials

- If a blood smear is not immediately available and there is no other obvious cause of illness treat as malaria.
- In Africa and many other regions quinine is the drug of choice for severe malaria. In South East Asia and the Amazon basin quinine is no longer always effective.
- Initially give treatment intravenously, if possible; if not, intramuscularly.
- Change to oral therapy as soon as possible.

Severe malaria

First-line antimalarial: intravenous quinine

- Give 20 mg/kg of quinine salt (maximum 1.4 gm) in 5% glucose in a concentration of 1 mg of quinine to 1 ml of 5% glucose over 4–6 hours. Use an in-line infusion chamber (100–150 ml) to ensure that the loading dose does not go in too quickly. There is a major risk of cardiac side effects if this happens. If safe control over the rate of infusion of intravenous quinine is not possible, give it intramuscularly (with initial doses of 10 mg/kg 1M at 0 and 4 hours).
- Then 10 mg/kg in 10 ml/kg fluid every 12 hours for 24 hours or longer if child remains unconscious. These latter doses can be given over 2 hours.
- Never give bolus infusion.
- As soon as child is able to take orally, switch to quinine tablets 10 mg/kg every 8 hours for a total of 7 days.
- For intramuscular injections, dilute the quinine solution for better absorption and less pain.

Side effects

- Common: cinchonism (tinnitus, hearing loss, nausea and vomiting, uneasiness, restlessness, dizziness, blurring of vision).
- Uncommon: hypoglycaemia, although this is a common complication of severe malaria.
- Serious cardiovascular problems (QT prolongation) and neurological toxicity are rare.
- If overdosed by mistake with quinine tablets give activated charcoal orally or by nasogastric tube as a suspension in water (1 g/kg).

Second-line antimalarials

Second-line drugs include pyrimethamine with sulfadoxine (fansidar), amiodaquine, metakelfin and halofantrine. Artemether and mefloquine are currently designated as reserve drugs for multidrug-resistant malaria. However, this varies considerably from country to country. For example, Fansidar has become first-line treatment where chloroquine resistance is high.

Always check local guidelines on drug sensitivities.

- **Sodium artesunate IV**
  2.4 mg/kg as a bolus
  then 1.2 mg/kg after 12 hours and 24 hours
  then 1.2 mg/kg every 24 hours for 6 days (give same dose orally as soon as possible)
- **Artemether IM**
  3.2 mg/kg immediately, then 1.6 mg/kg IM once daily for 4 days
- **Quinidine gluconate IV**
  15 mg/kg IV loading dose over 4 hours.
  After 12 hours infuse 7.5 mg/kg over 4 hours, then repeat 8 hourly. Change to oral quinine as above as soon as possible.
  Quinidine is more cardiotoxic than quinine. ✔
- **Chloroquine IV**
  No longer dependably effective in most endemic areas. Avoid, as resistance in children is relatively high.
  5 mg base/kg every 6 hours for a total of 25 mg base/kg (five doses) as infusion in 5% glucose (give over 2–4 hours).

Change to an oral therapy when the child can tolerate it.

Side effects

Nausea, vomiting, headache, uneasiness, restlessness, blurred vision, hypotension and pruritus.

Poisoning: coma, convulsions, dysrhythmias and hypotension.

Uncomplicated malaria

Fever and a positive blood smear but none of the following:
- Altered consciousness
- Severe anaemia (haemoglobin <6 g/dl)
- Hypoglycaemia (blood glucose <2.5 mmol/litre)
- Respiratory distress
- Jaundice

If blood smear is not possible, and fever occurs in an endemic region, treat as for malaria. Drug treatment depends on resistance patterns. The following drugs are used for children:

- **Chloroquine tablets (150 mg base)**
  Still the first-line treatment in many African countries because it is inexpensive and partially effective.

Day 1	10 mg/kg/day
Day 2	10 mg/kg/day
Day 3	5 mg/kg/day

- **Chloroquine tablets (if weight not available)**
  Check local formulary with tablets as chloroquine comes in different strengths

Age	1–4 years	1/4 adult dose
Age	4–8 years	1/2 adult dose
Age	8–12 years	3/4 adult dose

- **Chloroquine injection (if child is vomiting)**

Day 1	5 mg/kg/dose then repeat after 6 hours (SC or IM max. 200 mg/dose)
Day 2	repeat day 1 injection dose or use tablets in dose above
Day 3	5 mg/kg/dose or tablets at same dose

- Fansidar (not if allergic to sulphanomides) (each tablet contains pyrimethamine 25 mg plus sulfadoxine 500 mg)

Age	Dose (single dose given daily for 7 days)
3–6 months	1/4 tablet
6 months–1 year	1/2 tablet
1–3 years	1 tablet
4–6 years	1.5 tablets
6–12 years	2 tablets
>12 years	3 tablets

  NOT IF <3 months of age

Primaquine (tablets 7.5 mg base)
250 micrograms/kg once daily for 14 days or 21 days for infections in SE Asia or W Pacific. For children with G6PD deficiency 750 micrograms/kg once **weekly** for 8 weeks. **After food**.

- **Quinine (usually reserved for severe malaria)**
  Oral therapy for uncomplicated malaria is 10 mg/kg/dose every 8 hours for 7 days. Shorter regimens, i.e. 3–5 days should be combined with fansidar or metakelfin.
- **Other drugs for *P. falciparum* resistant to other antimalarial drugs (not less than 1 year of age)**
  - **Mefloquine (tablet 250 mg)**

  Dose: 25 mg/kg as a single dose, max. 1 g (4 tablets) Must not be combined with quinine or related drugs such as chloroquine or halofantrine.
  - **Halofantrine** (250 mg tablets, 100 mg/5 ml suspension)

  Dose: 8 mg/kg every 6 hours for three doses.

Infection with *P. vivax* and *P. ovale*

Both of these malaria strains can relapse. This can usually be prevented by giving a course of primaquine 250 micrograms base/kg daily for 10–14 days (tablets contain 15 mg base per tablet).

Prophylaxis

Most important is the prevention of mosquito bites. Drugs for prophylaxis depend on the region and sensitivity of the malarial parasite.

IMPORTANT FOR:
- Children with sickle cell disease – chloroquine 5 mg/kg weekly.
- Pregnant mothers in an endemic area.
- Child or adult who returns to an endemic area after an absence of over one year even if he/she is originally from that region.
- Non-immune individuals: people from non-endemic areas.

4.28 Helminth infections: “worms”

Ed Cooper

Minimum standards requirements

- Faecal microscopy and egg count
- Anoscopy
- Eosinophil count and chest *X* ray
- Mebendazole, albendazole, ivermectin
- Topical thiabendazole

Introduction

These parasites cause the most prevalent infections on earth of humans and of other animals. Although it is never desirable for a child to have worms, most children seen at the hospital have only a few worms in their intestine, or a few juvenile forms (larvae) in other organs, and these are tolerated without identifiable symptoms. However, actual, measurable ill health or the risk of serious complications are directly related to the number of parasites in a child. Although the children bearing these heavy loads of parasites are in a minority, because the prevalences are so great the numbers of heavily infected children are also great. About 2000 million children and young people have worms, so if we cut off only the most heavily infected 5% and suggest they are definitely made significantly unwell by their parasites, that would still amount to 100 million young people. These will commonly be seen in hospitals and clinics, often presenting for other reasons, without their heavy infections being recognised.

Diagnosis

The main clues to heavy parasitosis are in growth and nutrition. Gastrointestinal symptoms also occur. Symptoms and signs may be suggestive of worm infections but only one or two are pathognomonic. In addition, there have been an increasing number of studies on the effects of helminth infections on cognitive function and general physical fitness: these have added to the case for community control of these infections as an important public health measure. However, these effects do not generally give rise to symptoms which present in hospitalised patients.

Table 4.28.1 Diagnosis of helminth infections due to the presence of adult worms

Symptom/sign	Suggestive of this species of adult worm in intestine
Short stature, not growing	*Trichuris* or hookworm
Mild/moderate muscle wasting	*Trichuris* or hookworm
Anaemia, microcytic hypochromic	Hookworm or severe trichuriasis; not *Ascaris*
Hypoproteinaemia, possible oedema	Hookworm or severe trichuriasis or disseminated strongyloidiasis; not *Ascaris*
Pica, especially eating soil (geophagia)	Any or all helminths
Colicky abdominal pain	*Ascaris:* common but a weak correlation
Intestinal obstruction	*Ascaris:* quite common surgical emergency
Jaundice and/or pancreatitis	*Ascaris:* uncommon
Laryngeal obstruction	*Ascaris:* rare
Vomiting up worms	*Ascaris:* common
Chronic diarrhoea	*Trichuris* or severe hookworm or strongyloidiasis
Defaecating during sleeping hours	*Trichuris*
Blood and mucus in stool	*Trichuris*
Rectal prolapse	*Trichuris*
Finger clubbing	Intense trichuriasis or hookworm; not *Ascaris*
Perianal itching	*Enterobius*
Vulvovaginitis	*Enterobius*

Investigations

Investigation for adult worm in intestine

Except for *Enterobius*, this depends on the examination of stool. Full laboratory details are beyond the scope of this manual, but for *Ascaris*, hookworm and *Trichuris* examination by the Kato (modified Kato or Kato–Katz) method is recommended. This requires only microscope slides, a standard hole in a flat spatula so that a 50 mg stool sample is squashed on to the slide, cellophane, glycerol and a stain such as malachite green. The microscopist's count of eggs per gram of stool gives an indication of the intensity of infection. This should be used to guide the clinician in giving weight to the likelihood of helminth infection as a major cause of the symptoms listed in Table 4.28.1. Other causes will also be in the picture with most of the symptoms because of their lack of specificity, given that other infections and other nutritional conditions are present in the same locality.

Enterobius (pinworm) eggs are only occasionally seen in stool because they adhere to perianal skin where the female worm has deposited them. They can be picked up on sticky tape and transferred to a glass slide. Specific diagnosis of *Enterobius* is quite unrewarding anyway as it is reasonable to treat the family when it is suspected without proving the presence of the worm (see below).

The most effective way to establish that *Trichuris* infection is intense is to see the worms on prolapsed rectal mucosa or to do anoscopy. An otoscope can be used for anoscopy in young children. This is because the worms are usually confined to the caecum, so if they have reached the lower rectal mucosa the infection must be intense.

Strongyloides is a rare cause of illness in young children although it becomes more significant in adolescence in some regions. Microscopy has a low sensitivity and the stool requires culture by special techniques. Serology is not widely available and also lacks specificity.

Table 4.28.2 Illness due to larvae rather than adult worms

Symptom/sign	Suggestive of this species of larvae in viscera*
Cough and wheeze	*Toxocara canis/cati* (dog/cat roundworm) and also *Ascaris* and hookworm
Hepatomegaly	*Toxocara*
Lymphadenopathy	*Toxocara*
Leucocytosis with extreme eosinophilia	*Toxocara*
Epilepsy/encephalopathy	*Toxocara* (rare)
Uveitis or proliferative retinitis	*Toxocara* (younger children escape in endemic areas: naive strangers are more susceptible)

* Larvae in/under skin (itchy area with red wiggly line, moving from day to day, often with pyoderma) suggests *Ancylostoma braziliensis* (dog hookworm).

Investigation for migrating larvae

Eosinophilia is characteristic. Sometimes 20–50% of the leucocytes are eosinophils. By contrast, eosinophilia is not a constant feature of established infection with adult worms and so is a useless diagnostic marker for intestinal infection.

The chest *X* ray may show a flaring shadow spreading out from the hila.

Serology is diagnostically useful in visceral larva migrans (**Toxocara *infection***) but is only ***done in special centres or research laboratories***.

Diagnosis of cutaneous larva migrans (dog hookworm infection picked up from skin-ground contact) is purely clinical. The key is to think of it when looking at a patch of itchy pyoderma – the red line has often disappeared under the scratching.

It is not clear how much of the total burden of coughing, wheezing and dyspnoea in a child population in an endemic zone is due to the pulmonary migration of helminth larvae. Factors which make the symptoms more severe are migration of children naive to *Ascaris* or hookworm infection into the endemic area, and zoonotic larvae (*Toxocara*) which cannot complete their migration but die in their human hosts.

Treatment

The broad-spectrum anthelmintics mebendazole and albendazole are drugs which combine great efficacy with an almost complete absence of side effects in ordinary use. They are the drugs of choice for ascariasis, hookworm infection, trichuriasis and enterobiaisis. Albendazole is as effective as thiabendazole for visceral larva migrans and with less side effects. However, visceral larva migrans is a self-limiting condition and symptoms and signs resolve in three months. Thiabendazole is still useful for cutaneous larva migrans in a topical preparation. Ivermectin is recommended for strongyloidiasis, but albendazole remains useful and is preferable to thiabendazole because it is less toxic.

Mebendazole

This is most commonly available as 100 mg tablets, but is also produced as a 20 mg/5 ml liquid and a 500 mg tablet. The tablets are chewable and pleasant enough to take. The 500 mg tablet is useful for mass campaigns against *Trichuris* or hookworm. Standard treatment for *Trichuris* infection or symptomatic hookworm infection is 100 mg twice daily for three days. It is not approved for use in children under 2 years, but clinical judgement should be used in a symptomatic child. It is considered unsafe in pregnancy or lactation.

Albendazole

This drug is closely related to mebendazole with similar pharmokinetics. It has superior efficacy to mebendazole in systemically invasive conditions: it is more effective against

migrating larvae. It is available as 200 mg tablets or 200 mg/5 ml liquid. Standard treatment for *Trichuris* infection is 400 mg daily for three days. Cautions are as for mebendazole, noting its greater systemic absorption.

Regimens for specific infections

- *Ascaris*: mebendazole 100 mg twice daily for 3 days or single dose of albendazole (400 mg).
- Hookworm: for reliable expulsion use as for trichuriasis, (above). For useful effect in a mass campaign, a single dose can be given.
- *Trichuris*: see above.
- *Enterobius*: single dose of mebendazole or albendazole. However, repeated doses are necessary (for example, once weekly for three weeks) and it is also advisable to treat all young children in a household as spread is person to person.
- Cutaneous larva migrans: thiabendazole tablets can be crushed and mixed with hydrocortisone ointment. This is effective topically.
- Strongyloidiasis and rarer infections such as capillariasis: more prolonged treatment with higher dosage is necessary.

In places or situations where only older drugs are available

Details and dosages are not given here: manufacturers' recommendations may be followed but these drugs are inferior to mebendazole and albendazole and should be replaced if possible.

- Piperazine is effective against *Ascaris* and *Enterobius*. No action on *Trichuris* or hookworm, and toxic in children prone to epileptic seizures.
- Levamisole is effective against *Ascaris* and is useful enough in hookworm infection (especially *Necator americanus*) to be used in mass control programmes.
- Thiabendazole has limited effectiveness in trichuriasis and is useful in strongyloidiasis, toxocariasis and cutaneous larva migrans.
- Pyrantel is effective against *Ascaris* and *Enterobius* with some action against *Necator americanus* and less against *Ancylostoma duodenale*. Only if combined with oxantel does the preparation affect *Trichuris*.

Parasitology of “worms”

- *Ascaris lumbricoides* is the common human roundworm. Adults are up to 30 cm long and live for years in the small intestine. They are often seen in stool or vomit. Transmission is only by ingestion of embryonated eggs in soil and the larvae migrate through the portal vein, liver and lungs before being reswallowed and developing into adults which reproduce sexually and shed eggs in faeces. The ill effects of ascariasis are largely accidental, for example a worm in the common bile duct, but the probability of this accident is a function of the worm load.
- *Trichuris trichiura* is the whipworm, up to 4 cm long and of the thickness of a hair except at the tail end. It is a geohelminth like *Ascaris* and transmission occurs in similar conditions. Maturation occurs only in the gut without tissue invasion beyond the mucosa. Its ill effects are a direct, predictable function of the worm load and amount to a form of colitis with systemic secondary effects of chronic inflammation. This intensity of infection occurs only in a small minority of children.
- Hookworms are of two species, *Necator americanus*, the New World hookworm, and *Ancylostoma duodenale*. Both adult forms are hair-like, about 1 cm long, with cutting plates at the mouth end. *Ancylostoma* is generally the more virulent pathogen, but both species occur widely with overlap and the differences between them can be ignored by clinicians without a special interest in this subject. They have invasive larvae and both skin penetration and ingestion of embyonated eggs are involved in transmission. Sustained blood loss in the small intestine leads to an accumulating risk of anaemia, but in children protein-losing enteropathy and systemic secondary effects of chronic inflammation are equally important.
- *Enterobius vermicularis* is intestinal but not a geohelminth. There is no larval invasion. It is spread as embryonated eggs by personal contact among children, for example under fingernails, but also in dust. It is largely harmless but can cause secondary infection in the vaginal introitus.
- *Strongyloides stercoralis* is capable of independent existence in the soil. It is an opportunistic parasite of several mammals including man. Person-to-person spread occurs with probability related to intimacy of contact. Although it is acquired in childhood its most devastating (often fatal) effects occur only when filariform larvae become disseminated through asexual reproduction in the host. This happens if host immunity breaks down, for example with severe malnutrition or malignancy in later life. Surprisingly, disseminated strongyloidiasis is not associated with HIV infection, although it is associated with another retrovirus, HTLV1.

4.29 Hydatid disease

George Wyatt

Minimum standards requirements

- Ultrasound and chest *X* ray
- Eosinophil count
- Albendazole
- Surgical excision with scolicides

Echinococcus granulosus eggs are transmitted from dogs and occasionally other carnivores (foxes and wolves) by finger or contaminated water or food. Sheep are intermediate hosts and so do not have eggs in their faeces. Because of the slow rate of growth of hydatid cysts, symptoms from infection in childhood often present in adult life. Many cysts remain asymptomatic, eventually calcify and become sterile.

Epidemiology

Widespread in sheep-rearing countries and wherever there is intimate contact between humans and dogs or other canids and where dogs scavenge dead animals or uncooked offal. High incidence in Turkana area of Kenya.

Clinical features

- Cysts may occur in virtually any organ.
- Many cysts are asymptomatic. The liver will be palpable due to a symptomatic cyst of a fair size.
- Rupture may cause anaphylactic reactions and spread of infection by daughter scolices (heads of immature worms).
- Abdomen
  - Liver (60% of all cysts), also intra-abdominal cysts.
  - Abdominal pain.
  - Communication with biliary tract: cholangitis, rigors, jaundice.
  - Rupture from trauma.
- Chest
  - Lungs (25% of cysts).
  - Pleuritic pain and cough.
  - Often asymptomatic, detected on chest *X* ray.
- Other areas
  - Brain: space-occupying lesions (3–5% in some countries).
  - Bone cysts: pathological fractures, respond poorly to chemotherapy.

Diagnosis

- Ultrasound is effective in detecting liver and abdominal cysts. Presence of a separated membrane or daughter cysts makes diagnosis highly likely. Needs to be differentiated from simple hepatic cysts.
- Lung cysts are visible on plain *X* rays.
- Eosinophilia is only present in around 20% of cases.
- ***Serology; ELISA has 90% sensitivity but may be negative with completely walled-off or extrahepatic cysts. Cross-reactions may occur with other helminths, especially cysticercosis.***

Treatment

Asymptomatic or calcified cysts usually require no treatment, but patients should be followed up with repeat ultrasound scans.

Albendazole

- Cycles of albendazole 7.5–10 mg/kg orally 12 hourly for 28 days followed by 14 drug-free days. If possible give at least two courses before surgery.
- Re-examine cysts by ultrasound after three courses and give further treatment as necessary.
- Liver damage and bone marrow depression are rare but important complications.
- Albendazole reaches therapeutic peak levels within 4 hours so even a single dose before interventions may have prophylactic value
- Avoid in pregnancy (teratogenesis).

Surgery

- Surgical removal is standard treatment; avoidance of spillage of cyst contents is essential.

- High rates of recurrence and of surgical complications are recorded in inexpert hands.
- Scolicides which destroy the worms, such as hypertonic 10% saline, 95% alcohol or cetrimide are injected into cysts before removal.
- Important to avoid scolicides entering bile ducts which may cause sclerosing cholangitis.

Aspiration under ultrasonic control

- Replacement of cyst contents with a scolicide and reaspiration is increasingly advocated. Each daughter cyst must be punctured.
- Risk of anaphylaxis, biliary spillage and recurrence or spread. **Epinephrine must be immediately available** ✔.
- Long-term follow up studies of effectiveness are not available.
- ***Endoscopic retrograde cholangiopancreatography (ERCP) to detect cyst/bile duct connections before aspiration can reduce risk***.

4.30 Schistosomiasis

Brian Coulter

Minimum standards requirements

- Public health measures to improve water and sanitation
- Urine and faecal microscopy ± rectal biopsy
- Hydration and nutritional support
- Praziquantel

Schistosomiasis occurs in areas of the world where there is the combination of warm fresh water containing specific snails and urinary and/or faecal excretion of *Schistosoma* eggs by humans.

Parasite and life cycle

- Eggs are passed in stool or urine into fresh water containing snails viz *Bulinus* (*S. haematobium*) and *Biomphalaria* (*S. mansoni*).
- Miracidia hatch from the eggs, penetrate the snail, replicate into cercariae which are then released into the water.
- Cercaria penetrates the skin (or pharyngeal mucosa) of humans, loses its tail and becomes a schistosomula which is transported to the lung capillaries. It reaches the left side of the heart and is distributed throughout the body. Those that reach the portal system develop into mature worms in the liver.
- Adult males and females copulate and migrate in pairs to their professed egg-laying sites, viz *S. haematobium* to the vesical veins and pelvic plexus and *S. mansoni* to the superior and inferior mesenteric veins.
- Females produce eggs daily throughout their average 3–4 years lifespan. Most eggs pass through the vessel wall and about 50% reach the lumen of the urinary tract or intestine and are excreted. Those that remain in the tissues provoke an immune reaction which causes the disease. Some eggs are transported to the liver and some reach the general circulation.

Pathogenesis

Pathogenesis can be divided into four stages.

1. **Dermatitis**. An itchy papular rash "swimmers itch" lasting 1–2 days may develop as a result of humoral immune reaction to invading cercariae and schistosomulae. However, it is more likely to be due to avian schistosoma (non-pathogenic to man). Older children and adults develop a degree of resistance to this stage of invasion.
2. **Katayama fever** (2–8 weeks). Humoral reaction to adult worms and eggs results in an acute illness associated with formation of immune complexes. Symptoms include fever, rigors, malaise, diarrhoea, cough, hepatosplenomegaly and marked eosinophilia. Self-limiting disease.
3. **Established disease** (usually after 2 months). T-cell delayed-hypersensitivity response to eggs deposited in tissue resulting in granuloma formation. If worm load is reduced by drug therapy at this stage granulomata may resolve leaving little disease.
4. **Fibrotic complications**. Repeated infections without treatment eventually result in fibrosis, i.e. of the ureter/bladder (*S. haematobium*) and liver (*S. mansoni*). Little response to drug therapy at this stage.

Table 4.30.1 Schistosomiasis: geographical areas

Schistosoma	Disease	Area
S. haematobium	Urinary tract	**Africa**, Middle East
S. mansoni	Intestines, liver	**Africa**, Middle East, **South America**
S. intercalatum	Intestines, liver	Central and West Africa, uncommon
S. japonicum	Intestines, liver	**China, Indonesia, Philippines**
S. mekongi	Intestines, liver	Laos, Kampuchea Small number of foci

Major *Schistosoma* and main areas are in bold.

Epidemiology

- Schistosomiasis is associated with communities living near swamps, rivers, irrigation canals and rice fields who have poor hygiene and sanitary facilities and lack of ready supply of clean water.
- Infection is highest in children (5–14 years) who are an important reservoir of infection because of their indiscriminate excretion habits near and in water.
- Infections decrease after puberty but adults are still at risk when farming or washing clothes.

Clinical features

Table 4.30.2 Symptoms and complications of *S. haematobium* and *S. mansoni* †

Initial stage	*S. haematobium*	*S. mansoni*
Swimmers' itch Katayama fever*	Terminal haematuria Obstructive uropathy	Bloody diarrhoea Anaemia Hepatic fibrosis**
	Calcification of bladder and lower ureters	Portal hypertension Ascites Colonic polyposis
	Bladder calculi	Nephropathy

† Similar complications occur with *S. japonicum*.
* More severe with *S. japonicum.*
** May occur with *S. haematobium.*

S. haematobium

- Terminal haematuria, there may be dysuria.
- In a minority of children frequent untreated infections eventually lead to structural disorder of bladder and lower ureter resulting in obstructive uropathy, hypertension and chronic renal failure.
- Obstruction can be demonstrated by ultrasonography and intravenous pyelogram. Adequate treatment in the early stages may be followed by resolution of ureteric lesions.

S. mansoni

- Bloody diarrhoea. In long-standing cases severe iron-deficient anaemia, and even heart failure (due to anaemia).
- Protein-losing enteropathy with hypoalbuminaemia may result from colonic granulomatous disease and polyps.
- Left lobe of liver is enlarged more than right. Ascites may occur. Liver function is usually well preserved.
- Marked splenomegaly due to portal hypertension is associated with pancytopenia.
- Haematemesis from oesophageal varices is the final event which influences prognosis.
- Ultrasonography is useful for grading degree of periportal fibrosis and in differential diagnosis from other liver diseases.
- Acute and long term management of oesophageal varices requires endoscopy and decisions regarding sclerotherapy (see Chapter 3.10 on liver disease).
- Nephropathy due to immune complex disease may manifest with microscopic haematuria and proteinuria or nephrotic syndrome. Nephrotic syndrome has a poor prognosis, especially if associated with amyloid disease.

Salmonella infection

Schistosoma worms may harbour *Salmonella* species including *S. typhi* which cannot be eradicated until the schistosomiasis is treated. Occurs in both *S. haematobium* and *S. mansoni* infections. *Salmonella* may cause a reversible nephritis in *S. mansoni* infection.

Ectopic complications common to *S. haematobium* and *S. mansoni*

- Spinal cord myelopathy†
- Brain granulomata*†
- Pulmonary hypertension†
- Chronic *Salmonella* infection

† Less common with *S. haematobium.*
* More common with *S. japonicum.*

Diagnosis

Microscopy of urine or faeces

S. haematobium

A mid-day specimen is best. Urine should be sedimented or filtered. Viability of the eggs (and thus requirement for treatment) can be established by looking for miracidia which hatch when eggs are put in boiled water which has been cooled.

S. mansoni

If stool smear is negative on microscopy a concentration method must be undertaken. Miracidial hatching techniques are also available.

Rectal biopsy

Rectal biopsy to demonstrate the presence of eggs is undertaken if urine and faeces are negative.

Serology

Serology is of little value for diagnosis in indigenous patients but may be useful in the non-immune, for

example tourists to an endemic area. Antigen tests are being developed.

Management

Praziquantel is effective against all human *Schistosoma* species. If unavailable, metriphonate may be used for *S. haematobium* and oxamniquine for *S. mansoni*. Praziquantel is given in a dose of 40 mg/kg in 2 divided doses given 4–6 hours apart on one day. For heavy *S. mansoni* infection and for *S. japonicum* 60 mg/kg is advised, given in two doses 4–6 hours apart. Repeat urine or stool examination should be done at 3–4 months. Metriphonate is given over 6 weeks as 3 doses of 7.5–10 mg/kg orally: each dose at 2 weekly intervals.

Prevention

Control of schistosomiasis is very difficult. Measures include regular mass treatment of communities and improvement in water supply, sanitation and hygiene. Molluscicide (use of chemicals to kill the snails) is usually impractical and too expensive for general use.

4.31 Rickettsial diseases

James Bunn

Minimum standards requirements

- Supportive care and hydration
- Early treatment with doxycycline, chloramphenicol
- Public health measures and vector control

- Rickettsial diseases are caused by intracellular Gram-negative organisms similar to bacteria.
- Illnesses are restricted by geography to places where both the natural animal host and its insect vector are present, and the vector has contact with humans.

Clinical presentation

- Illnesses are generally characterised by fever, rash and malaise. They are often misdiagnosed as measles, meningococcaemia, typhoid, or rheumatic fever, or investigated as a pyrexia of unknown origin (see Chapter 2.14).
- Disease is caused by a vasculitis of small blood vessels, which on the skin is seen as a petechial or haemorrhagic rash. The vasculitis may affect many organ systems, and explains the wide range of symptoms seen.
- There are features specific to individual rickettsia, including meningoencephalitis (in Rocky Mountain spotted fever), myocarditis and cough (Q fever) or lymphadenopathy and hepatosplenomegaly (scrub typhus).
- An eschar at the site of the infecting bite is helpful in the diagnosis of tick-borne and mite-borne rickettsial infections, and is recognised as a necrotic black papule.
- The severity of illness varies with the organism, and age of patient. For example, in Rocky Mountain Spotted fever, the untreated acute illness has a case fatality rate of 20%, with two-thirds of cases occurring in children under 15 years. In contrast, louse-borne typhus may only cause mild symptoms in children, with deaths occurring mainly in adults.

Diagnosis

- Diagnosis is usually clinical, with serological confirmation as appropriate.
- ***In humans most rickettsia cause a non-specific serum antibody cross-reaction to Proteus vulgaris antigen, and this is used in the Weil–Felix agglutination test.***
- ***The Weil–Felix reaction does not occur for rickettsial pox, Q-fever or ehrlichosis for which specific diagnostic serological tests are available.***
- Treatment need not await serological diagnosis, as this is often delayed.

Treatment

- In older children tetracyclines, particularly doxycycline (2.2 mg/kg twice daily to 100 mg twice daily maximum). Not advised under 8–10 years because of dental staining.
- Oral chloramphenicol (25 mg/kg/ four times daily to 3 g/day maximum) is also effective.
- Treatment should be for 7–10 days.
- Fluoroquinolones (for example ciprofloxacin) may be effective and are being evaluated.

Table 4.31.1 Some major Rickettsia and their distribution

Disease	Agent	Vector	Reservoir	Distribution
Rocky Mountain Spotted fever	*R. rickettsii*	Ticks	Rodents, dogs, rabbits	USA, South America, Canada
Rickettsial pox	*R. akari*	Mite	Mouse	Worldwide
Louse-borne typhus	*R. prowazekii*	Lice	Human	Worldwide
Murine typhus	*R. typhi*	Flea	Mouse (urban)	Worldwide
Scrub typhus	*Orienta tsutsugamushi*	Mite	Rodents	Australia, India, SE Asia
Q-fever	*Coxiella burnetti*	None	Cattle, goats	Worldwide

Control

- Insect vector control is important, for human louse-borne typhus, which occurs in cold mountainous areas where people live close together, or in internally displaced or refugee populations. In these situations, delousing of individuals with insecticides prevents and controls epidemic typhus. For scrub typhus, mite bites can be prevented with topical insect repellents.
- A vaccine is available for Rocky Mountain spotted fever.

Health education

This may include

- Community education on the risks of living in very close proximity to animals.
- The need for regular refacing of mud walls and floors.
- For human louse-borne typhus, the importance of washing/sunning clothes and bedding.

4.32 Scrub typhus

Pornthep Chanthavanich and Brian Coulter

Minimum standards requirements

- Serology
- Chest *X* ray
- Doxycycline, tetracycline, chloramphenicol

Epidemiology

- Geographical distribution: Asia, Australia, Pacific Islands.
- Agent: *Orientia tsutsugamushi* (Rickettsia tsutsugamushi).
- Hosts: Rodents are reservoir hosts. Humans are accidental hosts. Common age group is 5–14 years and more common in boys.
- Vector: Larva of trombiculid mite. Mites live on jungle grass and become infectious by biting and sucking tissue fluid of infected rodent or by transovarian transmission to the next generation of mites.

Clinical manifestations

- Incubation period 5–18 days.
- Abrupt onset of fever, severe headache, myalgia, cough, suffused conjunctiva, dark red papular or maculopapular rash (5–7 days after fever) on trunk, arms and thighs.
- Eschar (19–28% in children, 46–82% in adults) may be seen at site of mite bite especially in perineum or axilla or trousers' belt region. Eschar is a firmly adherent black scab, 3–6 mm in diameter, with raised red margin.
- Regional or generalised lymphadenopathy, hepatomegaly and sometimes maculopapular rash. Moderate leucocytosis may be seen and occasionally thrombocytopenia.
- **In severe cases**, complications include meningoencephalitis, myocarditis, pneumonitis, respiratory distress syndrome or rarely renal failure.
- **In non-severe cases**, fever subsides within two weeks. Indigenous people in endemic areas usually have mild illness without rash or eschar.

Diagnosis

- Diagnosis is based on clinical manifestations, geographical distribution and history of contact with jungle grass exposure in the bush.
- Confirmation is by serology or polymerase chain reaction. Weil–Felix test titre ⩾1:160 (or fourfold rise after 2–4 weeks) occurs in only 50% of cases. ***More sensitive serologic tests are the indirect immunoperoxidase test and the indirect immunofluorescent tests. For individuals living in endemic areas the positive titre is ⩾1:400 or a four fold rise in acute and convalscent sera. The positive titre indicating infection may be lower in non-endogenous children.***
- Routine blood examinations are unhelpful but are required to rule out other diseases such as Dengue haemorrhagic fever, malaria and leptospirosis.
- Blood culture to exclude septicaemia, for example typhoid.
- Chest *X* ray is indicated if there is cough and dyspnoea to detect pneumonitis, pleural effusion or respiratory distress syndrome.
- Perform lumbar puncture if there is meningism or severe headache to rule out other causes of CNS infection. CSF commonly shows a picture of aseptic meningitis.
- A fall in body temperature usually occurs within 24–48 hours after treatment.

Management

- Drug of choice: doxycycline orally 2.2 mg/kg initially followed by 2.2 mg/kg 12 hours later, then 1.1 mg/kg every 12 hours until afebrile for 2–3 days **or** continue treatment for 5–7 days.
- Alternative drugs: tetracycline 250 mg orally four times a day for 7 days (>8 years) or chloramphenicol 15–25 mg/kg orally four times a day for 7 days – depending on severity.
  In a few cases fever returns 5–7 days later. Repeat dose of antibiotic.
  Tetracycline should not be given to oliguric patients. Doxycycline is safe in renal impairment.
- In severe cases, risk of dying outweighs risk of tooth discoloration from doxycycline or tetracycline.
- **Remember antimicrobial agents only suppress infection. Cure depends on host immunity.**
- Treatment should not be withheld pending laboratory confirmation for a clinically suspected infection.

4.33 Other infections caused by parasites

David Southall and Brian Coulter

Toxoplasmosis

- Infection by *Toxoplasma gondii* is usually acquired from the faeces of cats.
- The most common manifestation is a congenital infection acquired during pregnancy.
- See Chapter 4.18 for information on congenital toxoplasmosis. The key features are fever, rash, petechiae, lymphadenopathy, hepatomegaly, splenomegaly, jaundice, hydrocephalus or microcephaly, microphthalmia, epileptic seizures, cerebral calcification and chorioretinitis.
- Acquired infections are usually mild presenting with a glandular fever-like illness. Severe acquired infection occurs in immunosuppressed children particularly those with AIDS (see Chapter 4.18 for management).

Amoebiasis

Infection by *Entamoeba histolytica*

- This is acquired from human hosts via contaminated food, water or by direct contact.
- Most infected children are asymptomatic but some present with acute diarrhoea with colicky abdominal pains.
- A small proportion have bloody diarrhoea with a fever and systemic illness. This can last for many weeks.
- Rarely intestinal perforation with peritonitis or haemorrhage may occur.
- The diagnosis can be confirmed by observing the amoebae in a fresh stool or following a biopsy of the ulcers at sigmoidoscopy.
- Amoebic liver abscess(es) occurs in less than 1% of infected individuals. They present with fever, abdominal pain, and a tender liver sometimes with a palpable mass. The liver abscess often occurs without gastrointestinal symptoms and with negative stools. The diagnosis can be confirmed by ultrasound scan or ***computed tomography scan if available***.
- Treatment is required for those with systemic illnesses, those with diarrhoea due to invasive ulceration and those with liver abscesses.
- Metronidazole is the drug of choice and is well absorbed orally: 7.5 mg/kg three times daily for 5–10 days (maximum daily dose 400 mg).
- If the abscess is very large and particularly if there is concern that it may rupture, it may require aspiration under careful ultrasound support.
- After the acute treatment of a liver abscess, diloxanide should be used immediately following the course of metronidazole in order to remove all amoeba from the bowel.

  The dose of diloxanide is:

1 month to 12 years	7 mg/kg three times daily for 10 days
>12 years	500 mg three times daily for 10 days.

Cryptosporidiosis

- *Cryptosporidium parvum* can be acquired from infected human or animal hosts and from contaminated water and food.
- It causes an acute gastroenteritis, self-limiting in most children. The enteritis is associated with watery diarrhoea, nausea and colicky abdominal pains. It lasts for approximately two weeks. In otherwise healthy children it does not usually require treatment with antimicrobials unless it persists or is associated with systemic illness in which case azithromycin may be effective.
- In children with AIDS it can produce a protracted and severe illness involving major weight loss.(See Chapter 4.18 for details of its treatment under these circumstances (with azithromycin).

Azithromycin:

6 months to 12 years	10 mg/kg once daily for 3 days or longer in AIDS
12–18 years	400–500 mg once daily for 3 days or longer in AIDS

Avoid in patients with liver disease.

Giardiasis

- Infection by *Giardia lamblia* can be acquired from infected human or animal hosts and from contaminated water and food. They live in the duodenum.
- The infection may be asymptomatic.
- It can produce an acute gastroenteritis with watery stools, colicky abdominal pains and nausea.
- It can also produce a chronic diarrhoeal illness with malabsorption and colicky abdominal pain lasting for many months.

- Diagnosis is best made from examining a fresh stool. Sometimes more than one examination will be necessary.
- Metronidazole (doses as in amoebiasis above) is appropriate in the chronic form of the infection. The acute form usually resolves without treatment.

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Section 5

Injuries

How to use this book

This is a comprehensive text for all paediatricians caring for children in hospital. It can be used by those with limited resources and also where greater resources are available. We have identified the different levels of care in the following ways:

- **Minimum standards requirements** are given in a highlighted box at the the beginning of each clinical chapter.
- ***A standard of care*** where resources are not limited appears as bold, italicised text.
- **Key points** of particular importance in management of children are identified by a tick in the margin and bold text.

In this way we hope the book will act as a user-friendly, speedy reference on any paediatric ward.

5.1 Childhood accidents and their prevention

David Southall

Introduction

- Accidents are the commonest cause of death among children age 1–14 years.
- Accidents cause half of all deaths of children aged between 10 and 14 years.
- Accidents result in many children being permanently disabled annually.
- Accidents lead to one-fifth of all hospital paediatric admissions.

✔ **Most accidents can be prevented**.

Risk factors

Boys are more frequently injured than girls. The difference emerges at age 1–2 years.

Children's accidents are intimately related to development. A newborn baby can only fall if dropped, or if a parent falls holding the baby. An older baby can wriggle and roll off a changing table or a bed. A crawling baby can climb upstairs and fall back. A small child can climb and fall out of a window. An older child can climb a tree, or fall in a playground. Knowledge of development helps anticipate dangers.

Exposure to different circumstances also varies with age. Children under 5 years experience accidents at home. School-age children experience accidents at school, sport and play, and are especially at risk of accidental deaths as pedestrians.

Accidents are linked to inequalities in environments. Children in poor families are twice as likely to die from an accident as children in rich families and for some accident types, such as burns, the chances are six times higher. This does not mean that poor parents care less about their children than rich parents, or they do not know about accident risks. It may mean that there are many other pressures – overcrowding, lack of money, poor housing – and there is less power such as owning one's own home and being able to afford safety equipment to make changes for safety.

Accidents are more common in families where there is stress from mental illness, marital discord, moving home and a variety of similar factors.

Accident prevention

- **Accidents can be prevented completely**. This is termed primary prevention. An example is a fireguard preventing access to an open fire.
- The harm caused by an accident can be minimised. This is secondary prevention. For example, a seatbelt can reduce injury even if a car crash occurs.
- Finally, rapid attention to an injury can **reduce mortality and morbidity**. This is tertiary prevention. Examples are cold water on burns and scalds or pressing on a laceration.

There are three main approaches to accident prevention. These are the following:

- **Education**
  Increasing knowledge about a problem and the solutions, to change attitudes and eventually behaviour.
- **Engineering**
  Safe design of products and the environment, including the architecture of the home.
- **Enforcement**
  The role of legislation, regulations and standards in accident prevention.

  Countermeasures can be active, that is a conscious decision to use them has to be taken every time, such as putting pans on the back hobs of the cooker. Or they can be passive, that is, built in to the product: for example, junior formulations of paracetamol are sold in small bottles that do not contain a lethal dose.

  There are a variety of ways in which doctors and other healthcare personnel can participate in reducing children's injuries.
- **Be informed**
  Medical education before and/or after graduation.
- **Set a good example**
  Wear your seatbelt. Drive carefully past schools. Consider your own home and family with safety in mind.
- **Take opportunities**
  Can you offer safety advice to a family after an accident has happened? Do you know possible preventative strategies for that accident type? Have you developed the communication skills to listen to parents and advise them? Can you photograph that injured child in that setting, and use it to support a family in improving their household, or a neighbourhood in a media campaign?

- **Collaborate with others**
  Be prepared to participate in working groups and campaigns. You have special expertise and influence to offer.

Advice the hospital can give parents about accident prevention

- Always have a smoke detector in your house.
- Use a car seat. Do not nurse baby/child on your lap in a car.
- Keep domestic hot water temperature below 42°C.
- Use stair gates when there are children up to 2 years of age in the house.
- Use window locks. Your child can easily fall out.
- Young children should not play in the road. Teach the "stop, look, listen" code at the roadside whenever you are with him/her. Teach him/her to take you across the road.
- Children up to 3 years can drown in very shallow water (for example, garden ponds or paddling pools). Contact between their faces and water can lead to the stopping of breathing, inability to move and loss of consciousness.
- Do not leave children alone in the house.
- Teach him/her what to do if becoming separated from you in a shop, or busy place.
- Be particularly careful about saucepans and kettles containing hot liquids. They can easily be pulled on to children, resulting in severe burns.
- Use a fire guard at all times.
- Keep medicines out of reach of all children, ideally in child-proof containers.
- Keep household chemicals and kerosene out of reach – preferably in a locked cupboard.
- Keep matches out of reach.
- Do not use pillows with children under 3 years of age.
- Babies and young children can choke on small objects like peanuts, buttons and pins.
- Keep plastic bags away from children.
- Keep firearms away from children.

Suggested advice from parents to their children on preventing harm/accidents when they go out alone

- Never go off with an adult they do not know without first seeking permission from a parent or other person responsible for their care.
- Never go out alone without first telling the adult looking after them where they are going, who they are going with and when they will be coming home. They should know that they should always return on time or their parents can become very worried.
- They should always carry enough money to make a phone call home in an emergency.
- If they are being bullied at school:
  - Enlist the help of friends to help them confront the bullies. All bullies are cowards.
  - If this is not possible, or it does not work, inform their teacher and/or their parents or the person they live with.
- If they have to walk alone after dark, choose a busy, well-lit route and never take short cuts through secluded areas.
- They should trust their instincts about people they meet. If they are in doubt, do not go off with them, particularly if they will be alone with them.
- If they are attacked, it is okay to shout, kick, bite and do anything that will help them to escape.

Advice with respect to firearms and children

- Parents should never give or show arms/weapons to children even if the arms/weapons are unloaded or unusable.
- If arms/weapons must be kept at home, ensure they are secured and out of reach of children. For example: in a (wall) safe, locked drawer, inside and on the top shelf of a locked cupboard/wardrobe, or any other unknown safe place (hole in floor or wall).
- Before arms/weapons are put away, they should always be checked **twice** to ensure they are unloaded and unusable/not in a position to be fired.
- Always check the barrel of the gun and the bullet chambers are empty. Unchecked or unsafe weapons often result in serious accidents.
- Always keep the weapon and the ammunition separate.
- Try not to encourage children to play with weapons or weapon-like toys.

Acknowledgement

The first part of this chapter was written as a summary of the excellent paper in Advanced Life Support Group *Advanced Paediatric Life Support, 3rd edn.* London: BMJ Books 2001.

5.2 Child ill-treatment and abuse

Neela Shabde and David Southall

Minimum standards requirements

- Social services/police child protection system
- Forensic analysis
- Skeletal *X* rays
- Photography

Physical ill-treatment or abuse (non-accidental injury)

This rarely exists without emotional ill-treatment or abuse and sometimes accompanies sexual abuse.

Injuries include:

- Burns and scalds.
- Multiple bruises of different ages.
- Finger-tip bruising on the chest and arms (from gripping/pinching/shaking).
- Bruises on the cheeks.
- Torn frenulum (bottle/dummy forced into mouth).
- "Black" eyes or slap marks.
- Bony injuries:
  - fractures of long bones, skull and ribs
  - epiphyseal separation at the end of long bones
  - periosteal separation and haematomas
- (Especially in infants) tearing of the superficial veins over the brain and retinae causing subdural and retinal haemorrhages: it can kill, leave physical and mental handicap and visual loss.
- Bite marks.
- Failure to thrive due to neglect (category 2) or deliberate starvation (category 3). (See Chapter 1.30.)
- Induced illness including suffocation or poisoning.

Features of presentation suggesting ill-treatment or abuse

- Delay in seeking medical help.
- Often presents late at night inappropriately well dressed.
- History of "accident" too vague or too rehearsed with inconsistencies. History changes on retelling or questioning.
- No explanation at all.
- History of "accident" inconsistent with the injury observed or the developmental age of the child.
- Parents evasive or hostile.
- Child's interactions with parents abnormal.
- Child looks sad, withdrawn, anxious or frightened (frozen watchfulness).
- Child may indicate the aggressor.
- "Collusion of silence' or one parent may implicate the other.
- The compliant child – very anxious to please.

Patterns of injury which indicate non-accidental injury

- Some patterns of injury are diagnostic:
  - finger-tip bruising, especially multiple
  - cigarette burn
  - lash or belt marks
- Injury inconsistent with the history: for example symmetrical scalds of both feet up to the ankles.
- Injury inconsistent with the child's development: for example, head injury in a non-ambulant child.
- Multiple bruises of differing ages: (see Figure 5.2.1)
  - Site of bruising, for example behind or on ears
  - Burns and scalds
    child held and dipped in hot water – symmetrical burns of feet and lower limbs and often also of the buttocks
    cigarette burns – often multiple and can be confused with impetigo
    hands, bottom or face held against a hot object.

Neglect

- Persistent failure to meet a child's essential needs by omitting basic parenting tasks and responsibilities.
- Examples include failure to protect a child from dangers (including cold or sunburn), failure to thrive, failure to meet the child's emotional need for love, affection and stimulation.

Sexual abuse

A broad definition would be any use of children by adults for their sexual gratification. For example, pornography, exposure to indecent acts, inappropriate touching and fondling, masturbation of an adult, intracural, vaginal, oral or anal intercourse. The child may be dependent and/or developmentally immature.

General points

- Sexual abuse may present in several ways.
- Diagnosis of sexual abuse is achieved through a multi-agency assessment, the medical examination is only one part of this process.
- Physical examination is important as abnormal signs may be the first sign of sexual abuse and may also corroborate the child's disclosure. However, physical examination may be normal in victims of sexual abuse.
- Medical examination should be carried out with due respect and sensitivity.
- Physical abuse and sexual abuse are seen together in around 15% of cases.
- Management is based on the same principles as other abuse and is essentially multidisciplinary.
- Sexual abuse occurs in all social classes, in all cultures and is underdiagnosed. The incidence is unknown.

Presentation

This may occur following:
- Disclosure by the child.
- Symptoms of local genital or oral infection – redness, soreness, discharge.
- Symptoms of local genital or oral trauma – bleeding, pain, bruising.
- Emotional and behavioural problems – enuresis, encopresis, hyperactivity, poor attention, anorexia, attempted suicide.
- Sexualised behaviour, inappropriate sexual knowledge.

Physical signs in sexual abuse

These require careful interpretation in the context of a full medical assessment.

Clinical signs suggestive of abuse
- Grip marks or bruising on thighs, pelvis, peri-anal area.
- Bite marks.
- Bruising, swelling, erythema, abrasions, tears of the external genitalia and introitus or mouth – due to attempted or actual penetration.
- Damage to fourchette, peri-urethral structures, hymen and vaginal mucosa.
- Scarring of posterior fourchette or vaginal wall.
- Pregnancy.
- Sexually transmitted diseases, including HIV infection (see Chapters 4.2 and 4.18).
- Presence of sperm in the vagina or rectum.
- In boys – penile bruising and laceration – without a history of accidental trauma.
- Anal signs – are difficult to interpret unless there is acute anal trauma; for example, the tyre sign – peri-anal oedema associated with anal penetration, usually resolves within 36 hours.

Signs which may possibly indicate sexual abuse but which by themselves are not diagnostic.
It is important that these are documented in the notes.
- Thickening of hymen, bumps and notches of the hymen, perihymenal adhesions, hymenal tears and loss of hymenal tissue.
- Size of hymenal orifice.
- Presence of labial fusion.
- Peri-anal erythema or venous distension, anal fissures, anal dilation, reflex anal dilation and anal funneling.

Emotional ill-treatment or abuse

Persistent or severe emotional ill-treatment, abuse or rejection resulting in severe adverse effects on emotional and behavioural development. All abuse involves some emotional abuse. This category should be used where it is the main or only form of abuse.

Examples include:
- Persistently punishing for normal/desirable behaviours.
- Persistently acting towards a child in a negative way by ridiculing, humiliating, scape-goating or name-calling.

Consequences of emotional abuse

These vary with age
- Impaired physical development – often fail to reach their optimum potential in growth. Height can be well below the third centile for age and improves when the child is placed in a more nurturing environment.
- Behavioural abnormalities – anxious attachment, lack of social responsiveness, expressionless face, child is afraid to speak, eagerness to please, attention seeking, overactive/"hyperactive", no wariness of strangers, hunger for human contact, inability to form relationships, self-injurious or self-stimulating behaviours, hoarding and stealing of food, pica, enuresis and encopresis, bizarre behavioural patterns (sometimes autistic-like behaviour).
- Impaired psychological development, especially speech and language – aggression, emotional unresponsiveness, emotional instability, impaired social development, lack of social esteem, dependency, serious social difficulties, underachievement, negative self-evaluation, poor concentration, attention and school performance.
- Psychiatric disorder – emotional ill-treatment and abuse has been described in association with three psychiatric disorders of childhood:
  - Depression
  - Reactive attachment disorder of infancy
  - Multiple personality disorder

In general children become sad, dejected and withdrawn.
- Physical problems:
  - Failure to thrive
  - Recurrent and severe nappy rash
  - Generally unkempt appearance with poor hygiene
  - Recurrent minor infections
  - Recurrent attendances to accident & emergency department or recurrent hospital admissions without serious illness or injury.
- Children suffering abuse are often threatened by being told they will be to blame if the family is separated. Fear of what might happen to them may result in children

between the ages of 4 and 10 years colluding with the abusive parent.

Management/protection of children from ill-treatment or abuse

- The welfare of the child is paramount.
- Multi-agency collaboration (medical/nursing staff, social workers, police, courts).
- A legal framework within the country.
- Once protection is achieved, children will benefit from expert psychological intervention and emotional support.

Immediate action when ill-treatment or abuse is suspected

- A detailed history and **full medical examination** are required (including examination of the genitalia).
- If verbal, child should be asked how he was hurt (ideally separately from parent/carer).
- Consider other children in the family who may need to be examined.
- Check if family is known to police or social services.
- Admit child to hospital if observation or treatment is indicated or to a place of safety if the child is considered to be at risk. Staff can then have the opportunity of talking with the child.
- If parents refuse to allow child to be examined or admitted, urgent action to protect the child will be required within the legal framework of the country.
- This may mean referral to the duty social worker or the police.
- Meticulous, legible and full record of history, examination and subsequent actions must be recorded in the child's medical notes. It is important to record history as it is spoken and include evaluation of the parent's reactions and attitudes.
- All the injuries must be noted in detail and photographs taken if possible and with child's permission.
- Bruises can be recorded on a body chart (see Figure 5.2.1).

A thorough medical examination should include:

- Child's demeanour.
- Height weight and head circumference (in pre-school child) plotted on a centile chart.
- Examination of mouth, nose, ears, neck, genitalia.
- Inspection of skin surface for bruises, marks, cuts.
- Examination of eyes for retinal haemorrhages (may need pupil dilatation (see Chapter 3.34).
- Systemic examination.
- An assessment of the child's developmental age.

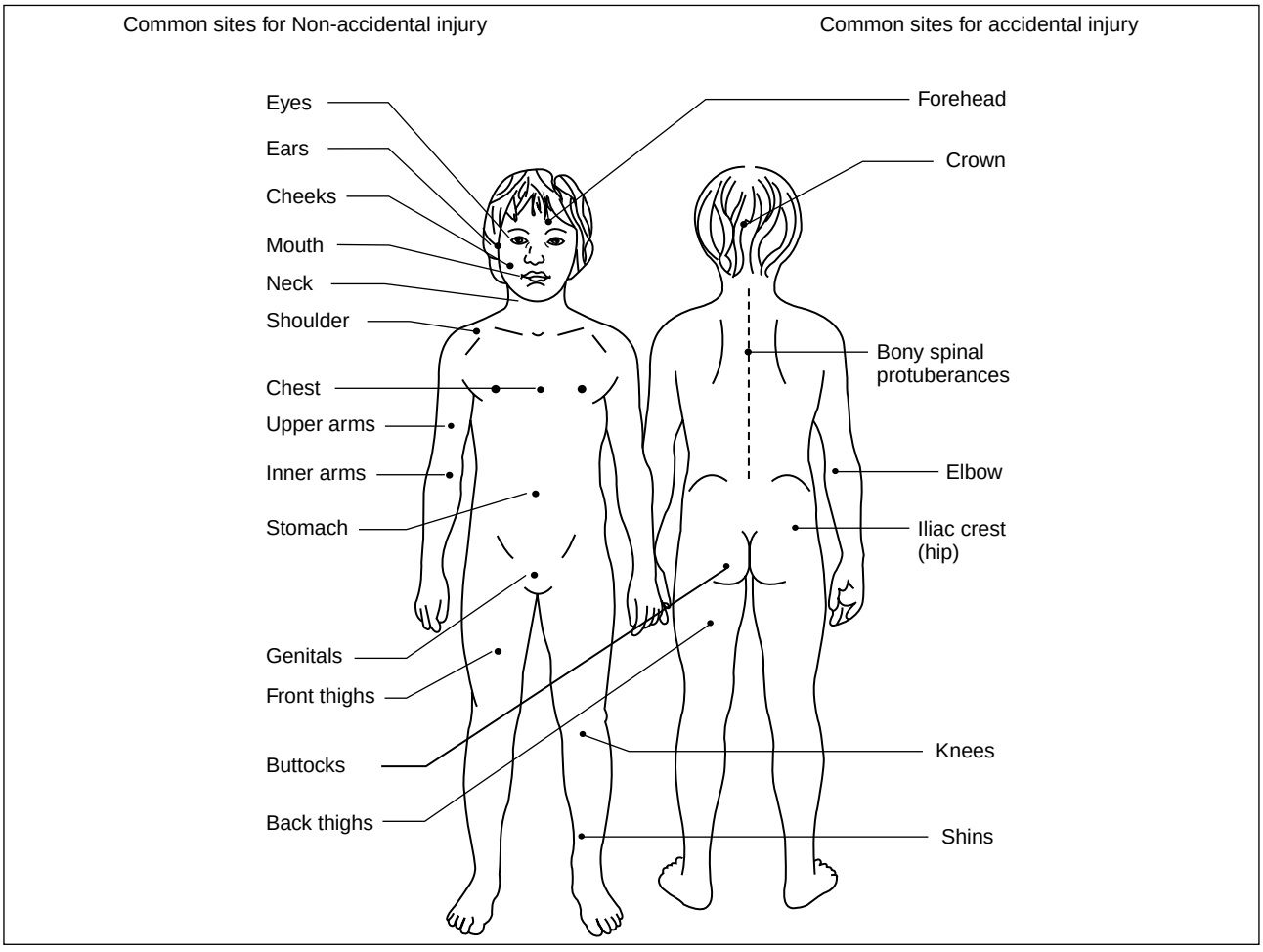

Figure 5.2.1 Common injury sites.

Subsequent management

Investigations to exclude medical causes include:
- Full blood count, platelets and clotting screen.
- A detailed skeletal survey should be performed (especially in children under 2 years) to look for new and old fractures.

Induced illness syndrome (IIS)

Factitious illness syndrome (IIS) or factitious illness syndrome (also known as Münchhausen syndrome by proxy), is thought to present the most severe end of a spectrum of "pathological healthcare seeking behaviours". These can be described as unusual, abnormal, unreasonable or excessive healthcare-seeking behaviour in which parents attempt to meet their own conscious or subconscious needs. There may also be attention from the media or financial benefits. It is probably more common in rich than poor countries.

Definition

Significant harm which is caused to a child by the actions of a parent or other carer who deliberately fabricates symptoms or induce medical symptoms in a child, which would not otherwise be present. The actions may be as a result of omission or commission and include such behaviours as:
- Deliberate poisoning
- Deliberate burning or other damage to the skin to induce symptoms
- Deliberate suffocation to induce symptoms
- Removal of or tampering with necessary equipment
- Introducing foreign material to tests or other behaviour which causes damaging or unnecessary tests to be performed on the child
- Deliberately fabricating fits in the child.

Criteria for diagnosis

- Illness in a child is fabricated by a parent, or someone who is acting as the child's carer.
- The child is presented for medical assessment and care, usually persistently, often resulting in multiple medical procedures.
- The perpetrator denies the aetiology of the illness in the child.
- Acute symptoms and signs of illness cease when the child is separated from the perpetrator, almost always the mother.

Prevalence

The exact prevalence is not known. In the UK, a two year prospective study identified an annual incidence of at least 2.8/100 000 in children under 1 year of age and at least 0.5/100 000 in children under 16 years of age. Less life-threatening, but nevertheless very damaging forms of deception are not uncommon and may either go unrecognised, or attempts to intervene may fail because of insufficient evidence.

It is important to note that a child with a known medical condition may also be subjected to such behaviour of their parent or carer resulting in excessive and inappropriate medical attention.

The perpetrator(s)

- The perpetrator is usually the mother, but the father or another carer may also be incriminated. On occasions, both parents, or parents **and** child may collude.
- Perpetrators are thought to have personality disorders but few have overt mental illness. There may be a personal history of pathological healthcare-seeking behaviours in the form of somatising disorder or factitious illness, self harm, eating disorder, compulsive lying, false accusation of physical or sexual assault or criminal behaviour.
- Perpetrators tend to be particularly skilled in concealing their activities and in misleading professionals. They are strongly motivated to continue or even escalate the abuse in order to "confirm" their case. They are very likely to deny their activities when challenged. They may become more devious and guarded and may seek alternative healthcare or move to another area, with consequent risks to the child.

Investigative process

This must involve collaboration between agencies. Having collected detailed information from the medical records of all the children and parents in the family, plus social services and police records, if available, on the parents, the next stage requires consolidation and evaluation of all the evidence to determine a critical threshold of concern and intervention. In some cases, forensic or witness evidence is obtained at an early stage, triggering immediate intervention. In some cases an admission to hospital will be required, particularly when a young child (<1 year) with an apparent life-threatening event (ALTE) for further observation and further medical investigations including covert video surveillance (CVS).

In less certain cases, time is required to collate and assess evidence, to consult with others and to assess risks before formally implementing child-protection procedures. If the risks are considered to be high, it may be necessary to separate the child from the suspected perpetrator either by voluntary or legal means. This provides the opportunity to assess whether the "illness" resolves in the absence of suspected perpetrator(s).

The Police Division of the Child Protection Unit (if it exists) may be able to provide information on parent/carer regarding relevant information pertaining to offences such as fraud, violence or deception.

It is important to note that because of the nature of the deception, true parental cooperation may be unachievable and the investigation may have to be carried out covertly either in part or throughout. This runs counter to the normal and more desirable approach of honesty and openness with parents. Nevertheless, adherence to child-protection procedure provides an appropriate framework for investigation and has to proceed with a timescale, which minimises the risk to the child.

The most difficult dilemma for professionals is how and when to confront the parents when it is safe to do so, to

be balanced against the need to protect the child from a potentially dangerous situation.

Strategy meetings

These involve all the key professionals, including police, social services, legal adviser, health professionals – consultant paediatrician, senior nurse and child mental health professionals with an interest in IIS. The main purpose is to maintain coordination, control the pace of the investigation, to plan further action, such as a need for child protection conference, a plan of action for anticipated difficulties, etc. Usually a senior member of the social services department will chair the meetings.

Court action may be necessary and will be addressed at a strategy meeting or child protection conference.

Outcomes

- Long protracted involvement between the family, the statutory agencies and courts. This may include a detailed and comprehensive assessment of the perpetrator and the family and possible rehabilitation depending on the nature of the abuse and the perpetrator's ability to admit and change the abusive behaviour.
- Retrospective research on recorded cases has revealed that criminal proceedings are uncommon – particularly for mothers. Some believe that perpetrators are unlikely to change their behaviour. There is a high level of comorbidity of siblings. Therefore, safety of other children in the family including those unborn may need consideration within child-protection framework.
- The consequences to the child of such abuse are very grave, particularly if abuse is not confirmed. These include death and brain damage in very young children and other physical morbidity. Research on detected cases has revealed both short- and long-term mental health problems in the victims due to other coexistent abuse, attachment disorder, emotional harm, educational and social disadvantage.

Key messages

- IIS is a relatively rare but serious form of child abuse with significant mortality and long-term physical and mental morbidity.
- Consensus amongst health professionals is essential in order to take effective action, but can sometimes be difficult to achieve.
- Early consultation with senior police and social service professionals is necessary even if suspicions remain unconfirmed.
- A well-coordinated multidisciplinary response must be triggered as soon as the potential harm to the child is recognised.

Perpetrators are challenging and devious and the child-protection investigation may need to be covert until there is sufficient evidence to confront the perpetrator and secure the safety of the child.

5.3 Wounds

Joan Robson

Minimum standards requirements

- Analgesia (see Chapter 1.27)
- Immunisation against tetanus (see Chapter 4.9)
- Human antitetanus immunoglobulin
- Sutures
- Adhesive strips/tissue glues

Definition

In a medico-legal context, to wound is to destroy, however superficially or minutely, a bodily surface, be it skin or mucous membrane. A contusion (bruise) is excluded.

Nature of injuries causing wounds

- Kinetic energy
- Heat
- Cold
- Chemical
- Electrical

Types of wounds

- Abrasion – friction injury, also known as "graze"
- Laceration – blunt injury
- Incision – injury from a sharp object
- Stab – injury from knife, scissors, screwdriver, poker, etc., usually penetrating
- Needlestick
- Bite – human or animal
- Firearm – shotgun, rifle, revolver or pistol (see Chapter 5.9)
- Blast
- Burn (see Chapters 5.10 and 5.11)

Note: A variety of types of wounds may coexist following a single incident.

Management of major injuries using APLS/ATLS system (see Chapters 1.19 and 5.4)

Primary survey and resuscitation

- Assess: **A**irway and cervical spine control
  Breathing
  Circulation and haemorrhage control
  Disability
  Exposure
- Identify and correct life-threatening abnormalities
- Resuscitate and stabilise vital functions

Secondary survey and emergency treatments

Remember that if simple resuscitative measures do not stabilise the child, then operative intervention may be necessary before a formal secondary survey is done. In the secondary survey:

- Determine the full extent of all injuries to the head, face, neck, chest, abdomen, pelvis, spine and extremities
- Have an emergency treatment plan to give emergency treatments in order of priority.

Definitive care

The definitive care of major injuries which include wounds is often carried out by teams which have not been involved in the resuscitation and emergency treatments. Good communications are essential using:

- Legible and detailed notes
- Prompt and efficient transfer to a unit which can give the definitive care – this may be an interhospital transfer
- Clear handover summary.

Management of minor wounds

Assessment of each wound:

- Nature of injury causing wound
- Type of wound
- Site

- Size
- Shape
- Position
- Depth
- Relevant motor function
- Relevant sensation
- Circulation distal to the wound.

Associated features

- Erythema (redness)
- Oedema (swelling)
- Contusion (bruise)
- **Surgical emphysema – needs immediate specialist care**
- Tenderness – if this extends beyond the area of the wound, a fracture may be present (see Chapter 5.6)
- Pain

General assessment

- Allergies
- Immunisation state
- Intercurrent illness
- Medication
- Past medical history
- Time of last meal

General principles

- After assessment of pain, give appropriate analgesia (see Chapter 1.27).
- If a radiopaque foreign body may be present, arrange *X* ray.
- The most important local treatment for all wounds is vigorous cleaning with sterile saline to remove dirt and possible pathogenic organisms (after analgesia).
- Local, regional or general anaesthesia may be needed to achieve optimal cleaning (see Chapter 1.13).
- Superficial, palpable foreign bodies should be removed as soon as possible.
- Removal of deeper foreign bodies may need specialist advice.
- **Dead or damaged tissue must be excised – specialist advice is needed if this involves more than a very small area of skin or mucous membrane**.
- **If tendons or nerves have been damaged then specialised care is needed**.
- Give tetanus prophylaxis if the patient is **not immunised** – ideally this should be human anti-tetanus immunoglobulin (HATI), 250–500 units IM. Wounds particularly prone to tetanus are those sustained more than 6 hours prior to presentation, those of puncture type, those with much devitalised tissue, and those contaminated with soil or dung.
- If the child has received antitetanus immunisation in the past, a single extra dose of tetanus toxoid IM should be given.

Specific injuries

Abrasions

- After thorough cleaning and debridement, leave wounds exposed or cover them for five days with vaseline gauze.
- If debris is left in a wound, epithelium will grow over it and "tattooing" will occur.

Lacerations and incisions

- Only clean, fresh wounds should be closed immediately – preferably only less than 6 hours old, certainly less than 12 hours old.
- **Distal-based flap lacerations may need specialist care if the blood supply is poor**.
- To close superficial wounds adhesive strips and special tissue glues are excellent but must not be used for deeper wounds, in which cavities will be created and healing will not occur.
- Close deeper wounds in layers without tension.
- Close skin with interrupted sutures, ideally using monofilament material.
- If the wound is compound (associated with a fracture) then an antibiotic should be given to prevent osteomyelitis (see Chapter 3.45).
- Arrange for removal of sutures at the times shown in Table 5.3.1.
- Younger patients heal more quickly. Malnourished patients take longer to heal.

Table 5.3.1 Times for removal of sutures

Site	Days
Face	4
Scalp and neck	5–7
Hand (flexor surface)	5–7
Trunk and arms (not extensor surfaces)	5–7
Legs (not extensor surfaces)	7–10
Hands (extensor surfaces)	7–10
Elbows and knees	10–14

Fingertip injuries

- Preserve maximum length.
- If the tip is amputated distal to the bone then regeneration will occur if the wound is kept clean and moist under paraffin gauze dressings changed weekly.
- Other principles of treatment are the same as for lacerations and incised wounds.

Tongue lacerations

- Most stop bleeding spontaneously and do not need sutures.
- Repair under general anaesthesia if there is profuse bleeding or the full thickness of tongue is involved.
- Use absorbable sutures.

Stab wounds

- **Stabbing may cause serious penetrating injuries to deep structures which may lead to rapid death from haemorrhage or air embolus**.

- The external dimensions of a stab wound may be deceptively small compared to the damage to underlying structures.
- Superficial stab wounds are treated in the same way as lacerations and incised wounds.
- Patients with penetrating wounds need resuscitation and emergency exploration under general anaesthesia.

Needlestick injuries (see Chapter 1.2)
- If there is **skin puncture** encourage bleeding and wash the wound thoroughly with plenty of soap and water. Dry the wound and apply a dry dressing if appropriate.
- If there is only **skin contact** wash with lots of soap and water but do not scrub. Scrubbing may damage the skin.
- If there is **splashing into the mouth** rinse with plenty of water.
- If there is **splashing into the eye** rinse with plenty of water. Obtain the help of a colleague to do this.
- If the identity of the donor (person whose blood is on the needle) is known, try to find out if that person has hepatitis B and/or HIV infection.
- Consider immunisation for hepatitis B and triple therapy for HIV if these are available.

Bites (see Chapters 4.22 and 5.13)

Firearm wounds (see Chapter 5.9)
- **Many of these will be major injuries**.
- Shotgun wounds often give severe disruption of the underlying tissues.
- Gas forced into the wound may cause the tissues along the track to go pink due to carbon monoxide.
- Rifle, revolver and pistol wounds give neat round holes and the amount of tissue damage is related to the velocity and range of the shot.
- The general principles of management are those for all types of wounds.

Blast injuries (see Chapter 5.8 on landmine injuries)

Burns and scalds (see Chapters 5.10 and 5.11)

Complications of wounds

Retained foreign body

- This will give swelling beneath the wound.
- Secondary infection is more likely if there is a retained foreign body.

Infection

- Tetanus – this is most likely to occur if the wound has been contaminated with soil and/or manure and the child is not fully immunised.
- Bacterial.
- Fungal.

Delayed healing

- This may be due to poor apposition of the edges, malnutrition and/or infection.
- Excision of the edges of the wound and secondary suture may be helpful, except in malnutrition.

5.4 Life-threatening trauma in children

Peter Oakley and Nicholas Coleman

Minimum standards requirements

- ABCDE (see Chapters 1.19 and 1.20)
- Shock (see Chapter 3.6)
- Intensive care (see Chapter 1.25)
- *X* rays, ultrasound (see Chapter 1.11)
- Cervical collars (see Chapter 5.7)
- Chest drain (see Chapter 6.12)
- Analgesia (see Chapter 1.27)
- Transport system (see Chapter 1.25)

Overview

After infancy, trauma is the commonest cause of death in children. It is also a major cause of disability, especially after head injury. Road accidents and falls predominate. Children are less likely to suffer from serious penetrating injuries, though in cities where stabbings and shootings are common, or in armed conflict, such violence overspills into childhood. Their inexperience makes them prone to accidents and their vulnerability exposes them to non-accidental injury (abuse – see Chapters 1.30 and 5.2). The patterns of injury and the physiological consequences can be quite different from adults, reflecting their different size and shape, the elasticity of their body tissues and the immaturity of their physiological systems.

In general, trauma is a disruption of anatomical structures with disturbance to physiological systems. This overview sets out the tasks required to treat the injured child: to control damaged anatomical structures and maintain physiological system control. When the injuries are life-threatening, little time is available and a structured, prioritised approach is necessary. Three phases of care emerge, with the focus shifting from physiology to anatomy and back to physiology.

- **First phase: rapid physiological assessment (primary survey) and resuscitation**
  Identify and treat immediately *life-threatening problems. This follows the classic* ***airway–breathing–circulation–disability approach*** of the primary survey and immediate resuscitation, as developed by the Advanced Trauma Life Support course. Children are especially prone to respiratory problems and their high metabolic rate, small airways and immature lung structures allow hypoxaemia to develop alarmingly quickly.
- **Second phase: anatomical evaluation (secondary survey) and definitive care of injuries**
  This consists of a systematic physical examination from head-to-toe, front-and-back, examining the body surface, the orifices, the major cavities (intracranial, intrathoracic and intra-abdominal) as well as the skeleton. In addition, investigations and imaging take place. Definitive care represents the specific therapeutic interventions for injuries identified and includes pain relief, reduction and splinting of fractures, wound care as well as surgical operations to repair or resect injured organs.
- **Third phase: intensive care**
  The focus shifts again to physiological control, but with a watchful eye kept on anatomical structures to identify any missed or latent injuries. This phase progresses, following physiological improvement, to acute ward care and rehabilitation, where residual anatomical and physiological problems are managed, the child's specific psychological needs are addressed and re-integration with normal life is planned.

A trauma system serves to integrate the various phases of care. The anatomical, physiological and psychological problems affecting the injured child and their impact on the family unit draw in a range of specialists. The system promotes multidisciplinary cooperation as the child moves from the field, through the resuscitation room in the Emergency Department, to the Operating Theatre and Intensive Care Unit. Rehabilitation is promoted at an early stage as a proactive process rather than as a route of disposal. On a regional/national scale, hospitals must cooperate to share specialist services, such as neurosurgery and paediatric intensive care. Some children will need to be transferred for specialist management after stabilisation in the initial receiving hospital. Another consequence of regional or national rationalisation is that hospitals that are less well equipped to receive injured children from the field will be bypassed unless precluded by geographical constraints.

The clinical personnel involved in paediatric trauma care should be specifically trained in dealing with children. Advanced Trauma Life Support (ATLS) and Advanced Paediatric Life Support (APLS) courses provide firm basis for training (see Chapter 1.19). In institutions

with a high incidence of major trauma in children, the trauma team members can maintain on-going experience with both children and major trauma. The catchment population will generally need to be greater than one million to achieve this. Otherwise, the trauma team should include a combination of trauma specialists and paediatricians. It should consist of core personnel from Accident & Emergency Medicine, Anaesthetics, Paediatrics (or Paediatric Intensive Care) and Surgery. Other specialists should be summoned promptly as problems are identified. Too many specialists at the outset affects the team's performance. It has been suggested that a team of five to eight persons, including two or three nurses, is optimal. In some parts of the world, limited resources and geographical isolation will preclude the development of multidisciplinary teams. Nevertheless, by training local doctors, nurses and ancillary workers in the principles of ATLS and APLS, an effective trauma response can still be achieved.

Primary survey

This rapid physiological assessment concentrates on the **identification and simultaneous management** of immediately life-threatening conditions. The assessment itself should take no more than a minute to perform, though it may be interrupted by life-saving interventions. It consists of:

- **A** Airway with cervical spine control
- **B** Breathing and ventilation
- **C** Circulation and haemorrhage control
- **D** Disability assessment
- **E** Exposure

Airway with cervical spine control

The injured child must have a clear, unobstructed airway. High-concentration oxygen should be provided through a reservoir mask or, if the breathing needs to be supported, through a self-inflating bag with an oxygen reservoir. In blunt trauma cases, where the mechanism of injury puts the cervical spine at risk, the head and neck should be kept in neutral alignment. This can be achieved immediately by manual in-line immobilisation with a clinician or a trained assistant holding the sides of the head. A correctly-fitting hard collar, side-supports and head blocks should then be used to maintain immobilisation until the spine has been cleared, though the manual in-line method should be resumed if airway manoeuvres such as intubation are necessary. **Normal *X* rays do not exclude spinal cord injury in children;** protection should remain in place until injury is excluded by clinical and radiological examination by a trained physician.

The airway may be compromised by injuries to the face, tongue or upper airway, the presence of blood or vomit, or by a decreased conscious level resulting from a head injury. Signs of obstruction include:

- Rapid respiratory rate
- Noisy breathing (in total obstruction may be silent)
- Recession/paradoxical breathing
- Cyanosis
- Agitation or drowsiness
- Decreased or absent breath sounds on auscultation.

The airway should be cleared of debris and a careful jaw thrust applied. If there is no improvement an oropharyngeal airway should be inserted. If the airway is still obstructed, the child requires a definitive airway. Other situations, such as severe head injury or hypovolaemia may also require a definitive airway.

The preferred method of achieving a definitive airway in children following blunt trauma is **orotracheal intubation under direct vision with manual in-line stabilisation of the cervical spine**. Unless the child is deeply unconscious, this will require the use of sedative drugs or general anaesthetic agents and muscle relaxants. Ideally, it should be performed by an experienced anaesthetist. The following sequence should be followed:

1. Cervical spine stabilisation by a dedicated operator, removing the collar to facilitate mouth opening.
2. Pre-oxygenation with 100% oxygen with manual lung inflation if required.
3. Administration of a carefully-judged, reduced dose of an anaesthetic induction agent.
4. Application of cricoid pressure.
5. Suxamethonium 1–2 mg/kg.
6. Intubation with a correctly-sized tracheal tube.
7. Replacement of the collar and blocks after confirming tube placement and relaxing cricoid pressure.

Confirmation of correct placement of the tube

Signs such as chest movement and auscultation remain helpful but are occasionally misleading, especially in inexperienced hands. The most important issue is to see the tube pass through the vocal cords. The correct size is a tube that can be placed easily through the cords with only a small leak. Intubation of the right main bronchus is best avoided by carefully placing the tube only 2–3 cm below the cords and noting the length at the teeth before checking by auscultation (best in the left and right lower axillae). ***Capnography, if available, is a useful adjunct to help confirm correct tube placement***.

If orotracheal intubation is not possible, needle cricothyroidotomy can be used for temporary oxygenation of the small child. Larger children over 11 years of age may be better managed with a surgical cricothyroidotomy. These procedures should also be accompanied by in-line immobilisation if the cervical spine is at risk (see Chapter 6.14).

Breathing

Assessment of adequacy of respiration:

- Rate
- Chest expansion
- Recession
- Use of accessory muscles
- Nasal flaring
- Inspiratory or expiratory noises
- Breath sounds
- Heart rate
- Colour
- Mental state.

Careful examination of the trachea, neck veins and chest may indicate the presence of pleural collections of air or blood. Tension pneumothorax should be treated immediately with needle thoracocentesis in the second intercostal space in the midclavicular line, (see Chapters 6.11 and 6.12) followed by tube thoracostomy. The early use of pulse oximetry is strongly advised. Doubts about airway patency require immediate reassessment and correction by orotracheal intubation. Patients who are intubated should have their respiration controlled manually or by a ventilator, rather than by allowing spontaneous breathing, particularly after head injury. ***Arterial blood gas estimations, if available, will allow oxygenation and control of carbon dioxide to be monitored***.

Circulatory assessment

- Systolic blood pressure
- Capillary refill (should be 2 seconds or less: measure on the sternum)
- Skin colour
- Temperature
- Mental state
- Respiratory rate

The blood pressure is initially well-maintained despite continuing bleeding, due to the child's exceptional ability to vasoconstrict. As an indicator of haemorrhage, it can be falsely reassuring; a tachycardia may be a more revealing feature. Obvious external haemorrhage should be controlled with manual pressure ideally using a sterile dressing. Cannulation of peripheral veins is preferred for volume resuscitation, although this can be very difficult in small children. Other options include:

- Intraosseous infusion (<6 years old) (see Chapters 6.10 and 1.21)
- Femoral vein catheterisation (see Chapter 6.6)
- Venous cutdown (saphenous vein) (see Chapter 6.7)
- Jugular or subclavian vein catheterisation. (see Chapter 6.6)

A useful strategy is to proceed with the method in which the operator is most experienced. Central access should be restricted to those highly skilled in its use, although the femoral route can be cannulated at little risk using the Seldinger approach. Intraosseous infusion into an uninjured extremity, usually the anteromedial aspect of the tibia 1–3 cm below the tubercle, is quick, safe, effective and easy to learn. Peripheral access can often be established once peripheral perfusion has been improved. Both femoral venous and tibial intraosseous access are best avoided if there is clinical evidence of a pelvic or abdominal injury. In such cases, it is better to secure vascular access above the diaphragm.

Blood should be drawn for typing and cross-matching, haemoglobin and full blood count, glucose and electrolytes. Fluid resuscitation should commence with a bolus of 20 ml/kg of crystalloid solution ideally warmed, for example 0.9% saline or Ringer's lactate solution. After repeating this twice, the need for surgical intervention and transfusion of packed red cells should be considered. **The most important aspect of fluid resuscitation is the child's response to the fluid challenge**. Improvement is indicated by:

- Decrease in heart rate
- Increased skin temperature
- Quicker capillary refill
- Improving mental state
- Increased systolic blood pressure
- Satisfactory urine output.

Failure to improve should prompt an urgent search for chest, abdominal, or pelvic haemorrhage, with the immediate involvement of an experienced surgeon.

In small children, it is useful to delegate the initial fluid bolus to a member of the trauma team, who attaches the warmed fluid bag to the intravenous cannula via a three-way tap to which is attached a 20 ml syringe. The child's weight in kilograms is obtained from a parent or estimated from the age or body length. The same number of syringefulls (as the number of kilograms) are serially administered, conveniently delivering the 20 ml/kg aliquot with minimal calculation.

Not all cases of hypovolaemia require aggressive fluid therapy. In adults, withholding fluids in penetrating trunk trauma before achieving surgical haemostasis has been associated with improved outcome. The concept is to avoid pushing up the blood pressure, which hinders clot formation and promotes further bleeding. Aggressive fluid replacement can lead to increased fluid requirements, hypothermia, dilution of clotting factors, excessive blood transfusion and its associated immunosuppression. The principle of hypotensive resuscitation has been extrapolated to children. On the other hand, in severe head injury, cerebral perfusion is critically dependent on maintaining blood pressure. If a child has both a severe head injury and major trunk bleeding, the apparently conflicting requirements are best managed by maintaining priorities in ABCD order and achieving prompt surgical haemostasis. Beyond this strategic conflict, it should be remembered that the normal blood pressure is lower in children, hypovolaemia mimics head injury and blood pressure itself is a poor indicator of organ perfusion.

Disability

A brief neurological assessment is vital for future comparison. Time constraints do not allow a detailed neurological examination during the primary survey. Pupil size and reactivity represent the only lateralising features to seek at this stage. Eye opening, verbal and motor responses should be noted, allowing an estimate of Glasgow Coma Score to be made (see Chapter 3.36). As children aged less than 4 years cannot be judged according to the adult verbal scale, a modified scale has been devised (see Chapter 3.36).

Exposure

The child should be undressed (use scissors to cut away clothes if appropriate) during the primary survey to allow the anatomical search for injuries, which follows in the secondary survey. **Prolonged exposure should be**

strictly avoided. The child's high surface area-to-weight ratio promotes rapid heat loss, predisposing to hypothermia and its attendant complications, such as arrhythmia, platelet dysfunction and a predisposition to infection. A radiant heater is suitable for small children; a warm ambient temperature and a convective air-heating blanket are useful at all ages.

At the end of the primary survey, the severely injured child should have the following:

- Clear airway, breathing 100% oxygen
- Cervical spine immobilisation in blunt trauma cases
- Adequate respiration, achieved by manual or mechanical ventilation and chest decompression when indicated
- Venous access and an initial fluid challenge if indicated on circulatory assessment
- Blood sent for typing and cross-matching
- The potential need for immediate life-saving surgery considered and preparations underway.

The following life-threatening conditions should have been excluded or identified and treated by the end of the primary survey:

- Airway obstruction
- Tension pneumothorax
- Open pneumothorax
- Massive haemothorax
- Flail chest
- Cardiac tamponade

Adjuncts to the primary survey and resuscitation include:

- ECG/oxygen saturation/blood pressure monitoring
- Gastric and urinary catheters
- ***Portable X*** rays of the chest and pelvis

Before the secondary survey begins, it should be remembered that the **ABCD components of the primary survey require constant re-evalulation as deterioration can be rapid**. **Emergency operative treatment to control life-threatening haemorrhage should be performed promptly, without waiting for non-urgent examination and imaging**. Identifying all anatomical injuries remains an important goal but may be overridden by pressing physiological requirements. **Analgesia is frequently forgotten, or mistakenly thought to precipitate cardiorespiratory deterioration**. **The careful titration of intravenous opioids is both safe and humane** (see Chapter 1.27).

Secondary survey

This involves careful examination from head-to-toe in a systematic way, including a **controlled examination of the back, avoiding spinal movement (by log-rolling)**. Clear documentation of all injuries should be noted to serve as the basis of the subsequent management strategy.

Head injury

This remains the commonest cause of death and disability in severe trauma in children and is dealt with in more detail elsewhere (see Chapter 5.5) The scalp and face are examined for bruising, abrasions, lacerations and evidence of fracture. Basal skull fracture is manifest by signs such as racoon eyes, bleeding from the ears or a visible haemotympanum, Battle's sign (bruising over the mastoid process, a relatively late sign) and cerebrospinal fluid leakage from the nose, mouth or ears. The Glasgow Coma Score is again evaluated (Chapter 3.36) allowing a dynamic comparison with the primary survey estimation, unless the child is now intubated and sedated. As infants and small children are prone to hypoglycaemia, it is important to consider this as a potential cause of altered consciousness (see Chapter 3.12).

Chest trauma

The majority of chest injuries result from blunt trauma and are usually associated with injuries in other organ systems. The increased compliance of the chest wall is protective, but can make interpretation of the severity of injury difficult. Rib fractures are uncommon but indicate significant blunt force has been applied. Moreover, serious chest injury can occur without obvious external signs of trauma. The energy that is not dissipated in breaking the elastic ribs may be transferred to the lungs to be manifest as pulmonary contusion. Respiratory failure can occur quickly in infants and young children with chest trauma, yet the majority of chest injuries require no more than the insertion of an intercostal drain.

- Thorough re-examination of the chest front and back, using the classical **inspection–palpation–percussion–auscultation approach**, is combined with a chest *X* ray.
- Particular attention is directed to the symmetry of chest movement and breath sounds, the presence of surgical emphysema and pain or instability on compressing the chest.
- Tracheal deviation and altered heart sounds are noted.
- On log-rolling the child, **it is important to reconsider flail chest** as a posterior floating segment is often poorly tolerated in children.

Pneumothorax

- A tension pneumothorax needs immediate decompression, followed by the insertion of an intercostal drain.
- A simple pneumothorax is a radiological diagnosis and, though not life-threatening, may be associated with significant underlying lung injury. All traumatic pneumothoraces require drainage rather than close observation alone.
- Open pneumothoraces, or sucking chest wounds, allow bidirectional flow of air through a chest wall defect. They are conventionally treated by applying an occlusive dressing, taped on three sides to serve as a flap valve, followed by inserting a chest drain remote from the site of injury. A better dressing is the customised Asherman chest seal, which consists of an adhesive ring, similar to that on a colostomy stoma bag, which projects into a pipe-shaped flap valve, resembling that in a Heimlich valve.

Pulmonary contusion is usually caused by blunt trauma and may occur in association with a flail segment. Treatment

involves supplemental oxygen, careful fluid management and particular attention to pain relief. Endotracheal intubation may be necessary in severe cases.

A traumatic haemothorax may represent a significant proportion of a child's blood volume. Intercostal drainage and appropriate volume replacement is usually all that is required. Myocardial contusion and injuries to the tracheobronchial tree, aorta and other great vessels are rare in children. Treatment follows the same principles as in adults. Diaphragmatic rupture may occur in association with "lap" seat belts and the child may develop respiratory failure due to the excursion of abdominal contents into the chest.

Abdominal trauma

Children are less well protected from solid-organ injury than adults and severe visceral injuries occur more frequently. Unexplained blood loss evident during the primary survey may be due to intra-abdominal haemorrhage.

✔ The **abdomen is a classical silent area after trauma**. **It has to be actively cleared of injury rather than simply noted to be soft and non-tender, especially in the face of altered consciousness**. Cardiovascular decompensation may occur late and precipitously.

- Thorough history-taking and a careful examination of the abdomen may give clues to as the origin of bleeding or perforation.
- Gastric distension may cause respiratory embarrassment and a gastric tube should be placed.
- In order to gain cooperation in a frightened child, the mother's hand may be used to permit palpation.
- In a severely injured child, a urinary catheter should be inserted after gentle rectal examination. This may be omitted in small babies and in less severely injured children. Small boys are particularly prone to urethral stricture after catheterisation. It remains important to exclude haematuria and renal tract injury if the mechanism of injury is of concern (by examining the first urine for red blood cells; see Chapters 3.7 and 6.25).

Abdominal ultrasound and computed tomography scanning

These have become invaluable adjuncts to the secondary survey for not only diagnosing intra-abdominal injury but for monitoring progress when a defined injury is being managed conservatively. Furthermore, bleeding from solid organs may not show up immediately in the resuscitation room and evidence of hollow-organ rupture may take 24 hours or more to show as free fluid on ultrasound. This commits the trauma team to a high index of suspicion well beyond the classical **golden hour**.

Children with refractory shock, penetrating injuries or signs of perforation require laparotomy. Other injuries may be managed conservatively. After initial fluid transfusion, an experienced surgeon may decide that bleeding from an injured spleen, liver or kidney does not require immediate operative intervention. ***Computed tomography (CT) scanning is an invaluable aid in this judgement.***

Splenic rupture is relatively common and can occur after relatively minor trauma, especially if enlarged following an infection or inflammatory process. Signs include left upper quadrant pain and tenderness and referred pain to the shoulder tip. Non-operative management is used frequently in many centres, but long-term problems of splenectomy are insignificant in comparison to the potential consequences of inadequate supervision of conservative management. Increasingly, liver injuries are also being managed conservatively. Unlike the relatively straightforward operation of splenectomy, operative liver repair or resection is hazardous and packing plays a major role in the operative management of uncontrolled hepatic bleeding.

Injuries to the retroperitoneal organs, such as the kidneys or pancreas, may present with vague or atypical signs, again requiring a high index of suspicion. A significant kidney injury does not always cause demonstrable haematuria. Ultrasound studies and ***dynamic contrast CT scans*** provide valuable information on renal structure and function. Intravenous urography remains useful for demonstrating the details of renal and ureteric injury, especially in centres without a CT scanner. Pancreatic injury may occur with a normal amylase level and the amylase may be raised in the absence of pancreatic damage. ***CT*** and ultrasound are sometimes helpful but false-negative reports are frequent.

Spinal trauma (see Chapter 5.7)

Spinal injury must be ruled out in any child who has been subject to a mechanism of injury capable of damaging the spine. This seemingly obvious statement draws attention to the fact that it is often surprisingly difficult to ascertain whether there has been an injury to the spine or not, particularly in the face of a concomitant head injury or in a child who is too young to communicate. Even in an alert older child, distracting pain from a limb injury may lead the patient to ignore and deny neck pain even when a spinal fracture exists. Radiological clearance in children is further complicated by the difficulty of interpreting *X* rays of immature bones and by the relative ligamentous laxity which gives rise to pseudosubluxation. Moreover, spinal cord injury without radiological abnormality (SCIWORA) has been recognised as a problem in children.

Spinal injury is less common in children than in adults, partly because of the elasticity of the bones and ligaments. This same elasticity contributes to the different patterns of spinal injury seen. In the cervical spine, for example, injuries tend to occur at a higher level than in adults and often span several segments rather than dissipating energy in fracturing a single vertebra.

The process of spinal clearance in children is similar to that in adults. The entire spine is palpated during a log-roll, when the patient is turned on to the side in a controlled way, keeping the spine in-line. The presence of palpable steps, bogginess or tenderness is noted. The limbs are examined for sensory and motor signs of focal or segmental deficit. A diaphragmatic breathing pattern, bradycardia, hypotension, peripheral vasodilatation and priapism indicate spinal cord injury. Throughout the primary and secondary survey, spinal precautions are maintained, using a hard collar and side-supports (blocks and straps or sandbags and tape), except for airway procedures

and local examination, when manual in-line immobilisation is reinstituted.

If the child is alert, able to communicate clearly and has no distracting pain from another injury, the spine can be cleared clinically without resorting to *X* rays. Otherwise, spinal precautions are maintained until radiological clearance is achieved and the child re-examined. Three views of the cervical spine are required: cross-table lateral view with arm traction to reveal the C7–T1 interface; anteroposterior view; transoral odontoid peg view. These must be assessed by an experienced traumatologist, orthopaedic surgeon or radiologist, paying particular attention to the soft tissues as well as the bony structures. If the mechanism of injury warrants it, thoracic and lumbar views are also required.

If the lower cervical spine is not adequately visualised on the lateral view, oblique views are requested. If the *X* rays are inadequate or show suspicious areas, ***CT scanning, if available, is recommended to confirm or exclude a fracture. The MRI scan provides a better examination of neural, ligamentous and other soft tissues, though its sensitivity reveals minor as well as major tissue injury, making interpretation more difficult. It remains expensive and is not universally available. It is a frightening environment for an unsedated child and the powerful magnetic field creates challenging logistical problems.***

Limb trauma

Pelvic injury remains a potentially life-threatening injury, especially if it is associated with a large retroperitoneal haematoma or if the fracture site communicates with the rectum. External fixation or embolisation are sometimes required to control major venous or arterial bleeding, respectively. The suitability of these techniques depends on the particular configuration of the fracture and distinguishing retroperitoneal haemorrhage from intraperitoneal bleeding needing a laparotomy can be difficult.

Not all pelvic trauma is serious. Some pubic rami fractures are minor injuries with little intervention required. Nevertheless, the pelvis is a ring structure that tends to break in two places. On inspecting the pelvic *X* ray, careful attention should be paid to the sacroiliac joints and sacral foramina to seek subtle evidence of a second break in the ring.

In general, limb fractures are more likely to be managed conservatively than in adults, reflecting the child's capacity to heal and the risk of interfering with growth plates. An understanding of the Salter–Harris classification of epiphyseal fractures is essential and access to a radiological atlas of developmental stages is helpful (see Chapter 5.6).

Continuing care

Transfer

Not every hospital has the resources and expertise to safely care for injured children. Ideally, children with serious injuries should be transported directly from the scene of the accident to a centre with such capability (if one exists in the country). Even then, geographical constraints may render transfer unsafe. Interhospital transfer requires careful planning to provide: trained medical and nursing escorts; simple, compact, robust equipment; drugs for resuscitation, sedation, pain relief and relaxation; fluids and blood products if indicated; and a suitable vehicle and ambulance staff.

Communication between the referring and the admitting clinicians is necessary not only to agree that transfer is indicated, but to establish guidelines for care in transit and to warn the receiving centre when the patient is expected to arrive. Some paediatric units offer retrieval by a specially-trained team from the receiving hospital as an alternative to direct transfer by the referring team. In trauma care, some transfers are time-limited, for example to evacuate an extradural haematoma. In such cases, the extra time taken for a retrieval team to reach the referring hospital may offset the benefit of their specialised skills. Even when the transfer is urgent, it is essential to try to achieve physiological stability before embarking on a hazardous journey in the isolated environment of the ambulance. In particular, a child with a head injury should not be transferred in a hypotensive condition from unrecognised intraperitoneal bleeding.

Perioperative care

In the operating theatre, definitive anatomical reduction, repair or resection of individual injuries takes place. While the surgical team focuses on anatomical correction, the anaesthetic team maintains physiological system control. The impetus and sense of urgency evident in the Emergency Department should be maintained, without losing the thoroughness necessary to manage all aspects of care.

If the child has a significant head injury, the anaesthetic agents should be chosen to avoid increasing intracranial pressure or cerebral blood flow. In general, this means avoiding high doses of volatile agents such as halothane or isoflurane. Ketamine has long been considered to be contraindicated in head injury, although there is recent evidence that challenges this view. It may be the only anaesthetic available. **If the child is undergoing lengthy extracranial surgery in the face of a severe head injury, it is wise to observe the pupils at frequent intervals and to monitor intracranial pressure intraoperatively, if this facility is available**. ✔

Maintaining the child's core temperature is a key aim during prolonged surgery. Hypothermia impairs platelet function and increases the risk of infection, though it has been claimed to help preserve brain function in severe head injury. The ambient temperature should be adjusted, without sacrificing acceptable operating conditions for the surgical and anaesthetic teams. Convective air heaters, applied to body parts not involved in the operative procedure, have transformed our ability to preserve body heat.

Intensive care

In the immediate management of the injured child, the focus was on physiological assessment and intervention

using an ABCD approach, followed by anatomical assessment and definitive care. When intensive care is instituted, physiological stabilisation again becomes the main concern, though it is important to remain alert to recognise any further injuries not evident in the secondary survey. Detailed physiological control is facilitated by invasive monitoring and one-to-one nursing. The intensive care team assumes an important role in the continuing coordination of multidisciplinary care.

In addition to the respiratory, circulatory and nervous system control, which parallels the earlier ABCD approach, metabolic control and host defence emerge as important aspects of intensive care. Metabolic control is concerned with biochemical processes, including feeding, fluid balance and hepatic, renal and endocrine control. Host defence considers the body as a whole and its interaction with the external environment. Not only is consideration given to wound and nosocomial infection and their prophylaxis, but further injury from physical positioning, temperature disturbances and gastric erosions is to be prevented.

Therapeutic targets may be set in accordance with established guidelines. *For example and if available, in severe head injury**, maintaining a $PaCO_2$ of 4–4.5 kPa by adjusting ventilation and a cerebral perfusion pressure of 70 mmHg using inotropes are widely recommended**.* Titration of drug and fluid therapy to effect remains the cornerstone of intensive care practice.

Step-down care and rehabilitation

High-dependency care, acute ward care and rehabilitation serve to minimise disability, rather than influence mortality, which is already largely determined by this time. The emphasis shifts towards integration back into normal life, physically and psychologically, though the course may be interrupted by further reconstructive surgery. If an unexpected complication occurs, the team managing the child must be alert to recognise when a return to intensive care is indicated.

Summary

Trauma is anatomical structural damage with physiological consequences. Its early management involves a proactive plan to control physiological systems and anatomical structures within a multidisciplinary environment. Though the emphasis changes as the child progresses from the emergency room to the operating theatre or intensive care unit and beyond, the overall aims are the same. Every link in the chain of care is important in optimising the ultimate outcome.

Further reading

Advanced Trauma Life Support Chicago: American College of Surgeons, 1997.

Pre-hospital Paediatric Life Support. Advanced Life Support Group. London: BMJ Books, 2000.

5.5 Head injuries

Jonathan Punt

Minimum standards requirements

- ABCD and maintenance of blood pressure and electrolyte balance
- Emergency burr-hole
- Parenteral antibiotics
- Mannitol and dexamethasone
- Anticonvulsants

Skull fractures

Most skull fractures resolve without treatment but should be observed for 24 hours in case of an intracranial haematoma. Fractures which are compound, either externally (i.e. the overlying scalp is broken) or internally (i.e. there is a fracture into a paranasal sinus or into the middle ear) require attention.

Externally compound fractures

- Like all wounds, should be explored to remove all dead tissue and foreign material. This is the most effective means of preventing infection. Operation should be performed as soon as possible; simple wounds under local anaesthetic, more complex wounds will require general anaesthesia.
- Depressed fractures will require elevation to ensure the full extent of the wound, including the brain substance, has been cleaned and that the dura is repaired if it has been torn. If the wound is less than 24 hours old and not heavily contaminated, then the bone fragments can be replaced. If the wound is older than 24 hours or is heavily contaminated, then it is safer to discard the bone fragments.
- Antibiotics are not generally required, as it is the mechanical debridement of the wound that is the crucial step. However, compound depressed skull fractures that have occurred in an agricultural or rural setting may be contaminated with *Clostridium tetani* and are best covered with five days of IV benzylpenicillin (for 1 month to 12 years 50 mg/kg every 6 hours by slow injection and for >12 years 2.4 g every 6 hours) and antitetanus active immunisation. Animal bites, especially from dogs, will be contaminated with *Pasteurella multocida* and should be covered with ampicillin (IV 40 mg/kg 8 hourly up to maximum of 4 g/day). If surgery is delayed for more than 24 hours, antibiotics should be given.

The most important of all of the brain's coverings is the skin! The scalp has excellent vascularity and every effort should be made to preserve scalp. **Once significant areas are lost, complex skin flaps will be required**. Split-skin grafts will not take on bare calvarial bone. If substantial areas of full-thickness scalp are lost, as in burns or assaults from large animals, a useful technique is to make multiple burr-holes, leaving the dura intact; in the course of a few weeks the florid granulation tissue that grows out of the burr-holes will form a satisfactory base to accept split-skin grafts.

Internally compound fractures

- Carry the danger of cerebrospinal fluid (CSF) fistula and meningitis.
- Prophylactic antibiotics are not indicated.
- Most CSF rhinorrhoea or otorrhoea will resolve spontaneously, but cases persisting for longer than two weeks will require formal repair. This will entail a major craniotomy, preferably preceded by imaging with ***computed tomography (CT) of the skull base with dedicated coronal bone sections***.
- Meningitis complicating traumatic CSF rhinorrhoea or otorrhoea is usually with *Streptococcus pneumoniae* and should be treated for two weeks with intravenous benzylpenicillin (dose above) or cefotaxime (for <12 years 50 mg/kg every 6 hours and for >12 years 1–3 g every 6 hours). **It is an absolute indication for surgical repair to prevent further episodes.**
- Some cases of traumatic CSF rhinorrhoea can be repaired by an ear, nose and throat (ENT) surgeon via an extracranial, endoscopic approach via the ethmoid sinuses.

Penetrating injuries

Children are especially prone to suffering penetrating injuries because of the thin nature of the immature skull, especially around the orbit. **Such wounds require exploration through their full extent to prevent brain abscess**.

Missile injuries require removal of all foreign material wherever feasible. High-velocity penetrating brain injuries from modern military weapons are invariably fatal because of the extreme forces involved, and these patients, along with those who are in deep coma following even low-velocity gunshot wounds, will not make a useful recovery: **only terminal care is appropriate**.

Intracranial haematomas

Only 6 in 1000 patients will develop a significant intracranial haematoma following a non-missile head injury. **The most useful guide to the development of an intracranial haematoma is deterioration in the level of consciousness**. ***The ideal investigation is CT***. If CT is not readily available, then burr-hole exploration is justified in the hope of finding an extradural or subdural clot.

Emergency, temporary reduction of raised intracranial pressure can be achieved by one or more of the following medical measures:

- Mannitol 20% by IV infusion over 20 minutes (250–500 mg/kg). This can be repeated as required
- Dexamethasone by slow IV injection (500 micrograms/kg and then 50 micrograms/kg every 6 hours
- ***Intubation and artificial ventilation to keep $PaCO_2$ around 4 KPa***.

An extradural clot will always be beneath the site of trauma. The place to make the burr-hole is therefore at the site of any external site of injury. This may be known from the history or may be found by shaving the entire scalp in search of bruises, grazes, lacerations or soft-tissue swelling. A plain skull radiograph, if available, may show a fracture, and if so the burr-hole should be made at the site of the fracture. If there are none of the aforementioned clues, then "blind" burr-hole exploration will be required. This should commence on the side of the dilated pupil, or on the side of the pupil that dilated first.

Three standard burr-holes are made: subtemporal, frontal and parietal. It is crucial to make the subtemporal burr-hole low enough in the middle cranial fossa; the correct place is immediately above the zygoma at the midpoint between the outer canthus of the eye and the external auditory meatus. If an extradural clot is found, then the burr-hole must be extended as either a craniectomy or a craniotomy. The margins should extend sufficiently far to uncover the entire clot that can then be evacuated by suction. Bleeding meningeal arteries can be controlled with diathermy or by underrunning with a suture. Bleeding from major venous sinuses can be controlled by haemostatic gauze and by hitching the adjacent dura to the surrounding pericranium with sutures. Diffuse meningeal oozing will stop spontaneously if it is not fiddled with; the application of hydrogen peroxide or warm saline packs may help. When the clot is evacuated and the bleeding is stopped, it is essential to hitch the dura around the perimeter of the bone opening to the adjacent pericranium in order to prevent recurrence. In very young children it may be better to pass sutures through small drill holes in the surrounding bone. If a craniotomy has been made, the bone flap is replaced.

If no extradural haematoma is found at any of the burr-hole sites, then the dura should be opened cautiously. If there is a subdural clot, then a craniotomy is necessary. It is safer to make multiple, short, dural incisions rather than a wide dural opening. It is difficult to be certain whether a tense dura is due to subdural clot or brain swelling. Most acute subdural clots are associated with quite severe brain injury, and a wide dural opening is very likely to be rewarded by massive, uncontrollable fungation of the brain.

Postoperatively, anaesthesia can be reversed unless the patient is to be evacuated to another facility. If a significant clot has been found, then there should be a prompt improvement in the level of consciousness.

In a baby with severe signs of rapidly progressive raised intracranial pressure following a closed head injury, it is reasonable to search for an acute subdural haematoma by passing an adult (18-gauge) lumbar puncture needle into the subdural space through the anterior fontanelle or through a diastased coronal suture. The baby is wrapped in a sheet and held supine by an assistant so as to secure the head, the arms and the trunk. The entry point is either at the most lateral extremity of the anterior fontanelle or at a point in line with the pupil, whichever is the further from the midline. **In a conscious child, local anaesthesia must first be applied**. The needle is passed at a shallow angle, in an anterior direction, through the skin and then through the relatively resistant dura. The obturator is removed from the needle and any subdural fluid allowed to drain spontaneously. The needle is then withdrawn and the puncture hole in the skin closed with a suture.

Major diffuse brain injury

The only measures that are of proven value are maintenance of adequate oxygenation and removal of intracranial haematomas. ***Artificial ventilation, tracheostomy and more sophisticated medical measures designed to control raised intracranial pressure may be of value, but require evacuation to a fully equipped and staffed children's neurointensive care unit***.

In the absence of such a facility, the best strategy is to concentrate on optimising the care of the unconscious patient with attention to:

- preservation of the airway, maintenance of adequate ventilation
- the maintenance of appropriate fluid and electrolyte balance, avoiding hypotonic IV fluids and hyponatraemia
- care of the skin, bladder and bowel
- avoidance of fever $>38°C$.

Fluid restriction is not indicated, but fluid overload should be avoided. Most unconscious children will absorb adequately via the gastrointestinal tract and nasogastric tube feeding will suffice.

If transfer or evacuation is required within the first 48 hours after injury, then endotracheal intubation and mechanical ventilation is desirable. Steroids are of no value and increase the risk of intercurrent infection. Antibiotics are reserved for patients with proven sepsis. Anticonvulsant drugs are only given if there are seizures.

Early traumatic epilepsy

Epileptic seizures in the first 48 hours after injury are common in children. Except in infants they do not, in isolation, indicate the presence of an intracranial haematoma. Most seizures are self-limiting and simply require airway protection. An antiepileptic drug should be given to prevent further fits. It is important to remember that the child with an acutely injured brain will be exquisitely sensitive to the respiratory depressant effects of diazepam or lorazepam. These are best avoided. The safest drug is paraldehyde administered per rectum (0.3 ml/kg up to 1 year, then 1 ml per year of age up to a maximum of 10 ml. Paraldehyde can be diluted with an equal volume of olive oil. It can be given using a plastic syringe if given immediately, otherwise by glass syringe. Do not give if it has a brown colour or smells of acetic acid). A longer-acting drug must also be given at the same time and maintained; the most appropriate are **phenobarbitone** for children aged less than 5 years (load 15 mg/kg slowly IV then a total of 5 mg/kg/day starting dose up to a maximum of 6 mg/kg/day IV or orally in two divided doses 12 hours apart) and **phenytoin** for those aged over 5 years, administered intravenously initially (load 15 mg/kg IV over 20 minutes followed by a further 10 mg/kg IV over a further 20 minutes if first dose is unsuccessful). Then 2.5 mg/kg every 12 hours IV over 20 minutes initially increasing up to a maximum of 7.5 mg/kg every 12 hours (each dose given over 20 minutes if IV). Phenytoin can also be given orally.

5.6 Fractures

Steve Mannion

Minimum standards requirements

- *X* rays and ultrasound
- Splints
- Plaster of Paris bandages
- Traction
- External and internal fixation
- Physiotherapy

Introduction

As any parent knows, all children are susceptible to injury. However, children in disadvantaged countries are probably more at risk than their rich world counterparts as they often live in less regulated and protective environments. Once injured there may be a considerable delay in their presentation to a healthcare facility, a situation that can complicate and restrict treatment options.

Scarce *X* ray resources and a limited range of treatment modalities can then further complicate treatment of paediatric fractures.

However, on a more optimistic note it can be said that paediatric fractures are often more "forgiving" when compared to those of the adult; they are often easier to reduce, less requiring of internal fixation, are quicker to unite and, due to the potential for remodelling with continued skeletal growth greater degrees of malunion can be tolerated.

Diagnosis

Certain features of the history and examination may suggest the presence of a fracture:

- History of a significant traumatic event
- Swelling
- Bruising
- Deformity
- Loss of function: inability to move, weightbear
- Bony crepitus at the fracture site
- Consider the possibility of non-accidental injury (NAI) if the fracture appears inconsistent with the history given (see Chapters 1.30 and 5.2).

Open fractures

Open fractures occur where the fracture site communicates with a laceration or break in the skin relating to it. There is the potential introduction of contaminants and resultant infection. Often they are the result of a greater degree of violence than in closed fractures.

Open fractures are graded according to the Gustilo classification:

I Skin wound of $<$1cm with minimal soft-tissue injury

II Skin wound of $>$1cm, with moderate soft tissue injury

III These wounds typically involve a far greater degree of violence and energy transfer.

- A: extensive wound $>$10 cm with crushed tissue and contamination but for which soft-tissue coverage is usually possible.
- B: extensive wound $>$10 cm, again with crushed tissue and contamination but where it is not thought that local soft-tissue coverage is possible and thus a **regional** or **free** flap may be necessary.
- C: any open fracture with an associated major vascular injury requiring repair for limb salvage.

Treatment is dictated by the extent of soft-tissue injury as reflected in the above scoring system. The initial priority is a thorough debridement and copious irrigation of the fracture site in order to reduce the burden of contamination and reduce the chance of infective sequelae. Once this has been done some form of stabilisation is necessary. Internal fixation of open fractures carries a considerable risk of infection and thus safer options are plaster application (with or without windowing to expose the wound) or external fixation.

Compartment syndrome

The associated soft tissue injury and subsequent swelling leads to an elevation of interstitial pressure within a closed fascial compartment which results in microvascular compromise. Left untreated, tissue necrosis will occur. The commonest site is in the lower leg, but compartment syndromes can also occur in the thigh, foot and upper limb.

The signs and symptoms of compartment syndrome are:

- A hard, woody, swollen extremity
- Severe pain on passive movement
- Loss of distal sensation and movement

- Pain out of proportion to the severity of the fracture and not relieved by splinting or analgesia
- Loss of distal pulses is a late sign.

Whilst it is possible to monitor intracompartment pressures such technology will rarely be available. The alternative is to have a high index of suspicion for fractures involving significant soft-tissue injury and regularly review the clinical condition of the limb.

Treatment of compartment syndrome is by prompt surgical fasciotomies to decompress the affected compartments. In the lower leg are four muscular compartments separated by strong fascia: 1) The lateral compartment containing the peroneal muscles, 2) The anterior compartment containing the dorsiflexor muscles of the ankle and toes, 3) The superficial posterior compartment containing the gastrocnemius and soleus muscles and 4) The deep posterior compartment containing the deep plantar flexors of the ankle and toes. The lateral and anterior compartments can be decompressed through the same anterolateral longitudinal incision. A single posteromedial incision can be used for the deep and superficial posterior compartments. In each case the fascial envelope containing the muscle group must be incised along its length in order to permit swelling and prevent the build up of pressure within the compartment.

X rays

The *X* rays are the most useful and specific diagnostic modality. Where possible two orthogonal *X* rays (at 90° to each other) should be obtained, ideally including the joints above and below the suspected fracture site.

Terms relating to fracture appearance on *X* ray:

- Transverse: fracture line at 90° to long axis of bone
- Oblique: other than the above
- Simple: involving a single fracture line
- Comminuted: involving bony fragmentation
- Greenstick: visible fracture at only one cortex on the *X* ray view. Only seen in paediatric fractures and due to flexible nature of paediatric bone; implies intact periosteum along opposite side to fracture and is a good prognostic sign.

Salter–Harris classification

This relates to the *X* ray pattern of fractures occurring around the epiphysis, or growth plate, of a bone. Such fractures make up about 15–20% of major long bone fractures and 34% of all hand fractures in childhood.

There are five grades with worse prognosis for fracture outcome with increasing grade:

I: Fracture across the epiphyseal line, not extending into the epiphysis or metaphysis. Occurs when the growth plate is very thick and thus tends to be seen in young children. Healing is rapid and complications rare.

II: Fracture across the epiphyseal line extending into the metaphysis, but not into the epiphysis. Usually occurs in children over the age of 10. Healing is usually rapid and there is rarely growth disturbance.

III: Fracture extending from the metaphysis through the growth plate and into the epiphysis. This type of fracture occurs when the growth plate is partially fused and has a poorer prognosis.

IV: Fracture extending completely across the epiphyseal line and into the epiphysis. Can occur at any age and also associated with a poor prognosis.

V: Crush injury to the growth plate. Caused by severe axial loading. Inevitably there is partial destruction of the epiphyseal plate and thus considerable chance of growth disturbance.

Treatment of fractures

- **Reduce** the fracture (if displaced)
- **Hold** the fracture whilst bony healing occurs
- **Rehabilitate**: restore function and range of motion

The potential for remodelling with continued skeletal growth is more marked in younger children. It occurs to a greater degree in the plane of movement of the affected joint. As a result of remodelling, angular deformity can gradually resolve with growth and thus accurate initial reduction is not mandatory. In contrast it is important to accurately reduce intra-articular fractures in order to prevent secondary arthrosis.

Children will often be unable to tolerate reduction under local anaesthesia. General anaesthetic will usually be required (see Chapter 1.13).

Once reduced, the fracture needs to be held in position whilst bony union occurs.

Splintage

Splintage of a fracture involves immobilising the fracture, thereby preventing relative motion of the bone ends. In the acute phase this will help relieve the pain associated with the fracture. In the longer term, the fracture stability conferred by the splint will help promote bony union.

The commonest form of splintage is using plaster of Paris bandages (see plastercraft below). If these are not available preformed then it is possible to make them from crêpe bandages and calcium sulphate. The bandages can be applied in the form of a complete (circumferential) cast or as a backslab, along only one side of the injured limb. **In any situation where swelling is anticipated a complete cast should either be bivalved or split down to skin along its length** ✔. In some circumstances, plaster of Paris may not be available. If this is the case then splints can often be fashioned from locally available materials. One example of this is using strips of bamboo and bandaging.

The splint should be applied with the limb in the position of function ✔. In this way if stiffness does occur the limb will still have some use. For the elbow this position is 90° of flexion, for the ankle a position of neutral plantar/dorsiflexion (the sole of the foot at 90° to the lower leg) is preferred.

Plastercraft

Before starting to apply a plaster, all equipment should be ready to hand. The limb should be covered in stockinette, if available, and then cotton wool. Bony prominences

(ankle malleoli, fibular head, wrist, olecranon) should be covered with extra padding to prevent pressure sores.

The plaster bandage should be immersed in water for about 5 seconds, by which point bubbles should have stopped arising from the plaster. Cold water is usually best, but hot water causes the plaster to set faster, so the temperature should be adjusted according to need.

For plaster slabs, the length required should have been premeasured and then the slab made up in readiness, most slabs requiring a thickness of between 5 and 10 layers of plaster bandage. Once dipped, the slab should be applied to the limb over the layer of cotton wool and then bandaged into place.

For circumferential casts the bandage should be unwound half a turn before dipping, with the roll held in one hand and the free end in the other. After immersion excess water should be allowed to fall from the plaster but it should not be wrung or squeezed as this will result in a plaster that is too dry to make a good cast. The plaster bandage can then be wound around the injured limb with each turn overlapping the previous by about two-thirds. Twists and turns in the plaster should be avoided as these can constrict the limb. Once applied the limb should be held still until the plaster sets.

If proprietary plaster of Paris bandages are not available it is possible to make them using gauze bandages and plaster. Medicinal grade plaster (Calcium sulphate PBC) is ideal, but failing this building plaster can be used. The plaster should be sprinkled onto an unrolled bandage that is **just damp** (in order that the plaster adheres). The bandage can then be rolled up and used in a similar way to a commercially available plaster bandage.

Once set, a useful technique is to write, with broad marker pen, the details of the fracture on the plaster cast as a so-called "fracture passport". The details can include the date of the fracture, the date plaster was applied, the intended date of removal and even a sketch of the fracture configuration itself. This information can be invaluable for subsequent care as notes/*X* rays can often be mislaid or lost.

Traction

An alternative to splintage is traction. By exerting a pull along the axis of the injured bone traction helps to effect reduction and maintain alignment. Traction can either represent a definitive mode of treatment and be maintained until bony union, or temporary, being maintained only until the fracture is stable enough to be treated in a plaster cast.

Several types of traction exist:

- **Skin traction**. Traction is exerted to the limb by means of a bandage (usually adhesive) applied around the limb.
- **Balanced traction**. Where traction of the more distal part of the limb is maintained by reaction against a more proximal structure. The classic example is the Thomas' splint for femoral fractures where the splint is braced against the ipsilateral ischial tuberosity.
- **Skeletal traction**. Where traction is exerted by means of a pin inserted into bone distal to the fracture. An example is a traction pin inserted through the proximal tibia as treatment for a femoral fracture. In the paediatric context, care should be exercised to avoid growth plates when inserting the traction pins.

Traction methods of treatment are most applicable to fractures of the lower limb but there are occasional circumstances where these methods are used in the upper limb. One example is temporary skin traction for a supracondylar fracture of the humerus.

External fixation

Stabilising the fracture by means of an external scaffold which is fixed to the bones proximal and distal to the fracture by means of threaded pins. It is relevant to unstable compound fractures, particularly those with extensive soft-tissue wounding. Several different types of fixator exist. The pins can be sited away from bone growth plates and the fracture reduced prior to the linking bar being tightened. Following application of the fixator, the pin tracks must be cleaned daily with saline in order to prevent the build up of crust, infection of the track and secondary osteomyelitis. The fixator can remain on until bony union or replaced with a plaster cast once the fracture becomes stable enough to tolerate this and/or the soft-tissue wound heals.

A variant of external fixation is percutaneous K-wiring, usually used in conjunction with plaster casting. Particularly relevant to peri-articular fractures this involves the insertion of smooth K-wires across the fracture line in order to prevent secondary displacement. The external ends of the wires should be bent to prevent migration. The wires can be removed once fracture stability permits, typically at 2–3 weeks. In the absence of K-wires, improvisation using long "spinal" type needles is possible.

Internal fixation

The use of screws, plates and other types of metalwork rigidly to hold the reduction of a fracture. Whilst these techniques permit accurate, stable reduction, there is an associated risk of infection of the fixation device. Thus, when considering this form of treatment, the following should be borne in mind:

- The fracture should warrant internal fixation as opposed to splintage, traction or external fixation
- There should be an adequate supply of the metalwork in a full range of sizes and the required instruments for their insertion. For a sustainable fracture treatment philosophy **resupply of fixation devices also needs to be available**
- The fixation devices and tools should be sterile
- The level of asepsis in the operating theatre must be high
- The surgeon should be trained in the application of the device and in the surgical approach necessary for it
- In some cases intraoperative *X* ray guidance (fluoroscopy) is necessary for accurate fixation
- Intramedullary methods of fixation, popular in rich countries for the fixation of adult long bone fractures,

are rarely appropriate to paediatric cases as they violate the epiphyseal plates potentially resulting in growth disturbance.

The decision to use this method of fixation will be based on a risk-benefit analysis with consideration given to the fracture configuration, age of the child, operative resources and training of the surgeon involved.

On-going fracture care

Once reduction and stabilisation of the fracture has occurred, on-going care is required to monitor the progress of the fracture to union. The treating physician should document the treatment provided and estimate the duration of immobilisation needed. Where the provision of notes and *X* rays is limited, one suggestion is to draw the fracture on the surface of the plaster cast along with the intended date of removal.

At initial follow up, the quality of the plaster cast should be inspected and *X* rays taken (where possible) to ensure secondary displacement has not taken place.

Overall duration of immobilisation needed is dependent upon the age of the patient and the fracture configuration. Determination of bony union involves the removal of the plaster cast or external fixation device (after an appropriate period by which union would have been predicted to occur) and the **gentle** stressing of the fracture site. The presence of persistent tenderness, swelling or abnormal movement are all indicative that union has yet to occur. The extent of fracture callus on *X* ray is also indicative of the state of union.

Rehabilitation

Children rarely need dedicated physical therapy following fracture healing. They should be encouraged to move their joints through a full range of motion and exercises should be prescribed to restore muscle bulk.

Specific fractures

Femoral shaft

Closed femoral shaft fractures in children are usually best treated with traction, with the type dependent on the age of the child. Typically the duration required is one week per year of age, but this can be shortened by transfer into a plaster hip spica once fracture stability permits.

- Age 0–2 with weight under 12 kg: Gallows traction, thighs in 45° flexion and hips 30° abduction. Limb length inequality seldom a problem as fracture does not shorten excessively. Shortening of up to 1.5 cm and angulation of up to 30° is acceptable. Early spica casting is often possible. **This fracture is associated with non-accidental injury in 50–80% of cases**.
- Age 2–10: Skin traction, either in the 90/90 position (hip and knee flexed to 90°) or Perkins type (straight traction). Alternatively, especially in the older members of this group, skeletal traction through a distal femoral traction pin again either in the 90/90 position or straight. Up to 2 cm of bayonet shortening can be tolerated with no adverse effects. Early spica casting can be used if position is acceptable. With skin traction weight used should not exceed 5 kg, but with skeletal traction up to 10% of body weight can be applied.
- Age 10–15: Skeletal traction, either in 90/90 position or straight. Much greater risk of shortening in this group and less potential for subsequent growth acceleration and length equalisation. Over the age of 15, can be treated as per adults.

Tibial shaft

Closed tibial shaft fractures in children are usually uncomplicated and can be treated satisfactorily with closed reduction and long leg cast application.

- The cast should be applied with the knee in 5–10° of flexion.
- In comparison to the femur there is less potential for overgrowth and thus it is important to maintain the fracture out to length that is, ensure that the length of the fractured limb is the same as the uninjured side. Acceptable degrees of shortening are 5–10 mm in the 0–5 years old age group and 0–5 mm in the 5–10 year-old age group.
- Acceptable axial alignment is less than 10° of recurvatum (where the apex of the fracture site points posteriorly) and less than 5° of varus or valgus angulation.
- As union progresses it may be possible to convert the long leg cast to a patellar tendon bearing cast after 3 weeks.
- Undisplaced fractures in children aged 1–5 can often be treated in below-knee casts or even below-knee plaster cylinders.

Distal humeral

Supracondylar fractures of the humerus have the highest rate of complications and some of the poorest results of treatment of all paediatric fractures. The peak incidence is at the age of 6–7 years.

- The vast majority (98%) are extension type, featuring a posteriorly displaced distal part. Only 2% are flexion type, resulting from a fall onto the point of the elbow.
- A careful assessment of distal vascularity should be made. In fractures with posterolateral displacement, the medial humeral spike can tether the brachial artery.
- If distal pulses are absent, then closed reduction should be attempted. If this fails to restore pulses, then immediate open reduction and surgical exploration of the brachial artery should be performed.
- In other displaced fractures closed reduction should be attempted, possibly combined with percutaneous pin fixation for unstable fractures.
- Acceptable reductions will have no more than 4° of varus as determined by Baumann's angle on the anteroposterior radiograph. Additionally, the axis of the capitellum should be at 45° to the humeral shaft.

- If an acceptable position is not obtained, then this may be an indication for open reduction and percutaneous K-wire fixation.
- Alternatively, the limb can be placed on traction in extension. As swelling subsides so it will become easier to effect a closed reduction (with or without K-wiring).
- Once reduced, an above-elbow plaster backslab should be applied with the elbow flexed. Flexion above 90° will assist in maintaining the reduction of extension-type fractures, but care should be exercised to ensure that distal pulses are maintained.
- Ideally check *X* rays should be performed on a weekly basis to ensure reduction is maintained. The plaster cast can be completed once swelling has resolved and percutaneous wires can be removed after three weeks.
- Typical duration of immobilisation necessary for union is four to five weeks in the 0–5-year age group and six to seven weeks in the 5–10-year-old age group.

Forearm fractures

- Both bone paediatric forearm fractures typically result from the indirect violence of a fall on an outstretched hand. They may be greenstick or complete. If the periosteal sleeve is disrupted the fractures may be unstable.
- *X* rays should include the wrist and elbow as the integrity of the proximal and distal radioulnar joints needs to be determined.
- In contrast to adult forearm fractures, the majority of these injuries can be treated by closed reduction and plaster immobilisation.
- Up to the age of 9, acceptable reduction can be defined as anything less than complete displacement, 15° of displacement and 45° of malrotation.
- Beyond 9 years at least bayonet apposition is required with less than 30° of malrotation, less than 10° angulation if the fracture is proximal or less than 15° if it is distal.
- Immediately following fracture union, there may be a cosmetic deformity if the above reduction criteria are utilised, but this deformity should remodel if there is over two years of skeletal growth remaining.
- Following reduction, an assessment of forearm supination and pronation should be performed to ensure there is no block.
- The arm should be immobilised in an above-elbow cast with the elbow flexed to 90°. Opinion varies as to the position of the wrist in the cast, some surgeons placing the wrist in neutral supination/pronation for all fractures, others placing it in supination for proximal third fractures, neutral for middle third and pronation for distal third.
- Follow up *X* rays should be taken at one- and two-week intervals following manipulation to ensure that secondary displacement has not occurred. If displacement does occur, remanipulation can be attempted.
- Some very unstable fractures may prove difficult to treat by closed methods. These may benefit from intramedullary pinning (Rush pins) or cross K-wiring if facilities exist for this (***intraoperative fluoroscopy is required***).

Distal radial ("wrist") fractures

- Children's distal radius fractures are usually the result of a fall on the outstretched hand and are rarely intra-articular.
- Common types are:
  - Galleazi fracture (isolated fracture of the distal radius, implies associated disruption of the distal radioulnar joint)
  - Physeal (pattern of injury described by the Salter–Harris classification)
  - Torus (buckling of the cortex on the compression side of the fracture without angulation)
  - Greenstick (incomplete fracture)
- In children these fractures can almost always be treated with closed reduction and plaster immobilisation.
- The reduction manoeuvre is to hyperextend the wrist followed by traction and "hingeing" of the distal fragment over the fracture site.
- Acceptable reduction can be defined as anything less than complete displacement and slight angulation. As in forearm fractures, cosmetic deformity should remodel if more than two years of skeletal growth is remaining.
- Check *X* rays should be performed at one and two weeks postreduction to exclude secondary displacement.
- Duration of immobilisation required depends upon the fracture configuration and age of the child, but typically three to five weeks is needed.

Conclusions

- Most paediatric fractures can be treated by closed methods.
- Very often the periosteal sleeve will be intact leading to enhanced fracture stability.
- Completely accurate reduction is not always necessary as children's bones have the potential to remodel with continued skeletal growth.

5.7 Spinal cord injuries

Waghi El Masri

Minimum standards requirements

- ABCD (see Chapter 5.4)
- Cervical collars
- Dexamethasone
- Physiotherapy
- Jewitt brace
- Urinary catheters
- Glyceryl trinitrate
- Laxatives
- Furosemide

Mechanism

- The following predispose to spinal injuries: achondroplasia, Klippel–Fiel syndrome, Down's syndrome and juvenile rheumatoid arthritis.
- Injuries can occur during birth and from abuse.
- Most occur in road traffic accidents, sports and from falls.
- Non-traumatic causes include transverse myelitis; for example epidural abscess, tuberculosis of the spine, neuroblastoma, astrocytoma, eosinophilic granulomata, lipoma, teratoma and aneurysmal bone cysts.

Diagnosis

- In the conscious patient, localised tenderness in the spine, impairment/loss of sensation, of voluntary motor power and of reflexes can help determine the level of vertebral involvement.
- In the semiconscious or unconscious patient, hypotension associated with bradycardia, dilated peripheral veins in the lower limbs, paradoxical respiration, lack of spontaneous movement of limbs, lack of response to painful stimuli applied by pressure over bony prominences at various levels, and urinary retention are all signs suggestive of a spinal cord injury.
- 10–20% of injuries are in more than one site, therefore *X* ray of whole spine is necessary.
- Associated other injuries are common and loss of sensation may delay their diagnosis.

Neurological deterioration

- From further mechanical damage and/or further non-mechanical damage to neural tissue during treatment.
- From hypoxia, hypotension and sepsis.

Management of spinal cord injuries

- Prevent complications related to multisystem dysfunction.
- To contain the "biomechanical instability" of the spinal column by preventing movement at the site of the fracture.
- Dexamethasone should not be given routinely to children with spinal injuries as there is no evidence that steroids improve the neurological outcome.
- Dexamethasone should only be considered if there are signs of neurological deterioration following acute spinal injuries. The recommended dose is 500 micrograms/kg immediately followed by 50 micrograms/kg every six hours for 48 hours.
- "Rehabilitation" should begin as soon as possible.
- Early counselling and psychological support to the child, parents and family members.
- Commencement of physiotherapeutic procedures to prevent contractures of paralysed muscles, chest infections and pressure sores.
- Train all systems of the body to function as safely and as near normal as possible.
- Psychosocial and physical reintegration of the child in the community without losing significantly on education.
- Teaching programme for the child and/or parents aimed at minimising the development of complications (medical, physical and psychological) in the medium and long term.
- Offer life-long regular annual or biannual assessment and treatment if necessary to maintain health and habilitation.

Acute spinal injury

- Keep spine in a neutral position (with pillow arrangement).

- For cervical spine, immobilise with cervical collar or sandbags at the side of the head for about six weeks, followed by bracing for six to eight weeks.
- In children below 6 years, the sagittal diameter of the skull exceeds that of the chest forcing the neck into flexion. A cut-out in the board or the mattress to recess the occiput should be made.
- Children with intact neurology can be adequately braced in a Minerva cast for cervical spine injuries and a body cast for thoracolumbar injuries.
- Minerva and body casts should not be applied to children with sensory loss because of the risk of pressure sores.
- A hard cervical collar for the tetraplegic and a Jewett brace for the paraplegic child are likely to provide adequate support until healing occurs.

Temperature control

The patient may not be able to control temperature, becoming pyrexial in a hot environment or hypothermic in a cold environment.

Cardiovascular and peripheral vascular system problems

- Spinal shock (autonomic areflexia) may cause bradycardia with hypotension.
- Care with intravenous hydration as circulatory overload and pulmonary oedema can easily occur.
- Hypoxia, hypothermia and tracheal suction can aggravate the bradycardia.
- Postural hypotension is most profound during the state of spinal areflexia. Early mobilisation can result in significant drop in blood pressure which may affect spinal cord blood flow and adversely affect neurological recovery.
- Following return of autonomic reflex activity, patients with cord lesions above T6 can develop autonomic dysreflexia: sudden onset of pounding headaches, flushing, blotchiness of the skin above the level of the injury, conjunctival congestion associated with sweating and high blood pressure. The commonest causes are urinary retention and constipation. Treat by placing patient in the upright position (usually sitting) and if >12 years the administration of sublingual glyceryl trinitrate (300 micrograms). If urinary retention is the cause, catheterisation following the liberal instillation of urethral lubricant with local anaesthetic will rapidly reduce the blood pressure and relieve the symptoms.

Hypercalcaemia

- Occurs in 10–20% of patients, especially in tetraplegia and complete spinal cord injuries. Onset is insiduous in the first few weeks following injury. Nausea, anorexia and vomiting, can mimic an acute abdomen. Polydipsia, polyuria, dehydration, lethargy and occasionally psychosis can occur.
- Adequate hydration and furosemide are the first line of treatment.

Respiratory system

- Children with injuries above C4 are unlikely to be able to breathe spontaneously.
- Children with lesions below C4 (most children with activity in the biceps) are able to breathe independently using their diaphragm, provided no major chest injury is present.
- Encourage deep-breathing exercises, postural drainage, assist coughing and monitor oxygen saturation if possible.

Gastrointestinal system

- All patients are at risk of developing paralytic ileus.
- The resulting abdominal distension can embarrass the diaphragm.
- Avoid oral intake in the first 48–72 hours from injury and until bowel sounds are audible.
- Risk of gastrointestinal bleeding from stress ulcers is high: therefore administer H_2-blockers or antacids for the first 3–4 weeks following injury (see Chapter 3.29).
- Risk of constipation is high causing haemorrhoids, anal fissures and mucosal tears. Consider regular senna tablets (7.5 mg sennoside) or liquid (7.5 mg sennoside in 5 ml) for children <6 years, 2.5–5 ml at night and for children >6 years, 5–10 ml or 1–2 tablets at night.
- A regular bowel regime consisting of glycerine suppositories (infant 1 g, child <12 years 2 g and child >12 years 4 g 15 minutes prior to digital evacuation of the bowel and at fixed and regular intervals not exceeding 48 hours, should be instituted initially by a nurse or parent; later by the child.

Locomotor system

- High risk of contractures of muscles, limitation of the range of movement in the joints of the paralysed limbs, excess spasticity and fractures of long bones which are preventable.
- Passive movements and good positioning in bed and early splinting (if necessary), should prevent contractures.

Urinary system

- Urinary retention occurs during the stage of spinal areflexia and is usually permanent in children with lower motor neurone lesions.
- Reflex micturition gradually develops in children with upper motor neurone lesions, usually from the sixth week onwards.

- Extra fluid intake should be encouraged.
- Up to the age of 2–3 years, 4 hourly gentle suprapubic pressure will empty the bladder.
- Children beyond the age of 3 years are best managed with intermittent catheterisation until effective reflex micturition occurs and the residual urine is consistently below 60 ml.
- Children with lower motor neurone lesions are likely to require intermittent catheterisation for the rest of their lives. Initially this should be done by an attendant. With teaching and training however, a child with good hand function can learn to do clean self-intermittent catheterisation. Intermittent catheterisation is the safest method of bladder drainage.
- Indwelling urethral catheters are not recommended beyond the first 48–72 hours.
- Antibiotics should be reserved for urinary infections with systemic manifestations.

Skin

- Sensory impairment loss, impairment of vasomotor regulation of skin blood flow associated with paralysis, double incontinence, possibly anaemia and urinary infections; all render the skin of patients with spinal cord injuries vulnerable to breakdown and infections.
- Skin breakdown is preventable.
- In the acute stage, regular log-turning of the child together with adequate management of the bladder and bowels and vigilance in maintaining cleanliness will prevent skin breakdown.
- In the rehabilitation stage, training of the patient in self-care, hygiene and adequate seating can all assist.
- Latex allergy can develop.

Sexuality and fertility

- Discuss situation with the child in early adolescence.
- Advice about contraception is necessary for girls since fertility is not affected regardless of the level and density of the spinal cord injury.
- Boys with upper motor neurone lesions will have reflexogenic but not psychogenic erections. Male fertility is significantly affected.

Psychosocial integration, education, vocational training, employment

- Continuing education, vocational training and employment must be pursued as the child grows.

Further reading

Betz RR, Mulcahey MJ eds. *The Child with a Spinal Cord Injury*. Rosemont, Illinois: American Academy of Orthopaedic Surgeons 1996.

Grundy D, Swain A. *ABC of Spinal Cord Injuries*. 4th ed. London: BMJ Books, 2002.

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5.8 Landmine injuries in children

Eddie Chaloner

Minimum standards requirements

- ABCDE (see Chapter 5.4)
- Shock (see Chapter 3.6)
- Analgesia (see Chapter 1.27)
- Antitetanus immunisation and immunoglobulin
- Penicillin
- Prostheses

Patterns

- Caused by stepping onto a buried blast mine: traumatic amputation of the detonating limb with fragment and minor blast damage to the other leg (most common injury).
- Caused by fragmentation landmine: widespread fragment injury to limbs and trunk.
- Caused by close-proximity detonation of a landmine in the hand or close to the face. Results in amputation of the hand/arm, plus damage to the face, eyes and head. Usually occurs in mine clearers or in those handling weapons.

Some mines are scattered from aircraft or by shells to lay on the surface of the ground. These weapons are unstable and likely to explode when handled. Unexploded ordnance, such as grenades, can also explode if handled resulting in the same pattern of injury.

Specific problems in children

- Sustain a greater level of injury per gram of explosive than adults because of their smaller body mass (a small antipersonnel mine of approx. 30 g which would normally require a below-knee amputation in an adult, may result in an above-knee amputation in a child).
- Susceptible to close-proximity detonation injuries, because of their tendencies to pick up and play with objects they find.

Treatment

- Initial surgical management follows the basic principles of resuscitation.
- In injury caused by stepping onto a buried blast mine, airway maintenance is not usually a problem, as the child is frequently conscious.
- As with all injured children, fear and bewilderment from pain and unfamiliar surroundings can be distressing for the child and for medical attendants.
- In close-proximity detonation injury, airway maintenance can be a problem. The patient is often unconscious and there may be damage to the upper airway from the blast. A tracheostomy may be required. Benzylpenicillin and antitetanus toxoid should be administered.
- Anaesthesia can be achieved using a ketamine infusion (see Chapter 1.13).

Injury from stepping onto a buried blast mine: technique of amputation

- On the operating table, a thorough wash with warm, clean water and a scrubbing brush will get rid of the gross contamination and general soiling of the limbs prior to formal skin preparation.
- ALWAYS use an above-knee orthopaedic tourniquet to minimise perioperative blood loss, which is proportionally greater in children than adults.
- Perform a standard amputation according to International Committee of the Red Cross surgical guidelines. Remember the following points:
  - The muscles are usually contused more proximally by blast damage than may be initially apparent.
  - Dirt and contamination can be propelled up tissue planes by the blast. An amputation through the blast damage can leave contamination in the wound.
  - Make a bulky myoplasty to cover the bone end using the medial gastrocnemius below the knee, or medial vastus above knee. Leave generous skin flaps as the muscle in the stump will swell considerably postoperatively.

- Make an anterior bevel to the bone when dividing it and file the edges down.
- Let the tourniquet down when the amputation is completed to check haemostasis before applying the dressing.
- Perform thorough wound toilet of the injuries to the other leg. Explore all wounds and excise contaminated tissue. Leave these wounds open to be closed or skin grafted at five days postoperatively.
- NEVER close the amputation stump primarily. Lightly pack the open stump with gauze and apply a bulky dressing. Write on the dressing the date for wound inspection (usually at five days postoperatively).
- Do not take the dressing down on the ward unless the patient manifests signs of systemic toxicity (fever, tachycardia, foul-smelling dressing).
- Give blood only if haemoglobin falls to less than 8 g/dl.
- Give IV benzylpenicillin for 48 hours (50 mg/kg 6 hourly), then orally for a further three days (12.5 mg/kg 4 times daily).
- Inspect the wound at five days. If the tissue is healthy and not infected, close with interrupted non-absorbable sutures over a drain. Leave sutures in for three weeks.
- Early physiotherapy is crucial to success, especially to eliminate flexion contracture of the below-knee amputation.
- Refer early to a prosthetic workshop for casting. Children will need several sets of prostheses as they grow.

5.9 Gunshot wounds

Steve Mannion

Minimum standards requirements

- ABCDE (see Chapter 5.4)
- Shock (see Chapter 3.6)
- Analgesia (see Chapter 1.27)
- Antitetanus immunisation and immunoglobulin
- Penicillin
- *X* rays and ultrasound
- Intensive care (see Chapter 1.25)

Introduction

Although the end of the Cold War led to a reduction of the risk of conflict in Europe, numerous conflicts continue to rage in the disadvantaged world. Many of these conflicts are between ill-disciplined or irregular armies who often specifically target civilian populations in defiance of the Geneva Conventions. In this process, children are inevitably susceptible to sustaining gunshot wounds.

The International Committee of the Red Cross has drawn attention to the global proliferation of weapons. For example, there are estimated to be as many as 125 million AK47 assault rifles in circulation worldwide. As conflicts resolve, these weapons become marketable commodities and spread to neighbouring states where they become the criminal's weapon of choice. The net result of this is injury to the civilian population including children.

Ballistics

The science of ballistics addresses aspects of missile and bullet flight and relates these to the potential for injury. The issues relevant to the mechanism of wounding are:

- When a bullet impacts tissue it will impart some of its kinetic energy into it.
- This will cause the tissue to accelerate away from the track of the projectile resulting in a **temporary cavity**.
- Once the bullet has passed, the inherent elasticity of the tissues will cause the temporary cavity to collapse, leaving some degree of **permanent cavity** along the track.

The extent to which cavitation occurs is governed by the amount of kinetic energy imparted to the tissues by the projectile. The equation governing this is

$$\text{Kinetic energy} = \tfrac{1}{2}\, m\, (V_1^2 - V_2^2)$$

where m is the mass of the projectile, V_1 the velocity on entering the tissues and V_2 the velocity on exiting.

The degree to which the projectile's velocity is attenuated in transiting the tissues is dependent upon the diameter of the bullet, its orientation and flight characteristics on impact, and the nature of the tissue itself.

Categories of gunshot wounds

In practice the masses of most commonly used bullets are similar and thus the velocity of the projectile largely defines the injury potential. In this regard, gunshot wounds can largely be broken down into three categories depending on the nature of the weapon used.

Handguns

- The commonest types of these weapons feature a bullet with a diameter of 9 mm and a muzzle velocity of around 1000 ft per second.
- Only a small temporary cavity is formed and the injury is essentially confined to the bullet track.
- Provided the bullet has not transected any major structures the degree of injury may only be slight.
- Some of the bullets for these types of weapon are designed to deform on impact. These are the hollow or soft- (lead) tipped bullets. On impact they tend to flatten, presenting a greater surface area to the direction of travel, thus resulting in an increased transfer of energy and greater wounding effect.

Shotguns

- The cartridge contains multiple pellets of a specified diameter.
- This diameter can be from 1 mm ("birdshot") to 10 mm ("buckshot").
- Once fired, the pellets disperse in a cone-shaped pattern.
- The degree and rapidity of dispersion is proportional to the size and number of pellets as well as the diameter of the shotgun barrel at the muzzle.

- Due to their aerodynamics, the velocity of individual pellets will attenuate over short distances even in air. Furthermore, the conical dispersion leads to a rapid decline in the number of pellets which will hit a particular target as range increases.
- The above factors lead to the weapon being virtually ineffective at ranges over 50 metres.
- A severe pattern of injury is seen at close range. Although each pellet may only be travelling at low ballistic velocity, the combined effect of multiple pellets is a formidable destructive force, shredding the tissues and causing massive disruption.

Military assault rifles

- These weapons typically have a bullet of 7.62 mm diameter that leaves the weapon at a speed of around 3000 feet per second.
- Rifling of the barrel sets the bullet spinning, which, combined with the increased velocity, leads to greater accuracy at long range.
- Rather than follow a uniform flight path, the bullet has a periodic motion, oscillating around its flight axis with the movements of precession, nutation and yaw.
- The very much greater kinetic energy of these bullets leads to a much bigger temporary cavity than seen in low-velocity munitions.
- The subatmospheric pressure in the cavity will tend to suck in clothing and other debris from outside the wound causing contamination.
- The shock front of accelerating tissue, propagating away from the point of impact, causes stretching and tearing of the tissues, cellular disruption and microvascular injury.
- The margin of tissue around the cavity, termed the "zone of extravasation", is full of haemorrhage, has little tendency to further bleeding and, if muscle, shows no tendency to contract when stimulated. This tissue is non-viable and will become a culture medium for infection if left in place.
- The shock wave itself can cause fracture of bone and intimal disruption of major vessels.
- The oscillating nature of the bullet trajectory can cause it to "tumble" on impacting with the tissues. When this occurs, due to the non-uniform motion, even greater proportions of the kinetic energy are transmitted. The resulting tissue acceleration can lead to the exit wound of such a bullet being very much larger than the entry.
- The nature of the tissue being transited has a great impact on the extent of damage occurring. Relatively elastic, compressible tissue such as lung propagates the shock wave to a much lesser extent than dense, fluid-filled tissue such as liver. Hence a high-velocity bullet may transit lung causing only contusion, whilst transiting solid organs causes gross disruption.

Treatment

Although it is clearly impossible to cover the treatment of gunshot wounds to every possible anatomical structure in the body, there are some themes common to all such injuries.

Most of the wounds encountered will be to the limbs as gunshots to the head, chest and abdomen have a high rate of on-scene mortality.

Protocols for treating gunshot wounds have been adopted and publicised by the International Committee of the Red Cross (ICRC) who have extensive experience of treating such injuries as part of their war surgery programmes.

Initial measures

The initial measures in the treatment of gunshot wounds are similar to those for any severe injury.

- General assessment and resuscitation of the patient, addressing potentially life-threatening conditions according to ABC priorities (airway, breathing, stopping haemorrhage).
- Application of dressings to the open wounds.
- Emergency splintage of fractures.
- Obtaining intravenous access.
- The degree to which fluid resuscitation should be carried out is controversial. ATLS teaching recommends an initial bolus of 20 ml/kg, after which the child should be carefully monitored with respect to the adequacy of organ perfusion and the response to this initial fluid challenge.
- Analgesia as required.
- Antibiotics – the ICRC recommend intravenous benzylpenicillin at a dose appropriate to the size of the child (usually 50 mg/kg IV 6 hourly).
- Tetanus toxoid and antitetanus serum.
- Appropriate radiographs of the injured areas.

Wound assessment

Before proceeding to surgical treatment, certain aspects of the wound need to be assessed.

- From the history, the nature of the weapon used (if known).
- Site of the entrance wound (and exit, if present).
- Sizes of entrance and exit wounds.
- Cavity formation.
- The anatomical structures which may have been transited.
- Distal perfusion.
- Presence of fractures.
- Degree of contamination.

Wound excision

This involves removal from the wound of any dead and contaminated tissue which if left would become a medium for infection. It is most relevant to high-energy transfer (high-velocity) wounds which feature large cavities and considerable amounts of dead tissue and contamination.

- Wound excision should be a planned procedure with prior consideration given to the position of the patient and type of anaesthesia required.
- For limb wounds a pneumatic tourniquet should be used where possible to reduce blood loss.

- Skin incision ("debridement") decompresses the wound and allows swelling of the tissues without constriction.
- Where possible, the incisions should be longitudinal and not cross joints.
- Skin is a resilient tissue, only minimal excision is usually necessary.
- Dead and contaminated tissue should be excised.
- Dead muscle is dusky in colour, shows little tendency to bleed and does not contract to forcep pressure.
- Foreign material should be excised from the wound. However, the obsessive pursuit of small metallic debris, such as that from a disintegrating bullet or shotgun pellets, is not worthwhile.
- Bone fragments denuded of soft-tissue attachment (muscle or periosteum) should be removed as, left in the wound, they will become infected and form osteomyelitic sequestrae.
- There should be no primary repair of nerve or tendon. Where obviously divided these structures should be marked (with suture) for later repair.
- At the end of the procedure the excised wound should be washed with copious quantities of saline and then a dry, bulky, sterile dressing applied.
- Some low-energy-transfer (velocity) wounds such as those from most handguns, because of the minimal cavitation and zone of extravasation do not need the extensive debridement and excision outlined above. These wounds can, in certain circumstances, be managed without surgery.

Delayed primary closure

Once wound excision has been undertaken the patient can be returned to the ward and the following regime followed:

- Continued analgesia
- Benzylpenicillin; IV 50 mg/kg every 4 hours for the first 24 hours and then orally for a further four days (12.5 mg/kg 4 times daily)
- Monitoring of the patient for signs of sepsis
- The dressing should be left in place on the ward and only removed when the patient returns to theatre after an interval period for **delayed primary closure**
- The ICRC recommend an interval period of five days, but latest practice tends towards shorter periods of 48–72 hours
- The only indication for return to theatre and dressing removal before this interval period has elapsed is an offensive dressing combined with signs of patient sepsis. The most common cause of this situation is an inadequate initial wound excision.

In the process of delayed primary closure:

- The dressing should be removed in theatre under appropriate anaesthesia
- If clean, the wound can be closed, or, if skin cover is deficient, split-skin grafted
- If there is evidence of infection, further debridement/excision can be undertaken and the process repeated, aiming for delayed closure after a further five days
- Following closure, rehabilitation of the injured part can commence.

Specific features relating to certain anatomical sites

- Wounds of the head and neck, by virtue of the enhanced vascular supply to these areas, can safely be closed or reconstructed at the initial operation.
- Wounds to major vessels need to be reconstructed primarily.
- Breeches of the dura, pleura and peritoneum should, where possible, be closed at initial surgery.
- Most gunshot wounds to the chest can be treated with tube thoracostomy alone.
- Penetration within 5 cm of the midline of the thorax or abdomen is associated with a risk of injury to the great vessels or heart.
- Gunshot wounds to the head that transit the cranial cavity carry a very poor prognosis, especially if from a high-energy transfer weapon.
- Penetrating gunshot wounds of the abdomen are associated with a >85% chance of bowel or major organ transit. Exploratory laparotomy is, therefore, virtually mandatory.

Conclusion

Gunshot wounds from whatever type of weapon represent a severe injury. Some understanding of ballistics can help in the assessment of these injuries and treatment according to basic principles, such as those recommended by the International Committee of the Red Cross, can lead to a satisfactory outcome even with limited clinical resources.

510 Ingestion burns

Anthony Roberts

Minimum standards requirements

- ABCD (see Chapter 1.19)
- Buffered phosphate
- Nasogastric feeding
- Intubation and tracheostomy (see Chapter 6.15)

Types of ingestion burns

Burns from hot fluids

- Burns from drinking hot fluids are relatively rare in developmentally normal children but can occur in those with learning difficulties.
- Normally only the mouth is burned.
- Swelling and blistering can be very rapid and require an oral or nasal (preferred) airway.
- Swelling has usually gone down within 48 hours and the necessity for further treatment is unusual.

Caustic fluids

- Burns from drinking caustic fluids are much more severe.
- In general, caustic alkali solutions are more dangerous than acids.

First aid

- The single most important aspect of first aid is dilution with water.
- DO NOT MAKE the child vomit as burning fluid causes further damage when passing up the oesophagus.
- Do consider passing a wide-bore nasogastric tube and remove the stomach contents AFTER further dilution or neutralisation.
- Specific neutralisation procedures are valuable, particular with alkalis which continue to burn.
- Dilute vinegar, 10:1, can be used for alkali burns.
- Baking soda can be used for acid burns, but remember that a large volume of carbon dioxide is produced and must be removed.
- Milk can also be effective.
- Buffered phosphate (in one litre the contents are disodium hydrogen phosphate dodecahydrate 180 g and potassium dihydrogen phosphate anhydrous 70 g) should be kept made up and used for any chemical burn: ingestion, skin or eyes.

Treatment

- Swelling of the soft palate may require an airway or intubation.
- Feeding with milk can be helpful orally or through a nasogastric tube.

Complications

- Serious burning, particularly of the oesophagus, can lead to perforation, and in later stages to strictures.
- Acute perforation of the oesophagus is frequently fatal; treat either by drip and suction or thoracotomy.
- Late stricture, during and after the healing phase, is a very common problem after ingestion of caustic fluids.
- Mild cases can be treated by dilatation of the oesophagus.
- ***More severe cases may require an oesphagectomy, followed by a stomach pull-up or small bowel replacement.***
- However, when the stricture reduces the ability of the child to eat, then a feeding gastrostomy tube passed through the abdominal wall directly into the stomach may be needed to provide nutrition.

Prevention

- Parents and teachers informed to keep dangerous fluids out of the reach of children.
- Never put chemicals in the wrong bottles or containers.

5.11 Management of burns

Anthony Roberts

Minimum standards requirements

- Analgesia (see Chapter 1.27)
- Shock (see Chapter 3.6)
- Antiseptic dressings
- Antitetanus immunisation
- Antibiotics: flucloxacillin, penicillin

Summary of actions

- Assess the airway. If signs of obstruction, insert oral or nasal airway or consider early intubation before swelling and respiratory arrest occur.
- Take a very brief history; could there be other injuries?
- Expose the body and make a rapid assessment of the burn area.
- If clearly more than 10%, establish an intravenous cannula and give intravenous analgesia (morphine 100 micrograms/kg as a loading dose, then see Chapter 1.27).
- Commence either 0.9% saline or Hartmann's solution at 3 ml/kg per % burn for the first 24 hours, backdated to the time of the burn. Half of this should be given (in hourly divided doses) during the first 8 hours, and the second half in the next 16 hours, again in hourly doses.
- Even if less than 10%, consider intravenous analgesia if the child is clearly distressed.
- Do not give oral fluids at this stage.
- Establish any other injuries or medical conditions.
- Make an accurate assessment of the area of the burn and draw it on a chart (see Figure 5.11.1).
- Estimate the depth of the burn.
- Establish, and if necessary update, the antitetanus status of the child.
- Consider and decide whether an escharotomy is necessary.
- Dress the burned areas, or treat any area which is going to be kept exposed.
- Consider and decide whether the child needs admission (with parent).
- Commence oral fluids. In burns >8% divide the calculated daily requirement by 24 and give it on an hourly basis. Free fluid can lead to vomiting.
- Decide if the child requires urinary catheterisation (>30%, or with complications).

Further information about all of these decisions will now be presented.

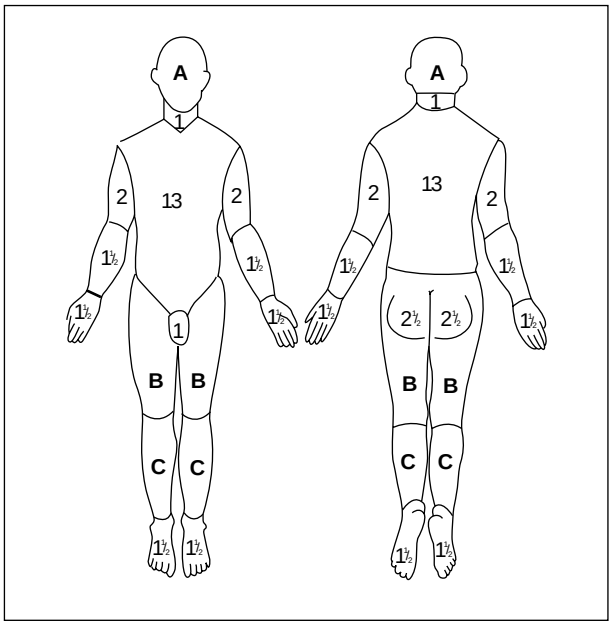

Figure 5.11.1 Burn area assessment.

	% Surface area at				
Area indicated	0	1 year	5 years	10 years	15 years
A	9.5	8.5	6.5	5.5	4.5
B	2.75	3.25	4.0	4.5	4.5
C	2.5	2.5	2.75	3.0	3.25

Legend for Figure 5.11.1

Introduction

- The severity of a burn depends on the area of the body involved and on its depth.
- The majority of burns in children are caused by hot fluids or flame.
- Other causes include electricity, chemicals, radiation and frostbite.

Definitions

Erythema. An increase in skin capillary blood flow. In pigmented skin it is often difficult to recognise, but is characterised by discomfort, a slight thickening and change of texture of the surface of the skin with later partial or complete desquamation occurring some days afterwards. **The important element is that blistering does not occur, and fluid is not lost**.

Superficial partial thickness burn. This is skin in which there is early (within one hour) blistering following the injury, associated with pain. If the blisters are removed, the exposed surface is shiny, loses pigmentation in pigmented races and is extremely painful. Pressure on the surface causes blanching which on release of the pressure instantly becomes red again.

Deep dermal burn. Red blood cells leave the capillaries and become fixed in the dermis. In non-pigmented skin therefore the redness does not blanch on pressure. This is much more difficult to diagnose in pigmented skin but the skin becomes thicker and harder in the area. Blistering occurs later, or may not occur at all. If the burn is in the deeper part of the dermis, the heat breaks down the red cells and the area becomes white with no blanching present. Removal of the blistering, if it has occurred, leaves a bed that is wet and shiny, but has mild discomfort only.

Deep burn. All elements of the skin and the skin hair follicles, sweat glands, etc. are destroyed. The skin is either white or charred brown. No blistering occurs. **Painless on examination**.

First aid

✔ **Cold water rapidly applied is the best first aid**

Seconds count. **The longer the skin is in contact with the flame or hot fluid the greater is the extent and depth of burning**. The best first aid in all situations, except with electricity, is cold water or other cold fluid (milk, etc.) applied as soon as possible. It is less important whether the water is sterile or not, and it should be applied before clothes are removed, as removing clothes can often take some time. Cold water reduces the severity of the burn and also reduces pain. It should ideally be applied for approximately 10 minutes. Following this, the burn should be covered with clean dressings or Klingfilm plastic wrapping.

If the cause of the burn is electricity, then it is important that the child is isolated from electricity or it is turned off before cold water is applied, otherwise greater damage could be done.

Following the period of cooling with water, the child needs to be kept warm; otherwise hypothermia can result, particularly in young babies.

Assessment

- Assessment of a burn must be carried out in the same way as the assessment of any other injured child.
- It is quite possible that the burn is not the major injury or problem when the child is seen. For instance, it may have been an epileptic attack that caused the burn, or the child may have fallen or jumped from a burning house, or been involved in a road traffic accident and hence have multiple fractures and/or a head injury.

ABC

Airway (see Chapter 5.10)

Breathing

- If either of these are compromised, early use of an oral or nasal airway, or endotracheal intubation may be required.
- If flame inhalation has occurred, then the airway tends to close very rapidly, making intubation very difficult. Apart from the history, the signs to observe are altered voice or stridor, singeing of the nasal hairs and deposition of soot in the throat or nose.

Circulation

- Fluid is lost through the capillaries following a burn. In minor burns this is a local phenomenon, but in severe burns then all of the vascular bed becomes leaky.
- A child with a burn of less than 10% of the total body surface area can normally cope by increasing the oral intake. This, however, is not an absolute figure, and in particular if the child is vomiting then intravenous fluid may be necessary for a smaller burn. Similarly, if safe intravenous fluid is not available, then a burn of up to 25% may have to be managed with increased oral fluids alone. When oral fluids are being used, either in combination with intravenous therapy or alone, then only small regular doses of fluid should be given.
- For larger burns >5% oral fluids should be an electrolyte solution (ORS).

Burn area and depth

Area of burn

Estimation of the area of a burn is based on Wallace's Rule of Nines, on charts (see Figure 5.11.1), and the fact that the area of the **patient's** extended hand with fingers closed is approximately 1% of the total body surface area.

Wallace's Rule of Nines is applicable to older children and adults. For newborn babies it can be modified by adding 9% to the area of the head and taking 9% from the area of the legs. For every year older that the child becomes, then 1% is taken from the head and added to the legs until, at the age of 10, approximately adult proportions have been reached. The area of the child's hand

can be used for estimating the size of small burns, but can also be used to estimate the areas unburned in extensive burns, and this can then be extracted from the Rule of Nines figures, i.e. if 2% of an arm is unburned then the area of burn on that arm will be 7%.

- It is very common for inexperienced people to overestimate the size of burn.
- Erythema MUST NOT be included – fluid is not lost.
- The decision whether or not to start intravenous fluids is dependent on this initial assessment.
- An overestimation will mean that far too much fluid may be given.

Depth of burn

The depth of the burn is based on history, appearance and examination.

- Flame or hot fat burns in children are almost always deep.
- Hot water burns (scalds) may be superficial or deep dermal.
- The appearance can be altered if more than a few hours old, or by the application of various first aid treatments.
- First: assess capillary return.
  Second: test sensation. Is it increased (in a superficial partial thickness burn), reduced (in a deep dermal burn) or absent (in a full-thickness burn)?

The test is done by using a sterile hypodermic needle. In older children, it is possible to ask whether they can tell the difference between the sharp and the blunt ends when lightly applied to the burn. In younger children the best way of doing the test is when the child is sleeping or has his/her eyes closed and very gently touching what appears to be the deepest part of the burn. If there is a sudden startle reflex then it is probably a superficial partial thickness burn, a slow awakening a deep dermal burn, and if it is possible to put the needle into the burn with no response then it is likely to be a deep burn.

Many superficial burns become deeper during the first 48 hours after their occurrence, and need reassessing at 48 hours.

Treatment

Analgesia

- In all cases of shock, or potential shock, then analgesia should be given intravenously (see Chapter 1.27).
- Oral analgesia does not work, and intramuscular analgesia can be very dangerous because when the circulatory volume is re-established and muscle blood flow recommences, the child can become overdosed. Opiate overdose can be reversed with naloxone given intravenously (see Chapter 1.27).

Intubation and IV resuscitation

The two major initial decisions that must be made for any patient with burns are:

- Whether to intubate
- Whether to commence intravenous resuscitation.

Intravenous fluids

- Ideally by peripheral vein; in emergency, intraosseous, intraperitoneal or central venous lines may be needed but these can increase risk of infection.
- Wherever possible, long lines should not be used as this increases the risk of septicaemia.
- Intravenous resuscitation is necessary in large burns because of the loss of the intravascular component of the circulation.
- Normal (0.9%) saline is probably the best fluid as additional potassium is not usually required because of the cell breakdown from the injury. Hartmann's solution is also appropriate.
- Glucose 5% and glucose in 0.18% saline are **inappropriate** and can lead to hyponatraemia and water overload. Both natural colloids, i.e. albumin solution, plasma and blood, as well as artificial colloids, i.e. Haemacel, and various starch derivatives are available. The former have risks of transmitting infection and are very expensive, and the latter have not been well studied for resuscitation of burns, but are cheaper.
- A combination of colloid and crystalloid resuscitation can be used. It is essential that not too much intravenous fluid is given since it may lead to pulmonary and/or cerebral oedema, together with an excessive extravascular deposition of fluid. Crystalloid resuscitation can also lead to "compartment syndrome" because of the increasing pressure within the muscular compartments and it is important to observe for pain, particularly in the lower legs.
- The amount of fluid loss from burns decreases over the first 48–72 hours after the injury. The amount of fluid to be given initially therefore depends on how long before admission the burn occurred. This calculated amount can be given as rapidly as possible as a bolus dose. Following this, the assessment of the resuscitation can be made by a combination of the clinical picture, i.e. degree of dehydration, the blood haematocrit and the urine output.
- It is essential throughout that accurate and updated fluid input and output charts are kept. For major burns (over 30%) hourly haematocrit (or haemoglobin) and urine outputs (ideally 1 ml/kg/hour) are helpful in the first 24 hours and then decreasing afterwards. For burns between 10% and 30% then 4-hourly tests are normally sufficient.
- In larger burns, i.e. greater than 30%, and burns involving the genitalia and in young normally incontinent female children, a urinary catheter is essential. In males, a urinary bag can be used. A catheter may also be necessary if resuscitation is not proceeding well. Catheters can lead to infection and should be removed as soon as possible.

Enteral fluids

- Start oral or nasogastric feeding of the child as soon as convenient after admission. If the child is being breastfed then this should continue.
- Although thirst is common, free fluid orally may induce vomiting which can make resuscitation more difficult.

- For burns between 5% and 10% the daily requirement of that child's oral intake should be increased by 50% to allow for the burn (given on an hourly basis).
- The normal oral requirement of a child can be calculated as 100 ml/kg for the first 10 kg; 50 ml/kg for the next 10 kg; and 25 ml/kg for any weight up to the total weight of the child.
- This may need to be increased by 10% or 20% in hot climates.
- For example, in a child of 1-year-old where the daily requirement is 800 ml, add 400 ml (i.e. 50% extra) for the burn making 1200 ml, divide by 24 and give 50 ml orally per hour.
- The oral fluid given should ideally be Dioralyte or a similar electrolyte solution suitable for dehydrated children. If this is not available, diluted milk or water is acceptable.
- If the child is being intravenously resuscitated then the normal daily requirement of fluid is **also** given orally, again on an hourly basis. If vomiting occurs, the daily requirement must be given IV.
- If all is well after 24 hours then free fluids can be given, but close input and output charting will continue to be required.

Feeding

- Early feeding reduces the risk of gastric ulcer formation and of stasis. It is recommended therefore that either small quantities of food are given orally or with a thin-bore nasogastric tube. A thin-bore nasogastric tube can be used to give milk or other similar high-protein foodstuffs.
- Intravenous feeding is strongly contraindicated as this will not aid the gastrointestinal tract, and leads to a high risk of septicaemia.

Treatment of the burn

Minor burns

The best definition of a minor burn is one that can be treated as an outpatient.

Admission

- A child with a burn should be admitted unless it is completely safe to be treated as an outpatient.
- If possible, isolate the child in a warm clean room.
- The following patients require admission:
  - All airway burns or children with a history of smoke inhalation.
  - Burns in children of more than 6% total body surface area.
  - Deep burns of greater than 5 cm diameter.
  - Moderate burns of the face, hands or perineum.
  - Where there is inadequate social support in the home.
  - Any suspicion of non-accidental injury.

Dressings

- Because a burn is normally caused by hot fluids or flame, the burn wound is sterile.
- **Hands should be washed and sterile gloves should be worn by all members of the team whenever the patient is being touched**. ✔
- Ideally plastic aprons should also be used to prevent cross-infection during dressings, etc.

Purpose and placing of dressings

The purposes of a dressing are:

- To maintain sterility
- To relieve pain
- To absorb fluid produced by the burn wound
- To aid healing.

As to placement of the dressing:

- The layer of the dressing closest to the wound should contain an antiseptic: chlorhexidine or povidone iodine (10%)
- On top of this dressing should be placed a layer of gauze and then sterile cotton wool to absorb fluid
- The whole to be held in place by a bandage.

Dressing changes

- Every time a dressing is changed, there will be pain, and the delicate reforming epithelium will be injured.
- **Dressings should not be done on a daily basis,** ✔ particularly in a superficial partial thickness wound. Therefore the initial change should be at 48 hours approximately after the burn when dressings come off easily, the maximum amount of fluid has been discharged from the wound, and it is possible to reassess the wound for area and depth.
- If at the first dressing change, the wound is still a superficial partial thickness burn, then the second dressing is left for eight further days, by which stage healing should have occurred.
- If the wound is deeper, then a decision whether to operate must be made (see below) but the second dressing can still be left for at least a week.
- When surgery is not possible or appropriate, dressings can be done initially on a weekly basis but increased to two or three times a week as greater infection and discharge develops.
- When any dressing is done, microbiological swabs should be taken.

Tetanus

- Antitetanus prophylaxis should be given at the earliest possible time.

Antibiotics

- Haemolytic *Streptococcus pyogenes* and *Pseudomonas aeruginosa* are the most common serious infections.
- In most burns *Staphylococcus aureus* is also present, but does not need treatment unless it is invasive. If it is, flucloxacillin is more appropriate than penicillin.
- Antibiotics should only be given when there are signs of invasive infection.
- An exception to this is *Streptococcus pyogenes* which should be treated with flucloxacillin (not

benzylpenicillin) as soon as it is found on a swab or suspected clinically (for example lymphangitis).

Surgery

The surgical treatment of burns can be divided into four time zones:

Immediate	within hours
Early	within days
Medium term	within weeks
Long term	within years

Immediate surgery

There are two operations which may need to be done within hours of the burn:

- Tracheostomy
  Whenever possible this operation should be avoided as an endotracheal tube usually gives better results and less mortality (depending on available intensive care).
  - An emergency tracheostomy for a severely swollen oral/pharyngeal/laryngeal airway is a very high-risk operation if the airway has not already been secured. It is better to use a mini-tracheostomy through the cricothyroid membrane.
  - Tracheostomy has a high mortality because of infection, displacement, lung volume loss and tube blockage.
- Escharotomy
  - A deep circumferential full-thickness burn of the limb, or even occasionally the trunk, can act as a tourniquet to that area.
  - Very early release (i.e. within two hours) is necessary to prevent severe and irrecoverable muscle and nerve damage. This can be done without any anaesthetic because the deep burn has no sensation.
  - The incisions should not overlie superficial bone or tendons but need to go down to the fascia.
  - For more severe burns, and in particular, high-voltage electrical burns, appropriate incisions are needed to decompress the deep compartments as well.
  - Urgent decompression of deep compartments may be required in severe high-voltage electrical burns, which can damage underlying muscle with no skin damage visible except at the entry and exit points.

Early surgery

- Early surgery for deep dermal and deep burns has been shown to give better functional and cosmetic results with less risk of infection than allowing the natural processes of the body to remove the dead tissue.
- However, it is a technique that is difficult to learn from books, often requires blood transfusion and therefore if tangential excision is to be used without previous experience, then only a small area should be attempted.
- Blood loss can be very rapid.
- An experienced anaesthetist is important.

Medium term surgery

When wounds are granulating then thin split-skin grafts can be taken to cover the granulating areas.

Late surgery

Reconstruction to release contractures, and to improve both function and appearance is best carried out, where possible, in a specialist centre.

Facilities and personnel

- **All serious burn patients are best cared for in specialist burn units with a trained team of personnel**. ✔
- For larger burns, ideally single rooms are most appropriate and these should be kept warm at all times. It is extremely important that they are clean and that insects, etc. are controlled.
- One of the most serious problems is cross-infection between patients, and adequate plastic aprons, gloves and hand-washing facilities must be available.
- In the early stages of burn resuscitation, and after surgery, nursing should be on a one-to-one basis.

Psychology

- There are frequently major psychological consequences to major burns. Firstly, there is a long and often painful stay in hospital. Secondly, there is the loss of function and appearance than can result from the burn injury.
- There are often psychological consequences for the parents, both as a result of the guilt from allowing the accident to happen, and from the acceptance of a child with often a major alteration of appearance and function.

Prevention

- The best solution to the problem of the burn injury is prevention.
- Use antenatal classes, posters in village halls and talks in school.
- The causes of burns in children will vary in different communities and prevention should be directed at local causes.
- If possible, limit the temperature of water coming from domestic taps.

512 Poisoning

Joan Robson, Luz Marina Lozano Chavarria and David Southall

Minimum standards requirements

- ABCD (see Chapters 1.19 and 1.20)
- Shock (see Chapter 3.6)
- Convulsions (see Chapter 3.38)
- Oxygen
- Glucose
- Naloxone
- Activated charcoal
- Paediatric ipecacuanha
- Wide-bore orogastric tubes
- Desferrioxamine
- N-acetylcysteine
- Sodium bicarbonate
- Vitamin K
- Exchange transfusion
- Atropine
- Pralidoxime
- d-penicillamine
- EDTA

- **Suspected poisoning** in children is common.
- **Accidental poisoning** is usually in the age group 18–36 months.
- **Intentional overdose** is often a cry for help in adolescent girls rather than a serious suicide attempt, but all children and adolescents who take intentional overdoses should be admitted to hospital and undergo full psychiatric and social assessment.
- **Drug abuse** may be misuse of alcohol, abuse of solvents or more potent recreational drugs such as LSD, ecstasy or opiates.
- Rarely children are **deliberately poisoned** by adults either as a result of depressive illness or as part of abuse (see Chapter 5.2).

Important symptoms and signs of poisoning are:

- Sudden unexplained illness in a previously healthy child
- Drowsiness or coma
- Convulsions
- Ataxia
- Tachypnoea
- Tachycardia or flushing
- Cardiac arrhythmia or hypotension
- Unusual behaviour
- Pupillary abnormalities

Remember that there may not be a history of poisoning so ask specifically about access to prescribed drugs, local medicines, household substances, berries or plants if the above symptoms or signs are present.

Management of poisoning

- Remove the patient from the source of the poison. This mainly applies to fumes, for example in a house fire. Contaminated skin and eyes should be washed with clean water.
- Assess and stabilise **A**irway, **B**reathing and **C**irculation. Next assess **D**isability.

This initial assessment of the child with suspected poisoning is done to recognise life-threatening emergencies and the early signs of a seriously ill or injured child. A useful system for rapid assessment is that used in the Advanced Paediatric Life Support (APLS) system:

- **A**irway and **B**reathing
  - Work of breathing
  - Respiratory rate/rhythm
  - Stridor/wheeze
  - Auscultation
  - Skin colour
- **C**irculation
  - Heart rate
  - Pulse volume
  - Capillary refill
  - Skin temperature
- **D**isability
  - Mental status/conscious level
  - Posture
  - Pupils

The whole assessment should take less than a minute.

Once **A**irway, **B**reathing and **C**irculation are recognised as being stable or have been stabilised, then definitive management of specific conditions can proceed. During

✔ definitive management, **reassessment of ABCD at frequent intervals will be necessary to assess progress and detect deterioration**.

- Test for hypoglycaemia and if present treat with 0.5 g glucose/kg body weight (IV if unconscious, otherwise orally.) If IV 2 ml/kg 10% glucose over 3 minutes then 0.1ml/kg/minute to keep blood glucose 5–8 mmol/litre.
- Treat convulsions with diazepam 100–200 micrograms/kg IV slowly or 500 micrograms/kg per rectum.
- If an opiate or methadone overdose is suspected give naloxone, 10 micrograms/kg IV repeated up to a maximum of 2 mg. Remember that naloxone has a short half-life and further boluses or an infusion of 10–20 micrograms/kg/hour or more may be required.
- Identify the substance ingested or inhaled if at all possible. Questions to be asked are:
  - What medicines, domestic products, berries or plants has the child had access to?
  - How much has been taken?
  - When did the child have access to the substances?
  - Is the container or a sample available as this will be helpful at the hospital?
  - Are other children involved?
  - What symptons has the child had?

Use National Poisons Information Centres or computer-based references to get information on the side effects, toxicity and treatment needed if these services are available.

- Anyone telephoning an emergency service for advice for a suspected poison should be asked to take the child to the nearest Emergency Care Centre with a sample of the substance ingested or the empty container.
- Remove, adsorb or neutralise the ingested substance as quickly as possible.
- If substance is non-toxic give liberal fluids orally.
- If substance is a corrosive, there may be serious injury to the mouth, throat, airway, oesophagus or stomach. The most dangerous are sodium or potassium hydroxide cleaning fluids. Others include bleach and other disinfectants. Serious oesophageal injury can result in perforations and mediastinitis, later leading to oesophageal strictures. The presence or burns within the mouth is of concern and suggests that oesophageal injury is possible. Stridor suggests laryngeal damage. ✔ **No emetic should be given**. **Milk or water given as soon as possible will help**.

  If there is a severe stricture it may be necessary to bypass the oesophagus with a gastrostomy tube. If available, ***endoscopy is essential for identifying injury***. A ruptured oesophagus will lead to mediastinitis and should be treated with gastrostomy and prophylactic antibiotics (cefuroxime and metronidazole).
- For all **other** poisons except heavy metals give activated charcoal if this is available (1 g/kg suspended in water). The sooner it is given the better (preferably within 1 hour of ingestion of poison). Repeat after 4 hours if a sustained release drug has been taken). If charcoal is not available and a potentially life-threatening dose of poison has been taken (particularly of iron), give paediatric ipecacuanha (10 ml for those aged 6 months to 2 years and 15 ml for >2 years plus a glass of water) to induce vomiting. **Do not give ipecacuanha if the child has a decreasing level of, or impaired, consciousness**. ✔ **Do NOT give if corrosive solutions have been ingested or if kerosene, turpentine or petrol have been ingested and could be inhaled following vomiting resulting in lipoid pneumonia**.
- Gastric lavage with a wide-bore orogastric tube should be used only if a potentially life-threatening dose has been taken and provided that the airway is protected. It should not be used if there is a decreasing level of, or impaired, consciousness without airway protection. **It should not be used for poisons containing hydrocarbons or corrosives**. Lavage cycles of 15 ml/kg are usually appropriate.
- In a few instances specific antidotes are advised. These should only be given when full information on the poison is available. **Never give salt to induce vomiting**. ✔
- Treat symptoms as they arise. ***If laboratory services are available take samples of blood, vomit or urine for drug levels as indicated under specific poisons***. **If the patient is comatose then the blood sugar should also be checked and *blood gases analysed*** ✔ if appropriate equipment is available.
- Give antidote if this is indicated for the specific poison (see below).
- Admit all patients with symptoms attributable to a poison and all patients who have ingested iron, pesticides, corrosives, paracetamol, salicylate, narcotic drugs or tricyclic antidepressant drugs.

  In addition, all patients who have deliberately taken poisons should be admitted.
- Always remember that an older child or adult may have given a child drugs intentionally. This is a form of child abuse and if there is the slightest suspicion of this, the appropriate child protection procedures should be instituted, if they are available. The child should be admitted (see Chapter 5.2).

Commonly ingested drugs

Local medicines

These are prescribed for diarrhoea and vomiting. They may give profound acidosis and respiratory distress. They can also give paralytic ileus.

Treatment

- Treat the metabolic disturbance
- Pass a nasogastric tube

Iron

- Usually the result of taking iron tablets prescribed for another family member.
- Produces severe gastrointestinal effects with vomiting, diarrhoea, gastrointestinal bleeding, metabolic acidosis. Subsequently after 12–24 hours there is encephalopathy, liver damage and circulatory collapse.
- Late effects include scarring of the stomach which may produce pyloric stenosis.

If available, a serum iron level at 4 hours > 300micrograms/dl indicates significant poisoning.

X ray may show the number of tablets.

✔ **Aim to remove as much as possible by vomiting.** Gastric lavage with a wide-bore orogastric tube may remove significant amounts of iron.

Desferrioxamine should be given by deep IM injection 1 g for <12 years and 2 g for >12 years. IM doses of desferrioxamine of 1–2 g should be repeated every 12 hours until serum iron is normal (serum iron less than iron binding capacity). If very ill, give IV infusion of desferrioxamine 15 mg/kg/hour to a maximum dose of 80 mg/kg in 24 hours. Usually reduce rate after 6 hours.

Paracetamol

- Can lead to liver and renal failure (see Chapter 3.9).
- Induce vomiting and if possible measure paracetamol level.
- Give *N*-acetylcysteine or methionine as soon as possible, ideally within 8 hours of ingestion. If child is conscious and tolerating oral fluids, and within 8 hours of ingestion, give methionine orally (under 6 years 1 g every 4 hours for four doses; 6 years and over 2.5 g every 4 hours for four doses).
- If child presents >8 hours after ingestion or cannot be given oral preparation, give IV *N*-acetylcysteine (initially as loading dose 150 mg/kg over 15 minutes, then IV infusion of 50 mg/kg over 4 hours, finally 100 mg/kg IV over 16 hours). An oral form of *N*-acetylcysteine is available (load 140 mg/kg and then 70 mg/kg every 4 hours for 16 doses).

Salicylates

- Produce acidotic-like breathing, vomiting and tinnitus.
- Hyperventilation is due to a direct stimulation of the respiratory centre and produces respiratory alkalosis but also there is a metabolic acidosis from ketosis. Consequently the hyperventilation is extreme.
- A fever may occur.
- There is peripheral vasodilatation.
- Moderate hyperglycaemia develops.

There is delayed gastric emptying and therefore give activated charcoal if available (1 g/kg and repeat after 4 hours) even if >4 hours after ingestion. If charcoal is not available induce vomiting.

- Give sodium bicarbonate 1 mmol/kg IV as 4.2% over 4 hours to correct acidosis and help excrete salicylate.
- Give sufficient IV fluids to compensate for hyperventilation and give sufficient glucose to minimise ketosis, but regularly monitor blood glucose.
- Watch electrolytes carefully and avoid hypokalaemia and hypernatraemia.
- In very severe cases, ***peritoneal haemodialysis if available is ideal.*** In its absence exchange transfusion may help.

Benzodiazepines

Flumazenil is a specific antagonist. Initial dose slow IV 10 micrograms/kg repeat at 1 minute intervals to a maximum of 40 micrograms/kg (2 mg maximim dose) If necessary can be followed by an infusion of 2–10 micrograms/kg/hour (not recommended in children who have received long-term benzodiazepine treatment for epilepsy).

Tricyclic antidepressants

Produce drowsiness, ataxia, dilated pupils and tachycardia. Severe poisoning results in cardiac arrhythmias (particularly ventricular tachycardia) and severe hypotension and convulsions.

Induce vomiting, perform gastric lavage and administer charcoal as above **but first protect airway if drowsy**.

- Treat convulsions as with any status epilepticus (see Chapter 3.38).
- Monitor the ECG continuously. Arrhythmias can be reduced by using IV phenytoin. Phenytoin is given as a loading dose of 15–20 mg/kg over 30–45 minutes and then 2.5–7.5 mg/kg 12 hourly. A lidocaine infusion (10–50 micrograms/kg/minute) is an alternative to phenytoin.
- Where there is severe cardiac toxicity, prolonged external cardiac massage may keep the child alive long enough for the drug to wear off.

Poisonous household and natural products

Bleach (3–6% sodium hypochlorite)
Do not induce vomiting ✔

- Symptoms: burning sensation, vomiting and abdominal discomfort.
- Treatment: liberal fluids and milk.

Corrosive agents
Do not induce vomiting ✔

- Oven cleaners (30% caustic soda)
  Kettle descalers (concentrated formic acid)
  Dishwashing powders (silicates and metasilicates)
  Drain cleaners (sodium hydroxide)
  Car battery acid (concentrated sulphuric acid)
- Symptoms: considerable tissue damage of skin, mouth, oesophagus or stomach – late strictures may occur.
- Treatment: wash skin and mouth to dilute corrosive.

Petroleum compounds such as kerosene, turpentine and petrol
Do not induce vomiting ✔

- If inhaled may give hydrocarbon (lipoid) pneumonia leading to a cough, respiratory distress with hypoxaemia due to pulmonary oedema and lipoid pneumonia. A chest *X* ray is essential in all cases.
- If large amounts are ingested may cause encephalopathy.
- Additional inspired oxygen may be required.
- An antibiotic may be needed but only for secondary chest infections.
- Dexamethasone may help if lipoid pnuemonia.

Organophosphorus compounds

Insecticides such as, malathion, chlorthion, parathion, TEPP and phosdrin can be absorbed through the skin, lungs or gastrointestinal tract.

Symptoms due to excessive parasympathetic effects due to inhibition of cholinesterase include excessive secretions of mucus in the lungs (bronchorrhoea) with ensuing respiratory distress and sometimes wheezing, salivation, lacrimation, bradycardia, sweating, gastrointestinal cramps, vomiting, diarrhoea, and convulsions, blurred vision and small pupils, muscle weakness and twitching progressing to paralysis, loss of reflexes and sphincter control.

Treatment

- Aim is to get rid of poison from:
  Eyes: copious irrigation.
  Skin: remove contaminated clothing and wash.
  Gastrointestinal tract: give activated charcoal 1 g/kg and repeat after 4 hours.
- Admit all cases as some effects are late.

✔ In severe cases particularly where there is bronchorrhoea give **atropine** 10–50 micrograms/kg IV or IM every 5–10 minutes until the skin becomes flushed and dry, the pupils dilate and tachycardia develops (that is atropinisation has occured). A specific cholinesterase reactivator can also be given as follows and ideally within 12 hours of ingestion (ineffective after 24 hours).

Pralidoxime 30 mg/kg diluted with 10–15 ml of water by IV infusion at a rate not exceeding 5 mg/minute. It should produce an improved muscle power in 30 minutes. It can be repeated once or twice as required and as is shown to be effective.

Assisted ventilation may be required if available.

Lead poisoning

This is usually a chronic form of poisoning. The lead can come from paint, from lead piping or, from car batteries. In some cultures, lead-containing substances can be applied for cosmetic purposes, for example Surma in India.

Early signs are non-specific for example, vomiting, abdominal pain and anorexia. Anaemia is usually present. Prior to encephalopathy with raised intracranial pressure, there may be headaches and insomnia. Peripheral neuropathy may be present. *X* rays may show bands of increased density at the metaphyses. Harmful effects on the kidneys result in hypertension, aminoaciduria and glycosuria. There is a microcytic hypochromic anaemia with punctate basophilia. The diagnosis is made by showing a marked increase in urinary lead after d-penicillamine and elevated blood lead levels.

Treatment

- Treat by first removing the source of ingested lead.
- A diet rich in calcium, phosphate and vitamin D (plenty of milk) should be given if possible.
- In cases of lead encephalopathy, use IV infusion of edetate calcium (EDTA) in 250 ml of 5% glucose 15–20 mg/kg every 6 hours for 5–7 days. A repeat course may be needed two weeks later.
- Boluses of mannitol 250–500 mg/kg IV over 30–60 minutes may also be required for raised intracranial pressure whilst the above is given.
- An alternative approach is as follows:
  The standard chelating agents are BAL (dimercaprol), sodium calcium edetate (EDTA), d-penicillamine and 2–3 dimercaptosuccininc acid (DMSA).

  If encephalopathy or blood lead >100micrograms/dl give BAL 450 mg/m^2/day in 6 divided doses IM for 2 days plus EDTA 1.5 g/m^2/day in 4 divided doses by slow IV infusion every 6 hours.

  If encephalopathy is absent but a severe degree of poisoning is present give BAL 300 mg/m^2/day in 4 divided doses IM for 2 days and EDTA 1 g/m^2/day in 4 divided doses by slow IV infusion every 6 hours.

  If poisoning is chronic and blood lead is <45 micrograms/dl give ORALLY DMSA 10 mg/kg three times daily for 5 days then 10 mg/kg twice daily for 14 days. Repeat after an interval >2 weeks if required (do not give DMSA if there is lead visible on abdominal *X* ray; it increases lead absorption).

Poisonous plants

- Usually only small quantities are ingested.
- Treatment: activated charcoal and supportive therapy given.

Carbon monoxide poisoning

Toxic effects are due to hypoxia. Move the child from the source and give 100% oxygen as soon as possible (half-life of carbon monoxide is 5 hours in room air but 1.5 hours in 100% oxygen). The child may look pink but is hypoxaemic and therefore guide the duration of oxygen on other clinical signs of hypoxia rather than cyanosis which will be masked. For similar reasons, pulse oximeters will give falsely high readings. ABCD management as per APLS may be required.

Cerebral oedema may develop.

Hyperbaric oxygen treatment may be helpful if available.

Volatile substance abuse ("sniffing")

This mainly occurs in the age range 11–17 years and is a group activity. Substances that are sniffed or sprayed into the respiratory system are numerous. The commonest are solvent-based adhesives ("glue sniffing"), butane gas, cleaning fluids, aerosols and fire-extinguisher substances.

Clinical features

Sores around mouth and nose. Smell of solvents on clothes and breath. All features of ethyl alcohol intoxication plus extreme disorientation, hallucinations and sudden "unexplained" death. Accidents can occur secondary to volatile substance abuse, for example falling from height, drowning, suffocation and inhalation of vomit.

Management

Remove from atmosphere of solvent. Admit to hospital. Try to confirm diagnosis by ***blood and urine tests***. Treat symptomatically. Arrange expert psychological and emotional support.

Laboratory investigations in poisoning in children

These are often expensive and/or are very time consuming to perform. They should only be requested if the result will alter the management of the patient. Many hospitals will not have these facilities.

Alcohol

- *Blood alcohol estimations are useful if:*
  There is an indication that methyl alcohol has been ingested.
  The patient is very drowsy or comatose and there is doubt that sufficient alcohol has been ingested to explain the symptoms.
- Blood sugar estimations should be done in all cases of alcohol ingestion in children. Do blood glucose stick test first and if this is low, a quantitative glucose analysis should be requested. If in doubt give glucose (see above for doses).

Interpretation
Peak blood levels of alcohol occur 30–60 minutes after ingestion.

Barbiturates

Only phenobarbitone can be detected by the hospital laboratories. Tests should be requested on comatose patients suspected of taking phenobarbitone. Other barbiturates have to be assessed at a supraregional laboratory. Results are not obtained for at least three days.

Interpretation
Plasma concentrations of phenobarbitone in adults greater than 90 mg/litre are consistent with severe poisoning; return to consciousness is expected at 45–90 mg/litre.

Carboxyhaemoglobin

Should be estimated if there has been significant exposure to carbon monoxide and the patient is showing symptoms of poisoning.

Interpretation
In adults, symptoms are not usually apparent until carboxyhaemoglobin reaches 30%; normal levels are 1.5%, but smoking can increase the level to 6.5%.

Cyanide

Appreciable quantities of cyanide can be formed during the burning of plastics including foam-filled furniture. Blood levels are unlikely to be obtained in time to be of clinical relevance.

Iron

Children ingesting iron should ideally have a plasma iron estimated before desferrioxamine is given. Serum levels of 300 micrograms/dl are associated with moderate toxicity, 500 micrograms/dl with serious toxicity and 1 mg/dl with death.

Interpretation
Patients with acute iron poisoning have significant increase in plasma iron within 2 hours of overdosage. Initial serum levels less than 90 micromols/litre are supportive but not absolute evidence of mild poisoning. Normal serum iron levels are in the region of 10–30 micromols/litre (80–180 micrograms/dl).

Paracetamol

Take blood at least 4 hours after ingestion of paracetamol.

Interpretation
A plasma level which falls above a line joining 200 mg/litre at 4 hours and 50 mg/litre at 12 hours when plotted on a semilog/timescale indicates moderate-to-severe poisoning. Lower thresholds for treatment are indicated if the patient is on enzyme-inducing drugs or alcohol is taken habitually.

Salicylate

Take blood on presentation and at intervals while level is still rising – remember absorption can continue up to 24 hours.

Interpretation
Therapeutic levels are 15–30 mg/dl.

Tricyclic antidepressants

A qualitative screening test is available for confirmation of diagnosis. This is done on stomach aspirate or urine.

Paraquat

A test kit for paraquat in urine is available. Paraquat is reduced to a blue radical ion with sodium dithionite under alkaline conditions.

We are very grateful to Dr Alex Proudfoot for his help with this chapter.

513 Envenoming

David Lalloo and R D G Theakston

Minimum standards requirements

- Mono- and polyspecific antivenoms
- Epinephrine
- Chlorpheniramine
- Anticholinesterase (only if appropriate for region)
- Analgesia (see Chapter 1.27)
- Prazosin (only if appropriate for region)
- Heart failure treatment (see Chapter 3.5)

Envenoming by snakes, scorpions, spiders or marine venomous animals is common in many areas of the tropics. Children are particularly at risk; they may be attracted to venomous creatures and do not recognise the danger that they represent. Envenoming is often more severe and more rapid in children as ratio of amount of venom to bodyweight is much higher.

A clear-cut history of envenoming is often not present. Some bites are not recognised at the time of the event, other children will be too young to explain what has happened. Envenoming should always be considered in any child with an unexplained illness, particularly if there is severe pain, swelling or blistering of a limb or if a child is bleeding or has signs of neurotoxicity.

Prevention

Discourage children from handling snakes/scorpions or spiders or touching marine animals. They should be taught to avoid putting their hands down holes and to carefully check shoes and clothing before dressing. Keeping grass short around dwellings, use of sensible footwear, keeping dwellings insect-free and taking care when swimming can all help to prevent injury by venomous animals.

Snakebite

There are a large number of species of venomous snakes throughout the world. These can be divided into three main categories: **vipers**, **elapids** and **sea snakes**. The pattern of envenoming depends upon the biting species; clinicians need to know about snakes present in region in which they work. **Only 50–70% of patients bitten by venomous snakes develop signs of envenoming**.

Major clinical effects following snakebite can be divided into:-

- **Local effects**: pain, swelling or blistering of the bitten limb. Necrosis at site of the wound may sometimes develop.
- **Systemic effects**
  - Non-specific symptoms: vomiting, headache, collapse.
  - Painful regional lymph node enlargement indicating absorption of venom.
  - Specific signs:
    Non-clotting of blood.
    Bleeding from gums, old wounds, sores.
    Neurotoxicity: ptosis, bulbar palsy and respiratory paralysis.
    Rhabdomyolysis: muscle pains and black urine.
    Shock: hypotension, usually due to hypovolaemia.
- **Vipers** most commonly cause local swelling, shock, bleeding and non-clotting blood.
- **Elapids** cause neurotoxicity and usually minimal signs at the bite site (with the exception of some cobras which also cause necrosis).
- **Sea snakes** cause myotoxicity and subsequent paresis. Exceptions to this general rule do occur; for example, some vipers cause neurotoxicity and some Australasian elapids also cause non-clotting blood and haemorrhage.

First aid outside hospital

- Reassure the patient. Many symptoms following snakebite are due to anxiety.
- Immobilise and splint the limb. Moving the limb may increase systemic absorption of venom.
- Wipe site with a clean cloth.
- Avoid harmful manoeuvres such as cutting/suction/tourniquets.
- Apply a pressure bandage if tissue necrosis is rare following snakebite in your region, particularly if rapid transport to hospital is not possible. This is especially important for snakes that cause neurotoxicity. Apply a crêpe bandage over the bite site and wind firmly up

the limb. **Note**: This can only be recommended on a regional, not clinical basis, as necrosis is not apparent initially.
- Transport patient to hospital as soon as possible.
- If snake has been killed, take it with the patient to hospital.

Diagnosis and initial assessment

- Carefully examine bitten limb for local signs.
- Measure pulse, respiration rate, blood pressure and urine output. Blood pressure and other signs of shock (see Chapter 3.6) must be watched for if children are unwell, are bleeding or have significant swelling; shock is common in viper bites.
- Look for non-clotting blood. This may be only sign of envenoming in some viper bites. The 20-minute whole-blood clotting test (WBCT20) is an extremely easy and useful test. Perform on admission and repeat 6 hours later.

WBCT2O test

- Place a few millilitres of freshly sampled blood in a new, clean, dry glass tube or bottle.
- Leave undisturbed for 20 minutes at ambient temperature.
- Tip vessel once.
- If blood is still liquid (unclotted) and runs out, patient has hypofibrinogenaemia ("incoagulable blood") as a result of venom-induced consumption coagulopathy.

- Look carefully for signs of bleeding which may be subtle (gums/old wounds/sores). Bleeding internally (most often intracranial) may cause clinical signs.
- Look for early signs of neurotoxicity; ptosis (children may interpret this as feeling sleepy), limb weakness, or difficulties in talking, swallowing or breathing.
- Check for muscle tenderness and myoglobinuria in sea snake bites.
- Take blood for:
  - Haemoglobin, white cell count and platelet count.
  - Prothrombin time, APTT and ***fibrinogen levels if available***.
  - Serum urea and creatinine.
  - ***Creatine phosphokinase (CPK) (reflecting skeletal muscle damage) if available***.
- ECG if available.

Hospital or health centre management

General management

- Observe in hospital for at least 24 hours, even if there are no signs of envenoming initially. Review regularly; envenoming may develop quite rapidly.
- Nurse patients on their side with a slight head-down tilt to prevent aspiration of blood or secretions.
- Avoid intramuscular injections and invasive procedures in patients with incoagulable blood.
- Give tetanus prophylaxis. Routine antibiotic prophylaxis is not required unless necrosis is present.

Antivenom

Antivenom is indicated for signs of systemic envenoming. Evidence for its efficacy in severe local envenoming is poor, but it is usually indicated if swelling extends over more than half the bitten limb. Monospecific (monovalent) antivenom may be used for a single species of snake, polyspecific (polyvalent) for a number of different species. Dose of antivenom depends upon manufacturers' recommendations and local experience. **Children require exactly the same dose as adults (dose is dependent upon amount of venom injected, not bodyweight)**.

- Dilute antivenom in 2–3 volumes of glucose/0.9% saline and infuse over an hour or so. Infusion rate should be slow initially and gradually increased. **Note**: doses of antivenom vary considerably.
- Draw up epinephrine in a syringe ready for use.
- Observe patients closely during antivenom administration. Common early signs of an antivenom reaction are urticaria and itching, restlessness, fever, cough or feeling of constriction in the throat.
- Patients with these signs should be treated with epinephrine 10 micrograms/kg IM and if a nebuliser is available, 5 ml 1 in 1000 epinephrine. An antihistamine, for example chlorpheniramine (200 micrograms/kg IM or IV) should also be given.
- Unless life-threatening anaphylaxis has occurred, antivenom can cautiously be restarted after this treatment. Routine prophylaxis against antivenom reactions is currently unproven and should not generally be used.
- Monitor response to antivenom. In presence of coagulopathy, restoration of clotting depends upon hepatic resynthesis of clotting factors. Repeat WBCT20 and other clotting studies if available, six hours after antivenom; if blood is still non-clotting, further antivenom is indicated. After restoration of normal clotting, measure clotting at six-hourly intervals as a coagulopathy may recur due to late absorption of venom from bite site.

Response of neurotoxicity to antivenom is less predictable. In species with predominantly postsynaptically-acting toxins, antivenom may reverse neurotoxicity; failure to do so is an indication for further doses. However, response to antivenom is poor in species with presynaptically-acting toxins.

Other therapy

- Excise sloughs from necrotic wounds. Skin grafting may be necessary. Severe swelling may lead to suspicion of a compartment syndrome. Fasciotomy should **not** be performed unless there is definite evidence of raised intra-compartmental pressure ***(>45 mmHg) if measurable***, and any coagulopathy has been corrected. **Note**: **clinical assessment for compartment syndromes often is misleading following snakebite**: **therefore objective criteria necessary**. ✔
- Blood products are not necessary to treat a coagulopathy if adequate antivenom has been given.

- Endotracheal intubation or even tracheostomy should be considered to prevent aspiration if bulbar palsy develops; often obvious when difficulty in swallowing leads to pooling of secretions.
- If antivenom is not available, then give vitamin K 300 micrograms/kg IV and ***fresh frozen plasma 10–20 ml/kg if available***.
- Paralysis of intercostal muscles and diaphragm requires ***artificial ventilation***. This can be performed by manual bagging and may need to be maintained for days, using relays of relatives if equipment is not available.
- Anticholinesterases may reverse neurotoxicity following envenoming by some species.
- Maintain careful fluid balance to treat shock and prevent renal failure.
- Some cobras spit venom into the eyes of their victims. Rapid irrigation with water will prevent severe inflammation. 0.5% epinephrine drops may help to reduce pain and inflammation.

Scorpion stings

In some areas of the world, scorpion stings are more common than snakebites and cause significant mortality. The stinging scorpion is often not seen. A number of different species have broadly similar clinical effects. The major feature of envenoming is severe pain around the bite site, which may last for many hours or even days. Systemic envenoming is more common in children and may occur within minutes of a bite. Major clinical features are caused by activation of the autonomic nervous system.

Clinical features of scorpion stings

- Tachypnoea
- Excessive salivation
- Nausea and vomiting
- Lachrymation
- Sweating
- Abdominal pain
- Muscle twitches and spasms
- Hypertension
- Pulmonary oedema
- Cardiac arrhythmias
- Hypotension
- Respiratory failure

Severe hypertension, myocardial failure and pulmonary oedema are particularly prominent in severe envenoming.

Management

- Take patient to hospital immediately; delay is a frequent cause of death.
- Control pain with infiltration of 1% lidocaine around wound or systemic opiates (with care) (see Chapter 1.27).
- Scorpion antivenom is available for some species. Give intravenously in systemic envenoming, but IM injection has been used with good effect.
- Prazosin is effective for treating hypertension and cardiac failure (5–15 micrograms/kg two to four times a day increasing to control blood pressure to a maximum of 500 micrograms/kg/day <12 years and 20 mg/kg/day >12 years). The child should be lying down for the first 4–6 hours of treatment in case of a sudden fall in blood pressure.
- Severe pulmonary oedema requires aggressive treatment with diuretics and vasodilators (see Chapter 3.5).

Spider bites

Three genera of spiders cause significant envenoming in the tropics. Each cause different clinical effects, but fatal envenoming is rare.

- **Widow spiders** (*Latrodectus* spp.) are found throughout the world. Severe pain at bite site is common. Rare cases develop systemic envenoming with abdominal and generalised pain and other features due to transmitter release from autonomic nerves. Hypertension is characteristic of severe envenoming (see use of prazosin above or Chapters 3.5 and 3.7). Antivenom is available in some regions and is effective for relief of pain and systemic symptoms. Opiates are also useful for treatment of pain (see Chapter 1.27).
- **Recluse spiders** (*Loxosceles* sp.) have a wide distribution and cause bites in which pain develops over a number of hours. A white ischaemic area gradually breaks down to form a black eschar over seven days or so. Healing may be prolonged and occasionally severe scarring occurs. Efficacy of antivenom and other advocated treatments (dapsone, steroids, hyperbaric oxygen) remains uncertain.
- **Banana spiders** (*Phoneutria* sp.) occur only in South America. They usually cause severe burning pain at the site of the bite, but in severe cases may cause systemic envenoming with tachycardia, hypertension, sweating and priapism. Polyspecific antivenom is available.

Marine envenoming

Venomous fish

Many different venomous fish may sting children if they stand on or touch the fish. Systemic envenoming is rare. Excruciating pain at the site of sting is the major effect.

- Regional nerve blocks and local infiltration of 1% lidocaine may be effective (see Chapters 1.13 and 1.27).
- Most marine venoms are heat-labile. Immersing stung part into hot water is extremely effective for relieving pain. Care should be taken to avoid scalding; the envenomed limb may have abnormal sensation. Clinicians should check water temperature with their own hand. Asking patient to immerse the non-bitten limb may help to avoid scalding.

Jellyfish

Venomous jellyfish have large number of stinging capsules (nematocysts) on their tentacles which inject venom when tentacles contact skin. Pain and wheals are the usual effects, but rarely, systemic envenoming can be life-threatening. Many of the nematocysts will remain undischarged on

tentacles that adhere to the victim; rubbing area of the sting will cause further discharge and worsen envenoming.

- In box jellyfish stings, pouring vinegar over the sting will prevent discharge of nematocysts. For most other jellyfish, seawater should be poured over stings and adherent tentacles gently removed. Ice is useful for pain relief.
- Box jellyfish stings may occasionally be rapidly life-threatening. Antivenom is available and can be administered intramuscularly.

514 Drowning and near drowning

Christiane Ronald

Minimum standards requirements

- ABCD (see Chapter 1.19)
- Radiant heat/hot water bottles
- Low-reading thermometer
- Intensive care if available

Definitions

- "Drowning" is defined as an immersion injury that results in death within 24 hours. Drowning is a leading cause of death in children <5 years.
- "Near drowning" is an immersion injury with survival for at least 24 hours, irrespective of outcome: 40–50% of submersion injuries result in death and the prognosis for recovery is worst if submersion >9 minutes or resuscitation >25 minutes.

✔ **REMEMBER**:
- Small children can drown in small volumes of water, for example in a bucket or shallow pool.
- Not all drowning is accidental (abuse/neglect).
- Other injuries may be present.
- Other illnesses may have resulted in the drowning, for example epilepsy

Properties of water

- Water can be fresh (hypotonic) or salty (hypertonic)
- Water can conceal hidden dangers: trauma, entrapment, tide and flow, contamination
- Water can act as a solid at high-impact velocity
- Water may be only one of several problems afflicting the child: alcohol, drugs, child abuse, epilepsy, trauma, etc.

Risk factors for drowning

- Access to water by unsupervised children.
- Highest mortality (78%) in rivers, canals, lakes.
- Lowest mortality (6%) in public swimming pools.
- Epilepsy and other medical disorders
- Alcohol, substance abuse, "bravado"

Problems which may be present at drowning/near drowning

- Hypothermia
- Hypoxia/pulmonary oedema/(adult respiratory distress syndrome)
- Hypotension/ventricular dysrhythmias/cardiac arrest
- Cerebral depression/coma/hypoxic ischaemic brain injury
- Other injuries, especially spinal and head injuries
- Electrolyte disturbances
- Ingestions such as alcohol, anticonvulsant drugs
- Pre-existing epilepsy

Assessment and resuscitation (see Chapter 1.20 on resuscitation for more detail)

Airway and cervical spine control, gastric decompression
Breathing – intubation (with positive and expiratory pressure), high-concentration oxygen
Circulation and control of external haemorrhage
- feel for brachial/carotid pulse
- capillary refill time (over sternum)

Disability and neurological examination (AVPU)
Exposure and temperature control – core temperature measurement (best taken with low-reading thermometer 10 cm into rectum)

Rewarming

Beware rewarming shock. (see Chapter 3.6)
- Do not allow temperature to rise >37°C.
- Prevent further heat loss: remove cold wet clothes.

External rewarming if >32°C
- Radiant heater.
- Dry warm blankets.

Core rewarming if <32°C
- Warmed IV fluid to 39°C.
- Gastric, peritoneal or bladder lavage with 0.9% saline at 42°C.
- Heated humidified oxygen (42°C).

Monitor

- Core temperature
- Vital signs: ECG tracing
- Glucose, electrolytes, ***blood gases if available***
- Chest *X* ray
- Urine output and urinalysis
- Blood culture

When to stop?

- Immersion time: most children who do not recover have been submerged for more than 3–8 minutes.
- Take a detailed history of the rescue.
- If the first gasp occurs between 1 and 3 minutes after cardiopulmonary resuscitation, the prognosis is good.
- Intact survival has been reported after cold submersion for 66 minutes.
- Survival has been reported after 6.5 hours of cardiopulmonary resuscitation.
- A child has been revived from a body temperature of 15°C.
- BUT: Cool-water drowning does not have the protection offered by ice-cold water.
- Failure to restore a perfusing rhythm within approximately 30 minutes of rewarming to 32–35°C makes further efforts unlikely to be successful.
- **Resuscitation should not be discontinued until ✔ the core temperature is at least 32°C or cannot be raised**.

515 Heat stroke

David Southall

Minimum standards requirements

- ABC (see Chapters 1.19 and 1.20)
- Shock (see Chapter 3.6)
- Ice packs
- Fans

Clinical signs

- Confusion
- Tachycardia
- Fever (>40°C)
- Hot dry skin
- Tachypnoea
- Hypotonia

Pathophysiology

- Neurological impairment.
- Renal insufficiency.
- Disseminated intravascular coagulation.
- Acute respiratory failure.
- May have underlying infection predisposing to the heat stroke.

Treatment

- **URGENT COOLING**: Aim to cool within 30 minutes. Remove clothes, spray with cool water, fan if available, ice packs to neck, axillae and groins. Especially important to cool head.
- Provide system support as necessary.
- Give fluids intravenously especially if respiratory failure.
- Give oxygen.
- In hot climates, each hospital should have a cool room (ice or air-conditioned) for emergency treatment.

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Section 6

Procedures

How to use this book

This is a comprehensive text for all paediatricians caring for children in hospital. It can be used by those with limited resources and also where greater resources are available. We have identified the different levels of care in the following ways:

- **Minimum standards requirements** are given in a highlighted box at the the beginning of each clinical chapter.
- ***A standard of care*** where resources are not limited appears as bold, italicised text.
- **Key points** of particular importance in management of children are identified by a tick in the margin and bold text.

In this way we hope the book will act as a user-friendly, speedy reference on any paediatric ward.

6.1 Assessing nutritional status and growth

Nutritional status

Calculating the child's weight-for-length is the most relevant measurement in nutritional assessment.

Measuring length

At 2 years and under

Ideally two people are needed with the child supine on a flat surface.

One person should:

- Assist in positioning the child face-up on the measuring board, supporting the head and placing it against the headboard
- Position the crown of the head against the headboard, compressing the hair
- Check that the child lies straight along the centreline of the board and is not slanted, and does not change position.

It is usual for this person to stand or kneel behind the headboard.

The second person should:

- Support the trunk as the child is positioned on the board
- Lay the child flat along the board
- Place one hand on the shins above the ankles or on the knees and press down firmly. With the other hand, place the footpiece firmly against the heels
- Measure the length (to the nearest 0.1 cm) and record immediately.

The measuring board should be checked for accuracy every month.

At 3 years and older

- Without shoes.
- Child standing with heels and back in contact with an upright wall.
- Head is held to look straight forwards with lower eye sockets in line with ears. The nose must not be tilted upwards.
- A weighted block at right angles to the wall is then lowered onto the head and a scale fixed to the wall is read.
- During measurement the child should be asked to stretch the neck to be as tall as possible BUT the heels must not leave the ground. The measurer should help to stretch the neck by firm pressure upwards under the mastoid processes.
- Measure height to within 0.1 cm and record immediately.

Measuring weight

At 2 years and under

- Leave a cloth in the weighing pan to prevent chilling the child.
- Adjust the scales to zero with the cloth in the pan.
- Place the naked child gently on the cloth in the weighing pan.
- Wait for the child to settle and the weight to stabilise.
- Measure the weight (to the nearest 10 g) and record immediately.

Standardisation of the scales should be performed weekly or whenever the scales are moved.

At 3 years and older

- In the nude or if pants are worn remove 0.1kg.
- The bladder should be emptied.

Determining the child's percentage weight-for-length or standard deviation weight-for-length

Refer to Table 6.1.1 below

- Locate the row containing the child's length in the central column of Table 6.1.1.
- Look to the left in that row for boys, and to the right for girls.
- Note where the child's weight lies with respect to the weights recorded in this row.
- Select the weight closest to that of the child.
- Look up this column to read the weight-for-length of the child.

Example 1: Boy: length 61 cm, weight 5.3 kg
This child is −1 SD weight-for-length (90% of the median).

Example 2: Girl: length 67 cm, weight 4.3 kg;
This child is less than −4 SD weight-for-length (less than 60% of the median).

Monitoring weight gain

Calculating weight gain

The example below is for weight gain over three days, but the same procedure can be applied to any interval.

- Subtract the child's weight (in grams) as it was three days earlier from the current weight.
- Divide by 3 to determine the average daily weight gain (g/day).

Table 6.1.1 WHO/NCHS normalised reference weight-for-length (49–84 cm) and weight-for-height (85–110 cm), by sex

Boys' weight (kg)						Girls' weight (kg)				
–4 SD 60%	–3 SD 70%	–2 SD 80%	–1 SD 90%	Median	Length (cm)	Median	–1 SD 90%	–2 SD 80%	–3 SD 70%	–4 SD 60%
1.8	2.1	2.5	2.8	3.1	**49**	3.3	2.9	2.6	2.2	1.8
1.8	2.2	2.5	2.9	3.3	**50**	3.4	3	2.6	2.3	1.9
1.8	2.2	2.6	3.1	3.5	**51**	3.5	3.1	2.7	2.3	1.9
1.9	2.3	2.8	3.2	3.7	**52**	3.7	3.3	2.8	2.4	2
1.9	2.4	2.9	3.4	3.9	**53**	3.9	3.4	3	2.5	2.1
2	2.6	3.1	3.6	4.1	**54**	4.1	3.6	3.1	2.7	2.2
2.2	2.7	3.3	3.8	4.3	**55**	4.3	3.8	3.3	2.8	2.3
2.3	2.9	3.5	4	4.6	**56**	4.5	4	3.5	3	2.4
2.5	3.1	3.7	4.3	4.8	**57**	4.8	4.2	3.7	3.1	2.6
2.7	3.3	3.9	4.5	5.1	**58**	5	4.4	3.9	3.3	2.7
2.9	3.5	4.1	4.8	5.4	**59**	5.3	4.7	4.1	3.5	2.9
3.1	3.7	4.4	5	5.7	**60**	5.5	4.9	4.3	3.7	3.1
3.3	4	4.6	5.3	5.9	**61**	5.8	5.2	4.6	3.9	3.3
3.5	4.2	4.9	5.6	6.2	**62**	6.1	5.4	4.8	4.1	3.5
3.8	4.5	5.2	5.8	6.5	**63**	6.4	5.7	5	4.4	3.7
4	4.7	5.4	6.1	6.8	**64**	6.7	6	5.3	4.6	3.9
4.3	5	5.7	6.4	7.1	**65**	7	6.3	5.5	4.8	4.1
4.5	5.3	6	6.7	7.4	**66**	7.3	6.5	5.8	5.1	4.3
4.8	5.5	6.2	7	7.7	**67**	7.5	6.8	6	5.3	4.5
5.1	5.8	6.5	7.3	8	**68**	7.8	7.1	6.3	5.5	4.8
5.3	6	6.8	7.5	8.3	**69**	8.1	7.3	6.5	5.8	5
5.5	6.3	7	7.8	8.5	**70**	8.4	7.6	6.8	6	5.2
5.8	6.5	7.3	8.1	8.8	**71**	8.6	7.8	7	6.2	5.4
6	6.8	7.5	8.3	9.1	**72**	8.9	8.1	7.2	6.4	5.6
6.2	7	7.8	8.6	9.3	**73**	9.1	8.3	7.5	6.6	5.8
6.4	7.2	8	8.8	9.6	**74**	9.4	8.5	7.7	6.8	6
6.6	7.4	8.2	9	9.8	**75**	9.6	8.7	7.9	7	6.2
6.8	7.6	8.4	9.2	10	**76**	9.8	8.9	8.1	7.2	6.4
7	7.8	8.6	9.4	10.3	**77**	10	9.1	8.3	7.4	6.6
7.1	8	8.8	9.7	10.2	**78**	10.2	9.3	8.5	7.6	6.7
7.3	8.2	9	9.9	10.7	**79**	10.4	9.5	8.7	7.8	6.9
7.5	8.3	9.2	10.1	10.9	**80**	10.6	9.7	8.8	8	7.1
7.6	8.5	9.4	10.2	11.1	**81**	10.8	9.9	9	8.1	7.2
7.8	8.7	9.6	10.4	11.3	**82**	11	10.1	9.2	8.3	7.4
7.9	8.8	9.7	10.6	11.5	**83**	11.2	10.3	9.4	8.5	7.6
8.1	9	9.9	10.8	11.7	**84**	11.4	10.5	9.6	8.7	7.7
8.3	9.4	10.5	11.7	12.8	**88**	12.5	11.4	10.3	9.2	8.1
8.4	9.6	10.7	11.9	13	**89**	12.7	11.6	10.5	9.3	8.2
8.6	9.8	10.9	12.1	13.3	**90**	12.9	11.8	10.7	9.5	8.4
8.8	9.9	11.1	12.3	13.5	**91**	13.2	12	10.8	9.7	8.5
8.9	10.1	11.3	12.5	13.7	**92**	13.4	12.2	11	9.9	8.7
9.1	10.3	11.5	12.8	14	**93**	13.6	12.4	11.2	10	8.8
9.2	10.5	11.7	13	14.2	**94**	13.9	12.6	11.4	10.2	9
9.4	10.7	11.9	13.2	14.5	**95**	14.1	12.9	11.6	10.4	9.1
9.6	10.9	12.1	13.4	14.7	**96**	14.3	13.1	11.8	10.6	9.3
9.7	11	12.4	13.7	15	**97**	14.6	13.3	12	10.7	9.5
9.9	11.2	12.6	13.9	15.2	**98**	14.9	13.5	12.2	10.9	9.6
10.1	11.4	12.8	14.1	15.5	**99**	15.1	13.8	12.4	11.1	9.8
10.3	11.6	13	14.4	15.7	**100**	15.4	14	12.7	11.3	9.9
10.4	11.8	13.2	14.6	16	**101**	15.6	14.3	12.9	11.5	10.1
10.6	12	13.4	14.9	16.3	**102**	15.9	14.5	13.1	11.7	10.3
10.8	12.2	13.7	15.1	16.6	**103**	16.2	14.7	13.3	11.9	10.5
11	12.4	13.9	15.4	16.9	**104**	16.5	15	13.5	12.1	10.6
11.2	12.7	14.2	15.6	17.1	**105**	16.7	15.3	13.8	12.3	10.8
11.4	12.9	14.4	15.9	17.4	**106**	17	15.5	14	12.5	11
11.6	13.1	14.7	16.2	17.7	**107**	17.3	15.8	14.3	12.7	11.2
11.8	13.4	14.9	16.5	18	**108**	17.6	16.1	14.5	13	11.4
12	13.6	15.2	16.8	18.3	**109**	17.9	16.4	14.8	13.2	11.6
12.2	13.8	15.4	17.1	18.7	**110**	18.2	16.6	15	13.4	11.9

Notes:

1. SD = standard deviation score or Z-score; although the interpretation of a fixed percent-of-median value varies across age and height, and generally, the two scales cannot be compared, the approximate percent-of-the median values for −1 and −2 SD are 90% and 80% of median, respectively (*Bulletin of the World Health Organization*, 1994, **72**:273–283).
2. Length is measured below 85 cm; height is measured 85 cm and above. Recumbent length is on average 0.5 cm greater than standing height, although the difference is of no importance to the individual child. A correction may be made by deducting 0.5 cm from all lengths above 84.9 cm if the standing height cannot be measured.

- Divide by the child's average weight in kilograms to calculate the weight gain as g/kg/day.

Monitoring charts

Charts can be used to monitor the weight gain of a severely malnourished child (see example as Figure 6.1.1). The horizontal X axis represents the number of days after admission, while the vertical Y axis represent the weight of the child in kilograms. Notice that the weight in kilograms is stepped in 0.5 kg increments. This means the chart can be photocopied and used for any child, irrespective of their starting weight. Fill in the starting weight at the appropriate level (such as 5 kg, 5.5 kg, 6 kg ... etc.). Choosing an appropriate starting weight like this is preferable to using a chart with weights marked from 0, because this more flexible chart gives a larger scale which shows the pattern of change more clearly.

A blank intake and output chart for recording the food given to an individual patient, the amount consumed, and any losses through vomiting or diarrhoea is shown in Figure 6.1.2.

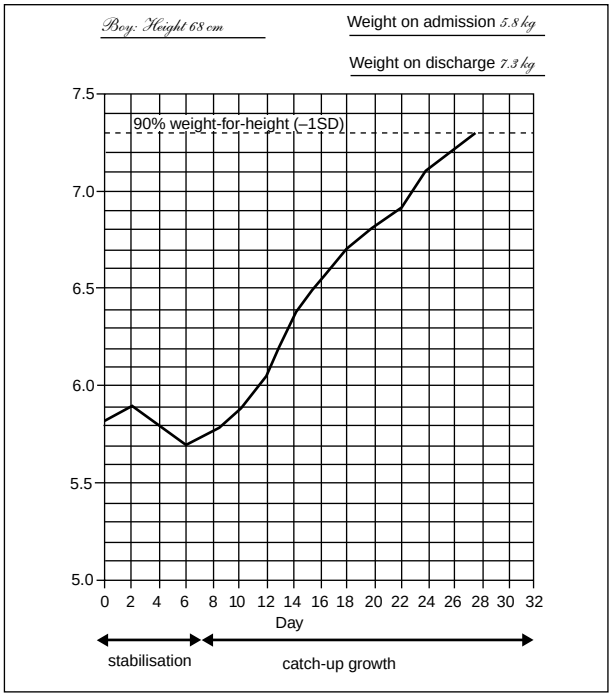


Figure 6.1.1 Weight chart: example (boy weighing 5.8 kg on admission).

Name _____ Ward _____

Hospital number _____

Age _____ Weight _____ Date of admission _____

Date ..... Feed: ..... feeds of .....ml each = ...ml per day

Time	Type of feed	Volume offered (ml)	Volume left in cup (ml)	Amount taken by child (ml)	Vomit estimate (ml)	Watery diarrhoea (Yes/No)
Totals:			Sub-total			Total taken in 24 hrs

Figure 6.1.2 Example of 24-hour food intake chart.

Additional measurements for assessing nutritional status

Measuring mid upper arm circumference

- Measure with non-stretchable tape placed around the arm midway between the elbow and the shoulder.
- The tape should be gently tightened but not to compress the underlying tissues.
- Measurement includes fat and muscle.
- Normal for child 1–5 years = 14–16.5 cm.

In summary, for a child between 1 and 5 years < 12.5 cm is definitely malnourished and between 12.5 and 13.5 cm is probably malnourished.

Measuring triceps skinfold thickness

Special skinfold callipers measure the double layer of skin and subcutaneous fat overlying the triceps muscle when the skinfold is lifted.

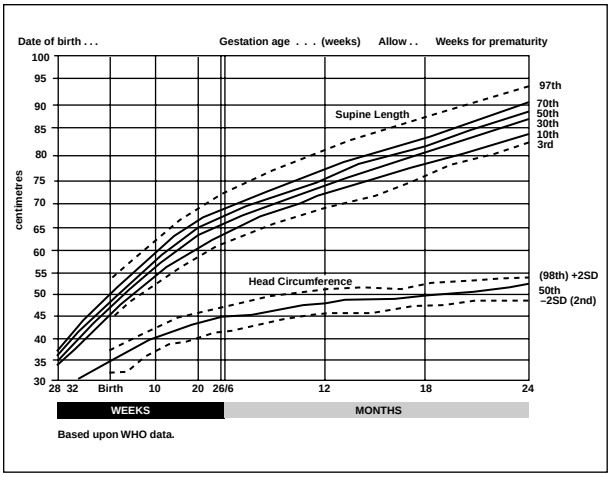


Figure 6.1.4 Girls' supine length and head circumference. Birth to 2 years.

Measuring growth and development

Individual measurements of weight and height/length can be plotted sequentially on charts to identify any failure of growth.

Figures 6.1.3 to 6.1.10 show charts of height and weight for boys and girls. These charts also include data for infants born prematurely and also head circumference measurements.

Measurement of head circumference

- Use a non-stretchable tape.
- Measure around the forehead above the eyebrows to the maximum occipital point.
- Measure twice for accuracy.

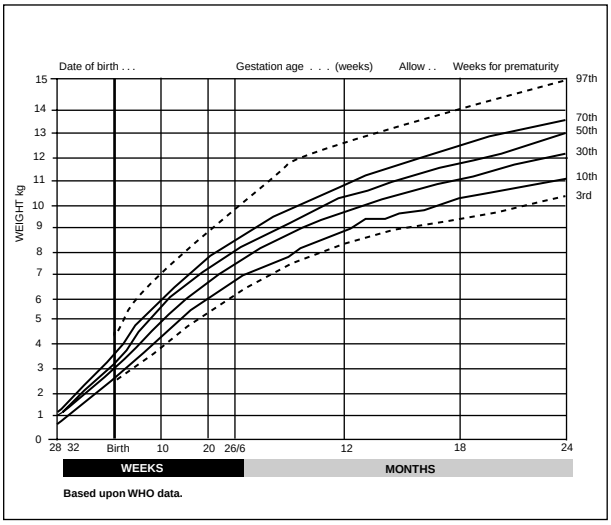


Figure 6.1.5 Boys' weight. Birth to 2 years.

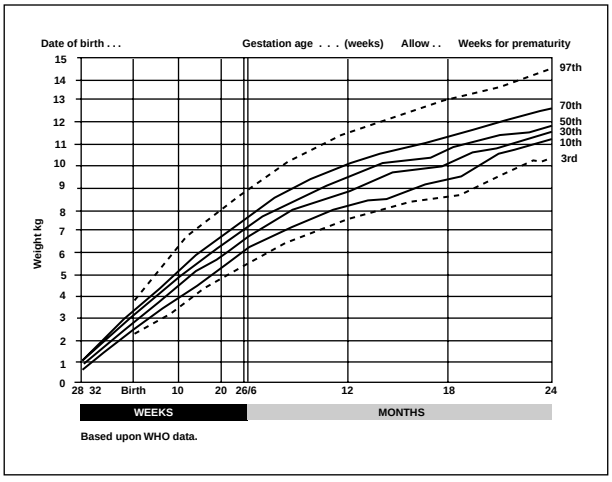


Figure 6.1.3 Girls' weight. Birth to 2 years.

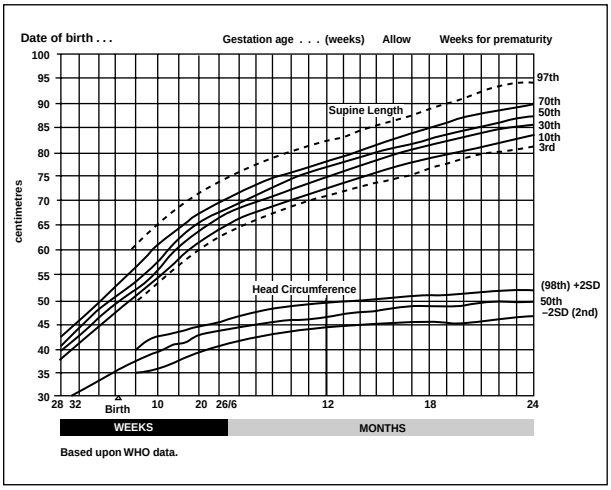


Figure 6.1.6 Boys's supine length and head circumference. Birth to 2 years.

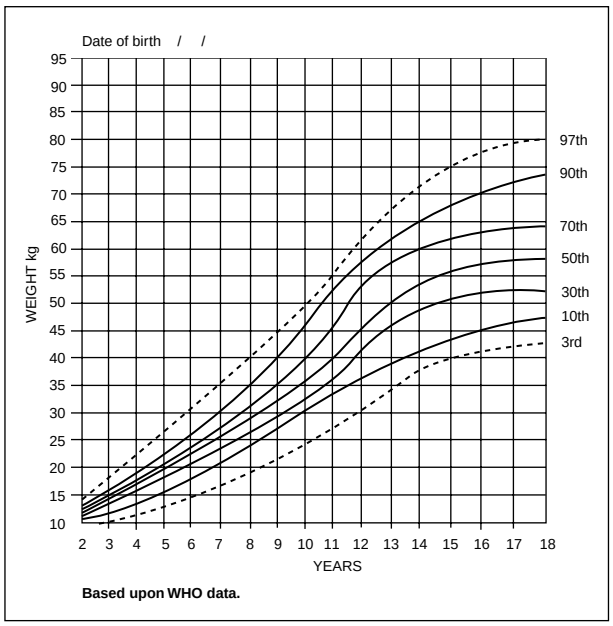


Figure 6.1.7 Girls' weight, 2–18 years.

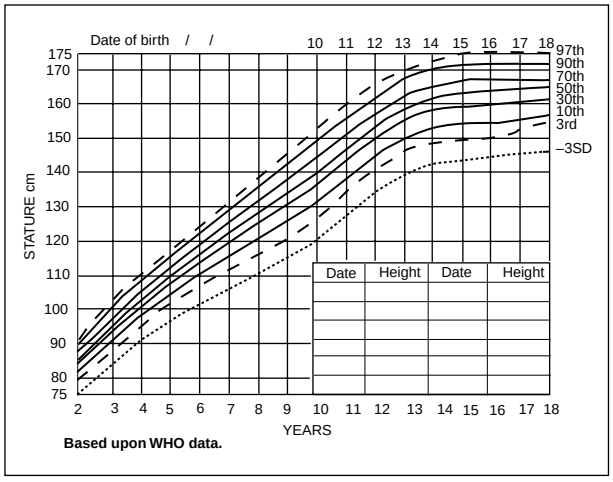


Figure 6.1.8 Girls' stature, 2–18 years.

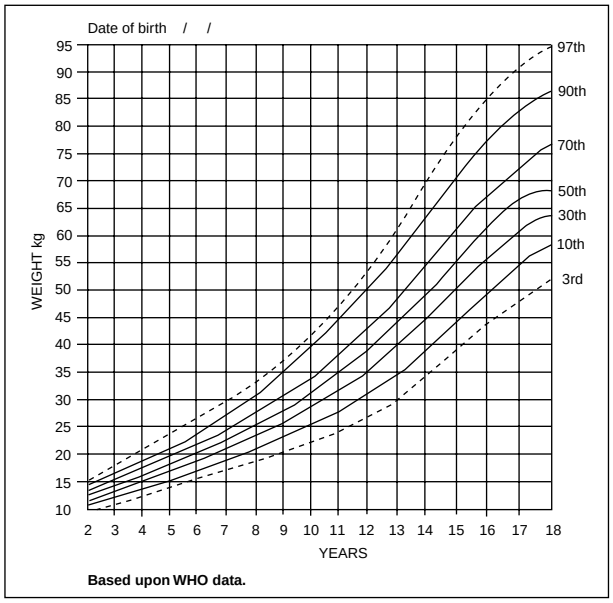


Figure 6.1.9 Boys' body weight, 2–18 years.

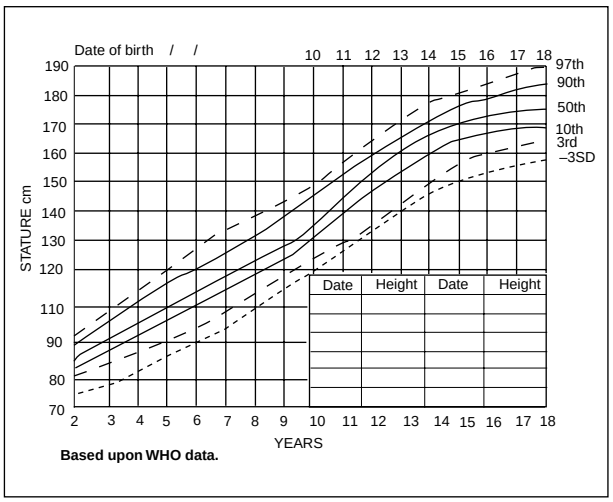


Figure 6.1.10 Boys' stature, 2–18 years.

Assessment of pubertal state

Girls

Record the following

Breast development

Stage 1 Pre-adolescent: elevation of nipple only.

Stage 2 Breast bud stage: elevation of breast and nipple as small mound. Enlargement of areola diameter.

Stage 3 Further enlargement and elevation of breast and areola, with no separation of their contours.

Stage 4 Projection of areola and nipple to form a secondary mound above the level of the breast.

Stage 5 Mature stage: projection of nipple only, due to recession of the areola to the general contour of the breast.

Pubic hair

Stage 1 Pre-adolescent: the vellus over the pubes is no further developed than that over the abdominal wall, i.e. no pubic hair.

Stage 2 Sparse growth of long, slightly pigmented downy hair, straight or slightly curled, chiefly along labia.

Stage 3 Considerably darker, coarser and more curled. The hair spreads sparsely over the junction of the pubes.

Stage 4 Hair now adult in type, but area covered is still considerably smaller than in the adult. No spread to the medial surface of thighs.

Stage 5 Adult in quantity and type with distribution of the horizontal (or classically "feminine") pattern. Spread to medial surface of thighs but not up linea alba or elsewhere above the base of the inverse triangle (spread up the linea alba occurs late and is rated stage 6).

Menarche–age at first period.

Boys

Genital (penis) development

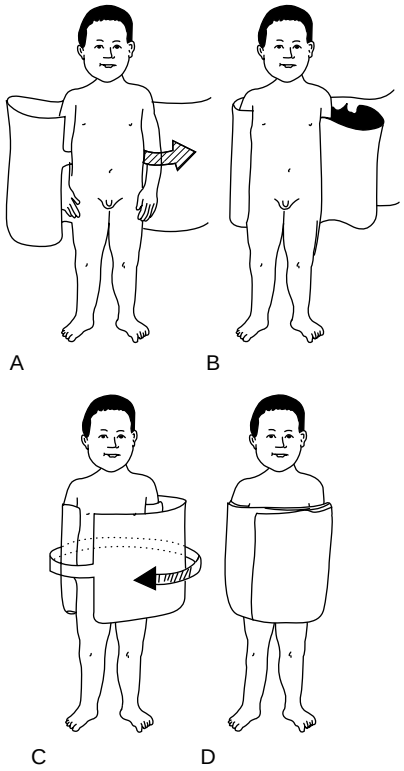
Stage 1 Pre-adolescent, testes, scrotum and penis are of about the same size and proportion as in early childhood.

Stage 2 Enlargement of scrotum and testes. Skin of scrotum reddens and changes in texture. Little or no enlargement of penis at this stage.
Stage 3 Enlargement of penis, which occurs at first mainly in length. Further growth of testes and scrotum.
Stage 4 Increased size of penis with growth in breadth and development of glans. Testes and scrotum larger; scrotal skin darkened.
Stage 5 Genitalia adult in size and shape.

Pubic hair
Stage 1 Pre-adolescent: the vellus over the pubes is no further developed than that over the abdominal wall, i.e. no pubic hair.
Stage 2 Sparse growth of long slightly pigmented downy hair, straight or slightly curled, chiefly at the base of the penis.
Stage 3 Considerably darker, coarser and more curled. The hair spreads sparsely over the junction of the pubes.
Stage 4 Hair now adult in type, but area covered is still considerably smaller than in the adult. No spread to the medial surface of thighs.
Stage 5 Adult in quantity and type. Spread to medial surface of thighs but not up linea alba or elsewhere above the base of the inverse triangle (spread up linea alba occurs late and is rated stage 6).

*Testicular volume**
Stage 1 1.5–3 ml
Stage 2 4–6 ml
Stage 3 6–10 ml
Stage 4 10–12 ml
Stage 5 15–20 ml
*Approximate volume at each genital stage

6.2 Restraining children for procedures



Wrapping the child to hold securely during a practical procedure
One end of a folded sheet should be pulled through under the arms on both sides (A and B). The other end is then brought across the front and wrapped around the child (C and D).

Restraining the child for examination of eyes, ears or mouth

Figure 6.2.1 Restraining children.

6.3 Systems to minimise errors in drug/infusion administration in hospital*

General points

1 Drug vials once reconstituted do not contain preservatives/antiseptic. Multiple sampling from them is potentially hazardous.

2 For infants, dilute drugs to ensure volumes can accurately be measured. For example do not use doses <0.1ml for a 1 ml syringe.

3 Serious errors can occur if the dead space in the hub of the syringe is overlooked during dilution. For example, if the active drug is drawn into a 1 ml syringe up to the 0.1ml mark, the syringe will contain between 0.19 and 0.23 ml. If the syringe is then filled with diluent to 1 ml the syringe will contain approximately twice as much drug as intended. Dilution must involve first half filling the syringe with DILUENT and then adding active drug by using the distance between 2 graduations on the syringe. Mix the two by moving the plunger and then finally add further diluent to total planned volume of active drug and diluent. For dilutions >10 times use a small syringe to inject the active drug connected by a sterile 3-way tap to a larger syringe. Then add diluent to the large syringe to reach desired volume.

4 Many drugs are equally as effective orally as parenterally. Oral administration is safer and less expensive. The following antibiotics are as effective orally as IV in a baby who is taking feeds: amoxycillin, ampicillin, chloramphenicol, ciprofloxacin, co-trimoxazole, erythromycin, flucloxacillin, fluconazole, isoniazid, metronidazole, pyrimethamine, rifampicin, sodium-fusidate, and trimethoprim.

5 If a drug is given down an oro/nasogastric tube, a proportion will remain in the tube unless flushed through.

6 Rectal drugs are less reliably absorbed than oral drugs. Liquid formulations are better than suppositories in infants.

7 When giving IV drugs, do so slowly in all cases. After being injected into the line (ideally through a 3-way tap) the normal IV infusion rate of the fluid going into the cannula can be used to drive the drug slowly into the patient. If there is no background infusion give sufficient follow-up (flush) of fluid, 0.9% saline, sterile water or 5% glucose, to ensure the drug does not remain in the cannula or T piece. Give flush over 2 minutes to avoid sudden surge of drug, (remember the hub). If the IV drug needs to be given rapidly, for example adenosine, use a 2 ml bolus of 0.9% saline via a second syringe not by temporarily increasing the infusion rate (sometimes temporarily becomes prolonged and dangerous).

8 Do not mix incompatible fluids IV.

9 For IV drug infusion (using a syringe/infusion pump) given in addition to background IV infusions:
- Adjust total 24 hour IV fluid intake
- Never allow a surge of a vasoactive drug such as dopamine or epinephrine
- Never put more drug or background IV into syringe or burette than is needed over a defined period of time
- Check and chart rate of infusion and confirm this by examining amount left every hour.

10 Intramuscular injections need special precautions:
- IM injections are unsafe in shock and especially with opiates, for example a high dose can be released once recovery of the circulation occurs
- To avoid nerve damage, only the anterior aspect of the quadriceps muscle in the thigh is safe in a small wasted infant <1 year of age
- Alternate between legs if multiple injections required
- Do not give IM injections if a severe bleeding tendency is present
- It is essential to draw back the plunger to ensure that the needle is not in a vein before injecting potentially dangerous drugs (for example epinephrine or lidocaine).

Care of intravascular lines

- Placement of central venous lines: check with a lateral *X* ray that line is placed well into a major vein and if near heart with tip ideally in SVC at the entrance to the right atrium.
- Placement of an umbilical arterial line should either be above the diaphragm in the thoracic aorta or below the 2 renal arteries (at L4) to minimize the risk of renal or mesenteric artery thrombosis.
- All arterial lines can result in life threatening haemorrhage or occlusion leading to ischaemia. Procedures to ensure these complications do not occur should be in place.
- Never give a drug into a drip that has started to issue. Some drugs for example those containing calcium, can cause severe scarring. Inspect the cannula tip before and whilst injecting any drug IV.
- Local infection can become systemic, especially in neonates or in the immunosuppressed. Always remove cannula if there is erythema in tissue around it and if lymphangitis is seen. If lymphangitis is present always take a blood culture from a separate vein and start IV or IM antibiotics. Always place cannulae aseptically and keep the site clean. There is no evidence that frequent changes of cannula site or infusion kit are of benefit. However, it is a good idea to change the giving set after blood transfusion or if a line of blood has entered the infusion tubing from the vein and clotted there. It can act as a site for bacterial colonization. Otherwise change lines every 3 or 4 days (lipid solutions every 48 hours).
- Air embolism – if air reaches the heart, unlike blood it stays there, especially if the patient is lying flat. Unless

*Adapted from *Neonatal formulary(3). The Northern Neonatal Network*, 11th ed. London: BMJ Books, 2000.

immediately aspirated, air in the heart can block the circulation. Umbilical venous and other central venous lines are particularly dangerous. There must be a tap or syringe on the catheter at all times, especially during insertion. An alternative source of air embolus is through the giving set, especially when pumps are being used.

- Blood loss. In neonates this can occur from the umbilical stump. From central venous or arterial lines, it can rapidly be fatal, and therefore all connections must be luer locked and the connections to the cannula and its entry must be observable at all times. Ideally arterial lines should be connected to a pressure transducer and alarm.

Use of IV/IA (intra-arterial) access

- When sampling from an IV/IA line, clear the dead space first (by 3 × its volume). Blood glucose levels cannot be accurately measured from any line through which a glucose solution is infused, even if many times the dead space has been cleared.
- For blood culture, always use a separate, fresh, venous needle, stab sample.
- Never add anything to a line carrying total parenteral nutrition (TPN).
- Certain infusions, such as glucose >10%, epinephrine, and dopamine are better through a central vein. In an emergency, dopamine and epinephrine infusions can be given through a peripheral vein.
- When a continuous infusion is not required, a peripheral cannula can be stopped off with a sterile bung after flushing the drug in with 0.9% saline, sterile water or 5% glucose to clear the dead space (there is no evidence that a heparin lock is needed for cannula in peripheral veins).
- Central venous catheters must be firmly anchored to the skin so they do not migrate into or out of position. After individual drug injections and without continuous infusion, a heparin lock is appropriate to prevent clotting of the line (10 units of heparin per 1 ml of 0.9% saline) particularly in double, triple or quadruple lumen catheters (always use luer lock connections to minimise extravasation).
- Peripheral artery lines should never be used for giving drugs. To maintain patency a continuous low rate (0.5 – 1.0 ml/hour) infusion of heparinised 0.9% or 0.18% saline is useful (heparin at 1 unit/ml). Clear the 1 ml dead space of the catheter before and after sampling, which must be done aseptically.
- In neonates and infants, frequent flushed can result in sodium overload and therefore consider using 0.18% saline or sterile water to achieve this.
- Central arterial lines (usually in the aorta) can be safely used to give glucose or TPN if the catheter tip site is checked radiologically (not near mesenteric or renal arteries). Most drugs (except inotropes) can also be safely given by this route by slow infusion (not by boluses).
- Do not add drugs to any line containing blood or blood products.
- Most IV drugs can be given into an infusion containing 0.9% saline or up to 10% glucose (exceptions include amphotericin B, phenytoin, and erythromycin).
- If only one line is being used for an infusion and more than one drug needs to be given, try to wait 10 minutes between them. If not possible, separate by 1 ml of 0.18% saline/4% glucose, 0.9% saline or sterile water for injections. This is very important with an alkaline drug such as sodium bicarbonate. Always give the flush slowly over at least 2 minutes to ensure that the drug already in the line/vein does not move forward in the patient in a sudden rapid surge, (especially if the catheter/vein contains an inotrope or vasoactive drug such a aminophylline, cimetidine, phenytoin or ranitidine which can cause an arrhythmia).
- When two IV drugs need to be given together and there is only one IV catheter, terminal co-infusion using a T or Y connector next to the catheter can be used. It is important to know whether this is safe for the drugs in question.

Minimising IV infusion and IV drugs errors

Errors of commission and omission occur. For example excess IV fluids can be dangerous by circulatory overload and inadequate IV fluids can be dangerous through hypoglycaemia (especially in the neonate and commonly when a blood transfusion is being given and the infant is relying on IV glucose). Extravasation can also result in the absence of a vital drug (such as morphine infusion for pain).
Errors will always occur with human actions, and systems in place to minimise these are essential.

Steps to reduce errors and their impact

1 Prescribe or change infusion rates as infrequently as possible: once or twice daily.
2 Never have > one IV infusion line running at the same time unless absolutely necessary.
3 Use a burette in which no more than the prescribed volume is present (especially in infants and young children).
4 Record hourly the amount given (from burette, syringe or infusion bag) and the amount left.
5 Check the infusion site hourly to ensure extravasation has not occurred.
6 Ensure that flushes are only used when essential and are given slowly over at least 2 minutes.
7 Ensure that flushes do not overload the patient with sodium.
8 Be particularly careful with potassium solutions given IV (use enteral route wherever possible).
9 Check and double check the following:
   - Is it the right drug? Check ampule as well as box
   - Is it the right concentration?
   - Is shelf life within expiry date?
   - Has it been constituted and diluted correctly?
   - Is it for the right patient?

- Is the dose right (2 persons to check the prescription chart)?
- Is it the correct syringe (deal with one patient at a time)?
- Is the IV line patent?
- Is a separate flush needed? If so has the flush been checked?
- Are sharps disposed of (including glass ampules)?
- Has it been signed off as completed (ideally countersigned)?

Writing a prescription

- Use block capitals.
- Use approved names.
- Dosage should be in grams (g) milligrams (mg) or micrograms. **ALWAYS WRITE MICROGRAMS IN FULL**.
- Volumes should be in millilitres (ml).
- Avoid decimal places whenever possible. If not possible preface by a zero, thus write 500 mg not 0.5 g. If decimal is used write 0.5 **NOT** .5 ml.
- Write times using 24 hour clock.
- Routes of administration can be abbreviated to: IV (intravenous), IM (intramuscular), PO (orally), SC (subcutaneous), NEB (nebuliser), RECT (rectally).
- As required prescriptions must be specific as to how much, how often and for what purpose (indicate maximum 24 hour dose).
- Each drug should be signed for individually by a registered doctor.
- Stop dates for short course treatments should be recorded when first prescribed.

This chapter has been adapted from the following book:
The Northern Neonatal Network
Neonatal Formulary (3) London: BMJ Books, 2000

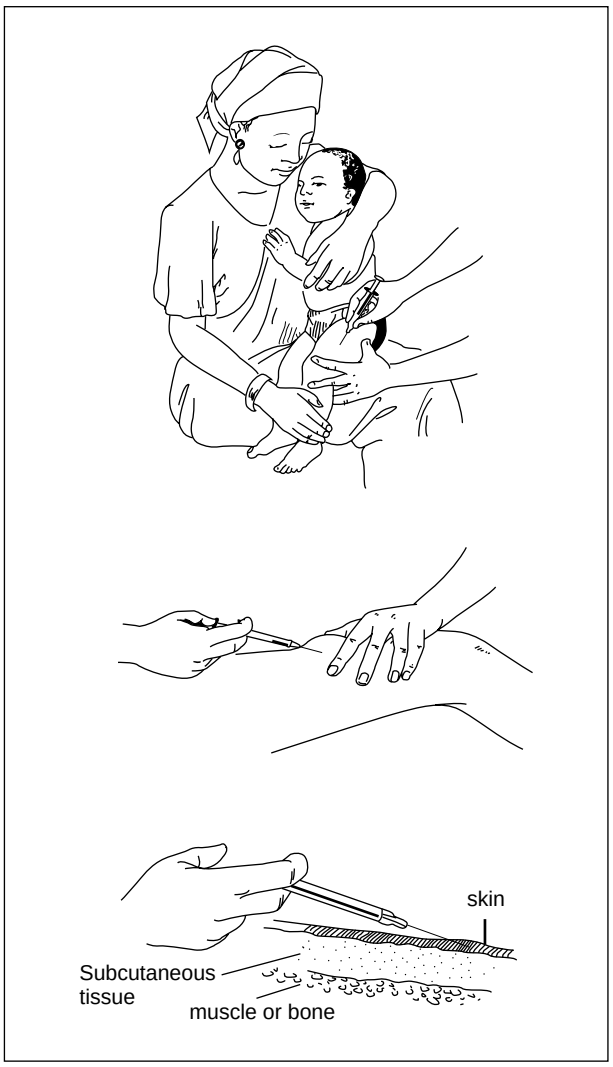


Figure 6.4.1 (Top) Intramuscular injection. (Middle) Subcutaneous injection. (Bottom) Intradermal injection.

6.4 Giving injections

Intramuscular injections

In children over 2 years old, give the injection in the upper, outer quadrant of the buttock. Choose the site carefully, well away from the sciatic nerve. In younger or severely malnourished children, use the outer side of the thigh midway between the hip and the knee, or over the deltoid muscle in the upper arm. Hold the muscle at the injection site between the thumb and first finger and push the needle (23–25 gauge) into the muscle at a 90° angle (45° angle in the thigh). Draw back the plunger to make sure there is no blood (if there is, withdraw slightly and try again). Give the drug by pushing the plunger slowly till the end. Remove the needle and press firmly over the injection site with a small swab or cotton wool.

Subcutaneous injections

Select the site, as described above, for intramuscular injection. Pinch up skin and subcutaneous tissue between finger and thumb. Push the needle (23–25 gauge) under the skin at a 45° angle into the subcutaneous fatty tissue. Do not go deep to enter the underlying muscle. Draw back the plunger to make sure there is no blood (if there is, withdraw slightly and try again). Give the drug by pushing the plunger slowly till the end. Remove the needle and press firmly over the injection site with cotton wool.

Intradermal

Select an area of skin, which has no infection or damage for the injection (for example over the deltoid in the upper arm). Stretch the skin between the thumb and forefinger of one hand; with the other, slowly insert the needle (25 gauge), ***bevel upwards***, for about 2 mm just under and almost parallel to the surface of the skin. Considerable resistance is felt when injecting intradermally. A raised, blanched bleb showing the surface of the hair follicles is a sign that the injection has been given correctly.

6.5 Peripheral venous cannulation

- Prepare kit:
  - 18–25 gauge intravenous cannula or butterfly needles
  - 2 ml syringe and "T-piece" containing 0.9% sodium chloride for flushing
  - Tape or plaster of Paris for scalp veins
  - Small splint (can be made from wooden spatula covered with gauze)
  - Alcohol swabs for skin cleaning
  - Local anaesthetic cream if available
  - Tourniquet (or assistant).
- Cannula size:
  - Neonate 24–25 gauge
  - Infant 22–24 gauge
  - Child 20–22 gauge
  - Adolescent 18–20 gauge.

  Apply tourniquet to distend vein.
- Choose vein:
  - Forearm
  - Long saphenous (anterior to medial malleolus)
  - Back of hand or front of wrist
  - Scalp.

Useful sites to cannulate include dorsum of feet and hands. The saphenous and antecubital veins are larger but can be useful for percutaneously inserted "long lines". The antecubital veins are also useful for venepunctures for laboratory studies. If possible, place cannula close to bone where it is more fixed.

- Decide direction of blood flow.
- Clean skin with antiseptic.
- Fix and slightly stretch skin with other hand.
- Pass cannula through skin at slight (10–20°) angle – ***be decisive***.
- Stop once through skin.
- Flatten cannula to skin and advance with long axis of cannula in the same direction as the vein – ***be decisive***.
- Aim to pass into vein at first attempt with steady advancement.
- Always watch for blood appearing in hub of cannula.
- As soon as blood seen *stop* (larger cannulae: advance a couple more millimetres).
- Hold needle still, advance cannula over needle until hub at skin.
- Hold cannula still.
- Withdraw needle.
- Connect connector, flush and fix. No subcutaneous swelling should be seen and there should be no resistance to injection.
- If no blood seen on advancing cannula but felt to be beyond vein, stop.
- Gently pull cannula back in same direction as advancement; if blood appears, stop once again.
- Follow procedure as if blood seen on first advancement (transfixion technique).
- Connect the T-piece and flush the cannula gently with saline to confirm that it is in the vein.
- If the cannula is satisfactorily inserted, tape it in place by looping a thin piece of the tape under the hub and round to form a "V" shape fixing it to the skin.
- When splinting try to "double back" the tape, i.e. put a short and a long piece back to back leaving just the ends of the longer piece sticky. This helps prevent excessive amounts of tape sticking to the baby, particularly important in more immature babies whose skin is easily damaged.

Note on flushing lines: the smaller the syringe used, the greater the pressure exerted on fluid in the line. Therefore avoid using 1 ml syringes to flush a blocked line – the line may rupture or damage tissue by infiltration.

Blood sampling from intravenous cannulae

If the patient needs blood samples at the time of cannulation it is often possible to take these as the cannula is inserted. Blood can be dripped from the end of the cannula into the appropriate bottles or a syringe can be used to ***gently*** aspirate blood from the cannula. If the cannula has been flushed with saline prior to insertion, the first 0.5–1 ml of blood should be discarded.

Special sites for intravenous cannulae

Scalp veins

Procedure

1 Restrain the child.
2 Shave the appropriate area of the scalp with a *sterile* razor.
3 Clean the skin.
4 Have an assistant distend the vein by holding a taut piece of tubing or bandaging perpendicular to it, proximal to the site of puncture.
5 Fill the syringe with 0.9% saline and flush the butterfly set.
6 Disconnect the syringe and leave the end of the tubing open.
7 Puncture the skin and enter the vein. Blood will flow back through the tubing.
8 Infuse a small quantity of fluid to see that the cannula is properly placed and then tape into position.

Scalp drips are generally more precarious than ones in the limbs and need to be carefully observed. Infiltration into the soft tissues of the scalp can spread quickly and cause extensive necrosis if irritant. Shave the hair from an area about 2–3 cm around the site selected in order to allow for fixation by tape. Always ensure that the tip of the needle is not covered by dressings, so that infiltration is quickly seen.

External jugular vein

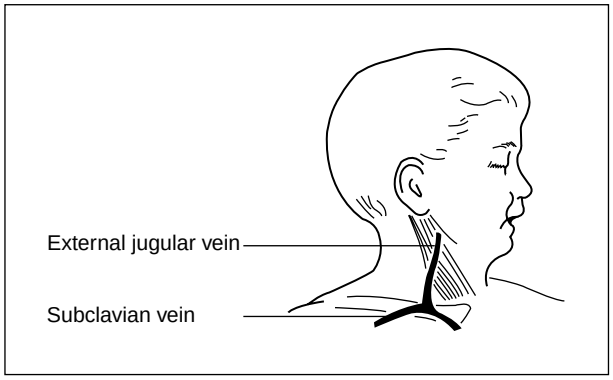


Figure 6.5.1 External jugular vein and the subclavian vein.

Procedure

1 Place child in a 15–30° head-down position (or with padding under the shoulders so that the head hangs lower than the shoulders).
2 Turn the head away from the site of puncture. Restrain the child as necessary in this position.
3 Clean the skin over the appropriate side of the neck.
4 Identify the external jugular vein, which can be seen passing over the sternocleidomastoid muscle at the junction of its middle and lower thirds.
5 Have an assistant place his or her finger at the lower end of the visible part of the vein just above the clavicle. This stabilises it and compresses it so that it remains distended.
6 Puncture the skin and enter the vein.
7 When free flow of blood is obtained, ensure no air bubbles are present in the tubing and then attach a giving set.
8 Tape the cannula securely in position.

Safe intravenous infusions where no burettes are available

- Mark the infusion bottle with tape for each hour to be given and label each hour.
- Alternatively empty until only the necessary amount to be given is left in the bottle.

6.6 Central venous cannulation

Aims

- Venous access when peripheral cannulation is not possible (in an emergency, intraosseous cannulation is faster and easier).
- Monitor central venous pressure
- Prolonged vascular access
- Large-bore vascular access
- Administer certain drugs or concentrated glucose solutions
- During resuscitation

Procedure

Several routes are possible but most widely used are the femoral and internal jugular. The femoral approach is easiest in the emergency situation. A subclavian approach may be useful in the older child.

Basic technique

- Sterile pack
- Sterile Seldinger wires
- Cannula: single 16–22G cannula
- Single, double or triple lumen if available (5 FG 5–8 cm length for neonate, 7 FG 8–15 cm length child)
- Syringe and N saline
- Suture and tape to fix
- Local anaesthetic with fine 25 G needles

Prepare child

- Explanation (if conscious)
- Position
- S*terilise skin and maintain sterile technique*
- Local analgesia to skin (if conscious)

Two insertion techniques are available namely:

- As peripheral cannulation
- Seldinger technique (wire).

Some use cannula for both techniques (improved safety, improved identification of vein). Others use only needle to enter vein for Seldinger method.

Seldinger method

- Identify vein with cannula over needle attached to syringe (same approach as for peripheral cannulation) or just needle; keep drawing back on syringe as needle is advanced. As vein is entered, blood flows back.
- Stop, pass cannula over needle or disconnect syringe.
- Disconnect syringe.
- Pass wire through cannula to ensure far enough into vein (any resistance, stop; withdraw wire with needle and start again); holding wire firmly, withdraw cannula over wire. Consider more local anaesthetic along first 1 cm of wire.
- Pass dilator over wire (sometimes a need to make a small cut at the skin); holding wire firmly, withdraw dilator.
- Pass cannula/catheter filled with 0.9% saline over wire (passage of cannula should be smooth, meeting no resistance).
- Hold cannula, withdraw wire gently, if sticks do not force.
- Confirm correct placement by aspiration of blood.

- Suture and fix with antiseptic ointment over entry site.
- Confirm position with *X* ray.

Femoral cannulation

This is adequate for almost all needs, technically much easier and with lower complication rates, particularly in neonates and infants. However, if not sterile there is a risk of causing septic arthritis in the hip joint.

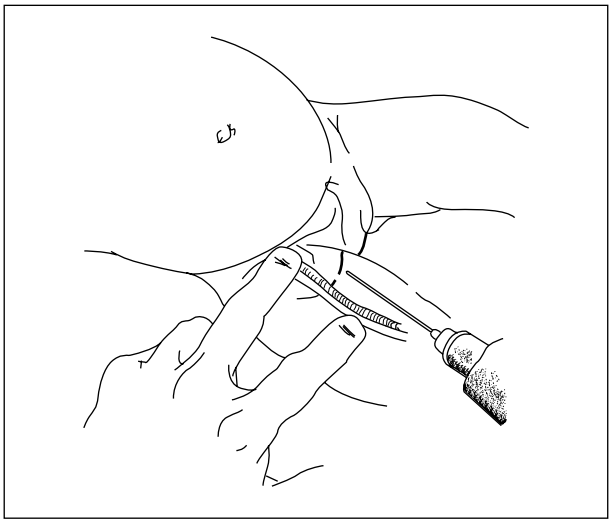


Figure 6.6.1 Femoral cannulation.

1 Position the patient supine with the leg slightly abducted. Place towel under buttocks to raise pelvis.
2 Clean the skin and drape with sterile towels. Locate the vein by finding the femoral arterial pulsation 2 cm below the midpoint of the inguinal ligament. The vein lies immediately medial to the artery. Infiltrate the skin with local anaesthetic.
3 With a finger on the femoral artery, introduce the cannula needle with syringe attached at 45° to the skin along the line of the vein pointing towards the umbilicus. Advance the needle whilst aspirating.
4 When blood "flashes back" into the syringe, stop advancing and remove the syringe from the needle. Feed the Seldinger guide wire through the needle cannula keeping hold of one end of the wire at all times.
5 Withdraw the needle over the wire, then after dilatation (see above) feed the catheter over the wire into the vein.
6 Withdraw the wire and aspirate for blood to confirm position. Then flush the catheter with saline.
7 Suture the catheter in place.

Note: If unsure whether vein or artery consider transducing pressure waveform.

Internal jugular cannulation

Head down for internal jugular and subclavian approaches – increases vein distension and reduces risk of air embolism.

1 Position child 30° head-down and turn head to left-hand side for the right-sided approach which avoids the lymphatic duct. Place towel or roll under shoulders to extend the neck.
2 Clean the skin and drape with towels exposing the neck to the clavicle.
3 Identify the apex of the triangle formed by the two heads of the sternocleidomastoid and clavicle and infiltrate local anaesthetic (if conscious). Alternatively, identify carotid pulsation medial to sternomastoid at level of lower border of thyroid cartilage; vein is just lateral to this (usually); aim needle at ~30° to skin and towards the ipsilateral nipple (the neck is very short and vein is superficial in the very young). Estimate the length of catheter from the skin entry to the nipple.
4 Direct the needle at 30° to the skin pointing towards the right nipple and puncture the skin at the apex of the triangle.
5 Holding this position, advance the needle, aspirating all the time. If blood "flashes back" stop advancing and remove the syringe from the needle. (If you do not cannulate the vein, withdraw the needle (but not out of the skin) and advance again slightly more laterally).
6 Feed the Seldinger guide wire through the needle, always having control of one end of the wire.
7 Withdraw the needle over the guide wire and then, after dilator, feed the catheter over the wire into the superior vena cava (ensure all outlets are in the vein).
8 Withdraw the wire, aspirate for blood and attach the infusion set. **Do not leave the catheter open as this may lead to an air embolism**.
9 Suture the catheter in place and obtain a chest *X* ray (if possible) to check for a pneumothorax and the position of the catheter tip which should be in the superior vena cava, ideally at the superior vena cava/right atrial junction, but not in the right atrium.

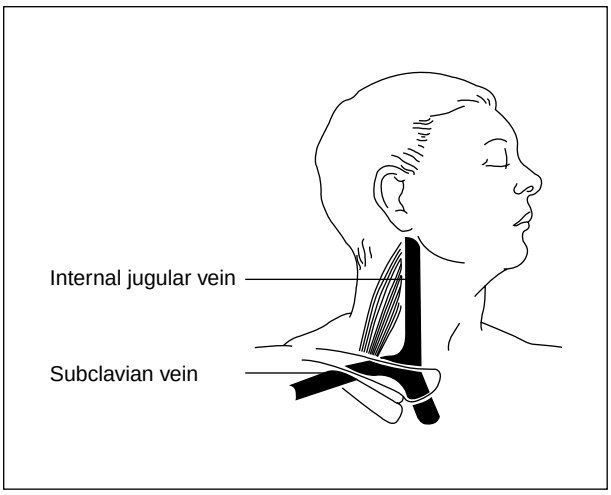


Figure 6.6.2 Internal jugular vein and subclavian vein.

Subclavian cannulation

- Place supine, turn head to contralateral side, put roll to extend neck a little, identify midpoint of clavicle.

- Aim for suprasternal notch, pass needle just beneath clavicle at midpoint (more medial in older child); the vein lies anterior to the subclavian artery and is closest at the medial end of the clavicle.
- Subclavian artery puncture is not uncommon (cannot compress to stop bleeding but is rarely a problem unless coagulopathy is present).

Complications of central venous cannulation

- Fewer and less severe in femoral cannulation
- Arterial puncture
- Nerve damage
- Pneumothorax
- Extravasation of administered fluids/drugs
- Septicaemia if not sterile or if cannula in place for more than five days

6.7 Cut down venous cannulation

Indication

Continuous intravenous access where percutaneous attempts have failed. (In the emergency situation, intraosseous access is faster and easier.)

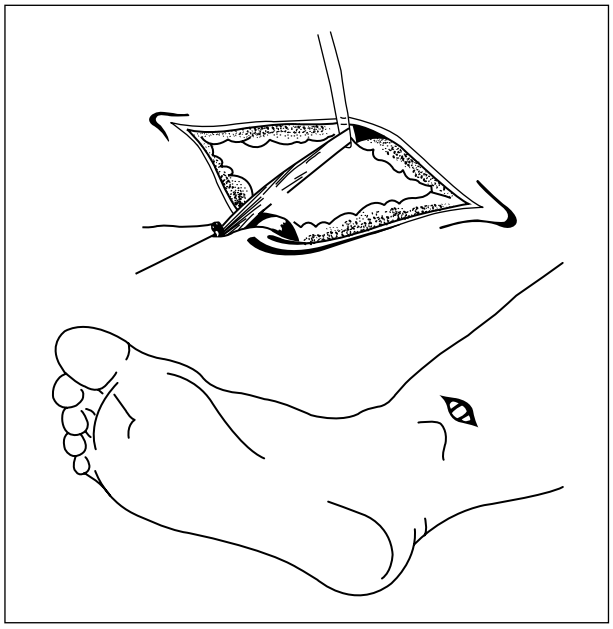


Figure 6.7.1 (Top) Tethering of vein. (Bottom) Long saphenous cut down site.

Equipment

Skin prep (iodine, alcohol)	Local anaesthetic
Scalpel	Curved artery forceps
Suture	Syringe and hypodermic needle
IV cannula	Sterile drapes

Procedure

- Identify landmarks.
  - *Brachial*

    Infant — One fingerbreadth lateral to the medial epicondyle of the humerus

    Small child — Two fingerbreadths lateral to the medial epicondyle of the humerus

    Older child — Three fingerbreadths lateral to the medial epicondyle of the humerus
  - *Saphenous*

    Infant — Half a fingerbreadth superior and anterior to the medial malleolus

    Small child — One fingerbreadth superior and anterior to the medial malleolus

    Older child — Two fingerbreadths superior and anterior to the medial malleolus
- Immobilise limb and apply tourniquet above site at pressure between venous and arterial.
- Clean skin and drape with sterile towels.
- Infiltrate local anaesthetic into skin after marking the site of the vein (if conscious).
- Incise the skin perpendicular to the long axis of the vein.
- Bluntly dissect the subcutaneous tissues with the curved artery forceps (tips pointing downwards) parallel to the vein. With tips pointing up, scoop up the tissues and open the forceps – you should have picked up the vein! Clear about 2 cm of vein from surrounding tissue.
- Pass a proximal and distal ligature around vein. Tie only the distal ligature and use for traction.
- Make a small hole in the vein with the scalpel proximal to the tied ligature and feed the catheter into the vein proximally (ideally up to the hub). Tie the proximal ligature around vein and catheter.
- Aspirate blood (if blood does not aspirate you may be against the vein wall so pull back a little and repeat) and flush with normal saline.
- Close incision with interrupted sutures, place antiseptic ointment (for example iodine) over wound, and suture the catheter to the skin (ensure local anaesthetic at suture site if conscious). Cover with dressing.

6.8 Umbilical vein catheterisation

Indications

- Where there is urgency during resuscitation of the newborn to give intravenous fluids and drugs.
- Temporarily for exchange transfusion – the catheter should not be left in position after exchange.

Time of insertion

Catheterisation is usually easy in the first seven days of life.

Equipment

- Gown and gloves.
- Sterile instruments including:
  Fine scissors
  Forceps
  Scalpel
- Silk suture for retaining.
- 5 French gauge umbilical catheter (a sterile feeding tube may be satisfactory if this is not available but measure the length first so that you will know how much you have passed by measuring the length from the hub to the umbilical insertion. Cannulae designed for use as UVCs are usually marked in 5-cm steps).
- Three-way tap.
- 0.5% chlorhexidine or 10% povidone-iodine for cleaning skin.
- Sterile cotton wool balls.
- Sterile towels or drapes to cover baby's abdomen.
- Sterile 2 ml syringe and connector filled with 0.9% saline.

Procedure

- Assemble the syringe, three-way tap and catheter. Flush and fill the catheter with sterile 0.9% saline.
- Clean the umbilical cord and surrounding skin with 0.5% chlorhexidine or 10% povidone-iodine.
- Cut back cord cleanly with scalpel to about 2 cm from base.
- Cover with towels to from a sterile working surface.
- Hold the cord at an edge with forceps.
- Identify the vein – usually gaping, larger, and well separated from the two small thicker-walled arteries (see Figure 6.8.1).

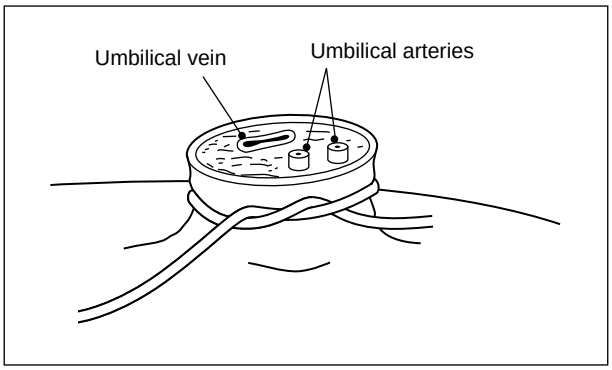


Figure 6.8.1 Cross section of cord.

- Hold the catheter approximately 2 cm from the end with forceps and insert the tip into the vein. Gently advance the catheter which should pass easily.
- For resuscitation and exchange insert 4–6 cms.
- For longer term place the catheter into the IVC at junction with right atrium. It should be advanced {approximately (2 × weight in kg) + 5 + length of stump} in centimetres (usually same length as from umbilicus to internipple line). Check blood can be drawn back easily – if not withdraw slightly until blood flows.
- Sometimes the catheter will not pass easily through the ductus venosus or deviates into a lobe of the liver. Therefore (if available) obtain an *X* ray and check the position of tip before catheter is used. Ideally it should be at the IVC/RA junction, but a position below the liver will also be acceptable for short-term use (4–6 cms in). The catheter should **not** be in the liver. Either L_3–L_4 or T_8 positions.
- The catheter can be secured by winding a suture round it several times and then passing a stitch through the cord base. An additional safeguard is to form two wings of tape which can then be taped to the abdominal wall – always remembering that it is preferable to use as little tape as possible in smaller babies. It is essential that the catheter does not fall out (see Figure 6.8.2).

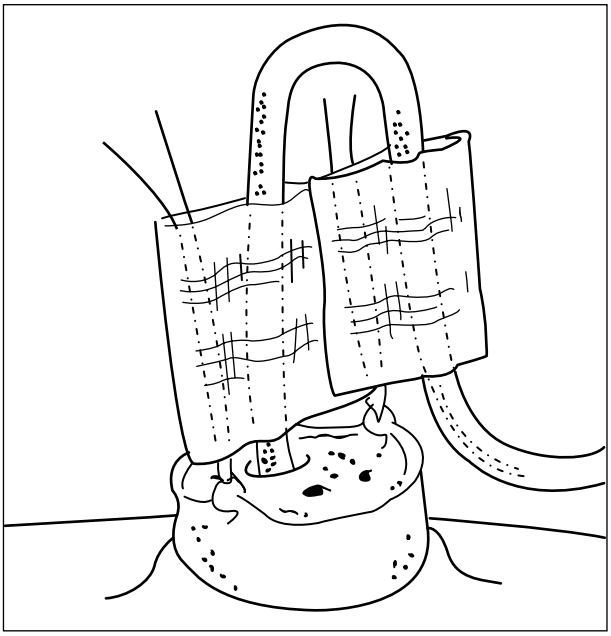


Figure 6.8.2 Fixation of catheter in the umbilicus.

Occasionally the umbilical vein is kinked and advance of the catheter is blocked at 1–2 cm beyond the abdominal wall. Gentle traction on the cord usually relieves this.

If obstruction occurs at more than 2 cm, and only partly gives way with pressure, the catheter is probably either wedged in the portal system or coiled up in the portal sinus. It is advisable to withdraw the catheter part way and reinsert.

Care of indwelling catheters

Leave the cord exposed to air. Remove blocked catheters.

Removal of catheter

- Sterile technique.
- Remove a specimen of blood for culture.
- If possible place a purse string suture about the vessel at the base of the umbilicus and withdraw the catheter slowly.
- Tighten purse string suture.
- Compress with dressing for 10 minutes.

Time of removal of catheter

Remove the catheter as soon as possible as dictated by the clinical state of the baby. The infection rate rises after 24 hours. Complications are more common with venous than arterial catheter, so venous catheters should rarely be left in.

Complications

- Thrombosis: survivors may develop portal vein thrombosis.
- Embolism.
  - From clots in catheter.
  - From injected air.
- Vascular perforation.
- Vascular damage from hypertonic solutions.
  More common when the tip is in the portal system.
- Haemorrhage from disconnected catheter.
- Necrotising enterocolitis or bowel perforation may occur as a complication of exchange transfusion.
- Infection.
  There is no evidence that prophylactic antibiotics are of any value.

6.9 Exchange transfusion

Equipment

- As for UVC (see Chapter 6.8).
- O rhesus-negative blood or blood cross-matched against **maternal** antibodies.
- Ideally, a blood warmer (especially for low birth weight infants) otherwise warm under mother's dress next to skin.
- Syringe of suitable size (see below).
- Four-way (or two three-way) tap(s).
- Bottle or bag for waste blood.

Technique

- **Note**: although the potassium concentration of the blood is often 8–10 mmol/litre this does not usually cause significant hyperkalaemia.
- Plan to spend **at least 2 hours** on an exchange transfusion. You will need an experienced observer to monitor the baby and record each aliquot of blood withdrawn and replaced.
- Pass the UVC as described above and check its position with an *X* ray (if possible). Ideally it should be positioned at the IVC right atrial junction, but a position below the liver is also acceptable if the line will sample and flush easily. A line positioned **in the liver** should not be used.
- Connect the four-way tap to the UVC – put a syringe on the second port and then run one line to the donor blood infusion set and another to the waste bottle.
- Decide on the size of aliquot you will be exchanging with each draw and infusion. Roughly:
  Baby under 1500 g 5 ml
  Baby 1500 to 2500 g 5 to 10 ml
  Baby over 2500 g 10 to 15 ml

  If you use small aliquots remember to add an allowance for the "dead space" in tubing between the syringe and the baby.
- You should draw out each aliquot over 2–3 minutes and replace over 3–4 minutes with the observer keeping a running total.
- Aim to exchange double the baby's blood volume: that is ($2 \times 80 \times$ wt. in kilograms) millilitres.
- Send the first aliquot for bilirubin, electrolyte and calcium concentration.
- Half-way through the procedure check the blood glucose, calcium and potassium concentrations, pH and base deficit.
- Measure them again, together with the bilirubin concentration, at the end of the procedure. Sometimes it is necessary to exchange more than once in quick succession.

6.10 Intraosseous needle insertion

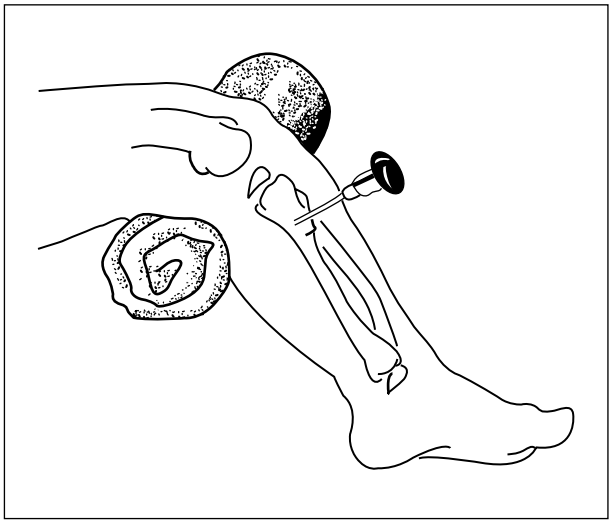


Figure 6.10.1 Intraosseous needle insertion.

Indication

In emergency when other attempts at intravenous access have failed.

Equipment

- Skin prep (iodine, alcohol)
- 18 gauge intraosseous needle (at least 1.5 cm in length)
- Syringes (5 ml and 20 ml)
- Infusion fluid

Procedure

1 Suitable site (flat anteromedial surface of tibia, 2–3 cm below tibial tuberosity or anterolateral surface of femur, 3 cm above the lateral condyle). (Avoid bones where there are fractures proximal to the insertion site.)
2 Clean skin and position knee flexed at 30° over a towel. Grasp the limb firmly.
3 Insert needle at 90° to the skin with a rotating action. When you feel a sudden "give" you have entered the medullary cavity. The needle should stand up by itself.
4 Withdraw trochar and aspirate with 5 ml syringe to confirm position. Send aspirate for cross-matching of blood if needed. Flush with saline to expel clots and observe for subcutaneous swelling. Infuse fluid boluses with 20 ml syringe.
5 Secure intravenous access as soon as possible. When needle is removed cover with sterile dressing.

6.11 Emergency thoracocentesis

Needle thoracocentesis can be life saving and can be performed quickly with minimum equipment. It should be followed by chest drain placement.

Minimum equipment

- Swabs for disinfecting the skin.
- Large over-the-needle intravenous cannula (16 gauge or 20–22 gauge in preterm infants).
- 20 ml syringe.

Procedure

- Identify the second intercostal space in the midclavicular line on the side of the pneumothorax (the ***opposite*** side to the direction of tracheal deviation).
- Swab the chest wall with surgical preparation solution or an alcohol swab.

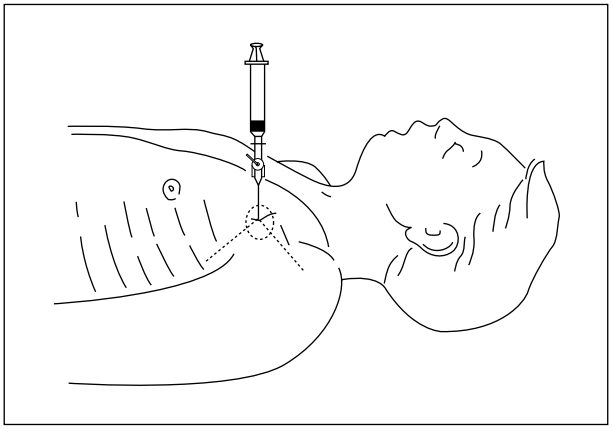


Figure 6.11.1 Needle thoracocentesis.

- Attach the syringe to the cannula.
- Insert the cannula vertically into the chest wall, just above the rib below, aspirating all the time (Figure 6.11.1).
- If air is aspirated remove the needle, leaving the plastic cannula in place.
- Tape the cannula in place and proceed to chest drain insertion as soon as possible.

If needle thoracocentesis is attempted, and the patient does not have a tension pneumothorax, the chance of causing a pneumothorax is 10–20%. Patients who have had this procedure should ideally have a chest radiograph, and may require chest drainage if subsequently ventilated.

6.12 Insertion of a chest drain

Chest drain placement should be performed using the open technique described here. This minimises lung damage. In general, the largest size drain that will pass between the ribs should be used.

Minimum equipment

- Skin prep and surgical drapes
- Scalpel with fine straight blade
  - Blunt forceps
  - Artery forceps
  - Large clamps × 2
- Suture
- Local anaesthetic if child is conscious
- Scissors
  - Chest drain tube
  - Underwater seal or Heimlich flutter valve

Procedure

1 Wash hands and arms to elbows, and wear mask, surgical hat (bonnet), sterile gown and sterile surgical gloves.
2 Prepare the underwater seal with an assistant and take the sterile end of the tube, ready to connect to the chest tube once inserted. The "seal" end should be covered by no more than 1–2 cm H_2O.
3 Decide on the insertion site (usually the fourth or fifth intercostal space in the anterior or midaxillary line) on the side with the pneumothorax (Figure 6.12.1).
4 Swab the chest wall with surgical prep or an alcohol swab.
5 Use local anaesthetic if the child is conscious. Morphine (100 micrograms/kg IV over 10 minutes) should also be given if the child is conscious but in the preterm infant who is not ventilated this may precipitate apnoea. Facilities to provide bag and mask

ventilation and/or intubation should be immediately available together with staff trained in their use.

6 Make a 1–3 cm skin incision along the line of the intercostal space, **immediately above the rib below to avoid damage to the neurovascular bundle which lies under the inferior edge of each rib**.

7 Bluntly dissect using artery forceps through the subcutaneous tissues just over the top of the rib below, and puncture the parietal pleura with the tip of the forceps.

8 Put a gloved finger into the incision and clear the path into the pleura (Figure 6.12.1). This will not be possible in small children.

9 Holding the chest drain about 1 cm from the end pass it into the hole you have made – it should now thread in easily. Pass about 3 cm into the pleural cavity then connect to the underwater seal. Ideally advance the chest drain tube into the pleural space during expiration.

10 Ensure the tube is in the pleural space by listening for air movement, and by looking for fogging of the tube during expiration.

11 Connect the chest drain tube to an underwater seal.

12 Secure the tube using a suture passed through the skin at the incision site (ensure adequate local anaesthetic) and tie around the tube.

13 Cover the puncture site in the chest wall with a sterile dressing and tape the chest tube to the chest wall – cotton gauze under "opsite" may provide an optimal occlusive dressing.

14 Obtain a chest radiograph if at all possible.

If the chest tube is satisfactorily positioned and working, occasional bubbles will pass through the underwater seal. The water level in the tube may also rise and fall slightly with the respiratory cycle.

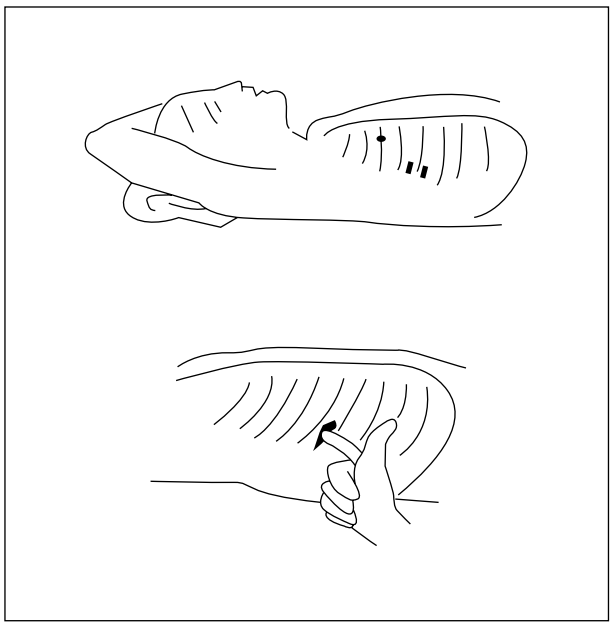


Figure 6.12.1 (Top) Insertion site. (Bottom) Insert finger into the incision and clear path into pleura.

6.13 Intubation

Aims

- Secure airway
- Protect airway
- Prolonged ventilation
- Intraoperative ventilation
- Tracheobronchial toilet
- Application of high airway pressures and PEEP
- Cardiopulmonary resuscitation (all of the above)

Which tube?

Uncuffed under 25 kg body weight. Narrowest part of airway is below the glottis at the circular non-distensible cricoid ring.

Correct tube is that which passes easily through the glottis and subglottic area with a small air leak detectable at 20 cm H_2O (= sustained gentle positive pressure).

The correct size of tube is one that can just fit into the nostril.

- In preterm neonates 2.5–3.5 mm internal diameter.
- In full-term neonates 3.0–4.0 mm internal diameter.
- In infants after neonatal period 3.5–4.5 mm internal diameter.
- Children over 1 year
  - Internal diameter in mm = age/4 + 4
  - Length of tube in cm = age/2 plus 12 for oral tube
    = age/2 plus 15 for nasal tube

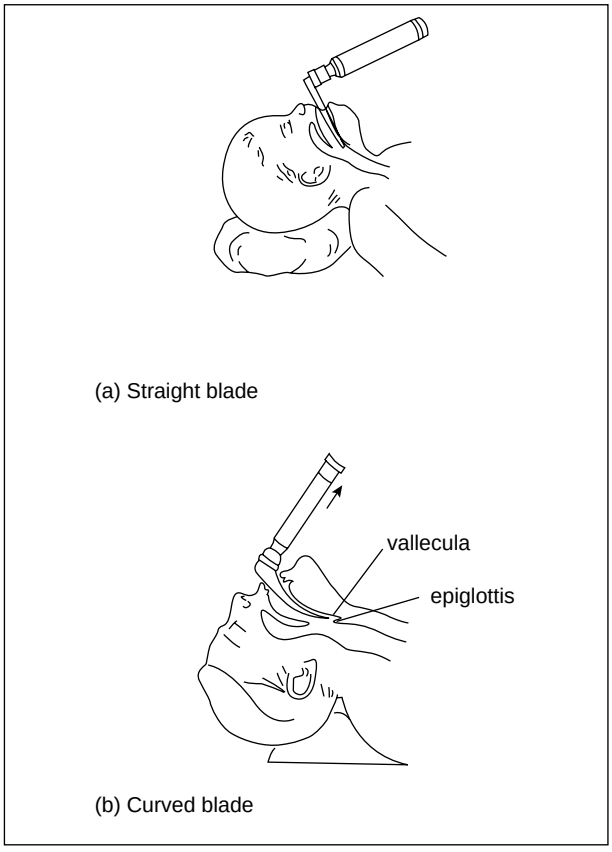


Figure 6.13.1 Blades for laryngoscopy.

Aids to intubation

- Laryngoscope: blade (straight for neonates and infants because of long, floppy epiglottis, curved for older children), intact bulb
- Magill forceps
- Introducer (not protruding further than end of tube itself)
- Gum elastic bougie (over which tube can pass)
- Cricoid pressure (can help visualisation of larynx)
- Suction apparatus must be available

Predicting difficulty

Likely to be difficult where there is:
- Difficulty in opening mouth or reduced neck mobility
- Laryngeal/pharyngeal lesions (for example large tonsils, diphtheria, epiglottis)

Congenital airway abnormalities:Pierre-Robin syndrome, mucopolysaccharoidoses, Down's syndrome,
Acquired airway abnormalities:burns, trauma
Look from the side: if small chin = difficult

Complications

- Displacement: into oesophagus or endobronchial
- Obstruction: kinking, secretions, mucous plug
- Trauma: lips to larynx
- Hypertensive response
- Spasm: laryngeal, pharyngeal
- Aspiration: gastric contents
- Vagal response resulting in profound bradycardia

How to do it

Prepare and check equipment

- Choose appropriate tube size with one size above and below also available
- Prepare tape ready to fix tube
- Suction must be available
- Induce anaesthesia and give muscle relaxant unless completely obtunded
  Do not attempt in semiconscious child

Position child's head and airway

- >3–4 years: "sniffing morning air" position (head extended on shoulders, flexed at neck, pillow under head)
- <3 years (especially neonates and infants): neutral position (large occiput)
- Keep in neutral position with in-line immobilisation if unstable cervical spine (trauma, Down's syndrome)

Oxygenate child

By bag and mask for 1–2 minutes with 100% oxygen (use reservoir)
- Introduce laryngoscope into right side of mouth
- Sweep tongue to the left
- Advance blade until epiglottis seen
- Curved blade:
  - Advance blade anterior to epiglottis
  - Lift epiglottis forward by moving blade away from own body
- Straight blade:
  - Advance blade beneath epiglottis, into oesophagus
  - Pull back, glottis will "flop" into view

Must identify glottis/cords

- Insert endotracheal tube gently through vocal cords
- Stop at predetermined length
- Must see tube go through cords

Confirm correct placement

- Chest moves up and down with ventilation
- Listen to breath sounds in axillae and anterior chest wall
- Confirm no breath sounds in stomach and air not building up into mouth around tube
- Oxygen saturations do not go down
- ***Carbon dioxide is measured from expired gases*** ✔
- Secure tube
- Proceed to nasal intubation if skilled (best for long-term ventilation)

Intubation of the newborn infant without a laryngoscope

This can be very helpful if a laryngoscope is not available, or a child has facial or oral deformities that interfere with ability to insert a laryngoscope or to see the larynx; for example severe micrognathia.

1 Insert the index finger of the left hand into the baby's mouth, with its palmar surface sliding along the tongue. Use the little finger if the baby is small.
2 Slide the finger along the tongue until it meets the epiglottis. This feels like a small band running across the root of the tongue.
3 Slide the finger a little further until the tip lies behind and superior to the larynx and the nail touches the posterior pharyngeal wall.
4 Slide the tube into the mouth between the finger and the tongue until the tip lies **in** the midline at the root of the distal phalanx of the finger.
5 At this point place the left thumb on the baby's neck just below the cricoid cartilage in order to grasp the larynx between the thumb on the outside and the fingertip on the inside.
6 While the thumb and finger steady the larynx against side to side motion the right hand advances the tube a short distance; about 1–2 cm.
7 A slight give can sometimes be felt as the tube passes into the larynx but **no force is needed for insertion**.
8 When the tube is in the trachea the laryngeal cartilage can be felt to encircle it. If it has passed into the

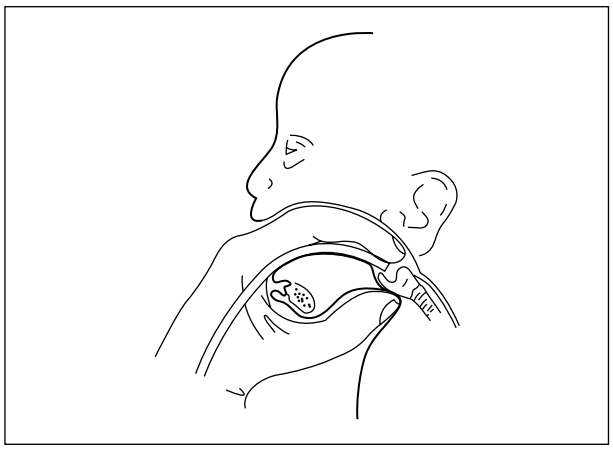


Figure 6.13.2 Intubation of the newborn baby without a laryngoscope.

oesophagus it can be felt between the finger and the larynx.

It is worth practising this technique on a stillborn baby.

Fixation of endotracheal tubes

✔ ● **Have at least two people available to do this. One should hold the tube at all times**.
- Sometimes sedation is required to do this.
- Cut two strips of sticky zinc oxide tape (see Figure 6.13.3). Together they should reach from just in front of the ear across the cheek and above upper lip to the opposite ear.

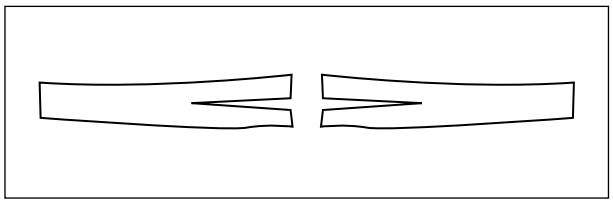


Figure 6.13.3 Zinc oxide tape cut for fixation of endotracheal tube (See figure 6.13.4)

- If available, apply some benzoin tincture to the cheeks, above the upper lip and under the lower lip. (This makes the tape stick better.)
- Make sure the endotracheal tube is clean.
- Start with the broad end of the tape: stick this onto the cheek, then wrap one of the thinner ends carefully around the tube. It is useful still being able to see the endotracheal tube marking at the lips.
- The other half gets taped across the upper lip to the opposite cheek.
- The second tape starts on the other cheek, and the thinner half gets stuck across the chin, the other half is also wrapped around the tube (see Figure 6.13.4).

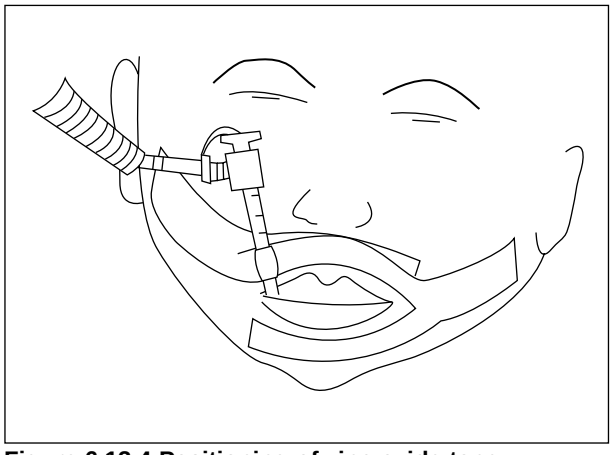


Figure 6.13.4 Positioning of zinc oxide tape.

6.14 Creation of an emergency surgical airway

Cricothyroidotomy is indicated if a patent airway cannot be achieved by other means. It must be performed promptly and decisively when necessary.

In children under the age of 12 years, needle cricothyroidotomy is preferred to surgical cricothyroidotomy. In the adolescent, either technique can be used but the surgical technique allows better protection of the airway. The relevant anatomy is shown in Figure 6.14.1.

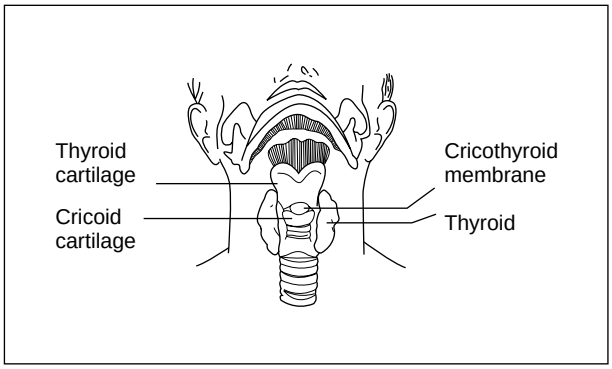


Figure 6.14.1 Anatomy for cricothyroidotomy.

In a very small baby, or if a foreign body is below the cricoid ring, direct tracheal puncture using the same technique can be used.

Needle cricothyroidotomy

This technique is far from easy in practice. In an emergency situation the child may be struggling, and attempts to breathe or swallow may result in the larynx moving up and down:

1 Attach a cricothyroidotomy cannula-over-needle (or if not available, an intravenous cannula and needle) of appropriate size (16–20 gauge) to a 5 ml syringe.
2 Place the patient in a supine position.
3 If there is no risk of cervical spine injury, extend the neck, perhaps with a sandbag under the shoulders.
4 Identify the cricothyroid membrane by palpation between the thyroid and cricoid cartilages.
5 Prepare the neck with antiseptic swabs.
6 Place your left hand on the neck to identify and stabilise the cricothyroid membrane, and to protect the lateral vascular structures from needle injury.
7 Insert the needle and cannula through the cricothyroid membrane at a 45° angle caudally, aspirating as the needle is advanced (Figure 6.14.2).
8 When air is aspirated, advance the cannula over the needle, being careful not to damage the posterior tracheal wall. Withdraw the needle.
9 Recheck that air can be aspirated from the cannula.

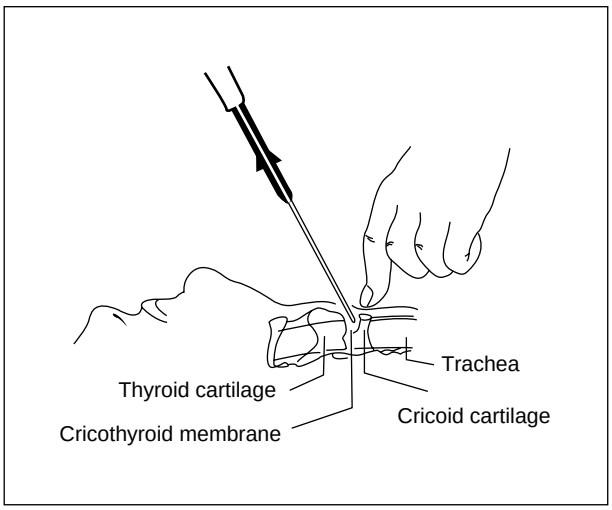


Figure 6.14.2 Insertion of needle and cannula through the cricothyroid membrane.

10 Attach the hub of the cannula to an oxygen flowmeter via a Y-connector. Initially the oxygen flow rate (in litres) should be set at the child's age (in years).
11 Ventilate by occluding the open end of the Y-connector with a thumb for 1 second, to direct gas into the lungs. If this does not cause the chest to rise, the oxygen flow rate should be increased by increments of 1 litre, and the effect of 1 second's occlusion of the Y-connector reassessed.
12 Allow passive exhalation (via the upper airway) by taking the thumb off for 4 seconds.
13 Observe chest movement and auscultate breath sounds to confirm adequate ventilation.
14 Check the neck to exclude swelling from the injection of gas into the tissues rather than the trachea.
15 Secure the equipment to the patient's neck.
16 Having completed emergency airway management, arrange to proceed to a more definitive airway procedure, such as tracheotomy.

Important notes

There are two common misconceptions about transtracheal insufflation. The first is that it is possible to ventilate a patient via a needle cricothyroidotomy using a self-inflating bag. The maximum pressure from a bag is approximately 4.41 kPa (45 cm H_2O) (the blow-off valve pressure) and this is insufficient to drive gas through a narrow cannula. In comparison, wall oxygen is provided at a pressure of 4 atmospheres (approximately 392 kPa or 4000 cm H_20). The second misconception is that expiration can occur through the cannula, or through a separate cannula inserted through the cricothyroid membrane. This is not possible. The intratracheal pressure during expiration is usually less than 2.9 kPa (30 cm H_2O) (less than one-hundredth of the driving pressure in inspiration). Expiration must occur via the upper airway, even in situations of partial upper airway obstruction. Should upper airway obstruction be complete, it is necessary to reduce the gas flow to 1–2 litres/min. This provides some oxygenation but little ventilation.

Nevertheless, insufflation buys a few minutes in which to attempt a surgical airway.

Surgical cricothyroidotomy

This should only be considered in the older child (12 years or over):

1 Place the patient in a supine position.
2 If there is no risk of neck injury, consider extending the neck to improve access. Otherwise, maintain a neutral alignment.
3 Identify the cricothyroid membrane.
4 Prepare the skin and, if the patient is conscious, infiltrate with local anaesthetic.
5 Place your left hand on the neck to stabilise the cricothyroid membrane, and to protect the lateral vascular structures from injury.
6 Make a small vertical incision in the skin, and press the lateral edges of the incision outwards, to minimise bleeding.
7 Make a transverse incision through the cricothyroid membrane, being careful not to damage the cricoid cartilage.
8 Insert a tracheal spreader, or use the handle of the scalpel by inserting it through the incision and twisting it through 90° to open the airway.
9 Insert an appropriately sized endotracheal or tracheostomy tube. It is advisable to use a slightly smaller size than would have been used for an oral or nasal tube.
10 Ventilate the patient and check that this is effective.
11 Secure the tube to prevent dislodgement.

Complications of cricothyroidotomy

These include:

- Asphyxia.
- Aspiration of blood or secretions.
- Haemorrhage or haematoma.
- Creation of a false passage into the tissues.
- Surgical emphysema (subcutaneous or mediastinal).
- Pulmonary barotrauma.
- Subglottic oedema or stenosis.
- Oesophageal perforation.
- Cellulitis.

6.15 Needle pericardiocentesis

Indication

- To reduce a pericardial effusion causing haemodynamic compromise.
- To diagnose pericarditis.

Equipment

- ECG monitor: Local anaesthetic
- Syringe: Over needle cannula (16/18 gauge)
- Skin prep: Sterile drapes

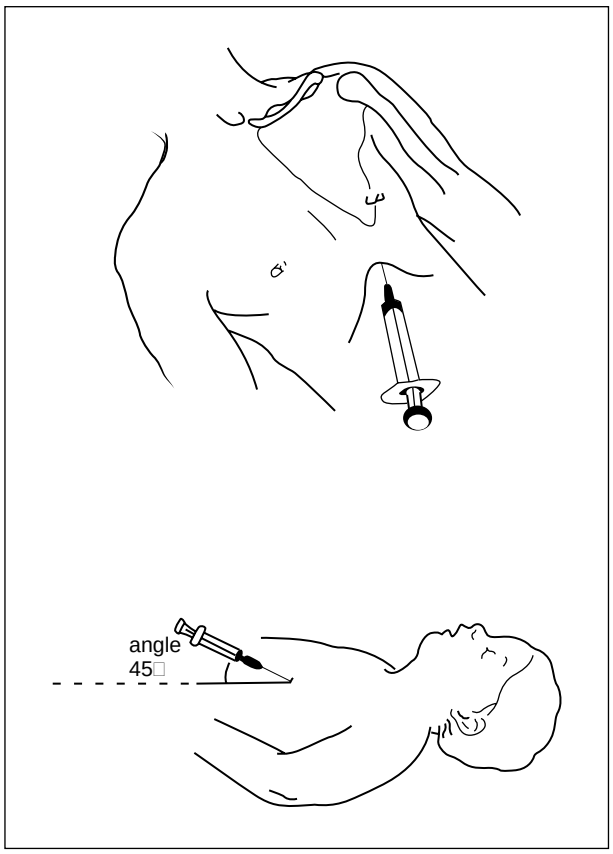


Figure 6.15.1 Needle pericardiocentesis.

Technique

1 Position the patient supine and attach the ECG. Stand on the patient's right with the ECG monitor at the patient's head so you can see it easily.
2 Clean the skin from nipples to umbilicus and drape with sterile towels to expose the perixiphoid region. Infiltrate local anaesthetic at the costal margin just below the xiphoid process.
3 Attach the cannula to the syringe. Insert the cannula just below and to the left of the xiphoid process. Angle the needle at 45° to the skin and pointing towards the tip of the left scapula.
4 Advance the needle, holding this position, aspirating all the time and watching the cardiac monitor. As the distended pericardial sac is entered fluid will flow back into the syringe. If the myocardium is touched the ECG pattern will change (arrhythmia, ectopics, "injury" pattern). If large amounts of bright red blood are aspirated the ventricle has been entered; withdraw slightly.
5 If successful, cardiac function should improve immediately. Withdraw the needle attach a three-way tap and secure the cannula for further aspirations.
6 This is a temporary procedure and some patients will require a formal pericardiotomy. Note pericardial aspiration may not work well for viscous fluids (for example clotted blood) in the pericardial sac.

6.16 Intracardiac injection of drugs in the resuscitation of the neonate (see Chapter 1.22)

Intracardiac injections of epinephrine, sodium bicarbonate or THAM, and 10% glucose may be a reasonable risk to take under the circumstances of neonatal cardiac arrest which is unresponsive to external cardiac massage, and combined with adequate lung inflations and an inability to gain access to the umbilical vein.

1 Sodium bicarbonate or THAM by reversing intracardiac acidosis may improve cardiac output. 1 ml/kg of 8.4% sodium bicarbonate or of 0.6 M THAM. Give a few cardiac compressions after the injection. If no increase in heart rate within 2 minutes repeat the dose.
2 10% GLUCOSE 2 ml/kg
3 **Epinephrine**: (0.1 ml/kg of the 1 in 10 000 solution) before abandoning resuscitation.
4 **Volume**: On rare occasions profound bradycardia will respond to volume expansion. A bolus of 10–20 ml/kg of 0.9% saline or colloid is usually sufficient.

Technique of intracardiac injection in the neonate

The risk of pneumothorax is remote if the guidelines below are followed because there is virtually no lung tissue between the heart and chest wall in the newborn baby.

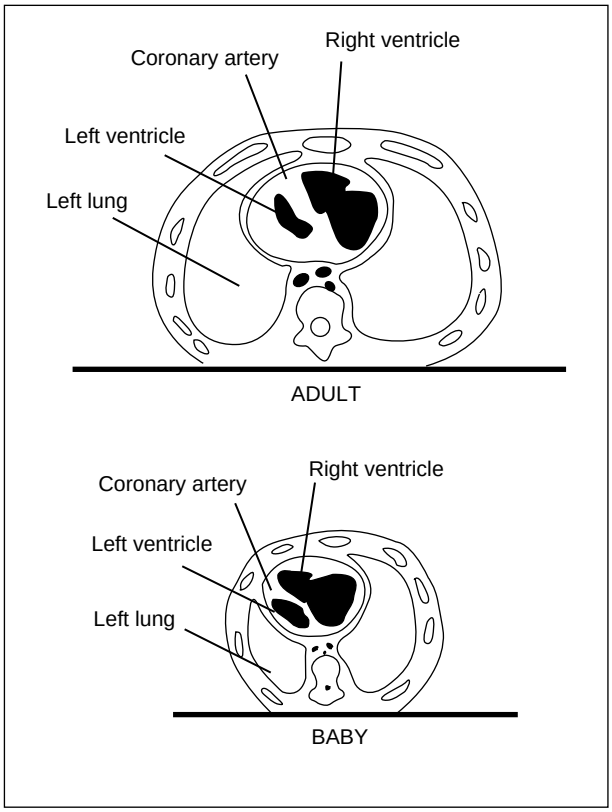


Figure 6.16.1 Intrathoracic anatomy.

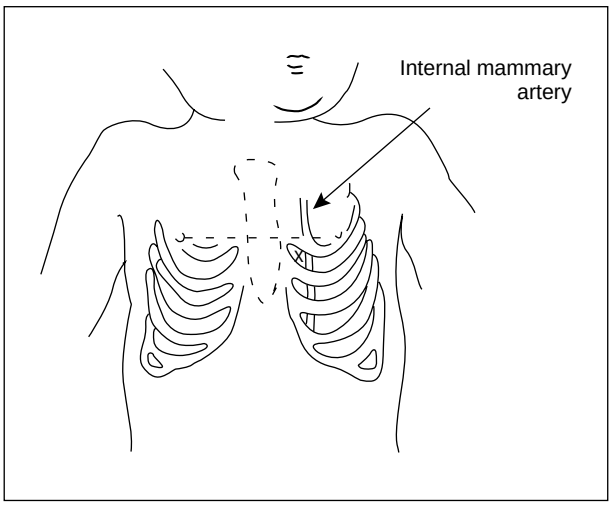


Figure 6.16.2 Insert the needle as close as possible to the sternum to avoid the internal mammary artery. Insert at point X.

The anterior descending branch of the left coronary artery is also more lateral in babies and therefore much less likely to be damaged from an anterior approach than it would be in adults.

Use a 21G (green) needle attached to a syringe. Choose a point at the left sternal edge and just below a line joining the nipples (ideally 4th or 5th intercostal space). Keep as close as possible to the sternal edge to avoid the internal mammary artery. Point the needle towards the spine. You will usually enter the right ventricle or atrium, as these are anterior to the left heart chambers. Check that the needle is in a cardiac chamber by aspirating blood into the syringe. Never inject anything unless blood can first be aspirated freely.

6.17 Defibrillation

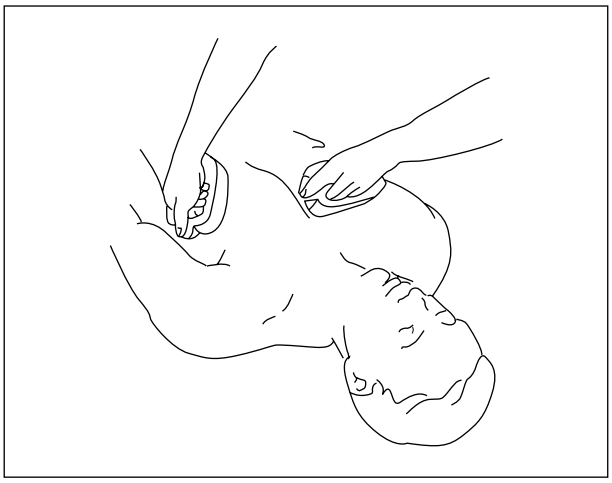


Figure 6.17.1 Position of paddles for the use of a defibrillator.

Safety

A defibrillator delivers enough current to **cause** cardiac arrest. The user must ensure that other rescuers are not in physical contact with the patient (or the trolley) at the moment the shock is delivered. The defibrillator should only be charged when the paddles are in contact with the child.

Procedure

Basic life support should be interrupted for the shortest possible time (during 5–9 below)

1 Apply gel pads or electrode gel
2 Select the correct paddles (paediatric paddles for those <10 kg)
3 Select the energy required
4 Place the electrodes onto (see below) gel, and apply firm pressure
5 Press the charge button
6 Wait until the defibrillator is charged
7 Shout “Stand back!”
8 Check all other rescuers are clear
9 Deliver the shock

Correct paddle placement

The usual placement is anterolateral. One paddle is put over the apex in the midaxillary line, and the other is placed just to the right of the sternum, immediately below the clavicle.

Good paddle contact

Gel pads or electrode gel should always be used (if the latter, care should be taken not to join the two areas of application). Firm pressure should be applied to the paddles.

Correct energy selection

The recommended levels are 2 joules/kg for the first two shocks and then 4 joules/kg.

6.18 Non-invasive ventilatory support

Continuous positive airway pressure (CPAP)

Continuous positive airway pressure (CPAP) has several beneficial effects on the airway and lungs of the pre-term and full-term infant. These include prevention of alveolar collapse, increased functional residual capacity (FRC), and splinting of the airway. It is therefore of most value when used early in the course of respiratory disease (i.e. before too much alveolar collapse has taken place). Several units around the world use it as first-line ventilatory support in even the smallest infants (<750 g birthweight) with respiratory failure with success.

Indications

- Signs of significant respiratory distress (tachypnoea, recession, grunting, nasal flare).

- Diseases with low FRC (respiratory distress syndrome, transient tachypnoea of the newborn, pulmonary oedema).
- Meconium aspiration syndrome.
- Apnoea and bradycardia of prematurity.
- Tracheomalacia.

Requirements

(See Figures 6.18.1 and 6.18.2)

- Low-resistance delivery system:
  - Large-bore tubing.
  - Short, wide connection to patient
- Consistent, reliable pressure generation:
  - Aim for +5 to +10 cm H_2O.
  - Appropriate, snug-fitting, nasal prongs or nasal mask.
  - Well-positioned and secured nasal prongs or nasal mask.
  - Prevention of leaks, mainly via mouth, with a chinstrap.
- Optimally maintained airway:
  - Warmed, humidified gas.
  - Prevention of neck flexion/overextension with a neckroll. (Keep head in neutral position.)
  - Remove secretions with suction only if appropriate.
- Meticulous, consistent technique.
- Good nursing

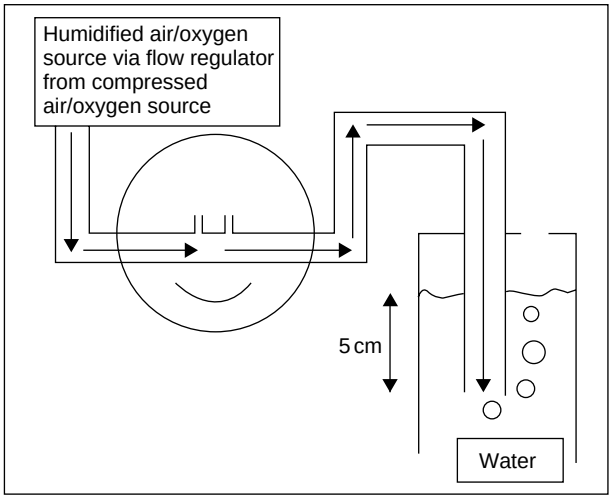


Figure 6.18.2 Simplified diagram of a simple water manometer controlled nasal prong CPAP (set here at +5 cm H_2O).

Complications

- Nasal septum erosion/necrosis. This is a result of ill-fitting nasal prongs and can be avoided by the fitting of snug, but not tight prongs (blanching of the overlying skin suggests the prongs are too large) that are held firmly in place to prevent rubbing as the child moves.
- Pneumothorax. All methods of artificial ventilation are associated with this problem. However the more effective the CPAP is, the less the work of breathing and therefore the lower the risk of pneumothoraces should be. Any pneumothorax which does occur should be drained appropriately. It is inappropriate to discontinue the CPAP.
- Gastric distension from swallowed air – this is a benign finding and is easily overcome by the venting of any such air via an orogastric tube.

Continuous negative extrathoracic pressure (CNEP)

Continuous negative extrathoracic pressure (CNEP) is a method by which subatmospheric pressure is applied to the outside of a child's chest by nursing him/her in a specially designed chamber. The patient's head is kept outside the chamber, thereby allowing the nose, mouth and all the

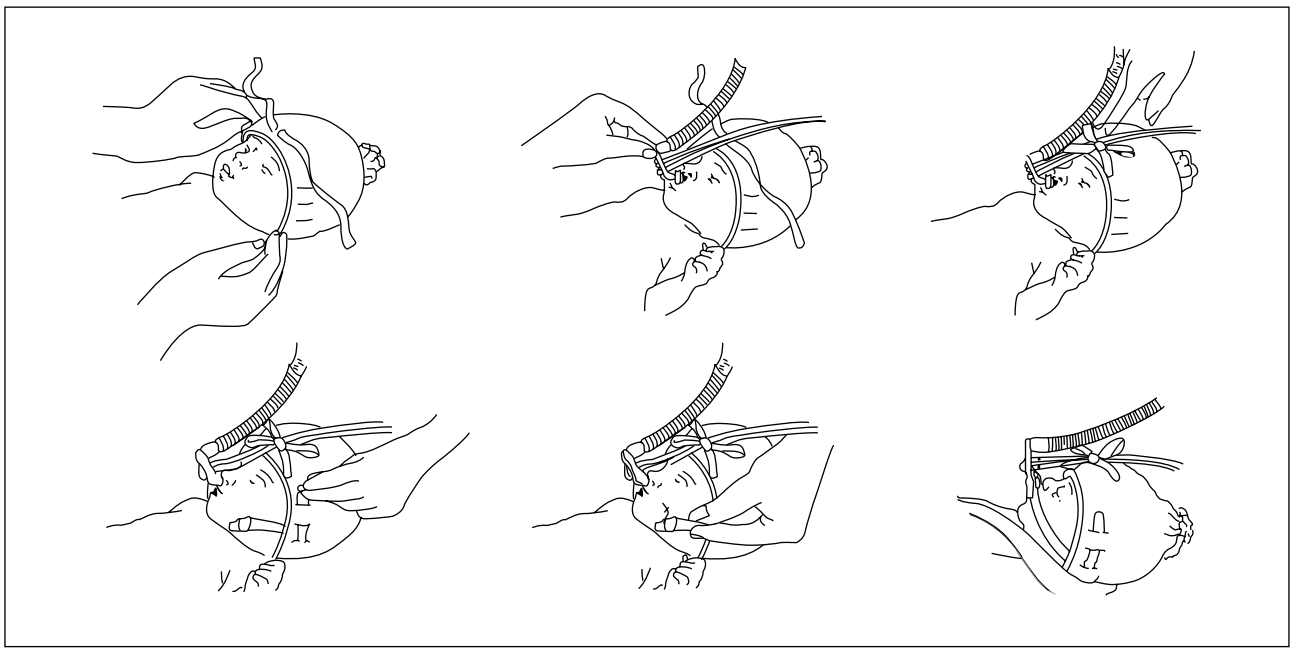


Figure 6.18.1 Attaching nasal prong CPAP to the infant using a bonnet.

airways into the lungs to remain at atmospheric pressure. As a result of this pressure difference, the chest is expanded and air is encouraged to enter the lungs. Areas of lung that were previously poorly inflated may be expanded and this allows more chance for recovery from of lung disease. There is more certainty of distending pressure reaching the lungs than with CPAP.

The use of CNEP depends upon continued breathing efforts by the child to move air into and out of the lungs. When inside the chamber, the breathing rate falls, the effort of breathing is reduced and thus less energy is needed for breathing.

Indications for negative pressure

- Neonatal respiratory failure
  Bronchiolitis
  Other acute lower respiratory infections
- Respiratory failure due to weakness of the respiratory muscles

Advantages of negative pressure

- Absence of airway invasion
  - Avoids trauma to the airways
  - Reduces the need for suctioning
  - Lowers the risk of introducing infection into the lungs
  - More comfortable for the patient and less need for sedation
- Less complex equipment
  - Can be managed by nurses or parents at home
  - Not difficult for staff/parents to learn how to use
  - Less utilisation of intensive care resources
  - Quick to institute, not requiring medical or anaesthetic staff
- Physiological (compared to positive pressure ventilation)
  - Does **not** increase pulmonary vascular resistance
  - Less likely to significantly reduce cardiac output
  - Enhances lung perfusion, as well as ventilation

Disadvantages of negative pressure

- Negative pressure generated in the upper airway on inspiration may be increased, thus exacerbating **pre-existing** upper airway obstruction.
- Less suitable where need for ventilation is critical and continuous (unless supported by IPPV).
- Maintenance of body temperature in newborn infants may require specific attention.

Components of the negative-pressure system

The negative pressure system includes:

- The chamber (Figure 6.18.3)
- The neck seal (Figure 6.18.4)
- The neck protector
- The suction unit (which can produce either continuous negative pressure or cycles of pressure which can ventilate the baby)
- The pressure monitor

The neck protector

This is a piece of two or four thicknesses of ribbed cotton tubular stockinet. Two holes cut in the sides allow this to be fitted like a polo-necked vest over the infant.

The suction unit

This incorporates an electrical fan with a valve which provides variable levels of suction. The valve is adjustable by a pressure control knob. A suction hose connects from the suction unit to the base of the chamber.

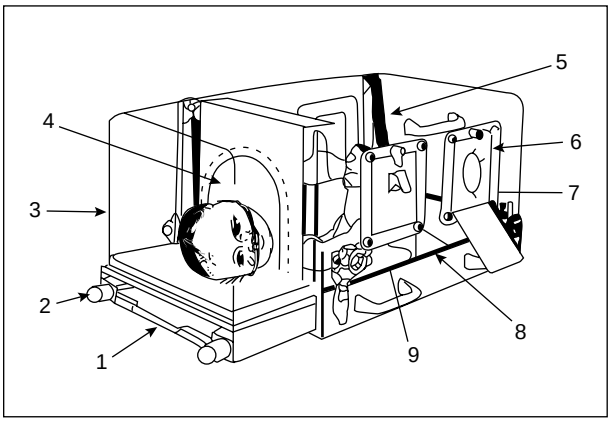


Figure 6.18.3 The chamber.
For low birthweight infants, the chamber is built onto an incubator base incorporating a cabinet and heater, the latter providing a servo-controlled circulation of hot air into the interior of the chamber. Particular features of the chamber shown in the illustration include the following: 1. Release for head section. 2. Rods upon which the head section slides out away from the chamber base. 3. Headbox. 4. Latex rubber neck seal fixed onto the arch of the lid. 5. Gas strut hinges. 6. Foam gasket or cuff on the porthole. 7. Porthole for infusions, monitoring leads, etc. 8. Footplate to support the infant when the chamber is tilted head up. 9. Rubber strips below which leads, etc. can enter chamber.

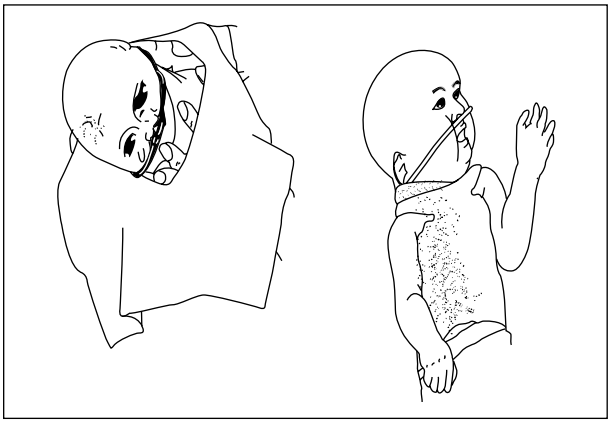


Figure 6.18.4 The neck seal. This is a piece of stretchy latex rubber with a circular hole 2–5 cm in diameter. This is stretched and fitted around the infant's neck overlying the neck protector. (Stockinet possibly also with a silicone rubber protective circular pad.)

The pressure monitor
This can be a simple calibrated U-tube containing coloured alcohol or other fluid or a more sophisticated pressure monitor.

Safety of negative-pressure ventilation

- Special care must be taken not to trap fingers or toes when closing the lid.
- To avoid damage to monitor leads, these must pass through either (1) a diaphragm at the base of the chamber, or (2) between a rubber strip at the foot end of the chamber and a second rubber strip on the lower edge of the lid. Monitor leads will be damaged and pressure lost if leads are brought out through the sides of the chamber between the unprotected perspex edge of the lid and the rubber seal on the base.

Care of a baby receiving negative-pressure ventilation

Feeding
Whilst receiving negative pressure, the patient would not usually be fed orally. Feeding is usually given by a nasogastric tube. This tube should be clamped off when not in use. Do not leave the tube open to air or the stomach may become distended.

If the baby develops problems with abdominal distension, the stomach should not be left on free drainage in the conventional way. Either frequent aspiration of the stomach contents should be performed with the tube clamped off in between times, or the end of the tube can be put through the neck seal (in between the silicone gel and the vest) and the tube left on free drainage inside the chamber.

If the clinical condition allows, the baby may come out of the chamber at regular intervals for breast or cup feeds.

Procedures
Most procedures such as the re-siting of an intravenous line can be carried out whilst the infant is receiving negative pressure. When the arms of the doctor or nurse are inserted through the portholes, subatmospheric pressure can be maintained by the close-fitting cuffs around the forearms.

Neck care
Particular attention is paid to the baby's neck. Observe the neck for soreness frequently at 6–8 hourly intervals if possible when the baby is turned. It is important to ensure that all the layers of the neck protector are in place and that the latex does not come into contact with the baby's skin. It is not necessary to replace the neck seal components unless they are soiled or damaged. If they are in place for a prolonged period, especially in a newborn preterm baby whose skin may scale and shed, it is probably better to wash and dry the neck at regular intervals when the clinical condition allows preventing colonisation by skin commensals.

It is also important frequently to check the neck to ensure that the latex does not become too tight if the baby becomes oedematous. If this happens it is important to replace the latex with a sheet containing for a larger sized aperture.

Controlling body temperature
An important principle to follow in the control of body temperature in babies receiving negative pressure, is to **prevent hypothermia**. Due to convective cooling around the neck seal and radiant heat loss the baby (particularly if preterm and low birth weight) is more likely to cool rapidly and be more difficult to warm up than in conventional incubators.

Plastic sheeting or bubble wrap, placed over the infant's body to create a "microenvironment", may be required.

An overhead radiant heater may be used as an additional heat source over the headbox and chamber.

Trouble-shooting

Problem: inadequate pressure
- Excess leak at neck
  - Slacken latex into the arch in the lid.
  - Move the baby upwards.
  - Re-position the latex.
  - Double the thickness of stockinet collar under the latex.
  - Use latex with a smaller hole (if large leak present).
- Excess leak between chamber and base
  - Tighten quick release lid and base catches.
  - Replace rubber strip gasket around the chamber base.
- Excess leak at the portholes
  - Renew cuffs or foam gaskets.
  - Tighten or secure the iris diaphragm porthole.
- Inadequate suction pressure
  - Check that the hose is plugged in at both ends.
  - Check that the access hole for suction inside the chamber is not blocked by, for example sheet.
  - Check pressure achieved by suction unit after directly occluding hose at end.

Problem: unsettled baby
- Baby working hard
  - Inadequate negative pressure or rate.
- Upper airway obstruction
  - Check for stridor, tracheal tug, CO_2 retention. Different method of respiratory support may be needed.
- Anxiety
  - Reassurance/sedation to make baby comfortable.
- Sore neck
  - Check neck and treat sore areas to relieve discomfort

Problem: abdominal distension
- Air swallowing or via nasogastric tube
  - Close nasogastric tube.
  - More frequent suction of nasogastric tube or free drainage inside the chamber.

Problem: cold baby
- Excess leak

See "Inadequate pressure".
- Cold environment
  - Warm and humidify headbox O_2.
  - Overhead heater.
  - Introduce warmed humidified air directly into chamber using humidifier.

Problem: neck soreness
- Pressure or contact allergy
  - Ensure that there is at least a fourfold thickness of stockinet between the latex and the skin.
  - If the latex is stretched too tightly, the baby may be suspended at the neck. Release and pleat it as described above.
  - The hole in the latex may be too small, revise if necessary.

Problem: inadequate oxygen in the headbox
- Excess leak at the neck: see "Inadequate pressure".
- Inadequate oxygen flow/concentration: seal the top and sides of the headbox, for example with clingfilm. Two supplies of high-flow humidified 100% oxygen may be needed to provide a high concentration in the headbox.

6.19 Insertion of oro/nasogastric tube

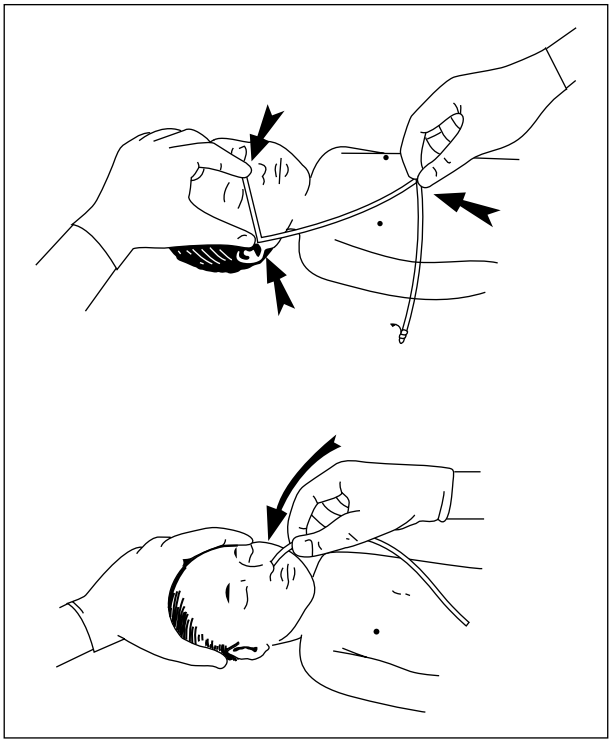


Figure 6.19.1 Inserting a nasogastric tube. The distance is measured from the nose to the ear and then to the epigastrium, and then the tube is inserted to the measured distance.

Indication

To feed any child who is unable to take food by mouth.

Equipment

- Nasogastric tube
- Lubricant
- Litmus paper
- Syringe
- Stethoscope
- Adhesive tape

In preterm infants:
- 4 French gauge tube (weight 1000 g or less)
- 6 French gauge tube (weight over 1000 g)
- 8–10 French gauge tube – used for abdominal decompression, for example baby with ileus or receiving continuous positive airway pressure (CPAP).

Procedure

1 Place the child supine with head in the "sniffing" position.

2 Measure length of tube from nose via earlobe to the mid-point between xiphoid and umbilicus.

3 Feed the tube lubricated with KY jelly or 0.9% saline through either nose or mouth directly backwards. (The neonate is a nose breather and therefore the oral route is preferred.) Try to advance the tube as the child swallows. If the baby has respiratory distress, a gastric tube is best passed through the mouth. If passed through the nose it may increase upper airway resistance.

4 Check the position of the tube by aspirating stomach contents and checking a change in the litmus paper (blue to pink) or flush the tube with 2–3 ml air (only 1ml in the neonate) and listen over the stomach. If in doubt *X* ray chest/abdomen. (Note: the acidity of the gastric fluid may be reduced in preterm infants.)

5 Secure the tube by taping it to the cheek and record the length of tube outside the nose or mouth.

6.20 Incision and drainage of abscess

Indication

- The collection of a mass of infection anywhere.
- If unsure of an abscess, aspirate for pus before proceeding to incision and drainage.
- Multiple/recurrent abscesses may be associated with HIV, tuberculosis, malnutrition, diabetes mellitus, anaemia, foreign bodies.

Equipment

- Skin prep
- Scalpel
- Microbiology swab
- Curette
- Sterile gauze

Procedure

- If systemically unwell take blood cultures (before giving antibiotics).
- Antibiotics are only indicated if systemically unwell or if spreading cellulitis is present.
- Use general anaesthesia for most sites (peri-anal, breast, cervical etc.). Regional blocks may be used for limbs in older children. (Note: local infiltration produces poor anaesthesia in inflamed tissue.)
- Clean the skin.
- Incise over the most superficial tender point in the direction of skin creases. Take a sample of pus for culture and staining including Ziel-Nielsen if indicated. (The commonest error is to make the incision too small.)
- Insert curette spoon or finger to break down any loculi. Send a sample of the wall of the abscess for tuberculosis if indicated.
- Irrigate the cavity with saline to flush out necrotic material.
- If a large cavity exists, loosely pack with sterile gauze. For a small cavity place a "wick" (piece of rolled gauze) into the wound forming a track. Cover the wound loosely with absorbant dressing. Change the gauze packing after 24 hours with analgesia beforehand. Remove the wick after 48 hours.
- As the cavity discharges pus, it should heal from deeply to superficially through the open skin incision.

6.21 Liver biopsy

Indications

Most commonly used when a histological diagnosis is required for example chronic active hepatitis and unexplained hepatomegaly. It is also used in the assessment of metabolic disorders, pyrexia of unknown origin, drug effects on the liver and storage disorders.

Liver biopsy may be undertaken blind but it is safer when done with ultrasound guidance. Ultrasound is essential for focal lesions. **Ultrasonography should first** be undertaken to exclude focal liver disorders such as bile duct obstruction, hydatid cysts and arteriovenous malformations.

Equipment

- Skin prep
- Marker pen
- Scalpel
- Assistant
- Local anaesthetic
- Biopsy needle (Menghini or Tru-cut)
- Sterile drapes

Procedure

1 Ensure the patient has no contraindications
- Platelets $<80 \times 10^9$/litre
- Abnormal clotting (prothrombin time >3 seconds above normal, INR >1.3)
- Ascites
- Extrahepatic biliary obstruction
- Uncooperative patient

If clotting is abormal give vitamin K 300 micrograms/kg for three days parenterally and recheck. ***If it is essential to proceed then give fresh frozen plasma and platelets one hour before and after the procedure (if available).***

2 Cross-match blood and consent before proceeding.

3 In a cooperative child, biopsy may be done under sedation, otherwise under general anaesthetic.

4 Child lies on edge of bed with right hand behind his/her head. Outline of the liver is percussed along midaxillary line (or imaged by ultrasonography) and a site marked with iodine one space below upper limit of liver dullness, usually in 7–8th intercostal space while child is in expiration. If young or unable to cooperate, the child is held with both arms above the head and by the feet.

5 Clean and drape the area.

6 Aspiration needles, for example Menghini are simple to use. Cutting needles such as Tru-cut increase probability of obtaining adequate tissue for histology. Technique for using Menghini needle is described.

A size 1.4×40 mm needle is commonly used. The 3-cm nail is inserted to prevent aspiration of specimen into syringe. The needle is attached to a 20 ml syringe half filled with normal saline. The area is anaesthetised down to the capsule of the liver with adequate amounts of 1% lignocaine. A small nick is made in the skin with a scalpel blade. In young children, an attendant presses gently on the chest just above the liver to immobilise it. If possible, ask the child to hold his/her breath in an expiratory position. In young children who may already be crying, this will itself help to fix the lung in expiration. The needle is held with the left thumb as a guard to prevent too deep insertion. It is inserted down to the parietal pleura and 1 ml of saline is injected to clear any tissue. Aspiration is commenced and the needle is then rapidly pushed in and withdrawn from the liver in one swift movement.

The core of liver tissue is then gently expelled on to a glass surface. Specimens for routine histology are placed in buffered formal saline at −4°C. Specimens for electron microscopy are immediately put in a petri dish containing gluteraldehyde fixative and cut up. Specimens are then stored in 0.15 M isotonic phosphate buffer at −4°C until embedded.

7 Do not take more than three attempts.
8 Cover the skin incision with a dressing and lie the child on the right side for at least 3 hours.
9 Monitor blood pressure, pulse, respirations and temperature hourly for 6 hours. Keep the patient overnight.
10 Complications
- Pain (incision site and shoulder), bleeding, biliary peritonitis, sepsis.
- The commonest complication is pain which usually responds to analgesia. If it does not, then further investigation is needed.

6.22 Abdominal paracentesis

Indications

- To detect intra-abdominal injury after blunt trauma in the haemodynamically unstable child. (Haemodynamic instability after **penetrating** trauma always requires a laparotomy.)
- To identify peritonitis.
- To identify ruptured bowel.

Equipment

- Local anaesthetic (ideally with epinephrine)
- Over-needle catheter 16–20 gauge
- 20 ml syringe
- Warmed normal saline and infusion set
- Urinary catheter and nasogastric tube
- Sterile drapes
- Skin prep (iodine/alcohol)

Procedure

MUST BE A STERILE PROCEDURE.
1 Decompress bladder and stomach with urinary catheter and nasogastric tube.
2 Prepare the abdomen (costal margin to pubis). Drape the area with sterile towels exposing the peri-umbilical region.
3 Infiltrate local anaesthetic in the midline (a third of the distance between the umbilicus and the pubis). If pelvic trauma is suspected, infiltrate above the umbilicus.
4 Insert catheter over needle. Remove needle and aspirate.
5 If in the aspirate,
more than 10 ml fresh blood
turbid,
bile-stained fluid,
faeces or food debris,
there is a serious problem, probably indicating the need for a laporatomy.
6 If none of the above abnormalities is seen on aspiration, instill 10 ml/kg of warm 0.9% saline into the abdomen and allow 5 minutes to circulate. Retrieve the fluid.
7 Interpret the results of analysing the retrieved fluid. Abnormal if:
- red blood cell count (unspun) >100 000 per ml – may need laparotomy if unstable
- White cell count (unspun) >500 per ml
- Bile staining
- Faeces
- Gram stain/microscopy positive

8 If laporotomy is indicated, withdraw the catheter and cover the wound with a sterile dressing, then transfer to theatre.

6.23 Lumbar puncture

Figure 6.23.1 (Top) Holding a lying child for lumbar puncture. Note: the spine is curved to open up the spaces between the vertebrae. (Bottom) Restraining an older child in sitting position in order to carry out a lumbar puncture.

Equipment

- Iodine
- Sterile gloves
- Sterile dressings pack
- Spinal needle with stylet
- Collodion
- Small adhesive dressing

Indications

- As part of a septic screen
- Investigating seizures
- Investigating apnoea
- As therapy in posthaemorrhagic hydrocephalus

PRECAUTIONS

See sections on coma, meningitis and seizures for contraindications.

- **Lumbar puncture is dangerous in the presence of raised intracranial pressure**.
- Avoid in the very sick (may not tolerate procedure).
- Avoid in bleeding diathesis.
- Excessive neck flexion when positioning can lead to hypoxaemia and acute respiratory deterioration.

If a spinal needle is unavailable and a normal (non-stylet) needle is used, the needle bore may become blocked with skin on insertion and hence not flow. There is also the risk of tissue implantation leading to a dermoid cyst.

Advance the needle slowly. The subarachnoid space is only 0.5–0.7 cm below the skin in premature infants and 1 cm in babies, hence overpenetration is an easy mistake. Overpenetration leads to puncturing of the anterior vertebral venous plexus and a bloody sample, so that CSF microscopy is less informative or impossible.

Procedure (see figure 6.23.1)

- Employ a full aseptic technique.
- Position the patient on the edge of the examination table in lateral decubitus or sitting up. An experienced assistant to hold the patient is very helpful. Flex the spine maximally but avoid excessive neck flexion.
- Clean the lumbar area with iodine. Drape with sterile towels.
- Identify site of insertion: L4, L5 lumbar space (on level with iliac crests).
- Slowly insert spinal needle in the midline, aiming towards the umbilicus.
- Stop advancing when "give"/puncture sensation is felt on entering the subarachnoid space (often not felt in neonates). Frequent stylet withdrawals may be necessary during the procedure to see if CSF flows, that is when the subarachnoid space has been successfully entered.
- Withdraw stylet. Allow six drops of CSF to drip into each sample container.
- Replace stylet.
- Withdraw needle and swab puncture site with collodion.
- Cover site with adhesive dressing.
- Send samples for:
  - Microbiology (Gram stain, *Mycobacterium* culture if suspected, microscopy, cell counts, culture and sensitivity, virology)
  - Biochemistry (glucose, protein)

6.24 Suprapubic aspiration of urine

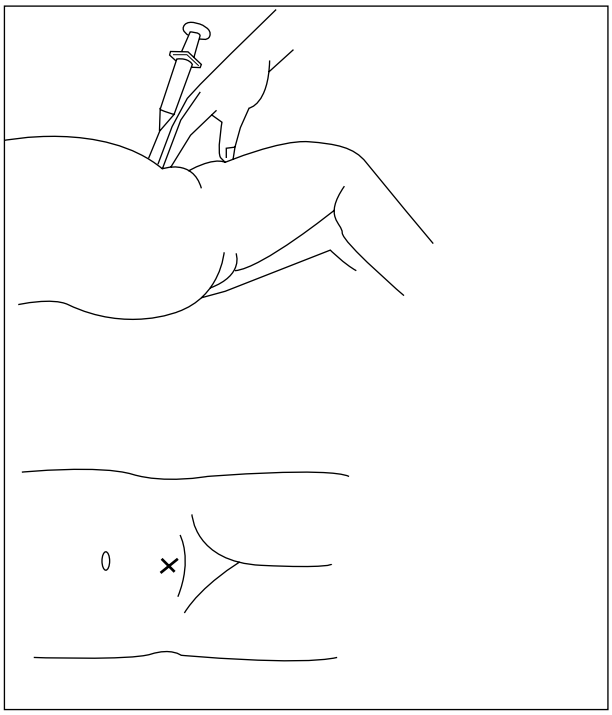


Figure 6.24.1 Position for carrying out suprapubic aspirate. This is a side view. Note the angle of insertion of the needle.

Indications

Usually in sick infants where urgent diagnosis is required and there is a palpable bladder which does not respond to manual expression for a clean catch.

Procedure

Use a sterile technique throughout. Advance a 23–24 gauge needle attached to a syringe to a depth of 3 cm in the midline at the proximal transverse crease above the pubis.

6.25 Microscopy of the urine

- Urinary tract infections (UTIs) are common in children.
- Though many are not serious, some cause kidney damage and lead to scars.
- Kidney scars can lead to high blood pressure, and to kidney failure later in life.
- A child with a UTI can develop kidney damage very fast – in just a few days! The only way to prevent this is to make the diagnosis and treat at once.
- Urine microscopy is the only way to diagnose UTIs immediately and reliably.

Urine

In a UTI the urine has:
- One species of bacteria at a concentration of at least 100 000/ml.
- An excess of white blood cells (WBC).

Bacterial numbers

Most children with a UTI have between 10 and 1,000 million bacteria/ml. 100 000/ml is actually a very small number of bacteria indeed. When urine is collected from children, it often becomes contaminated with a very small number of bacteria, and these are often of just one species. This means that if you rely on laboratory culture to make the diagnosis of UTI, you are likely to have many false-positives – perhaps one for every genuine case. Remember that every child diagnosed as having a UTI in this way will undergo investigations, sometimes invasive.

White cells

Children frequently have extra-urinary WBCs without a UTI:
- 10% of febrile children have hundreds of extra WBCs.
- Girls void some urine into the vagina, so vaginal WBCs are readily washed into the urine (and so are epithelial cells seen in the urine of most girls after puberty).

Children with UTIs often have no excess of WBCs, because:
- WBCs do not last long in urine, especially if it is alkaline and so urine must be examined soon after collection.
- Ill infants may be unable to mount a WBC response.

White cells are thus an unreliable and potentially misleading sign.

How should we count bacteria?

Laboratory culture

This is the widest used, and the traditional method. It remains acceptable, but if you use it:
- You have to accept that some positive reports will be false.
- You will wait at least 48 hours for the result. In reality it is often several days or a week before a positive lab report reaches the doctor and treatment starts. **Remember that kidney damage can become permanent in three days**.
- You will have to recall patients a few days later if the culture grows a mixture of bacteria. This is usually caused by the contamination of urine as it is collected, and is common. It must be repeated in case a UTI was present as well.
- You will miss the occasional UTIs caused by anaerobes.

Urine microscopy

If you use it:
- You can discard sterile urines, and reassure families at once.
- You can repeat contaminated urine sample at once.
- You can treat children with UTIs instantly.
- You can diagnose anaerobic UTIs as easily as aerobic ones.
- You can save time and money because it is cheaper and quicker than urine culture.

Which microscope?
Light microscope with an ordinary lens: bacteria are only easy to see after being gram-stained. An alternative is not to stain the urine but to use a counting chamber with mirrored surface.

Phase-contrast microscopes: these enable you to see unstained bacteria very easily, just using a drop of fresh urine on a glass slide. They look and work exactly the same as ordinary light microscopes, except that the lens (objective) and the condenser (underneath) are specially modified.

How do you microscope urine?
You can microscope fresh urine on a slide with a counting chamber. There is no need to stain or spin the urine.

The slide has two "chambers". Each has a grid etched onto the glass surface. In certain clinical situations, such as examining peritoneal dialysis fluid for suspected peritonitis, the grid can be used to make accurate counts of the concentrations of elements present.

Usually, this degree of accuracy is unnecessary. However, the grid is always useful because it tells you that you have got the microscope focused on the urine – if you examine a specimen with no cells or bacteria on a plain slide it is impossible to be certain otherwise.

Clean the slide and a coverslip with a tissue. Breathe over the slide to create a "mist" on it, and quickly push the coverslip into place. This creates a chamber 0.1 mm deep with a grid etched on the bottom.

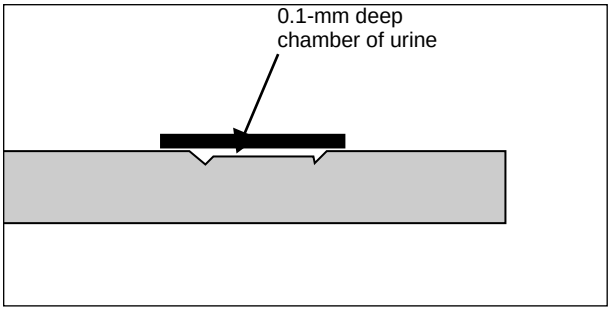


Figure 6.25.1 Microscope slide with coverslip.

Test the urine with a dip-stick (to check for blood, protein and glucose). Then touch the tip of the dip-stick on the slide so that a small amount of urine is drawn into the chamber by capillary action – you are now ready to go!

What are you looking for?

Bacteria

Most bacteria that cause UTIs are bacilli, that is rod-shaped. They are easy to identify – they look like straight lines, usually look about 3 mm long. Mostly they remain still, or just move a little, like a shimmer. This is due to Brownian motion (caused by them being hit by water molecules), and is not them swimming. Rarely you see motile bacteria – they hurtle about like crazy!

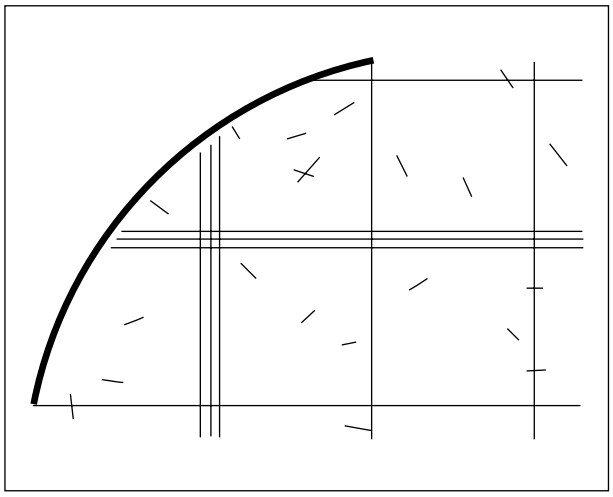


Figure 6.25.2 Bacteria (rods).

Infections also occur sometimes with streptococci, that is bacteria like strings of beads. There are always some strings that are four or more cocci long. If you think you see "cocci" individually, or in clumps, these are phosphate crystals – don't be fooled! Ignore the fact that they may be moving – just Brownian motion again!

White blood cells

These are round, and look between 3 and 5 mm in diameter. All have a 'granular' appearance to their cytoplasm.

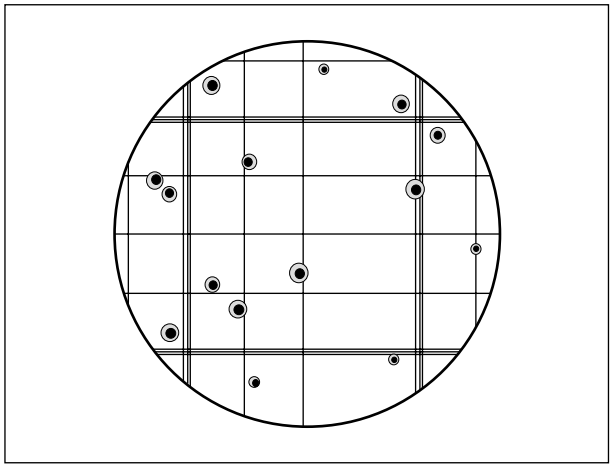


Figure 6.25.3 White blood cells.

With the larger ones you can often make out the individual granules shimmering and moving within the cell, and the nucleus (lobulated in neutrophils).

Red blood cells

These are smaller than WBCs, and do not have any content or granular appearance.

If the red cells are present because of trauma (for example after an injury, postsurgery) or a UTI, they will either look just like red cells in the blood – biconcave discs – or will all look a bit shrunken and wrinkled, or a bit swollen. The important thing is that they all look the same.

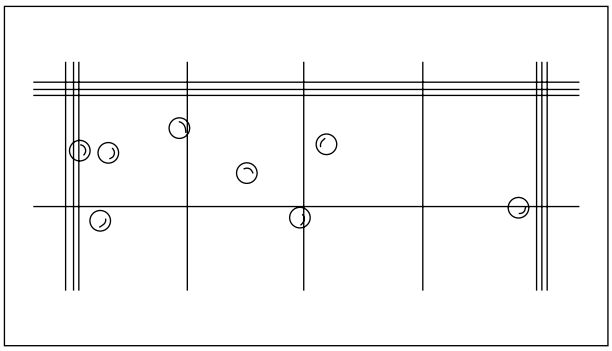


Figure 6.25.4 Red blood cells.

If the red cells are in the urine because of kidney inflammation (glomerulonephritis), they are usually smaller, but are also all different shapes. This is probably because they get damaged as they pass down the tubules of the kidney. Sometimes the red cells are very bizarre shapes. They are referred to as "glomerular" red cells.

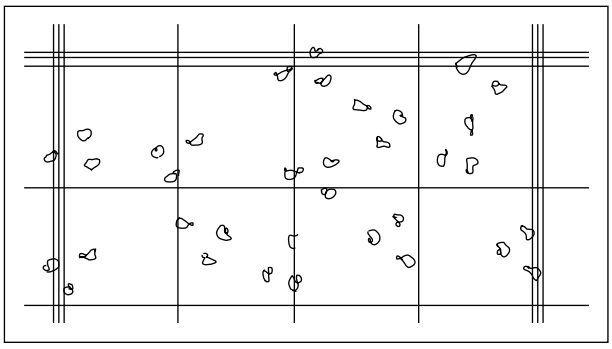


Figure 6.25.5 Glomerular red cells.

Epithelial cells

These are very large, flat cells with an easily seen, round nucleus. They are from the vagina, and are only seen in

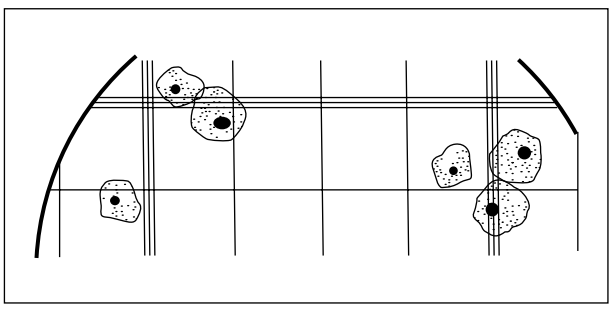


Figure 6.25.6 Epithelial cells.

the urine of older girls, where they are common. If you see large numbers it suggests particularly heavy vaginal contamination.

Casts
These indicate kidney inflammation (glomerulonephritis). They consist of abnormal kidney tubule contents that have solidified, and have retained the shape of the tubule as they have been passed into the urine.

- Pure protein casts look glass-like, and are called hyaline.
- Those consisting of debris (dead tubule cells in acute tubular necrosis) are called granular casts.
- Some casts are made up of red or white cells. Many casts consist of a mixture of these.

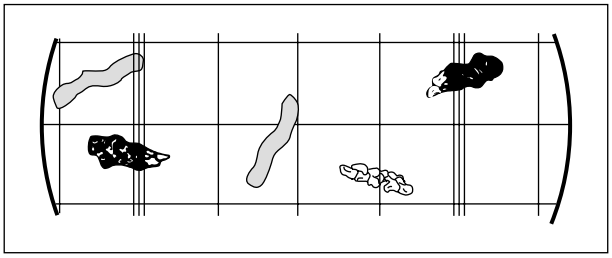


Figure 6.25.7 Casts.

Debris
Contaminated urine samples often contain a variety of debris. Some elements have an obvious origin, such as cotton fibres, but others cannot be identified.

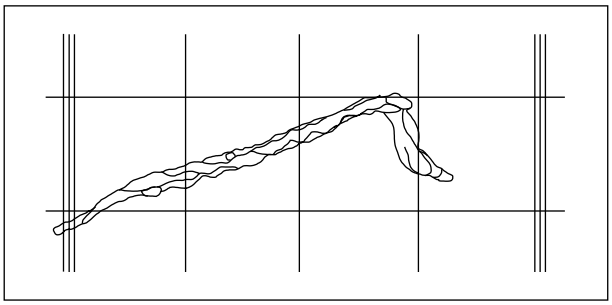


Figure 6.25.8 Debris.

Crystals
Often urines contain obvious crystals, whose shape allows their chemical origin to be identified. However, this is rarely of clinical significance.

The commonest "crystals" do not look much like diamonds; instead, phosphate crystals look like small black dots, either singly, or in clumps (and even in casts). They move slightly (shimmer) from Brownian motion, and can be mistaken by the unwary for small round bacteria (cocci).

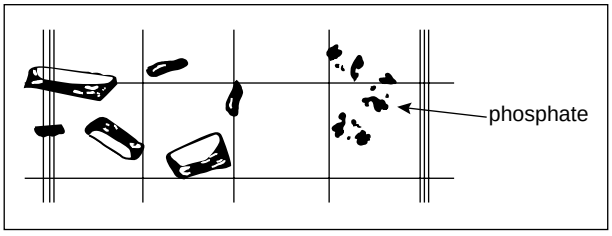


Figure 6.25.9 Crystals.

Diagnosing UTIs

Primarily, UTIs are diagnosed by looking for bacteria.

Infected urines

- About 99% of urine infections are caused by rod-shaped bacteria, bacilli.
- In most UTIs, every field you look at will have many bacteria, in some cases thousands – and they all look the same.

Therefore, when you see many bacteria in a fresh urine, all looking the same, you can be sure the child has a UTI.

If you see a urine which has at least one rod, but less than 10 in the centre of the grid (square "5"), you have to think of contamination – so collect another sample to see if the finding persists (and think about vaginal lactobacilli – see below).

What to do with a positive microscopy?

- You can start treatment at once with the best-guess antibiotic.
- Also, send the urine for culture with direct sensitivities.
- The lab will grow the bacteria to confirm which species they are, and to test which antibiotics they are sensitive to. Without direct sensitivity testing this takes two days, but with it you will usually get the result the next day.

Sterile urines

- Most urines will be sterile.
- If you see no bacteria or cells, check by looking at five 'size-A' squares (that is, about five fields).
- If you see nothing in that area, then you can be certain that the urine is not infected. **Throw it away!**
- Even if you see other elements, if there are no bacteria, it is not a UTI. Remember that you see WBCs in many children with fevers, for example with tonsillitis, or pneumonia.
- Remember that many girls have WBCs in their urine from the vagina (and often epithelial cells too).

Contaminated urines
If you see:

- More than one shape of bacteria
- Some bacteria, but also lots of debris (for example cotton fibres or many epithelial cells)
- Many bacteria in a urine that was collected some hours ago, or from a nappy that had been on the baby for hours, then collect a repeat sample – it is likely to have been contaminated. If necessary, you need to go on collecting repeat urines until one is either definitely sterile, or definitely infected.

Vaginal contamination
It is physiological for girls to void some of their urine into the vagina, so normal female urine will contain "vaginal washings".

In young girls this makes little difference to the microscopy findings. In older girls it is normal to see some epithelial cells.

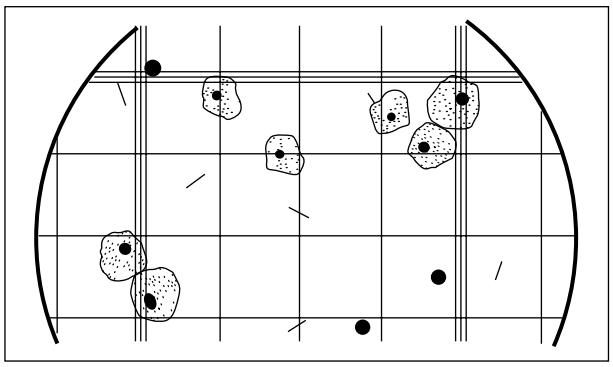


Figure 6.25.10 Urinary lactobacilli.

Also, many older girls wash lactobacilli into the urine. These are long rods, up to 4 mm or more. It is unusual for there to be large numbers, but they can cause confusion with a UTI. If you are uncertain, either ask the lab to Gram-stain them, or to culture them. Unlike the bacteria that cause UTIs, they are Gram positive.

They do not grow in conventional UTI culture media, so the lab will report a sterile urine. If you want to be absolutely certain, ask the lab to culture the urine anaerobically.

Recording the results

Labels can be printed to stick into the clinical notes. This is important because negative urines will be discarded, and this will be the only record of the test.

URINE PHASE-CONTRAST MICROSCOPY

Name: ..... Date: .....

MICRO Bacteria.....
WBC ..... RBC .....
Casts, etc .....

STICKS Protein ..... Blood .....
Glucose ..... Other .....

ACTION (tick one of the three options)

Urine not infected, sample discarded ☐

Urine contaminated, sample repeated ☐

UTI; urine sent for culture and direct sensitivities, and antibiotics started ☐

SIGN & PRINT NAME: .....

Figure 6.25.11 Labelling microscopy results.

Practise with saliva

Saliva contains bacteria, usually rods of varying lengths, and cocci. Saliva usually has some white cells present, and epithelial cells, especially if you scrape your buccal mucosa – and red cells if you scrape it hard enough.

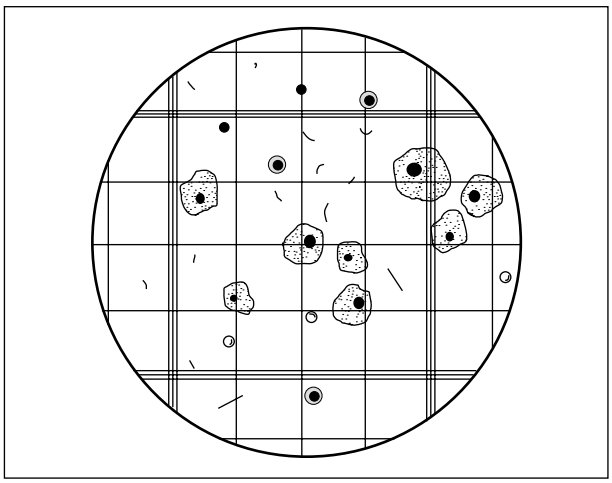


Figure 6.25.12 Microscopy of saliva.

Microscopy with saliva provides a useful technique to practise and learn with.

Counting what you see

Why?

For most clinical purposes it is not necessary to count the exact concentrations of cells or bacteria you see – estimates such as many or few are enough. Sometimes it is helpful to quantify more carefully, for example to monitor the numbers of casts in a child with glomerulonephritis.

Occasionally it is essential to count exactly the numbers seen, for example, the number of WBCs is critical to diagnose and treat peritonitis in children on peritoneal dialysis.

How?

- Calculate all the counts per microlitre.
- Count at least 10 of each element you are interested in – the number and size of the squares you need to count will therefore depend on their concentration in the urine.
- Figure 6.25.13 shows the etched grid.
  One of the central squares ("3") is 1 × 1 mm.
  With the cover-slip on, the chamber is 0.1 mm deep, so the central square has a volume of 0.1 microlitre.
- The whole grid of nine similar squares has a total volume of 0.9 microlitres.
- Note that one microlitre is one-thousandth of an millilitre.
- Thus, 100 000 bacteria/ml is equivalent to 100/microlitre, so a "significant" UTI culture would mean at least 100 bacteria per microlitre, or 10 in a central square of the grid.

How to count?

- Very infrequent elements
  Count all those in squares 1,2,3,4 and 5, and multiply by 2.
- Infrequent elements
  Count all those in large square 5, and multiply by 10.

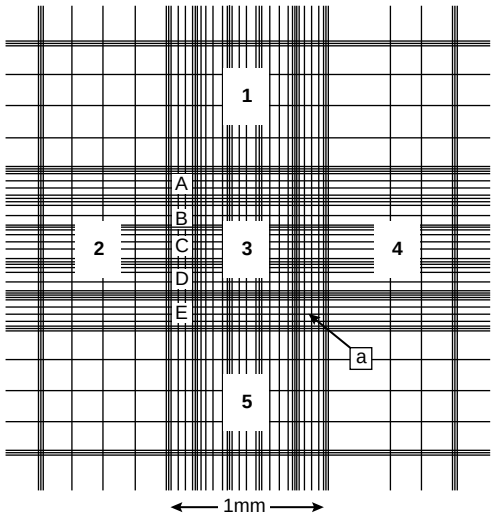


Figure 6.25.13 Counting grid.

- Frequent elements
  Count five smaller squares, for example A, B, C, D and E, and multiply by 50.
- Very frequent elements
  Count all those in square A, and multiply by 250 (for ease of calculation, multiply by 1000 and divide by 4).
- Overwhelmingly frequent elements (usually bacteria)
  Count all those in one of the smallest squares, for example square a, and multiply by 4000.

6.26 Handwashing

- Why? Not washing hands is the major cause of nosocomial infection.
- When? Before and after treating patients (injections, dressings and any personal contact, including handshaking), after using the toilet, before and after preparing/eating food.
- How?
  - Remove watches and jewellery, except wedding rings.
  - Wash with antiseptic soap for at least 20 seconds.
  - Pay special care to fingernails, around ring and between fingers.
  - Rinse thoroughly under clean water and dry fully.
  - Cover any wounds with a waterproof dressing/plaster.
  - Look after your hands by using hand cream.

We thank Francis Calder and Susern Tan for the following figures: 6.6.2; 6.7.1; 6.10.1; 6.15.1.

The figures 6.5.1, 6.6.2, 6.7.1, 6.10.1, 6.11.1, 6.12.1, 6.13.1, 6.14.2, 6.15.1, 6.17.1 were reproduced with thanks from Advanced Life Support Group. *Advanced Paediatric Life Support, 3rd ed.* London: BMJ Books, 2001.

Section 7

Appendix

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7.1 Fluid and electrolyte management

Normal requirements

Volume

Blood volume is about 100 ml/kg at birth falling to about 80 ml/kg at one year. Total body water varies from 800 ml/kg in the neonate to 600 ml/kg at one year and thereafter. Of this about two thirds (400 ml/kg) is intracellular fluid, the rest being extracellular fluid. Thus initial expansion of vascular volume in a state of shock can be achieved with relatively small volumes of fluid: 20 ml/kg will usually suffice. However, this volume is only a fraction of that required to correct dehydration if the fluid has been lost from all body compartments which it is. Clinically, dehydration is not detectable until >3–5%.

Fluid requirement can be divided into four types:

- For replacement of **insensible losses** through sweat, respiration, gastrointestinal loss etc.
- For replacement of **essential urine output**, the minimal urine output to allow excretion of the products of metabolism etc.
- Extra fluid to maintain a **modest state of diuresis**.
- Fluid to replace **abnormal losses** such as blood loss, severe diarrhoea, diabetic polyuria losses etc.

A formula for calculating normal fluid requirement is given in Table 7.1.1. It is useful because it is simple, can be applied to all age ranges and is easily subdivided. The formula gives total fluid requirements, that is for insensible losses plus essential urine output plus a modest state of diuresis.

Table 7.1.1 Normal fluid and electrolyte requirements

Body weight	Fluid requirement per day	Fluid requirement per hour
First 10 kg	100 ml/kg	4 ml/kg
Second 10 kg	50 ml/kg	2 ml/kg
Subsequent kg	20 ml/kg	1 ml/kg

Examples using table 7.1.1:

6 kg infant would require 600 ml per day

14 kg child would require 1000 + 200 = 1200 ml per day

25 kg child would require 1000 + 500 + 100 = 1600 ml per day

An alternative calculation is shown in Table 7.1.2.

Table 7.1.2 Fluid management: Maintenance fluid requirements

Body weight of child	Fluid (ml/day)
<10 kg	100–120 ml/kg
10–19 kg	90–120 ml/kg
>20 kg	50–90 ml/kg

Electrolytes

There are obligatory losses of electrolytes in stools, urine, and sweat, and these require replacement. Any excess is simply excreted in the urine.

Table 7.1.3 Electrolyte contents of body fluids

Fluid	Na (mmol/l)	K (mmol/l)	Cl (mmol/l)	HCO_3 (mmol/l)
Plasma	135–141	3.5–5.5	100–105	24–28
Gastric	20–80	5–20	100–150	0
Intestinal	100–140	5–15	90–130	15–65
Diarrhoea	7–96	34–150	17–164	0–75
Sweat	<40	6–15	<40	0–10

Table 7.1.4 Normal water, electrolyte, energy and protein requirements (provided excessive loss is not present)

Body weight	Water (ml/kg/day)	Sodium (mmol/kg/day)	Potassium (mmol/kg/day)	Energy (kcal/day)	Protein (g/day)
First 10 kg	100	2–4	1.5–2.5	110	3.00
Second 10 kg	50	1–2	0.5–1.5	75	1.50
Subsequent kg	20	0.5–1	0.2–0.7	30	0.75

Table 7.1.5 Commonly available crystalloid fluids

Fluid	Na^+ (mmol/l)	K^+ (mmol/l)	Cl^- (mmol/l)	Energy (kcal/l)
Isotonic crystalloid fluids				
Saline 0.9%	150	0	150	0
Saline 0.45%, glucose 2.5%	75	0	75	100
Saline 0.18%, glucose 4%	30	0	30	160
Glucose 5%	0	0	0	200
Saline 0.18%, glucose 4%, plus 10 mmol KCl/500 ml	30	20	50	160
Hartmann's solution* (Ringer's lactate)	131	5	111	0
Hypertonic crystalloid solutions				
Saline 0.45%, glucose 5%	75	0	75	200
Glucose 10 %	0	0	0	400
Saline 0.18%, glucose 10%	30	0	30	400
Glucose 20%	0	0	0	800

* Ringer's lactate Hartmann's also has bicarbonate as lactate 29 mmol/l and calcium 2 mmol/l.

Table 7.1.6 Commonly available colloid fluids

Colloid solutions	Na^+ (mmol/l)	K^+ (mmol/l)	Ca^{++} (mmol/l)	Duration of action (hours)	Comments
Albumin 4.5%	150	1	0	6	Protein buffers
Gelofusine	154	<1	<1	3	Gelatine
Haemaccel	145	5	12.5	3	Gelatine
Pentastarch	154	0	0	7	Hydroxyethyl starch

Some useful information

1. Percentage solution = number of grams in 100 ml
   for example 10% glucose = 10 g in 100 ml

2. One millimole = molecular weight in milligrams

3. Some atomic weights:

hydrogen	1.0
carbon	12.0
nitrogen	14.0
oxygen	16.0
sodium	23.0
phosphorus	31.0
chlorine	35.5
potassium	39.1
calcium	40.1

therefore for example:

1 mmol NaCl	= 58.5 mg
1mmol $NaHCO_3$	= 84 mg
1mmol KCl	= 74.6 mg

4. The equivalent weight of an electrolyte = molecular weight/valency
   for example Ca = 40/2
5. Useful figures to know:

30% NaCl	= 5 mmol/ml each of Na and Cl
0.9% NaCl	= 0.154 mmol/ml each of Na and Cl
15% KCl (15 g/100 ml)	= 2 mmol/ml each of K and Cl (also called concentrated or **strong KCl**)
10% Ca Gluconate (10 g/100 ml)	= 0.225 mmol/ml (note: 1 ml of CaCl 10% is equivalent to 3 ml CaGluconate 10%)
8.4% $NaHCO_3$	= 1 mmol Na/ml and 1 mmol HCO_3/ml
1ml/hour 0.9% saline	= 3.7 mmol Na in 24 hours

7.2 Example of prescription chart

NAME		WEIGHT		AGE				
DATE	TREATMENT	DOSE	REGIMEN	SIGN	NURSES: TIME GIVEN/SIG DOSE: 1.	2.	3.	4.

Figure 7.2.1. Example of prescription chart.

7.3 WHO Treatment Plan A (modified)

Home therapy to prevent dehydration and malnutrition

Mothers should be taught how to
- Give the child more fluids than usual, to prevent dehydration
- Continue to feed the child, to prevent malnutrition
- Take the child to a health worker if there are signs of dehydration or other problems

Rule 1: Give the child more fluids than usual, to prevent dehydration

What fluids to give

Many countries have designated recommended home fluids. Wherever possible, these should include at least one fluid that normally contains salt (see below). Plain clean water should also be given. Other fluids should be recommended that are frequently given to children in the area, that mothers consider acceptable for children with diarrhoea, and that mothers would be likely to give in increased amounts when advised to do so.

Suitable fluids

Most fluids that a child normally takes can be used. It is helpful to divide suitable fluids into two groups:

A) Fluids that normally contain salt, such as:
- ORS solution
- salted drinks (for example salted rice water or a salted yoghurt drink)
- vegetable or chicken soup with salt.

B) Fluids that do not contain salt, such as:
- plain water
- water in which a cereal has been cooked (for example unsalted rice water)
- unsalted soup
- yoghurt drinks without salt

- green coconut water
- weak tea (unsweetened)
- unsweetened fresh fruit juice.

Unsuitable fluids

AVOID drinks sweetened with sugar, which can cause osmotic diarrhoea and hypernatraemia. Some examples are: soft drinks, **sweetened** fruit drinks, sweetened tea. Other fluids to avoid are those with stimulant, diuretic or purgative effects, for example, coffee, coca cola and some medicinal teas or infusions.

How much fluid to give

The general rule is: give as much fluid as the child wants until diarrhoea stops. As a guide, after each loose stool, give:

- children under 2 years of age: 50–100 ml (a quarter to half a large cup) of fluid
- children aged 2 to 10 years: 100–200 ml (a half to one large cup)
- older children: as much fluid as they want.

Rule 2: Continue to feed the child, to prevent malnutrition

✔ Feeding should be continued during diarrhoea and increased afterwards. Food should never be withheld and the child's usual foods should not be diluted. **Breastfeeding should always be continued**. The aim is to give as much nutrient rich food as the child will accept. Most children with watery diarrhoea regain their appetite after dehydration is corrected, whereas those with bloody diarrhoea often eat poorly until the illness resolves.

When food is given, sufficient nutrients are usually absorbed to support continued growth and weight gain. Continued feeding also speeds the recovery of normal intestinal function, including the ability to digest and absorb various nutrients. In contrast, children whose food is restricted or diluted lose weight, have diarrhoea of longer duration, and recover intestinal function more slowly.

What foods to give

This depends on the child's age, food preferences and pre-illness feeding pattern; cultural practices are also important. In general, foods suitable for a child with diarrhoea are the same as those required by healthy children. Specific recommendations are given below.

Milk

- Infants of any age who are breastfed should be allowed to breastfeed as often and as long as they want. Infants will often breastfeed more than usual; this should be encouraged.
- Infants who are not breastfed should be given their usual milk feed (or formula) at least every three hours, if possible by cup. Special commercial formulas advertised for use in diarrhoea are expensive and unnecessary; they should not be given routinely. Clinically significant milk intolerance is rarely a problem.
- Infants below 4 months of age who take breast milk and other foods should receive increased breastfeeding. As the child recovers and the supply of breast milk increases, other foods should be decreased (and given by cup, not bottle). This usually takes about one week. If possible, the infant of this age should become **exclusively** breastfed.

There is no value in routinely testing the stools of infants for pH or reducing substances. Such tests are oversensitive, often indicating impaired absorption of lactose when it is not clinically important. It is more important to monitor the child's clinical response (i.e. weight gain, general improvement). Milk intolerance is only clinically important when milk feeding causes a prompt increase in stool volume and a return or worsening of the signs of dehydration, often with loss of weight.

Other foods

If the child is at least 6 months old or is already taking soft foods, he or she should be given cereals, vegetables and other foods, in addition to milk. If the child is over 6 months and such foods are not yet being given, they should be started during the diarrhoea episode or soon after it stops.

Recommended foods should be culturally acceptable, readily available, have a high content of energy and provide adequate amounts of essential micronutrients. They should be well cooked, and mashed or ground to make them easy to digest; fermented foods are also easy to digest. Milk should be mixed with a cereal. If possible, 5 to 10 ml (1 teaspoon to 1 tablespoon) of vegetable oil should be added to each serving of cereal. Meat, fish or egg should be given, if available. Foods rich in potassium, such as bananas, green coconut water and fresh fruit juice are beneficial.

How much food and how often

Offer the child food every three or four hours (six times a day). Frequent, small feedings are tolerated better than less frequent, large ones.

After the diarrhoea stops, continue giving the same energy rich foods and provide one more meal than usual each day for at least two weeks. If the child is malnourished, extra meals should be given until the child has regained normal weight-for-height.

Rule 3: Take the child to a health worker if there are signs of dehydration or other problems

The mother should take her child to a health worker if the child:

- starts to pass many watery stools
- has repeated vomiting
- becomes very thirsty
- is eating or drinking poorly
- develops a fever
- has blood in the stool
- the child does not get better in three days.

7.4 History and examination sheet for severe malnutrition

HISTORY

Reg. No ..... Parent's name: ..... First name: .....

Age ..... Date of birth (d/m/y) ...../...../..... Sex .....

Date of examination: ...../...../..... Examiner's name ..... Status .....

Who is giving the history? *patient/mother/father/sister/grandmother/aunt/other*

Is this person the main caretaker for the patient at home? yes/no If no, who is the caretaker? .....

HISTORY OF PRESENT ILLNESS

How long has the patient been ill? ..... (h/d/wk/mo/yr)

What are the complaints – in the patient's own words – and how long has each been present?

1 .... (h/d/wk/mo/yr)

2 .... (h/d/wk/mo/yr)

3 .... (h/d/wk/mo/yr)

4 .... (h/d/wk/mo/yr)

Describe the details of the complaints, how they have progressed, and the factors associated with each one

.....

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.....

.....

SYSTEMATIC QUESTIONS (GIVE ADDITIONAL DETAILS OF ABNORMALITIES ABOVE)

Appetite hungry/ normal/ poor/ very poor **Weight** is decreasing/ steady/ increasing ..... (d/ wk/ mo)
Swelling: none/ feet/ legs/ face/ all over ..... (d/wk/mo) **Eyes** sunken no/ recent/ longstanding
Diarrhoea N/Y ..... (h/d/wk/mo) **Stools per day** ..... Normal/ watery/ soft/ blood/ mucus/ green/ pale
Repeated episodes of diarrhoea: N/Y
Vomiting N/Y ..... (h/d/wk/mo) No. per day .....
Breathing: normal/ fast/ noisy/ difficult for ..... (h/d/wk) **Cough:** N/Y for ..... (d/wk/mo)
Fever N/Y **Convulsions** N/Y **Unconsiousness** N/Y
Treatment: Patient has already seen Dr/ Clinic/ Hospital/ Traditional healer ..... times for this illness.
Treatment given .....

PAST AND SOCIAL HISTORY

Past diseases: describe .....
Mother/ father absent N/Y reason..... .....(wk/mo/yr) **Patient:** twin/ fostered/ adopted/ orphan
Gestation: early/ normal or.....(wk/mo) **Birth weight:** large/ normal/ small or ..... (kg/lb)
Mother's age .....yr No. live births ..... No. Living children .....
Family eating together: No. adults ..... No. children .....
Resources (food/income/crops/livestock).....

DIET HISTORY

Breastfeed alone for .....(wk/ mo) **Age stopped breastfeeding** ..... (wk/mo)
Food before ill breast/ milk/ porridge/ family plate/ fruit/ leaves/ drinks/ other
Food since ill breast/ milk/ porridge/ family plate/ fruit/ leaves/ drinks/ other
Describe: .....
Last 24h – describe: .....

EXAMINATION

Reg. No. ..... Parent's name ..... First name ..... Age ..... (d/m/y) Sex .....

General does the patient look: not ill/ ill/very ill/comatose
Mood and behaviour normal/apathetic/inactive/irritable/repeated movements
Development/regression Patient can: sit/crawl/stand/walk

EAR NOSE & THROAT

Eyes normal/ conjunctivitis/ xerosis/ keratomalacia mild/mod/severe
Mouth normal/sore/red/smooth tongue/candida/herpes/angular stomatitis
Membrane colour: normal/pale/jaundiced/cyanosed **Gums** normal/bleeding
Ears normal/discharging **Teeth** number __/__ normal/caries/plaque

RESPIRATORY SYSTEM AND CHEST

Breathing normal/noisy/asymmetrical/laboured/wheeze/indrawing
Rate ..... **/min or** more/less than 50/60 **Chest** normal/asymmetric/pigeon/sulcus

CARDIOVASCULAR SYSTEM AND HYDRATION

Oedema none/+/++/+++/uncertain feet/pretibial/hands/face/generalised
Hydration normal/ dehydrated/shock/uncertain **Passing urine** N/Y
Eyes normal/ sunken/ staring **Peripheries** normal/ warm/ cold
Pulse rate ..... /min normal/strong/weak **Heart sounds** normal/gallop/murmur

GASTRO-INTESTINAL

Stool not seen/normal/soft/watery/green/pale/mucus/blood
Abdomen: normal/distended/ tender/visible peristalsis
Bowel sounds: normal/active/quiet/absent **splash** N/Y
Liver ..... cm below costal margin normal/firm/hard smooth/irregular
Spleen not felt/felt/large – normal/firm/hard – tender/painless

NERVOUS SYSTEM

Tone normal/stiff/floppy
Meninges normal/stiff neck/Brudzinski/fontanelle bulging
Reflexes normal/increased/decreased/absent

SKIN HAIR BONE LYMPH NODES

Skin change none/mild/mod/severe peeling/raw/ulcers infection/cuts/bruises
Perineum normal/rash/raw/candida **Purpura** N/Y
Hair black/brown/red/blond normal/easily plucked/balding
Scabies none/local/generalised **Eyelash** normal/long
Lymph nodes none/groin/axilla/neck Tender/painless Soft/firm/hard/fixed
Ribs ends normal/swollen/displaced **Gynecomastia** N/Y

Describe abnormalities below

.....

.....

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7.5 Formulas and recipes for severely malnourished children

Table 7.5.1 Formula for home-made ReSoMal: rehydration solution for severely malnourished children

Ingredient	Amount
Water	2 litres
WHO-ORS	One 1-litre packet*
Sucrose	50 g
Electrolyte/mineral solution**	40 ml

* 3.5 g sodium chloride, 2.9 g trisodium citrate dihydrate, 1.5 g potassium chloride, 20 g glucose.

** See below for the recipe for the electrolyte/mineral solution. If this cannot be made up, use 45 ml of KCl solution (100 g KCl in 1 litre of water) instead.

ReSoMal properly formulated contains approximately 45 mmol Na, 40 mmol K, and 3 mmol mg and 125 mmol glucose/litre.

For the use of ReSoMal in the management of dehydration in severely malnourished children, follow the guidelines given in Chapter 3.16. Its osmolarity is 300 mosmols/litre.

Formula for concentrated electrolyte/mineral solution

This is used in the preparation of starter and catch-up feeding formulas and ReSoMal. Sachets containing pre-mixed electrolytes and minerals are produced by some manufacturers. If these are not available or affordable, prepare the solution (2500 ml) using the ingredients shown in Table 7.5.2.

Table 7.5.2 Formula for concentrated electrolyte/mineral solution

Ingredient	gram	mmol/20 ml
Potassium chloride:	224	24 mmol
Tripotassium citrate	81	2 mmol
Magnesium chloride:	76	3 mmol
Zinc acetate:	8.2	300 micromol
Copper sulphate:	1.4	45 micromol
Water: make up to	2500 ml	

If available, also add selenium (0.028 g of sodium selenate) and iodine (0.012 g of potassium iodide) per 2500 ml.

- Dissolve the ingredients in cooled boiled water.
- Store the solution in sterilized bottles in the fridge to retard deterioration. Discard if it turns cloudy. Make fresh each month.
- Add 20 ml of the concentrated electrolyte/mineral solution to each 1000 ml of milk feed.

If it is not possible to prepare this electrolyte/mineral solution and pre-mixed sachets are not available, give K, Mg and Zn separately. Make a 10% stock solution of potassium chloride (100 g in 1 litre of water) and a 1.5% solution of zinc acetate (15 g in 1 litre of water).

For the oral rehydration solution ReSoMal, use 45 ml of the stock KCl solution instead of 40 ml electrolyte/mineral solution.

For milk feeds F-75 and F-100, add 22.5 ml of the stock KCl solution instead of 20 ml of the electrolyte/mineral solution to 1000 ml of feed. Give the 1.5% zinc acetate solution by mouth 1 ml/kg/day. Give 0.3 ml/kg of 50% magnesium sulphate intramuscularly once to a maximum of 2 ml.

Table 7.5.3 Recipes of re-feeding formulas F-75 and F-100

Ingredient	F-75[ab] (starter)	F-75[c] (starter: cereal-based)	F-100[d] (catch-up)
Dried skimmed milk (g)	25	25	80
Sugar (g)	100	70	50
Cereal flour (g)	–	35	–
Vegetable oil (g)	27	27	60
Electrolyte/mineral solution (ml)	20	20	20
Water: make up to (ml)	1000	1000	1000
Contents per 100 ml			
Energy (kcal)	75	75	100
Protein (g)	0.9	1.1	2.9
Lactose (g)	1.3	1.3	4.2
Potassium (mmol)	4.0	4.2	6.3
Sodium (mmol)	0.6	0.6	1.9
Magnesium (mmol)	0.43	0.46	0.73
Zinc (mg)	2.0	2.0	2.3
Copper (mg)	0.25	0.25	0.25
% energy from protein	5	6	12
% energy from fat	32	32	53
Osmolality (mOsm/litre)	413	334	419

[a] A comparable starter formula can be made from 35 g whole dried milk, 100 g sugar, 20 g oil, 20 ml electrolyte/mineral solution, and water to make 1000 ml. If using fresh cows' milk, take 300 ml milk, 100 g sugar, 20 ml oil, 20 ml electrolyte/mineral solution, and water to make 1000 ml.

[b] Isotonic versions of F-75 (280 mOsmol/litre) are available commercially, in which maltodextrins replace some of the sugar, and in which all the extra nutrients (K, Mg and micronutrients) are incorporated.

[c] Cook for 4 minutes. This may be helpful for children with dysentery or persistent diarrhoea.

[d] A comparable catch-up formula can be made from 110 g whole dried milk, 50 g sugar, 30 g oil, 20 ml electrolyte/mineral solution, and water to make 1000 ml. If using fresh cows' milk, take 880 ml milk, 75 g sugar, 20 ml oil, 20 ml electrolyte/mineral solution, and water to make 1000 ml.

Mix the milk, sugar, oil and electrolyte/mineral solution to a paste, and then slowly add the warm boiled water. Make up to 1000 ml. If available, use an electric blender or hand whisk.

Table 7.5.4 Volumes of F-75 per feed.

Child's weight (kg)	2-hourly (ml/feed)	3-hourly (ml/feed)	4-hourly (ml/feed)
2.0	20	30	45
2.2	25	35	50
2.4	25	40	55
2.6	30	45	55
2.8	30	45	60
3.0	35	50	65
3.2	35	55	70
3.4	35	55	75
3.6	40	60	80
3.8	40	60	85
4.0	45	65	90
4.2	45	70	90
4.4	50	70	95
4.6	50	75	100
4.8	55	80	105
5.0	55	80	110
5.2	55	85	115
5.4	60	90	120
5.6	60	90	125
5.8	65	95	130
6.0	65	100	130
6.2	70	100	135
6.4	70	105	140
6.6	75	110	145
6.8	75	110	150
7.0	75	115	155
7.2	80	120	160
7.4	80	120	160
7.6	85	125	165
7.8	85	130	170
8.0	90	130	175
8.2	90	135	180
8.4	90	140	185
8.6	95	140	190
8.8	95	145	195
9.0	100	145	200
9.2	100	150	200
9.4	105	155	205
9.6	105	155	210
9.8	110	160	215
10.0	110	160	220

7.6 Examples of care charts

NAME ..... AGE ..... DATE OF ADMISSION..... HOSPITAL No. .....

DATE							
TIME							
TEMPERATURE							
PULSE							
RESPIRATIONS							
BP							
SaO_2%							
AVPU							
WEIGHT							
OXYGEN THERAPY							
CONVULSIONS							
FLUIDS – RECORD AMOUNT AND TYPE							
INTRAVENOUS	1						
	2						
NASOGASTRIC	1						
	2						
ORAL	1						
	2						
24 HOUR TOTAL INPUT							
OUTPUT – URINE							
OUTPUT – STOOL							
OUTPUT – VOMIT							
BLOOD PRODUCTS							
POSITION CHANGE							
SKIN CARE							
EYE CARE							
MOUTH CARE							
PAIN RELIEF							
GENERAL ASSESSMENT							
HEALTH EDUCATION							

OUTCOME .....

Figure 7.6.1. Example of care chart for sick children.

NAME ..... D.O.B..... UNIT No. .....

DATE															
		AM	PM	AM	PM	AM	PM	AM	PM	AM	PM	AM	PM	AM	PM
TEMPERATURE															
PULSE															
RESPIRATIONS															
CONVULSIONS															
WEIGHT															
MILK FEEDS NG/ORAL	1														
	2														
INDICATE	3														
IF	4														
OTHER	5														
THAN	6														
MILK	7														
	8														
IV FLUIDS															
BLOOD PRODUCTS															
24 HR INPUT TOTAL															
OXYGEN THERAPY															
PHOTOTHERAPY															
OUTPUT – URINE															
OUTPUT – STOOL															

Figure 7.6.2. Example of care chart for neonatal unit.

7.7 Estimating body surface area

Body surface area is commonly used in paediatrics to calculate drug dosages. This is because in children beyond the neonatal period, metabolic rate, renal clearance, and some other bodily functions vary more closely with surface area than they do with weight.

In practice, using surface area as the basis of prescribing means that smaller children receive relatively more drug than they would if weight was being used.

For many drugs, the therapeutic margin is wide enough for it not to matter which method of dosage calculation is used, but for some it makes a significant difference, and avoids ineffective underprescribing in smaller children. Examples where it should be used are most cancer chemotherapy agents, and corticosteroids.

Though there are several widely used formulae and nomograms that relate surface area to body weight and height, Boyd's self-adjusting power equation that relates it to body weight alone has been shown to be the most reliable method of estimation. A major advantage is that for any particular weight, it is merely necessary to read the surface area from a table (7.7.1). This is quicker and reduces the chance of making an error almost to zero.

Table 7.7.1 Boyd's equation for estimating body surface area

Weight kg	SA m^2	Weight kg	SA m^2	Weight kg	SA m^2
0.7	0.07	12	0.56	38	1.23
1.0	0.10	13	0.59	40	1.27
1.6	0.14	14	0.62	42	1.32
2.0	0.16	15	0.65	44	1.36
2.6	0.19	16	0.68	46	1.40
3.0	0.21	17	0.71	48	1.44
3.6	0.24	18	0.74	50	1.48
4.0	0.26	19	0.77	52	1.52
4.5	0.28	20	0.79	54	1.56
5.0	0.30	22	0.85	56	1.60
5.5	0.33	24	0.90	58	1.63
6.0	0.35	26	0.95	60	1.67
7.0	0.38	28	1.00	65	1.76
8.0	0.42	30	1.05	70	1.85
9.0	0.46	32	1.09	75	1.94
10.0	0.49	34	1.14	80	2.03
11.0	0.53	36	1.19	90	2.19

7.8 Normal values for vital clinical signs and laboratory measurements

For those children who are seriously unwell

- Is the child alert, sleepy, irritable?
- Is there an increased breathing rate?
- Is there a rapid/slow heart rate – is the pulse weak? is it bounding?
- Examine depth of breathing – is it shallow?
- Is breathing noisy? – stridor, wheezing, grunting?
- Is there nasal flaring (nares moving in and out with breathing)?
- Is there tracheal tug (marked inward movement at trachea when breathing) or the use of accessory muscles to help breathe or intercostal/subcostal recession?
- Is the skin mottled?
- Look at colour of skin, lips, nail beds for cyanosis (going blue)
- Check for capillary refill, should be <3 sec (5 sec pressure on sternum)
- Check O_2 saturations if possible.

For those with anaemia

- Is there tiredness/lethargy?
- Is there pallor of the skin, mucous membranes, gums, insides of eyelids, fingernails?
- Is there shortness of breath?
- Ask if the child's stools are black – ? blood or on iron supplements

For those with blood clotting disorders

- Are there bleeding gums when eating/brushing teeth?
- Are there nose bleeds?
- Is there excessive bruising?
- Is there bruising in unexpected places (exclude non-accidental injury)?
- Are there petechiae (tiny, flat, red or purple spots on skin or mucous membranes caused by local haemorrhage)? Do a tourniquet test (Chapter 4.15). <5–10 petechiae in 2.5 cm circle on forearm (pressure halfway systolic to diastolic for 5 minutes)

Normal heart rates

In an infant irregular heart rates and one consistently <90/minute or >180/minute always merits an ECG. Heart rates during crying can be up to 220/minute in healthy infants. Heart rates down to 40/minute can occur during sleep in normal/healthy children.

Table 7.8.1 Average respiratory rates

Age	Breaths per minute
Newborn	35
1 to 11 month	30
2 yr	25
4 yr	23
6 yr	21
8 yr	20
10–12 yr	19
14–16 yr	17–18
18 yr	16–18

Table 7.8.2 Normal core body temperatures

Centigrade	Fahrenheit
Infant: 36.5–37.5°	97.7–99.5°
Child: 36.0–37.2°	96.8–98.6°

Table 7.8.3 Normal blood pressure

Blood pressure	Systolic (mmHg)	Diastolic (mmHg)
Birth (12 hr, less than 1000 g)	39–59	16–36
Birth (12 hr, 3 kg)	50–70	25–45
Neonate (96 hr)	60–90	20–60
Infant	87–105	53–66
Toddler	95–105	53–66
7 yr	97–112	57–71
15 yr	112–128	66–80

Laboratory blood test values

Table 7.8.4 Haemoglobin (normal values)

Age		Hb g/dl
1–3 day		14.5–22.5
2 week		14.5–18.0
6 month		10.0–12.5
1–5 yr		10.5–13.0
6–12 yr		11.5–15.0
12–18 yr	male	13.0–16.0
	female	12.0–16.0

Table 7.8.5 Platelets

Age	Platelets 10^9/litre
Newborn	84–478
Child	150–400

Table 7.8.6 White blood cells and ESR

	Age	Values
ESR	All ages	0–10 mm/hr
WBC	1–2 days	9.0–34.0 × 10^9/litre
	Neonates	6.0–19.5 × 10^9/litre
	1–3 yr	6.0–17.5 × 10^9/litre
	4–7 yr	5.5–15.5 × 10^9/litre
	8–13 yr	4.5–13.5 × 10^9/litre
Lymphocytes	>1 yr	median 4.1–6.0 × 10^9/litre

Table 7.8.7 Blood chemistry

Substance	Age	Value range	
Albumin	Preterm	18–30g/l	
	Full term – <1 week	25–34g/l	
	<5 yr	39–50g/l	
	5–19 yr	40–53g/l	
Amylase	All ages	30–100 units per 1	
ASO titre	2–5 yr	120–160 Todd units	
	6–9 yr	240 Todd units	
	10–12 yr	320 Todd units	
Bicarbonate	All ages	Arterial: 21–28 mmol/l Venous: 22–29 mmol/l	
Bilirbuin conjugated	>1 yr	0–3.4 micromol/l	
		Total	Ionised
Calcium	0–24 hr	2.3–2.65 mmol/l	1.07–1.27 mmol/l
	24 hr–4 days	1.75–3.0 mmol/l	1.00–1.17 mmol/l
	4–7 days	2.25–2.73 mmol/l	1.12–1.23 mmol/l
	Child	2.15–2.70 mmol/l	1.12–1.23 mmol/l
Chloride	Neonate	97–110 mmol/l	
	Child	98–106 mmol/l	
Creatinine	Neonate	27–88 micromol/l	
	Infant	18–35 micromol/l	
	Child	27–62 micromol/l	
Glucose	Preterm	1.4–3.3 mmol/l	
	0–24 hr	2.2–3.3 mmol/l	
	Infant	2.8–5.0 mmol/l	
	Child	3.3–5.5 mmol/l	
Iron	Child	4–33 micromol/l	
TIBC	Infant	17.9–71.6 micromol/l	
	Child	44.8–71.6 micromol/l	
Lead	Child	<0.48 micromol/l	
Magnesium	0–7 days	0.48–1.05 mmol/l	
	7 days–2 yr	0.65–1.05 mmol/l	
	2–14 yr	0.60–0.95 mmol/l	
Osmolality	Child	Serum 276–295 mosmol/l	
Alkaline phosphatase	<9 yr	145–420 units per 1	
Inorganic phosphorus	0–5 days	1.55–2.65 mmol/l	
	1–3 yr	1.25–2.10 mmol/l	
	4–11 yr	1.20–1.80 mmol/l	
	12–15 yr	0.95–1.75 mmol/l	
Potassium	<2 months	3.0–7.0 mmol/l	
	2–12 months	3.6–6.0 mmol/l	
	>1 yr	3.5–5.0 mmol/l	
Sodium	Newborn	134–146 mmol/l	
	Infant	139–146 mmol/l	
	Child	138–146 mmol/l	
Retinol/ Vitamin A	1–6 yr	0.70–1.5 micromol/l	
	7–12 yr	0.9–1.7 micromol/l	
	13–19 yr	0.9–2.5 micromol/l	
Urea	Child	2.5–6.6 mmol/l	
Zinc	Child	9.8–18.1 micromol/l	

Oxygen saturation

95%–100% (depends on altitude and corrections will be needed for those living >1000 metres above sea level. (see chapter 3.19)

Blood gases (normal arterial range)

pH	7.35–7.45
P_{CO_2}	4.5–6.0 kPa (35–45 mmHg)
O_2	10–13 kPa (75–98 mmHg)

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7.9 Clinical tests for brain stem death

Clinical prerequisites for testing

Preconditions

- Cause of the coma is known.
- Cause of the coma is irreversible.
- ***Patient is deeply unconscious on a ventilator***.
- Coma is not due to suppressant drugs or alcohol.
- Patient is cardio-vascularly optimised.

Exclusions

- No hypothermia (below 35°C).
- No endocrine or metabolic disturbances.
- Muscle relaxants excluded (***use peripheral nerve stimulator if necessary***).
- Patient has no circulating or therapeutic levels of any drugs that could cause coma.

Clinical tests

1 The pupils are fixed and dilated and do not react to light.
2 Absent corneal reflex.
3 The vestibular ocular reflexes are absent (no eye movements occur during injection of 50 mls of ice cold water into each external auditory meatus in turn).
4 No motor responses within the cranial nerve distribution can be elicited on stimulation of any somatic area.
5 There is no cough or gag reflex.
6 Positive apnoea test (refer to apnoea test specifics, box below).

Specifics of the apnoea test

- The P_{CO_2} should be >5.3 kPa (>40 mmHg) prior to testing and should rise to at least 6.7 kPa (50 mmHg) during the test.
- The patient should be preoxygenated with 100% oxygen for 10 minutes prior to testing and baseline arterial blood gases taken.
- Disconnect patient from ventilator but continue oxygenation by administering 6 l/min of oxygen via a fine bore catheter down the endotracheal tube.
- Observe patient for 10 minutes for any respiratory effort and ensure P_{CO_2} has risen above 6.7 kPa (50 mmHg).
- Reconnect patient to the ventilator.
- Discontinue testing if any hypotension, cardiac arrhythmias or hypoxia occurs.

Time of death

Death is pronounced at completion of the second test. In the UK legal time of death is when the first test indicates brain stem death.

- The declarations of brain death must be recorded in the medical notes with the date and time. The death certificate may be issued at this time if the coroner is not involved. (UK practice)
- Spinal reflexes – the spinal cord may continue to function after the death of the brain stem. The resulting limb movement may cause distress to both family and staff caring for the patient.
- After the second set of brain stem death tests are completed and the patient has been certified dead, muscle relaxants may be given to suppress spinal reflexes to prevent further distress to the family.

Timing of tests

- At least 6 hours must have elapsed since the onset of coma prior to testing for brain death.
- If cardiac arrest is the cause of coma or the patient has received a general anaesthetic, 24 hours must have passed since the onset of coma prior to testing for brain death. (UK practice)
- The tests are performed twice and the interval between the tests varies between 1–4 hours or at the discretion of the consultant in charge of the patient.

Who can perform brain stem death tests?

- The consultant in charge of the patient and one other clinically independent consultant or senior doctor, both of whom should have been qualified for at least five years.
- The doctors conducting the tests must not be members of a transplant team.

Paediatric guidelines for brain stem testing (British Paediatric Association 1991)

- Child should be 37 weeks' gestation plus 2 months of age.
- The tests should be conducted by senior doctors.

7.10 Low cost technology for neonatal care (Dr S Rahman)

Regular antenatal care, regionalisation of neonatal/perinatal services, development of community based neonatal services, establishment of highly technical neonatal ICU's and the provision of highly skilled neonatal human resource and infrastructure have been the mainstay of increased neonatal survival in rich countries.

Unfortunately the cost of neonatal care is very high. A major component is incurred in the care of low birth weight (LBW) babies. Although maternal interventions such as prenatal care, are relatively inexpensive, each normal birth that avoids a VLBW baby saves $59 700 in first year medical expenses in rich countries. For babies with birth weight more than 750 g, a shift of 250g at birth saves an average of $12 000–$16 000 in first year medical costs and a shift of 500 g generates $28 000 in savings. Unfortunately there has been no significant decline in the overall birth rate of VLBW babies even in rich countries like the USA where the LBW rate in 1996 was 7.4%. The situation is even more dismal in disadvantaged countries, where not only is per capita income meager, but countries are also facing the problem of high fertility and growth rates (3.2% in Pakistan). Babies of birth weights $<$2500 g account for approximately 25% of the births in some disadvantaged countries. The Infant Mortality Rates (IMR) remain very high (95/1000 in Pakistan) with up to 58% of IMR being due to Neonatal Mortality.

The meagre resources of disadvantaged countries need to be diverted towards more cost effective programmes of neonatal perinatal care such as improvements in maternal nutrition, antenatal care, prevention of sepsis, hypothermia, birth asphyxia and the management of neonatal jaundice. Looking at the burden of neonatal patient's load, even establishment of enough numbers of level 1 and 2 neonatal nurseries is a nightmare. Public funds are rarely able to fund this investment and private resources are limited only to perhaps 5–10% of the population. The development of low cost neonatal care equipment and Kangaroo baby care can contribute significantly to improving health care of newborn babies in poor countries.

Low cost equipment

Many poor countries have invented a large number of low cost gadgets to resuscitate their newborns, prevent hypothermia, treat jaundice etc. The cost difference between the locally produced and imported equipment is enormous. For example an imported resuscitator costs Rs. 350 000 in Pakistan while the resuscitator produced locally by the Neonatal Unit, Khyber Teaching Hospital, Peshawar costs only Rs. 12 000 and it provides all the functional capabilities of the imported resuscitator. Locally made phototherapies are ten times less costly than imported machines. Various centres in India and Pakistan have prepared their own infant warming beds to prevent hypothermia, resuscitation equipment for use in the community, meconium aspirators and simple oxygen supply systems. This aspect of neonatal care in the developing countries needs investment at the commercial level, which will further reduce the cost of locally produced gadgets.

Kangaroo baby care

This is also called "Skin to skin contact", a method of care of premature babies developed in Bogota, Columbia in 1979. Faced with a large number of premature babies and

very limited number of costly incubators, the pediatricians decided to use the mothers of these babies as "Human Incubators" through this technique. The method consists of positioning undressed, stable premature infants, between their mother's breasts, where they receive early skin-to-skin contact, warmth and mutual stimulation from their mothers. Typically dressed only in diapers, infants are placed in a head up vertical position that permits direct eye to eye contact, skin to skin sensations, and close proximity for auditory stimulation between mother and infant. Mothers typically wear loose fitted, front opening blouses or dresses that serve as a modified marsupial-like pocket carrier, for the baby. This low cost method has surprisingly not only improved the survival of premature babies in disadvantaged countries, but has also been tested and used in modified form in rich countries like Sweden, England, Germany, Italy and the USA.

Kangaroo Care is therapeutic for both mother and baby. The mother's body temperature and its regulatory mechanism helps to stabilize the baby's body temperature. The baby's heart rate is more stable, depth of breathing becomes more even, apnoeic spells decrease and the length of any apnoeic episodes diminishes. The oxygen saturation becomes more stable and slightly increased. Babies sleep more frequently and for longer periods during Kangaroo Care. They may enter deep sleep and stay there for 13–26 consecutive minutes. They go to sleep twice as often and have more than 2.5 times sleep when they are being held rather than when placed in a cot or Isolette. This spares more of their caloric intake for weight gain. Frequent Kangaroo Care markedly increases the baby's alertness and sociability. The periods of agitated state are diminished; the number of crying episodes and the duration of each episode are significantly reduced.

Kangaroo Care improves breast-feeding by increasing access to the breast, immediate availability of nutrition when required, attraction by the scent of breast milk, improved vigor of the baby and enhanced let down reflex. The parents feel more intimacy to their baby that positively increases their happiness and excitement. Mothers feel a very valuable positive contribution to the life and survival of their baby. There is enhanced feeling of control and sense of confidence. The level of anxiety is reduced and the ability to handle and care for very small fragile infants is markedly increased. Kangaroo Care can be offered to stable premature babies on the ventilator and has shown to improve their weaning and weight gain.

Resuscitation equipment

One of the most important ways of preventing disability in children is through effective neonatal resuscitation. Inflation of the lungs is the key and can be achieved with inexpensive equipment (see Figure 7.10.1). Although oxygen is desirable, a supply of air can be used to inflate the lungs safely provided there is a pressure relief system. In fact the equipment shown in the figure is preferable to the self-inflating bag and mask systems since it can deliver sustained inflationary pressures for 2–3 seconds, an advantage when trying to open an infant's lungs for the first time. Any neonatal unit should be able to make the system shown in the figure.

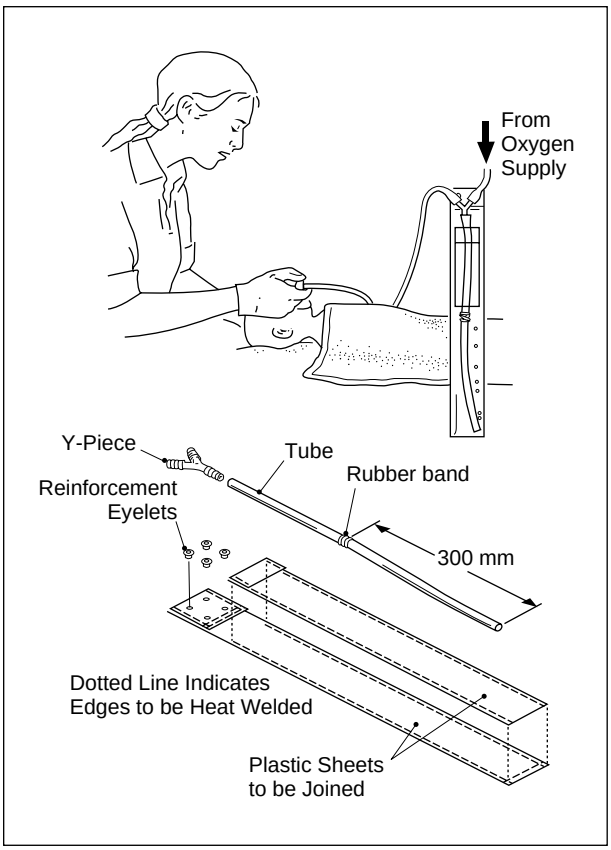

Figure 7.10.1 Low cost equipment for mask ventilation in the resuscitation of the newborn (with many thanks to Drs Ed Hey and S Richmond)

For examples of low cost equipment manufactured by Dr Rahman's department see: www.childfriendlyhealthcare.org

Further reading

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7.11 Life support training courses: equipment list

Basic airway management

Stethoscopes × 2
Paediatric bag-valve-mask with reservoir and O_2 tubing × 2
Child bag-valve-mask with reservoir and O_2 tubing × 2
Infant bag-valve-mask with reservoir and O_2 tubing × 2
T-piece with O_2 tubing × 2
O_2 Mask with reservoir and O_2 tubing × 2

O_2 mask with O_2 tubing × 2
Child mask size 03 × 2
Child mask size 02 × 2
Infant mask size 01 × 2
Infant mask size 00 × 2
Tongue depressors × 2
Nasal airway size 6.0 with safety pin × 2
Oral pharyngeal airway size 000 × 2
Yankeur suckers × 2
Suction catheters × 2

Advanced airway management

Stethoscopes × 2
Endotracheal tubes size 3.0 × 2
3.5 × 2
4.0 × 2
4.5 × 4
5.0 × 2
5.5 × 2
6.0 × 2
Laryngoscope handles × 2
Laryngoscope blades size 0 × 6 (including spares)
1 × 6 (including spares)
Extension connectors for ETT × 2
Magills forceps × 2
Suction catheters size 6 × 2
8 × 2
Yankeur suckers × 2
Naso-gastric tubes size 10 × 2
Introducers × 2
Strapping × 2
Nasal airway size 6.0 with safety pin × 2
Tongue depressors × 2

Intraosseous equipment

Intraosseous needles × 4
Cannulae size 14 Gauge × 1 (demonstration only)
Syringes 5 ml × 12
IV giving set × 1
3-way tap × 1
Syringe 20 ml × 1
Extension set × 1
0.9% NaCl 500 mls × 1
Chamois leather to cover chicken thighs
Aprons × 6
Gloves
Tablecloth
Plastic bag
Chicken thighs × 12

Basic life support equipment

Disinfectant wipes or resusci face shields or cotton wool and disinfectant
Pocket mask × 2
Spare valves × 12

Umbilical catheterisation equipment

Forceps × 10
Disposable scalpels × 10
Cotton ties × 6
Sutures
Scissors × 6
Strapping × 2
Naso-gastric tubes size 8 × 6
Aprons × 6
Gloves
Tablecloth
Plastic bottles × 6
Glove tips × 6
Plastic bag
Umbilicus × 6

Chest drain equipment

Chest drains × 4
Forceps × 3
Syringes 20 ml × 3
Sutures
Scalpels × 6
Cannulae 17 gauge × 6
Needles 23 gauge × 6
Syringes 5 ml × 6
Aprons × 6
Gloves
Tablecloth
Plastic bag
Goat's/sheep's rib cage

Defibrillation equipment

Defibrillator
Electrodes
Gel pads
Electrode gel

Scenario equipment needed

Mannikins
Bag-valve-mask × 3 sizes
Masks – all sizes
O_2 mask
O_2 mask with reservoir
Torch
Defibrillator pads
Naso-gastric tube
Suction catheters
Yankeur suckers
Syringes – large and small
Cannulae
Intraosseous needles
Monitor and defibrillator
Endotracheal tubes

Laryngoscope with 2 blades
Stethoscope
Strapping
Towels
Airways – all sizes
IV fluid giving sets
3-way taps
Tongue depressors

Cervical collars

Purple × 1
Blue × 3
Pink × 3

Extra equipment for scenarios

IV fluid giving sets with burettes × 2
Syringes 50 ml × 2
5 ml × 2
Cannulae × 2
3-way taps × 2
Torches × 2
Yankeur suckers × 2
Suction catheters × 2
Tongue depressors × 2

Mannikins

Resuscitation training baby × 5
child × 1
airway × 1
doll spares × 1 box

Miscellaneous equipment needed

Spare bulb for slide projector
Spare AA batteries
Spare AAA batteries
Flip chart paper
Note paper
Pens for board writing
Pencils/pens
Name badges
Diagrams of cardiac rhythms
Scenarios 1–12
Sellotape/Blutak
Slide projector (borrowed from PG room)
Sheets for curtains

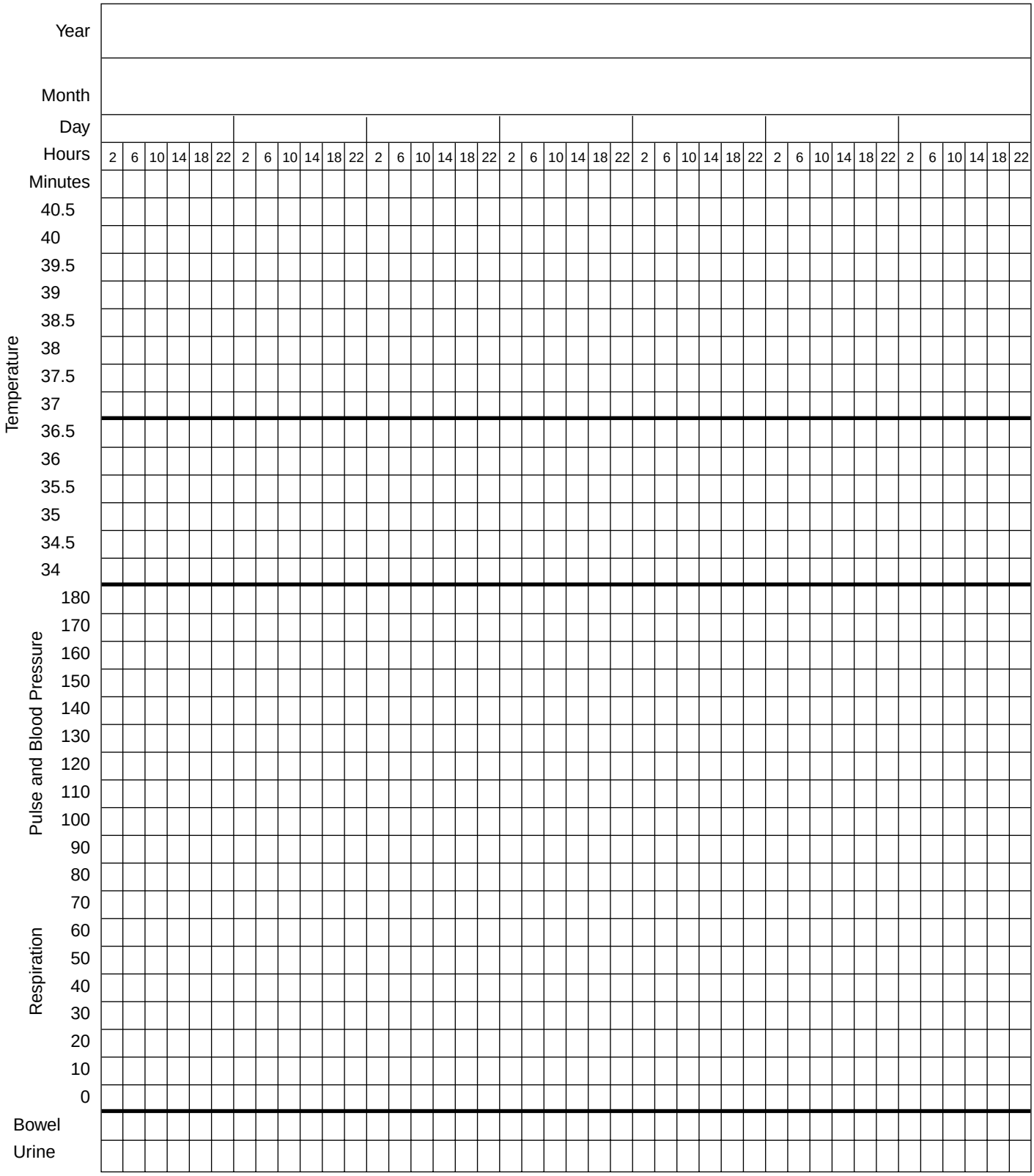
7.12 Example of a programme used for a one day paediatric pain management course

08.30–09.00	**Registration and Introduction**
09.00–10.00	**Lecture 1:** The Physiology of Pain
10.00–10.30	**Lecture 2:** The Pharmacology of Pain
10.30–10.45	*Coffee*
10.45–11.30	**Lecture 3:** The Use of Opioids
11.30–12.30	**Lecture 4:** Pain Control Teams
12.30–13.30	*Lunch*
13.30–15.30	**Workshop Scenarios** Acute Pain Procedural Pain Post-Operative Pain Chronic Pain
15.30–15.45	*Tea*
15.45–17.30	**Group Work** Pain Assessment Tools Children vs. Adults Non-Pharmacological Methods of Pain Control Cultural Aspects of Pain
17.30–18.00	**Evaluation** **Questions, Discussion** **Close**

7.13 Example of vital signs nursing chart

CHILD'S NAME
FATHER'S NAME
MOTHER'S NAME

4 hourly chart for
temperature, pulse and BP



CHILD ADVOCACY INTERNATIONAL

Figure 7.13.1 Example of vital signs nursing chart.

CHILD ADVOCACY INTERNATIONAL

FEEDING AND PROGRESS CHART

CHILD'S NAME:..... FATHER'S NAME: .....

DATE: . . . . . .
AGE: . . . . . .
FLUID REQUIREMENT: . . . . . .
FEEDING METHOD: . . . . . .

		BREATHING			FEEDING: FEEDS								IV THERAPY					HYGIENE				ELIMINATION		
TIME	COLOUR AND ACTIVITY	PHYSIO AND SUCTION	O_2	SaO_2	VOMIT	NG ASPIRATE	TYPE OF FEED	ROUTE	OFFERED	TAKEN	BM STIX	COMMENTS	FLUID	ML'S/HR	PRESSURE	VOLUME INFUSED	TOTAL	EYES	MOUTH	SKIN	TURNS	B.O.	P.U.	COMMENTS
0800																								
0900																								
1000																								
1100																								
1200																								
1300																								
1400																								
1500																								
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0400																								
0500																								
0600																								
0700																								
				TOTALS					TOTAL								TOTAL				TOTAL			

Figure 7.14.1 Example of feeding and progress chart.

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