

HANDBOOK TWO OF HOSPITAL CARE FOR INFANTS, CHILDREN AND ADOLESCENTS IN LOW RESOURCE AND EMERGENCY SETTINGS



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The authors welcome any suggestions for the next edition and for any corrections. These will be addressed in future printed and online versions.

Spine



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**HANDBOOK TWO OF HOSPITAL CARE
FOR INFANTS, CHILDREN AND
ADOLESCENTS IN LOW RESOURCE
AND EMERGENCY SETTINGS**

Handbook 2 PAEDIATRIC HOSPITAL HANDBOOK

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Instructions on the use of the two handbooks

Those parts of the chapters highlighted in light blue on the PDFs and greyscale in the printed versions, represent activities that may not be applicable to, or be able to be undertaken by, hospitals in certain low resource countries.

Editing and writing this book has been challenging for the editors and authors. We have identified what we regard as the acceptable minimum standards of treatment for all major diseases and injuries that affect infant and child, wherever they are cared for. 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 some of the best standards currently available. However, readers will notice that for most of the treatments recommended, the minimum and gold standards are identical because there are certain treatments that should be provided as essential hospital care, whatever the pressures. Key points, especially those where **inappropriate actions might be dangerous**, are presented in bold black or red font.

These handbooks should ideally be supplemented by scenario- and skill-based short training courses, combined with apprenticeship and small group teaching on the wards and in the operating theatre.

We believe in the continuing value of printed books to the practical application of healthcare, recognising that with time and improved access to high-speed internet in low resource settings, electronic materials, particularly videos and an easily accessed internet, will introduce major benefits. In the meantime, books should be available for all health workers regardless of their ability to pay for them. We hope that you will find these in-depth handbooks helpful.

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Section 1 Hospital management including infection control

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. These should be clearly defined within written personnel procedures, good training systems, and written policies and guidelines for all staff functions.

The services and facilities discussed in this text are only a basic overview. Refer to the guidelines and protocols produced by the Ministry of Health in your country or by the World Health Organisation. If these services and facilities are in place, and are managed efficiently, supported and maintained, mainline healthcare delivery has the best possibility to be effective.

Giving 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 situated. 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, direct clinical patient care and support services (laboratories, radiology, therapies, pharmacy, etc.), training for all staff and the administrative services necessary to support all of these activities. There should always be a head nurse, a head of support services and a senior doctor within the management team.

Staff management

Staff motivation and retention (human resource management) is an essential component of hospital management.

In order to provide good-quality essential health services to the people whom they serve, hospitals must put in place strategies and mechanisms to retain staff and help them to provide the best possible care for patients. The reasons for the healthcare worker crisis in hospitals in resource-limited countries include inadequate numbers of healthcare professionals, who are poorly distributed due to an unplanned 'brain drain' both regionally and internationally (attrition). According to the World Health Organization (2006), this phenomenon is caused by workers experiencing low salaries, poor, unsafe work environments, a lack of defined career paths, and poor-quality education and training.

Another important issue is the arbitrary transfer of health workers within a health facility that causes demotivation, or to a health facility in a different place, that can affect a health worker's family. Hospital officials and Ministries of Health should not transfer health workers internally or away from their families, without their full and freely given agreement.

In the light of the above factors that face health services and compromise hospital-based care, managers must endeavour to motivate the limited human resources available to ensure retention.

A systematic review of six papers evaluating the management and leadership strategies that promote healthcare worker retention in resource-limited countries has identified a number of key lessons, which can be summarised as follows.

Payment of financial incentives to healthcare professionals

This particularly refers to professionals working in unpopular rural areas. Hospitals may be run by boards which, as such, should be able sometimes to autonomously initiate better financial incentives for their staff. The above-mentioned review found that 86% of the studies

showed payment of an attractive salary and allowances was a key motivational strategy for maintaining healthcare workers in their posts. Often what made most healthcare workers leave their jobs, particularly in the public health sector, was being unable to provide basic support for their families on the salaries provided. According to a study conducted in South Africa, an increase in salary of healthcare workers has resulted in many health professionals who had previously left the public health sector, to work in private facilities, returning to the public sector.

Appreciation of healthcare workers

The community loyalty, personal commitment, and willingness to make personal sacrifices that are shown by healthcare workers must be recognised and encouraged. This means that both the hospital management and the communities that they serve must demonstrate their appreciation of health workers.

Staff must be respected for and thanked for the work they do. Personal appraisal and recognition followed by periodic awards is a motivating factor for staff retention. The views of all staff should be listened to, and they must be involved in decision making to enable the best problem-solving approaches to be identified and implemented. They are stakeholders in their hospital and their community but are often not included in decision-making.

Orderlies, porters and cleaners are just as important in providing patient care as doctors and nurses, and this needs to be recognised and made clear to all staff. It can be helpful for doctors, nurses and hospital managers to participate and help the cleaners during, for example, the monthly deep cleaning of a ward. An annual awards ceremony can be very helpful. For example, each department could be awarded certificates for:

- the most punctual member of staff
- the most improved member of staff
- the best dressed member of staff
- an award of excellence for the best all-round member of staff.

The awards could also include special categories, such as:

- the most long-serving member of staff (e.g. the refuse collector)
- an award for providing services above and beyond the 'call of duty'.

This allows awards to be made to staff who might not be in a position to further their education and to receive a certificate. Receiving such recognition in front of management and invited guests who are prominent in the hospital's catchment area is a huge honour and boost to morale.

Training and supervision

Studies conducted on human resource management for health services in Africa indicate healthcare workers' frustration at having to be assigned to responsibilities and functions for which they have limited or no training. This can be effectively managed by providing 'on-the-job' support through the provision of simple and clear guidelines on clinical procedures. Although resources may be limited for specialised advanced training, priority should be given to locally conducted ongoing training that is cost-effective, does not take health workers away from clinical areas for long periods, is sustainable, and aimed at equipping healthcare staff with the knowledge necessary to provide efficient and good-quality patient care.

Providing a programme, space and encouragement for healthcare workers to take turns to train and update their peers (e.g. an internal continuing medical education programme) can also be a low-cost and effective way for healthcare workers to share new or updated practice, as well as to develop their own teaching skills. Similarly, provision of basic information technology, computers and an Internet connection where possible is an important way of reducing professional isolation, helping healthcare workers to remain updated in their practice and to connect with the wider health community.

Some hospitals have benefited greatly by training the locally recruited nurse attendants (healthcare assistants) to second level (state-enrolled nurses) at the local nurse training school. Such nurses are usually born locally, and their families live nearby, which often commits them to continuing to serve the community in which they live. In one site that has used this approach, the hospital has been able to train over 30 nurse attendants to the second level.

The introduction of an on-call support service to nurses working out of hours can be valuable. Senior nursing staff who are knowledgeable and experienced have volunteered to help with difficult health or social problems that arise. This provides a link between management and the nursing and clinical staff, facilitating resource mobilisation and ensuring that staff are on duty at the right time and filling gaps where necessary. Many social problems for both staff and patients can be heard and addressed appropriately.

Similarly, it is important to have a suggestions box or online mechanism that allows any member of staff to air their views; anonymously if they wish to do so.

Provision of essential equipment and supplies

The lack of or inadequate provision of medical supplies, drugs and equipment in hospitals is one of the most difficult situations that healthcare workers have to cope with. Research has shown the demotivating situation that healthcare workers face when trying to treat patients without the necessary drugs and equipment. The provision of adequate and regular medical supplies, drugs and equipment is part of the answer to the ongoing question of how health systems in developing countries can best retain their health workforces. Such provision should be management and Ministry of Health priorities.

Provision of social and family amenities

Provision of basic facilities such as housing and good accommodation for healthcare staff, particularly those working in rural or remote areas, is found to have contributed immensely to retention in many resource-limited countries where they have been supported as part of a retention package.

For example, this is evident in Bansang Hospital, Gambia, West Africa, where staff retention for the past 5 years has been well recognised by authorities. Healthcare workers in Bansang Hospital are given fully furnished accommodation with water and electricity at no cost to the staff. This helps staff to increase their savings and thus boost their income, as they do not have to pay rent or utility bills. This initiative has not only enabled the hospital to retain its staff but has also served to attract other healthcare workers to come and work there.

A particular challenge for recruiting and retaining experienced healthcare professionals in remote regions is the provision of education for their children, particularly at secondary level. Arrangements for children to be educated and looked after elsewhere are offered in some countries, but this remains a barrier to retention.

Supporting and fostering a strong collective team culture is an important managerial role and a sign of good management. It helps to increase the mutual support and resilience of health workers, aides staff retention, and can bring benefits to both staff and patients. As an example, in Bansang Hospital in Gambia all staff are encouraged to grow their own vegetables and fruit for both patients and staff. Health workers there have formed their own 'Charitable Farming Association' improving their own access to fresh foods and the food of patients in the hospital. Farming activities have increased further in recent years as the hospital has been given 20 hectares of land.

A social centre for staff (with a television, sports facilities, etc.), particularly those who are not living close to or with their families, can be very helpful.

In conclusion, financial incentives can contribute to retention, but other non-financial incentives are equally likely to lead to sustainable retention. Given the economic situation in most resource-limited countries, the wages paid by Ministries of Health to public healthcare workers in prosperous economies might not be realistic in low-income countries. However, the implementation of cost-effective human resource strategies is a more realisable step forward.

There is a need to adopt both financial and non-financial strategies to retain healthcare staff.

Essential services and facilities

Hospital security and access

The security and accessibility of the hospital are of paramount importance, especially given the relative lack of police resources in many resource-limited countries. There is also a responsibility for governments, international agencies and all participants to ensure that hospitals are protected and do not become targeted during armed conflict.

At the local level, the hospital should have a perimeter fence with secure entrances where all persons attending have to demonstrate a legitimate reason for entry. All people that work in the hospital or health facility should have an official identity badge. No weapons should be allowed into the hospital, and in some countries, it may be necessary to have a metal detector to screen all visitors. The perimeter fence should be of a construction that will keep out animals.

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.

Fire safety

Regular assessments and inspections of all buildings should be undertaken in coordination with the local Fire authorities.

There should be clear written evacuation and fire policies, together with regular training for all staff, practice exercises and appropriate equipment easily accessible (e.g. fire extinguishers). Smoke and fire alarm systems should be in place and maintained. Fire retardant materials should be used for furnishings in the hospital wherever possible.

Communication systems

Good communication systems for staff, visitors and patients are essential. Ideally both outside and internal telephone systems should be available. If telephone systems are not feasible, alternative effective reliable systems of communication should be used. Keeping a regularly updated central record of health workers mobile phone numbers, and, if appropriate depending on mobile phone possession, a hospital paging system for doctors, senior nurses and managers aids communication in emergency situations.

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.

Effective communication between groups of staff in the external and internal environment improves the effectiveness and efficiency of care. This includes clear communication between referring and referral health facilities and protected time at the beginning and

end of each shift for health workers to handover details about current patients, Regular meetings should discuss individual patients, debriefs following deaths and clinical incidents, and auditing of specific aspects of clinical and unit management, such as infection control. The outcome of audit, particularly any changes in practice, needs to be available to those staff it affects, but such meetings should be constructive and educational and not used for apportioning blame.

Utilities

Water and sanitation

Hygiene within the hospital is paramount, and is dependent on a constant and high-quality water supply and adequate sanitation and washing facilities (i.e. bathrooms, showers, toilets and accessible sinks with an effective, functioning drainage system), all of which are vital if hospital-related infection is to be minimised.

Electricity

An electricity supply within the hospital, which functions independently of any power losses to the rest of the area, is mandatory. Therefore, a generator of sufficient power should be an essential item of equipment together with adequate fuel or other means to run it. The generator size can be calculated from bed dependency and operating theatre requirements. In resource-limited countries where an erratic power supply is common due to high fuel costs, solar back-ups are needed for hospitals to function efficiently and effectively. There should be special emergency circuits. Power-cut simulations should be carried out regularly to test the system. Regular maintenance is essential for safety.

Heating and ventilation

Ideally there should be a functioning 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 and in cold seasons, electric heaters should be available, when appropriate, in all areas where there are patients.

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

Laundry service (see Infection Control below)

Cleaning services (see Infection Control below)

Waste disposal systems (see Infection Control below)

Facility and utility maintenance services

Buildings, utilities and equipment

It is essential for buildings and equipment to be maintained regularly by suitably trained engineers, builders and other maintenance staff, in order to provide effective and safe care.

There is no point in having expensive medical and surgical equipment if it cannot be maintained or used. A sufficient number of bioengineers trained in the equipment present in the hospital are therefore essential.

All equipment that is used in the hospital should be robust, compatible if at all possible, suitable for the conditions and level of expertise available, and, when new, should be purchased with accompanying staff training and servicing arrangements. Any equipment or supplies

offered by donation, should only be accepted if they are compatible with existing systems and conditions, and can be safely maintained.

Porters

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

Caterers

Hospital food must be prepared under scrupulously hygienic conditions, and by staff who do not have gastroenteritis or superficial skin infections. Nutritious food should be provided free of charge to all patients and in the case of children to the family member staying with the child (particularly lactating mothers). Special diets for malnourished children and children with other specific dietary needs should be available (see Sections 29 and Sections 55, 56, 63 in handbook 1).

Administrative support

Rather than diverting away the skills of a trained nurse, dedicated reception and other administrative support staff need to be employed to aid facility managers and other non-clinical and clinical staff. There must be a staffed system for storing and processing medical and nursing records. There should be strict rules about who has access to these records, where they are stored and for how many years they are kept.

Human resource issues

Hiring and dismissing staff

There should be transparent procedures for advertising for, interviewing and employing staff. These must include non-discriminatory policies, in particular with regard to gender, age, and ethnic and religious status.

Employment and financial issues

It is essential that the medical and nursing professions in all countries are highly regarded and respected, so it is important that the salaries for doctors and nurses in the national health services reflect this. If not, the staff may have to undertake other jobs during the day and will not feel valued for their work. A lack of funding for salaries also increases the risk of corrupt practices, with some doctors taking supplies and equipment from their hospital to use in private clinics, thus depriving the poorest and most needy in the community.

Individual job descriptions and responsibilities should be agreed between healthcare professionals, their professional organisations and hospital management.

Arbitrary and compulsory transfer of staff from one place to another, at short notice and without consultation, is damaging both to morale and to the effectiveness of health services and should be avoided.

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 contracted number of hours worked (part- or full-time). On-call emergency work and its payment should also be part of the contract.

There should be a professional registration system for each country, which ensures a basic level of training, as well as a system that validates experience and ability at specific intervals after initial registration.

Concern about individual performance should be addressed sensitively and confidentially by a senior staff member on a one-to-one basis and recorded. Written guidelines should be used in a transparent way. Sometimes a period of supervised practice or retraining is appropriate.

Training and continuing staff education (see also Section 1.3 in *International Maternal and Child Health Care - A practical manual for hospitals worldwide* (2014). https://12dec1c3-e3f4-e784-7f3a-6e7903610962.filesusr.com/ugd/dd2ba4_c5284b1b150f4d70a72fe1cd710d530d.pdf Accessed 30.03.2021)

Induction training concerning hospital policies should be mandatory for all staff. Governments in well-resourced countries could encourage a support system of education for those working in less well-resourced regions.

New teaching techniques, such as skill- and scenario-based teaching (e.g. EMNCH courses) and internet based audio-visual tutorials using screen sharing and video should be introduced. Section 1.3 in: *International Maternal and Child Health Care - A practical manual for hospitals worldwide* (2014). https://12dec1c3-e3f4-e784-7f3a-6e7903610962.filesusr.com/ugd/dd2ba4_c5284b1b150f4d70a72fe1cd710d530d.pdf Accessed 30.03.2021

Professional registration requirements for healthcare workers

These will vary from one country to another. However, some form of governmental regulatory system and registration is essential. There should also be procedures governing the employment of expatriate staff in the health service.

Vetting of healthcare workers

There should be a system to ensure that all staff who are working with patients, whether they are local or from abroad, should be checked to ensure that they are suitably trained and have not been involved in the abuse of children or other criminal activity. This is also important with regard to expatriate staff. (see the Preventing Sexual Exploitation and Abuse (PSEA) program of UNICEF: PSEA assessment and PSEA toolkit for CSO partners <https://sites.unicef.org/about/partnerships/files/Information-Brief-PSEA-Assessment-PSEA-Toolkit-for-CSO-Partners-24-Jan-2020.pdf>)

Staff health (see also Section 1.17 *International Maternal and Child Health Care - A practical manual for hospitals worldwide* (2014). https://12dec1c3-e3f4-e784-7f3a-6e7903610962.filesusr.com/ugd/dd2ba4_c5284b1b150f4d70a72fe1cd710d530d.pdf Accessed 6.4.2021))

There should be a system for supporting staff health problems that may be detrimental to health workers and adversely affect patient care. Staff with physical or mental health-related problems need access to a supportive occupational health service.

There should be systems in place to protect patients from staff who are ill. This is a difficult but extremely important issue, particularly with regard to illnesses such as TB, HIV and hepatitis. Sometimes other support is necessary so that a healthcare worker's performance can be restored in the interests of all.

An important example of protecting staff health relates to Needlestick injury

Although the risk of infection depends on the nature of the pathogen, a policy should be in place to deal with this issue urgently, especially in hospitals where there are many patients with HIV infection and hepatitis. Needlestick injuries are the commonest type of sharps injury, although other contaminated sharp instruments may also cause injuries. All healthcare

workers must be educated about the potential exposure that can occur during their duties and should have appropriate vaccinations. The risk of hepatitis B, hepatitis C and HIV infection should be assessed and appropriate immunisation or chemoprophylactic steps taken after an incident. Immediate treatment of such injuries should encourage washing thoroughly with running water and an antiseptic solution. Consult the infection control team for further advice and follow their basic protocol. An incident-reporting system should be in place. This should not be seen as punitive; active support by managers should encourage prompt and accurate reporting.

Exposure to human immunodeficiency virus (HIV)

The route of transmission of HIV is from person to person via sexual contact, sharing of needles contaminated with HIV, infusions that are contaminated with HIV, or transplantation of organs or tissues that are infected with HIV. The risk of a healthcare worker acquiring HIV after a needlestick or other 'sharps' injury is less than 0.5%. Risk reduction must be undertaken for all bloodborne pathogens, including adherence to standard precautions using personal protective equipment, appropriate safety devices, and a needle disposal system to limit sharps exposure. Training for healthcare workers in safe sharps practice should be ongoing.

Information on preventive measures must be provided to all staff who may potentially be exposed to blood and blood products. Policies that are in line with the local and national guidelines must include screening of patients, disposal of sharps and wastes, use of protective clothing, management of inoculation accidents, and sterilisation and disinfection procedures. Hospital policy must include measures to obtain serological testing of source patients promptly where necessary, usually with the patient's informed consent. Post-exposure prophylaxis should be started as per local or national guidelines.

A suggested strategy for use when a healthcare worker has been potentially exposed to HIV

- 1 Discuss with the patient (or in the case of a child, the family) what has happened, and ask whether the patient's HIV status is known. If it is not, discuss the possibility of testing, if the injury occurred during normal working hours. Remember that anyone undergoing an HIV test has the right to counselling. If the injury occurred out of hours, or the family decline testing, proceed to Step 3.
- 2 If the patient has negative HIV ELISA and is over 18 months of age, infection is extremely unlikely. If they are under 18 months of age, a positive ELISA may reflect maternal antibodies. However, any positive test result should lead to Step 3. If the result is negative, the healthcare worker is not at risk of HIV infection. However, further testing of both the child and the healthcare worker for hepatitis B and C may be warranted.
- 3 Arrange a baseline HIV ELISA for the healthcare worker after appropriate counselling. If the result is positive, they will need to discuss further treatment with their own doctor.
- 4 If the healthcare worker's baseline serology is negative and the patient is HIV positive, antiretroviral prophylaxis should be started urgently. Current recommendations advise 1 month of treatment. The healthcare worker will need a repeat ELISA after 3 to 6 months to check their status.

Exposure to hepatitis B virus

The route of transmission of hepatitis B virus is through body fluids such as blood, saliva, cerebrospinal fluid, peritoneal, pleural, pericardial and synovial fluid, amniotic fluid, semen, vaginal secretions and any other body fluid containing blood, and also through blood products. It is important to follow standard precautions, but immunisation is the best way of preventing transmission to healthcare staff. All healthcare workers who are in contact with patients or body fluids must be vaccinated against hepatitis B.

Staff who are infected with bloodborne pathogens may transmit these infections to patients, and therefore require careful evaluation with regard to their duties. This status should not be used to discriminate against them.

Exposure to hepatitis C virus

The route of infection is mainly parenteral. Sexual transmission does occur but is far less frequent. No post-exposure therapy is available for hepatitis C, but seroconversion (if any) must be documented. As for hepatitis B viral infection, the source person must be tested for hepatitis C virus infection. For any occupational exposure to bloodborne pathogens, counselling and appropriate clinical and serological follow-up must be provided.

Confidentiality

There should be a policy to promote and ensure that confidentiality of patient and health worker information is protected and is also reflected in daily practice.

Systems need to be in place to ensure that patient records and the personal files of employed staff are kept confidential.

Training about the principles and practice of confidentiality should be mandatory for all workers in the health facility.

Other services for patients and their relatives

Health information should be available. Health promotion information should be visible and available in all parts of the health facility in commonly understood languages and in pictorial form (see the Maternal and Child Healthcare Initiative (MCHI) manual. A manual for health workers based on a medical ethics approach to healthcare.

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Toilets should be available for visitors, and these and all facilities and areas of the health facility should be accessible to people with a disability.

If possible, telephones should also be available for visitors.

Ideally there should be written policies concerning the rights and responsibilities of patients, resident parents/carers and visitors. These policies should be prominently displayed around the hospital and should include issues such as the prevention of smoking, the effects of alcohol, violence (verbal and physical) and weapons in the hospital.

Smoking is particularly important in relation to children's health, but in the case of stressed parents it may be inappropriate to ban it altogether. Instead, it should be limited to defined areas.

Family-centred care

The role and rights of families in caring for patients alongside and in partnership with professional staff is vital but must be handled extremely carefully. Families must not be exploited, but equally in resource-limited countries hospital care would not be possible without their assistance. Families must be made welcome and good understanding of roles and effective communication are of paramount importance (see also Section 8 and the MCHI manual

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Play, sensory stimulation and support for children's wards

The importance of play and developmental support in children's well-being cannot be over-emphasised particularly for children that need health care. A friendly and stimulating

environment helps the child to understand and cope with their hospitalisation and to get better far more quickly (as advocated in the World Health Organization recommendations for the recovery management of children with malnutrition). It also helps to support the parents and can provide them with additional skills that they can continue to use at home once the patient has been discharged. Many parents cannot afford to stay at the hospital for long periods because of pressures to return to their home village, where they are pivotal to the daily routine, farming, or looking after other children. Mothers can be supported by passing on the knowledge of play as taught by a play worker. Giving a sick child access to play and information facilities in hospital also helps to reduce loneliness and fear.

Some well-resourced countries have training programmes and qualifications for play specialists. However, much can be achieved by recognizing the importance of play and information and integrating this into the regular practice of health workers. It is effective, as both an adjunct and core part of treatment, in the hands of a health worker or a dedicated play worker, using resources made of local and low-cost materials.

All health workers for children, including any dedicated play workers, need to have good communication and empathy skills with children and families. They also need to have a good understanding of child development and the needs of children in hospital, especially children who are alone and/or who have disabilities or other additional needs.

In addition, health workers and play workers need to be trained in how to deal with the sensory needs of children in specific situations, such as the comatose child, ie that these children can hear and have feelings when touched, and how to encourage the parents to talk and play with the child.

Conclusion

The provision, organisation and financing of services, facilities and functions, and the management of the human resources, are as important as those needed to provide the clinical and clinical support services. A sound hospital infrastructure and management are of paramount importance for the provision of good-quality care.

Further information on other work-related issues concerning healthcare staff can be found in International Maternal & Child Health Care : A practical manual for hospitals worldwide (MCAI 2014) https://12dec1c3-e3f4-e784-7f3a-6e7903610962.filesusr.com/ugd/dd2ba4_c5284b1b150f4d70a72fe1cd710d530d.pdf
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Prevention of hospital infection

Introduction

Nosocomial or hospital-acquired infection is a major problem not only in terms of cost but also, more importantly, because it increases morbidity and mortality in patients. Such infections may affect up to 10% of all patients. Nosocomial infection requires a source of microorganisms and a chain of transmission. It is essential that all healthcare staff examine their own practice to ensure that they are not part of this chain of transmission.

The combination of the use of powerful antibiotics and poor hygiene also predisposes to the development of antibiotic-resistant microorganisms, which are difficult both to eradicate from the environment and to treat.

Pregnant women are at high risk of infection. However, not all infections are related to their disease process, but rather they may be caused by failure of both hospital management and individual healthcare workers to introduce and adhere to strict infection control policies.

Requirements and procedures

The following measures are essential to minimise the risks of infection and cross-infection.

A clean and adequate water supply

Just as water and sanitation are of central importance in the prevention of cross-infection in emergency refugee camps, they are also of vital importance in hospitals, particularly where there are vulnerable patients. Running water (both hot and cold) is preferable. Hot water should be stored at 65°C, distributed at 60°C, and the temperature then reduced to 43°C to be used from the taps. This process helps to ensure that water-borne infections such as Legionnaire's disease are not passed on to staff or patients and reduces the risk of burns for staff.

Accessible sinks in all areas

These should preferably be equipped with elbow-operated taps, and there should be adequate washing and toilet facilities for staff and patients.

Effective cleaning policies

Sufficient staff should be employed over the 24-hour period to keep all areas of the hospital and grounds clean at all times. Written cleaning policies and training for cleaners should be in place, and a supply of appropriate cleaning materials and disinfectants readily available.

Toilets, bathrooms, and other facilities needed for personal hygiene of health workers, patients and visitors, and for cleaning equipment, are of particular importance, and these areas should always be kept scrupulously clean.

Clean hospital grounds, pathways and entrances reduce the risk of dirt being transmitted to the ward and other patient areas by staff, relatives, and other visitors.

Stray animals must be kept away from the hospital premises. Vermin must be kept away from the hospital buildings. Professional advice must be sought as soon as any signs of vermin are found.

Areas such as operating theatres, as well as certain items of equipment, must always be aseptic.

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There should be a central sterilizing service. If this is not possible there should be suitable sterilisers and a supply of appropriate disinfectants at a range of dilutions. Wherever possible the manufacturers' instructions for cleaning specific items of equipment should be followed.

Staff appointed as cleaners should be given adequate status and salaries to reflect the importance of the work they are doing, as well as training in how to keep the hospital clean and why this is so important.

Effective services for disposal of human and other waste

An incinerator that operates 24 hours a day is essential for the safe disposal of clinical waste. A system for handling and disposal of all clinical and non-clinical waste, including 'sharps', is also needed.

Written policies and procedures are necessary for the different types of waste disposal, and initial training and updates should be mandatory for all staff.

Human and other waste should be disposed of and collected separately.

Foot-operated bins are preferable as they reduce risks of injury and cross-contamination, and frequent waste rubbish collections are essential.

Laundry service

Bedding and other items must be frequently washed. Therefore, the hospital must have a staffed laundry service, ideally with enough industrial washing machines and drying facilities. Where hand washing is the only option, staff should wear protective clothing and high-quality thick gloves. Clean bedding, towels and nappies must be available. A small supply of nightwear and other clothing may be needed on the wards for families who do not have a change of clothes with them.

Strict hand-washing policies

Viruses and bacteria can commonly survive on the hands for up to 3 hours. **Correct hand-washing technique for all staff, visitors and patients is the most important factor in the prevention of cross-infection.** It is easily taught, and frequently an improvement in practice is demonstrated in the short term. However, when examined over a longer period, old habits and short cuts 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 (see Figure 1.1). Where it is impossible to provide liquid soap and paper towels, some simple innovative solutions have been found to enable good practice. Bar soap suspended in a net bag over the sink area and individual cloth towels for each patient, changed every 24 hours or at the discharge of the patient and kept within their bed space, can be effective. Added emollient protects the hands. Antiseptics can be added to liquid soap to improve antimicrobial activity, and chlorhexidine is a cheap and effective antiseptic that is widely available throughout the world. 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, a 70% alcohol gel is useful. This is a new but expensive product that has a significant part to play in the prevention of introduction of cross-infection in high-risk areas. When rubbed on and allowed to dry, it is effective in disinfecting the hands. After initial conventional hand washing it can be used between each patient contact, but further hand washing is still recommended after every five to six rubs.

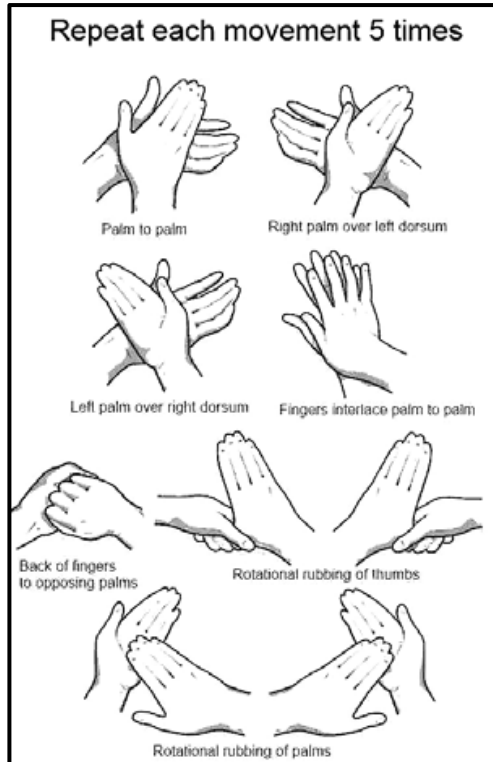


Figure1.1 Effective hand washing

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

All staff have a personal responsibility for hygiene, but every ward should also identify an individual (ideally a nurse with the support of a microbiologist, if available) to be responsible for the education of all staff in techniques to prevent the spread of infection, particularly effective hand washing and drying. Education and behaviour change will need to be continuous and ongoing, as even in the best health facilities training is only effective for a short period of time and frequent refreshers are needed. The identified staff member will need support from the hospital management to reinforce that all grades and members of staff comply with good handwashing and infection prevention practices (especially senior doctors and nurses who should act as role models). The identified staff member, no matter how junior they are, should have an agreed mandate to cross traditional hierarchies and be recognised as the expert in their unit, and anyone who is asked to carry out hand washing should immediately comply with this request.

Disposal of body fluids

Each ward or unit must have an area set aside for this purpose. It, and all the equipment that it contains, must be kept scrupulously clean and body fluids disposed of quickly, with any spillage removed immediately. If there is likely to be a risk 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 family members who will be assisting with the toileting of patients.

Care must be taken with sharp objects such as hypodermic needles, to protect the patient, their family, other unit visitors and staff.

Cleaning, disinfection and sterilisation of equipment and furniture

The manufacturer's instructions for individual items of equipment must always be followed. These will usually clearly state which items need to be sterilised and where disinfection will be sufficient. They will also indicate appropriate dilutions for disinfectants. All equipment should be cleaned before being sterilised or disinfected.

a) Sterilisation

This is the complete elimination and destruction of all forms of microbial life. This is frequently achieved by steam under pressure, dry heat, gas or liquid chemicals. Such a sterilisation system must be available in every ward where invasive procedures are undertaken, and such systems are also required for instruments and towels used in the operating theatre.

b) Disinfection

This is a process that eliminates most 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 a diluted form (usually 0.1% solution) for domestic use are contained in household cleaners available worldwide. These household cleaners can be used in the hospital environment for general cleaning, but stronger solutions (0.5% chlorine solution) must also be available, particularly for the disposal of body fluids, for initial cleaning of bloodstained instruments, and following outbreaks of notifiable infections. A 0.5–1% solution is recommended for the treatment of blood and body fluid spills, and 0.05–0.1% solution can be used for all surfaces. Hypochlorites are available as tablets, which makes the process of dilution easier.

How to prepare high-level disinfectant solutions

The best compound for the preparation of chlorine solutions for disinfection is household bleach (also known by other names such as Chlorox[®] and Eau de Javel). Household bleach is a solution of sodium hypochlorite which generally contains 5% (50 g/litre or 50 000 ppm) available chlorine.

Thick bleach solutions should never be used for disinfection purposes (other than in toilet bowls), as they contain potentially poisonous additives.

When preparing chlorine solutions for use, the following points should be noted:

1. Chlorine solutions gradually lose strength, and freshly diluted solutions must therefore be prepared daily.
2. Clear water should be used because organic matter destroys chlorine.
3. A 1:10 bleach solution (0.5%) is caustic. Avoid direct contact with the skin and eyes.
4. Bleach solutions give off chlorine gas, so must be prepared in a well-ventilated area.
5. Use plastic containers for mixing and storing bleach solutions, as metal containers are corroded rapidly and also affect the bleach.

Two different dilutions of bleach are used for disinfection. 1:10 bleach solution (containing 0.5% chlorine) is a strong disinfectant, which is used to disinfect the following:

- excreta
- bodies
- spills of blood or body fluids
- medical equipment (e.g. delivery sets, kidney dishes, suture instruments, catheters, speculum).

To prepare a 1:10 bleach solution, add one volume (e.g. 1 litre) of household bleach to nine volumes (e.g. 9 litres) of clean water.

Always wear gloves.

Immediately after delivery or examination, clean the instruments below the level of solution in the plastic bucket using a brush. Leave for 10 minutes and then place them in soapy water, wash with a brush, and flush every catheter with a 10–20 mL syringe. Next rinse with clean water and air dry, and then sterilise or boil for 20–30 minutes. Store dry in a metal bowl.

Change the solution after 24 hours or when it becomes bloodstained.

Label buckets with tape indicating the date and time when the solution was prepared and when it needs to be changed.

The above 0.5% solution can also be used to prepare 1:100 bleach solution (containing 0.05% chlorine)

This is used for the following:

- disinfecting surfaces
- disinfecting bedding
- disinfecting reusable protective clothing before it is laundered
- rinsing gloves between contact with different patients (if new gloves are not available)
- rinsing gloves, aprons and boots before leaving a patient's room
- disinfecting contaminated waste before disposal.

To prepare 1:100 bleach solution, add one volume (e.g. 1 litre) of 1:10 (0.5%) bleach solution to nine volumes (e.g. 9 litres) of clean water.

Note that 1:100 bleach solution can also be prepared directly from household bleach by adding 1 volume of household bleach to 99 volumes of clean water (e.g. 100 mL of bleach to 9.9 litres of clean water) but making it up from 1:10 bleach solution is easier.

c) Cleaning

This is often the most neglected of the three processes, and it must precede sterilisation and disinfection. When undertaken using a disinfectant detergent, cleaning alone will effectively reduce the number of microorganisms and make safe those items that come into contact with the intact skin (e.g. blood pressure cuffs, bed rails, intravenous poles).

Isolation of patients with specific infections

For isolation procedures to be effective they need to be instituted early. Different isolation techniques will be needed, and the use of gowns, gloves and masks will be necessary if the infection is very contagious and/or very serious. In some cases, nursing the patient in a cubicle or single room until medical tests are complete is all that is necessary. When there is a need for Personal Protective Equipment (PPE); gowns, gloves, face shields, goggles, and masks, these will require frequent changing or washing to ensure their efficacy and must be used by everyone who comes into contact with the patient, including medical staff and carers. Ideally, they should be used only once and then 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.

Infection control measures following the death of a patient

When a patient dies, the amount of time that family members can spend with them 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, in particular the belief that something of the individual survives death, either to be reborn through reincarnation or to fulfil their spiritual destiny in the afterlife. It is important that the correct funerary procedures, if any, are followed to ensure that the bereaved are not distressed by any omission which they consider important.

All societies, whether religious or not, have to deal with the problem of the death of their patients 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), and 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 perform post-mortem examinations to establish an exact cause of death. Effective hand-washing procedures remain of paramount importance.

In countries where the climate is characterised by extremes of temperature, refrigeration of 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, staff working in the mortuary will need to be provided with separate clothing for use in that department. The use of two pairs of gloves, or thick rubber gloves and protective clothing, will be necessary for the post-mortem examination if there is suspected infection of the body with life-threatening bacteria or viruses.

The mortuary department will need to have facilities for families to see and spend time with their dead relative, and a separate comfortable area where documentation can be completed and any necessary interviews with local government officials can be conducted. The mortuary department not only provides facilities for post-mortem examination, but also, in large centres, it can be part of the government facilities for forensic post-mortems, which may provide additional resources for the 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 that involves families in the consent procedures for all examinations performed after the patient's death.

Conclusion regarding infection control

Every person that works in a hospital or health facility has a role to play in the prevention of hospital-acquired infections. The greatest responsibility lies with the healthcare professionals, particularly midwives, neonatal and obstetric clinicians, nurses, and doctors, who in the hospital setting are in contact with patients and their families 24 hours a day; and because of this they have the highest risks of causing cross-infection. However, they can also be the catalysts for change, and improve the education of other hospital staff and families.

Section 2 identifying and managing children who have been abused

The basic principles of the investigation of child maltreatment are that:

- The welfare of the child is paramount
- Multi-agency/multi-sectored collaboration is needed
- Agencies must work together within the legal framework of the country (where this is in place).

In practice, most low resource settings have minimal services available to identify and protect children from neglect and abuse.

Child Rights

Article 19 of the United Nations Convention on the Rights of the Child states that children (people less than 18 years of age) have a right to be protected from being hurt and maltreated physically and mentally. It goes on to state that governments should ensure that children are properly cared for and should protect them from violence, abuse and neglect by their parents or anyone who looks after them.

Child abuse

Child abuse results in actual or potential harm to the child's health, survival, development, or dignity in the context of a relationship of responsibility, trust or power with the abuser.

Healthcare workers have a major responsibility in contributing to the prevention and recognition of childhood ill treatment. This poses challenges for healthcare workers when they work with children and families from different belief systems and cultural backgrounds. They may find that they must care for street children, child soldiers, and children separated from their parents by civil strife and unrest and find themselves making difficult judgements about how a child can be best protected when they have few if any points of reference, and only limited contact with other agencies.

Child maltreatment or abuse involves acts of omission and commission which result in harm to a child. It can occur in the family, in the community or in institutions (e.g., schools, hospitals, churches, mosques, temples, clubs, orphanages or other institutions). It encompasses:

- Exploitation through trafficking for sexual or other forms of slavery
- Exploitation through enforced prostitution
- Physical abuse
- Emotional abuse
- Neglect
- Sexual abuse
- Fabricated or induced illness (FII)
- Conscripted as child soldiers.

Presentation of a child to hospital which suggests possible ill treatment or abuse

- Delay in seeking medical help for an injury or serious clinical symptoms or signs (e.g. bleeding).
- A history that is vague or rehearsed, with inconsistencies and significant changes on re-telling or following questioning.
- No explanation of the cause of the injury.
- Repeated attendance at healthcare facilities (this may suggest fabricated or induced illness, FII; see below).
- Parents or carers being evasive or hostile.
- A history of injury that is inconsistent with the child's age and/or developmental skills.
- A 'collusion of silence', or one parent implicating the other.

- Accusations that the child is a witch, or that witchcraft has been perpetrated by others.
- The presence of other injuries, or a previous history of unusual injury.
- Child appearing sad, withdrawn, anxious or frightened ('frozen watchfulness'), or over-compliant.
- Child may indicate the abuser.

Consideration needs to be given to children with disabilities who may be unable to communicate about their ill treatment, and where their presentation may be misattributed to their disability.

Children who suffer abuse are often threatened by being told that 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.

Physical Abuse/III Treatment (Non- Accidental Injury)

Physical abuse can be defined as any act resulting in a non-accidental physical injury, including, not only intentional assault, but also the results of excessive or violent punishment. Physical abuse occurs when a person deliberately injures a child or young person.

Around 25 - 50% of all children report being physically abused, according to the World Health Organization (WHO). Physical abuse **usually coexists with emotional abuse, and sometimes accompanies sexual abuse**. However, in some settings, physical chastisement (especially of older children) continues to be considered part of 'good parenting' and important for instilling discipline in a community's children. In many countries, laws define which childhood punishments are considered excessive or abusive.

Some classifications define physical abuse as an injury that produces a mark. However, this does not take into consideration the emotional effects of physical abuse. The number and size of the bruises are helpful in distinguishing between mild and serious abuse. Any assault on a child is unacceptable and constitutes child abuse. A small bruise in a baby may predict future serious or fatal abuse.

Typical injuries include the following:

- Lash marks, especially on the trunk, legs, and hands
- Bruises, especially on the face, scalp, and on or behind the ears and on the buttocks
- Certain patterns of bruises, such as fingertip marks or bruises in the shape of the implement used, or multiple bruises of different ages. However, current scientific evidence concludes that we cannot accurately date a bruise from clinical assessment or from a photograph.
- Burns, including branding and scalds, especially when these are bilateral and/or symmetrical E.g., buttocks or face held against a hot object such as a radiator, or both hands or feet or buttocks scalded because of the child being immersed deliberately in hot water).
A pattern suggesting a cigarette burn or burns is also important, but be careful about the possibility of impetigo, which may mimic such burns (impetigo heals quickly and without scarring with antibiotic treatment, topical if a small area and systemic if widespread, whereas burns heal more slowly and may scar)
- Injuries to the mouth (especially a torn frenum)
- Bleeding from the mouth or nose in an infant (indicating the possibility of intentional suffocation)
- Adult bite marks

- Bony injuries, especially in non-ambulant children; skull fractures, spiral fractures of the humerus, rib fractures in young children and multiple fractures of different ages, epiphyseal separation at the end of long bones, periosteal separation and haematomas
- Inflicted head injury (especially in infants) involving tearing of the superficial veins over the brain and retina, causing subdural and retinal haemorrhages and often major generalised brain injury with features suggesting cerebral hypoxia or ischaemia. This condition can be fatal or may cause permanent physical and mental impairment and visual loss.
- Failure to thrive due to neglect (category 2) or deliberate starvation (category 3)
- Induced illness, including suffocation or poisoning.

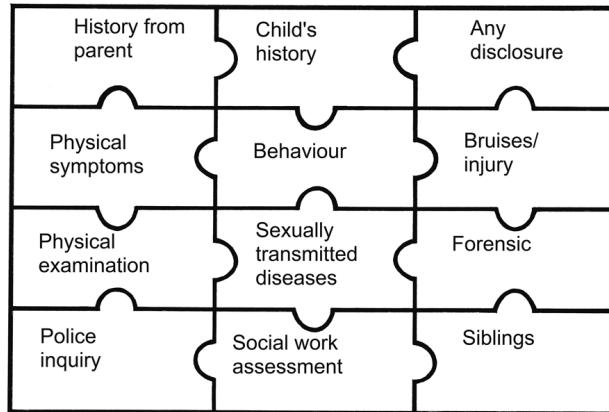


FIGURE 2.1 The Jigsaw of Child Abuse. (From Hobbs CJ and Wynne JM (1990) The sexually abused battered child. Archives of Disease in Childhood 65: 423–427.)

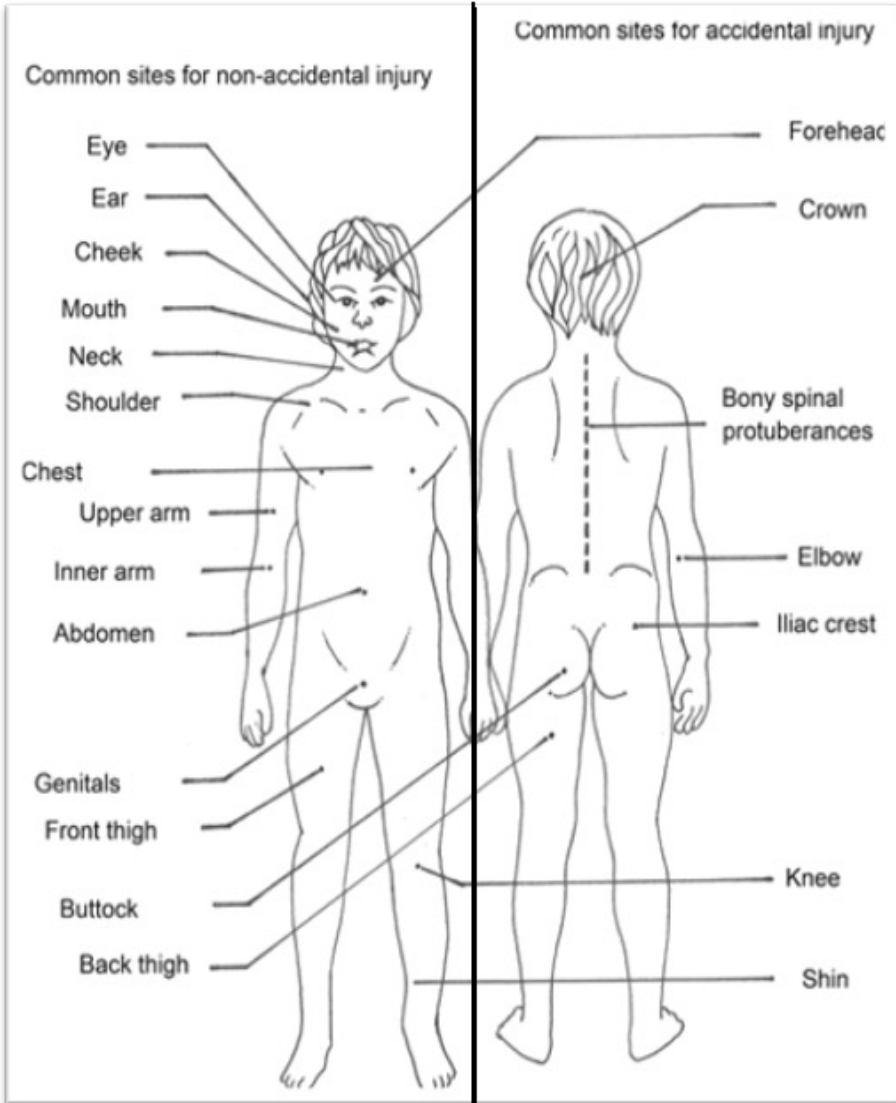


FIGURE 2.2 Common sites for non-accidental (abusive) injuries LEFT COLUMN and accidental injuries RIGHT COLUMN

Particular care needs to be taken when interpreting skin marks in settings where traditional practitioners use cupping, coining, scarification or tattooing treatments.

Children are more likely to be killed or to experience violence in their own home than outside it. The triad of violence against a partner (usually the female partner), mental ill health and substance misuse (drugs and/or alcohol) are identified as common features of families where significant harm to children have occurred. There is a strong correlation between domestic violence and child abuse.

There is also a strong relationship between animal abuse and child abuse.

Emotional and Psychological Abuse

This can occur as isolated incidents, as well as within a pattern of failure over time on the part of a parent or caregiver to provide a developmentally appropriate and supportive environment. This has a high probability of damaging the child's physical, mental, and emotional health, or their physical, mental, spiritual, moral or social development.

This category should be used where it is the main or only apparent form of abuse.

Emotional and psychological abuse includes:

- The restriction of movement
- Patterns of belittling
- Blaming
- Threatening
- Frightening
- Name-calling
- Scapegoating
- Persistent criticism
- Discriminating against the child compared with their siblings
- Ridiculing them, and other non-physical forms of rejection or hostile treatment.

This type of abuse can be difficult to recognise. Concerns are frequently raised by a child's extended family, neighbours, or nursery or school staff. All abuse involves some emotional abuse, but emotional abuse may exist independently. Emotional abuse occurs when, for example, there is:

- Emotional unavailability of the parent or carer (e.g., when they are preoccupied with their own needs because of mental health problems, substance abuse problems, or work commitments)
- Failure to allow the child to interact normally socially with others.

The consequences of emotional abuse vary with age and with its duration, and may include the following:

- Impaired physical development:
 - These children often fail to reach their optimum potential in terms of growth; this improves when the child is placed in a more nurturing environment.
- Impaired cognitive development
 - Including speech and language delay, poor concentration, and academic underachievement.
- Behavioural abnormalities
 - Such as anxious attachment
 - Lack of social responsiveness
 - Expressionless face
 - Fear of speaking
 - Eagerness to please
 - Attention seeking
 - Overactivity or 'hyperactivity'
 - No wariness with 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

Impaired psychological development, especially with regard to speech and language: aggression, emotional unresponsiveness, emotional instability, impaired social development, low self-esteem, dependency and separation anxiety, serious social difficulties, underachievement, negative self-evaluation, poor concentration, and poor academic performance or school attendance can also be present.

Emotional maltreatment and abuse have been described in association with three psychiatric disorders of childhood:

1. Depression
2. Reactive attachment disorder of infancy
3. Multiple personality disorder.

In general, these children become sad, dejected and withdrawn.

Medical problems include the following:

- Failure to thrive
- Recurrent and severe nappy rash
- Generally unkempt appearance, with poor hygiene
- Recurrent minor infections.

Neglect

Neglect includes both isolated incidents and a pattern of failure over time on the part of a parent or other family member to provide for the development and well-being of the child (where the parent can do so) in one or more of the following areas.

Neglect is persistent failure to meet a child's essential needs by inattention or omitting basic parenting tasks and responsibilities in all aspects of their needs (health, hygiene, clothing, education, and social, emotional, mental, spiritual, and moral needs). It also includes failure to provide appropriate nutrition, shelter, and safe living conditions.

Examples include lack of supervision, with failure to protect the child from dangers (e.g., cold temperatures, sunburn, drowning) due to poor supervision and attention to safety in the home (e.g., not providing stair gates or locks on windows), failure to thrive, and failure to meet the child's emotional needs for love, affection, and stimulation.

It results in serious impairment of the child's physical health, psychological well-being, and development. It may coexist with other forms of ill treatment.

The parents' or carers' own problems (e.g., learning difficulties, mental and physical health problems, poverty, inappropriate housing) can all contribute to this situation. In unstable settings, such as armed conflict, there may be significant security issues that contribute to their inability to provide a safe environment for their children.

Child Sexual Abuse

This is the involvement of children in sexual activity to which they cannot comprehend and therefore cannot consent. Another definition could be any activity in which an adult or older child uses a younger child in a sexual way.

In addition to direct sexual contact between adult and child (including intra-crural, oral, vaginal or anal sex and the masturbation of an adult), it includes the use of penetrative instrumentation, the production of pornographic imagery of children, exposing a child to indecent acts or pornography, and other voyeuristic practices. Very young children may also be trafficked for use in the sex industry, and older children may be groomed for prostitution.

Sexual abuse is a serious global problem that transcends economic or social barriers. Poverty, emotional deprivation and lack of education often mean that young people are powerless to avoid being trapped both in sexually abusive situations and in domestic violence. However, it is universally true that sexual abuse is most often suffered at the hands of a neighbour, family friend or a trusted person, including a parent. A significant power differential usually exists between victim and abuser. This fact is important when examining situations of sexual behaviour between children themselves.

In legal terms there are two types:

- When a stranger or someone the child knows abuses the child
- Incest:
 - When a relative by blood or by law abuses the child.

Some facts about Child Sexual Abuse

- Child sexual abuse is a problem in all socio-economic classes.
- Boys are also sexually abused.
- Abusers are usually people whom the child knows.
- It is never the child's fault if they are sexually abused.
- Women can also be abusers.
- Abusers cannot be recognised by their physical appearance.
- Sharing the experience of abuse with someone who is supportive and understanding helps the victim deal with it.
- Young children and even infants can be abused in this way.
- A child who is abused at a very young age may also be affected by it, even if they forget the episode.
- CSA is rarely accompanied by violence and physical force.
- The main motive behind sexual abuse is not sexual frustration but to gain 'power' and 'control'.
- some children who are being sexually abused, may not know this is wrong but see this as the norm.

Worldwide, 40 - 47% of sexual assaults are reported to involve girls under the age of 15 years. In Tanzania, almost 30% of adolescents undergoing abortions had been impregnated by men aged 45 years or older. In India, a study in 2007 reported that over 50% of children had been subjected to sexual abuse, and in 21% of cases this was of a severe nature. That study also reported that children living on the street or in institutional care were most vulnerable to sexual abuse.

Rape and impregnation as a weapon of war is now well recognised as a distressing and complex issue arising in countries where conflict is endemic.

Physical Symptoms and behavioural effects of Child Sexual Abuse

Sexual abuse may go undetected for years because the symptoms are vague and easily attributed to other factors, such as the arrival of a new sibling in the home or family breakdown. Abuse may come to light abruptly when a child says something inappropriate or makes a direct allegation (disclosure) when in a situation in which they feel safe, sometimes to a friend or teacher at school.

Physical symptoms are unusual and vague. Unexplained episodic dysuria and frequency or genital soreness are most often recalled in retrospect. Sexually transmitted disease and pregnancy may make the diagnosis irrefutable in some cases. Obvious pain, bleeding and signs of acute physical trauma to the genital and anal area are rarely presenting features, and usually accompany violent and reckless assaults, often by a stranger.

Other symptoms include difficulty in walking, gastrointestinal disturbances (including nausea), eating disorders, abdominal pain, and in the genital area pain, itching, visible injury, discharge, infection or difficulty urinating.

Bruises, cuts, and other injuries on any part of the body for which the cause is not clear and the child cannot give a full explanation may be present. Sexually transmitted diseases may occur at the time of abuse or lie dormant for months or even years, only to flare up in adolescence or adulthood.

There may be a noticeable fear of a particular person or place, sudden bedwetting or soiling, preoccupation with sexual acts, and a change of language or re-enacting their experiences with other children. Nightmares, withdrawn behaviour, changes in appetite and a decline in school achievement are not uncommon.

Other behavioural effects include the following:

- Persistent and inappropriate sexual play with peers, toys, animals, or self
- Sexual themes and fears in the child's artwork, stories, and play
- Sexual understanding or behaviour beyond that expected for the child's developmental stage
- Self-harm or hurting others, including fire setting and cruelty to animals
- Fear of being alone, of going home, or of places and people
- Running away from home
- Drug and/or alcohol use
- Adolescent prostitution or sexual promiscuity
- Suicidal feelings and attempts.

Parents should be concerned if a child appears to have unexplained expensive objects or financial resources. The grooming process often involves making a child feel cherished and creating dependency by offering them quality time, treats and gifts; in older children, the grooming process often includes encouragement to indulge in and provision of cigarettes and alcohol.

These effects may be symptoms of something other than Child Sexual Abuse, but the possibility of it should always be explored.

Most of the symptoms relate to long-term psychological damage. Young adults find it difficult to make intimate relationships and to trust others. Alcohol and drug dependency, anxiety disorders, self-harming and suicidal ideation may be linked to sexual abuse, especially at the hands of a trusted adult such as a parent. The trauma genesis of sexual abuse has been extensively described by Finklehor. He proposed four dynamics: traumatic sexualisation, powerlessness, betrayal, and stigmatisation.

Immediate Action when Ill Treatment or Abuse is suspected

A detailed history and full medical examination are required (including inspection of the genitals). Where possible, obtain the consent of the parent or carer and the child to carry out the medical examination.

If consent is withheld, work urgently within the legal framework of the country concerned to examine and protect the child in conjunction with police, social services or civil society organisations.

Ensure that the child (if they are old enough and able to speak) is given the opportunity away from their parent or carer to say how they were hurt (disclosure). At the same time, avoid asking leading questions to the child or parent as this might interfere with any police investigation.

When Responding to Disclosure by a child that possible abuse has occurred:

- Remain calm. An over-reaction will make the child feel even more frightened and ashamed.
- Believe the child.
- Listen in a non-judgmental way.
- Use the child's language.
- Tell the child that you are glad they have told you what happened.
- Reassure the child that they did nothing wrong.
- Explain to the child that abuse is an unfair thing that happens to children, without condemning the offender.
- Determine the immediate need for safety.
- Don't make promises that you cannot keep.
- Let the child know what you will do.
- Set in motion the process of getting help for the child.
- Take care of yourself.

Do's and Don'ts of Disclosure

Do use phrases like this:

- 'I believe you';
- 'You did the right thing by telling someone';
- 'I'm so sorry this has happened to you';
- 'It's not your fault';
- 'I will try to help you so that it won't happen again.'

Don't use phrases like this:

- 'Don't say such things!';
- 'Are you sure it happened/is happening?';
- 'Are you telling the truth?';
- 'Why are you telling me?';
- 'Why didn't you stop it?';
- 'What did you do to make this happen?'

- Consider whether other children in the family may need to be examined and protected.
- Record a full history as it is spoken and include an evaluation of the child–parent interaction.
- Carry out a careful examination in a well-lit room.
- Record the details of the history and examination legibly and contemporaneously in the child's medical notes. A form of the type available in the Appendix can be helpful.
- Include details of the child's demeanour and presentation, and their height and weight plotted on a centile chart. Ensure that an examination of the child's mouth, nose, ears and neck is undertaken, and complete a full systemic medical examination.
- Document any injuries on body diagrams (see Figures 2.2 and Appendix 2.3 to 2.7).
- Consider photo documentation (with the child's consent, if possible) of injuries, and ensure that the images can be stored safely and confidentially.
- Check whether the family is known to the police and/ or social services.
- Consider whether any additional medical investigations need to be carried out (e.g. X-rays for bony injuries, a skeletal survey in children under 3 years of age, clotting studies) (see Table 2.1).
- Admit the 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 to talk further with the child; alone if appropriate to do so.

- If the parents refuse to allow the 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 (if a referral has not already been made) or the police.

A thorough medical examination to be undertaken when all kinds of possible abuse are suspected and should include the following:

- Observation of the child's demeanour
- Height, weight and head circumference (in a preschool child) plotted on a centile chart
- Examination of the mouth, nose, ears, neck and genitals
- Inspection of skin surface for bruises, marks and cuts
- Examination of the eyes for retinal haemorrhages (pupil dilatation may be needed) (see Section 20).
- Systemic examination
- An assessment of the child's developmental age.

TABLE 2.1 Investigations exploring the differential diagnosis of signs of injury that may be due to child abuse

Injury	Differential diagnosis	Investigation
Bruising	<ul style="list-style-type: none"> • Coagulation disorder • Idiopathic thrombocytopaenic purpura/Henoch–Schönlein disease • Haemorrhagic disease of the newborn • Septicaemia • Connective tissue disorders • Birth marks • Dyes / Tattoos • Drug reactions • Self-inflicted injuries • Traditional treatments 	<ul style="list-style-type: none"> • Full blood count, • Blood film, • Coagulation studies, • Chest X-ray • Consider skeletal X-ray survey in children under 3 years of age • Opinion of expert in skin diseases (if available)
Bites	<ul style="list-style-type: none"> • Animal or human • Adult or child 	<ul style="list-style-type: none"> • DNA skin swab (if available) • Photography • Forensic dental assessment (if available)
Fractures	<ul style="list-style-type: none"> • Accidental injury • Birth injury • Infection • Malignancy • Osteogenesis imperfecta • Osteopaenia • Nutritional deficiencies (including rickets) 	<ul style="list-style-type: none"> • Chest X-ray • X-ray skeletal survey in children under 3 years of age Bone scan (if available) • Radiology advice • Blood calcium, phosphate, alkaline phosphatase if available • vitamin C and D levels (if available) • CT scan for head injury (if available)

Injury	Differential diagnosis	Investigation
Scalds & Burns	<ul style="list-style-type: none"> • Other skin pathologies (e.g. staphylococcal and streptococcal infection) • Drug reactions • Allergic reactions to plants (e.g. euphorbias) 	<ul style="list-style-type: none"> • Skin swabs • Dermatology opinion (if available)
Head injury/ Unexplained fits or coma	<ul style="list-style-type: none"> • Coagulation disorder • Epilepsy or febrile convulsion • Cerebral malaria • Meningitis • Poisoning 	<ul style="list-style-type: none"> • Retinal examination after pupil dilatation (see Section 20) • CT scan (if available) • Covert audio/video surveillance
Intentional suffocation Intentional poisoning	Naturally occurring near death episodes	<ul style="list-style-type: none"> • Covert audio and covert video surveillance • Forensic attention to collection of specimens for analysis

Clinical Examination and Physical Signs where child sexual abuse (CSA) is suspected

Clinical assessment of the sexually abused child should ideally be conducted by trained and experienced professionals, and preferably a specialist forensically trained doctor or nurse. The environs of the clinical space should be child friendly and the examination unhurried. All examinations should consider the global health needs of the child first; the needs of law enforcement should not be paramount.

Careful history taking is the first step, and if prosecution is being sought, the history should be obtained without risk of contamination, either directly from the child before a recording witness or in the child’s absence from the adult who knows first-hand what the child has said. The history should be elicited with free recall and by posing indirect questions (e.g., ‘Tell me why you are here’ or ‘Is there something that has upset you? Can you talk about it?’). The child’s words should be recorded as stated.

Children will feel less threatened if other aspects of their health are also examined at the same time. The normal child health enquiry about growth, diet, systems enquiry, and school function is important. Clinical examination of the child should also be holistic, and incidental findings should be relayed back to the parent and child appropriately. A whole-body approach is more likely to promote a sense of healing than a prolonged focus on the genitals and anus.

Physical abuse and sexual abuse are seen together in around 15% of cases. Physical signs of sexual abuse are uncommon even in long-standing, intrusive and painful abuse. Such subtle signs as may be present will be missed if the examiner does not encourage the child to be fully relaxed. The use of a high-quality lighting source is critical, as is the child’s posture. The gold standard is the use of photo-colposcopy, which provides magnification, light and a recording of the findings.

Small children are best examined in the frog-leg position, assisted by someone whom they trust. The knee–chest position may have to be used to define the hymenal free edge. Gentle anterior traction on the labia usually suffices to open or stretch the orifice. Older and pubertal

girls may benefit from the use of stirrups and a damp swab to identify deep clefts. This should be done after appropriate specimens have been collected for forensic analysis.

Criminal prosecution from sexual abuse allegations and forensic evidence gathering is a demanding process that requires strong links between law enforcement agencies and forensic examiners. Doctors and nurses who expect to provide such a service require training from experts in the field.

Essential reading for forensic practitioners should include *The Physical Signs of Child Sexual Abuse*. An evidence-based review and guidance for best practice. May 2015 produced by the Royal College of Paediatrics and Child Health in the UK. This document presents a review of all the substantial research into individual physical signs. It also presents guidance on best practice relating to examinations and healthcare. See further reading at the end of this section from the Faculty of Forensic and Legal Medicine, Royal College of Paediatrics and Child Health (2012) Guidelines on Paediatric forensic examinations in relation to possible child sexual abuse. Interim guidance regarding numbers of examinations and maintenance of competence was issued in 2020.

Diagnosis of Child Sexual Abuse

This is usually achieved following multi-agency assessment, the history and medical examination being only a part of the process.

Acute Sexual Assault Findings (within hours or a few days)

- Bruising and swelling, abrasions and lacerations to the external genitals without a history of accidental trauma.
- Grip marks and bruising around the limbs.
- Cigarette or lighter burns around the breasts and pubic area.
- Bite marks, including suction bites around the breasts, abdomen, and thighs.
- Petechiae around the eyes, tears to the oral frenulum, petechiae over the posterior fauces.
- Visible petechiae over the hymen, hymenal tears, haematomas, and bleeding.
- Petechiae or bites over the glans penis and scrotum.
- Bruising, oedema, and lacerations around the anal area (oedema usually resolves within 48 hours).
- Semen may be found in the vagina or rectum.
- Pregnancy is a major and not uncommon result.

Non-acute / historic Sexual Abuse Findings (the most common presentation):

- Hymenal transections, deep clefts and notches in the posterior hymen, and a complete absence of posterior hymenal tissue are signs of healed trauma and very rarely seen in girls who have not reported penetrative abuse. .
- Significant lacerations / tears can heal completely without scarring but may also heal to leave a notch or full-width transection on the hymen.
- The size of the hymenal orifice and the posterior hymenal width cannot be measured accurately and are non-discriminatory for sexual abuse.
- Scar tissue formation is an infrequent finding and most likely to be seen on non-hymenal tissue, only in girls who sustain a deep laceration. Recent studies have not identified scars on the hymen in pre-pubertal or pubertal girls.
- Hymenal injuries are never acceptable from a history of a straddle or other fall unless there is convincing evidence of direct penetration by an object.
- Small superficial notches, bumps and labial adhesions are not uncommon in non-abused girls.

- Anal findings are uncommon. Signs such as reduced tone, anal laxity, deep and poorly healing fissures and venous congestion may be seen in non-abused children and must be considered in the context of the history given.
- Most anal injuries heal without sign of previous trauma although extensive injuries heal to leave scar tissue and / or skin tags.
- Dynamic anal dilatation or dilatation of both internal and external anal sphincters in the absence of stool is associated with anal abuse.

TABLE 2.2 Differential diagnoses of genital and anal findings

Concerning Sign	Differential Diagnoses
Vaginal bleeding	<ul style="list-style-type: none"> • Accidental injury • Especially straddle injury • Urethral prolapse • Precocious puberty and other hormonal causes • Lichen sclerosis et atrophicus • Foreign body in genital tract • Severe vulvo-vaginitis • Tumours of the genital tract
Rectal bleeding	<ul style="list-style-type: none"> • Anal fissures caused by hard stool • Inflammatory bowel disease • Infective diarrhoea • Rectal polyps • Rectal prolapse
Vulvo-vaginitis	<ul style="list-style-type: none"> • Poor hygiene • Skin disease (e.g. eczema, lichen sclerosis) • Allergies to detergents/bath products

Additional considerations when performing examination of the genital and anal areas

- Ensure that a good light source is available, including the use of colposcopy, if this is available.
- Conduct interviews and examinations of children with another professional person present.
- Instrumental examination is not normally required in pre-pubertal girls. Assessment of the hymen in post- pubertal girls may require use of a cotton tip swab or other techniques.
- Knowledge of local practice regarding female genital cutting and male circumcision is important when interpreting clinical findings.
- Interpretation of anal signs is difficult and needs to be undertaken in conjunction with a careful history of the child’s bowel pattern.
- If forensic facilities are available, ensure that clothing items and relevant swabs are taken in line with local protocols, and that a chain of evidence is maintained to the forensic laboratory.
- Assess whether swabs and treatment for sexually transmitted infection (see Section 47) need to be taken immediately or at a follow-up review. Discuss with sexual health expert if available.
- Consider whether a pregnancy test needs to be carried out.
- Consider whether emergency contraception is needed.

- Consider the risks of HIV infection and whether post-exposure prophylaxis is needed in line with local protocols. This will vary depending on knowledge of the assailant, the nature of the injuries, and the country's HIV prevalence rates.
- Consider whether hepatitis B immunisation (if available) is indicated.

Forensic sampling in acute sexual assaults

A decision to undertake forensic sampling depends upon the following:

- Has contact abuse been reported?
 - There is always a possibility of transferred material if there has been direct contact. Even if a condom was used, relevant lubricant or saliva may be detected.
- How long is it since the assault? The Faculty of Forensic and Legal Medicine together with the RCPCH (Further reading) have published guidance intended to help the decision-making process when deciding if a forensic medical examination is warranted in 'child-unfriendly' hours, or can wait and be done during the day.
 - If it is less than 72 - 96 hours, there is a possibility of trace material being found, especially within skinfolds.
- Has there been bathing or washing?
 - Material may still be available, but this is less likely if the child has been washed thoroughly.
- How active is the child or adolescent?
 - Children who are immobile for reasons of illness or disability may retain trace material well beyond the standard time, as drainage from the vagina is erratic.

What samples should be collected?

The history should guide the practitioner in deciding where trace material is likely to be found. Any clothing worn by the child during the assault should be collected for forensic examination. The history may direct one to unusual sites (e.g. swabs may identify traces of adhesive from a victim who has alleged being strapped down with masking tape; microscopic rope fibres may be recovered from around the ankles or wrists). It is sensible to collect duplicate swabs from each area sampled.

In general, swabs lightly moistened with sterile distilled water should be used to collect material that is visibly dried on or speculatively present. Dry swabs are used in moist areas such as the mouth, glans penis, anus and vagina.

If the child is very young or an infant, semen or saliva may be present over a wide area (e.g. in the hair, armpits, abdomen, thigh creases). Damp swabs may be collected over all these areas in a young baby, whereas an adolescent is more likely to carry evidence over the breasts and in and around the vaginal area.

The use of an ultraviolet (UV) light in a dark room may help to identify both deposits of semen and saliva, and areas of deep trauma within the skin. These latter areas fluoresce because of disturbance of melanin, haemoglobin and collagen tissue. The UV light should be used with caution, as there is a risk of material denaturing with extended use.

An example of systematic head-to-toe trace evidence gathering could be as follows:

- Hair combings over a sheet of white paper, then folded and placed in a special plastic bag*
- Cut areas of hair if dried material is visible
- Specialised tooth brushings between the teeth and gum swabs if there has been oral ejaculation
- Fingernail scrapings if there was violent resisted assault *

- Damp swabs pressed firmly and rolled over any bites noted around the neck or breasts
- Swabs from the axillae and from within the umbilicus in a small infant
- Pubic hair combings*
- External vulval damp swabs
- Dry high vaginal swabs
- Swabs from the glans penis, behind the foreskin and over the shaft for saliva
- Damp peri-anal swabs
- Dry rectal swabs
- All clothing bagged individually as removed
- Tampons and sanitary towels similarly bagged.

Every item collected should be fully labelled, bagged and sealed by the receiving witness, most commonly a police officer. It is sensible to allow a brief interval between samples to ensure that earlier samples have been correctly dealt with.

* Forensic material obtained in this way is only relevant if the alleged perpetrator denies all contact with the victim,

Differential Diagnosis of sexual abuse

Several common naturally occurring conditions not due to abuse may give rise to a suspicion of sexual abuse but must be excluded.

Non-specific vulvo-vaginitis is the commonest. A frequent presentation in the pre-pubescent child, symptoms are of intermittent mild dysuria, redness and a sticky discharge. Symptoms are likely to relate to withdrawal of the maternal oestrogen effect, which makes some children intolerant of the use of strong detergents and poor hygiene practices. The use of loose-fitting underwear, gentle cleansing and the regular application of simple emollients usually provide relief of a condition that tends to recur until early puberty.

The presence of pinworms can cause genital symptoms, as can localised eczema in the napkin area. These require appropriate treatment.

Infective vulvo-vaginitis presents with significant inflammation and pain, sometimes associated with upper airways infection, and often streptococcal in origin. If possible, bacterial cultures should be obtained before appropriate antibiotics are offered.

Lichen sclerosus et atrophicus is an uncommon skin disorder which may be associated with other autoimmune disease, including morphea in adults, but it tends to be a stand-alone diagnosis in children. It presents with fragility, haemorrhaging and bruising of the skin of the labia, dysuria and occasional urinary retention. Diagnosis is made easy in longstanding cases by the classical picture of de-pigmentation in a figure-of-eight configuration associated with obvious skin fragility, and easy bleeding on stretching. Vigorous treatment with emollients is often adequate in mild cases, but topical steroids may be required to control severe signs and symptoms.

Retained foreign bodies can be the cause of intermittent bloodstaining and purulent or offensive discharge in very young children. It should be recognised that repeated insertion of foreign bodies into the vagina by a young child may be the presenting sign of learned or disturbed behaviour.

Constipation can give rise to intermittent anal bleeding and discomfort.

Inflammatory bowel disease may present with anal fissures, bleeding and discharge.

It is important to communicate to parents and other professionals that sexual abuse may not result in any physical findings, and that there are few signs which are absolutely diagnostic. Nevertheless, a medical examination following an allegation of sexual abuse may provide valuable forensic information as well as an opportunity for reassurance, treatment of infection, and access to wider therapeutic support.

Difficult judgements about how to proceed may have to be made in settings where female genital cutting is practised, and where legislation and/or community action against this practice is weak.

For sexual abuse, attention must be given to detection and treatment of all acquired sexually transmitted infection, including HIV, preferably within 2 weeks of an acute assault and possibly at the same time as the examination for long-standing abuse. Pubescent girls should be offered emergency contraception where indicated.

All incidental findings (for examples: anaemia, rashes, heart murmurs) and reported health problems should be attended to and followed up where necessary.

Sexually Transmitted Infections (STIs)

A sexually transmitted infection may be the presenting feature in sexual abuse. Children who have experienced contact sexual abuse should be screened for STIs. The screening programme should take local prevalence factors into account, as should the decision to offer prophylactic antibiotics or antiviral treatment (see also Section 47).

Neisseria gonorrhoeae (especially non-conjunctival gonococcus) is not an expected infection outside the neonatal period and is strong evidence for sexual abuse. *Chlamydia trachomatis* similarly usually implies sexual abuse. There is evidence for vertical transmission at birth, and limited research evidence for the persistence of asymptomatic colonisation beyond the first year of life.

The presence of either of these organisms, especially if symptomatic in mid-childhood and beyond, should raise the strongest suspicion of abuse, regardless of the presence of maternal infection. The evidence does not help to establish the age at which the possibility of vertical transmission can be excluded.

Trichomonas vaginalis may cause an offensive discharge in adolescents, and is a strong marker for sexual activity, consensual or otherwise. It is not known to infect the pre-pubescent child, although it may be found colonising newborns from infected maternal secretions.

Human papillomavirus is a very common infection, and hand-to-skin transmission is so frequent that it is usually difficult to use this infection to resolve issues of sexual transmission. Transfer may be perinatal, and lesions may be seen in early childhood in the mouth and larynx. However, the presence of ano-genital warts should mandate careful examination and screening for other STIs.

Herpes simplex virus (HSV) raises similar issues. Auto-innoculation and benign transfer cannot be excluded. Adolescents presenting with symptomatic genital HSV are likely to have had sexual contact. They need careful evaluation and screening for other STIs.

Treponema pallidum (syphilis) may be acquired through the placenta, and signs or symptoms of infection are highly unpredictable. Most studies in older children suggest transfer through sexual contact. The infection is exclusively sexually transmitted in adults. Sexual transmission should always be considered in a child presenting with symptoms. Infection in the parent does not exclude abuse.

Hepatitis B and hepatitis C are recognised as being sexually transferred in adults. There is insufficient research in children; however, screening of the sexually abused child is mandatory in high-risk situations, such as multiple or violent assaults and in high-prevalence regions.

Human immunodeficiency virus (HIV) see Section 36: The frequency of infection acquired through abuse will reflect the prevalence of HIV in the local population, and screening is strongly advised where the prevalence is high. The possibility of mother-to-child transmission or blood contamination for the blood-borne viruses should be excluded.

Screening for hepatitis and HIV will need to be timed to take account of time to seroconversion, which is usually a period of 6 or more weeks. Repeat screening for late conversion may be considered up to 12 weeks.

Most STIs can be screened for at 2 weeks from the date of contact. *Neisseria gonorrhoeae* should be screened for at the first possible opportunity, preferably at the time of assessment if feasible, or within 48 hours. Consideration should be given to hepatitis B vaccination if the child presents within a week or two after a penetrative assault. Similar consideration may need to be given to HIV prophylaxis on occasion.

Fabricated or Induced Illness (FII)

This is the severe end of a spectrum of unusual or abnormal health-seeking behaviours in which significant harm is caused by a parent or carer (usually the mother), who deliberately fabricates signs or symptoms or induces illness in a child. Sometimes the abuse is the direct result of inappropriate and often invasive and unnecessary investigations or treatment by healthcare workers responding to the parent's fabricated accounts of non-existent illness. The child is frequently brought for multiple medical assessments and investigations, the perpetrator (often the child's mother) denies knowledge of the causation of the illness, and the acute signs and symptoms cease when the child is separated from the perpetrator.

There is ongoing debate regarding terminology, but the Royal College of Paediatrics and Child Health in the UK (February 2021) recommend the use of Medically Unexplained Symptoms (MUS), Perplexing Presentations (PP) and Fabricated or Induced Illness (FII). In MUS, a child's symptoms, of which the child complains, and which are presumed to be genuinely experienced, are not fully explained by any known pathology. The term PP was introduced to describe the commonly encountered situation when there are alerting signs of possible FII (not yet amounting to likely or actual significant harm), when the actual state of the child's physical, mental health and neurodevelopment is not yet clear, but there is no perceived risk of immediate serious harm to the child's physical health or life. The alerting signs include the presence of discrepancies between reports, presentations of the child and independent observations of the child, implausible descriptions and unexplained findings or parental behaviour.

FII is a clinical situation in which a child is, or is very likely to be, harmed due to parent(s) behaviour and action, carried out in order to convince doctors that the child's state of physical and / or mental health and neurodevelopment is impaired (or more impaired than is actually the case). FII results in physical and emotional abuse and neglect, because of parental actions, behaviours, or beliefs and from doctors' responses to these.

Healthcare workers in hospital are often the first professionals to suspect FII in a child based on concerns about:

- Being given erroneous or misleading information, eg reported physical, psychological or behavioural symptoms and signs not observed independently

- Parents' insistence on continued investigations, instead of focusing on symptom alleviation when results of examination and investigations have already not explained the reported symptoms or signs in the child
- The introduction of foreign material into investigative tests.
- Deliberate fabrication of fits leading to dangerous treatments
- Unusual results of investigations (eg biochemical findings, unusual infective organisms)
- Removal of or tampering with medical monitoring equipment
- Deliberate poisoning
- Deliberate burns or damage to the skin
- Deliberate suffocation

Additional considerations when investigating fabricated and induced illness (FII)

The investigative process must involve early and continuing collaboration between all agencies, with detailed information sharing. Strategy planning meetings involving professionals from health, social services, police, education, and legal departments can be very helpful.

- Draw up a health chronology using all accessible sources of information.
- Gather forensic or witness information.
- A decision must be made by the multi-agency team as to whether it is necessary to separate the child from the suspected perpetrator by voluntary or legal means to determine whether symptoms or signs that might be fabricated or induced cease during separation.
- Decisions with the child protection division of the police about the potential value of and potential dangers of undertaking covert video surveillance
- A decision must be made by the multi-agency team as to how and when to confront the parents.
- Ensure the child's safety throughout the investigative process.
- Work within the country's legal framework which **must ensure mandatory reporting**.

Special issues concerning the management of child abuse

The child's immediate needs are as follows:

- To feel believed and acknowledged:
 - All interaction with the child should convey this message.
 - Intrusive questioning, especially questions that imply some measure of blame (e.g. 'Why didn't you tell earlier?') can cause a child to refuse to speak further and even retract previous statements.
 - The protective parent or carer should be briefed about this risk.
- To be safe from further harm:
 - Protection usually involves the statutory services (if they exist).
 - Police and social workers should be part of the multidisciplinary process that assesses the child's safe custody.
 - The safety of siblings must also be considered at this stage, if not before.
- To have all their health needs met:
 - Their immediate needs are for reassurance that they are healthy, and that any changes in the genital area will heal.
 - Care should be taken to use the right language to inform the parent and the child about these changes.
 - For sexual abuse the use of phrases such as 'no longer a virgin' is highly inappropriate in this context, and indeed anatomically inaccurate in most cases.
- **Mandatory reporting** by health and social care workers of suspicions of child abuse need to be established in every country. The welfare of the child is paramount, and every professional has a duty to protect children from harm.

Medical aftercare following childhood maltreatment

It is important that these children are offered follow-up in order to:

- Monitor the child's overall progress
- Ensure healing of injuries
- Investigate and treat any acquired infection
- Facilitate access to psychological therapeutic support.

Healthcare workers involved in the hospital care of children who have been abused may be asked to provide a police statement and to attend court as a witness.

Long-Term Needs

Those children who are going through the criminal justice process will need extensive support and counselling.

In well-managed sexual abuse allegations, most young children appear to function well. However, the disruption to their lives and the loss of familiar adults and objects around them does have an impact on many, and it is important that their carers are briefed.

Children who have been brutally sexualised may not recover and may seek inappropriate affection and contact which makes them difficult to parent. Others may suffer nightmares, phobias and other symptoms of anxiety. Long-term psychological input may be necessary.

Abuse involving special circumstances

Child trafficking

In 1994, the United Nations General Assembly defined trafficking as the 'illicit and clandestine movement of persons across national and international borders, largely from developing countries and some countries with economies in transition with the end goal of forcing women and girl children into sexually or economically oppressive and exploitative situations for the profit of recruiters, traffickers, crime syndicates, as well as other illegal activities related to trafficking, such as forced domestic labour, false marriages, clandestine employment and false adoption.'

It is estimated that in the last 30 years, trafficking in women and children in Asia for sexual exploitation alone has victimised over 30 million people.

Children are trafficked for several purposes, including:

- Sexual Exploitation
- Adoption
- Child Labour
- Child Soldiers
- Forced Marriage
- Body Parts
- Ritual Sacrifice.

Parents are promised education or jobs for their children. Some children are simply captured, then traded for whatever commodity is in demand (domestic work, sex work, drug carrying or beggary).

Children who are displaced are highly vulnerable to sexual and physical abuse. They fear seeking help and often do not have the language to do so.

Different cultural situations produce different types of exploitation. In Asia, for example, girls as young as 13 years may be exported as mail-order brides, and in Thailand around 100 000 women and girls from border countries are imported into the sex trade. Large numbers of children are being trafficked in West and Central Africa, mainly for domestic work but also for

sexual exploitation, to work in shops or on farms, or to be scavengers or street hawkers. Nearly 90% of these trafficked domestic workers are girls. Many of these girls are traded on into the Middle East and Europe.

The International Organisation for Migration (IOM) has produced an extensive document that comprehensively deals with all aspects of victim management (see Further reading section at the end of this section).

Advice, both for the country of origin and for the receiving country, is structured based on two principles:

- The child's interests are paramount.
- Above all, do no harm.

The starting point is the assessment of risk both in the receiving country and in the country of origin if repatriated. Risk depends on numerous factors, including the following:

- The extent to which trafficking is controlled by organised criminal groups
- Their known or estimated capacity to plan and implement reprisals against the victims and/or service delivery organisation staff
- The capacity of the local law enforcement agencies
- The extent of endemic corruption and how it adds to the level of risk.

It is critical that children have an appointed independent guardian who will act solely in their interest. Family members, including parents, may well be responsible or collusive in the trafficking, and drawing them in could greatly increase the risk of serious harm, including death, or re-trafficking. In some cultures, it may be socially acceptable for the family to shun or even kill a girl for having brought disgrace on her family.

Trafficked women may give birth to children within their repressive conditions. These children will be at very high risk of emotional abuse and neglect and early introduction into commercial sex. Babies are at risk of homicide.

Services provided for trafficked children should reflect the following needs (adapted from the IOM Report):

Approaches that demonstrate respect and promote participation (e.g. children being allowed to express their views in the language they speak best).

An understanding of the complex ways in which their experience has harmed them. Children who are trafficked are subjected to a persistently threatening and dangerous environment. In the face of this type of chronic abuse and stress, children and adolescents develop a personality that is suited for survival, but that is ill adapted to cope in normal non-threatening situations. Healthcare practitioners are responsible for employing health-promoting strategies, programmes and activities that recognise the child's level of development and help children and adolescents to reclaim and further develop their competencies for an active and meaningful life. This involves addressing a range of needs, including nutritional, physical and psychological development and education needs.

Tailoring services to meet the needs of each age group and in ways appropriate to the age and characteristics of the child concerned, never merely following programmes designed for adults. They should be assessed and managed by professionals trained in child development. Medical assessments need to be child friendly and provided by people with expertise.

Implementing strategies aimed at mitigating the effects of past trauma and fostering healthier patterns of development. One example of such strategies is stepwise early re-integration into education and into a peer group.

Rape as a Weapon of War

The rape of adults and children of both sexes is a common phenomenon in conflict zones (see latest reports from Ethiopia: 2020). As long ago as 1949, Article 29 of the Geneva Convention explicitly forbade degrading treatment, stating that 'Women shall be protected against any attack on their honour – especially rape, enforced prostitution and indecent assault.'

Rape as a crime against humanity was first prosecuted in the International Criminal Court in 2001 when three Bosnian Serbs were convicted of systematic sexual violence against Muslim women.

However, prosecution of these crimes by the relevant states has been negligible. In Rwanda, the mass rapes of the Tutsi women and girls permanently destroyed the capacity to child bear for some and forced others to bear children outside their ethnic group. Vast numbers of women and girls were rejected by their communities and became outcasts. Thousands of children witnessed the violence on their mothers and sisters. As rape as a weapon of war demoralises and destabilises entire communities, it weakens ethnic communities and ties, and affects populations with the exploitation of the reproductive rights and abilities of its victims. When rape is employed instead of a bullet, the weapon continues to wield its power beyond the primary victim. The battlefield may be the body, but the target is civil society. 'Rape, as with all terror-warfare, is not exclusively an attack on the body – it is an attack on the "body-politic." Its goal is not to maim or kill one person, but to control an entire socio-political process by crippling it. It is an attack directed equally against personal identity and cultural integrity.'

Thus in 1998, rape as an act of genocide was the decision of the International Criminal Tribunal for Rwanda. Despite these major precedents, prosecution of sexual crimes by the relevant states has been negligible. Rape has been a major feature of the war in South Sudan and in the Democratic Republic of Congo. Children as young as 5 years of age have been deliberately targeted. There is also strong anecdotal evidence that young soldiers barely in their teens have been ordered into gang rape to prove their 'manhood'.

The provision of care for victims of mass sexual abuse at this level is a daunting task and should involve major planning and resources. Emergency care for severe physical wounding during the assaults is logistically difficult. Pratt and Werchick recommend expanding access through 'mobile teams of rape specialists' that could not only provide treatment themselves, but also transport medical supplies and transfer knowledge to any staff already on the ground. Medications, including emergency contraception, hepatitis vaccine, STD prophylaxis and antiretroviral drugs, need to be available. Such teams will need to have access to surgical facilities, especially when very young children are involved.

Gang rape, the use of instruments and other violence increases the risk of HIV/AIDS transmission significantly; intercourse is accompanied by injuries and bleeding which increases the transmission of the virus compared with transmission during consensual sex. Internal vaginal and rectal injury can be very serious, and in the very young may be fatal.

According to Human Rights Watch, 'children were reportedly forced to hold their mothers down while they were raped'. It is not difficult to see that a significant range of service provision is required at several levels to deal with such traumatic damage in childhood.

Therapeutic services in isolation without intensive educational programmes and a whole-community approach are probably doomed to fail. Rape as a weapon of war activates cultural beliefs that result in the marginalisation of its victims, especially women and children, thus preventing those victims from receiving psychosocial support, and depriving them of

income. Women and girls are considered to be damaged and 'contaminated'. Wives may be denounced by their husbands, blamed for the rape, and regarded as 'married' to their rapists. Thus, communities see their raped women and children as enemies and place them outside their sphere of moral obligation. Some communities may demand that their wives and children leave their villages.

Empowering young children and their mothers by providing education, and the teaching of new skills leading to longer-term economic stability, are also areas that need careful planning. Provision of safe housing and basic needs may be all that is possible in the immediate aftermath of sexual violence.

Female Genital Mutilation/Cutting (FGM/FGC)

Female genital cutting (FGC), also known as female circumcision or female genital mutilation, refers to all procedures involving partial or total removal of the external female genitalia, or other injury to the female organs for non-therapeutic reasons. It ranges from very simple to radical, and may be carried out between birth and puberty, or can be performed just before marriage or childbirth. FGC/FGM varies across cultures, ethnic groups, and tribal affiliations. Globally, over 200 million women and girls are affected by FGM, most of them in Africa. <https://www.28toomany.org>

FGC is commonly performed by traditional medicine practitioners, including traditional birth attendants, local women or men, or female family members. Such individuals usually perform cutting without anaesthesia or sterilised instruments, using kitchen knives or razor blades. Sometimes, it is performed with corrosive substances. It is not uncommon for those who perform FGC to cut or damage more of the genital area than they intended to. Increasingly, doctors are also undertaking these procedures which is not considered ethical by WHO and in some countries will be illegal.

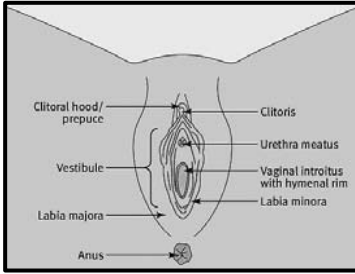


Figure 2.3 Normal female external genitalia..

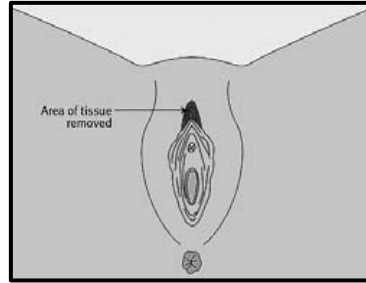


Figure 2.4 Area of tissue removed in type 1 female genital cutting.

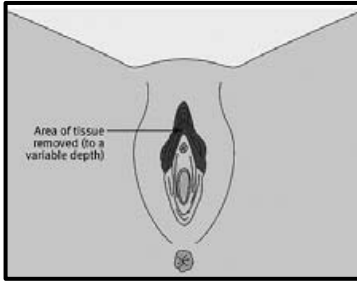


Figure 2.5 Area of tissue removed in type 2 female genital cutting

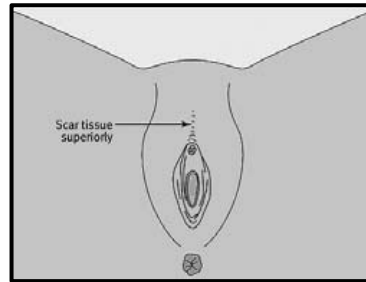


Figure 2.6 Appearance of genitalia after type 2 female genital cutting.

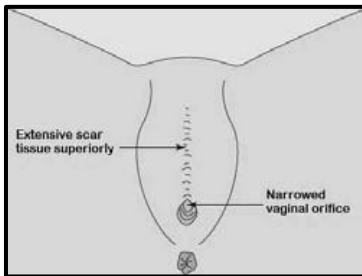


Figure 2.7 Appearance of genitalia after type 3 female genital cutting.

Implications and complications of FGC

The health problems associated with FGC are life-threatening haemorrhage, sometimes death during or shortly after the procedure (from haemorrhage or infection), death during pregnancy, the need for assistance during childbirth due to interference with normal delivery, and the spread of HIV/AIDS and hepatitis due to the frequent use of unclean and unsterile instruments. There are also links to mental illness in the victims and to intimate partner violence.

FGC is dangerous to girls' and women's health and psychological well-being. It can cause urological, gynaecological and obstetric problems. Around 10% of girls and women are estimated to die from the short-term complications of FGC, such as haemorrhage, shock and infection. Another 25% die in the long term because of recurrent urinary and vaginal infections, as well as complications during childbirth, such as severe bleeding and obstructed labour.

Short-term complications

1. haemorrhage and anaemia
2. severe pain (it is almost always the case that no local anaesthetic is given)
3. shock (due to haemorrhage and/or pain)
4. death from shock (due to haemorrhage and/or pain)
5. difficulty passing urine or faeces
6. urinary tract infection
7. urethral meatus injuries, prolonged micturition, and dysuria
8. injury to adjacent tissues
9. damage to other organs
10. fractures or dislocation due to restraint during the procedure
11. infection due to tetanus, and bloodborne viruses such as HIV, hepatitis B and C
12. vulval abscess.

Long-term complications

- 1) chronic pain
- 2) chronic pelvic infection
- 3) haematocolpos (obstruction to menstrual flow, leading to dangerous swelling of the vagina)
- 4) keloid scarring
- 5) vulval epithelial inclusion cysts
- 1) decreased quality of sexual life, including pain on intercourse
- 2) complications in pregnancy and childbirth, including obstructed labour (see below)
- 3) psychological damage, including fear and anxiety during labour and delivery, as well as post-traumatic stress disorder and depression
- 4) psychosexual effects; fear of, and anxiety about, sexual intercourse, difficulties with penetration, marital break-down and divorce.

Safeguarding children who are at risk of FGC/FGM

- The safety and welfare of the child is paramount.
- All agencies must act in the best interests of the rights of the child as stated in the UN Convention on the Rights of the Child (1989).
- In some countries, FGC/FGM is illegal, and it should be known in the country where you are practicing what the law actually states.

It is acknowledged that some families see FGC/FGM as an act of love rather than of cruelty. However, FGC/FGM causes significant harm in both the short term and long term and constitutes physical and emotional abuse of children.

The situation for FGC/FGM in Liberia

“President Ellen Johnson Sirleaf left office in January 2018 with a tremendous, if overdue, parting gift for the girls of Liberia. During her final hours in office, Africa’s first woman elected head of state signed an executive order abolishing female genital cutting, an ancient practice that had been endured by more than half of Liberia’s girls.

The fight is not quite over. Lawmakers have a year to enshrine the ban into law, and it may be many years before the law is properly enforced. But it is a momentous step that seemed unthinkable just six years ago, when an explosive newspaper article propelled the issue onto the national agenda”.

https://www.cjr.org/first_person/liberia-female-mutilation.php

Appropriate care for women and girls who have been subjected to FGC/FGM

- Provide access to information, support and services.
- Provide care pathways and guidelines for professionals.
- Ensure that information is accurate and up to date.
- Empower women and girls and encourage them to speak out and seek help.
- Engage and mobilise all concerned and develop an understanding of cultural diversity.
- Be open and supporting, sensitive and non-judgmental.
- Encourage alternative rites to FGC/FGM. This is a strategy that retains all of the rites of passage or initiation that the girls would traditionally undergo, except for the genital cutting. The girls are still encouraged to learn essential domestic duties that would be useful when they are married.

FGC/FGM is a violation of human rights. It is essential to empower women and girls, to encourage women to have a voice, and to raise awareness of the dangers of FGC/FGM. Engagement with all concerned local communities is crucial, including community and religious leaders. It is essential to work with all professionals. We all have a duty and a responsibility to safeguard girls who are at risk of FGC/FGM, as the welfare of children is paramount.

Further reading relevant to child protection in low resource settings

MCAI Improving care for children living in institutions

https://12dec1c3-e3f4-e784-7f3a-6e7903610962.filesusr.com/ugd/dd2ba4_5f66bc7d510b067dfd38f1a928fc32a7.pdf

Accessed 30th March 2021

MCAI Interagency child protection guidance handbook for low resource settings

https://12dec1c3-e3f4-e784-7f3a-6e7903610962.filesusr.com/ugd/dd2ba4_a6d7a20f903479dc0ebd096f66d4b083.pdf

Accessed 30th March 2021

PSEA assessment and PSEA toolkit for CSO partners

<https://sites.unicef.org/about/partnerships/files/Information-Brief-PSEA-Assessment-PSEA-Toolkit-for-CSO-Partners-24-Jan-2020.pdf>

Accessed 30th March 2021

Child Protection and Safeguarding in the UK. <https://childprotection.rcpch.ac.uk/>

Accessed 30th March 2021

Faculty of Forensic and Legal Medicine / Royal College of Paediatrics and Child Health joint publications on child sexual abuse can be found on -

<https://www.rcpch.ac.uk/resources/faculty-forensic-legal-medicine-fflm-guidance>

Accessed 30th March 2021

The Child-Friendly Healthcare Initiative (CFHI): Healthcare Provision in Accordance With the UN Convention on the Rights of the Child

https://12dec1c3-e3f4-e784-7f3a-6e7903610962.filesusr.com/ugd/dd2ba4_a1f7170e7d16f53ad143be8a4bcee8a6.pdf

Accessed 30th March 2021

Child friendly healthcare A manual for health workers MCAI

https://12dec1c3-e3f4-e784-7f3a-6e7903610962.filesusr.com/ugd/dd2ba4_f1fda93a47c64e19a058893659cc015c.pdf

Accessed 1st April 2021

Child Maltreatment & Abuse: Special Issue of Paediatrics and International Child Health.
https://12dec1c3-e3f4-e784-7f3a-6e7903610962.filesusr.com/ugd/dd2ba4_335d3062fa1d452da844a25a6e61d531.pdf
 Accessed 30th March 2021

Classification of child abuse by motive and degree rather than type of injury
https://12dec1c3-e3f4-e784-7f3a-6e7903610962.filesusr.com/ugd/dd2ba4_1f7e4250d9c2a29c1e90f34e823aa208.pdf
 Accessed 30th March 2021

The Maternal and Child Friendly Healthcare Initiative (MCFHI)
 A manual for health workers based on a medical ethics approach to healthcare.
https://12dec1c3-e3f4-e784-7f3a-6e7903610962.filesusr.com/ugd/dd2ba4_75777218b5234c91b817072eb1433ee4.pdf
 Accessed 30th March 2021

Appendix on child protection issues

Examination under child protection procedures: Suspected physical and/or sexual abuse
 Was consent sought/obtained by the child/young person or the parents/guardian.

CONSENT: YES/NO From who: Name:

Although child protection procedures may override consent, it is good practice to document that it was sought in the first place.

Patient details (circle correct information)

Name:	Date:
Date of birth.....	Time:
Age:	Place of examination:
MALE/FEMALE	
Address (prior to examination):	

Professionals involved in the assessment

Doctor's or nurse's name:	Police officer's name:
Social worker's name:	
School:	

Why was this examination undertaken?

--

Family and Social History
(including names, dates of birth, ages, occupations/schools, relationships)

--

History of any known medical problems

--

Persons present during examination

1.
2.
3.
4.

Examination of child

Age:	Years: Months:
Height/percentilecm/.....

Section 2 Identifying and managing children who have been abused. Dr. Jacqui Moq, Dr. Comfort Momoh, Clarissa Cupid, Dr. Neela Shabde, Dr. Martin Samuels and Prof. David Southall

Weight/percentilekg/.....
Head circumference/ percentilecm./.....

General appearance of child (any obvious neglect):

Significant comments made by the child or the parent/carer (record as accurately as possible):

Developmental assessment (circle correct answers):

Delayed development: Yes / No If Yes Severe / Moderate / Mild

Level of puberty: Pre-pubertal / Post-pubertal

General examination (use body maps for any injuries)
[Include inspection of oral frenum and palate and scalp]

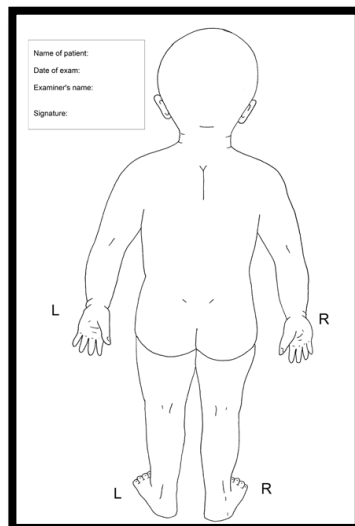
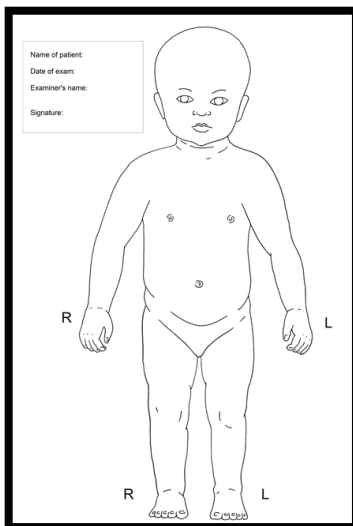


FIGURE 2.3 Diagrams on which to mark signs of injury to the front and back of the body. Reproduced with permission from Southampton City Primary Care Trust.

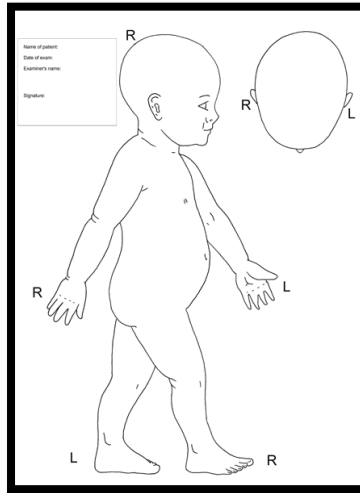


FIGURE 2.4 Diagram on which to mark signs of injury to the right side of the body. Reproduced with permission from Southampton City Primary Care Trust.

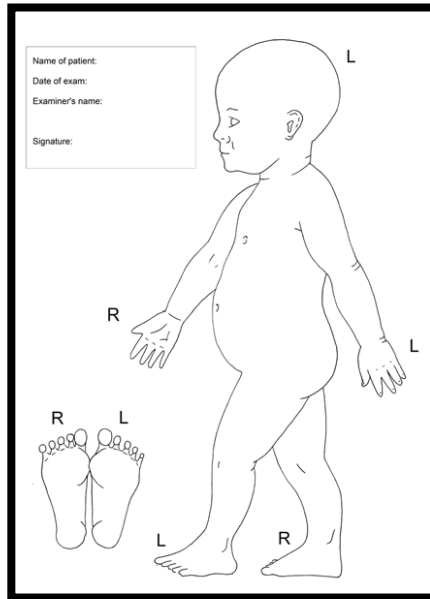


FIGURE 2.5 Diagram on which to mark signs of injury to the left side of the body. Reproduced with permission from Southampton City Primary Care Trust.

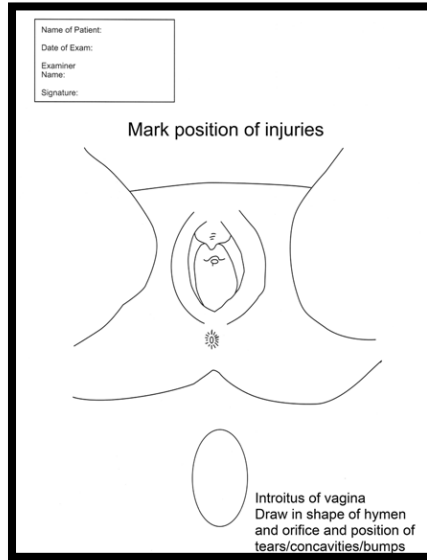


FIGURE 2.6 Diagram on which to draw the shape and position of any lesion on the female genitalia or anus. Reproduced with permission from Southampton City Primary Care Trust.

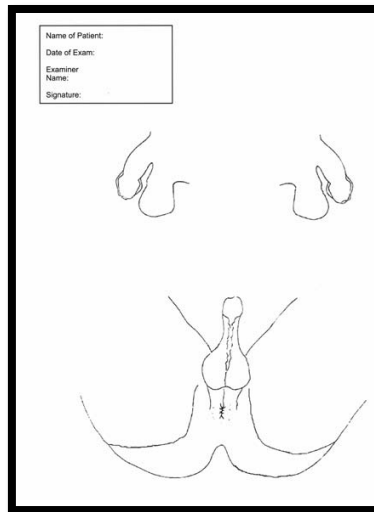


FIGURE 2.7 Diagram on which to draw the shape and position of any lesion on the male genitalia or anus. Reproduced with permission from Southampton City Primary Care Trust.

Examination of the anus

Issue/area examined	Details
Anal laxity/anal grip	
Anal folds	

Anal margin	
Surrounding tissues	
Examination position	

Examination of female genitalia

Issue/area examined	Details
External genitalia	
Pubertal signs?	
Labial separation or traction used?	
Labial fusion?	
Urethral opening	
Labia minora	
Peri-hymenal tissues	
Posterior fourchette	
Perineum	
Hymenal opening	
Hymen	
Examination position	

Examination of male genitalia

Issue/area examined	Details
Frenulum	
Urethral meatus/ Any discharge	
Signs of genital injury? Location	
Testicular swelling?	
Warts or skin disorders?	
Examination position	

Photographs Taken of Injuries

By whom	
Of what	

X-Rays Taken

Of what	
---------	--

Forensic Samples Collected

By whom	
Handed to whom	
List samples collected	

Screening for Sexually Transmitted Infections STIs

Date of Tests				
Tests Taken				
Results and Dates				

Any other clinical investigation:

--

Summary and interpretation of significant abnormal findings:

--

Conclusions and Doctor's or Senior Nurse's opinion:

--

Points discussed with social worker and parent/carer (and their opinion if applicable):

--

Arrangements for health follow-up for child (including investigations):

--

Signature: **Date:**

Circulation list for report:

Social Worker	
Police	
Head-Teacher at School	
Others (Please Specify)	

Name(s) of other children possibly at risk of abuse:		
Surname	First name	Date of birth

Checklist After Examination:

1. Have you been able to give a clear opinion on the case?
2. Have you considered alternative explanations for the findings?
3. Does the social worker understand your findings and opinion?

4. If the injuries are serious or indicate serious risk, have you considered the need for police involvement?
5. Are you happy with plans for the immediate safety of the child?
6. Are you in agreement with the proposed long-term management?
7. Is it important for you to attend the case conference? If so, make sure that the social worker knows this.
8. Have you recorded your discussions?
9. Have you written a care plan?

Section 3 Nursing sick children

Introduction

The range of professional nursing care is vast, varying from the challenges of meeting basic human needs, key assessment, and monitoring roles, to complex case management. Irrespective of the arena of nursing, a priority is to ensure that healthcare retains a focus on humane actions that increase the quality and effectiveness of care.

Children are unique individuals with rights enshrined by the UN Convention on the Rights of the Child (UNCRC) 1989. They have physiological, psychosocial, and emotional needs and responses that are different from adults. Effective nursing requires knowledge of these differences (recording vital signs means little without an understanding of the normal range). Much can be achieved by a **skilled 'child-friendly' approach** that leads to minimal intervention and trauma and includes the family and child in their own care and decisions.

In recent years there has been increased global focus on improving the quality of care provided. This recognises that increasing access to health care is of limited value if the quality of care is poor. Clinical factors such as guidelines, medical supplies and equipment are important contributors to quality of care; but improving children's 'Experience of Care' is also now recognised as vital to make health care more effective and to help children have better outcomes. Nurses are central and pivotal to efforts to improve quality and experience of care.

Role and education of nurses

The role of nurses, together with the education and status that they receive, varies widely, although there are steps to increase standardisation. Many countries provide no specific training in the nursing of sick children. Good care is often provided despite many challenges. The demands of healthcare sometimes result in an overlap of roles between nursing and medical staff, and in many situations the nurse may be the most experienced or skilled healthcare professional present. However, high quality care is much more likely to be consistently achieved by enabling nurses to undergo education and professional accreditation that relates specifically to the holistic physiological and psychological care of the child and their family.

The importance of involving families in care

Hospitalisation is often a frightening experience for children where the family no longer have their usual control over everyday life. Nurses should aim to return much of that control back to the child and family, and ensure that they receive the best care possible, given the environment and resources at their disposal.

The benefits and importance of involving parents cannot be overestimated. They, or another carer, should be supported to participate in the child's care and should be listened to. They can provide valuable information about what is normal for their child and how this may have changed. Since they know their child best, they are often the first to notice small changes in his or her condition that may prove significant. However, this critical knowledge can only be utilized if the contribution of families is recognised and valued.

Nurses as communicators and advocates

Nurses are central to communication between the multi-disciplinary team and the family. They have a key role to act as an advocate for the child.

Inappropriate and unnecessary painful interventions and investigations, long hospitalisations without good reason and unnecessary separations from the family cannot be justified and are an abuse of children's rights under the UNCRC. Hospitals and other healthcare settings present numerous potential risks to children.

It is the responsibility of every healthcare facility to be a place of safety, and of every nurse and healthcare worker to protect and defend the best interests of children. The hierarchical cultures in some healthcare systems can make this difficult, often to the detriment of the child. It is the responsibility of all healthcare professionals to promote an environment in which the views of all involved in the child's care, but especially the views of the child and their parents, are heard.

Basic nursing care

Organising care

In many places a small number of nurses and assistants have to look after many children. This makes it difficult to provide good and safe care. It is a major challenge for nurses and is another reason for working closely with parents and families.

It is important to organise nursing care well and efficiently, particularly when resources and the number of nurses are low. Even when there are good resources, poor organisation will result in suboptimal and inequitable care.

A system of allocating several patients to each nurse or grouping children together by the acuteness of their needs, can help to make the best use of available resources and ensure some continuity of care. However, any system of care is likely to be better than no system at all.

In busy settings it is also important to place the sicker children (particularly children without a parent present) in beds where they can be easily and most frequently seen – for example, near to the nurses' station.

The importance of basic care

Although patients may sometimes require complex treatment, there are basic needs that will always have to be met. Unfortunately, these are sometimes viewed as being of secondary importance, but providing good basic care often reduces the need for further intervention, and can enhance other therapies (for example the beneficial effect of improved nutrition on wound healing), whereas poor basic care has the opposite effect (e.g., the adverse effects of stress on respiratory and cardiac function). Where therapeutic options are limited due to lack of availability, limited financial resources, etc., or perhaps where no further curative treatment is possible, the provision of good basic personal care is particularly important. It is invariably viewed as part of the nursing role, but the underlying approach required is common to all healthcare professionals, and provision of good basic care is rarely a simple task. It requires understanding, skill and patience, and is a subject better covered in expanded publications; this manual only provides a simple overview.

Information, participation, and comfort

Hospitalisation often represents a traumatic change in the life of a child. Much that was previously familiar and predictable in their life has been replaced by an unfamiliar environment and fears about an uncertain immediate (and maybe long-term) future over which they have little control. Effective communication plays a vital role in the care of children and has a dramatic impact on their experience of and response to treatment. The words and expressions of nurses and other health workers can convey kindness, reassurance, information and support, that significantly helps children cope. Sometimes the power of this is undervalued in comparison to more clinical aspects of care, but even in the busiest situations it is possible and important to provide care with warmth and kindness.

Both the child and their family have a need to trust those who are caring for them, to be told what is wrong and also to know what is going to happen to them. This is a major factor

in helping them to adjust to the situation, develop coping strategies and make decisions about their own care.

The 'information needs' of children are often neglected, sometimes because it is assumed that their understanding is limited or because local traditions may not be very inclusive of children. However, even young children have a need to be given information in a language that is understandable to them. In the absence of reliable information, a child's fantasy and anxiety may be far more distressing than the reality.

Information-giving is particularly important in preparing the child (and their family) for procedures. They need to be told truthfully and sensitively what will happen, particularly if it might be painful. If the child is not warned, their trust in those around them can be destroyed, future procedures will be feared, their anxiety will be increased. The procedure may also become more difficult to do or less effective.

Children and their families should be allowed and encouraged to participate in decisions that affect them. This requires a willingness and ability to engage with them, actively listen, and to interpret non-verbal signs that may indicate something different to the words that are spoken. Children and families need to be able express their worries and concerns. Young children do not always have sufficient vocabulary to say exactly how they feel, so the use of play and other activities, such as drawing, can be a good way to enable them to express themselves.

Issues that may appear trivial to an adult may be important to a child. There may be issues causing distress that can be easily resolved, and in cases where there is not a solution, the feeling of isolation that may be experienced by children can be helped by having someone to share their anxieties with.

Where the situation is grave, families sometimes struggle to discuss distressing subjects with their child. Although these are difficult and emotional situations, even very young children are extremely sensitive to the distress and anxiety of their family. They sometimes have a much greater level of awareness than is recognised by their families or health workers. However, failing to acknowledge to a child the reality of their position may intensify a child's feelings of isolation by denying them the opportunity to express how they feel or to ask questions.

Time available may be short in busy settings (another reason for encouraging family involvement), so it is important to make the most effective use of time with a child. The way you approach, talk to and touch a child when providing care can make a big difference to the way they feel. It helps to build trust and influences compliance. Touch is a powerful tool to convey comfort and reassurance. Having their basic personal needs attended to 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.

Personal hygiene and protection of privacy and dignity

Personal care 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 given in a manner that protects the dignity of the child. Even very small children can 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 encouraging the family to be involved, as well as helping to fulfil the natural wish of many parents to participate in this way.

When washing the patient, extra attention should be paid to skin folds, the neck, the back, the ears and the genitals, and the child should be encouraged to do as much as they are able for themselves. Children who are malnourished, who have been sick for a long time or who have malignancies can have fragile skin that easily breaks down and requires special

attention. An effective method that can be used by both nurses and family carers is to gently change the patient's position at frequent intervals. This relieves the pressure on parts of the body and prevents reddening and breakdown of the skin. Pain relief should be given to prevent discomfort (see Section 9 Handbook 1) and, when possible, pressure-relieving measures are also helpful.

Good mouth care is very important, and the child 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, and help needs to be given to those who are unable to do this. For children who are very sick or who are dying, mouth care can help to prevent many problems, including bad breath, bleeding, infection, ulceration and pain, that can significantly add to their suffering. For these children the following measures can be helpful:

- using a soft toothbrush or mouth sponge to clean the child's mouth regularly
- using wet mouth swabs if the child's oral intake is low
- using lip balm for dry lips.

Hydration and feeding

Children can quickly become dehydrated, particularly if they suffer from diarrhoea, vomiting and/or fever, or when they are too tired and lethargic to drink. Any caregiver should be able to recognise the observable signs of dehydration (for signs and symptoms, see Sections 60 and 61 Handbook 1).

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

- To 1 litre of boiled water that has been allowed to cool add 6 level teaspoons of sugar and half a level teaspoon of salt (the solution should taste no saltier than tears).
- Or Dhal water, rice water, bean broth, fruit juices and thin porridge cereal are also helpful
- Breastfeeding mothers should be advised to breastfeed their child more often during episodes of diarrhoea.

Trying to persuade a child, particularly a sick young child, to drink is not always easy:

- Encourage the child to drink small amounts and often.
- Give them an age-appropriate explanation of why this is important.
- Involve the child and decide together with them how much they will try to drink each hour.
- A child under 2 years of age needs to drink between a half and a quarter of a 250-mL cup for every watery stool.
- A child over 2 years of age needs to drink between a half and one full 250-mL cup for every watery stool.

For small children or those who are too tired to drink by themselves, the following measures can be helpful:

- Use a small cup, spoon or syringe.
- Encourage the child to play, or to participate by using the syringe him- or herself.
- Give small rewards for drinking.
- Give praise when fluid is taken.

Children often lose their appetite when they are ill. In short acute episodes, the main priority is fluid intake and the replacement of salts. However, in longer periods of illness it is essential to ensure an adequate nutritional intake.

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

Feeding difficulties often cause distress to families. The parents of a child who is dying may feel that a lack of nutrition will contribute to or hasten their death. These anxieties should be understood. It should also be explained to the parents 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 influence oral intake, such as a sore mouth, nausea, vomiting and constipation.

Try the following:

- Provide familiar food for the patient and let them choose the food if possible.
- Encourage children to feed themselves if they can do so and encourage the parents to help.
- Avoid performing invasive procedures immediately before or during a meal.
- Try giving small amounts of food often, rather than two or three large meals a day.
- Avoid giving the child highly spiced or strong-smelling foods unless such foods are the cultural norm for them.
- Give food at familiar times for the child and try to make mealtimes fun.
- Praise the child when they eat, but do not criticise or punish them when they cannot.

Where possible, keep an accurate written record of all the patient's fluid and dietary intake, and also their output (urine, stool and vomit) (for an example of a chart, see *Figure 65.6 and Figures 9.2 and 9.4 International Maternal & Child Health Care: A practical manual for hospitals worldwide*. MCAI 2015. https://12dec1c3-e3f4-e784-7f3a-6e7903610962.filesusr.com/ugd/dd2ba4_c5284b1b150f4d70a72fe1cd710d530d.pdf Accessed 30.03.2021).

Compare the total intake and total output over a given time period (usually 24 hours) and add an amount for insensible losses through perspiration and breathing (approximately 15 mL/kg/day, or more if the child has fever or is in a hot environment). Alongside clinical observations, this will give an indication of the patient's hydration level and will also give warning of a patient who is becoming dehydrated. Written records are often inaccurate and frequent weighing (for example once each day at the same time, wearing clothes of similar weight) can provide a valuable guide to fluid balance.

Elimination

For the management of constipation and diarrhoea, see Section 17 in this Handbook and 60 and 61 in *Paediatric Handbook 1*

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

- Maintain an adequate level of hydration.
- Obtain information about the child's normal elimination pattern.
- Children may be too frightened to go to the toilet in a strange place.
- Pain (for example from a urinary tract infection, anal fissure or post-delivery in pregnant adolescent girls) 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 salty water can help to ease the pain and encourage urination or defecation.
- A child who is passing bloodstained stools may be frightened and need reassurance.
- Praise and encouragement are important and effective.

Further reading and resources

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Section 4 Nursing care for adolescents including those who are pregnant

Introduction

Adolescents are not small adults. Between the ages of 10 and 19 years their needs are complex as they make the difficult transition from dependency to responsible and independent adulthood. They are feeling their way towards this independence but at the same time may not feel confident, even if they are reluctant to admit this. When they are ill enough to require hospital care, frequently because of pregnancy and delivery, these pressures are even more marked.

Changes in adolescence that are relevant to hospital care

Sexual risk in adolescence

Adolescents, particularly girls, are vulnerable to sexual exploitation, especially if they are living in poverty. They may not know how to negotiate whether to have sex or not, or how to protect themselves from sexually transmitted infections (STIs) and pregnancy. They may have no caring adult to turn to. There are many stories of adolescent girls being coerced into unwanted sexual activity by those in positions of responsibility. They may be promised high school marks, or clothing, or money they need to help their families. They may fear reporting this, feeling that they will be shamed or not believed. They may be raped by men whom they know or to whom they are related, or by strangers, particularly in times of conflict and displacement. They may be endangered when rape is seen as bringing shame on the family. It may be difficult or impossible to prevent pregnancy, or they may not know how to do so. They may fail to report a pregnancy until it is too late for much of the antenatal care that is available. They may attempt to abort the fetus with extreme risk to their own life because they do not know what else to do. At the very least, these young women are vulnerable to STIs, including HIV and human papilloma virus (HPV) (the latter being a cause of cervical cancer). Such diseases (see Section 36) may be silent, or these young women may not know what to do or who to tell about symptoms that they experience. There is a high risk that pelvic inflammatory disease will result in infertility.

Pregnancy and childbirth

For an adolescent girl, pregnancy itself brings additional threats. She may not yet have completed her own growth and physical development. Her pelvis may not be fully developed, and the risk of obstructed labour and its consequences, namely fetal damage or death, maternal death, or fistulae, is greater than for older women. She needs good nutrition for her own growth but must provide for the fetus, and then for lactation, at the same time. The potential for malnutrition is high unless she has access to all the nutrients she requires. For this reason, it is important that her nutrition status is monitored during pregnancy. She should also be given advice, help and micronutrient supplementation. Other health dangers await pregnant adolescent girls. They are at greater risk of developing anaemia and pregnancy-induced hypertension, including pre-eclampsia and eclampsia. Stillbirth and neonatal mortality rates are higher in this age group, and babies are often of low birth weight.

These girls may also have undergone female genital cutting (FGC) (see Obstetric Handbook Section 29+), with the many and lifelong health problems this may cause, and even social exclusion because of resulting fistulae or death from obstructed labour.

https://12dec1c3-e3f4-e784-7f3a-6e7903610962.filesusr.com/ugd/dd2ba4_3f90a11b6ec844cb9969c7c75e7f9bab.pdf

Accessed 18th April 2021

Overall, complications of pregnancy and childbirth are the leading cause of death among female adolescents according to the World Health Organization (WHO).

Despite the dangers, many girls achieve a successful pregnancy outcome and become competent mothers. They may in fact be ambivalent about pregnancy, welcome or have planned it, especially if they see it as giving meaning to life or increasing their status. They may of course have to do this without the support of the baby's father or even their parents and will have their education disrupted or even ended. They will often have to bring up the child in poverty, perhaps also being excluded socially. Others are married at a very young age, not necessarily willingly. Any adolescent mother (and father) will need help with parenting and the development of life skills to prevent their circumstances from spiralling downward. Family planning advice will be very important to enable these young women to avoid further pregnancies too soon, which would add to the downward spiral.

Mental health problems

Mental health problems are not confined to adults. Apart from having psychiatric disorders such as schizophrenia, young people may be depressed and confused, bullied, abused emotionally and psychologically as well as physically, and must take part in activities that cause great stress. They may be forced to become child soldiers, they may have to take responsibility for other children in their family and even care for adult relatives such as parents with AIDS, and they may experience personal loss. An unhappy home life may lead to self-harming, to life-threatening eating disorders and to attempted suicide, any of which may result in hospital admission.

Malnutrition

It is not just in pregnancy that young people become malnourished. Malnutrition may occur because of poverty, or lack of access to healthy foods, or because of peer pressure and habit. This has substantial implications for their growth and development, their general health in the future, and their ability to recover from adolescent illness.

Implications for hospital care

Hospital care for adolescents is particularly difficult. Providers need to be sensitive to the needs and fears of these young people and avoid being judgmental. A bad experience or poor-quality services may lead to adolescents failing to return, and possibly spreading the word to their friends. For a girl to have to attend an antenatal clinic alongside older and obviously married women is hard. To have to sit in a family planning clinic alongside women from her own community, and to have to tell a nurse (who may be a relative or who may know her parents) that she is sexually active is even harder. Young men will find it equally difficult to attend for contraceptive advice, and for adolescents of both sexes, attending an STI clinic could be an ordeal that they do not wish to repeat. At the same time, the pressures on young people are often so great that bad experiences are not effective deterrents to risky behaviours.

Dignity, privacy, and confidentiality

Hospital care can be an experience of dignity and acceptance, or of rejection and embarrassment. Adolescents are unlikely to have special areas or wards except in the most well-resourced units. However, arrangement is needed to ensure that they are not nursed in the same space as very much older people, or with young children, and certainly not in mixed-sex areas. Facilities for privacy are needed, such as curtains around beds and inter-view spaces, and some arrangement should be made to ensure that interviews and conversations cannot be easily overheard.

Preservation of dignity, privacy and confidentiality is as important for adolescents as it is for adults. Going through puberty can make adolescents particularly self-conscious and even traumatised when examinations, such as those of their genitals, are performed insensitively or roughly. This may be especially difficult for young women (or men) who have experienced violence or sexual assault. Adolescent males may also feel great shame that an assault has happened and that they have been unable to prevent it.

Therefore, every effort needs to be made by care providers, from healthcare professionals to cleaners and porters, to maintain the privacy, dignity and confidentiality of these young people.

Confidentiality can be a particular issue when patients are minors, under the age of consent for the country in which they live. Parents or guardians may need to be given information that young people would prefer was not shared. There is no clear answer about how much those responsible should be told if the adolescent indicates that information should be withheld. Carers will need to be guided by their ethical codes and make decisions according to the best interests of each individual young person, as well as to the prevailing laws.

Parents or guardians may have legal powers to decide what treatment adolescents should receive, such as giving consent for surgery, but every effort needs to be made to take the young person's wishes and views into account. In some countries, even a court of law will take a child's wishes into account as they near the age at which they have the right to make decisions.

Services for young people with a disability

Care providers need to be particularly vigilant in providing services for young people living with disabilities, especially mental or learning disabilities. These people are vulnerable to abuse, to finding services inaccessible or inappropriate, and to misunderstanding of their needs. When they are away from their normal environment, as when in hospital, they may become withdrawn, confused and possibly uncooperative. Ensuring that young people with a disability feel secure and well treated is the responsibility of all grades of staff.

Emergency services

Emergency services for young people should not be neglected. Young women may need gynaecological assistance, for example, for the consequences of female genital cutting or sexual assault. They may need access to post-exposure prophylaxis for HIV, or to emergency contraception where this is acceptable, or to post-abortion care. Mental health problems may be acute and arise as emergencies, especially following major emotional stresses, when illnesses such as schizophrenia first arise, or when young people self-abuse or attempt suicide.

Building relationships

Trust is of primary importance for young people. Services that consider the need to build trusting relationships are likely to be more acceptable, and therefore more effective and better used. Continuity is important, so that adolescents do not see a new face every time they attend for healthcare or counselling, but instead can develop a relationship of trust. This is a significant management issue, but it is important in terms of service uptake, effective use of human and other resources, and overall effectiveness of treatment programmes.

Advocacy

Healthcare providers have a strong advocacy role for adolescents, particularly in the context of working together, as for example in professional associations. This may involve campaigning for better services, talking to colleagues about their behaviours, or providing professional development opportunities. It may involve listening to young people, working with them, or campaigning against harmful local practices such as female genital cutting, early and/or forced marriage, ritual sexual initiation by older men, and erroneous and damaging beliefs (for example that having sex with young girls will cure HIV).

Hospital care summarised

The WHO suggests that services need to be accessible, acceptable, equitable, appropriate, and effective. This means that young people need:

1. appropriate, acceptable, accessible and gender-sensitive hospital and community services
2. carers who have approachable, accepting, and non-judgmental attitudes
3. confidentiality, preservation of dignity and privacy
4. accurate and honest information
5. choice and some control over what happens to them
6. avoidance of inappropriate hospital inpatient facilities where possible
7. targeted health promotion
8. targeted services, for example: for young people with long-term physical disability or illness; for young people with learning and developmental disability; for family planning; for STIs and HIV; for pregnant adolescents, including post-abortion care, emergency contraception (if permitted), ante-natal, labour and postnatal services, and nutrition services for young mothers, whether supported or not, and for young fathers, for displaced adolescents (e.g. refugees, internally displaced persons, the homeless) emergency health and counselling services for support in crisis
9. advocates who understand their needs and can support them both as a group and individually (for example with families).

Finally, nurses, midwives and doctors may be the only people whom adolescents feel able to talk to, especially as others, such as parents, teachers and religious leaders, may be seen as authority figures. This vital role goes way beyond simply providing medical, surgical or obstetric care.

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Natalia Kanem, executive director of the UN Population Fund, is among experts warning about disrupted health services and a surge in gender-based violence. Sophie Cousins reports.

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Section 5 Managing the child with a disability in hospital

Introduction

Around 10% of children in most resource limited countries are disabled in some way. All children who are hospitalised for illness or injury are at risk of becoming disabled due to their condition and/or their management.

Disabled children: definitions

Disorder is a medically definable condition or disease.

Impairment is the loss or abnormality of physiological, anatomical or psychological structure or function.

Disability is any restriction, due to an impairment, in the child's ability to perform an activity in the normal way for a child of that age.

Handicap is the impact of the impairment or disability on the person's pursuits or achievement of goals that are desired by him or her or expected of him or her by society

Special needs refers to children who have needs greater than the normal needs of children of their age.

In 2001, the WHO introduced a new system of classification, The International Classification of Functioning, Disability and Health (commonly known as ICF)

<https://www.who.int/standards/classifications/international-classification-of-functioning-disability-and-health> Accessed 30.03.2021 which uses two lists; a list of body functions and structure, and a list of domains of activity and participation. Since an individual's functioning and disability occur in a context, the ICF also includes a list of environmental factors. The ICF replaces the previous classification based on 'impairment', 'disability' and 'handicap', and shifts the emphasis to functioning. The ICF puts the notions of 'health' and 'disability' in a new light. It acknowledges that every human being, through illness or injury, may develop a disability. It 'mainstreams' the experience of disability and recognises it as a universal human experience. Furthermore, the ICF considers the social aspects of disability and does not see disability only as a 'medical' or 'biological' dysfunction, but includes contextual factors, such as environmental factors. The ICF encourages assessment of the impact of the environment on the person's functioning. For example, hospital staff should be vigilant about preventing the development of contractures in a comatose child because they are concerned to ensure the best functioning of the child when he or she recovers.

Children's rights

Article 23 of the UN Convention on the Rights of the Child <https://www.unicef.org.uk/what-we-do/un-convention-child-rights> Accessed 30.03.2021 defines the right of disabled children to special care, education and training designed to help them to 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 findings 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 resource-limited countries have a greater degree of acceptance of disabled people than is found in more well-resourced countries. However, some cultures regard disability as a punishment or as a cause of shame. Accepting, encouraging and supportive behaviour of healthcare providers towards children with disabilities will go some way towards dispelling these attitudes.

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.

The aims of a service for disabled children

These are as follows:

- to provide health services which ensure that children reach their maximum potential, optimising their independence and ability to lead a high-quality life
- to ensure that disabilities are promptly identified and treated where possible
- to promote active involvement of disabled children and their families in all aspects of healthcare, working in partnership with healthcare professionals
- to promote access to the healthcare facility for families with disabled children
- to provide comprehensive, integrated and coordinated services both in the healthcare facility and in the community, utilising outreach, and community-based services, including community-based healthcare workers
- to enable health services to work with other key agencies, such as social services and education and training services.

The prevention of impairment and disability in children

This is the main issue in resource-limited 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 Neonatal Handbook), the prevention of cerebral oedema due to inappropriate fluid management (see Sections 66 and 73 Paediatric Handbook 1), the prevention of hypoxic-ischaemic cerebral injury (see Obstetric Handbook), provision of adequate nutrition, the avoidance of accidents and protection from abuse (see Section 2). Adequate immunisation to combat poliomyelitis, measles, malaria, TB and meningitis, which are major causes of disability, is also mandatory.

Antenatal, peripartum, infant and childcare

1. 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. This will involve promotion of early attendance at antenatal clinics. Detection and treatment of diseases such as syphilis and malaria, which can cause intrauterine growth retardation (IUGR) and prematurity, management of HIV and prevention of mother-to-child transmission, and the detection and management of intestinal worms (Section 44) and nutritional deficiencies are all essential.
2. Every attempt should be made to provide good nutrition for adolescent girls and women who may become pregnant. Folic acid supplements at the time of conception are vital for preventing spina bifida and other congenital abnormalities.
3. Iodination of salt is inexpensive and should be universal.
4. Iron-deficiency anaemia during pregnancy is associated with low-birth-weight 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 or, if contracted, be treated vigorously (see Obstetric Handbook). Ministers of Health should be persuaded of the value of providing malaria prophylaxis or intermittent preventive treatment (IPT) for all pregnant women in endemic areas.
5. Immunisation against tetanus (see Neonatal Handbook) is essential.
6. Obstetric care within hospitals should aim to prevent impairments due to complications of labour and delivery. Crucial to this is the availability of oxygen, obstetric surgery and anaesthesia, and an effective blood transfusion service.
7. Effective neonatal resuscitation should be available 24 hours a day in every maternity unit and for all home deliveries. Staff must be trained and should have the basic equipment (see Neonatal Handbook) necessary to prevent those causes of birth asphyxia which arise after the delivery of the baby.

8. Simple interventions such as not bathing immediately after birth, prevention of hypothermia, and 'skin to skin care' for low-birth-weight babies should be taught to village health workers and traditional birth attendants in regions where they play an important role in home deliveries.
9. Recognition of danger signs and the setting up of community-based referral systems to deal with emergencies should be implemented at village level.
10. Breastfeeding must be encouraged (see Neonatal Handbook), and special support must be given to help mothers provide breast milk for babies with developmental impairments that make sucking or attachment difficult.
11. Adequate training and facilities for the correct management of dehydration in gastroenteritis (see Section 60 and 61 Handbook 1), hypoxic-ischaemic injury (e.g., in major trauma) (see Section 81 Handbook 1) and severe anaemia from malaria (see Section 31 Handbook 1) all reduce the frequency of preventable brain damage.
12. Paediatricians in hospitals should advocate for programmes of injury prevention and the prevention of injuries to children resulting from conflict, displacement, or other social factors.

Management of disabled children: identification and primary diagnosis

- All babies should be systematically examined at birth and, if possible, also at 6 weeks of age to detect preventable disabilities such as dislocated hips and congenital cataracts.
- In regions where most babies continue to be born at home, community health workers (CHWs) should be trained to detect these problems or to encourage mothers to attend for postnatal checks at a clinic where these can be undertaken.
- Postnatal services should be established in all healthcare facilities that provide antenatal care and delivery services.
- Protocols for postnatal care should be developed based on WHO guidelines.
- Signs or symptoms of an emerging disability should be actively sought. Findings which suggest that the child may be disabled should be communicated to the parents in a culturally sensitive manner in accordance with locally developed guidelines. This communication must include information about the local availability of services and social support.

Comprehensive interdisciplinary assessment

- This should always include the child's strengths as well as their weaknesses, and an assessment of their home circumstances and educational needs.
- It should result in decisions about management, including any immediate surgical or medical treatments available to alleviate the condition.
- It should include an assessment of sensory, motor, behavioural and intellectual capabilities as outlined below.

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 if they are available.
- A care manager or key worker should be appointed, who will act as a liaison between professionals and the parents to ensure that the child fully benefits from the available resources.

There is a big mismatch between this ideal and the level of resourcing in many low resource settings. It may be more realistic to suggest that a named Community Health Worker be a key part of a discharge planning meeting.

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 their 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 that 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 the diagnosis and expected prognosis should be given by a senior clinician who has experience in this area and is aware of local attitudes and beliefs regarding disability and the services available to the child and their family.
- Services should be developed as resources allow.
- Policies about the intensity of resuscitative treatment given to children with various impairments should be developed by doctors, other healthcare professionals, representatives of the local community, including disabled people, and politicians. These policies must consider ethnic and cultural issues and local support available for the care of severely disabled children. Such policies must be reviewed frequently. A hospital ethics committee can be valuable in this respect (see Section 8).
- Development of services for and the rights of disabled people should be promoted wherever possible. Frontline staff should feel confident that they know and can work within the framework of the policy.

Diagnosis

All newborn babies should be examined before leaving hospital by a member of staff (usually a nurse or midwife, or a paediatrician if one is available), who has been trained to perform 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, namely family history, female gender, and breech presentation. Dislocated or dislocatable hips should be referred to an orthopaedic specialist.
- **Jaundice:** Any jaundice in the first 24 hours should be taken seriously and monitored appropriately. Causes of jaundice, such as blood group incompatibilities, glucose-6-phosphate dehydrogenase deficiency and sepsis, should be diagnosed and treated. Severe jaundice can lead to deafness and cerebral damage (see Neonatal Handbook).
- **Cardiovascular system:** This should be examined looking for cyanosis and equality of pulse volumes and listening to heart sounds. (see Section 40 handbook 1).
- **Hearing:** Behaviour should be observed, 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 Section 20).

Comprehensive assessment of disabled children

Most children in resource-limited countries are not born in hospital and therefore children with disabilities are more likely to present later in life.

History

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 for establishing the prevalence and distribution of various disabilities:

- 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 of 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 2-year-olds: 'Can he/she name at least one object (for example, an animal, a toy, a cup, a spoon)?' and 'Compared with other children of his/her age, does he/ she appear in any way to have difficulties in learning?'
- 10 For 3- to 9-year-olds: 'Is his/her speech in any way different from normal (not clear enough to be understood by people other than his/her immediate family)?'

Examination

A full clinical examination of all physical, sensory, and psychological systems should be undertaken.

Additional issues

- Determine how the child and their family have adapted their lives in response to the child's difficulties.
- Determine the extent to which the available treatment, training and management will improve the situation.
- Evaluate the emotional adjustment of the child and their family to the disability.
- Investigate the educational facilities available to the child and how they may be adapted to his/her needs.
- Determine the child's and family's strengths, abilities and positive personality traits which can be encouraged to help them to 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 that they can reach their maximum potential. For example, a protocol for a child with Down's syndrome could include the following:

- full medical examination
- chromosome studies (if facilities are available)
- ECG and chest X-ray with echocardiography (if available)
- development of a care plan with the parents/carers as partners
- audiological assessment
- visual assessment
- assessment by a speech therapist (if available) to promote communication skills
- assessment by an occupational therapist (if 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 (see Section 20)

Most newborn babies can focus on and follow the mother's face and large brightly coloured objects. 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 for neonates, toys for older infants, and pictures (whose dimensions correspond to Snellen letters) for older children.
- Determine whether visual impairment is an isolated problem or associated with other developmental defects (e.g. cerebral palsy) by undertaking a detailed history and physical examination.
- Check for the red reflex as follows. Shine a light on the pupil from arm's length. Normally it will appear red because of light reflected from the retina. If it appears white, consider the possibility of dense cataract, severe retinopathy of prematurity, or retinoblastoma. If the red reflex is normal, check the pupillary response to light. If the latter is normal, a local cause (i.e., optic nerve or retinal degeneration) is unlikely, and impaired vision is then most probably due to occipital lobe damage.
- Check the retina and optic nerve by fundoscopy to exclude optic atrophy and retinal degeneration. If in doubt, refer the child to an ophthalmologist. A low-cost adapter used with a mobile phone can be extremely helpful (please see the excellent images in the article referred to here:

<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4244748/> Accessed 30.03.2021

Causes of visual impairment

- Common causes of blindness in children include optic atrophy, congenital cataracts, and retinal degeneration, and in resource-limited countries they include vitamin A deficiency, measles, onchocerciasis and meningitis.
- Trachoma remains a major cause of blindness in many developing countries. Early detection and treatment can prevent blindness. Prevention activities should include hygiene. Mass drug administration in affected areas can be considered.
- Close liaison between paediatricians and ophthalmologists is required to develop policies to detect and treat visual defects as early as possible.

Management of impaired vision

Treatable causes:

- Cataract: Children with congenital cataract should be referred to an ophthalmologist as soon as possible for early treatment. If no treatment is available, the 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 Section 55 Handbook 1 for doses at different ages).
- Eye infections (see Section 20).

Community healthcare 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:

- Perform a visual assessment and provide suitable visual aids.
- Surgical correction of squints should be undertaken (when possible).
- Mobility training should be provided 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 healthcare workers are not vigilant. Because hearing defects often lead to lack of development of speech and language, the child is sometimes assumed to have learning difficulties and may be further isolated from their family and society because of this. All children who present with failure to develop language should have a good-quality hearing assessment.

Hearing is essential for language development, so early detection of hearing impairment is essential. A newborn infant responds to sudden noise with 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 or oto-acoustic emissions. The distraction test is used at 4 to 8 months of age, and audiometry is used in older children. *There are two types of hearing loss:*

1) Conductive hearing loss: The commonest cause is recurrent/chronic infective otitis media (see Section 37 Handbook 1).

2) Sensorineural hearing loss: The commonest causes are meningitis, cerebral malaria, genetic defects, drugs (e.g. excessive doses of aminoglycosides) and intrauterine infections. A hearing aid is required, and the child may need to learn a sign language.

Children with the following are at risk of hearing impairment:

- family history of sensorineural hearing loss
- dysmorphic features
- abnormalities of the pinnae
- severe birth asphyxia
- severe neonatal jaundice
- other neurological abnormalities
- postnatal infections (e.g., meningitis, measles)
- treatment with ototoxic drugs (e.g., gentamicin, streptomycin).

It is most important to identify and treat causes of conductive deafness, such as chronic otitis media (see Section 37 Handbook 1).

Treatable causes of sensorineural hearing loss are very rare.

Hospitals in association with community health services and education authorities should seek to develop services for early identification and prompt treatment of children with irreparable hearing problems. These should include 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 enhance functioning (e.g., information on prevention 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 Sections 6 and 49).
- Local people are often resourceful in developing appropriate equipment for their own children out of locally available materials (see *Disabled Village Children* referenced). Advice from occupational therapists and physiotherapists is very useful (if 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. Hospital staff, community health workers and family members should receive training on safe feeding techniques to improve the nutritional state of these children.
- Feeding may require the placement and management of a nasogastric tube, and parents or carers 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 also section on paraparesis and incontinence below.)

Where possible, neural tube defects should be prevented by adequate maternal nutrition, including folic acid and vitamins at the time of conception.

- Children born with neural tube defects should be treated urgently to prevent worsening of their condition (see Section 58 Handbook 1).
- 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 5.1 and 5.2)

Delayed development presupposes knowledge of normal development. Development proceeds in an orderly fashion, but there is considerable variation in the age at which milestones are achieved.

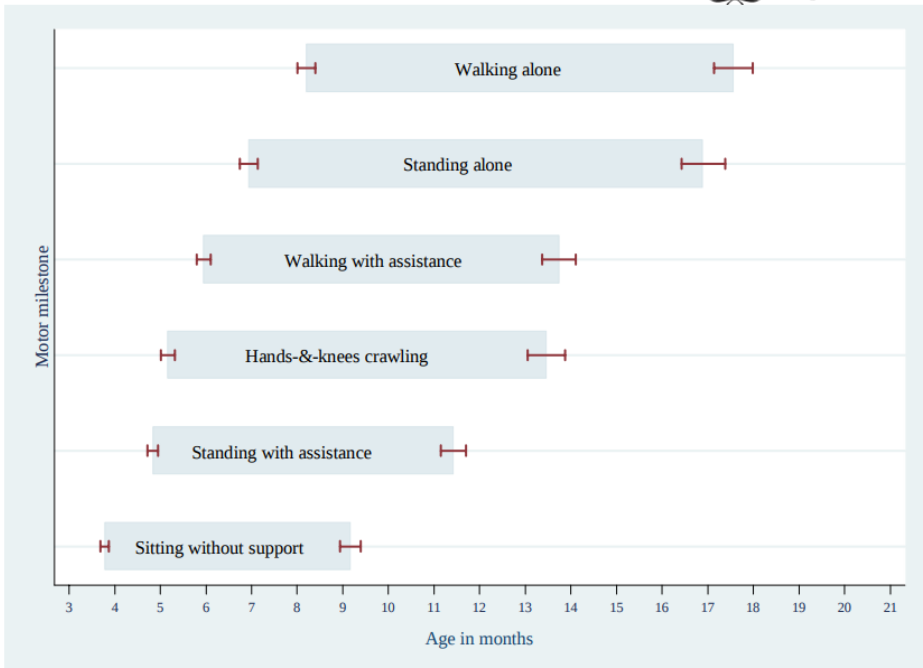
TABLE 5.1 Normal milestones in development

Age	Milestone
Birth	Focuses with eyes and responds to sound
4–6 weeks	Social smile
6–7 months	Sits without support, transfers objects from one hand to the other
9–10 months	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	Says ten words, feeds with spoon
2 years	Runs, draws straight line, says two-word sentences
3 years	Draws circle, draws cross, says three-word sentences, dresses in simple clothes without assistance
4 years	Stands on one leg, fluent speech <i>Note: in societies where access to pens and paper is very limited, even adults find drawing a line or a circle difficult as such an action is outside their experience</i>

TABLE 5.2 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 or toys, does not laugh or smile, has squint, hand preference, primitive reflexes still present
9–12 months	Not sitting, not saying 'baba' or '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 or other objects Passive – not moving about exploring, 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 commands
4 years	Unintelligible speech
At any age	Parental concern, regression of acquired skills

Windows of achievement for six gross motor milestones



Reference: WHO Multicentre Growth Reference Study Group. WHO Motor Development Study: Windows of achievement for six gross motor development milestones. Acta Paediatrica Supplement 2006;450:86-95.

https://cdn.who.int/media/docs/default-source/child-growth/child-growth-standards/indicators/motor-development-milestones/graph--windows-of-achievement-for-six-gross-motor-milestones04ac44c38d96466498e7633d9d44c7e6.pdf?sfvrsn=ea3a0241_0

Developmental assessment

The purpose of developmental assessment is threefold:

- 1 to confirm normal or delayed development
- 2 to identify possible causes of delayed development
- 3 to plan a strategy for intervention.

To achieve these aims, a detailed history and physical examination are essential. Emphasis is placed on perinatal and developmental history. As well as looking for signs and symptoms of severe malnutrition or micronutrient deficiencies, allowance must be made for prematurity. Evidence of microcephaly, dysmorphic features and signs of neglect must be looked for, and a detailed neurological examination, including primitive reflexes, undertaken. The following questions must be addressed:

- Does the child have global delay (i.e., delay in all areas of development)?
- Is the delay confined to one area of development? If it is confined to the motor area, this suggests a possible neuromuscular disorder. Delayed speech development with normal motor and social skills could suggest a hearing disorder.
- Has the child lost previously acquired skills, and if so, has the loss been progressive? This suggests a neuro-degenerative disorder.

Delayed walking (not walking by 18 months)

- Family history of late walking and otherwise normal: give reassurance.
- Global delay (especially in language and social skills): the child probably has mental impairment.
- Child failing to thrive and showing signs of malnutrition and poor nurture: this suggests neglect.
- Cerebral palsy with upper motor neuron signs (spasticity, clonus, brisk reflexes) or dystonia, ataxia, and involuntary movements.
- Neuromuscular disorders (see Sections 24 and 25) with flaccid weakness, wasting or fasciculation of muscle, absent or diminished reflexes.
- Congenital dislocation of the hips or rickets can cause delayed walking.

Delayed language development

For meaningful speech to develop, the infant must be able to hear, and have intact language pathways and normal oropharyngeal structures. The child must also receive verbal communication.

The following approach to 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 or defect of the mouth and pharynx (obvious on physical examination)?

If the child cannot communicate and has normal intelligence, they will try to compensate by using gestures and/ or signs. They are also likely to be frustrated and angry. The child whose language delay is part of a general learning difficulty is likely to be more passive and less frustrated.

Does the child have a problem with social interaction? Consider autism, signs of which include loss of social interaction, little or no non-verbal communication, no eye contact, and repetitive ritualistic behaviour.

Cerebral palsy (see Table 5.3)

Cerebral palsy refers to the disturbance of movement and/or posture that results from a non-progressive lesion of the developing brain. The commonest causes are hypoxic-ischaemic insult to the brain occurring prenatally or perinatally, or occasionally postnatally (e.g., meningitis, head injury). There are several different types of cerebral palsy, including the following:

- spastic diplegia (common with prematurity)
- spastic quadriplegia and spastic hemiplegia
- dyskinetic 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 concerning the motor system (this is best performed by a physiotherapist) and identify associated problems.

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 the parents to move and handle the child in their daily activities to improve mobility and aim to prevent contractures. Parents need support in ensuring both that the educational needs of the child are met, and that the child is integrated as fully as possible into society.

Deterioration in children with cerebral palsy

Children with cerebral palsy usually remain stable. If a child shows apparent deterioration consider the following possible causes:

- pain from dislocation of the 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 examples following trauma, infection or malignancy). Some causes are treatable if diagnosed early (for example TB of the spine). Burkitt’s lymphoma with paraparesis is a sign of advanced disease and is often associated with a poor prognosis. Both thorough clinical assessments to establish the level of the lesion, and reassessment to look for changes, are essential. Any suspected space-occupying lesion needs surgical advice.

TABLE 5.3 Problems in children with cerebral palsy

Problem	Action
Visual and hearing impairment	Refer to appropriate specialist
Epilepsy	Anti-epileptic drugs
Contractures	Physiotherapy and (rarely) surgery

Problem	Action
Dislocation of hips	Surgery to relieve pain
Feeding difficulties, failure to thrive	Monitor intake Correct positioning for feeding Increase energy content of food Consider gastro-oesophageal reflux
Recurrent aspiration	Close attention to feeding position, pacing of feeds, and positioning
Respiratory infections	Antibiotics
Gastro-oesophageal reflux	Feed thickener (starch), proton- pump inhibitor (e.g. omeprazole) Exclude oesophageal stricture and/ or aspiration
Constipation	Diet and stool softeners
Learning difficulties	Additional help with education

Many children with paraparesis will suffer preventable complications unless carers and staff are aware of the risks of the following:

- Poor nutrition: many children with paraparesis find it difficult to eat and drink. They need good food to enable them 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: these are prevented by ensuring that the child is moved regularly. The child can often learn to do this by using their 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. They must therefore 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, empty incompletely and may lead to reflux nephropathy, 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 or 4 hours. This technique can be easily learned by a carer or by the older child. (See Section 58)

Other children have bladders that are not full, and which 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 their relative immobility. A healthy 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 that is passed up the rectum for a washout of the bowel contents with saline.

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 (e.g. 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 referred to as mental retardation) are suspected when there is global developmental delay especially in language, social and fine motor skills. Gross motor milestones may be normal. Causes include fetal alcohol syndrome, hypoxic-ischaemic injury to the brain, Down's syndrome, fragile X syndrome and neurocutaneous syndromes, among others. Treatable causes should be excluded (for examples hypothyroidism, phenylketonuria).

The parents will need considerable support in coming to terms with the diagnosis and its implications. They should be encouraged to stimulate the child's 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 supported in special schools (if available). Their progress must be continuously monitored, and associated problems dealt with. They deserve the same care as normal children.

Autism and communication disorders

- Autism usually presents in the second or third year of life.
- It is primarily a communication disorder associated with an absence of or disordered speech and language development.
- It is often associated with obsessional behaviours or interests.
- It may or may not be associated with mental retardation.
- It is often associated with learning difficulties because of inability to understand social situations.

The following should arouse the suspicion of autism: no babbling by the age of 1 year, no pointing by the age of 1 year, no single-word utterances by 16 months, no spontaneous two-word utterances by 24 months, and any regression in social skills and language.

Developmental coordination disorder

About 5% of children have difficulties with coordination which may affect their ability to perform motor tasks such as writing or sport. It is important to exclude a serious neurological cause and identify that the child cannot do these activities well, so that teachers and others do not conclude that the child is 'lazy'.

Attention deficit disorder

This is a major problem associated with the following:

- difficulty with concentration
- impulsivity
- difficulty in predicting the outcome of actions, so the child does not learn from their mistakes

- a strong association with hyperkinesia/hyperactivity
- poor listening skills.

Attention deficit disorder improves with maturity.

Treatment is difficult. The most important points are to recognise the disorder, explain it to the parents and provide them with family and/or other support to cope with it. Stimulants, such as methylphenidate, used by an experienced health worker, may be very helpful.

Behaviour disorders

- Exclude attention deficit disorder and other developmental impairments.
- Try to exclude child abuse, although a behaviour disorder may coexist with abuse (see Section 2).

Psychiatric disorders (see Section 22)

- These are rare in young children.
- Severe malnutrition, deprivation and abuse can lead to depression and the signs of frozen awareness/watchfulness (see Section 2 on child abuse).

Surgically treated disabilities such as hare lip and cleft palate are addressed in the section on surgical problems (see Section 15 Neonatal Handbook).

Transition to adulthood for children with disabilities: a human rights perspective

The 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 at higher risk of abuse and exploitation and are likely to be more vulnerable as they pass 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 adulthood.

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 of young men and young women, and deep-seated prejudices and cultural taboos that cause further disablement and devaluing of young disabled people unless the human rights perspective is paramount.

Child-friendly and child-safe environments

- All buildings that are used as healthcare 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 needed ramps should also be provided.
- If the building has several floors, lifts should (when possible) be in place. If this is not possible, clinics serving disabled people should be located 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.

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 resource-limited communities are expected to work on the land or in industry, to look after livestock or to take responsibility for the care of their younger siblings at an age 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.

Disabled children may have similar experiences or worse (for example being used as beggars), which deprive 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, this is not the sort of independence that young people need.

At the other extreme, disabled children worldwide are often overprotected by their families, who may feel ashamed, or there may be cultural taboos and beliefs about the origins of conditions. The parents may wish to do everything for their disabled child, but this can result in the child not learning from experience. The result may be that the disabled young person does not get the opportunities for education and training that would 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.

Enabling the disabled child to become an integrated member of adult society is a challenge that requires the following:

- imagination, resources, and flexibility on the part of the health, education and social services
- active engagement with the young person, their family, and their community
- a real commitment to working with the strengths of the young person and minimising their weaknesses by reducing the barriers to their 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 commonly 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 healthcare 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 can understand the facts about their own bodies. Even when there are some physical problems that will affect sexual experience (e.g., 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 adulthood

Healthcare facilities that provide services for children with disabilities should develop expertise in enabling children to make the transition to adulthood. This expertise is likely to be achieved by developing shared knowledge among a group of relevant professionals working in partnership with young people. The service should be able to offer the following:

- information that 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 (1987) *Disabled Village Children: a guide for community health workers, rehabilitation workers and families*. Palo Alto, CA: Hesperian Foundation.

Section 6 Facilities in hospital for children with special needs and learning difficulties

Introduction

The most valuable asset is healthcare staff who can spend time with these children and their families, and preferably who are also able to visit them at home.

For children presenting at the hospital with established disabilities, the challenge is to ensure that they make the best use of their abilities and do not develop further disabilities.



FIGURE 6.1 Lying aids.

TABLE 6.1 Equipment for assessment

Equipment for physical examination	Equipment for skills assessment
Tape measure	20 brightly coloured wooden cubes, 2.5 cm in diameter
Auroscope	Threading beads of various sizes, and string
Ophthalmoscope	20 culturally appropriate pictures of common domestic objects, some of which have similar sounding names in the local language
Tendon hammer	Soft ball, approximately 10 cm in diameter
128-Hz tuning fork	Denver Developmental Screening Test
Simple audiometer	

Aids for disabled children

This section outlines the types of aids that should be available for disabled children in all hospitals. 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.

David Werner’s book, *Disabled Village Children*, is an excellent source of ideas and advice.

General principles

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- ‘*Look first at my strengths and not at my weaknesses*’: the preservation of best function is primarily achieved by education of the child and their 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 (for example in some regions 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 that you think will help, and ask yourself the following questions:
 - How will this aid help this child to function in their 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 who are recovering from illness spend most of their time lying on their back or on their side. 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 such as stiff foam plastic. Some children who need to have their legs separated because of adductor spasm will need a leg separator or pillow, also made of similar material.

Sitting aids

The type of sitting aids used will depend on the difficulties and developmental stage of the child. Most children with cerebral palsy benefit from being seated in a position in which their ankles, feet and hips are at 90 degrees and their 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 (for example a ‘steam engine’).

Children with spasticity also often benefit from a slight tilt backwards. The position and amount of head support needed depend on the amount of head control and extensor tone.

Standing aids

These may be useful for children who are showing improvement in their motor skills and can be expected to learn to stand independently but are also useful for children who may never stand independently, because the standing position aids the circulation and bone growth and strength, particularly of the hip joints. Some children find standing frames difficult to get used to at first and may need encouragement to use them.

Walking aids

There are a wide variety of these aids available. Perhaps the most useful is a walking frame that goes behind the child, and which can have a variety of attachments depending on the child’s balance and arm strength. Some parallel bars are also useful and will need to be set at different heights depending on the size of the child.

Section 6. Facilities in hospital for children with special needs and learning difficulties.

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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.

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 their buttocks as a result of spinal cord damage, they will be at risk of developing pressure sores if they remain seated in the same position for long periods of time. They can learn ways of taking the pressure of their buttocks. If pressure sores have developed, getting around the hospital may be better using a gurney.

Eating and drinking aids

Utensils with thick handles and cups with handles on both sides may be easier to use for children who find gripping difficult. It may be helpful to put a non-slip material underneath a bowl or plate to stop it sliding while the child is eating (a damp cloth works very well). Eating and drinking aids must be easy to wash. Assessments by occupational and speech and language therapists (if available) are invaluable for children with complex feeding difficulties.

Toileting aids

For details, see Section 58

Communication aids

Children who are unable to communicate verbally because of deafness and/or inability to use their oro-motor 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 in children with weak legs is the development of foot drop. This should not happen in your hospital. Regular exercises to move the ankles through their full range of movement should be done at least twice a day. The use of tight or heavy bed covers should be avoided, as they may hold weak feet in a bad position. It is best for the feet to rest with the ankles at 90 degrees. This is easily ensured by positioning a roll of blanket or similar material so that the feet are braced in this position.

Preventing knee and hip contractures

Regular exercises that take 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 their hips and knees extended.

Preventing scoliosis

This is achieved by symmetrical positioning of the child so that attention must be paid to both lying and sitting positions. With excessive and asymmetrical muscle tone it is often

Section 6. Facilities in hospital for children with special needs and learning difficulties.

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difficult to prevent scoliosis, and once it has developed, it often gets worse, particularly 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 develop more quickly if the circulation is poor. There is no substitute for good nutrition and regular moving and turning of the child. The skin should regularly be gently cleaned, and dried, and moisturising lotion used. Prevent bony areas from pressing on each other and on the mattress by using pillows or foam wedges, for example, between the knees or under the heels (see Section 58).

Further reading

Werner D (1999) *Disabled Village Children: a guide for community health workers, rehabilitation workers and families*. Palo Alto, CA: Hesperian Foundation.

A 2018 version can be found here but costs £27

https://hesperian.org/wp-content/uploads/pdf/en_dvc_2018/en_dvc_2018_fm.pdf

Accessed April 1st 2021

Section 7 Palliative care for children in communities in resource-limited settings

Introduction

Most children who need palliative care in resource-limited countries will require identification and treatment in the community rather than in hospital. Moreover, in the presence of effective care and support networks, home has frequently been demonstrated to be the best setting for palliative care for both the child and the family. These include children with cancer (post or during chemotherapy), HIV/AIDS, neurological conditions, and other Non-Communicable Diseases. Many patients are in the community and in Uganda are referred by Community Volunteer Workers (CVWs), trained at the Hospice.

In LMIC's there are often local solutions, to many challenges, to improve independence and quality of life. These are often cheaper, available, and more appropriate than expensive western recommended aids. They should not be rejected if suitable to the individual patient.

A high proportion of children do not reach hospitals in Africa. This percentage ranges from approximately 57% in Uganda to 85% in Ethiopia. Also, hospitals need to be aware that most families would wish for their child to die at home, where they can look after them and they can be buried with their ancestors near to the home. The cost of transporting a body is very high, so economic factors also play a part.

Therefore, any treatment that is given in the hospital must be of a kind that can be continued at home, otherwise the child will never be able to leave the hospital. Healthcare workers in hospital, with the support of Ministries of Health and community leaders, must set up systems to help community health workers to provide care in the community, including the safe management of morphine treatment when it is required.

Allowing the family and child to choose the setting for palliative care is of great importance. However, it is recognised that the necessary resources may be minimal or absent in many locations, and local conditions will determine what options are available.

WHO definition of palliative care

Palliative care is an approach that improves the quality of life of patients and their families facing the problem associated with life-threatening illness, through the prevention and relief of suffering by means of early identification and impeccable assessment and treatment of pain and other problems, physical, psychosocial, and spiritual.

Palliative care:

- Palliative care for children is the active total care of the child's body, mind and spirit, and also involves giving support to the family.
- It begins when illness is diagnosed and continues regardless of whether a child receives treatment directed at the disease.
- Health providers must evaluate and alleviate a child's physical, psychological, and social distress.
- Effective palliative care requires a broad multidisciplinary approach that includes the family and makes use of available community resources; it can be successfully implemented even if resources are limited.
- It can be provided in tertiary care facilities, in community health centres and even in children's homes.

This section describes the use of affordable medications that have been proven to work in resource-poor settings. Here, it is vital that government funds are spent carefully on measures which work and are not too expensive thus ensuring that the poorest families can also receive their right to palliative care for their children.

Although palliative care can relieve symptoms during the care of all sicknesses at any stage of the illness, the term is often (mistakenly) associated with relieving 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 healthcare professionals and may evolve over a period of time. It is important, however, to state that even when the body cannot be cured, there is always something more that 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. The Caring Team, too, should be receiving support through what can be a distressing time.

Essential healthcare for the dying child

Include parents or familiar caregivers.

- Include parents or familiar caregivers.
- This matters always.
- Their familiar presence will comfort the child.
- Even apparently unconscious children may still know their parents' or caregivers' voices.
- Parents invariably want to be able to provide care for their child. This is a natural wish and can aid their own coping strategies.
- During the Covid epidemic, children in hospitals need a caring adult and they are usually allowed. However, siblings and other child visitors may not be encouraged.

Set realistic goals.

1. The art of end-of-life care is to know when both goal and treatment must change.
2. The goal is to help the child to enjoy and cope with what is left of their life.
3. It should be clearly and well communicated that resuscitation measures are not to be a feature at the end of life. Our aim now, is not to cure, and never to kill, but always to comfort and increase quality of life within the present condition of time and place.
4. The social needs and goals of a dying child include access to 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 imminent. This must be gently explained, and the parents' and child's questions answered. It is wise, especially with children, to clarify the real question that is being asked. Replies must be honest, but the truth should be shared sensitively, a little at a time.

- 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 results in greater trust and cooperation than saying something won't hurt when it will.

All of those 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 their family. Great comfort can be derived from acknowledging their expressions of anxiety, and this helps to dissipate the feelings of isolation that are frequently experienced.

Adolescents will also have particular concerns and worries, and often have spiritual needs as well. Spirituality is a major aspect of life even for younger children in Africa. All members of the team must be aware of this and ready to discuss it with them. We can learn so much from a child.

List and treat the child's symptoms.

In palliative care, symptom intervention and practical care are paramount.

- Even with limited resources, symptoms can often be helped. Problem lists are a useful key to active needs.
- The availability of drugs does not guarantee their skillful use, but when medication is used effectively it will make both life and death more bearable. It can be helpful to give the family a treatment chart showing times or relationship to sunrise and sunset (see Figure 7.3).
- The child and carer together should make a list of all the symptoms. This can guide palliation even when the cause is incurable.
- Ask the carer to chart extra doses required and any medication-related problems that arise.

The duration and nature of palliative care will be unique to each child and their particular disease. For those children who cannot be cured (sadly they are the majority in resource-limited settings), highly effective 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.

For palliative care issues concerning HIV infection, see Section 36.

Common symptoms

Pain see Section 9 Handbook 1 and below.

For pain assessment,

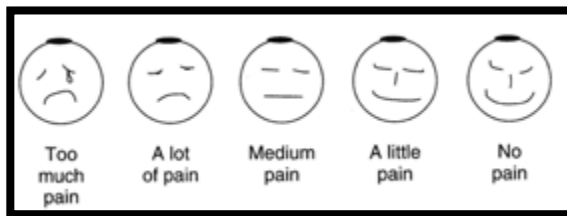
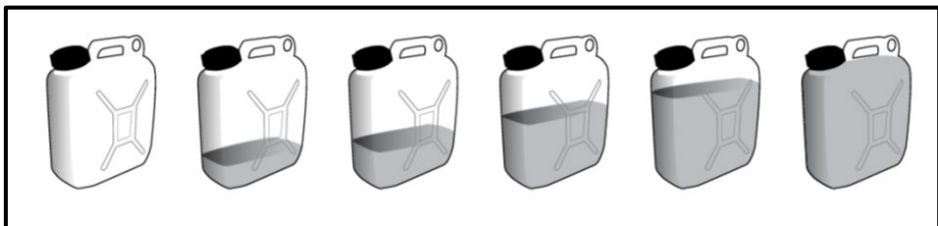


FIGURE 7.1 A commonly used faces pain scale for assessing the severity of pain in children.

NB When children have not learned to read or to look at pictures they see all these like pancakes without expression.

Jerrycan scales are used in Africa and other LMIC's. Children collect water in different sizes of jerrycan, from when they are small. They understand if empty or full.



0	1	2	3	4	5
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No pain	Mild pain	More pain	Moderate pain	Severe pain	Unbearable Overwhelming pain
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Figure 7.2 Jerrycan pain scale Blum et al. Health and Quality of Life Outcomes 2014, 12:118 Poster presentation: Johnston B, Hannan KE, Cercone K, Frazer T, Mwebesa E, Zirimenya L, et al., editors. Reliability and Acceptability of the Jerrycan Pain Scale for patients at Hospice Africa Uganda.

Principles of pain control

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 pain is relieved by opioids.
- Opioid-semi-responsive pain is relieved by the concurrent use of an opioid and an adjuvant drug that is a drug usually given for another purpose that also has analgesic action.
- Opioid-resistant pain (e.g., neuropathic and bone pain) is not relieved by opioids.

Neuropathic or nerve pain is more likely to fall into the semi-responsive or unresponsive groups. Bone pain falls into the semi-responsive group.

Analgesic approaches to pain relief

The optimal approach to pain management in children includes drug therapy, with analgesics usually being the mainstay of treatment.

Medicine	Reason	Morning		Afternoon		Evening		Night
		 On Waking	10 am	 12 noon	2 pm	 6 pm	10 pm	 Bedtime
Morphine 5 mg in 5 mL	Pain	2.5 mL	2.5 mL		2.5 mL	2.5 mL		5 mL
Senna	Constipation						2	
Ibuprofen 200 mg	Pain	2		2		2		2

Medication chart for Date.....

FIGURE 7.3 Example of treatment chart for family use in the community.

Correct use of analgesic drugs will relieve pain in most children, and should be based on the four key concepts recommended by the WHO:

1. by the ladder
2. by the clock (or by the sun if there is no clock!)

3. by mouth. Injections are not given at home because there are too few community health workers. Subcutaneous infusion pumps** are not always acceptable, and also need close monitoring, which is often not possible by the child or carer. They are not needed if oral morphine is available. Even if vomiting, the solution can be absorbed from the buccal mucosa.

By the ladder

Use the 'two-step' approach to analgesia, non-opioids and opioids. The second step in the three-step ladder that was proposed in 1986 by the WHO is now widely being omitted, and a two-step ladder is used instead (see Figure 7.4). This is because the middle step, namely codeine, is expensive and causes severe constipation, and if the child has cancer, they will need morphine, so can commence with a small dose that can then be titrated to the pain. Pain is classified as mild, moderate, or severe, and the analgesic choices are adjusted accordingly. The ladder approach is based on drugs that are, or must be, widely available in most countries. The sequential use of analgesic drugs is based on the child's level of pain, with a non-opioid analgesic usually being the first step.

Importantly, however, assessment of a child's pain may indicate the need for immediate use of a strong opioid. Morphine is the safest and most effective opioid, and the only affordable one in resource-limited settings.

There should be no hesitation in moving on to Step 2 (that is morphine) of the analgesic ladder if pain control is inadequate.

Only one drug from each pharmacological group should be used at the same time but remember that paracetamol plus a non-steroidal drug can be used together if there is no contraindication. Strong opioids can be increased until pain is relieved. Occasionally an alternative strong opioid (rarely affordable in resource-limited countries) may be substituted if side effects from the first opioid tried are intolerable.

The aim is for the child to be:

- pain free at night
- pain free at rest
- pain free on movement

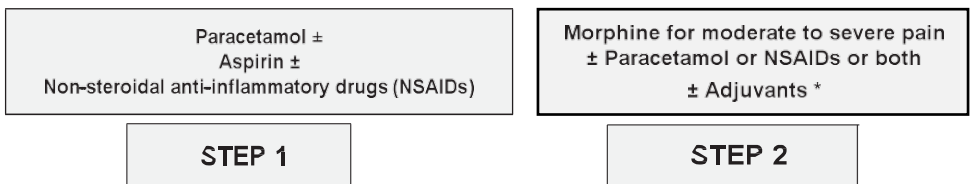


FIGURE 7.4 WHO two-step analgesic ladder. *An adjuvant is another drug (e.g. steroid or anxiolytic) or type of treatment (e.g. TENS or radiotherapy) that prevents but can also relieve pain.

It is recommended to avoid aspirin in children aged under 16 years due to hypersensitivity reactions, Reyes syndrome and peptic ulceration.

By the clock (or by sunrise/sunset)

Analgesia should be given regularly (e.g., every 4 hours or according to the half-life).

There is no place for 'when-requested' prescribing of analgesics in palliative care. The term PRN means "Pain Relief Never" in cancer. The dose must be titrated against that needed to control the pain of the individual patient.

Paracetamol and ibuprofen should be given at the recommended doses (see Section 9 Handbook 1), but the dose of morphine needed is estimated by titrating pain relief against the score at each visit, until pain, ideally, is down to zero, the patient is sleeping at night and active to that limited by any disability or weakness only.

Titration of pain versus morphine dose means giving an extra dose when pain breaks through and recording this. The next visit the clinician calculates the new total required in 24 hours and divides by 6 to give the 4 hourly dose and using for the 6th dose at night a double dose so that the child sleeps without interruption and without pain.

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 and explained to the family, so that these can be given as soon as breakthrough pain occurs.

The effectiveness of analgesia should be regularly reviewed, so that it can be titrated effectively against pain.

NOTE: Many countries have commenced without morphine and set up the service but have to be encouraged to continue advocacy with their Government after we have visited. They are not giving 'palliative' care but 'supportive' care only, which is so difficult when the child is in severe pain and the family is in distress. This occurs in the community, where most children have not reached cancer treatment. In 2021 there are over 10 countries in Africa that say they have palliative care but have not allowed, or even delayed, the import of morphine powder to reconstitute oral affordable morphine. Governments are reluctant to import it and even those who agree may have it stopped by incoming Ministers!

The alternatives to morphine on the market are not suitable and do not give smooth control and are also more expensive. Some are addictive. Remember oral morphine is too dilute to give intravenously. When given by mouth, and titrated against pain, oral morphine should not be addictive.

By the appropriate route

Children should receive drugs by the simplest, most effective, and least painful route. For this reason, the oral route is the preferred route.

Intramuscular injections should not be used. They are painful, and there is a risk of abscess and/or haematoma formation, particularly in children who may have low platelet counts or other blood-clotting problems. Also, use of the parenteral route means that the patient must be in hospital or a clinic and cannot go home. Children who are afraid of injections may deny that they are in pain and therefore suffer unnecessarily.

When selecting the best route of analgesic administration, it is important to consider 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

The doses of step 1 analgesics should be given according to recommended doses for their weight.

The doses of any analgesic must be based on the individual child's pain and circumstances. There is no single dose that will be appropriate for all children.

Regular reassessment of the child's pain and of the effectiveness of the analgesia is essential, so that the drug doses can be adjusted accordingly to keep the child pain free.

For some children, particularly those with cancer-induced pain, very large doses of opioids may be required to achieve satisfactory pain control. Therefore, it should be noted that some of the suggested dosage recommendations included in this section differ from those specified elsewhere in the handbook. This is appropriate in palliative care, and it reflects the differences in goals and priorities between the acute setting and the 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, rectally, or intravenously. It is available in an elixir, tablet, suppository and IV solution form, and can be given 4- to 6-hourly. Non-steroidal anti-inflammatory drugs (NSAIDs) such as ibuprofen and diclofenac are also helpful (for doses, see Section 9 Handbook 1).

It is now recommended that one should progress straight from paracetamol and NSAIDs to morphine.

CASE EXAMPLE

Haji, aged 3 years, presented with a clinical diagnosis of retinoblastoma. He was greatly distressed by severe pain. The lesion was too friable for a biopsy. Morphine was commenced immediately, based on weight, according to the WHO recommendation for children. This was titrated against the pain, and Haji's pain was eventually controlled on 100 mg 4-hourly and at night. He was then a happy child again.

Because of the friable nature of the growth which bled so easily, the radiotherapist allowed him to receive radiotherapy without a biopsy. The tumour disappeared. The morphine was reduced until he was pain free and well.

Today Haji is well, aged 16 years, and is still attending school.

Strong opioid analgesics (morphine)

Morphine is required either alone, or in combination with non-opioid analgesics and/or adjuvant drugs, to provide effective pain relief. Morphine does not have an analgesic 'ceiling effect' (i.e. there is no maximum dose), and children may require extremely large doses to obtain pain relief, but start at the recommended dose for severe pain (as described in Section 9 Handbook 1).

The strong opioid of choice internationally is oral morphine. This can be established locally in each country and is affordable. (see reference 5 at end of section)i. In resource-limited settings, children and their families may be alarmed by intravenous or subcutaneous infusions.

The oral route is preferred for morphine. Intravenous morphine has a ceiling due to limited change to M6G (the active ingredient) because it by passes the liver. Although a continuous infusion is commonly used in well-resourced countries, it is possible to achieve complete pain control with oral or rectal paracetamol or oral morphine in the palliative care setting. This approach is particularly useful in the community setting.

Children have been found to rapidly eliminate morphine metabolites, and this is most marked in younger children (under 9 years). This group of children may require more frequent dosing

and relatively higher doses to achieve pain relief. However, if oral doses are given at regular intervals, the most potent metabolite of morphine, M6G, accumulates and leads to smooth pain control.

Morphine must be available in all countries.

However, this is not the case at present. In Africa, only 15 out of 56 countries have oral morphine available for use at home, which is where most terminally ill patients want to die. Oral morphine that is made up in the country or within a district of the country is the affordable ideal. The drug is then immediately available, so pain can easily be controlled with it.

More complicated formulae and preparations may be available as immediate- or sustained-release preparations, including immediate-release suppositories. Once-daily preparations are commercially available, but there is little experience of their use in children, and they are too expensive for most resource-limited countries. Ideally, morphine should be free to all in need, and prescribed by a recognised prescriber. Usually, only doctors can prescribe. However, in Uganda, nurses can now prescribe after completing a Diploma in Palliative Care and clinical officers after a 9-week special training that emphasises prescribing methods and controls. Clinical officers have been trained for 4 years and can do more than nurses in most countries. In some African countries they are allowed to prescribe class A drugs after qualification.

Immediate-release morphine (from the list of essential medicines for children published by the WHO in 2010)

1. Morphine tablets (Sevredol): 10 mg, 20 mg and 50 mg.
2. Morphine sulphate mixture (Oramorph): 10 mg/5 mL.
3. Morphine sulphate mixture (Oramorph concentrate): 100 mg/5 mL.

The most affordable preparation is a morphine solution made from morphine powder in a pharmacy without the exorbitant profit taken by the 'middle man' (see Section 8).

The oral morphine starting dose is 150–300 microgram per kg body weight every 4 hours.

Immediate-release morphine should be given regularly every 4 hours. It may be useful to increase the night-time dose by 50–100% to eliminate night-time waking. Immediate-release oral morphine is the best choice in children because it is easier to titrate exactly against the pain.

Sustained-release morphine tablets (MST Continus) (5mg, 10mg, 15mg, 30mg, 60mg, 100mg and 200mg) and morphine granules for suspension (MST Continus) (10mg, 20mg, 30mg, 60mg, 100mg and 200mg), although available, are very expensive and therefore inappropriate for most situations in resource-limited countries. Those planning for a service must keep in mind the needs of the poor and spend the money available for morphine wisely so that there is enough for all in need. Occasionally a donation of these preparations make them available for a while. But very few donations continue forever and throwing a child back into pain must be avoided.

Breakthrough pain

Additional doses of Immediate-release morphine should be prescribed if the child experiences pain 'breaking through' (i.e., 16–17% of the total daily dose). This can be given up to hourly for breakthrough pain, and the parents should be advised to keep a record of all extra doses given so that the regular dose of morphine can be titrated accordingly, and more supplied as necessary.

Titration of the 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 doses plus 'breakthrough' doses) must be assessed. Usually increments of 20–50% of the previous total daily dose are required. Remember to increase the dose of breakthrough morphine accordingly when the regular dose is increased.

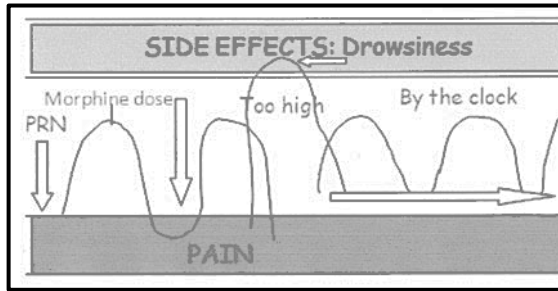


FIGURE 7.5 'As needed' (PRN) versus 'by the clock' versus 'high dose'. Initially, to the left, the PRN dosage regime results in episodes of unrelieved pain. In the middle, too high a dose produces drowsiness. To the right, the by-the-clock dosage regime results in constant relief of pain without drowsiness. (Diagram supplied by Dr Anne Merriman.)

Alternatives to oral route of administration for morphine

Indications for these include:

1. persistent vomiting
2. non-compliance with oral medication
3. dysphagia
4. bowel obstruction
5. physical deterioration that prevents oral intake
6. unsatisfactory response to oral medication.

Buccal Route

A stronger solution of morphine to match 4 hourly needs can be made to drip into the mouth inside the cheek where it is absorbed and metabolized similar to that swallowed.

Rectal route

This route may be acceptable for some children who are unable to take oral medication. Any oral preparation can be given rectally with similar effects:

- Paracetamol can be given as a suppository or tablet.
- Morphine solution (see above) can be easily given rectally and is very effective.
- Morphine suppositories (10 mg, 15 mg, 20 mg or 30 mg) can be given if available.

Although one can use the same dose and interval as for the oral route (i.e. 4-hourly), rectal administration is traumatic for the child, and generally a larger dose given in tablet or suppository form as half the daily dose 12-hourly is more acceptable.

Subcutaneous and intravenous routes

Avoid these in the community. It is possible to control nearly all pain by oral or buccal oral morphine solution.

As approximately 80% of children will die at home in resource-limited countries, oral or rectal morphine is likely to be the mainstay of treatment.

Diamorphine has less advantages in resource limited situations as it needs to be given by injection.

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. When appropriate, parents and children should be informed about the possible side effects and their management. Children on strong opioids should be assessed regularly.

Constipation

This is a common side effect, and laxatives such as bisacodyl (Dulcolax) or senna or sodium docusate must always be prescribed with strong opioids (see below). Advice should be given to increase intake of fluids and fibre (vegetables, fruit and cereals) in the child's diet where appropriate. If the child has loose stools, then this side effect may be welcomed. However, as soon as loose stools cease, laxatives need to be introduced.

Nausea and vomiting

Routine anti-emetics are not commonly needed but should be prescribed if required in case of opioid-induced nausea and vomiting. When such symptoms do occur, they normally resolve within 3 to 4 days.

Drowsiness and confusion

Daytime drowsiness, dizziness and mental clouding can occur at the start of treatment and sometimes following a dose increase. They almost always resolve within a few days. Cognitive and psychomotor disturbances are minimal once the patient is receiving a stable dose of opioid.

Pruritus

Itching is a not uncommon side effect of opioid treatment in children. Simple skin care alone may be effective. Also consider the following:

- Avoid hot baths
- Avoid using soap. Add Oilatum to the bath water and use aqueous cream as a soap substitute.
- Pat the skin dry rather than rubbing it.
- Avoid overheating and sweating.
- Use cool cotton clothing and bedding.
- Keep the fingernails short to reduce damage caused by scratching.

If itching is persistent, review the medication. If itching is opioid related and the drug cannot be changed, the addition of a systemic antihistamine such as chlorpheniramine may be beneficial.

Pruritus associated with obstructive jaundice will require good skin care plus systemic medication such as stanozolol, ondansetron or levomepromazine, if available.

Respiratory depression

Respiratory depression is uncommon in the conscious patient receiving oral morphine in order to control severe pain. If it does occur, management will be dictated by the child's overall condition and the place of care.

Nightmares and hallucinations

Both can occur. If they are distressing and not resolved by reassurance or resolution of other anxieties, try giving haloperidol at night (50–100 micrograms/kg). If needed in daytime also BNFC suggests 10-20 microgram/kg every 8-12 hours for restlessness and agitation in palliative care.

Urinary 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 for a short period.

Morphine toxicity

This can occur as a result of:

- too high a dose
- too rapid dose escalation
- pain that is not morphine responsive
- renal impairment
- previous therapeutic intervention to relieve pain (e.g. radiotherapy or nerve block).

Warning signs of morphine toxicity include:

1. drowsiness
2. confusion
3. pinpoint pupils
4. myoclonic jerks
5. hallucinations (auditory and visual)
6. vomiting
7. nightmares.

If toxicity occurs, consider reducing the morphine dose (several doses may need to be missed), then restart at a lower dose or stop morphine altogether.

Toxicity is rare when morphine is titrated against the pain. Constipation is the worst complication and can be prevented by introducing a laxative when morphine is started, unless the child has diarrhoea already, in which case the constipation would be beneficial for a few days, but the laxative needs to be introduced as soon as it ceases.

Watch carefully for breakthrough pain.

Address any side effects as discussed above. Escalating doses of opioids and metabolic disorders can exacerbate myoclonic jerks. Diazepam should be avoided. Just stop the morphine for a few days, ask the parent to watch carefully and then recommence with a lower dose.

Consultation with healthcare professionals who are experienced in palliative care is recommended.

Addiction and tolerance

Fear of addiction is not relevant when using opiates in palliative care, provided that a permanent source of opiates is available, **which must be the case**. In Uganda, in 2021, around 31,000 patients had been treated with affordable oral liquid morphine, without any abuse or addiction. Approximately 10% of these cases were children.

Prescribing opioids in patients with renal impairment

The active morphine metabolites are excreted by the kidney and accumulate in renal impairment, causing toxicity. When prescribing any opioid analgesics in children with renal failure, caution must be exercised, as patients with renal failure are extremely sensitive to

opioids. Renal failure is part of the dying process, and the team must be aware of this and reduce doses or increase time intervals as the child approaches death.

Suggested management strategies are as follows:

- Prescribe smaller doses of opioid analgesic.
- If problems with toxicity continue, consider giving smaller doses less frequently (i.e. 6- to 8-hourly).

Alternatives to oral morphine for severe pain

For information on approaches that can be used in well-resourced settings, see the Further reading at the end of this chapter.

Adjuvant therapy

Few children are truly morphine intolerant, and if the pain is not responding to morphine, always consider the aetiology of the pain and review the use of adjuvant therapy. The commonest causes of the family stopping oral morphine is advice from relatives, and where it is paid for, poverty. It is free for all prescribed by a registered prescriber (and that includes specially trained Nurses) in Uganda. We encourage African countries to urge their Governments to pay for this as it is so cheap, now US\$3 for 500mgs in 500mls.

Neuropathic pain

Co-analgesics such as an anticonvulsant or tricyclic antidepressants are essential, because this pain is only semi-responsive to opioids. The possibility of neuropathic pain should be considered if the pain has a burning or stabbing/shooting component. Burning pain requires amitriptyline in small doses slowly increasing if pain not improved in a week. For convulsive type stabbing/shooting pain, then anti-convulsant drugs are best. Phenytoin is the most affordable and available in LMICs.

Start at a low dose and gradually increase the dose to avoid sedation and toxicity.

However, some anti-retroviral drugs (ARVs) may interact with phenytoin so there is a need to check this out for children with HIV on ARVs.

Nerve compression pain

This may arise from compression of a nerve root, and morphine plus a trial of oral steroids should be tried. The result of adding a steroid can usually be seen within 24 hours, probably by reducing oedema around the tumour. If there is no improvement, steroid treatment should be discontinued. Set the lowest dose possible if continuing treatment.

Recommended doses for management of nerve pain:

Phenytoin

Age < 12 years: (1.5-2.5 mg/kg starting dose to target) and then 2.5-5 mg/kg twice daily (maximum 7.5 mg/kg twice daily or 300 mg once daily).

Age 12-18 years: 75-150 mg adjusted according to response up to 150-200 mg twice daily (maximum 300 mg twice daily).

Sodium valproate (often not available and expensive)

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/24 hours in divided doses.

Tricyclic antidepressants

- These drugs are useful for pain that is burning in nature.

- Give at night to avoid excessive sedation during the day. They can cause constipation.
- The analgesic effect begins after about 3-7 days of treatment but may take longer than this.
- Starting dose: amitriptyline 0.5 mg/kg at night increasing, if needed, to 1 mg/kg/day. Increase carefully to avoid excessive drowsiness. Lower doses are the most effective.

The BNFC doses for amitriptyline for neuropathic pain are:

Age 2-12 years: 0.2-0.5 mg/kg at night (maximum 10 mg), increasing gradually to a maximum of 1 mg/kg twice daily.

Age > 12 years: 10 mg at night, increasing gradually up to 75 mg at night if needed.

<https://bnfc.nice.org.uk/drug/amitriptyline-hydrochloride.html> Accessed 31.03.2021

Consider Gabapentin (if available)

Bone pain

Bone pain is generally resistant to opioids alone.

It usually responds to NSAIDs as the source of the pain is usually in the periosteum where the nerve endings are.

Although it works, take care giving *aspirin* for under 16-year-old children.

Bone pain not responding to NSAIDs in appropriate doses, should be supplemented commencing with low dose morphine, increased slowly.

Radiotherapy is particularly good for bone pain or metastases within the bone; not touching the periosteum. These areas may present with a pathological fracture. Consult a paediatric orthopaedic specialist if available.

Non-steroidal anti-inflammatory drugs (NSAIDs)

- NSAIDs have analgesic, anti-pyretic and anti-inflammatory properties. They are often effective in relieving musculoskeletal pain that is associated with bone metastases or soft tissue inflammation.
- Regular dosing is required for their full effect, but the maximum effect is usually seen within 2 weeks.
- It is worth trying another NSAID if there is no response to the first type.
- Damage to the gastrointestinal mucosa is the most frequent side effect. Gastric erosion and bleeding can be severe and difficult to control. If possible, ensure that NSAIDs are taken after food.
- NSAIDs are not usually appropriate for children with thrombocytopenia, because of their potential to cause gastric erosions and so increased tendency to bleed.
- According to the WHO there is no evidence for recommending the use of bisphosphonates (osteoporosis preventers) in children. In adults, modest improvements in pain have been observed, but also serious side effects such as osteonecrosis of the jaw.

For further information on NSAIDs, see Sections 9 and 65 in Handbook 1.

Nerve injury pain

This may arise either from tumour invasion of a nerve, oedema around a solid tumour, or as a side effect of radiotherapy.

Steroids in palliative care

Steroids have specific benefits in palliative care because of their ability to produce euphoria, improve appetite and increase weight gain. They also have an anti-inflammatory effect, which may be helpful in patients with nerve compression and raised intracranial pressure.

However, steroids should be used with caution in children, as the side effects of long-term steroid treatment can far outweigh its benefits. They include rapid weight gain, change in appearance, mood swings, behaviour changes and insomnia, which can be distressing for both the child and the parents, and the risk of gastric erosions. Most children experience symptom relief after short intensive courses, and if the prognosis is long, steroids should be withdrawn. If there is no improvement in symptoms within a short period of time (e.g. 5–7 days), steroids should be discontinued. If the initial symptom relief is not maintained, long-term use of these drugs should be avoided.

All steroids should be given in the morning in a daily dose to imitate the diurnal variation of natural steroid availability. Given in the evening steroids can cause insomnia.

Dexamethasone

High-dose dexamethasone is normally used to relieve pain associated with raised intracranial pressure, or spinal cord or nerve compression. Give steroids in the morning to avoid sleepless nights and to copy the normal diurnal rhythm of cortisol.

The initial dose is given in the morning as 25 mg for patients over 35 kg and 20 mg for patients less than 35 kg, followed by a sliding scale of reducing by 4 mg every 3 days until down to 10 mg per day, then continuing to decrease by 1–2 mg per day.

IM or IV in an emergency or until can swallow (usually once only):

- Age 1 month to 12 years: 100–400 micrograms/kg, once daily in the morning.
- Age 12–18 years: 8–24 mg daily.

Low-dose dexamethasone is normally used to improve appetite and well-being.

- Age 2–8 years: 0.5–1 mg, once daily in the morning.
- Age > 8 years: 1–2 mg, once daily in the morning.

Radiotherapy.

This therapy is only available in just over half the countries in Africa. Radiotherapy can be particularly useful for treating isolated sites of a disease if a tumour is radiosensitive. 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, if radiotherapy is available.

Non-pharmacological approaches to pain control

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
- diversional therapy with appropriate music, art or traditional games, according to the age of the child
- hypnosis and guided imagery
- massage and reflexology
- heat pads or cold packs
- transcutaneous electrical nerve stimulation (TENS).

Management of other symptoms

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(s) to implement an effective treatment plan.

Cancer-related causes include:

- raised intracranial pressure
- squashed stomach syndrome related to the presence of an abdominal mass
- irritation of the upper gastrointestinal tract
- gastric outflow obstruction
- anxiety
- uraemia
- pain
- blood in the stomach.

Treatment-related causes mainly involve the side effects of drugs, especially:

- opioids
- chemotherapy
- NSAIDs
- carbamazepine
- antibiotics.

Management of nausea and vomiting

Identify the cause(s) as described above (e.g., constipation, raised intracranial pressure, related to particular food), timing of meals and excessive encouragement to eat when naturally as condition deteriorates, appetite is reduced. Explain to parent/carer with appropriate advice and management.

Consider **stopping gastric irritants** such as antibiotics, NSAIDs and steroids if possible.

The proton pump inhibitor **omeprazole is particular effective:**

Age < 2 years, 700 micrograms/kg once daily increased to 3 mg/kg once daily, maximum dose of 20 mg once daily; body

Body weight 10–20 kg, 10 mg once daily, increased to 20 mg if needed;

Body weight over 20 kg, 20 mg daily increased to 40 mg once daily if needed. (Give the higher dose for 12 weeks only).

Prescribe an appropriate anti-emetic according to cause, availability, and cost.

- Review the therapy regularly and adjust it as required.
- IV fluids may be occasionally needed to counteract dehydration, but nasogastric tube insertion should be avoided where possible.

If treatment is unsuccessful, consider the following:

- Was the cause of the vomiting correctly identified and the appropriate anti-emetic prescribed?
- Has the anti-emetic had time to work at maximum dose?
- Is the route of administration appropriate for the child?

Anti-emetic therapy

Severe nausea and vomiting may require initial management by subcutaneous or IV infusion and then switching to oral medication when control is gained. The choice of anti-emetic depends on the cause of vomiting and the site of the anti-emetic action, so combinations of drugs with different sites of action are sometimes required, but to avoid side effects, avoid combining drugs of the same class. Extra-pyramidal side effects can occur with cyclizine, metoclopramide and domperidone (see Section 9 Handbook 1).

Haloperidol is the anti-emetic of choice for opioid-induced vomiting. It acts on the chemoreceptor trigger zone.

Dosage: 12.5–25 micrograms/kg twice daily by mouth, subcutaneously or IV. Haloperidol can be given orally at night.

By subcutaneous or IV continuous infusion ((rarely appropriate in the community)

1 month-11 years: 25-85 mcg/kg administered over 24 hours

12-17 years: 1.5-5 mg administered over 24 hours

<https://bnfc.nice.org.uk/drug/halperidol.html#indicationsAndDoses>

Accessed 5th December 2020

Cyclizine is used for nausea and vomiting caused by raised intracranial pressure or intestinal obstruction.

Dosage: all ages, by mouth, 1 mg/kg three times daily up to a maximum of 50 mg per dose. IV or subcutaneous route:

- All ages, 1 mg/kg 8-hourly

OR

Continuous IV/subcutaneous infusion (Avoid if possible, in the community):

- Age < 2 years: 3 mg/kg over 24 hours
- Age 2–5 years: 50 mg over 24 hours
- Age 6–12 years: 75 mg over 24 hours
- Age > 12 years: 150 mg over 24 hours.

<https://bnfc.nice.org.uk/drug/cyclizine.html> Accessed 31.03.2021

Dexamethasone

Dosage: use moderate doses (for example 100 micrograms/kg 12-hourly).

Metoclopramide

This acts on both the upper gastrointestinal tract and the chemoreceptor trigger zone, and speeds up gastric emptying. The extrapyramidal side effects are more common in children. It is useful for oesophageal reflux, gastric stasis, gastric irritation, gastric outflow, and high bowel obstruction. **Its use must be avoided in patients where there is partial or complete bowel obstruction.**

Dosage:

Oral route:

- Age 1–12 years: 100 micrograms/kg, two to three times a day
- Age > 12 years: 5–10 mg, two to three times a day.

Subcutaneous/IV route:

- Age 1–12 years: 500 micrograms/kg over 24 hours
- Age > 12 years: 15–30 mg over 24 hours.

If IV bolus injection give slowly over at least 3 minutes

Domperidone

This acts on both the upper gastrointestinal tract and the chemoreceptor trigger zone, and speeds up gastric emptying.

Dosage:

Oral route:

- Age 1–12 years: 200–400 micrograms/kg, three to four times a day
- Age > 12 years: 10–20 mg, three to four times a day.

Rectal route:

- Age 1–12 years: 15–30 mg, two to three times a day
- Age > 12 years: 30–60 mg, two to three times a day.

Constipation (see Section 17 for more advice)

Constipation is common in paediatric palliative care, and the causes may be multi-factorial. The prevention and relief of constipation in the terminally ill child is very important, as if left unresolved it can cause abdominal pain and discomfort, and nausea and vomiting.

Consider the following causes:

- drug induced (e.g. opioids, anticholinergics, antidepressants)
- reduced physical activity
- poor oral intake and general debility
- dehydration
- bowel obstruction
- spinal cord compression.

Management

- Treat the underlying cause where this is appropriate and possible.
- Constipation should be anticipated when opioid, anticholinergic or antidepressant drugs are being used, and laxatives should be prescribed prophylactically.
- Use laxatives appropriately and at the right doses, and avoid mixing two drugs from the same group (e.g., two stimulants).
- A good first choice is the combination of a stimulant laxative and a softening agent (e.g. senna plus sodium docusate).
- Titrate doses up as required, rather than adding a new laxative.
- If oral therapy fails, consider rectal measures such as suppositories/enemas.
- Affordable and useful in the tropics for the older child, are seeds of the paw paw fruit. These are dried and compressed to form a powder. Start with a tiny dose (1/2 teaspoon) and increase until going regularly. This has been researched by KEMRI.

Bowel obstruction

Bowel obstruction may be mechanical or functional, or both. The aim is to control pain and nausea. 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 appropriate, although for persistent vomiting due to obstruction a nasogastric tube may be helpful. However, insertion and their continued placement can be very uncomfortable, and should be avoided in a child if all possible.

Management

Elimination of pain and colic:

- For constant background pain, administer buccal morphine solution or morphine by continuous IV or subcutaneous infusion, using a portable syringe driver. Syringe drivers are rarely available and experience indicates that buccal liquid morphine, small volumes, works well.
- If colic is present, **do not use** prokinetic anti-emetics (such as metoclopramide or domperidone).
- Discontinue bulk-forming, osmotic and stimulant laxatives.

- Relieve associated constipation, continue to use 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 give IV as a single dose over at least 1 minute (age 2–5 years, 5 mg IV; 6–10 years, 10 mg IV; 11–15 years, 15 mg IV; 15–18 years, 20 mg IV). Repeat 8-hourly as required.

Elimination of nausea and reduction of vomiting when partial or complete bowel obstruction is present

- The choice of anti-emetic depends on whether colic is present.
- If colic is present, **cyclizine** is the first-line drug. Add **haloperidol** if nausea persists.
- If colic is absent and flatus is present, a trial of subcutaneous or IV metoclopramide is indicated. If this is ineffective, instigate management as described above.
- Dexamethasone may be of benefit in second-line management.

Dyspnoea

This is the most feared symptom and one of the most common reasons for admission. However, admission to hospital can exacerbate symptoms, due to the shock of rapid onset, and long travel. Rapid deterioration may occur when hospitalization demands a child being away from loved ones.

Shortness of breath associated with pulmonary complications in advanced paediatric cancer can be very distressing for both the child and the parents and requires effective management. The underlying pathophysiology needs to be considered when deciding on the management.

Common causes of dyspnoea include:

- metastases
- effusions
- pulmonary fibrosis
- anaemia
- infection
- superior vena cava (SVC) obstruction
- anxiety/fear
- increased secretions
- cardiac failure
- chest wall pain or constriction
- pulmonary embolus
- gross ascites.

Management

- Identify the cause.
- Give a clear explanation to the parents and the child.
- Treat the specific cause(s) or modify the pathological process (e.g. high-dose steroids and radiotherapy for superior vena cava obstruction).
- Non-drug measures are also important and include:
 - a calm approach
 - breathing exercises
 - an appropriate position
 - providing cool air (e.g., with fanning with the local newspaper or open a nearby window)
 - play therapy.

Drug treatment

Morphine has a complex effect on respiration, which is not fully understood. It can reduce the respiratory rate to a more comfortable level. This drug should be prescribed regularly in

children with continuous breathlessness at standard analgesic starting doses. If the child is already on morphine, increase the dose by 30–50%.

The anxiolytic and sedative effects of benzodiazepines also cause relaxation of the respiratory muscles. This may be helpful if the child or teenager is very anxious, and these drugs should be administered as a single dose and then at night or twice daily. The long half-life of benzodiazepines (around 36 hours) means that they should be avoided if possible.

Diazepam (oral route): Dosage:

- Age 4 weeks to 1 year: 200 micrograms/kg, two to three times daily
- Age 1–12 years: 2mg, two to three times daily
- Age > 12 years: 5–10 mg, two to three times daily.

Midazolam is rarely available in suitable preparations and should be avoided as even a slight overdose can trigger unconsciousness and earlier death,

Lorazepam is not always available but is well absorbed sublingually (so is useful for panic attacks), short acting, with a rapid onset of relief and a shorter half-life.

Dosage:

- Age 1–12 years: 50–100 micrograms/kg (maximum of 4mg per dose) (BNFC).
- Age > 12 years: 1–4 mg per dose. The dose may be repeated after 12 hours.

Corticosteroids may be useful, particularly in patients with superior vena cava obstruction and multiple lung metastases. Moderate doses of dexamethasone should be used, and the benefit should be apparent within 5 days. The dose should then be reduced to the lowest effective dose. Oxygen will be of benefit for hypoxic patients but is rarely available for home use.

Opening windows and doors, limiting people in a room, and fanning the patient may also be helpful.

Nebulised saline or salbutamol may provide subjective relief, especially if wheezing is present.

Steam inhalations or traditional medicines that are inhaled have sometimes given relief but may also be harmful. Commonly used in palliative care are lemon grass, ginger, rosemary and garlic. But this varies from county to county and tribe to tribe. A trial of such treatments needs to be carefully controlled (if possible).

Although steam inhalations and traditional medicines maybe helpful in some patients who cannot access health care outside the home, if there are significant respiratory symptoms, every effort must be made to admit the patient to a suitable hospital setting where oxygen, non-invasive respiratory support including nebulisers, and dexamethasone should be available.

Cough

Consider the following causes:

1. respiratory infection
2. airways disease
3. malignant obstruction
4. drug induced
5. oesophageal reflux
6. aspiration of saliva.

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 the following:

- simple linctus
- codeine linctus (this will cause constipation, so add a stool softener)
- opioids (as above)
- nebulised saline
- oral antibiotics (these are indicated if symptomatic chest infection with a productive cough is affecting quality of life).

Anxiety

Anxiety is common during palliative care. Talk to the child and give enough time to both the child and the parents or carers to discover the cause and give reassurance. Try to identify the cause of the child's anxiety (e.g., whether it is related to symptoms or fears about what is happening). Simple explanations, reassurance and a calm environment are important. Physical therapies such as relaxation and massage may be helpful. Try and avoid sedatives, especially diazepam, which has a long half-life of 36 hours. Lorazepam is preferable but often not available. Only use if other measures fail.

Anxiety and discomfort go together, so reassess the child's pain.

Massive external bleeding

Death from massive external bleeding is uncommon in children, but the risk of this is frightening and distressing for both the child and the parents, and prevention of such bleeding should be the aim of management, although this may not always be possible.

Causes of external bleeding include the following:

1. a low platelet count
2. clotting deficiencies
3. primary or secondary liver disease
4. disease progression
5. initial treatment (e.g. radiotherapy, chemotherapy).

Management

If there is a risk of massive haemorrhage, it is extremely valuable to have subcutaneous or intramuscular morphine and an appropriate sedative (e.g. rectal diazepam, buccal midazolam) readily available.

Persistent surface bleeding

This is not uncommon in children with leukaemia and can be alarming to both the child and their family but can be managed in the home environment.

Management

Topical treatment soaking gauze in adrenaline 1 in 1000 solution and applying it directly to the bleeding point.

Other haemostatic dressings can be used for persistent surface bleeding (e.g., in fungating tumours). These include crushed tranexamic acid tablets. If not available metronidazole sprinkled on to the area, or an alginate dressing such as Kaltostat if this is available.

Oral tranexamic acid can be useful if it is available (15-25 mg/Kg 2-3 times daily max single dose 1.5g), and can be used topically undiluted, applied directly to bleeding gums or nostrils, or used as a mouthwash. It can also be given IV (10mg/kg 2-3 times daily (max dose 1g) as prophylaxis.

The use of a dark-coloured handkerchief or towel at home to mop up the blood may help to reduce anxiety.

Use a dark coloured sheet or cover to prevent alarm at the sight of blood by relatives.

Spinal cord compression (see Section 58)

Consider spinal cord compression if the following signs and symptoms are present:

- localised pain in the spine, radiating around the chest
- sudden onset of weakness (e.g. of the legs)
- sensory disturbance
- sphincter dysfunction.

This is usually a clinical diagnosis, and action needs to be taken immediately.

Investigations such as computerised tomography (CT) and magnetic resonance imaging (MRI) are not usually available.

Management

- Patients with paraparesis have a better prognosis than those who are totally paraplegic.
- Loss of sphincter function is a poor prognostic sign.
- Rapid onset of complete paraplegia has a poor prognosis.
- The main therapeutic options are:
 - corticosteroids that can shrink the tumour and relieve spinal cord compression
 - radiotherapy.
- Steroids should be given in high doses initially and then reduced according to the response. These drugs often bring about an early improvement and relief of pain by reducing the peri-tumour inflammation. Give steroids in the morning to avoid insomnia and to copy the normal diurnal rhythm of cortisol.
- High-dose dexamethasone: The initial dose is given in the morning as 25 mg for patients over 35 kg and 20 mg for patients less than 35 kg, followed by a sliding scale of reducing by 4 mg every 3 days until down to 10 mgs per day, then continuing to decrease by 1–2 mg per day. The initial dose can be given IV if urgency required but oral doses should then follow. However, if symptoms recur, revert to a higher maintenance dose.
- In early stages, referral for concurrent radiotherapy should be considered if the prognosis is not very poor. If radiotherapy is available and if the patient lives far from the radiotherapy service with a vehicle needing to go over bumpy roads, damage to or complete transection of the cord may occur on the way. Every effort must be made to make the journey as smooth and comfortable as possible
- If radiotherapy is not available, as is the case in around 30% of African countries, and unless the parents have enough financial resources to take their child to another country, palliative support may be the best option.
- Once paraplegia is diagnosed, surgery, such as laminectomy, is only rarely indicated.
- Consider using a pressure-relieving mattress and give pressure area care.
- Pay attention to bowel function.
- Start physiotherapy to prevent contractures.
- Perform catheterisation if urinary obstruction is present.
- Avoid danthron-containing laxatives if the child is catheterised or incontinent, because of the risk of danthron burns.

Psychological support

Children experience significant psychological suffering as a result of loss of their ability to walk or run, as well as their inability to play and go to school. Other problems occur such as disfigurement, awareness they could die. Cultural beliefs that they are being punished imposed by parents blaming witchcraft by a neighbour, near relative or ancestor, need to be addressed with patient and parent. The child needs understanding and sympathetic advice from their healthcare provider and carers at this time.

Many children are deeply spiritual and find prayer and discussions such as “will I go to heaven”? “is God punishing me”? Reassure them that their God is a loving Father who forgives especially children. He is not to be feared. Be aware of their previous religious affiliation and approach and do not try to convert.

Convulsions

Convulsions 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. Keep in mind, the long half-life, as well as the failing renal function in many approaching the end of life. Do not use regular doses but ensure close observation of the child and obtain reports regularly (initially daily) by phone or directly from the child's carers.

Dosage:

- Age < 1 year: 2.5 mg (half of a 5 mg rectal tube/rectal solution)
- Age 1–4 years: one 5 mg rectal tube/rectal solution
- Age 5–12 years: 5 mg or 10 mg rectal tube/rectal solution
- Age > 12 years: 10 mg rectal tube/rectal solution.

For continuing severe seizures, consider giving midazolam by the buccal route, subcutaneously or by IV infusion if the child is in hospital (see Section 70 Handbook 1).

Care is required with midazolam as it may give permanent anaesthesia so that communication becomes impossible.

Muscle spasms

Muscle spasms can be severe in children with neurological and neurodegenerative disorders. They may occur alone, or be triggered by pain elsewhere (e.g. due to constipation).

Useful drugs for muscle spasm

Baclofen orally

- Age 1–10 years: initial dose 300 microgram/kg/day in four divided doses, increasing to usual dose of 0.75–2 mg/kg/day in divided doses
- Age > 10 years: 5 mg three times daily, increasing to 20 mg three times daily (up to a maximum dose of 100 mg/day).

Diazepam orally (initial doses are shown):

- Age 1 month to 1 year: 250 microgram/kg twice daily
- Age 1–5 years: 2.5 mg twice daily
- Age 5–12 years: 5 mg twice daily
- Age > 12 years: 10 mg twice daily up to a maximum dose of 40 mg/day (BNFC).

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 their skin.

Children with some degenerative conditions may have had faecal or urinary incontinence for a long time, whereas for others this may become a feature during the end stage of their disease (e.g. 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 (see Section 58).

Some useful suggestions

- Review laxatives where appropriate.
- Consider giving desmopressin tablets, 200–400 micrograms, or sublingual tablets, 120–240 micrograms, can be used. Care is needed as desmopressin can cause water retention and hyponatraemia, so start with lower doses. Keep a urinal or bedpan close to the bedside.
- Use cotton pads or towels (with plastic underneath) on top of the bed sheet. This will avoid the need to change all the sheets, and thus minimise disturbance to the child.
- Keep the area well ventilated (or keep a window open if appropriate).
- Try to ensure that the skin is kept clean, and use dimethicone, zinc and castor oil or other barrier creams if these are available.
- Help the child to wash regularly.
- Try to always preserve and maintain the child's dignity. Give reassurance and support to both the child and the parents.

Fungating wounds

Fungating wounds are rare in paediatric palliative care, but in resource-limited settings they are not infrequently encountered. They may occur with soft tissue sarcomas, often of the head and neck, and Kaposi's sarcoma which can be very distressing for the child and their family.

Useful management (where available) include the following:

- Soak any dressings with clean water, saline or Ringer-lactate or Hartmann's solution to ease removal, as these tumours may be friable and prone to bleeding.
- If possible, have available topical adrenaline 1 in 1000, or an alginate dressing (e.g. Kaltostat, or tranexamic acid), to apply topically to the tumour if it bleeds profusely (e.g. during a dressing change).

These tumours can cause offensive smells due to anaerobic microorganisms, which can be distressing to the child and their family. Metronidazole crushed and applied to the surface of the fungating area, is the treatment of choice. Oral metronidazole does not penetrate the fungating area, as the blood supply is often non-existent or poor. It is cheap and readily available in all resource-limited countries. This approach is very effective, and the smell is usually gone in 24 hours.

Charcoal dressings, if available, may help to absorb the odour.

The use of honey for dressings is also of benefit in controlling bacteria and odour.

Simple measures such as the use of aromatherapy oils around the home may be helpful, too.

The final days and hours of life

Terminal restlessness and agitation

These symptoms are not uncommon in the final stages of life. Useful drugs include buccal midazolam, and oral or rectal diazepam.

Midazolam (often not available in the community) is the sedative of choice, as it can be given via the buccal mucosa.

Dosage:

The initial regime is 30–100 micrograms/kg given as required. Titrate upwards as required (the upper dose may be limited by volume).

Rectal diazepam may also be useful.

Dosage:

5–10 mg rectal tube as required. The dose may be repeated if child remains very agitated and restless.

Increased secretions

- Increased secretions (the 'death rattle') can be more distressing for the parents and carers than for the child. It is important to explain this to those caring for the child.
- Good mouth care is essential.
- Anti-secretory agents are useful but can cause drowsiness and anti-cholinergic side effects.
- Start drug treatment early in order to avoid build-up of excessive secretions.
- However often the patient is unaware of the discomfort and the relatives are most upset as they think it is upsetting the patient. The excessive dryness of mucous membranes caused by hyoscine, can be worse for the patient. This should be explained to the relatives.

Hyoscine hydrobromide (scopolamine)

- This drug is anticholinergic.
- It reduces pharyngeal secretions which may bring discomfort when the patient is comfortable.
- It should be used with care as it dries up all secretions which can make the end of life very uncomfortable.
- It mixes with other commonly used drugs.
- Potential routes for administration: oral, as a sublingual tablet, IV or subcutaneous.

Dosage:

Oral/sublingual route:

- Age 1–12 years: 10 micrograms/kg/dose, four times a day
- Age > 12 years: 300 micrograms/dose, four times a day.

Subcutaneous or IV infusion:

- All ages: 10–50 micrograms/kg/24 hours.

Loss of the oral route for food and medication

As a child's condition deteriorates it may become difficult to use the oral route for medication. Buccal and rectal routes are the best options in this situation and work well. As discussed earlier, other routes that can be used at this point are the rectal, subcutaneous and (where already established) IV routes. Children who have been treated for cancer in private well-resourced hospitals may have central IV access, which can be used effectively in palliative care, but usually only in hospital.

Drugs that can be given via the subcutaneous or IV route (not common in some African countries) include analgesics, anti-emetics, sedatives, anxiolytics, and anticholinergic drugs. These can be combined in an infusion if they are compatible with each other.

If they are available, it is possible to use small portable infusion pumps (e.g. Graseby MS 26, WalkMed) to deliver combinations of medication over 24 hours. However, these devices are

unlikely to be available for home use in most resource-limited countries, and home palliative care teams would not generally be able to provide this form of treatment. Sometimes individual carers may be able to manage this form of treatment.

Note: Hospice Africa Uganda has rarely used injections or pumps, since 1994, and has completely controlled most pain and symptoms by using oral or buccal routes for medication.

Additional notes

- Avoid administering high concentrations of drugs in combination, especially when using cyclizine.
- Avoid mixing dexamethasone with other drugs if possible.
- Never give chlorpromazine, prochlorperazine or diazepam subcutaneously.
- More than two drugs can be combined in portable syringe drivers, although there is little supporting evidence in the form of clinical data. Always consult your local pharmacist before using any unusual combinations.

Psychological and spiritual support for the child, parents, and siblings

In many LMIC's, spirituality and prayer are important to the child and the family. Talking to the child about their spiritual needs is important and praying with or by them is usually acceptable. They need to know that they are loved by their God and by the clinicians and carers as well as the family. Touch, holding hands and hugs are acceptable (except in the Covid era) and go a long way to give comfort at this lonely and sad time for the child and family.

Care, that is child and family centred, is an essential principle of palliative care. The availability of an experienced key worker to coordinate the child's care with community healthcare professionals is essential, with good communication both between professionals and between professionals and the family being of paramount importance.

Initially, parents may need a lot of support when deciding whether to withdraw curative treatment and where to care for their child. Mostly they will be at home due to economic circumstances, and traditional preferences. Whatever the care setting, in hospital or at home, the parents will have many questions, fears and anxieties at this time, and, if possible, the opportunity to discuss their worries, changes in the child's condition and symptom management should be available 24 hours a day. Commonly asked questions include '*How long will it be?*' and '*How will my child die?*' These questions are not easy to answer and will also depend on the nature of the child's illness. For example, a child with leukaemia may have a very short period of palliative care, whereas 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 to emphasise that every child is different, and to guide the parents as the disease progresses. 'Days or weeks', 'weeks or months' or even 'hours rather than days' give adequate warning without being too precise.

Traditional practices are strong and may come into play. This is complicated because these vary from tribe to tribe, affecting the place of death, where they are buried and other rituals.

Parents may worry about their child being in pain, but also have anxieties about the use of strong medication such as morphine. A clear explanation of the use of analgesics is essential in this situation.

Many parents will want advice on talking to the dying child and their siblings. How to prepare the child's brothers and sisters will depend very much on their age and level of understanding, and on parental beliefs. For older children and teenagers, it is probably best to be honest, to prepare them gradually for what is happening and allow them to ask questions and participate in their

sibling's care, if appropriate. However, culture often does not allow the siblings to be involved at this time. And some are at boarding school and do not come home.

With younger children, the language used must be very simple and clear. For example, it is important to avoid using the phrase 'going to sleep' as the analogy for death as then parents are afraid of letting the child sleep! Or the older child is afraid to sleep! It is probably more appropriate to prepare younger children for a sibling's death when the end is obviously very close. Cultural preferences also need to be taken into account.

Talking to the child who is dying is a very personal matter for parents and will also be influenced by the child's age and understanding of the illness. For example, a teenager with cystic fibrosis may have anticipated death in adolescence or young adulthood, and a teenager who has had multiple relapses of cancer for many years may now realise that the 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 allowed to express themselves, they can become very anxious and agitated, or even withdrawn. Healthcare professionals can only try to encourage the parents to have an open and honest approach to their child's questions and wishes at this time.

The older children are becoming more computer literate and will look up their illness and know the prognosis, but the parents are keeping it from them. Telling the truth to an adult or a child patient is often not acceptable to them as they think it will accelerate death.

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 to prepare them for what may happen at the time of death if they wish to have this information. Changes in breathing are commonly distressing, and simple explanations of, for example, Cheyne–Stokes respiration or the 'death rattle' can avoid unnecessary distress. A single expiratory breath after death if the child is moved is not uncommon, and it should be explained to the parents that this does not mean that their child is still alive. Explanations of the changes in colour and very cold feel of the skin are important for parents and siblings. If they are not warned, parents may become distressed that their child was incontinent at the time of death. In the case of some diseases (e.g., leukaemia), the parents will need to be warned that their child may bleed from the nose or mouth at the time of or after death, and given simple practical measures for managing this situation.

Most families in LMICs have seen deaths so many times that they are more prepared than those who live where child death is rare. However occasionally some families require professional support around the time of the child's death, but this is rarely available in LMICs because of shortage of staff in hospitals.

After their child has died, the parents must be reassured that they need not rush to do anything but may spend some time with their child. Usually there is an all-night vigil as the family and friends gather around the body, pray and accept visitors bringing condolences. However, it is also important that any specific cultural or religious requirements are acknowledged and attended to. The parents should be encouraged, if they wish to do so, to hold, wash and dress their child. Some parents may want to take photographs, locks of hair, or hand and footprints, or organise favourite toys, photographs, letters, or other items for the child 'to take with them'. The participation of siblings in these activities can be very helpful.

Families have traditional ways of embalming the body in hot countries. They may use kerosene and inject it into the body cavities, especially if the body is to be taken on a journey to the family compound, for burial.

In countries where it is usually necessary for a child's death to be confirmed by a medical practitioner it is very rare for a post-mortem to be required. The death certificate then gives the authority for the death to be registered (according to each country's prevailing law) and the funeral arrangements to be made. However, in low resource settings many deaths are not recorded or properly documented and unless there is suspicion of a crime, the police are rarely involved in confirming death. The health workers in the community, usually in the neighbourhood, will help confirm deaths at home and burial arrangements will proceed from that point.

The specific cultural and religious beliefs of the family and the country in which they live will play an important role in the child's funeral. However, the parents may need advice about the choice between burial or cremation (uncommon in many countries), or about the funeral service itself.

Support after death

Support for parents, siblings, and the extended family around the time of the child's death and in the weeks and months afterwards will be very much influenced by the family's culture and family network, and by the support provided during the child's terminal care. Bereavement contacts from the professionals involved with the family should be offered wherever possible. A bereavement visit, and a card and small donation to the funeral expenses, will help them to understand they are not abandoned but thought of as friends. Follow up needs to explore how the family is coping and seeing if further support is required. In Africa cultural customs often take care of the bereaved. A telephone call to check on them is usually appreciated. Many Hospices and home care services have an annual memorial service for the deceased of that year, with the different faiths represented. This is a closure for the family and the team so involved with the departed and is really appreciated by the families.

Clinicians need to be aware of the cultural support and customs that affect bereavement, and refrain from imposing their own personal needs and values on others.

Ongoing bereavement support should be based on the family's specific 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 the anniversary of their death provide an opportunity for healthcare professionals and friends to show the family that their child has not been forgotten.

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Section 8 Medical Ethics and professional standards

Introduction

Ethics is the study of morality. Morality is defined as the values used to guide human behaviour and decision making.

Ethics of Health care

Medical ethics deals with moral issues in medical practice. Anyone who is involved in patient care uses ethics, despite most not having had any formal training.

Usually, law and ethics are closely related, but there are some differences. Ethical obligations usually take priority over legal duties.

There are four basic principles of ethics in healthcare which are important when making decisions

1 **Autonomy:**

If a patient is fully informed and competent (i.e. can understand the implications of having treatment or no treatment), they have the right to refuse or accept treatment. Such decisions must be respected, even if they are not thought by health workers to be in the patient's best interests.

2 **Beneficence:**

This means doing good and promoting well-being. This must be considered for the individual patient and may conflict with autonomy, although patient autonomy takes priority

3 **Non-maleficence:**

This means doing no harm. In healthcare, there is a risk of harm whenever investigations or treatment are carried out. Maleficence refers to harm inflicted with no intended benefit to the patient.

4 **Justice:**

This means equality or fairness. It refers to the fair allocation of scarce resources to patients, and the justification for money spent in the health service. This may mean equal access to healthcare, maximum benefit of resources available, or allowing people choice in their healthcare. This decision may not be able to be taken by an individual. In a society where justice prevails, the aim is for all citizens to have equal access to healthcare.

Healthcare ethics in different countries

Different cultures and societies have different expectations about the relative values of the individual ethical principles above. Some societies expect a beneficent or non-maleficent approach, whereas others expect an overriding respect for autonomy. It is essential that, as well as working within professional ethics, health workers respect the law in the countries where they practice, provided that the law does not harm the patient.

If a law or laws do harm patients or fail to protect them from harm, the healthcare workers should advocate for appropriate change in those law(s).

Some cultures put less weight on the individual and involve the family and/or community in decision making.

Gender may also affect decision making. In some societies, decision making is the man's responsibility, and the woman has no autonomy.

In some countries, health workers will not be forced to do anything unethical, whereas in others, there may be pressure from the police or the army to participate in torture or reveal the names of patients and their injuries and so break confidentiality.

In the USA, the emphasis is on the individual's autonomy, whereas in Africa beneficence and distributive justice maybe more important to communities.

Different healthcare associations have their own Ethical Code of Practices, References for International Codes of Ethics for Nurses and Midwives and for Nurse Anaesthetists are listed at the end of the chapter. Ethical dilemmas are common in clinical practice. The ethics of consent, confidentiality, end-of-life decisions and research will be discussed below.

The ethics of consent

Informed consent is the process of receiving information needed to make a decision.

Components of consent

- Assume the person has **capacity** unless there is evidence to suggest otherwise. This means s/he understands the information given, and can use it to make an informed decision
- All decisions must be **voluntary** and free from pressure from others.
- All decisions must be **informed** - the person must be given all of the information about what the treatment involves, including the benefits and risks, whether there are reasonable alternative treatments, and what will happen if treatment does not proceed.

Mental capacity is the ability to make decisions for yourself. People who cannot do this are said to 'lack capacity'. This applies to some children and some adults, due to illness, injury, a learning disability, or mental health problems that affect the way their brain works.

To have capacity a person must be able to:

- Understand the information that is relevant to the decision they want to make
- Retain the information long enough to be able to make the decision
- Weigh up the information available to make the decision
- Communicate their decision by any possible means, including talking, using sign language, or through simple muscle movements such as blinking an eye or squeezing a hand

The child's capacity and parental involvement: respect for autonomy

Where a child is not able to give or withhold consent to treatment, a person with parental responsibility must give permission for the child to have 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 by law in most countries. It is the doctor's responsibility, with input from other healthcare workers looking after the child, to assess a child's capacity to consent or refuse a proposed investigation or treatment before providing it.

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

'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.'

Children's capacity has a legal standing in some countries (such as the so-called 'Gillick' competence in English law).

Providing the information: an essential component of consent

Information should include details of the possible diagnoses and prognosis, possible management options, the purpose of a proposed investigation or treatment, and the likely benefits and probabilities of success, and information about any serious or frequently

occurring risks. The information should be given in a way that is clearly understood and remembered and may be supplemented by written or visual information.

All this information may be overwhelming for patients and their families. However, it is important that personal views about how much to disclose are not imposed on the patient when explaining an illness or treatment to them.

When providing information, it is essential that professionals find out about the patient's (and family's) needs and priorities. This is often the most difficult part of the communication process and involves answering any questions the patient or family raise honestly and as fully as possible. It is for the competent patient, not the doctor, to determine what is in the patient's own best interests.

In some societies, disease and pain are interpreted in terms of sin and retribution. This may make it difficult for health workers to explain diagnostic and management options in medical terms.

It may be inappropriate to discuss treatments that are not available.

Emergency situations

If the patient is unconscious or otherwise lacks capacity, urgent investigation and treatment may be carried out. This is sometimes called presumed consent, where the healthcare worker does what they believe is in the best interests of the patient.

The UK General Medical Council (1998) advice on consent for emergencies includes the following:

'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.'

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

These are as follows:

- 1 to participate in developing a plan of treatment
- 2 to receive an explanation of how each part of treatment will be provided
- 3 to receive clinically appropriate care and treatment
- 4 to be treated in a manner that is free from abuse, discrimination and/or exploitation
- 5 to be treated by staff who are sensitive to the family's cultural background
- 6 to be given privacy.

Confidentiality

This is not an ethical principle, but it involves respect for autonomy, beneficence towards the patient, and a desire to act with non-maleficence.

Confidentiality respects an individual's autonomy and their right to control information relating to their own health.

Patient information should only be disclosed without the patient's consent to individuals who **need** to know, and the recipient(s) of such information should keep it confidential.

Most countries have laws to enable the breaking of confidentiality in some circumstances – for example, to protect the safety of a third person.

- 1) to prevent a serious crime, as information may need to be disclosed to the police
- 2) to report suspected child abuse
- 3) to report someone who is HIV positive who is unwilling to inform their sexual partner(s) about this and does not consent to the healthcare worker telling the partner(s). The healthcare worker should inform the patient of his or her intention to inform the partner(s).

End-of-life issues

These include the following:

1. attempts to prolong the life of a dying patient
2. euthanasia and medically assisted suicide
3. care of terminally ill patients.

1. Attempts to prolong the life of a dying patient

Where there is no benefit to the patient, these attempts are unethical. Futile treatments are those that are assessed as bound to fail and which are prolonging the dying process. Withholding or withdrawing treatment is not the same as participating in assisted suicide or assisted euthanasia. This does not include palliative treatment, which must always be offered. The patient may decide to discontinue treatment for a life-threatening illness, while able to understand the information needed to make an informed choice. They must always be offered palliative treatment, whatever their choice.

The World Medical Association (WMA) is opposed to euthanasia and physician-assisted suicide because it is of the view that utmost respect has to be maintained for human life.

Euthanasia and assisted suicide are illegal in most countries and prohibited in most medical codes of ethics.

2. Euthanasia

Euthanasia (also known as 'assisted dying') means intentionally performing an act that is intended to end another person's life, and the patient has voluntarily asked for their life to be ended and is competent, informed and has an incurable illness

Assisted suicide

This is knowingly and intentionally providing a person with the knowledge or means to commit suicide, including counselling about lethal doses of drugs, prescribing or supplying drugs.

3. Care of terminally ill patients

Healthcare workers should provide compassionate end-of-life care, including relief of pain and suffering (see Section 7).

Withholding or withdrawing medical care

If the patient is a child, the healthcare team and the parents must serve the best interests of the child.

- 1) Although there is no significant ethical difference between withdrawing and withholding treatment, there are significant practical differences.
- 2) Optimal ethical decision-making concerning patients requires open and timely communication between members of the healthcare team, the patient and the family.
- 3) Parents must decide on behalf of a child who is unable to express preferences, unless healthcare workers caring for the child believe the parents are acting against the child's best interests. Cultural practices and religious beliefs may have an impact on this.
- 4) The wishes of a child who has sufficient understanding and experience should be given substantial consideration.
- 5) Resolution of disagreement should be by discussion, consultation, and consensus.
- 6) The duty of care is not an absolute duty to preserve life by all means.
- 7) A shift from life-sustaining treatment to palliation represents a change in aims and objectives and does not constitute withdrawal of care.
- 8) Health workers should never withdraw treatments that alleviate pain or promote comfort.

- 9) There is a difference 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. Treatments that may incidentally hasten death are justified if their primary aim is to relieve suffering.

Hospital ethics committees

Despite the growth of medical ethics and the publication of many professional codes of practice in recent years, it is still difficult for individuals to obtain guidance in resolving specific ethical dilemmas they face. Some hospitals have set up a hospital ethics committee or a clinical ethics forum to discuss these dilemmas.

Function of hospital ethics committees

- 1) **Education:** The committee should provide members of the hospital/medical staff with access to the language, concepts, principles and knowledge of ethics.
- 2) **Policy review and development:** the committee can assist the hospital and healthcare staff in the development of policies and guidelines regarding recurrent ethical issues and questions which arise in the care of individual patients.
- 3) **Case review:** the committee should be a forum for discussion of ethical questions that arise in the care of individual patients.

Appointment and membership

The committee should be multidisciplinary, and may include doctors, nurses, midwives, social worker, pastoral care, hospital director and chief of medical staff. A 30% membership from the general community has been suggested to ensure all views are represented.

Research ethics

Each year £35–40 billion is spent on healthcare research worldwide, but only 10% of this is aimed at the health problems of 90% of the world's population. This can lead to exploitation of people in the country where research is undertaken.

It is important that there are national guidelines in every country which set priorities for healthcare research.

There should be three main considerations for each research proposal:

- 1) relevance to healthcare priorities in that country
- 2) scientific validity
- 3) ethical acceptability.

There are several international guidelines and regulations with regard to research (for examples: World Medical Association, Council for International Organizations of Medical Science in collaboration with WHO, European Council and European Parliament), but these are often inappropriate for under-resourced countries.

Consent to research

For this to be valid, it should be given freely after full disclosure of all relevant information in a manner understandable by the research subject. Consent for the research may be withdrawn by the subject at any time without there being any change in other access to treatment or adverse effects on the subject. However, in some communities it is usual for male members of the family or a community to make decisions on behalf of women and children.

Level of care

The level of care provided to the control group (that is the group that is not having the active potential treatment) is controversial. Some argue that, if the research is externally sponsored, the people in the control group should receive the same standard of care as would be received in the sponsor's country. Others argue that this prevents some

research from being conducted. For example, if two treatments are being compared in the under-resourced country, it is more appropriate for the new treatment to be compared with the one currently available in that country, not one that is inaccessible there.

Post-research considerations

If an intervention is effective, should it be made available to the research participants and the community? The country concerned may not be able to afford this, but a decision should be made with the national government via its research ethics committee about what will happen after the trial period is over.

Healthcare worker relationships

As well as having an ethical duty towards patients, all healthcare workers have an ethical duty towards other healthcare workers, to the healthcare system, and to society. These are included in individual Ethical codes of Conduct for general and paediatric nurses, anaesthetic nurses, midwives and doctors and these Codes are generally similar.

The health worker–patient relationship

- 1) The healthcare worker's primary role is to be an advocate for each patient's care and well-being. They should always place the interests of their patients first. The healthcare worker also must accept responsibility for his or her clinical decisions.
- 2) The healthcare worker must treat each patient with honesty, compassion, dignity and respect. They should not exclude or discriminate against any patient because of ethnic origin, race, gender, age, socio-economic status, diagnosis, physical or mental disability, or sexual orientation.

The healthcare worker–healthcare worker relationship

Traditionally, doctors have been at the top of the caregiving hierarchy in the past, above nurses and other healthcare workers. This situation is gradually changing, with multidisciplinary input becoming more important in making decisions about patient care.

- 1) Healthcare workers have a responsibility to maintain moral integrity, intellectual honesty and clinical competence. They should be aware of the limitations of their expertise and seek consultation or assistance in clinical situations in which they are not expert.
- 2) Healthcare workers should work as a team, supporting each other and working together for the benefit of the patient.
- 3) Healthcare workers have an obligation to educate and share information with colleagues, including trainee healthcare workers.
- 4) There is an ethical obligation to report impairment or misconduct of colleagues to prevent potential harm to patients.

The relationship between the healthcare worker and the system of care

- 1) The healthcare worker's duty of patient advocacy should not be altered by the system of healthcare in which they practice.
- 2) If there are conflicts of interest, the patient's interests should take priority over those of others.
- 3) Healthcare professionals should campaign against unethical practices.
- 4) Healthcare professionals should not be influenced by commercial enterprises. The duty of the healthcare worker is to evaluate objectively what is best for the patient. Gifts designed to influence clinical practice are not acceptable.
- 5) Healthcare professionals should advocate to their departments of health for improved medical facilities and treatments for patients where acceptable basic facilities and treatments are lacking (for examples, oxygen, effective pain relief, blood transfusion services, access to vital skills such as surgery, basic life-saving drugs).

In many countries, there are huge divisions between the rich minority and the poor, exploited and disadvantaged majority. Healthcare professionals should be aware of this and aim to provide high standards of care independent of a patient's or family's ability to pay.

The relationship of the healthcare worker to society

Healthcare workers have a responsibility to society as well as to patients, and sometimes society's best interests may take precedence over those of the patient (for examples, mandatory reporting of patients with a designated disease, those who are unfit to drive motor vehicles, and those suspected of child abuse).

In other circumstances there may be requests from the police or the military to take part in practices that violate human rights (for example torture). Healthcare workers should report unjustified interference in the care of their patients, especially if fundamental human rights are being denied. If the authorities are not approachable or helpful, contacting a national medical or nursing association, the WMA or World Nursing Federation, or a human rights organisation may be needed.

The increasing mobility of society means that healthcare workers have a responsibility for global health, including preventing the spread of infectious diseases between societies and countries.

Another effect of globalisation is the mobility of healthcare professionals, and their migration from low-income to high-income countries. The shortage of healthcare workers is one of the biggest health problems facing low-income countries today.

The governments of low-income countries invest in the education and training of healthcare professionals, and therefore lose these resources and the contribution of these workers when graduates migrate. The factors considered by the migrant may be economic, social and/ or family related. Often in low-income countries there are low wages, poor working conditions, lack of leadership and very few incentives, as well as limited opportunities for their children. High-income governments encourage migration when there is a need, often with no compensation for the government where the migrant was trained. This presents an ethical dilemma whereby if emigration was prevented it would restrict the autonomy of the individual, but on the other hand the health of a society suffers if there is mass migration of healthcare professionals.

Codes of ethics for different healthcare professions and further reading

International Council of Midwives codes of ethics

Adopted at Glasgow International Council meeting, 2008 Reviewed and adopted at Prague Council meeting, 2014. Due for next review 2020

https://www.internationalmidwives.org/assets/files/general-files/2019/01/cd2008_001-eng-code-of-ethics-for-midwives.pdf Accessed March 31st 2021

The International Council of Nurses developed a code of ethics for the nursing profession in 1953. Last revised in 2012, due for next review 2020

https://www.icn.ch/sites/default/files/inline-files/2012_ICN_Codeofethicsfornurses_%20eng.pdf Accessed 31st March 2021

International Federation of Nurse Anaesthetists. First standards adopted 1991. Last revised and adopted 2016

https://www.ghdonline.org/uploads/IFNA_STANDARDS_FINAL_2016.pdf Accessed 31.03.2021

World Medical Association code of medical ethics First adopted 1949; revised and adopted 2006

<file:///Users/Admin/Downloads/wma-international-code-of-medical-ethics.pdf>

Accessed 31st March 2021

World Organization Against Torture

<https://www.kofiannanfoundation.org/speeches/world-organisation-against-torture/> Accessed 31.03.2021

Medical Foundation for the Care of Victims of Torture <https://www.freedomfromtorture.org/> (accessed 2nd March 2021).

Medical Ethics Manual 3rd edition 2015 – World Medical Association. www.wma.net/ publications (accessed 2 March 2021).

General Medical Council. GMC List of ethical guidance – General Medical Council. <https://www.gmc-uk.org/ethical-guidance> Accessed 31st March 2021

Nuffield Foundation. The Ethics of Clinical Research in Developing Countries. <https://www.nuffieldbioethics.org/publications/research-in-developing-countries> Accessed 31st March 2021

Naicker S, Plange-Rhule J, Tutt J et al. (2009) Shortage of healthcare workers in developing countries – Africa. *Ethnicity and Disease*, 19, S1 60–4.

<https://www.ethndis.org/priorsuparchives/ethn-19-01s1-60.pdf> Accessed 31.03.2021

Global Health Workforce Alliance (2011) Progress Report on Kampala Declaration and Agenda for Global Action.

<https://www.who.int/workforcealliance/knowledge/resources/kdagaprogressreport/en/> Accessed 31st March 2021.

Section 9 Medical records, history taking and clinical examination

- Records can be held by patients or parents, or by the hospital, or both.
- If they are patient or 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). Immunisation information, if included, should comply with national immunisation programmes.
- Hospital records need to be kept confidentially in a logical system for audit purposes, with easy access to previous notes.
- Discharge information and advice should be entered in the patient- or parent-held booklet.
- If possible, diagnoses should be coded and entered according to the International Classification of Diseases (ICD) or in accordance with local policy and coding.

History taking

- The medical history should, when age appropriate, include the child's own input. The source of the information may be the mother, the father, or the child him- or herself, and the source should be documented.
- It is important to listen, especially to the mother's worries about her child, considering her general frame of mind, her 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 the following:
 - pregnancy and previous deliveries (including stillbirths)
 - infant or young child feeding history
 - the immunisation record (best kept by the parents)
 - previous admissions or visits to hospital
 - existing medical problems
 - social circumstances at home, and the family history
 - the family's cultural beliefs and their religion and/ or tribe
 - medication taken by the patient, and any allergies
 - the patient's presenting complaints and current treatment, if any.
- Most patients and their families are anxious. They need reassurance, kindness and understanding.

Examination

The following basic equipment is required:

- stethoscope
- otoscope (if available)
- ophthalmoscope (if available)
- tendon hammer
- bright torch light (or mobile phone light)
- thermometer
- Pinard's stethoscope or Sonicaid (a hand-held Doppler)
- microscope (if available).

Conducting the examination

- A triage nurse (see Section 1 Handbook 1) can be helpful for making a preliminary assessment of patients. They can assess each patient and use the recorded body temperature, weight, general condition, and pain score of the patient to decide how urgently he or she should be seen by the doctor.
- Do not rush the examination. A thorough examination is often needed and taking time can help to gain the confidence of the patient and their family.
- If the patient 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 has been done. You want to avoid having a crying patient whom you cannot examine or auscultate.
- Small children and infants are best examined on the parent's lap; older 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 emergency examination checklist

Always check the following in the order shown:

- Airway and Bleeding needing immediate action
- Breathing
- Circulation
- Disability
- Exposure.

In the case of a critically ill patient, proceed to basic and/ or advanced life support using the structured approach (see Sections 11-13).

Patients who are not in need of immediate resuscitation

1. Introduce yourself to the patient and parent, if present.
2. Interact with any child throughout the examination.
3. General inspection: document dysmorphism, skin rashes or bruises, nutritional status, weight and height for age, jaundice, pallor, clubbing, (for child) relationship with parent, and state of consciousness.
4. Respiratory system: document chest wall expansion (is it symmetrical? is there recession?), respiratory rate, cyanosis, palpation, percussion, and auscultation.
5. Cardiovascular system: remember to feel all the pulses, particularly the femoral pulses. Measure the blood pressure (the cuff must cover two-thirds of the upper arm circumference), examine the jugular venous pressure, palpate the cardiac impulses (i.e. for left and right ventricles), and auscultate the apex, left sternal edge, pulmonary and aortic areas and carotids and over the back.
6. Abdominal system: if the patient is pregnant, assess the size of the uterus, the presentation of the fetus and listen for the fetal heart.
7. In an infant check the genitals for cryptorchidism, hernias and gender. Rectal examinations are occasionally necessary but need to be explained to the patient, parent and child (where appropriate). Inspect the mouth and teeth.
8. Neurological system: use the AVPU or Glasgow Coma Scale score (see Section 66 Handbook 1). Observe infants for their degree of responsiveness and rapport appropriate for age, social and motor skills, and 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.
9. Motor system: Always examine infants for dislocated/ dislocatable hips. Check the gait.
10. Urine: Test for protein, glucose, and blood, and ideally for infection using a microscope or appropriate stick tests.

Patients and parents have the right to be told any abnormal findings, and the actual process of the examination should be explained to the patient in age-appropriate language.

The history and examination findings, including the patient's weight and height, should be recorded, with daily entries on management and progress. (Be aware of the local guidelines on nutritional assessments, especially in settings where malnutrition is common.) When the

patient is discharged, they should be given discharge information about the admission and any further treatment and advice that needs to be shared with their primary care healthcare workers.

See Section 2 Handbook 1 and Appendix International Maternal & Child Health Care for examples of various charts, including those for vital signs, fluid balance, growth (2014) https://12dec1c3-e3f4-e784-7f3a-6e7903610962.filesusr.com/ugd/dd2ba4_c5284b1b150f4d70a72fe1cd710d530d.pdf (Accessed 31.03.2021)

Section 10 Traditional medicine and its relevance to hospital care and its dangers

Introduction

Traditional medicine comprises of diverse health practices, remedies, approaches, knowledge and beliefs incorporating plant, animal and mineral products, spiritual therapies, charms, manual techniques, exercises, and in fact any kind of salutary method applied singly or in combination to diagnose, treat and prevent illnesses or maintain well-being, which has been handed down by the tradition of a community or ethnic group. In contrast with conventional medicine, which focuses on experiment and disease-causing pathogens, traditional medicine claims that the human being is both a somatic and spiritual entity, and that disease can be due to supernatural causes arising from the anger of ancestral or evil spirits, the result of witchcraft, or the entry of an object into the body. It is therefore not only the symptoms of the disease that are taken into account, but also psychological and sociological factors. Thus, the holistic nature and culture-based approach to traditional healthcare is an important aspect of the practice and sets it apart from conventional western approaches.

World Health Organization (WHO) has described traditional medicine as the overall knowledge, skill and practices based on the theories, belief and experiences particularly to the different cultures used in the maintenance of health as well as prevention, diagnosis, improvement or treatment of physical and mental illness.

Traditional medicine is culturally treasured by various communities around the world. It thus plays an almost immeasurable role in healthcare delivery to the people. WHO estimate that traditional birth attendants assist in a majority of births in many African countries. Traditional medicine has shown in the past quality, safety, and efficacy which attribute to the purpose of ensuring that all people have access to care. Herbal medicines, traditional treatments, and traditional practitioners are the main source of health care for many people and sometimes the only source of care that is close to homes, accessible and affordable. It is also culturally acceptable and trusted by large numbers of people. The affordability of most traditional medicines makes them all more attractive at a time of seeking health-care and comfortable to the users.

Despite the promotion of biomedicine by international healthcare organizations, traditional medicine remains the primary form of healthcare for more than 80% of African populations. Traditional medical systems include not only traditional healers, but also the popular knowledge of local populations, known as domestic medicine or home remedies. Most rural and urban dwellers often supplement treatment by orthodox medical practitioners with treatment by traditional healers.

In Ghana, Mali, Nigeria and Zambia, it has been found that the first-line treatment for 60% of children with high fever from malaria is the use of herbal medicines at home. Cough and abdominal pains are other symptoms of children that are frequently treated with herbal medicine in Nigeria. Traditional medicine is extensively used in Latin America and Asia. In China, 30-50% of all healthcare is delivered by traditional health practitioners.

Nigerians are known to communicate with powerful cultural and religious beliefs and practices relating to health. Approximately 85% of the population use traditional medicine and consult its practitioners for healthcare. The majority (70%) of Nigeria's population is rural and relies almost exclusively on traditional medicine for its healthcare needs. The popularity of traditional medicine has been attributed to poverty, limited or no access to good quality orthodox medicine, illiteracy and ignorance. Other factors include affordability, availability, efficacy, costly or inefficient orthodox medical facilities, unfriendliness of hospital staff, poor communication (e.g. patients not being told the nature and cause of their illness), inadequate technical services leading to poor-quality care, treatment that is divorced from the patient's

culture, family and community, and the treatment only addressing biological aspects of the illness rather than also addressing spiritual aspects.

The **traditional healer**, as defined by the WHO (1976), is a person who is recognised by the community in which they live as being competent to provide healthcare by using vegetable, animal and mineral substances and certain other methods based on the social, cultural, and religious background, as well as the knowledge, attributes and beliefs that are prevalent in the community, regarding physical, mental and social well-being and the causation of disease and disability. They rely exclusively on practical experience and observations handed down from one generation to the next, whether verbally or in writing. For most countries of the world, a traditional healer may be able to perform many functions, thus being more versatile as a healer.

The elements of traditional medicine include, among others, herbal medicine, massage, homeopathy, mud baths, music therapy, wax baths, reflexology, dance therapy, hydrotherapy, mind and spirit therapies, self-exercise therapies, radiation and vibration, osteopathy, chiropractic medicine, aromatherapy, preventive medicine, radiant heat therapy, therapeutic fasting and dieting, spinal manipulation, and psychotherapy.

Relevance of traditional medicine

Many communities have developed various traditional systems using locally available resources for the alleviation of their health problems. This has resulted in the appearance of several different categories of healers, and a variety of healing methods, strategies and medicines or remedies. Most people who live in rural communities do not have access to orthodox medicine. For example, in Nigeria it is estimated that about 75% of the population still prefer to solve their health problems by consulting traditional healers. Furthermore, many rural communities have great faith in traditional medicine, particularly its inexplicable aspects, as they believe that it represents the wisdom of their forefathers which also incorporates their socio-cultural and religious background, which orthodox medicine seems to neglect. Although herbal medicines may have beneficial active ingredients, the dosage cannot be controlled as there is no assay system for defining potency, and this increases the risk to patients who receive such treatment.

In environments where illness is believed to have a magical/spiritual origin, people become involved in intense prayers and sacrifices to compensate for their frailty and powerlessness. Western explanations of illness are rarely taken seriously. Local people may embrace traditional medicine to the exclusion of all other approaches, or combine it with orthodox medicine. Adverse drug interactions may result from such combinations of approach. Medical practitioners working in places where traditional medicine is practised must be patient and respectful in their encounter with patients who are using traditional approaches. With health education and therapy that bring real health benefits, local people will become persuaded to accept effective evidence-based treatment.

Although the disadvantages of traditional medicine are numerous, it does also have a few advantages. The traditional healers and their drugs are available in these communities and their drugs are relatively easy to obtain compared with those of orthodox medicine. The healing system cares for the body, mind and soul of the patient in the context of the family, community, God or gods. The relationship between the practitioner and the patient can be close, encouraging and intense, with active participation of the family and neighbours. The practitioners are well known, trusted and respected in the community, and their methods fit very well with the culture and customs of the people. The drugs are cheap and readily available, and the healers accept payments either as a whole, in part or in kind, which also makes their treatments much more accessible for the people.

Traditional healers

There are various categories of traditional healers. Some of them may have areas of special interest.

Herbalists

An herbalist is a person who specialises in the economic or medicinal uses of plants. The whole plant may be used or parts of the plant, including the whole root, root bark, whole stem, stem bark, leaves, flowers, fruits, and seeds, which may be administered to the patient in the following forms:

- A powder that can be swallowed or taken with pap/ traditional porridge (cold or hot) or any drink
- A powder that is rubbed into cuts made on any part of the body with a sharp knife
- A preparation that is soaked for some time in water or local gin and decanted as required before drinking; the materials could also be boiled in water, cooled and strained.
- A preparation that is pounded with native soap and used for bathing; such 'medicated soaps' are commonly used to treat skin diseases
- Pastes or ointments, in a medium of palm oil or shea butter
- Soup which is consumed by the patient
- Herbal preparations may also be administered as an enema.

Herbal medicine is the use of only plants for medicinal and therapeutic purpose to treat the diseases and to improve human health. The plants are gathered from the environment and are therefore part of every cultural tradition and have helped the development and growth of herbalism. Some of the plants that are facing extinction due to drought, bush burning, rapid growth of communities, farming or other factors are specially cultivated by some herbalists to maintain a steady source of supply.

The use of herbal remedies has increased globally, it is predicted that this rate will be greater for the COVID-19 pandemic process. WHO reported nearly ~80% of the world's population uses and trust herbal products for treatment and about 2.9 million American children and teenagers have used herbs or their supplements. In China, the use of herbal medicine is changing from around 30–50% of the total drug consumption. In other developed countries, it is estimated that, more than 50% of the population use herbal products at least once in their life. The herbal medicines account for 60% of treatment at home in developing countries. In the children with a chronic illness or among inpatients and outpatients are higher use of the herbal medicine.

Families with children who have chronic medical conditions, such as autism, cystic fibrosis, rheumatoid arthritis, respiratory tract infections or asthma use herbal remedies as part of their treatment. Parents of children with asthma reported using a range of herbal products for self-care. The most common used herbal medicine for paediatric asthmatic patients were linden and ginger. Herbal medicine has traditionally been used in the treatment of symptoms for nocturnal enuresis or urinary incontinence.

The ginger, chamomile, mint, cardamom, garlic, and onion are used to prevent and treat nausea caused by chemotherapy. It has been reported some herbal products are effective in the management of ear pain in Otitis Media. Children with Attention Deficit Hyperactivity Disorder and Anxiety or Depression take herbal products a part of their treatments. The use of herbal medicine in children with medical comorbidities, excessive sleep problems or insomnia is 1.8 times higher than children without such difficulties.

Traditional birth attendants (TBAs)

A traditional birth attendant assists the mother at childbirth, and initially acquired her skills delivering babies by herself or by working with other birth attendants. TBAs are predominantly female. For example, around 60–85% of childbirth in Nigeria is overseen by

TBAs, especially in the rural communities. They therefore occupy a prominent position in the healthcare system. Their skills are wide ranging, including diagnosis of pregnancy, antenatal care, conduct of labour and postnatal care. They are quite acceptable to those living in rural communities because their practice is linked to socio-cultural practices. For this reason, some governments have started to train TBAs to reduce maternal and child morbidity and mortality.

MCAI introduced a training program in emergency care in The Gambia linking TBAs to the local district hospitals and including the provision of mobile phones and a vehicle provided by the President to transfer pregnant women with complications to the hospital during which time they were accompanied by their TBA (see Section 67 for the training manual developed for and a summary of this project).

Traditional bone setters

Traditional bone setters are knowledgeable in the art and skill of setting broken bones in the traditional way, using their skill to ensure that the bones unite and heal properly. They are involved in setting various types of fractures using wooden splints made from bamboo plants, and they use dry fibre from banana stems as bandaging. Wounds resulting from such fractures are usually cleaned and bleeding is stopped by the application of plant extracts. Some practitioners fracture similar bones in a bird and treat it alongside the fractured limb of the patient. This is used to determine the time that it will take for the patient's fracture to heal, and the correct time for removing the wrapped splints and clay cast. Importantly, some bone setters collaborate with orthodox medical practitioners who treat the open wounds, offer radiological services, and give advice on cases that require referral. This may help to reduce the number of complications occurring in their practices.

Traditional surgeons

These practitioners undertake minor surgery. The procedures that they perform include the cutting of tribal marks, male circumcision and female genital mutilation, ear piercing, and incision and drainage of abscesses, to name just a few. Complications such as haemorrhage, tetanus and sepsis have been reported in their practices.

Traditional psychiatrists

The traditional psychiatrist specialises mainly in the treatment of patients with mental disorders especially children with attention deficit disorder.

Those who are diagnosed as demon possessed are usually caned or beaten into submission and then given herbal hypnotics or highly sedative herbal potions to calm them. Such herbal preparations include extracts of the African *Rauwolfia* species. Treatment and rehabilitation of people with mental disorders usually take place over a long period of time. Incantations and various forms of occultism are often employed.

Practitioners of therapeutic occultism

These are traditional practitioners who use supernatural or mysterious forces, incantations, or prescribed rituals associated with the community's religious worship, and they adopt various inexplicable methods to treat a range of diseases. They are usually respected within the community because of their ability to deal with unseen and supernatural forces. They are regarded as witches and wizards.

Complications of traditional medicine

Contrary to popular opinion that traditional medicine, especially herbal medicine, is natural or safe, it can be hazardous to health if these preparations are taken in recommended or larger amounts, injected or combined with prescription drugs. Some Asian herbal products have been found to contain potentially dangerous concentrations of harmful substances such as arsenic, mercury and lead, many of which cause liver failure, haemorrhage, or heart failure.

Where confidence in conventional medical care is low, there is a tendency to resort to more risky traditional remedies which may be more toxic. Conversely, when confidence in conventional medical care improves, there is an increasing movement towards the use of less toxic remedies, even though the use of some traditional remedies may continue to satisfy cultural and social needs.

In many low-income countries, patients are subjected to traditional treatment as first aid therapy in emergency conditions at home. Caregivers may apply interventions that are ineffective, harmful, and have no pathophysiological basis. The application of traditional medical care that is ineffective may also lead to delayed presentation of potentially curable conditions to conventional care, resulting in unnecessary deaths and morbidity.

Some cultural practices are harmful to the health and survival of the new-born infant, and it is often young 'first-time' mothers who are most likely to follow these practices. The cultural practices involving giving a new-born infants cold baths, discarding colostrum, and providing food other than breast milk soon after birth are the common practices. The application of butter, ash or other substances, such as cow dung, to the umbilical stump increase the risk of life-threatening infection.

Examples of problems resulting from traditional medicine practices

Experience of the use of traditional medicine in Nigeria

In Nigeria, as in most other developing countries, children are subjected to unorthodox treatment as first aid therapy in emergency conditions at home. Caregivers may apply interventions that are ineffective, harmful, and have no pathophysiological basis. The use of traditional medicine is largely ethnocentric.

Crude oil

Crude oil is available in large amounts In the Niger Delta region of Nigeria. It is highly regarded locally as a remedy for a variety of ailments, including febrile convulsions, gastrointestinal disorders, burns, 'foot rot' and leg ulcers, and poisoning. It is also used in witchcraft. The oil is applied to the skin, mixed with alcohol or water as a drink, and instilled into body orifices such as the nostrils, ears, anus, vagina and urethra. The use of crude oil as traditional medicine in Nigeria has been reported to have an analgesic effect comparable to that of aspirin. Complications associated with its use have been reported in children with febrile convulsions.

Complications caused by crude oil have been reported to affect several organs, including the skin, lungs, liver and kidneys. Skin exposure may result in the formation of vesicles, blisters and even extensive epidermolysis. Ingestion of crude oil for treatment among children may result in nausea, vomiting and diarrhoea, and the aspiration of crude oil during vomiting results in chemical pneumonitis. Central nervous system symptoms range from vertigo and headache caused by ingestion of small doses, to lethargy, convulsions, coma and death with larger doses. Renal failure has been described as another toxic effect.

Cow's urine concoction

'Cow's urine' concoction (CUC) is a traditional medicine used in the management of convulsive disorders in childhood among the Yoruba speaking people of south-western Nigeria. It is prepared from leaves of tobacco, garlic and basil, lemon juice, rock salt and onion bulbs, which are soaked in cow urine, which acts as the vehicle in which the active principles of these constituents dissolve. Over 50 chemical compounds have been identified in CUC, the major ones being benzoic acid, phenylacetic acid, p-cresol, thymol and nicotine. These components are toxic and have harmful effects on the different systems of the body. The main effects are severe respiratory depression, effects on the cardiovascular system

and the central nervous system, and hypoglycaemia. These toxic effects acting singly or in combination are believed to be the cause(s) of death from CUC.

Cow dung

Globally, 60 million births occur outside healthcare facilities. Childbirth occurs in places where hygienic practices may be sub-optimal and it is estimated that 30–40% of infections resulting in deaths from neonatal sepsis are transmitted at the time of childbirth and have early onset of symptoms (developing during the first 72 hours after birth).

The unhealed umbilical cord is an important entry point for local and invasive infections during the neonatal period. It is rapidly infected by bacteria from the maternal genital tract and then from the environment. Infection can emanate from the bamboo stick that is used to cut the umbilicus, and from the cow dung (believed to have desiccating properties) that is used to dress the umbilical stump. Localised umbilical infection (omphalitis) emanating from these sources can spread to the abdominal wall, the peritoneum, or through the umbilical or portal vessels leading to systemic sepsis, which if untreated has a high fatality rate. Neonatal tetanus is a very important complication resulting from these practices, which are common among the Yoruba of south-western Nigeria and the Maasai people of Kenya. Cow dung is also used to anoint the heads of the sick among the Maasai people.

Traditional eye medications

The use of traditional eye medications among children revealed corneal opacities, staphyloma and corneal ulcers in 55% of the individuals studied. Other complications were pan-ophthalmitis, endophthalmitis, uveitis, cataract, and bullous keratopathy. Eleven individuals in one study underwent enucleation of the affected eye.

Traditional healers tend to prefer to use substances that cause irritation and pain, as these are perceived by both healers and patients to be more potent. Such substances may be acidic or alkaline, resulting in ocular burns. No particular attention is paid to concentration and sterility, as most of these concoctions (mixture of various substances, which may be plant or animal extracts) are prepared without regard for hygiene, including the use of contaminated water, local gin, saliva and even urine.

Most of these ocular conditions could have been adequately treated using standard medicines, which, were sometimes available.

Experience of traditional medicine in the Eastern Cape area of South Africa.

In another publication, traditional remedies were found to be regularly used in the home management of children in the Eastern Cape, and probably in the great majority of cases these remedies do little harm beyond delaying presentation to the healthcare system. However, serious effects were occasionally identified. Most often the traditional remedy was given to treat a symptom of an underlying disease, rather than being the cause of the condition or symptoms.

iYeza lo moya: commonly given to infants by mouth, with few problems reported. However, a traditional enema may also be given, which may have more toxic effects.

Senecio extracts: infusions of this weed with yellow flowers have been reported to cause veno-occlusive disease in a small number of children.

Impila: extracts from this root may cause fatal hepato-renal failure, often presenting with hypoglycaemia.

River onion: this is used both orally and rectally and causes hepato-renal failure in a significant number of children.

Jeyes fluid: this is sometimes added to rectal and oral remedies and causes local and systemic effects.

Surgical complications of traditional medicine in East Africa

A series of case histories in 2007 included a 6-year-old girl sustaining a spiral fracture of the humerus during a road traffic accident. The parents refused hospital treatment and took her to a traditional bone setter. Two weeks later she was brought back to the hospital with a gangrenous upper limb, which was the result of placing a tourniquet around the axilla. Debridement was undertaken but the child lost the whole of the skin of the forearm and most of the hand.

A second case involved an 18-month-old boy who underwent circumcision by a traditional practitioner. On subsequent admission to hospital, he was found to have partial amputation of the glans penis.

Management of suspected adverse effects of traditional medicine

These include a rapid assessment of:

Airway

Breathing

Circulation

Disability.

Regular assessment and treatment of these essential systems will ensure that management keeps abreast with progress and with the prevention of deterioration. However, when treating patients who have been given traditional medicines, first look for a medical cause of the symptoms and signs before assuming that the illness is due to the traditional remedy.

Table 10.1 Serious complications caused by traditional medicines, and their management

System affected	Symptoms and signs	Treatment
Cardiovascular	Increased cholinergic actions such as lacrimation, salivation, rhinorrhoea, diarrhoea, vomiting and miosis, severe bradycardia or heart block	Intravenous atropine may help
	Anticholinergic actions such as hyperthermia, tachycardia or tachyarrhythmias, mydriasis, constipation or acute urinary retention	Anticholinesterase drugs may help
Neurological	Weakness, epileptic fits, coma and intracranial bleeding	Check blood clotting Anticonvulsants
Pulmonary	Anaphylaxis, bronchoconstriction Severe interstitial pneumonitis, non-cardiac pulmonary oedema, acute eosinophilic pneumonia	Specific treatment for anaphylaxis and bronchoconstriction (see Sections 35-36 Handbook 1) Corticosteroids
Liver toxicity	Nausea, anorexia, vomiting, jaundice with elevated liver transaminases	Supportive treatment

System affected	Symptoms and signs	Treatment
Nephrotoxicity	Acute renal failure and tubular dysfunction	Supportive treatment
Heavy metal contamination with lead, arsenic, thallium or uranium	Gastrointestinal disorders, hepatitis, polyarthritis, encephalopathy (including ataxia and severe psychiatric disturbances)	See Section 87 Handbook 1

Clean any areas that are visibly affected with a topical application of sterile water and apply a non-adhesive dressing if necessary.

Laboratory investigations can be helpful for identifying organ systems that may be affected by a toxic traditional medicine. Take blood for a biochemical profile (urea and electrolytes, liver function tests, amylase and glucose) and a full blood count with indices. If there is any significant abnormality, refer the patient to the relevant specialist team.

Symptoms and signs such as convulsions should be treated with injection of benzodiazepines (see Section 70 Handbook1). Hypoglycaemia should be corrected with glucose infusion (Section 51 Handbook 1), and fluid and electrolyte disturbances should be corrected with appropriate oral administration or intravenous infusion (see Sections 7, 60 and 61 Handbook 1)

Appropriate antibiotics should be administered to patients with infective conditions.

Conclusion

Traditional medicine continues to represent a very large component of community healthcare, especially in resource-limited regions. Efforts to make traditional medicine safer are urgently required and might include official regulation to monitor the activities of traditional practitioners, standardise their practices and undertake toxicity studies on their products, in collaboration with scientists and recognised institutions. However, this will also require the traditional healers to be willing to work with such control of their practice, which could be a problem if the healers see this as an attempt to limit their practice, or to steal their secrets and remedies.

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Approach to emergencies

Training

Members of the clinical team must know their roles. They will ideally have trained together in:

- clinical situations and their diagnoses and treatments
- drugs and their use, administration and side effects
- emergency equipment and how it functions.

The ability of a facility to deal with emergencies should be assessed and reinforced by the frequent practice of emergency drills.

Initial management

1. Stay calm.
2. Do not leave the patient unattended.
3. Have a team leader in charge to avoid confusion.
4. Shout for help. Ask one person to go for help and another to get emergency equipment and supplies (e.g., oxygen cylinder and emergency kit). Ideally resuscitation equipment and drugs should be available on one dedicated trolley.
5. Assess and resuscitate in sequence using the structured approach – Airway, Breathing, Circulation, Disability (Neurological Status) (see below).
6. If the patient is conscious, ask what happened and what symptoms they have.
7. Constantly reassess the patient, particularly after any intervention.

Structured approach to any infant or child presenting as an emergency

Approach emergencies using the structured ABCD (Airway, Breathing, Circulation, Disability) approach, which ensures that all patients with a life-threatening or potentially life-threatening problem are identified and managed in an effective and efficient way whatever their diagnosis or pathology.

The structured approach to the seriously ill patient, which is outlined here, allows the health worker to focus on the appropriate level of diagnosis and treatment during the first hours of care.

Primary assessment and resuscitation are concerned with the maintenance of vital functions and the administration of life-saving treatments, whereas secondary assessment and emergency treatment allow more specific urgent therapies to be started.

Secondary assessment and emergency care require a system-by-system approach in order to minimise the risk of significant conditions being missed.

Following cardiac and/or respiratory arrest, the outcome for children is poor. Earlier recognition and management of potential respiratory, circulatory, or central neurological failure which may progress rapidly to cardiac and/or respiratory arrest will reduce mortality and secondary morbidity. The following section outlines the physical signs that should be used for the rapid primary assessment, resuscitation, secondary assessment and emergency treatment of infants and children.

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Primary assessment and resuscitation involves sequential assessment and resuscitation of vital functions Airway, Breathing and Circulation.

If there are no life-threatening signs, the primary assessment can be completed within less than 1 minute. If life-threatening signs are identified, resuscitation procedures are required.

If you are working on your own and have been unable to summon help, you must resuscitate Airway before Breathing, and Breathing before Circulation. This is because oxygen cannot be carried around in the blood to the vital organs if the blood is not oxygenated first, and the lungs cannot oxygenate the blood if there is no airway to allow air containing oxygen to enter the lungs.

If assistance is available, one person can deal with Airway, another with Breathing and a third with Circulation, all working simultaneously, but there must be a 'team leader' to take overall control.

During resuscitation, interventions that are either lifesaving or designed to prevent the patient reaching a "near-death" situation are performed (see below). These include such procedures as basic airway opening procedures, suction, oropharyngeal airway insertion, intubation, assisted ventilation, venous cannulation, and fluid resuscitation (when safe and appropriate). At the same time, **oxygen** is provided to all patients with life-threatening Airway, Breathing or Circulatory problems, vital signs are recorded, and essential monitoring is established.

This sequential primary assessment and any necessary resuscitation occur before any illness-specific diagnostic assessment or treatment takes place. Once the patient's vital functions are working safely, secondary assessment and emergency treatment can begin.

After each intervention, its effects should be tested by reassessment. **Regular reassessments** are a key component of the structured approach.

During secondary assessment, illness-specific pathophysiology is sought and emergency treatments are instituted. Before embarking on this phase, it is important that the resuscitative measures are fully under way. During the secondary assessment, vital signs should be checked frequently to detect any change in the patient's condition. If there is deterioration, primary assessment and resuscitation should be repeated in the 'Airway, Breathing, Circulation' sequence.

Primary assessment and resuscitation

Assessment and resuscitation occur at the same time. The order of assessment and resuscitation enables identification of immediately life-threatening problems, which are treated as they are found.

A rapid examination of vital ABC functions is required. **If at any stage a life-threatening A, B, or C problem is identified:**

FIRST CALL FOR HELP.

After ABC, always assess for neurological problems, and resuscitate their components (sometimes referred to as 'D' for disability of the ABC approach).

Primary assessment and resuscitation of airway

The first priority is establishment or maintenance of airway opening. If there is a need for resuscitation in a patient who is bleeding (e.g. in cases of massive trauma), try to stop this at the same time as you are opening the airway.

PRIMARY ASSESSMENT

LOOK – for chest or abdominal movement.

LISTEN – for breath sounds.

FEEL – for breath. Talk to the patient.

A patient who can speak or cry has a clear airway.

Signs associated with airway obstruction may include any of the following:

- an absence of breathing
- stridor, snoring, or gurgling in the throat
- cyanosis
- chest wall recession
- agitation, reduced consciousness, or coma.

Be alert for foreign bodies (see Section 32 Handbook 1 on choking).

Airway obstruction in an unconscious patient is most commonly due to obstruction by the tongue.

Resuscitation of the airway

Open the airway and keep it open.

If there is no evidence of air movement, open the airway using the following:

1. a head tilt, chin lift or jaw thrust manoeuvre (see Section 12 on basic life support). If this opens the airway and breathing starts, keep the airway open manually until it can be secured. Be careful when using head tilt if the cervical spine is at risk, but opening the airway is always the priority
2. suction/removal of blood, vomit or a foreign body.

If there is no improvement after adjusting the airway manually and trying different techniques, place an oropharyngeal airway, which may be helpful if the patient is unconscious and has no gag reflex. Do NOT insert a nasopharyngeal airway if there is any suspicion of base of skull injury.

If the airway is still not open, a definitive airway by intubation or surgical airway will be needed.

Give **oxygen** to all patients.

Be careful not to distress young children with partial upper airway obstruction due to infections such as epiglottitis and severe croup, as this may precipitate crying and acute worsening of their airway obstruction. Having a parent or other known adult present will help to keep the child calm.

Identify the 'at-risk' airway

Reassess the airway after any airway-opening manoeuvres. If there continues to be no evidence of air movement, then airway patency can be assessed by performing an airway-opening manoeuvre while giving rescue breaths. Proceed to Breathing (see below).

Advanced airway management (always call for an anaesthetist)

Advanced airway management techniques for securing the airway by intubation may be required in patients with any of the following:

1. persistent airway obstruction
2. altered level of consciousness, with failure to protect the airway, especially from vomiting

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3. facial trauma, including burns, penetrating neck trauma with expanding haematoma, and severe head injury (see Sections 79 and 81 Handbook 1).

Intubation should be performed by skilled professionals such as an anaesthetist (if available) (see Section 60 for details). The following sequence should be followed:

1. pre-oxygenation with 100% oxygen with manual lung inflation if required
2. administration of a carefully judged, reduced dose of an anaesthetic induction agent
3. suxamethonium 1–2 mg/kg
4. intubation with a correctly sized tracheal tube.

Confirmation of correct placement of the endotracheal tube

Signs such as chest movement and auscultation remain helpful, but are occasionally misleading, especially in inexperienced hands. The most important sign 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 (measuring expired carbon dioxide), if available, is a useful adjunct for helping to confirm correct tube placement.

If it is not possible to provide an airway using intubation, a surgical airway may be required.

NOTE: It is extremely risky to proceed to Circulation (and IV/IO cannulation) when partial upper airway obstruction is present in young children (e.g., due to epiglottitis, severe croup or a foreign body), as invasive procedures can precipitate complete airway closure. Stabilise the airway first. This will require help from an anaesthetist.

Emergency treatment situations

1. For severe croup, nebulised adrenaline can be helpful (5 mL of 1 in 1000). Always give oral steroid as soon as possible (150 micrograms/kg of dexamethasone or 1 mg/kg of prednisolone).
2. For upper airway obstruction due to anaphylaxis, nebulised adrenaline (5 mL of 1 in 1000) and IM adrenaline (1 mg IM in pregnancy and 10 micrograms/kg in children).
3. Inhaled foreign body (see Section 32 and 90 Handbook 1).
4. For severe bronchiolitis, clear the nasal airways by using gentle suction.

If the patient has major trauma and is obviously bleeding rapidly, to the point of exsanguination (see Section 45 Handbook 2 and 92 Handbook 1), measures to stop the exsanguination must be instituted at the same time as Airway resuscitation. Throughout primary assessment and resuscitation, protect the cervical spine with a collar, sandbags and tape if the patient is likely to have an unstable cervical spine and if subsequent surgical stabilisation is possible (see Section 58)

Primary assessment and resuscitation of breathing

An open airway does not guarantee adequate ventilation. The latter requires an intact respiratory centre and adequate pulmonary function augmented by coordinated movement of the diaphragm and chest wall.

Primary assessment

Assess whether breathing is adequate by:

1. assessing effort:
 - a. recession
 - b. rate
 - c. added noises

- d. accessory muscles
- e. alar flaring
- 2. assessing efficacy:
 - a. listening for reduced or absent breath sounds, or any wheezing, with a stethoscope or ear on chest wall
 - b. chest and/or abdominal expansion (symmetrical or asymmetrical)
 - c. abdominal excursion
 - d. SaO₂ if available
- 3. assessing effects on heart rate
- 4. assessing effects on skin colour (check the possibility of cyanosis)
- 5. assessing effects on mental status.

Evidence of life-threatening respiratory difficulty

This includes the following:

- 1. absence of breathing (apnoea)
- 2. very high or very low respiratory rates
- 3. gasping, which is a sign of severe hypoxaemia, and may indicate impending respiratory arrest and death
- 4. severe chest wall recession, usually with increased respiratory rate, but pre-terminally with a fall in rate
- 5. severe hypoxaemia (cyanosis)
- 6. signs of tension pneumothorax (respiratory distress with hyper-resonant percussion) (see Sections 79 and 91 Handbook 1)
- 7. major trauma to the chest (e.g., tension pneumothorax, haemothorax, flail chest) (see Sections 79 and 91 Handbook 1)
- 8. signs of severe asthma (severe respiratory distress with wheezing, **but a silent chest in severe asthma** can be a near-fatal situation).

Evidence of respiratory difficulty which can progress if not treated

This includes the following:

- 1. increased respiratory rate
- 2. inspiratory stridor
- 3. reduced or absent breath sounds on auscultation
- 4. expiratory wheezing
- 5. chest expansion (most important), and reduced abdominal excursion
- 6. pulse oximetry showing oxygen saturation (SpO₂) of less than 94% (allowing for a 1% error in pulse oximeters normal SpO₂ in a patient at sea level is 95–100% in air).

Fast breathing is caused by either an airway problem, lung disease or metabolic acidosis.

TABLE 11.1 Respiratory rates ‘at rest’ for different age groups

Age (years)	Respiratory rate (breaths/minute)
< 1	30–40
1–2	25–35
2–5	25–30
5–12	20–25
> 12	15–20

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Care should be taken when interpreting single measurements. Infants can show rates of between 30 and 90 breaths/minute depending on their state of activity. It is more useful to use trends in measurements as an indicator of improvement or deterioration.

WHO definitions of fast breathing in young children are as follows:

- < 2 months: ≥ 60 breaths/minute
- 2–12 months: ≥ 50 breaths/minute
- 12 months to 5 years: ≥ 40 breaths/minute

Slow breathing rates may result from fatigue or raised intracranial pressure or may immediately precede a respiratory arrest due to severe hypoxaemia.

Other signs of breathing difficulty

Chest wall recession

- Intercostal, subcostal or sternal recession reflects increased effort of breathing, which is seen in particular in infants, who have more compliant chest walls.
- The degree of recession indicates the severity of respiratory difficulty.
- In the patient with exhaustion, chest movement and recession will decrease.

Inspiratory or expiratory noises

1. Stridor, usually inspiratory, indicates laryngeal or tracheal obstruction.
2. Wheeze, predominantly expiratory, indicates lower airway obstruction.
3. Volume of noise is not an indicator of severity.

Grunting

- This is observed in infants and children with stiff lungs to prevent airway collapse (it represents the noise made by closure of the larynx during expiration, which is the body's attempt to increase lung volume).
- It is a sign of severe respiratory distress.

Accessory muscle use

- In infants, the use of the sternocleidomastoid muscle creates 'head bobbing' and does not help ventilation.
- Flaring of the alae nasi is also seen in infants with respiratory distress.

Exceptions

Increased effort of breathing does not occur in three circumstances:

1. exhaustion
2. central respiratory depression (e.g., from raised intracranial pressure, poisoning or encephalopathy)
3. neuromuscular disease (e.g., poliomyelitis).

Effects of breathing failure on other physiology

Heart rate: this is increased with hypoxia, but decreases when hypoxia is severe, when bradycardia is a sign of impending cardiorespiratory arrest.

Skin colour: hypoxia first causes vasoconstriction and pallor. Cyanosis is a late sign and may indicate impending cardiorespiratory arrest. In an anaemic patient it may never be seen, however hypoxic the patient is.

Mental status: hypoxia causes initial agitation, then drowsiness, followed by loss of consciousness.

Resuscitation of breathing

Always give high flow oxygen (10-12 litres /minute using face mask). Give as much oxygen as possible through a mask with a reservoir bag to any patient who is breathing but has respiratory difficulty or the other signs of hypoxia (e.g., cyanosis).

In the patient with **absent or inadequate breathing**, it is essential to breathe for the patient using:

- mouth-to-mouth or mouth-to-mouth-and-nose ventilation, *or*
- bag-valve-mask ventilation: if using oxygen, add a reservoir to increase the oxygen concentration.

Intubate (if skilled professionals (anaesthetists) are available) and provide assisted ventilation through the endotracheal tube if long- term ventilation is needed or bag-mask ventilation is ineffective.

DO NOT persist with intubation attempts without ventilating the patient intermittently with a bag and mask as necessary to prevent hypoxaemia during the intubation process.

Situations in which emergency treatment is given in addition to continuing respiratory support and oxygen.

1. Perform needle thoracocentesis if the diagnosis is tension pneumothorax (see Section 91 Handbook 1). This should be followed by a chest drain.
2. Consider inserting a chest drain if there is major trauma to the chest (see Section 91 Handbook 1)
3. Give nebulised salbutamol if the patient has severe, life-threatening asthma (2.5 mg for children < 5 years of age, or 5 mg for children > 5 years of age and pregnant mothers). If a nebuliser is not available, use a spacer and metered-dose inhaler (100 micrograms/puff; 10 puffs initially for all age groups).
4. Give nasal continuous positive airway pressure (CPAP) if a neonate has severe respiratory distress (see Section 91 Handbook 1).
5. Give IM adrenaline (1 mg in pregnancy and 10 micro- grams/kg in children) and nebulised salbutamol (see above) if wheezing is due to anaphylaxis.
6. Give anticoagulant (IV unfractionated heparin) if pulmonary embolus is diagnosed in pregnancy or post delivery (see Section 15 Obstetric Handbook).
7. Give calcium gluconate (10 mL 10% IV over 10 minutes) if respiratory arrest is due to magnesium toxicity in a patient treated for eclampsia with magnesium sulphate.

Primary assessment and resuscitation of circulation

Primary assessment

The circulatory system is more difficult to assess than airway and breathing, and individual measurements must not be over-interpreted.

If there is no palpable pulse, a very slow heart rate (<60 beats/minute in an infant, or < 40 beats/minute in a child or pregnant woman) or no 'signs of life' (e.g. movements, coughing, normal breathing), cardiac arrest or near-cardiac arrest is likely, and basic life support must be started (see Section 12).

Agonal gasps (irregular, infrequent breaths) do not provide adequate oxygenation and are not for these purposes a 'sign of life'.

In addition to cardiac arrest or near-arrest, shock and heart failure are additional life-threatening issues that it is important to identify.

Shock

The following clinical signs can help to identify shock (inadequate circulation) (see Sections 13 in Handbook 2 and 45 in Handbook 1).

TABLE 11.2 Heart rates 'at rest' at different ages

Age (years)	Heart rate (beats/minute)
< 1	110–160
1–2	100–150
2–5	95–140
5–12	80–120
> 12	60–100

Heart rate

Heart rate increases in shock and heart failure.

Severe bradycardia due to hypoxaemia may be a sign of near cardiorespiratory arrest.

The WHO definition of tachycardia is a heart rate of > 160 beats/min in children aged under 1 year, and > 120 beats/ minute in those aged 1–5 years.

Pulse volume

Absent peripheral pulses or reduced strength of central pulses can signify shock.

Capillary refill time (CRT)

- Pressure on the centre of the sternum or fingernail for 5 seconds should be followed by return of the circulation to the skin within 3 seconds or less. CRT may be prolonged by shock, cold environment, or the vasoconstriction that occurs as a fever develops.
- Prolonged CRT is not a specific or sensitive sign of shock and should not be used alone as a guide to the need for or the response to treatment.

Blood pressure

- The cuff should cover at least 80% of the length of the upper arm, and the bladder should be more than two-thirds of the arm's circumference. In pregnant mothers, the largest possible cuff should be used to avoid missing a raised blood pressure.
- Korotkoff phase 5 (K5, disappearance of sound) should be used to measure diastolic pressure. Korotkoff phase 5 (K5A, muffling or softening of sound) should only be used if the sound does not disappear until near to zero cuff pressure.
- In pregnancy the patient should ideally be sitting or lying in the lateral tilt positions when pressure is measured. In both of these positions, the cuff must be level with the heart.
- Hypotension is a relatively late sign of circulatory failure in both children and will be rapidly followed by cardiorespiratory arrest unless it is treated urgently.

TABLE 11.3 Systolic and diastolic blood pressure in children

Age (years)	Systolic blood pressure (mmHg) 5th centile	Systolic blood pressure (mmHg) 50th centile
< 1	65–75	80–90
1–2	70–75	85–95
2–5	70–80	85–100
5–12	80–90	90–110
> 12	90–105	100–120

Blood pressure may be difficult to measure and interpret, especially in infants and children under 5 years of age. The following formula can be used to calculate average systolic blood pressure in children (50th centile):

$$85 + (2 \times \text{age in years})$$

The cardiovascular system in children compensates 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 failure on other organs

Respiratory system: tachypnoea and hyperventilation occur because of the acidosis caused by poor tissue perfusion.

Skin: pale or mottled skin indicates poor perfusion.

Mental status: circulatory failure causes initial agitation, then drowsiness, followed by unconsciousness.

Urine output: a reduction in urine output to < 2 mL/kg/ hour in infants, < 1 mL/kg/hour in children indicates inadequate renal perfusion.

The WHO definition of shock is cold hands, *plus* CRT of > 3 seconds, *plus* a weak and rapid pulse.

Life-threatening shock is usually associated with:

1. severe tachycardia
2. a weak-volume pulse (ideally assess centrally: brachial, femoral, or carotid)
3. low blood pressure (this is a late sign, and very difficult to measure in young children)
4. extreme central pallor (if due to severe anaemia)
5. raised respiratory rate (due to acidosis)
6. poor skin circulation, with a CRT of > 3 seconds
7. reduced conscious level.

Remember that anaphylaxis is one cause of shock, and typically there is a relevant history and other signs such as angio-oedema and urticaria.

Remember that if shock is due to heart failure, fluid overload will be fatal (for information on how to recognise and manage shock caused by heart failure, see Section 45).

Resuscitation in shock

For cardiac arrest or near arrest, chest compressions should be undertaken (for information on basic and advanced life support, see Sections 12 and 13). Ensure that there is an open and secure airway.

Give high-flow oxygen to any patient who has an inadequate circulation (whether due to shock or to heart failure). This should be administered via a face mask with a reservoir bag (or an endotracheal tube if intubation has been necessary).

Venous or intra-osseous access should be obtained and blood for essential tests taken (haemoglobin, cross-matching, blood clotting factors, and urea and electrolytes if possible).

Fluids in shock

In most cases of shock, if obvious bleeding is the cause, then the first priority must be to stop this. IV or IO fluids are then required as the immediate resuscitation treatment once the airway has been opened and secured and oxygen is being given. However, different causes of shock require different approaches to treatment, as described below.

1. If loss of fluid causing hypovolaemia is the cause of shock: for infants and children give an immediate IV/IO bolus of 10–20 mL/kg of crystalloid (usually Ringer- lactate or Hartmann's solution) as appropriate for weight (see below), provided that heart failure is not present (see above).
2. For a child, weight can be estimated on the basis that birth weight doubles by 5 months, triples by 1 year, and quadruples by 2 years.
3. After 12 months of age, the following formula can be applied, but it needs to be modified according to whether the child is small or large compared with the average:

$$\text{weight (kg)} = 2 \times (\text{age in years} + 4)$$

Another formula, from the Advanced Life Support Group, is used in countries where children are becoming heavier:

$$\begin{aligned} 0\text{-}12 \text{ months weight in kg} &= (0.5 \times \text{age in months}) + 4 \\ 1\text{-}5 \text{ years weight in kg} &= (2 \times \text{age in years}) + 8 \\ 6\text{-}12 \text{ years weight in kg} &= (3 \times \text{age in years}) + 7 \end{aligned}$$

4. If the loss of fluid causing shock is due to severe gastroenteritis, there will usually be evidence of severe dehydration and a history of profound or long- standing diarrhoea. Give 20 mL/kg of Ringer-lactate or Hartmann's solution as an initial IV or IO bolus as rapidly as possible, reassess, and then repeat if necessary. In cases of cholera, up to 60 mL/kg might be required in children. Additional potassium will usually be required (see Section 7 Handbook 1).
5. If the loss of fluid causing shock is due to **bleeding**, give crystalloid immediately and then try to obtain blood for transfusion as rapidly as possible, ideally fresh blood. Give O-negative blood if this is available.

The concept of targeted crystalloid fluid resuscitation is important and requires urgent research into management if the cause of hypovolaemic shock is haemorrhage due to penetrating injury in trauma. Here the initial boluses of IV crystalloids required to treat shock would only be given to keep the vital organs (especially the brain, heart and kidneys) perfused before surgery and/or specific medical treatments to stop the bleeding have started to take effect. Fresh blood is particularly useful to combat the coagulopathy

that occurs in major blood loss if specific coagulation components such as platelets are unavailable.

Giving too much IV crystalloid can increase the blood pressure and theoretically increase bleeding by disrupting early clot formation. IV crystalloid also dilutes the red cells (and coagulation factors) in the circulation, but whether or not this could reduce oxygen-carrying capacity requires further research.

We suggest that when giving boluses of crystalloid in shock due to bleeding (before blood is available and before procedures undertaken to stop haemorrhage are effective) in patients with penetrating major trauma, only the amount needed to maintain the blood pressure at a level sufficient to perfuse the vital organs is given.

There is no clear evidence to indicate the precise blood pressure that should be achieved in children who are in shock due to haemorrhage. Adequate perfusion of vital organs may best be indicated by a radial pulse that can be palpated and a conscious level of A or V on the AVPU scale (i.e., the child is either awake or will respond by opening their eyes when spoken to).

In children under 2–3 years of age, the radial pulse may be difficult to feel, and the presence of a palpable brachial pulse may be the best available indicator at present.

In children, in order to maintain a radial or brachial pulse give 10 mL/kg IV boluses of crystalloid or ideally blood and reassess after each bolus.

In situations where there is brisk active blood loss and delay in obtaining blood or effective intervention to halt the bleeding, several boluses of crystalloids may be required. The importance of undertaking measures to halt the bleeding and obtaining blood for transfusion rapidly cannot be overstated.

6. Shock due to **septicaemia** maybe accompanied by purpura (meningococcus or Dengue: Sections 21 and 13 Handbook 1). Give penicillin or cephalosporin if meningococcus is suspected. If septic shock is suspected (Section 45 Handbook 1) give IV or IO boluses of Ringer-lactate or Hartmann's or 0.9% saline as fast as possible, 10 mL/kg in children, then reassess and repeat boluses as and when needed. Usually at least 40 mL/kg in children will be required to overcome septic shock (see Section 45 Handbook 1). In this situation, inotropes may be valuable if they are available and safe to use (see Section 45 Handbook 1).
7. If shock is due to anaphylaxis, give IM adrenaline (epinephrine), 1: 1000 (that is 1mg/1ml) in the following doses, Child 1 month-5 years 0.15 ml, child 6-11 years 0.3 ml, Child over 12 years and adult 0.5 ml in addition to IV or IO fluid. (see Reference at end)
8. If shock is due to diabetic ketoacidosis, there will usually be evidence of severe dehydration and coma. Give 10 mL/kg of 0.9% saline (or Ringer-lactate or Hartmann's solution) as an initial IV bolus as rapidly as possible, reassess, and then repeat if necessary. Once shock has been initially managed, give fluid more cautiously, as overloading can cause cerebral oedema and death in patients with this condition. Be careful to measure potassium level (if possible) before starting insulin
9. If shock is due to severe anaemia, IV crystalloid boluses such as Ringer-lactate or Hartmann's solution must be given with extreme care (due to the risk of heart failure). As soon as possible, give blood carefully (10 mL/kg in children over 15 minutes) and then reassess and repeat if it is safe to do so.
10. Partial exchange transfusion, under strict asepsis, may be helpful in this situation, especially if it is possible to access a large superficial vein in the antecubital fossa. Successively remove 20-mL aliquots of the patient's blood and replace each 20 mL with 40 mL of packed donor red blood cells until shock has resolved.

Heart failure

This life-threatening situation can be seen in severe anaemia, after fluid overload, in the presence of structural heart disease and with severe hypertension. It is important to distinguish heart failure from shock, as the resuscitation required is different. Some of the following signs will be present in heart failure:

1. tachycardia out of proportion to respiratory difficulty
2. severe palmar pallor (if anaemia is the cause)
3. raised jugular venous pressure
4. gallop rhythm on auscultation of the heart
5. some heart murmurs (if structural heart defect is responsible)
6. an enlarged, sometimes tender, liver
7. crepitations on listening to the lung bases
8. cyanosis that does not respond to oxygen in the case of infants with cyanotic congenital heart disease. Warning be careful giving oxygen to blue babies with congenital heart disease. Place pulse oximeter probe on right hand or arm and stop the oxygen if saturations fall (see Section 40 Handbook 1).

Resuscitation for heart failure

- 1 Sit the patient up.
- 2 Give oxygen.
- 3 Give furosemide 1–2 mg/kg by IV/IO injection in children
- 4 Consider giving morphine (50 micrograms/kg in children) and reassess. Morphine should be used with caution, especially in patients with altered mental status and impaired respiratory drive.
- 5 If the patient has severe anaemia, consider partial exchange transfusion (see above).

Specific situations where emergency treatment is given in heart failure with shock

1. Supraventricular tachycardia can cause both shock and heart failure. The heart rate will be 180 beats/minute, and in infants can reach > 220 beats/ minute (usually 250 to 300 beats per minute).

If available, ECG will confirm tachycardia. Treat by vagal manoeuvres, defibrillation if available, or adenosine if rapid IV access is available (see Section 49 Handbook 1).

2. In ventricular tachycardia defibrillation is needed if shock is present (see Section 13).
3. If congenital or rheumatic heart disease or cardiomyopathy is the cause of heart failure, inotropes or digoxin may be appropriate, but specialist advice will be needed.
4. If cyanotic congenital heart disease in the newborn is the cause of shock, give prostaglandin E₂, but specialist paediatric advice will be necessary (see Section 40 and Neonatal Handbook)

Primary assessment and resuscitation of neurological failure (disability) Always assess and treat Airway, Breathing and Circulation problems before undertaking neurological assessment.

Primary assessment

Conscious level: AVPU

Alert is the normal state for an awake child. If the patient does not respond to Voice (i.e. being spoken to and asked 'Are you all right?'), it is important that assessment of the response to Pain is undertaken next. A painful central stimulus can be delivered by sternal pressure, by supra-orbital ridge pressure or by pulling frontal hair. A patient who is Unresponsive or who only responds to pain has a significant degree of coma which can seriously interfere with vital Airway and Breathing functions.

Fits

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Generalised convulsions, also known as 'fits' or 'seizures', can seriously interfere with vital Airway and Breathing functions, both during the fit itself and immediately afterwards, when lowered levels of consciousness may be present.

Posture

Many patients who have a serious illness in any system are hypotonic. Stiff posturing, such as that shown by decorticate (flexed arms, extended legs) or decerebrate (extended arms, extended legs) posturing, is a sign of serious brain dysfunction. These postures can be mistaken for the tonic phase of a convulsion. Alternatively, a painful stimulus may be necessary to elicit these postures.

Severe extension of the neck due to upper airway obstruction can mimic the opisthotonus that occurs with meningeal irritation. In infants, a stiff neck and full fontanelle are signs that suggest meningitis.

Pupils

Many drugs and cerebral lesions have effects on pupil size and reactions. However, the most important pupillary signs to seek are dilatation, lack of reactivity to light and inequality, which suggest possible serious brain disorders.

Hypoglycaemia

Always check blood glucose levels or suspect hypoglycaemia in any unwell infant or young child, especially if they have impaired consciousness.

Hypoglycaemia with a blood glucose level of less than 2.5 mmol/L (45 mg/dL) can cause impaired consciousness, coma, or fits. Give 2 mL/kg of 10% glucose IV or IO (see Section 51 Handbook 1).

Respiratory effects of central neurological failure

The presence of any abnormal respiratory pattern in a patient with coma suggests mid- or hindbrain dysfunction.

Circulatory effects of central neurological failure Systemic hypertension with sinus bradycardia (Cushing's response) indicates compression of the medulla oblongata caused by herniation of the cerebellar tonsils through the foramen magnum. This is a late and pre-terminal sign.

Raised intracranial pressure (ICP) may cause:

1. hyperventilation
2. slow sighing respirations
3. apnoea
4. hypertension
5. bradycardia.

Resuscitation for neurological emergencies

1. If the patient is unconscious (P or U on the AVPU scale) but their airway and breathing are adequate, place them in the recovery position, so that if they vomit there is less likelihood of aspiration because when unconscious, the gag reflex may not be operative.
2. If the patient is unconscious or fitting, always give oxygen.
3. If hypoglycaemia is a cause of reduced consciousness (or a suspected cause, but immediate blood glucose measurements are not possible), treatment with glucose is urgently required. Give 2 mL/kg of 10% glucose IV or IO in children (see Section 51 Handbook 1)

If IV or IO access is not immediately available, give sublingual sugar, 1 teaspoonful moistened with 1 to 2 drops of water. Children should be monitored for early swallowing which leads to delayed absorption, and in this case another dose of sugar should be given. Continue to attempt IV or IO access, as parenteral glucose is a more reliable method of treating hypoglycaemia.

If sublingual sugar is given, repeat the doses at 20-minute intervals.

Recheck the blood glucose level after 20 minutes, and if the level is low (< 2.5 mmol/litre or < 45 mg/dL), repeat the IV/IO glucose (5 mL/kg) or repeat the sublingual sugar.

4. If fitting occurs in an infant or child and continues in your presence for more than 5 minutes and there is no hypoglycaemia, give buccal, IV or rectal anticonvulsants. Always make sure that a bag and mask are available in case the patient stops breathing, which is a strong possibility. Commonly used anticonvulsants in this situation are diazepam or, if there is no IV access, rectal diazepam, rectal paraldehyde or buccal midazolam (see Section 70 Handbook 1).

IV or IO diazepam: 250 micrograms/kg IV over 5 minutes

rectal diazepam: 500 micrograms/kg

rectal paraldehyde: 0.4 mL/kg (if available is a safe drug)

buccal midazolam: 300 micrograms/kg.

5. To gain time in acutely raised intracranial pressure (e.g., in cases of head injury), consider the use of IV hypertonic 3% saline 3-5 mL/kg over 15 mins) followed by a continuous infusion of 0.1-1.0 mL/kg/h of the same solution **or** mannitol, 250–500 mg/kg, which will draw fluid out of the brain for a short while, thereby temporarily reducing the ICP. Because the effect of 3% saline or mannitol is only short-lived (a matter of hours), it is used to gain time while definitive care is being set up (e.g., surgical intervention to drain an extradural or subdural haematoma).
6. In any case where meningitis or encephalitis is suspected, it is vital that suitable antibiotics and/or antiviral drugs are started IV or IO as soon as the condition is suspected (see Sections 67 and 68 Handbook 1). Antibiotic choices might include cefotaxime or chloramphenicol, penicillin, amoxicillin and gentamicin in the newborn. Consider adjunctive treatment with dexamethasone 150 micrograms/kg every 6 hours for 4 days starting before or with the first antibiotic dose.

Secondary assessment and emergency treatments

The secondary assessment takes place once vital functions have been assessed and the initial resuscitation of those vital functions has been started. Primary assessment and resuscitation can usually be undertaken in less than 1 minute if the patient does not have a life-threatening airway, breathing, circulation or neurological problem.

Secondary assessment includes a focused medical history, a focused clinical examination, and specific investigations. It differs from a standard medical history and examination in that it is designed to establish which emergency treatments might benefit the patient. Time is limited, and a focused approach is essential. At the end of secondary assessment, the practitioner should have a better understanding of the illness or component of injury likely to be affecting the patient and may have formulated a differential diagnosis. Emergency treatments will be appropriate at this stage – to treat either specific disorders (e.g., asthma) or conditions (e.g., raised intracranial pressure). Emergency treatments will be undertaken at this stage in addition to those given as part of resuscitation/life-saving treatments, to manage specific components of serious illnesses or injuries (e.g., steroids for asthma). The establishment of a definite diagnosis is part of definitive care.

The history often provides the vital clues. In the case of infants and children, the history is often obtained from an accompanying parent, although a history should be sought from the

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child if possible. Do not forget to ask any health worker who has seen the patient about the initial condition and about treatments and the response to treatments that have already been given.

Some patients will present with an acute exacerbation/ complication of a known condition, such as asthma or epilepsy. Such information is helpful in focusing attention on the appropriate system, but the practitioner should be wary of dismissing new pathologies in such patients. The structured approach avoids this problem. Unlike trauma (see Section 79 Handbook 1), illness affects systems rather than anatomical areas. The secondary assessment must reflect this, and the history of the complaint should be sought with special attention to the presenting system or systems involved. After the presenting system has been dealt with, all of the other systems should be assessed, and any additional emergency treatments commenced as appropriate.

The secondary assessment is not intended to complete the diagnostic process, but rather it aims to identify any problems that require emergency treatment.

An outline of a structured approach in the first hour of emergency management is given below and addresses most emergency conditions that are amenable to specific emergency treatments in this time period.

The symptoms, signs and treatments relevant to each emergency condition are elaborated further in the relevant sections of this handbook.

Examples of emergency treatment for airway and breathing

1. If in a young child there is a harsh stridor associated with a barking cough and severe respiratory distress, upper airway obstruction due to severe croup should be suspected. Nebulised adrenaline will already have been given as resuscitation, but now give oral prednisolone as emergency treatment (see Section 33 Handbook 1).
2. If there is a quiet stridor and drooling in a sick-looking child, consider epiglottitis or bacterial tracheitis. Intubation is likely to be urgently required, preferably by an anaesthetist, and is initial resuscitation if the airway is completely closed. Do not put the airway at risk by performing unpleasant or frightening interventions. Give intravenous antibiotics as emergency treatment, but only after the airway has been secured (see Sections 33 and 90 in Handbook 1 and 60 in Handbook 2). A surgical airway may also be needed as emergency treatment or as resuscitation if intubation is not possible, so contact a surgeon.
3. With a sudden onset and significant history of inhalation, consider a laryngeal foreign body. If the 'choking' protocol has been unsuccessful, the patient may require laryngoscopy (see Section 13). Do not put the airway at risk by performing unpleasant or frightening interventions but contact an anaesthetist/ENT surgeon urgently. However, in extreme, life-threatening cases, immediate direct laryngoscopy as part of resuscitation to remove a visible foreign body with Magill's forceps may be necessary.
4. Stridor following ingestion or injection of a known allergen suggests anaphylaxis (see Section 36 Handbook 1). Patients in whom this is likely should have IM and nebulised adrenaline (10 micrograms/kg IM for a child) as resuscitation treatment. IV or oral steroids would then be part of emergency treatment.
5. Patients with a history of asthma or with wheeze, significant respiratory distress and/or hypoxia should receive inhaled salbutamol and oxygen as resuscitation, but then need oral steroids and further inhaled bronchodilators as emergency treatment (see Section 35 Handbook 1).
6. Infants with wheeze and respiratory distress are likely to have bronchiolitis, and require oxygen, as well as clearing of nasal secretions as resuscitation, and IV or NG fluids as emergency treatment (see Section 34 Handbook 1).

7. In acidotic breathing, measure blood glucose levels to confirm diabetic ketoacidosis. A bolus of IV Ringer- lactate or Hartmann's solution will already have been given as resuscitation for any shock due to dehydration, and insulin can now be given as emergency treatment (see Section 50 Handbook 1). Be careful to measure potassium level (if possible) before starting insulin.

Examples of emergency treatment for circulation

1. Further IV/IO boluses of fluid should be considered in shocked patients with hypovolaemia from gastroenteritis or with sepsis who have not shown a sustained improvement in response to the first bolus given at resuscitation (see Sections 61 and 45 Handbook 1).
2. However, in trauma, if there is uncontrolled internal bleeding, early surgical intervention has priority, and too much IV fluid may be harmful. Continued blood transfusion is an emergency treatment after the initial resuscitation (see Section 54).
3. Consider inotropes, intubation, if available, as emergency treatment for shock (see Section 45 Handbook 1).
4. Consider IV broad-spectrum antibiotics as emergency treatment for shock in patients with no obvious fluid loss, as sepsis is likely. Antibiotics are essential if purpura is present, as a diagnosis of meningococcal infection is likely (see Section 21 Handbook 1).
5. If a patient has a cardiac arrhythmia, the appropriate protocol should be followed after initial resuscitation (see Section 13 Handbook 2 and 44 Handbook 1).
6. If anaphylaxis is suspected, IM adrenaline 10 micrograms/kg in children, in addition to fluid boluses, oxygen, steroids and antihistamines should be given as emergency treatment. (see Section 36 Handbook 1).
7. Surgical advice and interventions for certain gastrointestinal emergencies such as volvulus would constitute emergency treatment. The following symptoms and signs may suggest intra-abdominal emergencies: vomiting, abdominal pain, abdominal tenderness and/or rigidity, lack of bowel sounds, rectal bleeding, abdominal mass (see Section 74 Handbook 1). Consider pain relief in intra-abdominal emergencies as an emergency treatment.

Examples of emergency treatment for neurological failure

1. If hypoglycaemia with a blood glucose level of less than 2.5 mmol/L (45 mg/dL) is a possible diagnosis, it will have been treated as part of resuscitation, but the prevention of further hypoglycaemia by IV glucose infusion represents emergency treatment. Remember that there will be a reason for the hypoglycaemia, so further monitoring and treatment are needed until the child is drinking appropriate fluids or has an IV infusion in place through which dextrose can be given.
2. If convulsions persist after initial anticonvulsant drugs, treatment with further doses of anticonvulsants (see Section 70 Handbook 1) represents emergency treatment.
3. If there is evidence of raised intracranial pressure (i.e., decreased conscious level, abnormal posturing and/ or abnormal ocular motor reflexes), the patient should receive oxygen and bag-valve-mask ventilation as resuscitation, if they have apnoea or slow or poor breathing. Emergency treatment could include:
nursing with head in-line and 20–30 degrees head-up position (to aid cerebral venous drainage)
Repeat IV infusion with hypertonic 3% saline or mannitol (see above); however, the treatment becomes less effective with each dose (see Sections 66 and 73 Handbook 1)
4. in more long-standing raised ICP, caused by tumours in the brain, dexamethasone will help to reduce raised ICP for a few days while specialist neurosurgical intervention is sought, or as palliation (see Section 73 Handbook 1). The initial dose is 25 mg for patients over 35 kg and 20 mg for patients less than 35 kg, followed by a sliding scale of

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4 mg every 3 hours for 3 days, then every 6 hours for 1 day, and continuing to decrease by 1–2 mg per day.

5. In patients with a depressed conscious level or convulsions, antibiotics are urgently required, but then consider encephalitis and give acyclovir as appropriate, as emergency treatment (see Section 68 Handbook 1).
6. In unconscious patients with pinpoint pupils, consider the possibility of opiate poisoning. After supporting breathing, if necessary, a trial of naloxone should be given as emergency treatment (see Section 87 Handbook 1).

Developmental and family history

Particularly in a small child or infant, knowledge of the child's developmental progress and immunisation status may be useful. The family circumstances may also be helpful and asking about these may sometimes prompt parents to remember other details of the family's medical history. Always be aware of possible child protection needs and look for bruises/burns/scars etc in all ages. If possible, especially in infants, examine the fundi for retinal haemorrhages (see section 2).

Drugs and allergies

Any medication that the patient is currently taking, or has taken, should be recorded. In addition, if poisoning is a possibility, ask about any medication in the home that a child might have had access to. And consider other poisons such as antidepressants, iron, organophosphates, traditional herbal remedies etc. (see Section 10). A history of allergies should be sought.

Reference for anaphylaxis management

Emergency treatment of anaphylaxis: Guidelines for healthcare providers
2021 The Resuscitation Council UK. <https://www.resus.org.uk/library/additional-guidance/guidance-anaphylaxis/emergency-treatment>

12. Basic Life Support

Introduction

Basic life support (BLS) is a technique that can be employed by one or more rescuers to support the respiratory and circulatory functions of a collapsed patient using no or minimum equipment.

Respiratory arrest, cardiac arrest or both during infancy and childhood are unusual but survival rates are very low. The cause of the arrest is not often reversible, and the physiological changes present are complex and hinder effective Cardio-Pulmonary Resuscitation (CPR). Prevention is the key.

Resuscitation from cardiac and respiratory arrest in children

The international guidelines for resuscitation from cardiac arrest (European Resuscitation Council, 2021) detail two approaches to basic life support. One is for adults and the other for children.

The 'adult' programme is predicated on resuscitation from a sudden cardiac event (e.g. ventricular fibrillation from a coronary occlusion) in a patient who was ventilating before the event and therefore has oxygen in their blood. In this group, chest compressions to move the oxygenated blood into the coronary and cerebral arteries are of prime importance, and therefore the rescuer's sequence of actions after assessment starts with chest compressions, not rescue breaths.

The sequence of actions in the 'child' programme is predicated on a hypoxic event (including any respiratory failure or obstruction, or hypoxia at a cellular level as seen in shock). In this type, re-establishing oxygenation is of prime importance, and moving the oxygenated blood to the coronary and cerebral arteries is the second step. Therefore, the rescuer's sequence of actions after assessment starts with rescue breaths and then moves on to chest compressions. The 'child'-type cardiac arrest is seen in almost all children (excluding those rare arrhythmic events in children with congenital or acquired heart disease and those in whom sudden, unexpected collapse is preceded by apparent normal respiratory and circulatory function), and in adults who have a terminal acute illness involving respiratory or circulatory pathology. This includes patients who have had convulsions, trauma (including drowning), poisoning, bleeding, sepsis, etc.

In addition, international guidelines on resuscitation from cardiac arrest agree that, where possible, guidelines should be simplified as there is evidence that complex guidelines cause 'provider paralysis', resulting in no or poor life-saving effort being made.

In view of the above, the Advanced Life Support Group (ALSG/Maternal Child Health Advocacy International (MCAI) programme for resource-limited countries teaches a programme of basic life support for infants and children which reflects the known pathologies in these patients (i.e., respiratory and circulatory causes of cardiac arrest) and recognises that the clinicians who provide resuscitation attend patients of all ages.

This sequence taught therefore includes five preliminary rescue breaths and a subsequent ratio of 15:2.

Because of minor differences in technique based on anatomical differences between the groups, children are classified into two groups:

- infants (< 1 year of age)
- children between 1 year of age and puberty.

Basic life support for infants and children (see Figures 12.1)

The first action must be to CALL FOR HELP.

The initial approach: the three S's

Safety: it is essential that the rescuer does not become a second victim. Therefore, each rescuer should approach the patient with care, and remove the patient from any continuing source of danger if necessary.

Stimulate: ask the question '**Are you all right?**' in order to establish the state of consciousness of the patient.

Shout: this is essential because help will be needed.

If more than one rescuer is present, one person should start basic life support. The second person should activate the Emergency Medical Services (EMS) system and then return to assist in the basic life support effort.

If the patient is an infant or pre-pubertal child, and there is only one rescuer and no help has arrived, the rescuer should open the airway, deliver the five rescue breaths and give 1 minute of cardiopulmonary resuscitation (CPR), and then activate the EMS system (if one is available) using a mobile phone if available so as to continue CPR. If a mobile is not available and the patient is a baby, the rescuer will probably be able to carry them to a telephone while continuing CPR.

Are you all right?

An initial simple assessment of responsiveness consists of asking the patient 'Are you all right?' and gently shaking them by the shoulder. Infants may make some noise or open their eyes.

In cases associated with trauma, or possible trauma, the cervical spine should be immobilised during this procedure by placing one hand firmly on the forehead while one of the patient's shoulders is shaken.

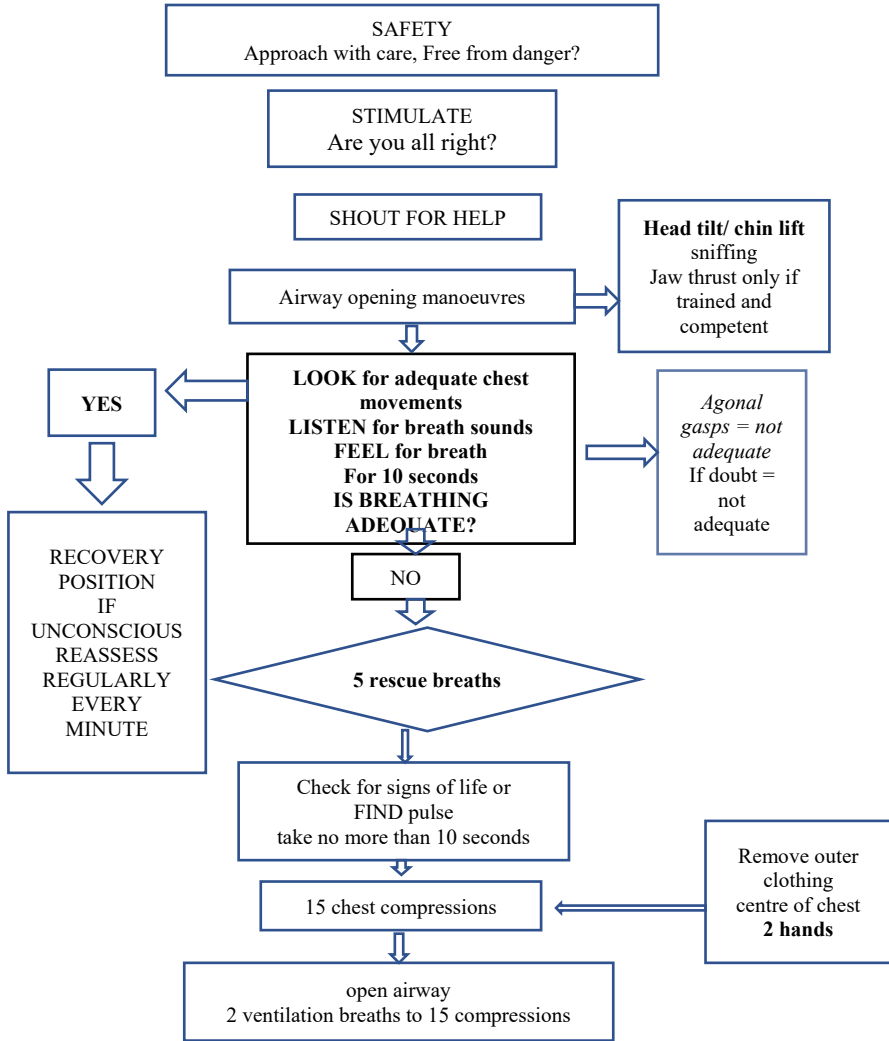


FIGURE 12.1 Algorithm for basic life support in infants and children. CPR, cardiopulmonary resuscitation; VF, ventricular fibrillation; VT, ventricular tachycardia; PEA, pulseless electrical activity.

Airway-opening actions

An obstructed airway may be the primary problem, and correction of the obstruction can result in recovery without the need for further intervention. If the patient is unconscious but breathing, the recovery position should be used.

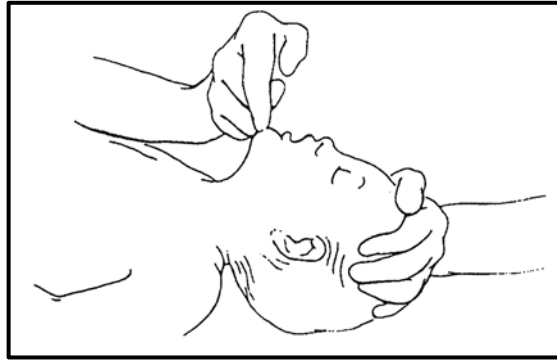


FIGURE 12.2 Head tilt with chin lift in neutral position for the infant.



FIGURE 12.3 Head tilt with chin lift in 'sniffing' position for the child older than 1 year

If the patient is not breathing, this may be because the airway is blocked by the tongue falling back and obstructing the pharynx. Attempt to open the airway using the head tilt/chin lift manoeuvre. The rescuer places their nearest hand on the patient's forehead and applies pressure to tilt the head back gently. The correct positions are 'neutral' in the infant (0–1 year of age) (see Figure 12.2) or 'sniffing' (nose up in the air) in the child (see Figure 12.3).

The fingers of the other hand should then be placed under the chin, and the chin of the supine patient should be lifted upwards. As this action may close the patient's mouth, it may be necessary to use the thumb of the same hand to part the lips slightly.

As an alternative to the head tilt/chin lift, the jaw thrust manoeuvre can be very effective, but requires more training and experience.

FIGURE 12.4 Jaw thrust to open airway.



Jaw thrust is achieved by placing two or three fingers under the angle of the mandible bilaterally and lifting the jaw upward (see Figure 12.4). This is potentially safer than the head

tilt/chin lift if there is a history of major trauma, as the latter manoeuvre may exacerbate a cervical spine injury. BUT airway opening is always the most important action which must be achieved and should always take precedence over concerns about a possible cervical spine injury.

The continued opening of the airway should then be assessed by:

- looking for adequate chest movements
- listening for breath sounds
- feeling for breaths.

This is best achieved by the rescuer placing their face above that of the patient, with the ear over the nose, the cheek over the mouth, and the eyes looking along the line of the chest. They should take no longer than 10 seconds to assess breathing.

If there is anything obvious in the mouth and it is easy to reach, remove it.

Do not perform a blind finger sweep in the mouth. A blind finger sweep can damage the soft palate, and foreign bodies may be forced further down the airway and become lodged below the vocal cords.

Breathing actions

If airway-opening techniques do not result in the resumption of adequate breathing within 10 seconds, and a self-inflating bag-mask system is not available, then the rescuer should commence mouth-to-mouth or mouth-to-mouth-and-nose exhaled air resuscitation.

Definition of adequate breathing

A patient may have very slow or shallow breathing, or take infrequent, noisy, agonal gasps. Do not confuse this with normal breathing.

Rescue breaths

If in doubt about the adequacy of breathing, five initial rescue breaths should be given. While the airway is held open, the rescuer breathes in and seals their mouth around the patient's mouth or mouth and nose (in the case of infants) (see Figures 12.5 and 12.6). If the mouth alone is used, the nose should be pinched using the thumb and index finger of the hand maintaining head tilt. Slow exhalation, 1–2 seconds, by the rescuer should result in the patient's chest rising. The rescuer should take a further breath him- or herself before the next rescue breath.



FIGURE 12.5 Mouth-to-mouth and nose breaths in neutral position for an infant.



FIGURE 12.6 Mouth-to-mouth breaths with pinched nose in sniffing position for a child or mother.

As children vary in size, only general guidance can be given regarding the volume and pressure of inflation (see Box 12.1).

BOX 12.1 General guidance for exhaled air resuscitation

- The chest should be seen to rise.
- Slow breaths at the lowest pressure reduce gastric distension.
- Firm gentle pressure on the cricoid cartilage may reduce gastric distension with air.

If the chest does not rise, the airway is not clear. The usual cause is failure to correctly apply the airway-opening techniques discussed earlier. The first step is to readjust the head tilt/chin lift position and try again. If this is not successful, jaw thrust should be tried. If two rescuers are present, one should maintain the airway while the other breathes for the patient.

Failure of both head tilt/chin lift and jaw thrust should lead to suspicion that a foreign body is causing the obstruction (see below).

While performing rescue breaths, the presence of a gag reflex or coughing is a positive sign of life (see below).

Circulation actions

Once the initial five breaths have been given successfully, circulation should be assessed and managed.

Check signs of life and/or pulse (take no more than 10 seconds)

Even experienced health professionals can find it difficult to be certain that the pulse is absent within 10 seconds, so the absence of 'signs of life' is the best indication for starting chest compressions. 'Signs of life' include movement, coughing, gagging or normal breathing (but not agonal gasps, which are irregular, infrequent breaths). Thus, the absence of evidence

of normal breathing, coughing or gagging (which may be noticed during rescue breaths) or any spontaneous movement is an indication for chest compressions.

Inadequacy of circulation is also indicated by the absence of a central pulse for up to 10 seconds, but it can be difficult and therefore time wasting to be certain about this – hence the current emphasis on assessing the presence of 'signs of life'.

In babies and young children, if a slow pulse (less than 60 beats/minute) is felt, this is still an indication for chest compressions. In older children, the carotid pulse in the neck can be palpated. However, infants generally have a short fat neck, so the carotid pulse may be difficult to identify. The brachial artery in the medial aspect of the antecubital fossa or the femoral artery in the groin should be felt in infants.

If there are no signs of life and/or a pulse is absent for up to 10 seconds, start chest compressions. Compressions should also be started if in an infant or young child there is an inadequate heart rate (less than 60 beats/minute), but only if this is accompanied by signs of poor perfusion, which include pallor, lack of responsiveness and poor muscle tone.

Start chest compressions if:

- there are no signs of life *or*
- there is no pulse *or*
- there is a slow pulse (less than 60 beats/minute in an unconscious infant or young child with poor perfusion).

Chest compressions

For the best output, the patient must be placed on their back, on a hard surface. The chest should be compressed by a third of its depth. Children vary in size, and the exact nature of the compressions given should reflect this. In general, infants (less than 1 year of age) require a different technique from pre-pubertal children, in whom the method used in adults can be applied with appropriate modifications for their size.

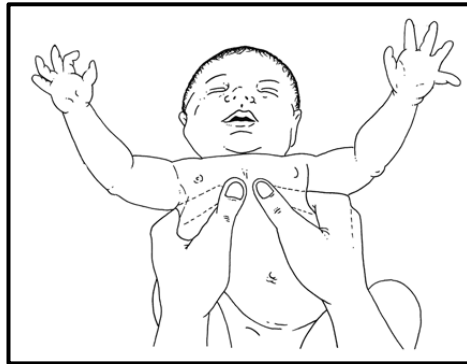


FIGURE 12.7 Two-thumb method for chest compressions in an infant (two rescuers).

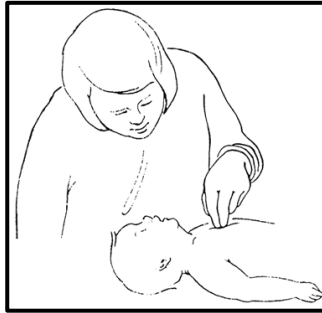


FIGURE 12.8 Two-finger method for chest compressions in an infant (one rescuer).

Position for chest compressions

Chest compressions should compress the lower half of the sternum.

For Infants: Infant chest compression can be more effectively achieved using the hand-encircling technique: the infant is held with both the rescuer's hands encircling or

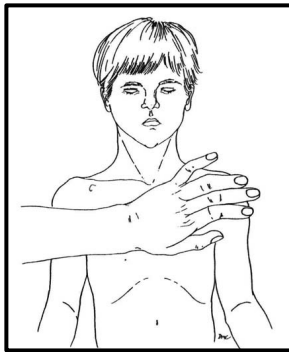


FIGURE 12.9 Chest compressions using one hand in a child.

partially encircling the chest. The thumbs are placed over the lower half of the sternum and compression is carried out as shown in Figure 12.7. This method is only possible when there are two rescuers, as the time needed to reposition the airway precludes the use of the technique by a single rescuer if the recommended rates of compression and ventilation are to be achieved. The single rescuer should use the two-finger method as shown in Figure 12.8, employing the other hand to maintain the airway position.

For small children: Place the heel of one hand over the lower half of the sternum. Lift the fingers to ensure that pressure is not applied over the child's ribs. Position yourself vertically above the child's chest and, with your arm straight, compress the sternum to depress it by approximately one third of the depth of the chest (Figure 12.9).

For larger children, or for small rescuers, compressions may be achieved most easily by using both hands with the fingers interlocked (Figure 12.10). The rescuer may choose one or two hands to achieve the desired compression of one third of the depth of the chest.

Once the correct technique has been chosen and the area for compression identified, 15 compressions should be given to 2 ventilations.

Technique for giving chest compressions in larger children and pregnant mothers

1. Kneel by the side of the patient, who must be positioned on a firm surface, the uterus having been displaced if appropriate (see below).
2. Place the heel of one hand in the centre of the patient's chest.
3. Place the heel of your other hand on top of the first hand.
4. Interlock the fingers of your hands and ensure that pressure is not applied over the patient's ribs. Do not apply any pressure over the upper abdomen or the bottom end of the bony sternum (breastbone).
5. Position yourself vertically above the patient's chest and, with your arms straight, press down on the sternum to a depth of 5–6 cm.
6. After each compression, release all the pressure on the chest without losing contact between your hands and the sternum.
7. Repeat at a rate of about 100–120 times a minute (a little less than 2 compressions a second).
8. Compression and release should take an equal amount of time.

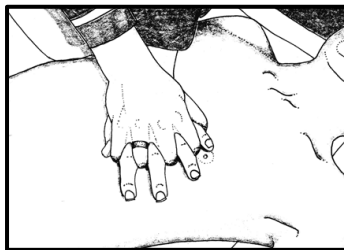


FIGURE 12.10 Chest compressions using two hands in a larger child

'Unnecessary' chest compressions are almost never damaging. It is important not to waste vital seconds before starting chest compressions after oxygenating the patient with the rescue breaths. If there are signs of life and the pulse is present (and has an adequate rate, with good perfusion), but apnoea persists, exhaled air resuscitation must be continued until spontaneous breathing resumes.

Technique for giving breaths and chest compressions in larger children (see Figure 12.10)

- After 15 compressions, open the airway again using the head tilt and chin lift (use the jaw thrust if you are experienced and capable of doing it properly and there are two rescuers).
- Pinch the soft part of the patient's nose closed, using the index finger and thumb of your hand on their forehead.
- Allow the patient's mouth to open but maintain chin lift.
- Take a normal breath and place your lips around the patient's mouth, making sure that you have a good seal. If you have a bag-valve-mask, this can be used instead of mouth-to-mouth basic life support in all age groups.
- Blow steadily into the patient's mouth while watching for their chest to rise; take about 1 second to make their chest rise, as in normal breathing; this is an effective rescue breath.
- Maintaining the head tilt and chin lift, take your mouth away from the patient and watch for their chest to fall as air is exhaled.
- Take another normal breath and blow into the patient's mouth once more to give a total of two effective rescue breaths. Then return your hands without delay to the correct position on the sternum and give a further 15 chest compressions.
- Continue with chest compressions and rescue breaths in a ratio of 15:2.
- Stop to recheck the patient only if they start breathing normally; otherwise, do not interrupt resuscitation.

- If your rescue breaths do not make the chest rise as in normal breathing, then before your next attempt:
 - check the patient's mouth and remove any visible obstruction
 - recheck that there is adequate head tilt and chin lift
 - try the jaw thrust if you are able to do this effectively.

Do not attempt more than two breaths each time before returning to chest compressions. If there is more than one rescuer present, a different person should take over CPR about every 2 minutes to prevent fatigue. Ensure that there is minimal delay during the changeover between rescuers.

Continuing cardiopulmonary resuscitation

The compression rate for all age groups is approximately 100–120 compressions per minute. A ratio of 15 compressions to 2 ventilations is maintained irrespective of the number of rescuers. With pauses for ventilation there will be less than 100–120 compressions per minute, although the rate is 100–120 per minute. Compressions can be recommenced at the end of inspiration and may augment exhalation.

If no help has arrived, the emergency services (if available) must be contacted after 1 minute of cardiopulmonary resuscitation. Apart from this interruption to summon help, basic life support must not be interrupted unless the patient moves or takes a breath.

Effective chest compressions are tiring for the rescuer. Continually check that the compressions and ventilations are satisfactory (they should be performed 'hard and fast') and, if possible, alternate the rescuers involved in this task. Any time spent readjusting the airway or re-establishing the correct position for compressions will seriously decrease the number of cycles given per minute. This can be a real problem for the solo rescuer, and there is no easy solution. In infants and small children, the free hand can maintain the head position. The correct position for compressions does not need to be measured after each set of ventilations.

The cardiopulmonary resuscitation manoeuvres recommended for infants and children are summarised in Table 12.1.

TABLE 12.1 Summary of basic life support techniques in infants and children

	Infants (< 1 year)	Children (1 year to puberty and teenagers)
Airway		
Head-tilt position	Neutral	Sniffing
Breathing		
Initial slow breaths	Five	Five
Circulation		
Pulse check	Brachial or femoral	Carotid
Landmark	Lower half of sternum	Lower half of sternum
Technique	Two fingers or two thumbs	One or two hands
CPR ratio	15:2	15:2

Call emergency services (if available)

If no help has arrived, the emergency services must be contacted after 1 minute of resuscitation has been delivered. A mobile phone can be used or an infant or small child may be carried to a static telephone or to get help while attempts are continued. Apart

from any necessary interruption to summon help, basic life support must not be interrupted unless the patient moves or takes a breath, or you are exhausted.

If recovery occurs and signs of life return, place the patient in the recovery position and continue to reassess them and ensure that specialist help arrives.

Basic life support and infection risk

Few cases have been reported. The most serious concerns are meningococcus and TB. In the case of meningococcus, rescuers involved in the resuscitation of the airway in such patients should take standard prophylactic antibiotics. There have been no reported cases of transmission of either hepatitis B or human immunodeficiency virus (HIV) through mouth-to-mouth ventilation. Blood-to-blood contact is the single most important route of transmission of these viruses, and in non-trauma resuscitation the risks are negligible. Sputum, saliva, sweat, tears, urine and vomit are low-risk fluids. Precautions should be taken, if possible, in cases where there might be contact with blood, cerebrospinal fluid pleural and peritoneal fluids. Precautions are also recommended if any bodily secretion contains visible blood. Devices that prevent direct contact between the rescuer and the patient (such as resuscitation masks) can be used to lower the risk. Gauze swabs or any other porous material placed over the patient's mouth are of no benefit in this regard.

Infection rates vary from country to country, and rescuers must be aware of the local risk. In countries where HIV/ AIDS is more prevalent, the risk to the rescuer will be greater.

If available, bag-valve-mask ventilation is always preferable to mouth-to-mouth ventilation.

If the rescuer is worried regarding cross infection, chest compressions alone will produce some ventilation of the lungs and should always be undertaken if there are no signs of life.

Minimising risks associated with BLS and COVID-19 is discussed in detail in Section 12 Handbook 1.

The recovery position

The patient should be placed in a stable, lateral position that ensures maintenance of an open airway with free drainage of fluid from the mouth, ability to monitor and gain access to the patient, security of the cervical spine and attention to pressure points (see Figure 12.11). The Resuscitation Council (UK) recommends the following sequence of actions when placing a patient in the recovery position:

- Remove the patient's spectacles (if present).
- Kneel beside the patient and make sure that both of their legs are straight.
- Place the arm nearest to you out at right angles to their body, elbow bent with the hand palm uppermost.
- Bring the far arm across the chest and hold the back of the hand against the patient's cheek nearest to you.
- With your other hand, grasp the far leg just above the knee and pull it up, keeping the foot on the ground.
- Keeping their hand pressed against their cheek, pull on the far leg to roll the patient towards you on to their side.
- Adjust the upper leg so that both the hip and knee are bent at right angles.
- Tilt the head back to make sure the airway remains open.
- Adjust the hand under the cheek, if necessary, to keep the head tilted.
- Check the patient's breathing regularly.

If the patient has to be kept in the recovery position for more than 30 minutes, turn them to the opposite side in order to relieve the pressure on the lower arm.

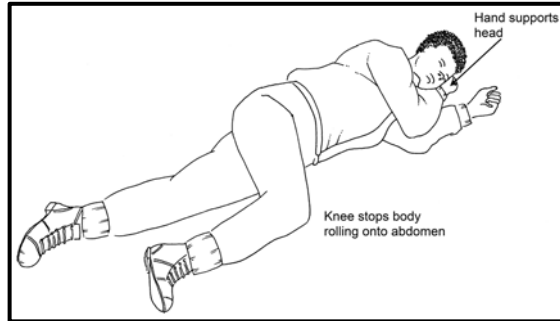


FIGURE 12.11 *The semi-prone or recovery position.*

Further reading

European Resuscitation Council Guidelines 2021: Executive summary

<https://www.cprguidelines.eu/assets/guidelines/RESUS-8995-Exec-Summary.pdf>

Accessed 20th April 2021

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Section 13. Advanced life support and cardio-pulmonary resuscitation in emergencies for infants and children (see Advanced Obstetric Handbook for management in adolescent girls who are pregnant)

Introduction

As described in Section 12 on basic life support, cardiac arrest in infants and children is usually the result of respiratory or circulatory collapse rather than a primarily cardiac event). This 'child' type of cardiac arrest is seen in almost all children (excluding those rare arrhythmic events in children with congenital or acquired heart disease, and those in whom sudden, unexpected collapse is preceded by apparent normal respiratory and circulatory function). This pattern of arrest includes patients who have had convulsions, trauma (including drowning), poisoning, bleeding, sepsis, etc.

In addition, there is international agreement that, where possible, guidelines on resuscitation of patients with cardiac arrest should be simplified, as there is evidence that complex guidelines cause 'provider paralysis', resulting in no or poor life-saving effort being made.

In view of this, the Advanced Life Support Group (ALSG)/Maternal Childhealth Advocacy International (MCAI) programme for resource-limited countries teaches a programme of basic life support (BLS) and advanced life support (ALS) for infants and children which reflects the most prevalent known pathologies in these groups (i.e., respiratory and circulatory causes of cardiac arrest).

Airway and breathing

Management of the airway (A) and breathing (B) components of the ABC must take priority in all situations. Resuscitation will fail if effective ventilation does not occur. Before effective resuscitation techniques can be applied, it is essential that the operator can:

1. understand the airway equipment available and how to use it
2. recognise respiratory failure and when it may occur
3. perform a systematic and prioritised approach (the structured ABC approach) to the management of the infant or child who has a problem of the airway or breathing (see Section 4 Handbook 1 and Section 11 this Handbook).

Airway: equipment and skills for opening and maintaining the airway

Essential airway and breathing equipment include the following:

- face masks (ideally with reservoirs)
- airways, including laryngeal mask airways (LMAs) if anaesthetic skills are available
- self-inflating bag-valve-mask devices
- tracheal tubes, introducers, and connectors
- laryngoscopes
 - Magill's forceps
 - suction devices
 - surgical airway packs for performing an emergency surgical airway.

This equipment should be available in all resuscitation areas, ideally on a resuscitation trolley. It is crucial to gain familiarity with it before an emergency occurs.

Pharyngeal airways

There are two main types of pharyngeal airway, namely oropharyngeal (see Figures 13.1 and 13.2) and nasopharyngeal.

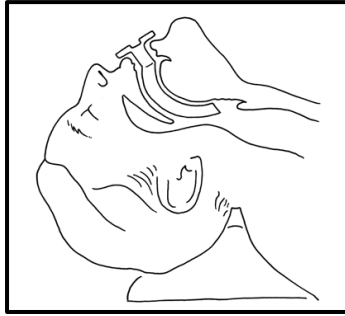


FIGURE 13.1 Oropharyngeal airway, showing position when inserted.

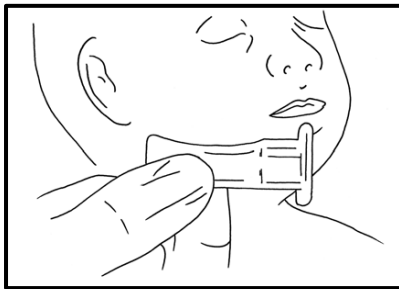


FIGURE 13.2 Oropharyngeal airway, showing sizing technique.

Oropharyngeal airways

The oropharyngeal or Guedel airway is used in the unconscious or obtunded patient to provide an open airway channel between the tongue and the posterior pharyngeal wall.

In the awake patient with an intact gag reflex, it may not be tolerated and inserting one may induce vomiting.

The oropharyngeal airway is available in a variety of sizes. A correctly sized airway when placed with its flange at the centre of the incisors, then curved around the face, will reach the angle of the mandible. Too small an airway may be ineffective, and too large an airway may cause laryngo- spasm. Either may cause mucosal trauma or may worsen airway obstruction. Reassessment following placement is therefore a vital part of safe insertion of an airway device.

There are two methods for inserting an oropharyngeal airway in children, depending on whether the child is small or large. However, there is no specific age of transition from one to the other – the choice of method depends on practicality and the skills of the operator. The important point is not to push the tongue back by inserting the airway carelessly. The twist technique is used for larger children. With this technique the convex side of the airway is used to depress the tongue as the airway is pushed into the mouth. The airway should be inserted upside down until the tip has passed the soft palate, and then rotated through 180 degrees so that the natural curve of the Guedel airway follows the curve of the tongue and pharynx (see Figure 13.3).

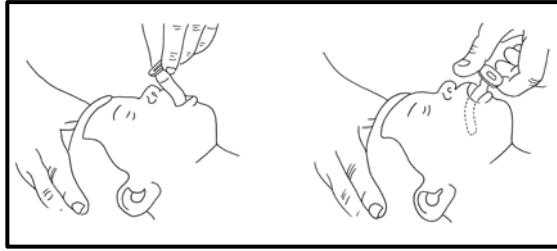


FIGURE 13.3 Oropharyngeal airway shown being inserted concave side up, then in place concave side down.

However, in infants and small children, as the tongue is larger relative to the size of the mouth, the airway cannot be rotated in the mouth without causing trauma. Therefore, the tongue is depressed with a tongue depressor and not by the convex side of the airway (see Figure 13.4).

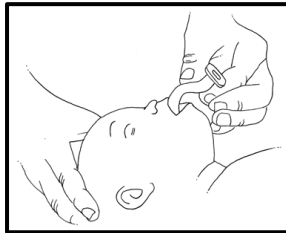


FIGURE 13.4 When inserting the airway without rotation, a tongue depressor can be helpful (not shown).

Nasopharyngeal airways

The nasopharyngeal airway is often better tolerated for longer term use than the Guedel airway. **It is contraindicated in fractures of the base of the skull.** It may also cause significant haemorrhage from the vascular nasal mucosa if it is not inserted with care, preferably with lubrication. A suitable length can be estimated by measuring from the lateral edge of the nostril to the tragus of the ear. An appropriate diameter is one that just fits into the nostril without causing sustained blanching of the alae nasi. If small-sized nasopharyngeal airways are not available, shortened endotracheal tubes may be used. Ensure that insertion of one or other of these devices results in an improvement in the patient's airway and breathing. If it does not improve the airway as shown by improved breathing, then a reappraisal of the choice or size of airway is urgently required.

Laryngoscopes

There are two principal designs of laryngoscope, namely straight bladed and curved bladed. The straight-bladed laryngoscope is usually employed to directly lift the epiglottis, thereby uncovering the vocal folds. The advantage of this approach is that the epiglottis is moved sufficiently so that it does not obscure the cords. The potential disadvantage is that vagal stimulation may cause laryngospasm or bradycardia.

The curved-bladed laryngoscope is designed to move the epiglottis forward by lifting it from in front. The tip of the blade is inserted into the mucosal pocket, known as the vallecula, anterior to the epiglottis, and the epiglottis is then moved forward by pressure in the vallecula. This may be equally effective for obtaining a view of the cords, and it has the

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advantage that less vagal stimulation ensues, as the mucosa of the vallecula is innervated by the glossopharyngeal nerve instead.

A laryngoscope blade appropriate for the age of the patient should be chosen. It is possible to intubate with a blade that is too long, but not with one that is too short.

Laryngoscopes are notoriously unreliable pieces of equipment which may develop flat batteries and unserviceable bulbs very quickly between uses. Therefore, it is vital that a spare is available at all times, and equipment must be regularly checked to ensure that it is in good working order.

Tracheal tubes

Uncuffed tubes should be used during resuscitation, by operators who do not have paediatric anaesthetic experience, for children up to approximately 10 years of age. If the operator is familiar with cuffed tube placement, both cuffed and uncuffed tubes are acceptable for infants and children undergoing emergency intubation, but not for neonates. Up until the age of around 10 years, the larynx is circular in cross section and the narrowest part of it is at the cricoid ring, rather than the vocal cords. An appropriately sized tube should give a relatively gas-tight fit in the larynx, but the fit should not be so tight that no leak is audible when the bag is compressed. Failure to observe this condition may lead to damage to the mucosa at the level of the cricoid ring, and to subsequent oedema following extubation.

The appropriate size of an uncuffed tracheal tube is estimated as follows:

internal diameter (mm) = (age in years/4) + 4 length (cm)

(age in years/2) + 12 for an oral tube length (cm)

(age in years/2) + 15 for nasal tube.

These formulae are appropriate for ages over 1 year. Neonates usually require a tube of internal diameter 3–3.5 mm, although preterm infants may need one of diameter 2.5 mm. Cuffed tubes should not be used in neonates.

For cuffed tracheal tubes, the appropriate internal diameter for children aged 2 years or older is estimated as follows:

internal diameter (mm) = (age in years divided by 4) + 3.5.

For infants of weight over 3 kg and up to 1 year in age a size 3 cuffed tube is usually acceptable, and for those aged 1–2 years a size 3.5 cuffed tube can generally be used.

The size of tracheal tubes is measured in terms of their internal diameter in millimetres. They are available in whole and half-millimetre sizes. The clinician should select a tube of appropriate size, but also prepare one a size smaller and one a size larger.

In the case of resuscitation in a young child where the lungs are very 'stiff' (e.g. in a cardiac arrest from severe bronchiolitis), a cuffed tube rather than an uncuffed tube may be used by a non-expert, but the risk of airway damage from the cuff must be balanced against the risk of failure to inflate the lungs.

Tracheal tube introducers

Intubation can be made easier using a stylet or introducer, which is placed through the lumen of the tracheal tube. There are two types – either soft and flexible or firm and malleable. The soft and flexible type can be allowed to project beyond the tip of the tube, so long as it is handled very gently. The firm and malleable type is used to alter the shape of the tube but

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can easily damage the tissues if allowed to protrude from the end of the tracheal tube. Tracheal tube introducers should not be used to force a tracheal tube into position. Bougies, which are flexible, deformable, blunt-ended gum elastic rods of different sizes, can be used to help to introduce a tracheal tube when access is difficult. A Seldinger-type technique is used. The bougie is introduced into the trachea using the laryngoscope, the endotracheal tube is then passed over it into the trachea, and finally the bougie is removed.

- 1 Lubricate the bougie with KY jelly.
- 2 Perform laryngoscopy. If the cords are not visible, identify landmarks to aid intubation.
- 3 Place the bougie into the pharynx and direct it into the larynx. If necessary, bend the bougie to negotiate the corner. Correct placement may be confirmed by detection of tracheal 'clicks' and 'hold-up' of the bougie (the absence of hold-up indicates oesophageal placement).
- 4 Hold the tube firmly in place and gently withdraw the bougie.
- 5 Remove the laryngoscope and confirm tube placement as usual.

Tracheal tube connectors

The proximal end of the tube connectors is of standard size, based on the 15-mm/22-mm system, which means that they can be connected to a standard self-inflating bag.

Magill's forceps

Magill's forceps (see Figure 13.5) are angled to allow a view around the forceps when they are in the mouth. They may be useful to help to position a tube through the cords by lifting it anteriorly, or to remove pharyngeal or supra-glottic foreign bodies.

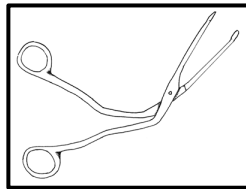


FIGURE 13.5 *Magill's forceps.*

Suction devices

These are used to remove blood, vomit and secretions from the mouth and throat, usually with a rigid suction tube (Yankauer suction tube; see below). In resuscitation areas, ideally the suction device should be connected to a central vacuum unit.

Portable suction devices are required for resuscitation when central suction is not available (as is the case in most resource-limited hospitals), and for transport to and from the resuscitation room. These are either manual, mains electrical or battery powered. A manual or battery-operated suction system must be available at all sites where resuscitation may be needed.

To clear the oropharynx of debris (e.g., vomit), a rigid sucker (e.g., Yankauer sucker) should be used with care not to damage delicate tissue or induce vomiting. The Yankauer sucker is available in both adult and paediatric sizes. It may have a side hole, which can be occluded by a finger, allowing greater control over vacuum pressure.

Tracheal suction catheters (see **Figure 13.6**) These may be required after intubation to remove bronchial secretions or aspirated fluids. In general, the appropriate size in French gauge which is numerically twice the internal diameter in millimetres (e.g., for a 3-mm tube the correct suction catheter is a French gauge 6).

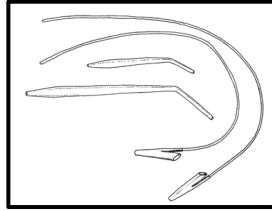


FIGURE 13.6 *Tracheal and oral or nasal suction catheters.*

Advanced airway techniques

Advanced airway techniques are used when the above techniques fail to maintain and protect an airway over the longer term, particularly if there is potential for it to become obstructed and thus prevent accurate control of oxygenation and ventilation. Advanced airway techniques (tracheal intubation, surgical cricothyroidotomy and surgical tracheostomy) are described in Section 90 Handbook 1.

Breathing: equipment and skills for helping the patient to breathe

The following equipment for oxygenation and ventilation should be immediately available in all areas where an emergency can occur and require treatment

- an oxygen source
- masks for those who are spontaneously breathing
- close-fitting face masks (for artificial ventilation)
- self-inflating bag-valve systems to be used with close-fitting face masks
- T-piece and open-ended bag systems (only to be used by those with anaesthetic skills)
- mechanical ventilators
- chest tubes
- gastric tubes.

Oxygen treatment

Indications

Give oxygen to patients:

1. with respiratory distress (severe indrawing of the lower chest wall, also known as recessions, raised respiratory rate, gasping, grunting with each breath, nasal flaring, head bobbing, etc.)
2. with cyanosis (blueness) that is central (around the lips and tongue, or inside the mouth in children with dark skin)
3. who are shocked
4. who are fitting
5. who are unconscious, with abnormal oxygen saturation (SaO₂) on a pulse oximeter.

Ideally, where the resources for this are available, oxygen therapy should be guided by pulse oximetry (see below). Give oxygen to children with an SpO₂ of < 94% and aim to keep SpO₂ at 94–98% (except at high altitude, where normal oxygen saturation levels are lower). If pulse oximeters are not available, the need for oxygen therapy has to be guided by clinical signs, which are less reliable.

Provision of oxygen

Oxygen must be always available. The two main sources of oxygen are cylinders and oxygen concentrators.

Oxygen cylinders contain compressed gas. A flow meter needs to be fitted to regulate flow. A hissing noise can be heard if gas is being delivered. Flow meters are used to ascertain how

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much oxygen is being delivered. Take the reading of flow rate from the middle of the ball. Always switch off the flow when the source is not in use (ensure that the indicator ball is at the bottom of the flow meter and not moving).

Do not leave anything inflammable near to the oxygen supply. Do not allow smoking near to the oxygen supply.

At least once a day, check that an adequate oxygen supply is available (use a signed logbook). If a gauge indicating the amount left in the cylinder is not available, switch on the flow and listen for a hissing noise. Replace empty cylinders promptly. Ensure that cylinders are stored and secured in an upright position in suitable containers so that they cannot fall over and cause injury. Cylinder keys to permit changes of regulator should be tied to each cylinder.

Oxygen concentrators may be available. They produce more than 95% oxygen with a flow of 1–8 litres/ minute but, unlike cylinders, they require a continuous electricity supply. For this reason, all areas where patients might need oxygen must have both cylinders and concentrators.

There are now small oxygen plants available that can provide oxygen for a defined area or even for the whole of a hospital or health facility. Some of them can be used to fill oxygen cylinders as well, thus providing a constant back-up (Diamedica)

Oxygen delivery

A mask with a reservoir bag (Figure 13.7) allows up to 100% oxygen to be delivered. Without a reservoir, it is only possible to deliver around 40% oxygen. If only low flow rates of oxygen are available, do not use a reservoir bag.

If an oxygen mask is being used, ensure that the mask is large enough to cover the mouth and nose. Both low- and high-flow oxygen (with a delivery rate of up to 15 litres/ minute) can be given. Hold the mask in place using the elastic strap around the back of the head or, in the case of a young child, ask the mother to hold it as close as possible to the child's face.



FIGURE 13.7 Oxygen masks with reservoir bags.

Nasal cannulae (also known as nasal prongs) (see Figure 13.8) are the preferred method of delivery in most circumstances, as they are safe, non-invasive, reliable and do not obstruct the nasal airway. Head boxes are not recommended, as they use up too much oxygen and deliver a low concentration. Face masks can be used for resuscitation purposes, ideally with a reservoir attached to deliver 100% oxygen.

Oxygen Monitoring

Nursing staff must know how to place and secure the nasal cannulae correctly. Check regularly that the equipment is working properly and remove and clean the cannulae at least twice a day.

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Monitor the patient at least every 3 hours to identify and correct any problems, including:

- SaO₂ values measured by pulse oximeter
- nasal cannulae out of position
- leaks in the oxygen delivery system
- incorrect oxygen flow rate
- airway obstructed by mucus (clear the nose with a moist wick or by gentle suction).

Pulse oximetry

Normal oxygen saturation at sea level in a child is 95–100%. Oxygen is ideally given to maintain oxygen saturation at 94–98%. Different cut-off values might be used at high altitude or if oxygen is scarce. The response to oxygen therapy in lung disease can be measured with the pulse oximeter, as the patient's SaO₂ should increase (in patients with cyanotic heart disease, SaO₂ does not change when oxygen is given). The oxygen flow can be titrated using the pulse oximeter as a monitor to obtain a stable SaO₂ of 94–98% without giving too much oxygen. This is especially important in pre-term babies with respiratory disease (see Section 91 Handbook 1 and Neonatal handbook).



FIGURE 13.8 Nasal cannulae delivering oxygen and taped in place.

Assessment of oxygenation at and above sea level

A systematic review in 2009 found an SpO₂ of 90% is the 2.5th centile for a population of healthy children living at an altitude of approximately 2500 m above sea level. This decreases to 85% at an altitude of approximately 3200 m.

See Section 55 for SpO₂ levels at different altitudes

Duration of oxygen therapy

Continue giving oxygen continuously until the patient can maintain an SpO₂ of 94–98% or higher in room air. When the patient is stable and improving, take them off oxygen for a few minutes. If the SpO₂ remains in the range 94–98%, discontinue oxygen, but check again 30 minutes later, and 3-hourly thereafter on the first day off oxygen to ensure that the patient is stable. Where pulse oximetry is not available, the duration of oxygen therapy has to be guided by clinical signs, which are less sensitive.

Breathing for the patient

Face masks with seal over nose and mouth for positive pressure ventilation (see Figure 13.9)

These face masks are used for either mouth-to-mask or, more commonly, bag-mask ventilation. Masks are available in various sizes, and the appropriate size to cover the mouth and nose should be chosen.

Face masks for mouth-to-mouth or bag-valve-mask ventilation in infants are of two main designs. Some masks conform to the anatomy of the patient's face and have a low dead

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space. Circular soft plastic masks give an excellent seal and are often preferred. Children's masks should be clear so that the child's colour or the presence of vomit can be seen. A pocket mask is a single-size clear plastic mask with an air-filled cushion rim designed for mouth-to-mask resuscitation. It can be supplied with a port for attaching it to an oxygen supply and can be used in adults and children. It can be used upside down to ventilate infants.

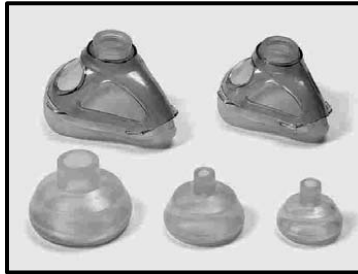


FIGURE 13.9 Face masks with cushioned rim for a leak-proof fit, and round shape for infants.

Self-inflating bags (see *Figure 13.10*)

This is one of the most important pieces of equipment, allowing hand ventilation by face mask **without** a supply of gas. The two appropriate sizes are 500 mL and 1600 mL (the smaller size for infants under 1 year of age, and the larger size for children and mothers). There is also a 300-mL version for small premature babies. These bags have pressure-limiting valves that operate at 30–45 cm H₂O. Test the valve by placing the mask on a surface and pressing the bag and ensuring that the valve opens. It can be overridden, if necessary, for stiff, poorly compliant lungs by loosening the screw at the top. The bag connects to the patient through a one-way valve to direct exhaled air to the atmosphere. The other end connects to the oxygen supply and can attach to a reservoir bag which allows high concentrations (up to 98%) of oxygen to be delivered. Without the reservoir bag, only concentrations of up to 40% can be delivered. The bag itself is easily dismantled and reassembled. It is important to realise that this system will operate without an attached oxygen supply, allowing resuscitation to be initiated before oxygen is available. However, if resuscitation is failing, check that oxygen is being delivered into the bag and to the patient and that the oxygen supply has not been disconnected.

Always use high-flow oxygen (if available) and a reservoir bag during resuscitation apart from at birth where room air is satisfactory for almost all babies (see Neonatal Handbook and section 11 here).

It is important to clean the system after each patient.

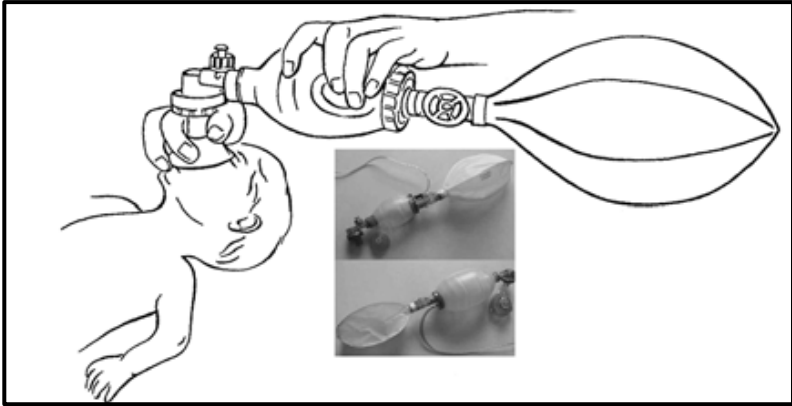


FIGURE 13.10 Two sizes of self-inflating bags and masks.

It is essential that the mask is properly sized and correctly placed over the mouth and nose of the patient (see Figures 13.11 and 13.12).

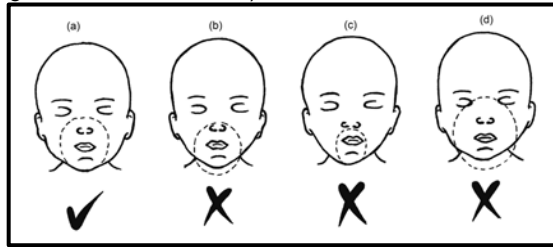


FIGURE 13.11 (a) Correct placement of infant mask. (b), (c) and (d) Incorrect placement of infant mask.



FIGURE 13.12 Two views of single-handed grip on mask.

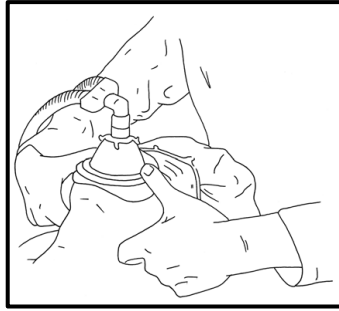


FIGURE 13.13 *Two-handed grip on mask incorporating jaw thrust.*

If the chest does not rise, the airway is not clear. The usual cause is failure to correctly apply the airway-opening techniques discussed previously. The first step to try is to readjust the head-tilt/chin-lift position and try again. If this is not successful, the jaw-thrust manoeuvre should be tried (see Figure 13.13). Failure of both the head-tilt/chin-lift and jaw-thrust manoeuvres should lead to suspicion that a foreign body is causing the obstruction.

Once breathing has restarted, replace the bag- valve-mask system with a simple face mask and reservoir. Because of the internal valves it is not possible to spontaneously breathe through the bag-valve-mask system.

Chest tubes

In cases with a significant haemothorax or pneumothorax (particularly tension pneumothorax), ventilation will be compromised, and insertion of a chest drain is mandatory (see Section 91 Handbook 1). ▼

Gastric tubes

Insertion of a gastric tube is essential after intubation and may also relieve respiratory distress in spontaneously breathing patients with abdominal emergencies or gastric stasis. It allows decompression of a stomach full of air from both bag and mask ventilation as well as air swallowed by a distressed patient. Without a gastric tube, the patient may vomit or there may be aspiration of stomach contents. In addition, venting of stomach gas will avoid diaphragmatic splinting. A nasogastric tube will increase airway resistance through the nose, which in a spontaneously breathing infant with respiratory failure can be significant. An orogastric tube has less effect on ventilation but is less readily tolerated and less easily fixed in position.

Further information

Additional breathing procedures are described in Section 35 Handbook 1 (on spacers and nebulisers).

Circulation: equipment and skills for maintaining the circulation

Details of how to undertake the following procedures are covered in Section 92 Handbook 1:

- peripheral venous cannulation
- blood sampling from an IV cannula
- intraosseous cannulation and infusion
- cutdown long saphenous venous cannulation
- insertion of central venous catheters
- needle pericardiocentesis.

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Management of cardiac arrest in children (see *Advanced Obstetric Handbook for management in adolescent girls who are pregnant*)

Cardiac arrest occurs when there is no effective cardiac output. Before any specific therapy is started, effective basic life support (as described above) must be established.

Four cardiac arrest heart rhythms can occur:

1. asystole
2. pulseless electrical activity (including electromechanical dissociation)
3. ventricular fibrillation
4. pulseless ventricular tachycardia.

These can be divided into two groups.

1. Asystole and pulseless electrical activity (PEA), which do not require defibrillation, are called '**non-shockable**' rhythms.
2. Ventricular fibrillation and pulseless ventricular tachycardia, which do require defibrillation, are called 'shockable' rhythms.

An electrocardiogram (ECG) is needed to make the diagnosis. ECG can be monitored directly or as part of an Automatic External Defibrillator (AED) see later in this Section.

Reversible causes of cardiac arrest

The causes of cardiac arrest are multifactorial, but the two commonest final pathways in childhood are through hypovolaemia and hypoxia. All reversible factors are conveniently remembered as the 4Hs and 4Ts (see below). Sometimes cardiac arrest is due to an identifiable and reversible cause, such as shock due to massive haemorrhage from major trauma, septicaemia or severe diarrhoea. In the trauma setting, cardiac arrest may be caused by severe hypovolaemia or tension pneumothorax or pericardial tamponade (see Section 79 Handbook 1).

It is often appropriate to give an early IV 10 ml/Kg bolus of Ringer-lactate/Hartmann's solution as this will be supportive in cases related to severe hypovolaemia. In addition, however, a tension pneumothorax requires definitive treatment by needle thoracocentesis. Continuing blood replacement and the prevention of further haemorrhage may also be required.

Rapid identification and treatment of reversible causes such as hypovolaemic shock, hypothermia, electrolyte, and acid-base disturbance, tension pneumothorax and pericardial tamponade are vital.

During CPR it is important to continually consider, and correct reversible causes of the cardiac arrest based on the history of the event and any clues that are found during resuscitation.

The 4Hs

1. **Hypovolaemia** is the most prevalent cause. Significant hypovolaemia may also be associated with trauma, gastroenteritis, anaphylaxis, and sepsis. Control of haemorrhage and urgent IV infusion of blood or in gastroenteritis intravenous crystalloid IV boluses must be given.
2. **Hypoxaemia** due to respiratory or heart failure is another major cause of cardiac arrest and its early reversal is key to successful resuscitation.
3. **Hyperkalaemia**, **Hypokalaemia**, **Hypocalcaemia**, and other metabolic abnormalities may be suggested by the patient's underlying condition (e.g., renal failure, severe gastroenteritis), tests taken during the resuscitation, or clues from the ECG.

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4. Hypothermia is associated with drowning incidents and requires particular care. A low-reading thermometer must be used to detect it.

The 4Ts

1. Tension pneumothorax in major trauma
2. Cardiac Tamponade in major trauma
3. Toxins/poisons, resulting either from accidental or deliberate overdose or from a medical mistake, may require specific antidotes.
4. Thrombo-embolic phenomena (pulmonary embolus: rare in children).

Non-shockable cardiac arrest

Asystole

This is the most common cardiac arrest rhythm. The response of the heart to prolonged severe hypoxia and shock (which are the usual pathologies) is progressive bradycardia leading to asystole.

The ECG will distinguish asystole from ventricular fibrillation, ventricular tachycardia and pulseless electrical activity. The ECG appearance of asystole is an almost straight line; occasionally P-waves are seen (see Figure 13.14). Check that the appearance is not caused by an artefact (e.g. a loose wire or disconnected electrode). Turn up the gain on the ECG monitor.

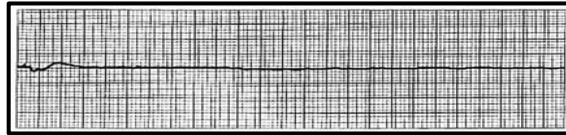


Figure 13.14 ECG appearance of asystole.

Pulseless electrical activity (PEA)

This is the absence of a palpable pulse or other signs of life despite the presence on the ECG monitor of recognisable ECG complexes that normally produce a pulse (see Figure 13.15). PEA is treated in the same way as asystole and often leads into asystole.

PEA can occur with major trauma, often with an identifiable and reversible cause such as severe hypovolaemia, tension pneumothorax or pericardial tamponade. PEA is also seen in hypothermic patients and in those with electrolyte abnormalities.

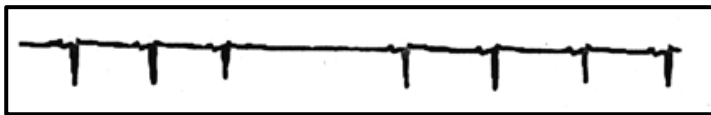


Figure 13.15 Pulseless electrical activity (PEA) in a patient with no pulse or signs of life.

Management of asystole/PEA

The first essential step is to establish ventilations and chest compressions effectively. Ensure an open airway, initially using an airway manoeuvre to open the airway and stabilising it with an oropharyngeal airway.

Ventilations are provided initially by bag and mask with high-concentration oxygen.

Provide effective chest compressions at a rate of approximately 100 per minute with a compression: ventilation ratio of 15:2. The depth of compression should be at least one-third of the antero-posterior diameter of the chest, and compressions should be given in the middle of the lower half of the sternum.

If asystole or PEA is identified give adrenaline 10 micrograms per Kg body weight intravenously or intra-osseous (IO). (0.1 mL/Kg of 1:10,000 solution/ kg). Adrenaline increases coronary artery perfusion, enhances the contractile state of the heart and

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stimulates spontaneous contractions. Where there is no existing IV access, the IO route is recommended as the route of choice, as it is rapid and effective. In each case, the adrenaline is followed by a normal IV flush of 2 mL of R/L or 0.9% saline.

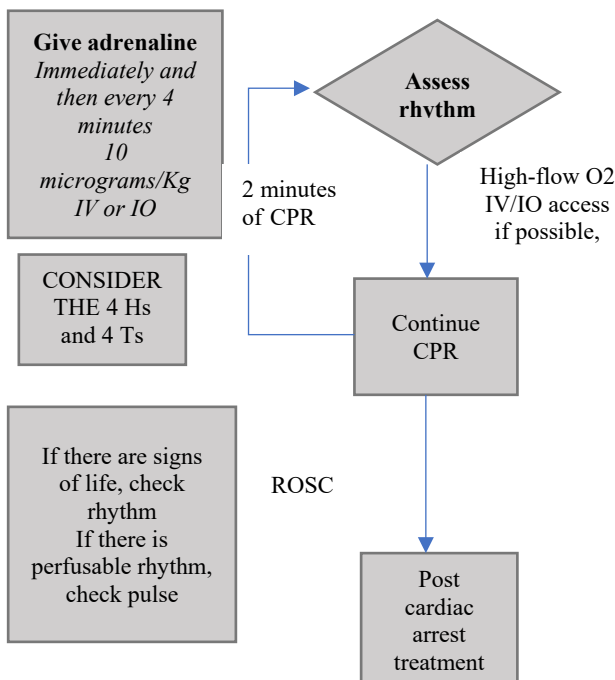
If available, and as soon as is feasible, a skilled and experienced operator (usually an anaesthetist) should intubate the patient's airway. This will both control and protect the airway and enable chest compressions to be given continuously, thus improving coronary perfusion. Once the patient has been intubated and compressions are uninterrupted, the ventilation rate should be around 10 to 12 breaths per minute. It is important for the team leader to check that the ventilations remain adequate when chest compressions are continuous. A guideline for non-shockable rhythms is shown in Figure 13.16.

During and following adrenaline treatment, chest compressions and ventilation should continue. The only reason for interrupting compressions and ventilation is to shock the patient, if necessary (see below under "shockable" rhythms), and to check the rhythm. A brief interruption may be necessary during difficult intubation. Giving chest compressions is tiring for the operator, so if enough personnel are available, change the operator frequently and ensure that they are achieving the recommended rate of 100 compressions per minute together with a depression of the chest wall by at least one-third of its antero-posterior diameter.

At intervals of about 2 minutes during the delivery of chest compressions, pause briefly to assess the rhythm on the ECG monitor or AED. If asystole persists, continue CPR while again checking the electrode position and contact.

1. If there is an organised rhythm, check for a pulse and signs of life.
2. If there is a Return Of Spontaneous Circulation (ROSC), continue post-resuscitation care, continuing the ventilation rate of around 10 to 12 breaths per minute.
3. If there is no pulse and no signs of life, continue the protocol.
4. Give adrenaline about every 4 minutes at a dose of 10 microgram/Kg

Figure 13.16 Guideline for the treatment of non-shockable (asystole and PEA) rhythms. CPR, cardiopulmonary resuscitation; IV, intravenous; IO, intra-osseous; ROSC, return of spontaneous circulation



Shockable cardiac arrest

Figure 13.17 ventricular fibrillation.

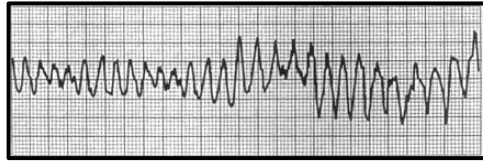
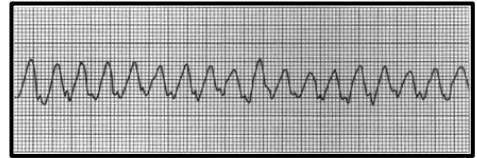
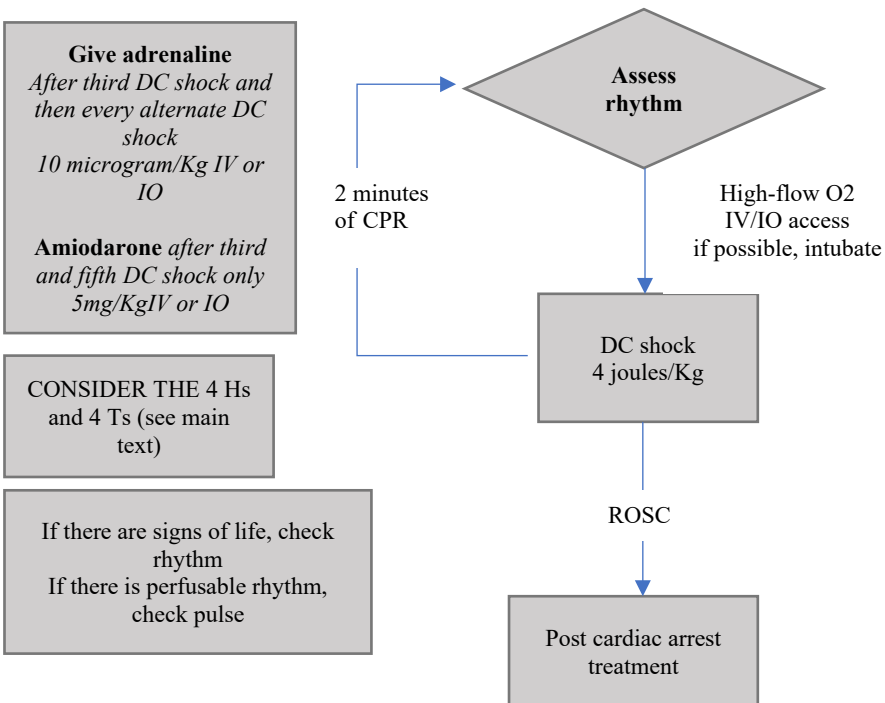


Figure 13.18 Pulseless ventricular tachycardia (pVT)



These arrhythmias are uncommon in children but either of them must be considered in patients with sudden witnessed collapse, hypothermia, poisoning by tricyclic antidepressants, congenital heart disorders or cardiac disease. The guideline for treating ventricular fibrillation (VF) (see Figure 13.17) and pulseless ventricular tachycardia (pVT) (Figure 13.18)

Figure 13.19 Algorithm for the treatment of shockable (VF and pVT) rhythms in children. CPR, cardiopulmonary resuscitation; IV, intravenous; IO, intra-osseous; ROSC, return of spontaneous circulation.



Undertaking ECG monitoring and placing ECG pads

For ECG or AED monitoring, and defibrillation, paediatric paddles (4.5 cm) should be used for children under 10 kg. One electrode is placed over the apex of the heart in the mid-axillary line, while the other is placed immediately below the clavicle just to the right of the

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sternum. If the paddles are too large, one should be placed on the upper back, below the left scapula, and the other should be placed on the front, to the left of the sternum.

For infants under 1 year of age, a manual defibrillator which can be adjusted to give the correct shock is recommended. However, if an AED is the only defibrillator available, its use should be considered, preferably with paediatric attenuation pads.

Automated external defibrillators (AEDs) are now commonly and publicly available in well-resourced countries and may well be a good way forward in low resource settings. The standard adult shock is used for children over 8 years of age. For children under 8 years, attenuated paediatric paddles should be used with the AED (if available). Many AEDs can detect VF/VT in children of all ages and differentiate between 'shockable' and 'non-shockable' rhythms with a high degree of sensitivity and specificity.

Undertaking defibrillation as part of CPR

If the patient's ECG is already being monitored, the rhythm might be identified before significant deterioration occurs. In unmonitored patients, basic life support will have been started in response to the collapse, and VF/pVT will be identified when the cardiac monitor or AED is put in place.

With immediate identification of VF/pVT, asynchronous electrical defibrillation of 4 Joules per Kg should be undertaken immediately, and the guideline continued as described below.

It is essential that during defibrillation, no person other than the patient receives the shock. It is the responsibility of the person giving the shock to ensure that other staff and/or relatives are not touching the patient.

First shock. An asynchronous shock of 4Joules/Kg in a child should be given immediately, and CPR immediately resumed without reassessing the rhythm or feeling for a pulse. Immediate resumption of CPR is vital because there is a pause between successful defibrillation and the appearance of an effective rhythm on the monitor. Cessation of chest compressions will reduce the likelihood of a successful outcome if a further shock is needed. However, no harm accrues from 'unnecessary' compressions.

If the first shock fails to defibrillate (convert the rhythm to produce an effective pulse), attention must revert to supporting coronary and cerebral perfusion as in asystole. Although the procedures for stabilising the airway and obtaining circulatory access are described above sequentially, they should be undertaken simultaneously under the direction of a resuscitation team leader. It is important for the team leader to check that the ventilations remain adequate when chest compressions are continuous.

The airway should be secured, the patient ventilated with high-flow oxygen, and effective chest compressions continued at a rate of 100 per minute, with a compression depth of at least one-third of the antero-posterior diameter of the chest, and a ratio of 15 compressions to 2 ventilations. As soon as is feasible, a skilled and experienced operator should intubate the child's airway (if available).

Intubation will both control and protect the airway and enable chest compressions to be given continuously, thus improving coronary perfusion. Once the patient has been intubated, compressions should be uninterrupted, and the ventilation rate should be around 10 to 12 breaths per minute. It is important for the team leader to check that the ventilations remain adequate when chest compressions are continuous.

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Obtain circulatory access. Whenever venous access is not readily obtainable, intra-osseous access should be considered early on in children, as it is rapid and effective. In each case, any drug given as described below is followed by a crystalloid flush (2–5 mL depending on size of the child).

Second shock Two minutes after the first shock, pause the chest compressions briefly to check the monitor. If VF/VT is still present, give a second shock of 4 joules/kg in a child and immediately resume CPR, commencing with chest compressions. Consider and correct reversible causes (the 4Hs and 4Ts) while continuing CPR for a further 2 minutes.

Third shock Pause briefly to check the ECG monitor/AED. If the rhythm is still VF/VT, give a third shock of 4 Joules/Kg.

Once chest compressions have resumed, give adrenaline 10 micrograms/Kg IV (0.1ml/Kg of 1 in 10,000 adrenaline) in a child and if available amiodarone 5mg/Kg IV or IO in a child, flushing after each drug. Continue CPR for 2 more minutes.

Fourth shock After completion of the 2 minutes of CPR, pause briefly to check the monitor, and if the rhythm is still VF/VT give an immediate fourth shock of 4 Joules/Kg and resume CPR. Continue CPR for 2 minutes.

Fifth shock After a further 2 minutes of CPR, pause briefly to check the monitor and if the rhythm is still shockable, give an immediate fifth shock of 4 Joules/Kg. Once chest compressions have resumed, give a second dose of adrenaline 10 micrograms/Kg in a child and a second dose of amiodarone of 5 mg/Kg IV or IO in a child.

Continued shocks After completion of the 2 minutes of CPR, pause briefly before the next shock to check the monitor. Continue giving shocks every 2 minutes, minimising the pauses in CPR as much as possible. Give adrenaline after every alternate shock (i.e. every 4 minutes) and continue to seek and treat reversible causes.

In addition, if at any stage there are signs of life (Return Of Spontaneous Circulation ROSC), such as regular respiratory effort, coughing or eye opening, stop CPR and check the monitor.

- If the rhythm is still VF/VT, continue with the sequence as described above.
- If the rhythm is asystole, change to the asystole/PEA sequence.
- If organised electrical activity is seen, check for signs of life and a pulse. If there is ROSC, continue post-resuscitation care.
- If there is no pulse (or a pulse of < 60 beats/minute) and no other signs of life, continue the asystole/PEA sequence.

In VT or VF that does not respond to the above sequence consider giving magnesium sulphate IV as a bolus of 25-50mg/Kg (up to an individual maximum dose of 2 grams in a child).

Sodium bicarbonate

If VF/VT is due to tricyclic antidepressant overdose or hyperkalaemia, sodium bicarbonate may be helpful. Give 1 mmol/Kg IV/IO (1ml/Kg of an 8.4% solution or 2ml/Kg of a 4.2 % solution) in a child.

Amiodarone

Amiodarone is the treatment of choice in shock-resistant ventricular fibrillation and pulseless ventricular tachycardia. The dose of amiodarone for VF/pulseless VT is 5mg/Kg by rapid IV/IO bolus.

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Lidocaine is an alternative to amiodarone if the latter is unavailable. The dose is 1mg/Kg 100 mg IV or IO as a bolus in a child.

It is DC shock that converts the heart back to a perfusing rhythm, not the drug. The purpose of the anti-arrhythmic drug is to stabilise the converted rhythm, and the purpose of adrenaline is to improve myocardial oxygenation by increasing coronary perfusion pressure. Adrenaline also increases the intensity of ventricular fibrillation, which increases the success rate of defibrillation.

Automatic external defibrillators (AEDs)

The use of the AED is now included in basic life support teaching for adults because early defibrillation is the most effective intervention for the majority of unpredicted cardiac arrests in adults. In children there may also be a primary cardiac cause of cardiac arrest, and the use of an AED may be lifesaving.

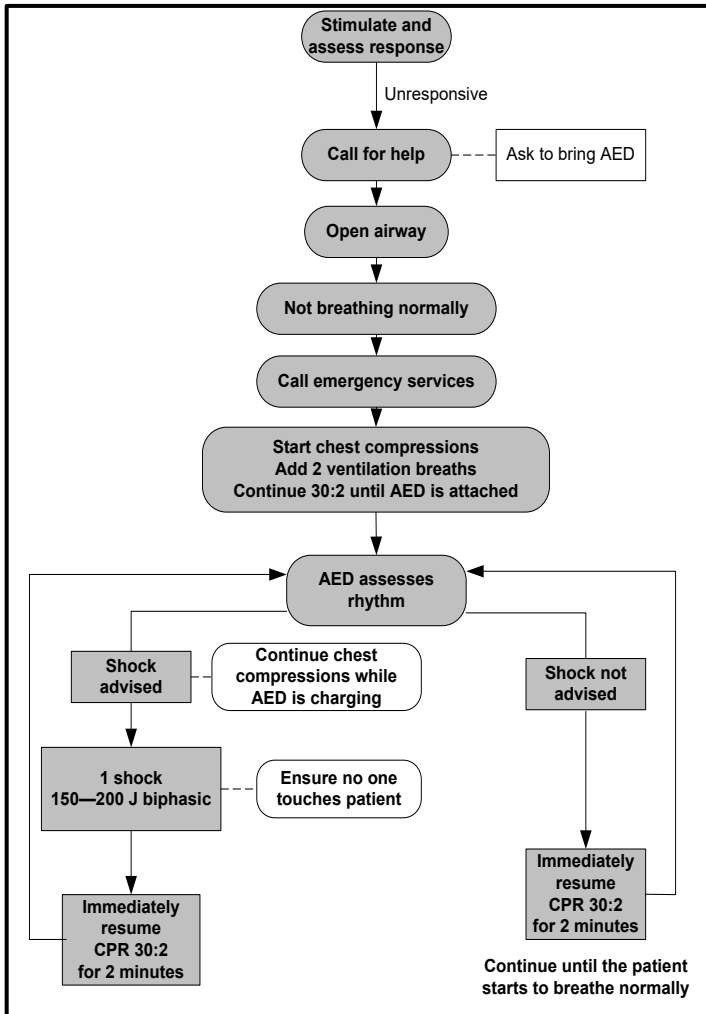
These devices are becoming widely available and are relatively inexpensive. They are lifesaving in cases where there is **cardiac arrest with a shockable rhythm** and have been designed for community use.

If defibrillation is to be successful, it must be performed within 15 minutes of the onset of fibrillation (and the earlier it is performed, the greater the likelihood of success). AEDs are also now widely used in the treatment of hospital cardiac arrests and are therefore included here.

Application of an AED

1. Expose the chest and place one adhesive defibrillator pad on the patient's chest to the right of the sternum below the right clavicle, and one in the mid-axillary line on the left side of the chest. Keep the axillary electrode vertical to maximise efficiency.
2. If a shock is indicated, most AED devices will do this automatically, but some will ask the operator to deliver the shock by pressing a button.
3. Immediately after the shock, resume compressions for 2 minutes, after which there will be a further prompt for a rhythm analysis.
4. **Always make sure any oxygen being given to the patient when activating the AED is switched off to avoid a fire or explosion.**
5. If defibrillation is not indicated, CPR should be continued for 2 minutes, at which stage the AED will prompt further analysis of the rhythm.
6. **It is essential that during defibrillation, no person other than the patient receives the shock. It is the responsibility of the person giving the shock to ensure that other staff and/or relatives are not touching the patient.**

Figure 13.20 Algorithm for automatic external defibrillator (AED) use (note adult rate of 30 compressions to two ventilations is illustrated but in children we recommend 15/2)



Drugs used in non-shockable and shockable cardiac arrest

Remember it is DC shock that converts the heart back to a perfusing rhythm, not the drug. The purpose of the anti-arrhythmic drug is to stabilise the converted rhythm, and the purpose of adrenaline is to improve myocardial oxygenation by increasing coronary perfusion pressure. Adrenaline also increases the intensity of ventricular fibrillation, which increases the success rate of defibrillation.

Oxygen

Although 100% oxygen must be used during the resuscitation process, once there is Return Of Spontaneous Circulation (ROSC) hyperoxia may be detrimental to tissues that are recovering. Pulse oximetry should be used to monitor and adjust for oxygen requirement after a successful resuscitation. SpO₂ should be maintained in the range 94–98 %. **Always**

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ensure that oxygen delivery is discontinued during defibrillation shocks, to avoid the risks of explosions and fire.

Adrenaline

Adrenaline is the first-line drug for treatment of cardiac arrest. Its effect is to increase blood flow to the brain and myocardium. It renders the myocardium more susceptible to defibrillation.

The initial IV or IO dose is 10 micrograms/Kg (0.1ml/Kg of 1 in 10,000 solution) in a child. In patients with no existing IV access, the intra-osseous route is recommended as the route of choice, as it is rapid and effective. In each case, adrenaline is followed by a 0.9% saline flush (2 to 5 mL).

Amiodarone

Amiodarone is the treatment of choice in shock-resistant ventricular fibrillation and pulseless ventricular tachycardia. The dose of amiodarone for VF/pulseless VT in a child is 5 mg/kg via rapid IV/IO bolus.

Lidocaine is an alternative to amiodarone if the latter is unavailable. The dose is 1 mg/kg IV or IO in a child.

Sodium bicarbonate

Good basic life support is more effective than sodium bicarbonate, which may be considered if spontaneous circulation has not returned after the first or second dose of adrenaline. Sodium bicarbonate is however, recommended in the treatment of patients with VT/VF due to hyperkalaemia and tricyclic antidepressant overdose (see above).

The dose is 1 mmol/Kg IV/IO (1ml/Kg of an 8.4% solution or 2 ml/Kg of a 4.2% solution) in a child.

Note that sodium bicarbonate inactivates adrenaline and dopamine, so the line must be flushed with 0.9% saline if these drugs have been or are subsequently given.

Also, sodium bicarbonate must not be given in the same intravenous line as calcium solutions as precipitation will occur.

Glucose

All patients, but especially infants and preschool children, can become hypoglycaemic when seriously ill. Blood glucose levels should therefore be checked frequently, and hypoglycaemia must be corrected. Hypoglycaemia is defined as a glucose concentration of less than 2.5 mmol/litre (45 mg/dL).

If hypoglycaemia is suspected but blood glucose levels cannot be measured, always give 2 ml/Kg of 10% glucose in a child preferably IV or IO or alternatively enterally (via a gastric tube). If blood glucose levels can be measured, avoid hyperglycaemia (maintain blood glucose concentration below 12 mmol/litre).

The decision to abandon CPR if it is unsuccessful. When to stop resuscitation?

Local guidelines should be in place. Resuscitation efforts are unlikely to be successful, and cessation can be considered, if there is no return of spontaneous circulation at any time after 20 minutes of life support and in the absence of recurring or refractory VF/VT.

The exceptions are patients with a history of poisoning or a primary hypothermic insult, in whom prolonged attempts may occasionally be successful. Prolonged external cardiac

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compressions during which central (femoral or arterial) pulses were felt have successfully resuscitated patients with tricyclic antidepressant overdose.

The presence of the parents at the child's side during resuscitation enables them to gain a realistic understanding of the efforts made to save their child's life. In general, family members should be offered the opportunity to be present during the resuscitation of their child.

A staff member (if available) must be designated as the parents' support and interpreter of events at all times.

The team leader, not the parents, decides when it is appropriate to stop the resuscitation.

If the presence of the parents is impeding the progress of the resuscitation, they should be sensitively asked to leave.

The team needs a debriefing session to support staff and reflect on practice.

Section 14. High-dependency care in children including those who are pregnant

Introduction

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

High-dependency care is a low-volume, high-demand specialty. Women and girls with complications of pregnancy and children under 2 years of age account for most of those who require high-dependency care. There also may be seasonal variation, with a peak in the winter months associated with respiratory-related illness, or a peak in the rainy season associated with malaria.

Dedicated 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 that provide this service. However, most patients who require high-dependency or intensive care will present to smaller peripheral hospitals rather than to large tertiary centres. Therefore, it is absolutely essential that staff in smaller district hospitals are able to recognise and appropriately treat sick children and pregnant women in the early stages of their illness (see EESS-EMNCH programme – Essential and Emergency Surgical Skills– *Emergency Maternal, Neonatal and Child Healthcare Manual and Pocket Book*, which can be found on the MCAI website: <https://www.mcai.org.uk> Accessed 28.03.2021

All medical and nursing staff who undertake high-dependency care should be well trained in emergency care so as to be able to stabilise critically ill or injured patients, and initiate appropriate medical therapy, which may involve intubation and ventilation. A proportion of such patients may then be safely transferred to an intensive care unit if this is still appropriate and a bed is available. Often, with good initial resuscitation and early diagnosis and treatment, the need for intensive care can be avoided. In a patient who requires intensive care, there should be early consultation with the regional/national intensive-care unit, usually by telephone or radio, so that further management can be jointly decided until a retrieval team, if available, arrives to collect the patient.

Transportation of critically ill patients particularly those receiving assisted ventilation, requires appropriately trained staff and equipment. Transportation is best thought of as ‘a *high-dependency care bed on wheels*’, and the aim should be that the patient does not deteriorate during transport. Before the patient is moved, proper resuscitation and stabilisation are essential.

Children and pregnant mothers exhibit fundamental differences that influence the training of staff and the type and size of equipment available. These differences extend across anatomy, physiology, pharmacology, and behaviour. However, both of these patient groups have less reserve and tend to decompensate early and quickly. They also have a greater capacity to make a full recovery.

Provision of high-dependency care is not just about equipment and facilities. The surrounding environment and contact with their family is crucial to the promotion of a patient’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 resources (see Table 14.1). In most resource-limited countries, only Level 1 care is likely to be available, and then only in the most well-funded hospitals, such as those in the capital cities or where medical students are trained.

Most patients can be managed at Level 1 with close monitoring, good nursing care and appropriate medical therapy. By providing optimal therapy it is often possible to prevent the deterioration of the patient (e.g., through good fluid management, early but appropriate treatment with antibiotics, and the use of oxygen).

Many hospitals will have poor outcomes if patients must be ventilated in sites where there is a lack of maintained ventilators, and no reliable oxygen source or blood gas analyser. As far as possible in countries with limited resources, intubation and ventilation should be avoided until they are necessary. Many patients can tolerate high pCO₂ levels with a compensated pH – it is hypoxia that is potentially fatal. It may be appropriate to develop and have available non-invasive modes of ventilatory support, such as nasal mask or cannula continuous positive airways pressure (CPAP), nasal or face mask intermittent positive pressure ventilation (IPPV), bilevel positive airways pressure (BiPAP), or negative pressure ventilation (CNEP or INPV). Similarly, the more invasive a procedure or monitoring process is, the greater the risk of complications.

Finally, it is essential that hospitals which provide high-dependency care have an on-site biomedical engineer to keep all the equipment serviced and safe.

TABLE 14.1 Levels of high dependency and intensive care

Level 3 (intensive care)
Multi-organ failure
Ideally one or more nurses per patient
Invasive monitoring
Examples: ventilation, hemofiltration
Optimise medical therapy
Level 2 (intensive care)
Single-organ failure
Ideally one nurse per patient
Non-invasive or invasive monitoring
Example: ventilation (intubation and extubation)
Optimise medical therapy
Level 1 (high-dependency care)
Requirement for closer observation and monitoring than is available on the standard ward
Ideally one nurse for every two patients
Non-invasive monitoring
Examples: after major surgery, non-intubated child with severe croup, pregnant woman with severe pre-eclampsia or eclampsia

Minimum standards for a lead centre providing intensive care

Medical staff

- Senior doctors or physician assistants with appropriate training in high-dependency care medicine.
- Training programme for junior medical staff specialising in high-dependency care.
- Provision of 24-hour cover at both senior and junior level.
- Resident junior cover for 24 hours by staff with skills in emergency care and resuscitation, whose only clinical responsibility is to the high-dependency care unit.
- Access on site to other specialist consultants (e.g., obstetrician, paediatrician, ENT surgeon, anaesthetist).

Nursing staff

- Nursing staff with training in high-dependency care and resuscitation.
- Ongoing training and support for nursing staff.
- Continuous 24-hour observation of each patient at all times by a nurse qualified in high-dependency care, with observations documented.

Support staff

- Availability of a physiotherapist.
- Availability of a pharmacist 24 hours a day.
- Availability of a dietitian.
- Availability of a biomedical engineer 24 hours a day.

Equipment and drugs

- Medical staff and nursing staff with training in how to use all equipment.
- Equipment maintained on a regular basis and according to manufacturer's guidelines by a biomedical engineer.
- Controlled drugs, especially morphine, available immediately and for 24 hours a day.

Retrieval service

- Available 24 hours a day from the community or other health facilities without high-dependency care (e.g. via an emergency ambulance service; see www.reproductive-health-journal.com/content/pdf/1742-4755-7-21.pdf Accessed 31.03.2021)
- Does not take staff from the high-dependency unit, leaving it uncovered.
- Usually an experienced doctor, midwife or nurse.
- Able to provide phone or radio advice.
- Equipped with portable battery-operated monitors (ECG, heart rate, respiratory rate, oxygen saturation) and suction. Possible to provide hand ventilation by bag and face mask or endotracheal tube rather than have a transport ventilator.

Clinical effectiveness and management

Protocols for admissions, discharges, retrievals, resuscitation, and stabilisation, and for treating major conditions.

Data collection and regular audit of deaths and near-miss cases to improve care provided.

Facilities for families

Access for carers of children and partners of pregnant mothers always.

Accommodation and food for families.

Maternal and Child Health Initiative (MCHI) environment https://12dec1c3-e3f4-e784-7f3a-6e7903610962.filesusr.com/ugd/dd2ba4_75777218b5234c91b817072eb1433ee4.pdf

Accessed March 31 2021

Essential equipment for the high-dependency care of children

1. Beds that are manually operated to tilt the head up or feet down.
2. Wedges for lateral tilt for children who are pregnant.
3. Suction systems and suction catheters, both electrical and manual (ideally wall suction).
4. Pulse oximeters and ECG monitors (one for each bed).

Section 14. High-dependency care in children including those who are pregnant. [Dr. Martin Samuels](#)

5. Resuscitation trolley containing drugs and equipment (particularly oropharyngeal airways, laryngoscopes with spare bulbs, endotracheal tubes and introducers, bag-valve-masks (child and adult), masks with reservoir bags).
6. Mobile screens.
7. Mobile oxygen cylinders and one oxygen concentrator for each bed, with face masks and nasal cannulae (infant, child and adult).
8. Wall sockets (six per bed).
9. One basic infant ventilator and one child/adult ventilator.
10. Nasal or mask CPAP systems (neonatal, child and adult).
11. Two automatic external defibrillators (AEDs).
12. Infusion pumps (if there are sufficient staff).
13. IV drip stands.
14. Basic CVP monitoring system.
15. Blood warmer (ideally).
16. Fridge for pharmacy drugs.
17. Fridge for blood for transfusion.
18. Lockable cupboard for drugs not needing refrigeration.
19. Metal lockable cupboard for controlled drugs.
20. Cupboard for storing IV fluids.
21. End-of-bed chart tables and specially designed high-dependency care charts.
22. One portable ultrasound scanner.
23. One portable fetal heart monitor.
24. Burette giving sets.
25. IV infusion monitors
26. Portable examination light.
27. Portable fans.
28. Suitable storage boxes (preferably easy to clean and label).
29. Blackboard for documenting priority issues for each patient.
30. Wall-mounted pathways of care.
31. Hand wash facilities × 3/4.
32. Separate sluice and patient and staff toilet and washing facilities.
33. Steriliser.

For details of procedures that are likely to be used in high-dependency care, see obstetric handbook and Sections 80 and 90-94 Handbook 1.

Section 15 Cancer in children

Introduction

- **More than 85% of all newly diagnosed children with cancer and 95% of deaths in children with cancer occur in low- and middle-income countries.**
- With an increasing global population, principally in resource-limited countries, the number of children will continue to increase both in terms of absolute numbers and proportionally in these countries.
- As malnutrition and infection decline, particularly in young children, the worldwide contribution to mortality from cancer will increase.
- Only a limited proportion of all children with cancer in resource-limited countries receive curative therapy, and most do not even receive any form of palliative care.
- A child diagnosed with cancer who lives in one of the poorest countries has an 80% probability of dying, compared with less than 30% in the most well-resourced countries (see Figure 15.1).

Epidemiology

Globally, the reported incidence rate of cancer in children (aged < 15 years) ranges from 80 to 150 per million per year. However, there are some differences between resource-limited and resource-rich countries. The incidence rates in children from low- and middle-income countries are towards the lower end of the range, which may partly be due to both under-diagnosis and under-reporting. It is currently estimated that globally approximately half of children with cancer are never diagnosed (and consequently do not get treatment) and that the true global burden of cancer in those aged <15 years is around 400,000 every year. Boys are around 20% more likely to develop cancer overall than girls. The ratio of boys to girls registered with childhood cancer increases with decreasing gross domestic product and with increasing infant mortality, suggesting a gender bias in diagnosing and registering cases in some resource-limited countries.

There are clear variations in the incidence of different childhood cancers around the world for example, a reported excess of retinoblastoma in India, Pakistan and sub-Saharan Africa. It is likely that some of this 'excess' is due to better diagnosis and recognition of retinoblastoma, which once at an advanced stage is easy to identify. On the other hand, the incidence of brain tumours and neuroblastoma is generally lower in resource-limited settings, and this may be due to varying levels of case ascertainment. In many countries, most noticeably those in sub-Saharan Africa, the HIV pandemic has been associated with a significant increase in cancers such as Kaposi's sarcoma and other tumours.

The cause of the majority of childhood cancers is unknown. Most cancers probably result from the interaction of environmental factors with a genetic predisposition. For example, African Burkitt 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 that induces immunosuppression, and frequent infections that cause B-cell proliferation are all likely aetiological factors.

Problems of treating children with cancer in resource-limited countries

The problems listed below are not exclusive to resource- limited countries, and not mutually exclusive (i.e. many factors interact, compounding the difficulties in treating cancer).

1. **Poverty:** national, regional, local and personal. This is often associated with low government expenditure on healthcare, absence of social care, and lack of insurance for medical illnesses.
2. **Lack of suitable treatment centres and training programmes.** Existing centres lack necessary resources like a steady supply of chemotherapy and supportive care drugs, access to radiotherapy, intensive care and other resources.
3. **Lack of trained staff, especially nurses,** but also lack of surgeons, pathologists, and paediatricians (especially paediatric oncologists).
4. **Healthcare professional resources**

- a) Staff morale problems (see Section 1)

Low morale may be due to low wages, overwork, and dirty crowded conditions, compounded by too many patients and by becoming accustomed to low patient survival rates. Solutions may include better training and remuneration, better working and living conditions, and making staff feel valued.

- b) It is important that all healthcare professionals recognise that **nursing care** is fundamental. Nurse bonus schemes for work effectively performed can be helpful (for example IV antibiotics and chemotherapy correctly administered for all patients).
- c) Nurses often rotate every few months between departments. Try to ensure that, for paediatric oncology, a cohort of nurses remains permanently on the ward, as much of the work is very specialised (for example chemotherapy administration).

- d) **Paediatric oncologists**

An effective service needs good leadership in a major centre. This can develop the service through training and the development of fellowship programmes. There can be support from overseas experts, perhaps with ‘twinning’ of hospitals from a well-resourced country to share decision making on complex cases and to supply visiting experts. Much useful work can be done in oncology with email and web conferencing to facilitate knowledge sharing in both directions. As there is an increase in the number of trained and training staff, round-the-clock expertise in paediatric oncology can be achieved with the development of on-call rotas.

5. Late presentation

- Patients’ families are often very poor.
- They may present to traditional healers first, leading to a delay in diagnosis and referral.
- They often cannot afford transport

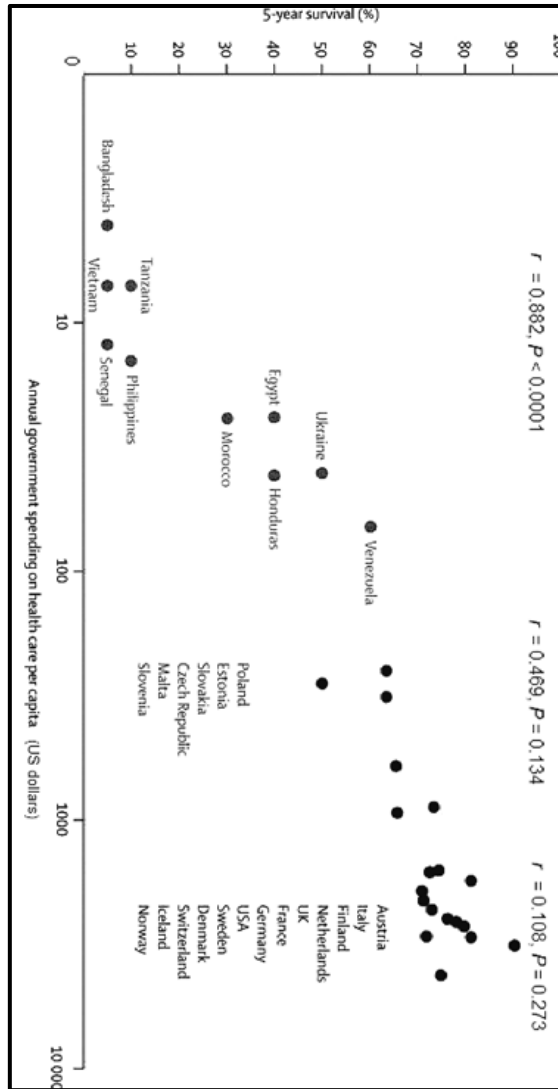


FIGURE 15.1 Correlation between annual government healthcare expenditure (US\$) per capita and childhood cancer survival (From Ribeiro RC, Steliarova-Foucher E, Magrath I et al. (2008) Baseline status of paediatric oncology care in ten low-income or mid-income countries receiving My Child Matters support: a descriptive study. *Lancet Oncology*. 9: 721–9.). Reproduced with permission from Macdonald S, Magill-Cuerden J (ed.) *Mayer's Midwifery: a textbook for midwives*. Elsevier Health Sciences; 2010. © Elsevier

6. High cost of treatment

- Expensive cytotoxic agents, counterfeit medications, quality control problems, cold chain difficulties (for example asparaginase is an enzyme and must be kept cool), restrictions (e.g. on oral morphine See Section 7).
- Cost of diagnostic imaging and pathology.
- Cost of supportive care: antibiotics and other antimicrobials, blood products.

- Cost of caring for critically ill children: high- dependency/intensive care, postoperative care.
 - There is a need for multiple support networks and institutions to develop the paediatric oncology service in the face of the poverty that causes the above problems. These will include individuals and their families, non-governmental organisations (NGOs), corporate business, public social responsibilities, twinning, and public private partnership. Government involvement is vital.
- 7. Inadequate provision of analgesics and other drugs**
- For all patients, and especially where cure is not possible, palliative care is a vital part of oncology treatment. Analgesics play a large part in supportive care and procedural sedation and analgesia, as well as in palliative care. The lack of these drugs is often coupled with a poor understanding and awareness of pain management in healthcare professionals.
- 8. There is often an interrupted supply and insufficient quality control of all drugs.**
- 9. Comorbidities**
- There is often a high prevalence of co-infections, malaria, anaemia, helminths, and malnutrition which can confound the diagnosis and cause decreasing tolerance of cytotoxic therapy.
- 10. Untimely and inappropriate cessation of treatment (i.e., abandonment of treatment)**
- A lack of education and knowledge about uncommon diseases among families, communities and healthcare workers leads to a lack of understanding of the need for treatment.
 - There is then a lack of financial and social support as treatment is lengthy, and the parent has to stay with the child in hospital. This makes it difficult to look after other children at home, and to work, leading to loss of employment.
 - Traditional beliefs include unrealistic preconceptions about cancer and reliance on traditional medicines. In Swahili, ‘the never healing sore’ refers to the fact that there is no expectation of cure, and therefore no point in treatment.
 - Adolescents frequently treated on adult wards.
 - Patients may be left on wards for months because of a lack of diagnostic facilities (for example children with brain tumours or osteosarcomas). It is important to search the wards for such patients and to alert colleagues to refer children to an oncologist where the diagnosis is unknown.
- 11. Impact on family structure**
- The loss of parental income may result in disruption and potential disintegration of the family, and at the very least to a change in family roles, especially where both parents need to work to maintain the family income.

Management of children with cancer in resource-limited countries

The following principles and practices should guide the management of children with cancer in resource-limited settings.

- Engage in twinning – that is, developing a link between a treatment centre in a resource-limited country and one in a resource-rich country, with the objective of sharing professional and technological expertise along with other resources.
- Initially, target curative treatment for cancers that are common and have a relatively good prognosis. When curative treatment is not an option or is not offered, it is essential

to provide palliative care to reduce suffering. **Both curative and palliative care must be seen as active forms of therapy.**

- If curative treatments are to be undertaken, then whenever possible they should be given in a specialist children's cancer centre (see below). There is potential for greatly increasing suffering by only offering 'half treatment' of cancer for children. It must be done fully and professionally, or alternatively the child must be given palliative care (see Section 7).
- Adapt treatment protocols in accordance with local infrastructure and facilities, maintaining a balance between treatment response and cure on the one hand and treatment toxicity and mortality on the other.
- Take steps to ensure compliance with and completion of treatment. Anticipate abandonment of treatment, and address the causes, which vary from country to country.
- Maintain a database of patients using free resources such as Resonance Technology <https://resonancehealth.org/RPC/login> Accessed 28.03.2021
- Engage in the education and training of healthcare professionals, including nurses and doctors, by using free resources such as cure4kids (www.cure4kids.org) <https://www.cure4kids.org/> Accessed 28.03.2021 as well as conducting in-house workshops.
- Aim to be part of regional, national and international collaborative groups to derive benefit from shared expertise, uniformity of treatment and supportive care, and participation in clinical trials.
- Develop parent support groups and provide resources for food, lodging and transport.

In countries where there is an improving infrastructure, the following cancers (which include the six index cancers proposed by the WHO Global Initiative) may have a good or reasonable chance of cure:

- **Acute lymphoblastic leukaemia (ALL) (WHO Index Cancer):** Standard risk ALL in children aged 2–10 years, with a white blood cell count of $< 50 \times 10^9/\text{litre}$, 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. However, **CNS-directed therapy with cranial radiotherapy plus limited intrathecal methotrexate or intrathecal methotrexate throughout therapy is required in all cases.**
- **Hodgkin's disease (WHO Index Cancer):** chlorambucil, vinblastine, procarbazine, prednisolone (ChIVPP) or cyclophosphamide, vincristine, procarbazine and prednisolone (COPP) or doxorubicin, vinblastine, dacarbazine, bleomycin (AVBD) six to eight courses..
- **Burkitt's lymphoma (WHO Index Cancer):** single-agent cyclophosphamide with intrathecal methotrexate and hydrocortisone may result in cure for low stage disease if resources are very limited. Alternatively, cyclophosphamide, vincristine, methotrexate, prednisolone (COMP) chemotherapy with intrathecal methotrexate and hydrocortisone.
- **Non-Burkitt/non-Hodgkin's lymphoma:** early stage – surgery plus COMP or cyclophosphamide, Adriamycin, vincristine, prednisolone (CHOP).
- **Brain tumours:**
 - Low-grade gliomas (**WHO Index Cancer**): surgery alone if resectable.

- Medulloblastoma and ependymoma (resectable/ non-metastatic): surgery followed by **radiotherapy**.
- **Retinoblastoma (WHO Index Cancer)**: enucleation (radiotherapy in some cases).
- **Neuroblastoma** (stage I and II): surgery alone or with 4 to 8 cycles of chemotherapy with vincristine, cyclophosphamide or carboplatin, etoposide.
- **Wilms' tumour (WHO Index Cancer)**:
 - Stage I: surgery plus 10 doses of vincristine (at weekly intervals).
 - Stage II : surgery plus vincristine/actinomycin for 6 months.
- **Resectable embryonal rhabdomyosarcoma** (certain sites): surgery plus vincristine/actinomycin D (four courses).
- **Germ-cell tumours**:
 - **Mature and immature teratoma**: surgery alone.
 - **Malignant germ-cell tumours (stage I)**: surgery alone.

Specialist cancer centres or units

Establishment

Specialist cancer centres or units, and the use of standard treatment protocols (discussed below), have both been fundamental to the ever-improving survival of children with cancer in resource-rich countries.

Cancer is a relatively rare disease, and its treatment is usually complex.

Management requires a dedicated and experienced multidisciplinary team.

Every country should aim to have at least one adequately equipped and funded centre, and then develop shared care or a satellite centre.

Advantages of a specialist children's cancer centre or unit

- 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 retention of staff, leading ultimately to accreditation as a principal care centre in paediatric oncology.
- Improved support, education and counselling of affected children and their families.
- Stimulus for the development of similar units in the same part of the world.
- 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 or unit

- Dedicated paediatric oncologist(s) and nursing staff supported by nutritionists, psychologists and social care workers.
- General surgeon and neurosurgeon trained in paediatric surgery.
- Access to radiotherapy and services of a radiotherapist.
- Blood and platelet banking facilities.
- Pathologist with experience of paediatric tumours with adequate histology and cytology facilities (immunohistochemistry is desirable; this can be in a centralised laboratory if it provides a service for more than one centre or country).

- Haematology, biochemistry, and microbiology laboratories with good quality control.
- Diagnostic imaging: X-ray, ultrasound; CT imaging is desirable, especially for brain tumours. Families may have to pay for these, which is often a limiting factor that determines whether a child is diagnosed and treated properly. Fine-needle aspiration is important, as are lumbar punctures performed under appropriate analgesia and sedation.
- It is vital to have good supportive care, including regular supplies of medication, good stockkeeping, and drug ordering (ensure that drugs are not stolen and sold on the 'black market'), IV fluid management systems to avoid tumour lysis syndrome, and good post-operative care.
- Adequate bed capacity. Most units are constantly overcrowded, with two patients per bed, and as units develop it is essential to build the capacity to cope with increasing numbers of patients.
- Computer facilities with Internet connections (for emailing to the link centre, Medline searches, patient database).
- Active involvement in auditing practice and participating in research.

Above all, there must be a keenness of all staff to work together to learn and make the unit successful.

Centre or unit database

All centres or units should keep a record of treatment, including details of patient demographics, diagnosis, treatment, side effects and survival. This will aid the identification of specific problems, the development of more effective treatment protocols for treatment and supportive care, and overall healthcare planning and development. The availability of free online and electronic resources such as [Resonance Technology](https://resonancehealth.org/RPC/login) (<https://resonancehealth.org/RPC/login>) Accessed 28.03.2021 makes this feasible.

Links with other centres or units and organisations

Provision should be made for communication and transportation for patients from remote areas. Satellite or shared-care centres can be developed by linking with healthcare facilities in other areas so that appropriate care can be continued (e.g. district hospitals).

Links with centres or units in resource-rich countries (twinning)

Links with an established unit in resource-rich countries can have the following advantages:

- sharing information and experience on how to raise awareness of cancer and reduce delays in diagnosis
- helping to speed up diagnosis and make it more precise
- development of locally affordable supportive, palliative, and curative care guidelines
- helping to train and retain staff
- helping to create patient data registration
- help to develop long-term sustainability
- providing support and advice for difficult problems (e.g., by email or web conferencing)
- pathology samples can be couriered for more complex testing (e.g., for immunophenotyping, special staining, VMMA, etc.)
- research collaboration.

In addition, links with international organisations are 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 or centre

Diagnosis

- A complete history and examination.
- Investigations to confirm histology, determine the extent of the tumour (staging) and identify any tumour-related toxicity (e.g. disturbance of renal, liver and/or bone-marrow function).

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 generally adequate for chest imaging, while **CT scan of the chest may be more definitive if it is available.**
- Ultrasonography affords good visualisation of the abdomen and pelvis, although **CT of the abdomen and pelvis may have advantages over ultrasonography in some patients.**
- For the brain, CT scanning is a necessary part of investigation and management. MRI of the brain has advantages over CT, but the availability of this technique is very limited.
- Nuclear imaging can further assist in accurate staging (for example technetium bone scan for bone and soft tissue sarcomas, and Meta-iodo benzyl guanidine (MIBG) scan for neuroblastomas).

Biochemical markers

These are 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).

Pathology

- Good histopathology is essential for the individual and is the only way to compile accurate incidence figures and survival data and to identify favourable histological subgroups.
- Close involvement of the pathologist is needed before biopsy or surgery so that the surgeon can obtain an optimal specimen in the right fixative.

Multidisciplinary team meetings

Following initial clinical assessment and investigation, all children with cancer should ideally be discussed at regular multidisciplinary team meetings that may include the oncologist, radiologist, surgeon and radiotherapist. Such discussions are also recommended at the time of significant events during treatment (e.g. progression or relapse). This ensures that the child benefits from the collective knowledge of the treating team and there is consistency in treatment. However, this can be difficult as staff will be very busy and may require extra funding.

Treatment protocols

- Each unit should use a standard protocol for each tumour type, with the 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.
- Such protocols have been developed for several of the childhood cancers by the SIOOP Paediatric Oncology in Developing Countries (PODC) Adapted Therapy Regimen Working Group.

Chemotherapy

- Late diagnosed childhood malignant tumours are 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 (e.g., 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

- This is important both for obtaining diagnostic material, and as local therapy to reduce tumour bulk. Surgeons should be specially trained and have experience in oncology.
- It is preferable for surgeons to have received specific training in operating on children and in tumour surgery.
- 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 pre-operative chemotherapy, which may reduce tumour size, and thus reduce peri-operative 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.
- 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 the older orthovoltage therapy in giving a more controllable beam and avoiding 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 avoided whenever possible.

Procedures

Bone-marrow aspiration

- This is needed in the diagnosis of leukaemia and lymphoma, and also to identify any bone-marrow infiltration with solid tumours such as neuroblastoma.
- **It is a painful procedure and must be done under analgesia and sedation (for example ketamine 2 mg/kg) (see Section 7 and Section 9 Handbook 1) along with infiltration of the skin and subcutaneous tissues down to periosteal level with a local anaesthetic.**
- Aspiration is preferably performed from the posterior iliac crest but can also be taken from the anterior crest.

Lumbar puncture

- This is needed 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 children should be done under analgesia and sedation along with local anaesthetic wherever possible (especially if multiple lumbar punctures are needed).

Venous access

- Venepuncture for administration of chemotherapy and blood sampling is painful and especially difficult in the young child (analgesia and sedative cover may be needed).
- Repeated venepuncture results in loss of venous access due to venous thrombosis and may significantly compromise therapy.
- Several agents, especially vinca alkaloids, are extremely damaging to tissues when extravasated.
- Short-term percutaneous placement of medium-length or long lines under local anaesthetic may provide an alternative means of venous access.
- The placement of a long-term peripherally inserted central venous catheters (PICC) is a good alternative in resource-limited setting if tunneled central venous catheters (e.g., Broviac line, Hickman line) are not available can be considered in children receiving intravenous chemotherapy. All these should be placed by an experienced surgeon/clinician, and its use is associated with an increased risk of infection, particularly from skin organisms such as staphylococci.

Psychological support

Cancer and its treatment are frightening experiences for many patients, and every attempt should be made to reduce the child's fears. An explanation of the diagnosis and treatment, including the likely outcome, should be given in clear understandable terms to the child's family and to the affected child or young adult wherever appropriate. Such information is best delivered over more than one conversation, allowing the family to understand it and come back to ask questions.

All aspects of treatment and associated side effects should be clearly explained, including details of supportive care, such as infection control, the importance of seeking a healthcare worker if a fever develops, mouth care, pain relief, care of lines, and procedures, such as surgery, bone-marrow aspirate and lumbar puncture. These conversations need to continue throughout treatment, thus establishing a relationship with the child and their family. The family must always be fully involved in the patient's care (e.g. by donating blood when it is

needed). Parents want their child’s doctor to focus on a potential cure and relief of symptoms, and then they can have faith in the doctor and derive hope for the future.

Side effects of the disease and/or its treatment

General side effects of chemotherapy

Acute effects

- Bone-marrow suppression
- Infection
- Bleeding
- Anaemia
- Nausea and vomiting
- Mucositis
- Alopecia
- Fatigue and cachexia
- Tumour lysis syndrome

Late effects

- Infertility
- Secondary malignancy

TABLE 15.1 Specific side effects of chemotherapy

Neurotoxicity	Vincristine (muscle weakness due to peripheral neuropathy, constipation, rarely encephalopathy)
Cardiomyopathy	Doxorubicin/daunorubicin
Respiratory system	Bleomycin
Urinary tract	Cisplatin (renal), ifosfamide (renal and bladder), cyclophosphamide (bladder)
Liver	Thioguanine, actinomycin D
Hearing	Cisplatin

Acute Side Effects

Infection

- Neutropenia, both at diagnosis in leukaemia and following most chemotherapy, produces a risk of significant bacterial and fungal sepsis derived from the patient’s own flora when the neutrophil count is $< 1.0 \times 10^9/\text{litre}$, and particularly when it is $< 0.2 \times 10^9/\text{litre}$.
- The greatest risk is from 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 $< 1.0 \times 10^9/\text{litre}$ who develop fever (e.g., $> 38^\circ\text{C}$ for 2 hours or $> 38.5^\circ\text{C}$ on one occasion).
- The antibiotic regimen should be determined by each centre depending on the prevailing flora, local resistance patterns 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, therapy with vancomycin or teicoplanin is recommended.
- For microbiologically proven septicaemia, antibiotics should be given for 5–7 days, the choice of drug depending on the antibiotic sensitivity of the isolated organism.
- If systemic fungal infection is proven or suspected (e.g., if fever fails to remit after 4–5 days of antibiotics), then intravenous amphotericin, despite its renal toxicity, is still the drug of choice and is widely available. Newer lipid-based formulations of amphotericin are less toxic but very expensive.
- *Pneumocystis carinii* pneumonia, especially in patients with leukaemia, requires prophylaxis with co-trimoxazole (calculated as a dose of 150 mg/m²/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 lifesaving but is rarely available.
- High dose aciclovir is the treatment of choice for zoster infections but is expensive and not yet widely available globally.

Bleeding and anaemia

- Adequate blood banking facilities with availability of blood component therapy such as packed red blood cells and platelets (see Section 54) are a fundamental part of therapy. Red blood cell transfusion should be reserved for symptomatic anaemia, or when the haemoglobin falls to a very low level (for example < 6 grams/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 of > 40×10⁹/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.

Nausea and vomiting

- Nausea and vomiting are 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 potential to produce vomiting, from very low (e.g., vincristine and etoposide) to very high (e.g. cisplatin). Anti-emetic therapy should be given wherever possible, preferably prophylactically, but certainly to patients with established retching and vomiting.

Anti-emetic agents

1. **5HT₃ antagonists (for example ondansetron):** these are the most effective anti-emetics, especially when combined with steroids. They have become increasingly available and affordable. Ondansetron dosage for ages six months–18 years either 5 mg/m² or 150 micrograms/ kg (max single dose 8 mg) IV before chemotherapy then repeated every 4 hours for two further doses, then give orally. Oral dose < 10 kg = 2 mg 12 hourly, > 10 kg = 4 mg 12 hourly for up to 5 days.

2. Steroids: the main effect is in combination with other agents (dexamethasone 6 mg/m² single dose).

If ondansetron or other 5HT₃ antagonists are not available, other drugs can be used such as:

- **Metoclopramide:** this is effective in high dose, but a greater risk of extrapyramidal side effects exists in children. Give 100 micrograms/kg for 1/12 to 1 year, 1–3 years 1 mg, 3–5 years 2 mg, twice daily and 5–9 years 2.5 mg, 9–18 years 5 mg thrice daily orally or slowly (over 2 minutes) by IV injection. Over 60 kg children can have 10 mg three times daily. **Avoid the IM route.**
- **Chlorpromazine:** orally or IV (the IV route can cause severe hypotension), child 1–12 years 500 micrograms/kg every 4–6 hours, maximum 75 mg daily. For 12–18 years 25–50 mg every 3–4 hours until vomiting stops.
- **Prochlorperazine:** orally or IV slowly over 10 minutes, 250 micrograms/kg 1–12 years every 8–12 hours (only if the child weighs over 10 kg or is over 1 year of age). 12–18 years 5–10 mg three times daily.
- **Benzodiazepines:** the main effect is sedation and amnesia. These drugs are useful for anticipatory nausea.

Oral mucositis

- This is a common side effect of many cytotoxic agents and radiotherapy.
- Scrupulous simple oral hygiene should be maintained. This can be achieved by regular thorough tooth brushing two to three times a day together with use of a mouthwash such as chlorhexidine if available.
- Oral fluconazole and oral acyclovir may be of benefit in oral mucositis with secondary infection from candida and herpes infection, respectively.

Alopecia

This is inevitable with most chemotherapy, but usually entirely reversible on completion of treatment. Some children are not upset by the appearance of alopecia, but for those who are distressed by it, a light but attractive head covering may be acceptable.

Nutrition

Maintenance of adequate nutrition is essential. 'Cancer wasting' or cachexia is a well-recognised complication of paediatric tumours and is subsequently associated with a decreased tolerance of chemotherapy and its side effects, and possibly an increase in cancer mortality.

Poor nutritional status may result from any of the following:

- stress
- pain
- increased metabolism (due to tumour or infection)
- anorexia
- altered sense of taste and smell
- chemotherapy-induced nausea and mucositis (e.g. stomatitis, oesophagitis)
- radiotherapy-induced mucositis and dry mouth (xerostomia)
- surgery-induced pain, bowel obstruction and appetite suppression.

In addition to this, an unacceptably high number of children in resource-limited countries who do not have cancer are malnourished. The effect of cancer and its treatment can be even more deleterious for such children.

Each child should have a nutritional assessment, including measurement of height or length, weight, mid upper arm circumference and triceps fold thickness (using callipers). Height and weight should be plotted on a standard percentile chart. (See Section 65).

Nutritional support should be given to children who consistently show a decrease across percentile lines. It may also be indicated in children with baseline malnourished status. A high-calorie diet with adequate protein should be given to all children with cancer, supplemented, if necessary, with specific additives to provide additional calories and protein.

If sufficient food cannot be taken orally, enteral feeding via a nasogastric tube (particularly overnight) should be considered. Total parenteral nutrition should be avoided, as it is expensive and associated with a high risk of complications, including infection and metabolic disturbance.

Tumour lysis syndrome (TLS)

This is a life-threatening complication that occurs when the rapid lysis of tumour cells, usually resulting from chemotherapy, leads to the release of excessive quantities of cellular contents into the systemic circulation, resulting in a metabolic disturbance characterised by the following:

- hyperkalaemia
- hyper-phosphataemia
- hyperuricaemia
- hypocalcaemia.

This metabolic derangement may lead to acute oliguric renal failure and cardiac arrhythmias.

TLS can occur spontaneously in tumours with a very high proliferative rate, as well as following initiation of treatment. It can be classified as laboratory TLS (with no clinical manifestations) or clinical TLS (with life-threatening clinical abnormalities).

Management of TLS

- Most importantly, anticipate and recognise patients who are at high risk of tumour lysis, i.e., those with leukaemia and lymphoma (particularly T-cell or Burkitt's phenotype, and those with a high white cell count $> 50 \times 10^9/\text{litre}$, hepatosplenomegaly or mediastinal mass, or high LDH).
- Intravenous hydration with potassium-free fluids, at least 2.5–3.0 litres/m²/day, should be commenced prior to treatment and then continued for the first few days of treatment. Ensure that there is adequate urine output ($\geq 1 \text{ mL/kg/hour}$).
- Regular allopurinol, 100 mg/m² dose every 8 hours, should be commenced prior to treatment and then continued for the first few days of treatment.
- Clinical and laboratory monitoring should be undertaken, including daily weight, input and output review, and assessment of blood biochemistry, with measurement of uric acid levels up to four times a day if needed.

Late Side Effects

Infertility

- This mainly occurs in males and is a consequence of specific 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 about infertility, and hormonal treatment may be offered.
- Sperm storage for adolescent boys before the start of treatment can be considered if this service is available.

Second tumours

- Chemotherapy results in a small but important risk of second tumours. These could be hematological especially acute myeloid leukaemia or solid tumours like sarcomas. This is particularly associated with alkylating agents such as cyclophosphamide (especially if used with radiotherapy), anthracyclines and topoisomerase-2 inhibitors (e.g., etoposide).

Treatment of individual tumour types

A detailed discussion of the presentation and management of every type of tumour is beyond the scope of this book. A brief overview for some of the major childhood cancers is given below.

Acute lymphoblastic leukaemia (ALL)

Approximately one-third of all children under 15 years of age with cancer have acute leukaemia, and 75–80% of these have acute lymphoblastic leukaemia, making it the most common childhood cancer in well-resourced countries.

Presentation

- Myelosuppression.
- Anaemia, infection (which can be life-threatening) and thrombocytopenia (bruising, bleeding, petechiae).
- Lymphadenopathy and hepatosplenomegaly.
- Bone pain and limp.

Diagnosis

- Full blood count.
- Blood film can be diagnostic for patients with very high white cell counts.
- Bone-marrow aspirates (these are always required).
- Morphology (e.g., FAB system), cytochemistry and immunocytochemistry (if available).
- Lumbar puncture: CSF cell count and cytospin for lymphoblasts.
- Chest X-ray (T-cell leukaemia).
- Ultrasound scan of the abdomen for assessment of the liver, spleen and kidneys.

Treatment

Rapid tumour lysis, which can sometimes be spontaneous, is a major risk, particularly for patients with high white cell counts leading to biochemical disturbances. Intravenous fluids, allopurinol and close monitoring of renal function are required at the start of treatment. The treatment of acute lymphoblastic leukaemia is divided into a number of phases, as described below.

Induction

The aim is to get the patient into remission (defined as the presence of < 5% blasts in bone marrow). 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 is too expensive, anthracyclines (e.g. doxorubicin) can be substituted.

CNS-directed therapy

This is needed in all patients to prevent CNS relapse. Standard therapy is to give several doses of intrathecal methotrexate throughout the treatment period and this, along with other aspects of treatment have slowly replaced the cranial irradiation (often not available in low income countries) particularly in the standard-risk (and increasingly in high-risk) patients. This has the added benefit of decreasing the late neuro-cognitive side effects seen as a result of cranial radiation.

Intensification therapy

The administration of periods of more intensive therapy (e.g. with drugs such as cyclophosphamide, daunorubicin and cytosine) has been associated with increased survival, although this treatment carries the risk of severe myelosuppression and should be used with caution unless a high level of supportive care is in place.

Continuation (maintenance) therapy

This essential part of treatment generally lasts for 2 to 3 years. Most regimens employ daily oral mercaptopurine and weekly oral methotrexate with or without 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. those with a white cell count at diagnosis of $< 50 \times 10^9$ /litre, and aged 2–10 years) to survive.

Acute myeloid leukaemia (AML)

This accounts for 15–20% of acute leukaemias in children.

Presentation

The presentation is the same as for acute lymphoblastic leukaemia, with more likelihood of tissue infiltration:

- gum hypertrophy: monocytic leukaemia
- skin involvement: myeloblastic leukaemia
- disseminated intravascular coagulation: promyelocytic leukaemia.

Diagnosis

See above section on acute lymphoblastic leukaemia.

Treatment

This is much less successful than for acute lymphoblastic leukaemia. Induction therapy is based on 8–10 days of intensive chemotherapy with drugs such as daunorubicin, etoposide,

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 acute lymphoblastic leukaemia. Lumbar puncture with triple intrathecal chemotherapy (methotrexate, hydrocortisone and cytosine) should be given with each course of chemotherapy.

Prognosis

Less than 50% of these children will be expected to survive long term, with a high risk of toxic death 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 therefore the presenting features are protean. Patients often 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. **Progression in size of Burkitt's lymphoma can be rapid, given its 48-hour doubling time.** Head tumours usually present with extensive involvement, with swelling of the jaw and tooth loosening, gum expansion, bleeding, ulceration and exophthalmos.
- The majority of non-Burkitt 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.

Diagnosis

The diagnosis is frequently suggested on clinical examination (e.g. classical features of Burkitt's or T-cell lymphoma). The diagnosis is supported by appropriate imaging (X-ray, ultrasound). Bone-marrow aspiration and lumbar puncture should be performed. Biopsy is necessary if the diagnosis cannot be made on a bone-marrow aspiration.

Treatment

Burkitt's lymphoma

This is an extremely chemosensitive tumour, and a high remission rate can be achieved with a single course of cyclophosphamide. Repeated courses of cyclophosphamide may be successful in some early-stage patients, but the success of therapy is further improved, particularly for patients with advanced disease, using multi-agent chemotherapy using combinations such as COMP (cyclophosphamide, vincristine, methotrexate, and prednisolone), for example, given over a 6-month period. This should be accompanied by administration of intrathecal methotrexate and hydrocortisone. As with acute lymphoblastic leukaemia, biochemical disturbance because of rapid tumour lysis is a major risk, and intravenous fluids, allopurinol and close monitoring of renal function are required.

Non-Burkitt B-cell NHL

Repeated courses of multi-agent chemotherapy with COMP or CHOP (cyclophosphamide, Adriamycin, vincristine, and prednisolone) are 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 the toxicity of these regimens is potentially high.

T-cell NHL

In contrast 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 or moderate-dose methotrexate with ongoing intrathecal methotrexate should be used.

Prognosis

Burkitt's lymphoma

The prognosis varies according to the stage of disease, although overall at least 85–90% of patients will be cured with modern therapy in well-resourced countries. Where ability to give chemotherapy is restricted, simpler therapy can yield 50–60% survival rates. However, CNS disease is associated with a poor outcome.

Non-Burkitt B-cell NHL

The prognosis is poorer than with Burkitt's lymphoma and depends on the stage of disease and the intensity of treatment. In low-stage disease a survival of at least 75% is expected. The prognosis is worse with extensive disease, particularly with bone-marrow or CNS involvement.

T-cell NHL

With modern leukaemia-type therapy, survival rates are around 65–70% or higher.

Hodgkin's lymphoma

Presentation

Unlike NHL, Hodgkin's lymphoma tends to be confined to the lymph nodes or spleen, although spread to other sites, such as the 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. Essential 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 utilised, often using extensive radiation fields (e.g., the 'mantle' or 'inverted Y' techniques) to cover all known sites of disease. Radiation is still used in localised disease, but generally **chemotherapy is preferred** for most patients, using regimens such as ChIVPP (Chlorambucil, vinblastine, procarbazine and prednisolone) or COPP (cyclophosphamide, vincristine, procarbazine and prednisolone). Six to eight courses are given every month. Such chemotherapy may be given on an outpatient basis, and is

relatively non-toxic, although the risk of infertility in boys is high. Some of the toxicity can be avoided by alternating COPP or with ABVD (doxorubicin, bleomycin, vinblastine and dacarbazine), although this may have the potential to cause cardiotoxicity.

Prognosis

Hodgkin's lymphoma generally carries a good prognosis. For patients with stages I and II tumours, over 80% are expected to be cured. Even with advanced disease, over 50% of patients would be expected to survive.

Brain tumours

These are a heterogeneous collection of several tumours that together represent around 25% of all childhood cancer patients in Europe and North America. The proportion in resource-limited countries is much lower, at least partly due to under-diagnosis because of limited availability of neuroimaging (CT and MRI), neurosurgery and neuropathology.

Presentation

About 60% of childhood brain tumours arise in the posterior fossa, and usually present 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. Imaging of the spine and examination of the spinal fluid is required to assess for CNS spread in high-grade bone tumours (for example medulloblastomas, high-grade gliomas).

Treatment

For most tumours, modern neurosurgery (see Section 73. Handbook 1) is vital to management. Prompt relief of raised intracranial pressure is often required and may be lifesaving. This is achieved with dexamethasone, which when used peri-operatively has also been shown to significantly reduce operative mortality.

Surgery may also be required to relieve hydrocephalus (e.g., with ventricular peritoneal shunting). The aim of definitive surgery is to provide a histological diagnosis and usually to shrink the tumour as much as possible. Tumour resection is required for most tumours, including all posterior fossa tumours (except the brainstem), tumours of the cerebral hemispheres and craniopharyngiomas. Some tumour types may be cured with surgery alone (e.g., cerebellar low-grade astrocytoma), although others (e.g., medulloblastoma) require adjuvant radiotherapy.

Generally, a large dose of radiotherapy is given to the tumour bed, while some tumours (e.g., medulloblastoma) require 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, although it can be used to try to delay radiotherapy in the very young. Radiotherapy to the

whole brain and spine has a very high risk of sequelae, particularly in young children. These include neuropsychological disability, growth failure (growth hormone deficiency and poor spinal growth) and hypothyroidism.

The following is a brief guide to the management and prognosis of individual tumour types.

Medulloblastoma

Prognosis

The prognosis is around 60% for children with non-metastatic disease and 30% for those with disseminated 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 to delay radiotherapy, but even then survival is only around 20%.

Cerebellar low-grade astrocytoma

Treatment

Surgical resection is performed, and post-operative radiotherapy is not required if the resection has been complete.

Prognosis

The prognosis is at least 80% following total resection.

High-grade glioma

Treatment

Surgical resection (as complete as possible) is performed, and post-operative focal radiotherapy is required.

Prognosis

Overall, the prognosis is very poor, at around 15%. Patients who undergo complete resection and have Grade 3 (anaplastic astrocytoma) tumours have a much better chance of survival than those who undergo subtotal resection and have Grade 4 tumours (glioblastoma multiforme).

Ependymoma

Treatment

Surgical resection (as complete as possible) is performed, and post-operative focal radiotherapy is required.

Prognosis

The prognosis is around 30–50%, mainly depending on the degree of tumour resection.

Brainstem glioma

Treatment

Focal exophytic tumours are treated with surgery followed by focal radiotherapy.

Prognosis

The prognosis is around 30–50%, mainly depending on the degree of tumour resection.

Diffuse (malignant) brainstem gliomas

Treatment

Palliative radiotherapy may possibly be used.

Prognosis

These tumours are fatal (less than 5% survival).

Craniopharyngioma

Treatment

Surgical resection is performed, although there is a high peri-operative mortality rate. Radiotherapy is sometimes used for recurrent tumours.

Prognosis

The prognosis is variable. All patients suffer from pan-hypopituitarism, which requires hormone replacement therapy.

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 50% of patients who present with bad biology and 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. Para-spinal masses that extend into the spinal canal causing cord compression, and thoracic primaries that cause airway obstruction, also occur. A significant number of patients (50%) present with metastatic disease that often causes bone pain and limp, with marrow infiltration mimicking leukaemia, skin infiltration or orbital masses causing proptosis or peri-orbital bruising.

Diagnosis

Ultrasound of the abdomen (or CT of the abdomen, if available), chest X-ray (or CT of the chest for thoracic tumours, if available), abdominal X-ray (calcification is often a feature of primary tumours), **24-hour urine collection or spot urine sample for catecholamine metabolites** (secreted in 85% of cases), bone-marrow aspirate and trephine (bilateral) are all helpful. MIBG scan and technetium bone scan are performed to investigate metastasis to the bones. 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 higher survival rate. For stage III patients, the prognosis is around 40%, with treatment including multi-agent chemotherapy with drugs such as cyclophosphamide, vincristine, Adriamycin, etoposide, and platinum, followed by surgical excision of the tumour. Stage IV disease has a very poor prognosis, and it is essential to provide palliative care to reduce the suffering of these patients.

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, namely an autosomal-dominant heritable form that may affect one or both eyes, and a sporadic (non-heritable) form that 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, sometimes facilitating preservation of vision and possibly preventing metastatic spread.

Wilms' tumour (nephroblastoma)

This tumour occurs in nearly all parts of the world and is one of the most curable of all childhood cancers. Approximately three out of four cases occur in children under 5 years of age.

Presentation

Most patients present with a large and 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, such as neuroblastoma and splenomegaly associated with malaria or haemoglobinopathy.

Diagnosis

The presence of a renal tumour can be confirmed by ultrasonography, which should also assess the presence of inferior vena cava involvement. Alternatively, intravenous urogram (with injection into the feet to perform a cavagram) can be used, as can CT scan (with contrast) (if available). The diagnosis can be made on the basis of clinical presentation and imaging findings. Histopathological confirmation is not mandatory, but is advisable, particularly in those under 6 months of age. A chest X-ray should look for evidence of lung metastases.

Treatment

The SIOP approach of up-front chemotherapy (4 weeks of vincristine and actinomycin D for non-metastatic tumours, and 6 weeks of the two drugs plus Adriamycin for meta- static tumours) followed by surgery is more suited to resource-limited countries. The duration and

type of further chemotherapy after surgery depend on the local staging of the tumour and the response to initial treatment. For stage I tumours, further vincristine and actinomycin may be given for 4 weeks or 6 months depending on the histological response. For stage II disease, vincristine and actinomycin should be given for 6 months, a regimen which may also be used for stage III tumours with the possible addition of radiotherapy. 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), particularly if the lung metastases persist after pre-nephrectomy chemotherapy.

Prognosis

For patients with stages I and II tumours (favourable histology), at least 80% should be cured. Stage III and IV tumours have survival rates of around 60–70% and 50–60%, respectively. However, the prognosis is poor for patients with unfavourable histology.

Liver tumours

The two main types of liver tumour are hepatoblastoma and hepatocellular carcinoma (HCC). Although both are rare in Europe and North America, in several parts of the world, such as 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 in the absence of coexisting cirrhosis.

Presentation

Hepatoblastoma generally presents in children under 3 years of age, whereas HCC is seen in older children and adolescents. The presentation in both hepatoblastoma and HCC is similar, with most patients presenting with abdominal distension 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 examination of the abdomen and CT scan (if available). The diagnosis may be confirmed by biopsy although radiological appearance with raised alpha-protein may suffice. Alpha-fetoprotein levels are elevated in nearly all cases of hepatoblastoma and in about 65% of cases of HCC. In these patients, the alpha-fetoprotein level may be used as a tumour marker to monitor progress. A chest X-ray should be taken to look for evidence of lung metastases.

Treatment and prognosis

Surgical excision is the definitive treatment for both tumours. Hepatoblastoma is a chemosensitive tumour, and pre-operative chemotherapy significantly improves the prognosis, facilitating surgical excision and the control of distant metastases. The most active agents are doxorubicin and cisplatin. Cisplatin monotherapy along with surgery is recommended for localised and non-metastatic tumours. The prognosis for patients with these 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 multi-centric

or extensively invasive, making resection possible in less than 30% of patients. Of these cases, only one-third survive long term.

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), the genito-urinary tract in both boys and girls, and the extremities. Two main histological types are recognised, namely 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) (this is particularly important for head and neck and orbital tumours), although other techniques such as tomography and ultrasound examination may be useful. For head and neck lesions, lumbar puncture with careful CSF examination is required. Parameningeal tumours are those in which CSF invasion is demonstrated or possible due to the proximity of the tumour to the meninges based on CT scanning (if available). Metastatic surveillance includes chest X-ray, abdominal ultrasound examination or CT scanning (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 and 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 (e.g. after three to six courses). Surgery is the usual local therapy for sites such as the extremities and genito-urinary system. 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 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, so careful consideration is needed 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 HIV pandemic. 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 50% of all sarcomas occur in the bone, the predominant 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. Around 95% of osteosarcomas arise in long bones, and about 50% occur in the upper tibia or lower femur. Around 50% 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 osteoblastic and/ or 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 the chest (if available) is used to detect lung metastases, the lung being the most common metastatic site for both tumours.

Treatment and prognosis for Ewing's sarcoma

Chemotherapy using vincristine, adriamycin, cyclophosphamide, ifosfamide and etoposide 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 (e.g. 45–56 Gy) should be given, although for long bone sites amputation may be more appropriate.

The overall prognosis is around 40% but depends on the site and the adequacy of local tumour control. The prognosis for patients with metastatic disease is very poor.

Treatment and prognosis for 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. Six courses of cisplatin and doxorubicin (three pre- and three post-surgery) may be feasible in many resource-limited countries. The current American and European protocols add high-dose methotrexate to the cisplatin and doxorubicin. Ifosfamide could be used as an alternative to high dose methotrexate.

Local control is either with amputation or (if available) with tumour resection and endoprosthetic bone replacement or rotation plasty.

Germ-cell tumours (GCTs)

Around 3% of tumours in children are GCTs, which are seen mainly in infants and adolescents. They include benign (mature and immature teratoma) and malignant (e.g. yolk sac tumour, germinoma) subtypes.

Presentation

In infancy, the usual presentation is a pelvic or sacrococcygeal mass often noticed after birth (or sometimes prior to birth on antenatal scans). In adolescents, GCTs present either as an enlarged mass in the gonads (testicular enlargement or a pelvic mass arising from the ovary) or in the mediastinum with signs of airway or superior vena cava obstruction.

Diagnosis

Initial assessment is by X-ray and CT (if available) for mediastinal masses, and ultrasound examination for abdominal and pelvic masses. Assessment of serum alpha-fetoprotein and β human chorionic gonadotrophin levels can assist in diagnosis and monitoring of the disease.

Treatment and prognosis

For mature and immature teratoma as well as malignant GCTs Stage I, surgery alone can be sufficient, with a survival of more than 90%. For more advanced malignant GCTs, four to six cycles of platinum-compound-based chemotherapy in addition to surgery can achieve a survival of around 70%.

Palliative chemotherapy and radiotherapy

As stated above, if curative treatment is not possible or has failed, the focus must then be on providing palliative care, particularly symptom control, including adequate pain relief (see Section 7 in Handbook 2 and 9 in Handbook 1). Occasionally, palliative chemotherapy may be appropriate, such as the use of steroids with or without vincristine in relapse or incurable acute lymphoblastic leukaemia and lymphomas. Steroids are also used in the control of symptoms such as headache due to certain brain tumours. Palliative radiotherapy may be useful for treating bone pain caused by tumour infiltration (e.g., in neuroblastoma) and by bone tumours themselves, and may be helpful in controlling symptoms caused by compression of nerves (including the spinal cord) or other vital organs.

Conclusion

Although in many resource-limited countries the curative treatment of children with cancer may not be achievable currently, children will present with often distressing symptoms, which we must strive to alleviate and palliate. As infections become more controllable in resource-limited settings, cancer starts to emerge as a major cause of morbidity and mortality. Some allocation of resources becomes inevitable, and as paediatric oncology requires a multidisciplinary approach, thinking about and acting on the problems faced by children with cancer can lead to improvement of care for all children in hospital.

Organisations working to advance paediatric oncology around the world

World Child Cancer (www.worldchildcancer.org) <https://www.worldchildcancer.org>

Accessed 28.03.2021: currently working in Mexico, Colombia, Cameroon, Ghana, Malawi, Mozambique, Bangladesh, the Philippines and the Pacific Islands.

Childhood Cancer International (CCI); <https://www.childhoodcancerinternational.org/>
Accessed 28.03.2021 an international network of parent support groups and survivor networks that provide psychosocial care for children and their families.

Maternal Childhealth Advocacy International – Cameroon <https://www.mcai.org.uk/> Accessed 28.03.2021.

St Jude Children’s Research Hospital based in the USA <https://www.stjude.org/> Accessed 28.03.2021 a paediatric treatment and research facility. It develops advanced cures for and means of prevention of paediatric cancer through research and treatment. It is involved worldwide in supporting projects through its International Outreach programme, including twinning. Through Cure4kids <https://www.cure4kids.org/> Accessed 28.03.21, it provides a free online education and collaboration resource dedicated to supporting the care of children with cancer and other catastrophic diseases worldwide.

International Society of Pediatric Oncology (SIOP) (Société Internationale d’Oncologie Pédiatrique) (<https://siop-online.org/> Accessed 28.03.2021): this organisation has a special focus on paediatric oncology in developing countries (PODC). Some of the relevant working groups include the following:

- twinning, collaboration and support
- adapted therapy regimen
- education and training
- abandonment of treatment
- palliative care
- essential drugs.

International Network for Cancer Treatment and Research (INCTR) <https://www.inctr.org/>
Accessed 28.03.2021 this organisation is dedicated to helping to build capacity for cancer research and treatment in developing countries, and it focuses on palliative care, cancer registration, research, training, nursing and pathology services.

Union for International Cancer Control (UICC) World Cancer Congress <https://www.uicc.org/>
Accessed 28.03.2021): this organisation focuses on raising awareness, education, and developing a global network of influence.

Franco-African Pediatric Oncology Group (GFAOP) (www.gfaop.org): this runs projects for children with cancer in Africa, including a recent Wilms’ tumour protocol trial.

The most recent addition to this global collaboration has been the entry of the WHO with announcement of the Global Initiative for Childhood Cancer in September 2018 <https://www.who.int/cancer/childhood-cancer/en/> Accessed 28.03.2021 In collaboration with others, the aim is to increase awareness and capacity at country level and thus reach at least a 60% survival rate for children with cancer by 2030, thereby saving an additional one million lives. The emphasis is on six index cancers (acute lymphoblastic leukemia, Hodgkin lymphoma, Burkitt lymphoma, low grade glioma, Wilms tumour and retinoblastoma), which together constitute 50-60% of the childhood cancer burden and are highly curable if

diagnosed in time and treated appropriately. Initially one country from each of the WHO regions was selected as a focus country for implementation of the WHO plan and this list of countries is now steadily expanding with the plan of >50 focus countries by 2025.

Further reading

Pizzo PA and Poplack DG (eds) (2020) *Principles and Practice of Pediatric Oncology*, 8th edn. Philadelphia, PA: Lippincott Williams & Wilkins.

Steven MCG, Caron HN and Biondi A (eds) (2012) *Cancer in Children: clinical management*, 6th edn. Oxford: Oxford University Press.

Section 16. Chronic renal failure

Background

Chronic renal failure (CRF) is more frequent in boys than in girls. Its commonest cause is congenital renal abnormalities such as dysplasia associated with severe antenatal vesico-ureteric reflux, and often also 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 of children with milder forms of CRF by simple treatments, especially in the case of 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 of a kind that has only limited availability worldwide at present. Each country should have a specialised centre that 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 with body growth, and renal function becomes outstripped by demand. Deterioration is likely to be quicker if the child has hypertension, or has recurrent urinary infections with continuing reflux, both of which require active treatment.

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 IV 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 well-being and growth. For each of these, 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. For bicarbonate, 1 mmol is equivalent to 84 mg, so 1 gram contains about 12 mmol bicarbonate. For intra-venous use, 8.4% bicarbonate solution contains 1 mmol/mL.
2. For sodium chloride (salt), 1 mmol is equivalent to 57 mg, so 1 gram contains about 18 mmol sodium. For intravenous use, each litre of 0.9% saline contains 150 mmol; a 30% saline solution contains 5 mmol sodium/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, indications to begin this treatment are often multiple, and include an intolerable diet or fluid restriction, and symptoms such as poor growth and lethargy as important factors, rather than just specific biochemical parameters.

Calcium and phosphate

CRF can lead to abnormalities of the plasma calcium and phosphate concentrations, and these can cause rickets and hyperparathyroidism (renal osteodystrophy), which 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 concentration, which in turn leads to a low plasma calcium level by mass action, and by suppressing the enzyme 1-alpha-hydroxylase, thus lowering the concentration of circulating activated 1-alpha-hydroxyvitamin D. A primary lack of 1-alpha-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 (reducing the intake of meat and dairy products), 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 lower end of the normal range. This commonly also results in a rise in plasma calcium levels into the normal range. Because of this, it is seldom necessary to treat mild CRF with 1-alpha-hydroxyvitamin D₃. If it is needed, in more severe CRF, start with about 20 nanograms/kg once daily, and titrate the dose up until the plasma calcium concentration 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 eventually leads to iron overload. Recombinant erythropoietin (if available) should be used, after adequate iron levels have been achieved.

Folate and vitamin B₁₂ supplementation is seldom required, but levels should be checked if possible).

Growth

Many factors lead to growth failure in children with CRF. In older children, attention to fluid and electrolyte intake, prevention of acidosis with bicarbonate supplements, and control of the bone biochemistry help considerably. Control of uraemia by encouraging a diet containing about 1 gram of 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.

Many also vomit excessively. Even when supplemented with tube feeds, very young children with CRF often remain small.

Transplantation

Renal transplantation (if it is available) from a living or deceased donor gives the best quality of life for children with end-stage renal failure.

Section 17 Constipation

Introduction

Definition

Constipation is defined as difficulty with, delay in or pain on defecation.

Normal defecation patterns

- Breastfed babies average three stools per day and formula-fed babies two stools per day. However, the range of normal stool frequency in breastfed babies is very wide, from one stool every few days to a stool with every feed.
- Children average one stool per day after 3 years, but the normal range is from once on alternate days to three times daily.

Pathophysiology

Most children with constipation have no underlying medical cause. An episode of constipation can be triggered by inadequate food or fluid intake, an intercurrent illness, or excessive intake of cow's milk.

Constipation cycle

The child passes a hard painful stool. On subsequent occasions they try to withhold the stool in order to avoid experiencing pain (faecal holding). The stool remains in the rectum, becoming harder still, and so causing even more pain when it is eventually passed.

If this cycle is allowed to continue, eventually the rectum may become enlarged, resulting in a 'megarectum'. The child by this stage has lost the normal urge to defecate, and the large rectal mass of stool holds open the anal sphincter, which leads to soiling with liquid faeces. This is involuntary and should not be confused with encopresis, which occurs when the child voluntarily passes normal stools in unacceptable places.

Diagnosis

Diagnosis can usually be made by taking a good history.

- On examination of the abdomen, faecal masses may be palpable. These are often in the left and right iliac fossae, but sometimes suprapubically. On inspection of the anus, anal tags and fissure may be seen in chronic constipation.
- On rectal examination, hard impacted faeces may be felt. Rectal examination is usually not necessary. If there is an anal fissure, rectal examination should be done with topical lignocaine jelly (1%) and terminated if it is too painful.
- Abdominal X-ray is not a useful examination for diagnosis of constipation.

Pathological causes

The vast majority of constipation is idiopathic, but there are a few uncommon causes that are important not to miss.

Hirschsprung's disease

Suspect this when there is infancy-onset constipation and a delay of more than 48 hours in passing meconium at birth. In more advanced cases there will be abdominal distension and sometimes failure to thrive and vomiting. There may be alternating constipation and diarrhoea and surprisingly little soiling for the degree of constipation. On rectal examination an explosive gush of faeces occurs when the examining finger is withdrawn.

Anal lesions that cause pain or create an obstruction

These include anal fissures, perianal skin infections and (rarely) congenital anterior anus and anal stenosis. One cause of painful anal lesions is sexual abuse, a rare but important cause which should not be missed.

Endocrine conditions

Hypothyroidism, renal tubular acidosis, diabetes insipidus and hypercalcaemia can be associated with constipation. There should be a high level of suspicion for a metabolic or endocrine cause if constipation and failure to thrive coexist.

Neurogenic constipation

Spinal cord lesions involving sensation in the rectum will cause neurogenic constipation. These can be excluded by a normal neurological and spinal examination.

Management of idiopathic constipation

Parental understanding of the aetiology and sequence of events in developing chronic constipation is crucial to successful physical and psychological management (see Figure 16.1). Each and every element of this flow diagram should be addressed and treated if management is to be completely successful.

Explanation

A careful and thorough explanation of the problem should be given to the parent and child. Emphasise that soiling is not deliberate, and that the child needs support, not condemnation. Assess the need for psychological as well as physical treatment.

Evacuation of hard impacted faeces

1. To soften and lubricate the retained faeces, initially give a softener. This could be a macrogol such as Movicol or another softening laxative such as docusate sodium. The dose will vary according to age.
2. Alongside the softener, in order to expel the retained mass, give a stimulant laxative (e.g. sodium picosulphate).
3. If sodium picosulphate is not available, a large dose of senna can be tried, but may need to be used for longer.
4. Only if the above fails give suppositories (glycerine) once daily (infant, 1 gram; child < 12 years, 2 grams; child > 12 years, 4 grams).
5. If the oral and suppository methods are unsuccessful, if excessive abdominal pain develops and/or there is vomiting, stimulant enemas will be required. Phosphate enemas should not be used in children under 2 years of age. For children aged 2–10 years give 60 mL (half a phosphate enema) and for those over 10 years of age give 120 mL (full enema). If phosphate enemas are 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 defecation. **The child should never be forcibly held down to receive an enema.** Give enemas **once a day** in the morning. Most children need two or three enemas to clear a faecal mass.
6. If these measures fail, the child should undergo manual evacuation of faecal mass under general anaesthetic, **but only if this is available and can be done safely.**

Maintenance laxatives to keep the stool soft, defecation pain-free and overcome faecal holding

- Softening agents such as Movicol or docusate sodium to keep the stool soft.
- Stimulant laxatives, usually senna or sodium picosulphate, to expel the soft stool. The aim is to produce loose stools initially and then subsequently reduce the dose to produce at least one soft stool per day. Often large doses will be required initially to overcome the child's faecal holding.

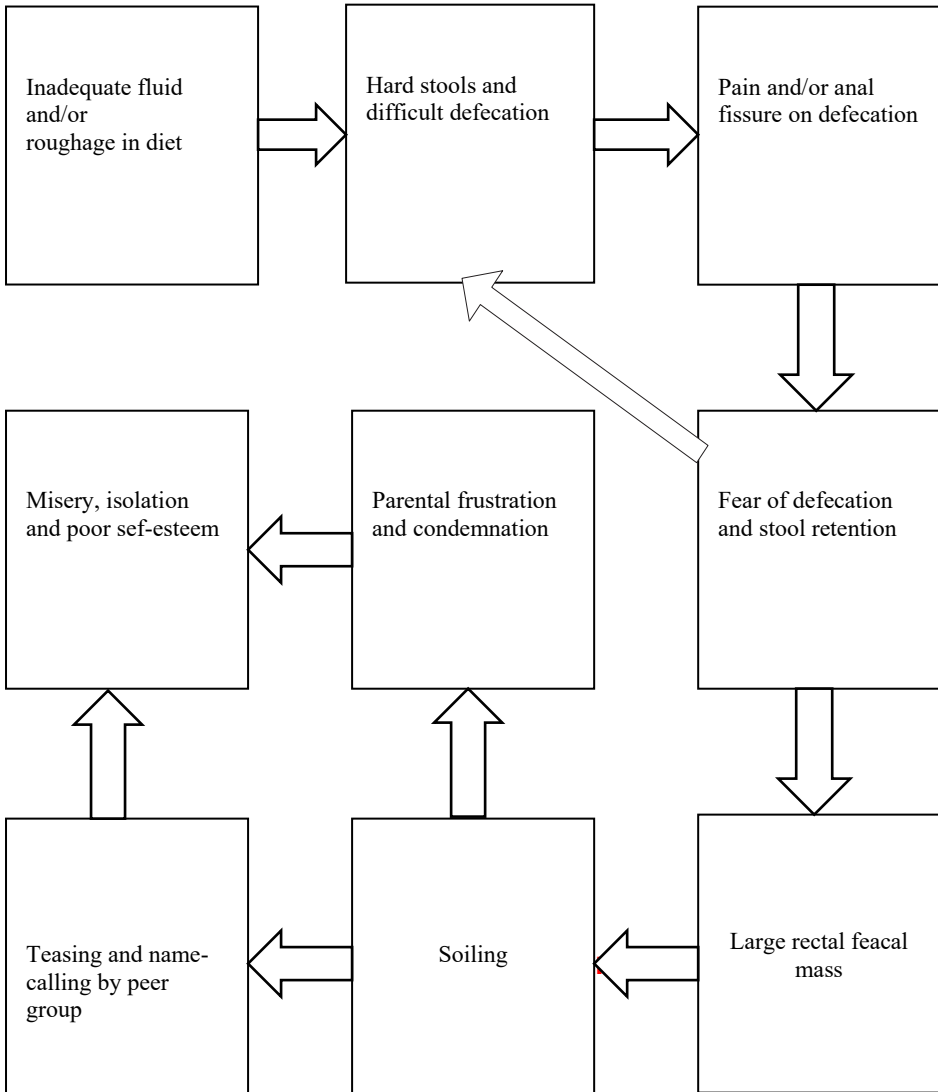
Behaviour changes

- Encourage increased fluid intake and a high-roughage diet (fruit, vegetables and cereals).
- Give positive praise and encouragement for regular toileting, and for passage of stool into the toilet.

Length of treatment

Children are likely to require several months of stimulant laxatives until their fear of defecation resolves, and often months to years of continuous or intermittent treatment with softening laxatives. A general rule of thumb is that the child will need laxatives for the same length of time that they were constipated before treatment started.

FIGURE 17.1 Sequence of events in faecal soiling.



Section 18. Cystic fibrosis

Cystic fibrosis is an autosomal-recessive genetic disorder that affects the lung, digestive system, sweat glands, liver, pancreas, and reproductive system. Most deaths from cystic fibrosis are caused by respiratory failure. In well-resourced countries, many patients now survive well into adulthood.

In the cells lining the airways of patients with cystic fibrosis (CF), chloride ions cannot leave the cell to enter the bronchial lumen. The cell cytoplasm has a high salt content, and water moves from the airway lumen into the cell by osmosis. The mucus within the lumen then becomes dehydrated.

Sticky mucus interferes with the action of the respiratory cilia, and this 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, causing poor fat digestion and very low fertility in male patients.

Incidence

The incidence of cystic fibrosis in countries such as the UK 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 the frequency of the disorder in resource-limited countries. Diagnosis relies on the sweat test, which is difficult to perform where laboratory facilities are limited. The incidence of cystic fibrosis among black South Africans is thought to be between 1 in 700 and 1 in 14000, with between 1 in 14 and 1 in 60 of the general population being carriers. In some well-resourced 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. This gene is as common in cystic fibrosis patients in North Africa as it is in those in Northern Ireland. Over 2000 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. As cystic fibrosis is a recessively inherited condition, two abnormal genes, one from each parent, are required for the disease to occur, and then this protein is defective and chloride transport is disrupted.

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 (e.g., Hirschsprung's disease, imperforate anus), and any sick newborn infant may develop non-specific abdominal distension. Around 15% of babies with cystic fibrosis present with meconium ileus (i.e., difficulty passing thick, sticky meconium, leading to small bowel obstruction).

Presentation in older children

This includes the following:

1. malabsorption (pale, greasy stools)
2. failure to thrive
3. rectal prolapse
4. chronic and recurrent chest infections
5. partially digested material with a high fat content may block the ascending colon (distal intestinal obstruction syndrome).

Diagnosing cystic fibrosis

The sweat test

This detects the high levels of chloride and sodium in sweat that occur in cystic fibrosis patients. The principle of the test is to allow the drug 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 are needed. Values highly suggestive of cystic fibrosis are concentrations of chloride and sodium of greater than 60 mmol/litre, with a higher concentration of chloride than sodium. False-negative and false-positive results are usually a consequence of faulty test technique, which is why there is a need for a specialised laboratory and experienced technician, which should be available in at least one hospital in every country.

Genetic tests

These can be performed on very small amounts of blood, collected as a dried blood spot on filter paper. It is possible to send dried blood spots to a genetics laboratory for analysis. A negative genetic test does not rule out cystic fibrosis (only common genes are tested).

Management

Treatment of children with cystic fibrosis in resource-limited countries has been identified as a priority area by the WHO. For practical reasons, children with cystic fibrosis can be seen regularly in a clinic alongside children with bronchiectasis. It is important that the child's parents understand that cystic fibrosis cannot be cured. However, these children can lead active lives with minimal symptoms initially, provided that daily treatment is given, and deteriorations are treated promptly.

Pancreatic enzyme supplements

Most children with cystic fibrosis will require pancreatic enzyme supplements (e.g. Creon, Solvay Healthcare, or Pancrease, Janssen-Cilag). Young infants are given half a capsule per milk feed. Older children may need over 10 capsules per meal. The capsules contain protease, lipase and amylase. The lipase is the most important component for preventing malabsorption. Most brands contain 5000–10 000 units of lipase per capsule. The correct dose of pancreatic enzyme supplements is not necessarily related to age, but rather it is the amount required to control symptoms of steatorrhoea (excessive fat in the stool) and achieve normal growth. The maximum dose (expressed as units of lipase) is 10,000 units/kg/day.

Fat-soluble vitamins

The child should be given extra fat-soluble vitamins. Appropriate doses are vitamin E, 50 mg once daily in infants and young children (aged 0–5 years), 100 mg/day in older children (aged 5–12 years), and 200 mg/day in patients over 12 years of age. Vitamin E may be given as vitamin E suspension 100 mg/mL or as 50 mg tablets.

Multivitamin drops, such as Abidec, which contains vitamin A 4000 units/6 mL and vitamin D2 400 units/6 mL, are also required. Abidec should be given as follows:

1. 0.3 mL/day for newborn infants
2. 0.6 mL/day for infants aged 1–12 months
3. 1.2 mL/day for children over 1 year of age.

Remember that an adequate calorie intake is vital. **Do not restrict fat in the diet.**

Chest physiotherapy

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 preschool children a foam 'wedge' helps the child to achieve the correct position for postural drainage. The percussion element of the treatment involves firm 'clapping' movements with the flat of the hand against the child's chest. 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', causing coughing, is suitable at this age. (For physiotherapy techniques, see Section 62)

Antibiotics

Children with cystic fibrosis have intermittent or chronic infection with *Staphylococcus aureus* in the first 2 to 3 years of life. *Haemophilus influenzae* is also seen in the early years and should be treated as a pathogen. In most children, chronic infection with *Pseudomonas aeruginosa* becomes established sooner or later. Later still, a variety of opportunistic organisms colonise the lungs. If the results of sputum or 'cough swab' cultures are available, 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. Ideally, the child's respiratory microorganisms should be monitored on a frequent and regular basis so that the appropriate antibiotic can be given promptly.

Antibiotic prophylaxis

In well-resourced countries, a continuous prophylactic oral antibiotic is often given to children with cystic fibrosis, up until 2 years of age. An antibiotic active against *S. aureus*, usually flucloxacillin, is chosen. In resource-limited countries this may not be an option, either because the diagnosis is made late or because continuous antibiotics are too expensive. However, flucloxacillin (125 mg twice daily) should be prescribed, if possible, for children under 2 years of age. Once mucoid *Pseudomonas aeruginosa* has become established, respiratory deterioration occurs, so in well-resourced countries various antibiotic regimes are used to reduce the bacterial burden in the lungs and slow down lung damage. Oral ciprofloxacin, inhaled nebulised colistin, or intravenous ceftazidime and tobramycin are variously used. Up-to-date details on antibiotic treatment for cystic fibrosis can be found on the website of the charity the Cystic Fibrosis Trust <https://www.cysticfibrosis.org.uk>

Treatment of exacerbations

If the cough worsens or the child produces more sputum, a full course of antibiotics should be started and continued for at least 2 weeks. Longer courses of antibiotics are given than in most other conditions. The following antibiotics are suitable.

Flucloxacillin

TABLE 18.1 Oral flucloxacillin doses

Age (years)	Dose	Number of doses/day
< 1 year	125 mg	4
1–6 years	250 mg	4
7–12 years	500 mg	4
> 12 years	500–1000 mg	4

Amoxicillin or ampicillin

TABLE 18.2 Oral amoxicillin or ampicillin doses

Age (years)	Dose	Number of doses/day
< 1 year	125 mg	4
1–7 years	250 mg	4
> 7 years	500 mg	4

Flucloxacillin, combined with amoxicillin, has good activity against *S. aureus* and *H. influenzae*.

An alternative is chloramphenicol, which is active against *S. aureus* and *H. influenzae*. Its activity against *P. aeruginosa* is poor.

Children with cystic fibrosis 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 cystic fibrosis.

The oral dose of *chloramphenicol* is 12.5 mg/kg 6-hourly.

If *P. aeruginosa* has been identified in sputum, or infection is suspected, use one of the following antibiotics.

Gentamicin

Dose: 7 mg/kg, once daily for 2 weeks.

Monitor gentamicin levels if possible. Peak is 5–10 micrograms/mL and trough is < 1 microgram/mL

Patients with cystic fibrosis often have more rapid renal clearance and have lower levels for a given dose than other patients. If possible, combine gentamicin with another antipseudomonal antibiotic, such as ceftazidime.

Ciprofloxacin

Dose:

Age < 1 year: 7.5 mg/kg/dose.

Age 1–3 years: 62.5 mg.

Age 3–7 years: 125 mg.

Age 7–12 years: 250 mg.

All age groups should have two doses of ciprofloxacin daily, and a course will last 2 weeks.

Ceftazidime

Dose: 50 mg/kg, three times daily, given over 30 minutes for 2 weeks.

Other manifestations and complications of cystic fibrosis

In addition to those features mentioned above under clinical presentation, the following may occur:

1. haemoptysis (not usually a major problem)
2. pneumothorax (usually small because of chronic pleural thickening)
3. bronchiectasis
4. biliary cirrhosis, portal hypertension and oesophageal varices
5. diabetes mellitus (requiring insulin)
6. infertility (in men)
7. children may become pregnant but will need careful management of their chest problems
8. 'meconium ileus equivalent' (obstructed bowel occurring in older children)
9. arthropathy.

With the best care or in those rare patients with mild disease, survival is possible into the fourth decade. Careful management will improve the quality of life greatly for children in resource-limited countries. Sadly, most patients with cystic fibrosis, in any part of the world without access lung transplantation, ultimately die of respiratory failure.

Section 19. Disorders of sexual development (DSD)

Uncertainty regarding a child's gender is a distressing emergency for the family. Most of these children are well unless associated with congenital adrenal hyperplasia (CAH) and salt loss (see above) or other major congenital abnormalities.

Avoid deciding the appropriate sex of rearing of the child until the results of diagnostic tests are available.

Support the parents during this difficult time.

DSD may be the result of excess androgens in females (the commonest situation, usually secondary to CAH of the 21-hydroxylase deficiency variety), lack of androgens (or the receptor) in males, or (rarely) mixed gonadal DSD with the presence of ovarian and testicular tissue. Minimum investigations are chromosome analysis and plasma for 17-hydroxyprogesterone (**which is** elevated in the commonest form of CAH). If a baby with a DSD becomes unwell with hypotension, hyponatraemia, and hyperkalaemia, assume an adrenal crisis and treat as described above.

Further investigation requires highly specialised tests (that is blood, radiology and ultrasound, fibroblasts, laparoscopy, or laparotomy). Treatment of non-CAH DSD is also complex but can often be deferred to allow appropriate transfer of care to a specialist centre. In specialist centres there need to be close working relationships between the different specialists. It is not appropriate for surgeons to operate without involving endocrinologists in working up these patients, and **it is essential that the members of the multidisciplinary team work closely together in the management of these cases.**

Congenital adrenal hyperplasia (CAH)

Congenital adrenal hyperplasia (CAH) is an autosomal-recessive condition, and therefore is more common in consanguineous relationships. Many forms exist, as several enzymes involved in the 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 over-masculinisation of the female (although under-masculinisation 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-beta-hydroxylase) causes salt retention and hypertension.

Females usually present as DSD (see above) and males with salt loss, which usually occurs 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 (see below).

Once the diagnosis is established, treat with mildly suppressive doses of hydrocortisone, 12–15 mg/m²/day in three divided doses and, if salt loss is demonstrated, fludrocortisone, 150–250 micrograms/m²/day in one dose. Infants may also require oral sodium chloride, 1 gram/10 kg/day (60 mg = 1 mmol).

Section 20. Eye problems

Introduction

Two of the most important eye disorders in children in resource-limited countries are vitamin A deficiency (xerophthalmia) and trachoma. Both can be prevented by appropriate action in the community, which is cheap and very effective for both disorders.

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 (see Figure 20.1).
- A bright torch light which can give a focused beam of light.
- An ophthalmoscope:
 - The ophthalmoscope is mainly used for examination of the ocular fundus (i.e. the retina, choroid and optic nerve).
 - It can also be used for examination of the ocular media (i.e. the cornea, lens, and aqueous and vitreous). Dial a small positive lens (about +2 or +3) in the ophthalmoscope 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 aqueous 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, sclera, iris, etc. To do this a very strong positive lens (about +20) is dialled in the ophthalmoscope, which is then held very close to the patient's eye.

There is an unacceptable lack of appropriate diagnostic instruments, and those that professionals have are liable to break down. An ultra-low-cost (12-14 USD) ophthalmoscope, otoscope and loupe which uses LED lights and is either solar powered or charged from a USB, so no batteries or bulbs to break down or be replaced, is now available <https://www.arlightscope.com/> Accessed 4th April 2021

- Mydriatic drops:
 - **Cyclopentolate 1%, or cyclopentolate 0.5%** in children less than 6 months old. **Atropine 0.5%** ointment is very long-acting. It can be given to parents to put into the eyes for 2 days prior to a clinic appointment, especially if an initial attempt at refraction and fundus examination has been unsuccessful because of the child becoming distressed when drops were used in the clinic.
- Local anaesthetic drops:
 - **Proxymetacaine 0.5% is ideal for children because it stings less than other topical anaesthetic drops. Tetracaine 0.5% or 1%** is an alternative which is less quickly degraded when not stored in the refrigerator.
- Sterile **fluorescein** paper strips.
- Binocular telescopic magnifying glasses (loupes) are very useful but not essential. Some magnification will be achieved by using a strong pair of reading glasses (+3.00–4.00 DS) perched as far down your nose as possible.
- More sophisticated equipment, such as a tonometer for measuring intraocular pressure, a slit lamp and a binocular indirect ophthalmoscope, may only be available in a specialist clinic. However, if available, they greatly add to the diagnosis and treatment that can be offered.

Gaining the confidence and trust of the child is the most important step in a successful eye examination, which should not be painful or unpleasant, except possibly unavoidably when drops are put in the child's eye. If the child finds it hard to cooperate, examine the parents' or older siblings' eyes first to gain the child's confidence. A general anaesthetic may sometimes be required in small children where a serious eye problem (for example retinoblastoma) is suspected.



FIGURE 20.1 Eye testing.



FIGURE 20.2 Position in which to examine the eye of a young child.

Three ways of examining the eyes of young children

Examining the eyes of babies and young children is often difficult. Patience and encouragement are required to gain the confidence of the child. If it is still difficult to get a good view, the following techniques may be helpful:

- Let the parent cuddle the child as he or she faces backwards over the parent's shoulder (see Figure 20. 2), especially if the parent's anxiety and sense of obligation to restrain the child is adding to the child's fears. You may then be able to attract the child's interest in participating in the examination from this secure position.
- In the case of infants, wrap the baby in a sheet or blanket, with their head on the examiner's lap, and their body on their mother's lap (see Figure 20.3). Gently hold open their 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.
- If it is difficult to get drops into the child's eye, try lying the child flat on their back, create a puddle of drops at the inner canthus, and wait while the child is held facing upwards (see Figure 20.4). The child will eventually open their eye, and the medication in the puddle of drops at the inner canthus will then go into the eye.
- In difficult cases, where a serious eye condition is suspected, it may be necessary to instil a drop of local anaesthetic and use a speculum to hold open the eyelids. However, this should only be done by an experienced professional in controlled circumstances and

must not be attempted in the face of determined resistance from any but the smallest child.



FIGURE 20. 3 Supine posture for eye examination.

Presenting symptoms of eye disease

These include the following:

- red, sore, irritable or discharging eyes
- impairment or loss of vision
- squint.

Red, sore, irritable or discharging eyes

- A sticky discharge with no redness, normal cornea and apparently normal vision in a child up to the age of 18 months (and occasionally older) is commonly caused by a blocked tear duct. Teach the mother to express the lacrimal sac with firm pressure to the side of the nose at the inner canthus.
- Bilateral sore red irritable eyes are usually caused by conjunctivitis. If the symptom is unilateral the usual cause is an ulcer or injury to the cornea or iritis. Evert the upper eyelid to inspect the upper tarsal conjunctiva. Apply fluorescein stain to the cornea to diagnose an ulcer or identify a foreign body. The green fluorescein dye will stain the ulcer. A foreign body, especially if lodged under the upper lid, may be associated with staining of the cornea.



FIGURE 20.4 Seated posture for eye examination.



FIGURE 20.5 Expressing the lacrimal duct. © www.medscape. Com

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.**
- The WHO-recommended treatment for severe neonatal conjunctivitis is a single IM injection of either **ceftriaxone 50 mg/kg** (maximum 125 mg) or **kanamycin 25 mg/kg**

(maximum 75 mg) and hourly **tetracycline ointment** or **chloramphenicol drops or ointment**.

- In presumed gonococcal infection, empirical treatment for possible co-infection with chlamydia – that is, ceftriaxone **and** erythromycin to prevent chlamydial pneumonia in the baby – should be strongly considered.

In addition, we recommend diagnosis and treatment of the mother for uro-genital disease due to gonococcus and/or chlamydia in order to prevent salpingitis.

- **Acute viral conjunctivitis** is a self-limiting disease that usually lasts for a week or so. Tear secretions are watery rather than mucopurulent. There is no specific treatment, but it is customary to give antibiotic drops.
- **Vernal conjunctivitis** is a chronic allergic conjunctivitis which is very common and causes recurrent severe itching of the eyes. Affected children are usually atopic (i.e. suffer from asthma and eczema). In addition to itchy eyes, there may be redness, watering, lid swelling and a mucus discharge. Typically, there are papillae of the conjunctiva under the upper lid. In some cases, these can be massive in size and may be associated with corneal ulceration in the upper third of the cornea. There may be nodular swelling and opacity at the corneo-scleral junction (i.e. the limbus). Anti-inflammatory drops such as cromoglycate relieve the symptoms, but in severe cases use topical steroids (e.g. hydrocortisone 1%, betamethasone 0.1%, or dexamethasone 0.1% eye drops). However, prolonged use of topical steroids has a high risk of causing steroid-induced glaucoma.

Trachoma

See Section 50

Corneal ulcers

- Corneal ulcers are usually **unilateral**. There is usually pain and photophobia. 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%** every 2 hours until the epithelium has healed.
- **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 2-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 one of these is the first choice.** Concentrated locally made antibiotic drops are very helpful if pre-prepared drops are not available. These can be made up by diluting antibiotic powder for injection in 5 mL of sterile water or 0.9% saline. These home-made eye drops should only be used for 48 hours and should then be discarded. The following are the recommended strengths: gentamicin 15 mg/mL or amikacin 50 mg/mL for Gram-negative organisms; cefuroxime, ceftazidime, or cefazolin 50 mg/mL for Gram-positive organisms. 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 are 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 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 either these or systemic antifungal agents in difficult cases.

Iritis

Iritis is a less common cause of 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 only affects malnourished children (see Section 55 Handbook 1).

- 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, which 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 the 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.
- Give vitamin A capsules (200 000 IU/day in children over 1 year of age, 100 000 IU/day for those aged 6–12 months, and 50 000/day for those under 6 months, for 2 days, then another dose in 2 weeks). Systemic antibiotics and rehydration may also be indicated.

The child who cannot see or who cannot see well

If only one eye is affected, the child and their 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 that is severe enough to cause serious visual impairment is most commonly a consequence of xerophthalmia and measles (both of which are preventable, by giving 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 sight-threatening congenital ocular abnormality. It may be present at birth or may develop in early childhood. It may be complete, presenting as a dense white opacity in the pupil, or be incomplete and less obvious. There will be a normal pupillary light reflex, so that the pupil constricts when a light is shone into the eye. In other causes of a white appearance of the pupil, including retinoblastoma, the reaction of the pupil to a light shone in the affected eye is usually lost.

Congenital cataracts require early expert surgical treatment, otherwise the child will develop 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 intra-ocular 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 (see Section 15. Neonatal Handbook). It is now particularly common in middle-income countries, such as Latin America, Eastern Europe, the Middle East and Asia. In countries with highly developed intensive neonatal care services, it is uncommon, and in resource- limited 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 that can be fatal in 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 following an infection such as typhoid or measles. There is no effective treatment.

Cortical blindness

Cortical blindness occurs following severe brain insults such as meningitis or cerebral malaria. The pupillary light reflex is normal, but the child cannot see. In some cases, the vision gradually improves with time.

Management of blindness

- Cataracts and glaucoma must be recognised and diagnosed early so that the child can be referred to a specialist centre promptly for surgery to preserve and save as much sight as possible.
- For most cases of retinal, optic nerve and cortical visual disorder, management is with rehabilitation and education rather than medical treatment.
- 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 opportunities for education. These aids may consist of a strongly positive lens worn as spectacles or used as a stand magnifier.

Squint

Squint, or misalignment of the eyes (also known as strabismus), is common in children. When assessing a child for squint, consider the following:

- **Does the child really have a squint?** Look at the corneal light reflexes. If the reflection of light is in the same position in each eye, 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 the 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?** Any disease which causes reduced vision in one eye is liable to cause a squint to develop. Test the pupillary light reflex and then dilate the pupils with mydriatic eye drops. Look for diseases such as cataract, retinal scar and in particular retinoblastoma. Refer the child for treatment if you find cataract or an abnormality in the retina. Treatment for retinoblastoma is urgent enucleation.
Retinoblastoma may appear as a creamy mass occupying all or part of the pupil or as a smaller lump arising from the retina. Retinoblastoma requires urgent referral to a specialist centre for life saving treatment. Prompt referral is of the utmost importance and is the principal reason why the mortality from retinoblastoma is close to zero in most well-resourced health care systems but close to 100% in many resource poor environments. Chemotherapy may be effective and affordable (see Section 15) or, if not available, urgent surgical enucleation may be the only way of saving the child's life.
- **Is there a refractive error, such as hypermetropia (long sight) or myopia (short sight)?** This requires refraction tests.
- **Is the squinting eye amblyopic (i.e. is there poor vision in the squinting eye)?** 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 1 hour a day for several weeks.

Amblyopia only develops in young children, and it can only be effectively treated in children under 5 years of age. Surgery may be required but should not be considered until eye disease and refractive errors have been excluded and amblyopia has been treated.

Further reading

Saul Rajak,, John Sandford-Smith Eye Diseases in Hot Climates Fifth edition

Section 21. Haemolytic anaemia

Definition

Haemolytic anaemias are disorders characterised by a reduction in the lifespan of red blood cells and may be congenital or acquired.

TABLE 21.1. The causes of congenital and acquired haemolytic anaemia

Congenital	Acquired
Haemoglobin defects: sickle-cell disease, thalassaemia	Infection: malaria, visceral leishmaniasis
Red cell enzyme defects: G6PD deficiency, pyruvate kinase deficiency	Alloimmune: haemolytic disease of the newborn, transfusion reactions
Red cell membrane defects: spherocytosis, elliptocytosis	Red cell fragmentation: haemolytic–uraemic syndrome
	Autoimmune caused by infection (e.g. EBV, CMV, HIV, mycoplasma), malignancies (lymphomas, leukaemias), immune deficiencies
	Drugs
	Burns

Clinical features of haemolytic anaemia

These include **pallor, jaundice, splenomegaly, and gallstones.**

The degree of splenomegaly can be a useful clue to the cause of haemolytic anaemia.

Intravascular haemolysis will give rise to haemoglobinuria.

Laboratory features of haemolytic anaemias: general

These include low haemoglobin, increased reticulocyte count, raised and predominantly unconjugated bilirubin, pink plasma after centrifuging of blood (due to free haemoglobin) in severe cases, reduced haptoglobin, raised LDH. Urinary products depend on whether there is intravascular or extravascular haemolysis.

Hereditary haemolytic anaemias

Red cell membrane defects (often dominant inheritance)

Hereditary 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 haemoglobin concentrations of 40–50 grams/L to asymptomatic individuals with normal haemoglobin levels. Neonates often will have jaundice and occasionally have a short period of transfusion dependency early on that later settles. Parents/patients need to be given advice regarding parvovirus B19 aplastic crisis, how to recognize the symptoms and how to seek medical help. It is common to have an increased haemolytic rate and reduction of haemoglobin at times of infection. These are usually self resolving but sometimes may need transfusion. The clinical phenotype often runs true within families, that is when parents are severely affected so are children. Some children will have a de novo mutation.

Diagnosis

- Along with a positive family history, the clinical features are mild jaundice, pallor and splenomegaly. Gallstones may occur in children.
- Laboratory features: blood film shows spherocytes, increased osmotic fragility of red cells, increased reticulocytes, negative antiglobulin (Coombs’) test. Next generation

sequencing is now used to identify the responsible genetic mutation though is not often available in resource-poor countries.

Treatment

- Folic acid 1 month—5 years 2.5mg daily. 5 years and older 5mg daily.
- Severely anaemic and symptomatic moderately anaemic children, and those who are transfusion dependent may benefit from splenectomy if the facilities available make this a low-risk procedure.
 - **Splenectomy carries a major risk of life- long increased vulnerability to infection with capsulated bacteria such as pneumococci, meningococci and Haemophilus influenzae type B. The risks and benefits need to be weighed up very carefully before splenectomy is undertaken.**
 - Delay splenectomy until after the age of 5–10 years if possible.
- Administration of **pneumococcal, meningococcal and HiB vaccine** prior to splenectomy, (pneumovax will need to be repeated 5 yearly), and lifelong prophylactic oral **penicillin V** thereafter (under 12 months of age, 62.5mg twice daily; 1–5 years 125 mg twice daily; over 5 years 250 mg twice daily).

Elliptocytosis

This condition is less common than spherocytosis. It is rare in European populations but is seen more often in West Africa. In South-East Asia there is a variant, South-East Asian ovalocytosis (SAO), which causes oval-shaped red cells and neonatal hyperbilirubinaemia, but little haemolysis later in life.

Diagnosis

- --Blood film shows 25–90% of oval, elliptical or rod-shaped red blood cells.
- --Heterozygotes often have only a very mild anaemia with very little haemolysis. Homozygotes tend to have severe haemolytic anaemia from infancy.
- Next generation sequencing can identify the mutation
-

Treatment

This is the same as for spherocytosis however, if the child does not have a high haemolytic rate then folic acid and long term follow up may not be necessary. It is rare to need splenectomy.

Stomatocytosis

Hereditary stomatocytosis is rare, but it can be acquired in several conditions, especially liver disease.

Diagnosis

Blood film shows erythrocytes with a central mouth-like slit (stomatocytes).

Treatment

This is the same as for spherocytosis, but **splenectomy is ineffective and may be harmful**, leading to a thrombotic tendency.

Metabolic defects

Glucose-6-phosphate dehydrogenase deficiency (G6PD) (X-linked)

There are two types of normal G6PD enzymes (types A and B). Worldwide, there may be 100 million people with diminished red cell G6PD activity. G6PD A deficiency is common in children of African descent, and their G6PD function is reduced to about 10% of normal. G6PD B deficiency (G6PD Mediterranean) is less common, and 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 causes episodic haemolytic anaemia and jaundice. Some patients have ongoing haemolysis but this is very rare.

Typical triggers of haemolysis are related to oxidant stress and include:

- favism due to ingestion of the fava broad bean or inhalation of its pollen. Lentils and soya are permitted.
- oxidant drugs such as antimalarial drugs, sulphonamides, high-dose aspirin, quinidine, quinine, nitrofurantoin, phenacetin. Note that the following are well tolerated: normal dose paracetamol and ibuprofen; immunisations; vitamin K given to newborns. A patient who needs a medication should check whether it is known to cause significant haemolysis. If primaquine is necessary, this can be given weekly for 8 weeks.
- other chemicals, such as those in mothballs (naphthalene), can also trigger an episode.
-

Diagnosis

- Blood film shows 'blister' and 'bite' cells. Heinz bodies may be seen on unstained blood film.
- Enzyme assay (if available) is needed to make the diagnosis (but this may be normal if reticulocyte numbers are raised). It may be necessary to wait several weeks after an acute episode before measuring enzyme levels.

Next generation sequencing can be used to identify the mutation and is helpful to identify carriers or those who have a high reticulocyte count.

Treatment

- Avoid drugs that cause oxidant stress (i.e. chloroquine, primaquine, sulphonamides, nitrofurantoin, quinolones, dapson, high-dose aspirin, phenacetin) or fava beans. If primaquine is necessary, this can be given weekly for 8 weeks.
- Patients usually recover spontaneously once the precipitating factors have been removed.
- Transfusion may be necessary if there is severe haemolysis. Treatment will also involve folic acid, at treatment doses, and may need iron replacement to reconstitute the normal haemoglobin level as there is haemoglobin and thus iron loss in the urine.

Pyruvate kinase deficiency (autosomal recessive)

This is the second commonest enzyme defect of the glycolytic pathway and affects mainly northern Europeans.

Clinical features

These are very variable.

- Neonates may have severe haemolysis and present with early jaundice (within 48 hours), anaemia and hyperbilirubinaemia.
- 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 and an increased tendency to long bone fractures.

Diagnosis

- Blood film shows increased reticulocytes, Heinz bodies and mild macrocytosis.
- Enzyme assay for pyruvate kinase.
- Next generation sequencing can identify the mutation.

Treatment

- Splenectomy (only if the facilities available make this a low-risk procedure).
- Transfusion if there is severe anaemia or an aplastic crisis.
- There are new agents that are available that can treat the disorder directly. Some of these are still in clinical trial

- Folic acid should be given to all patients with a raised haemolytic rate to prevent deficiency.

Haemoglobin defects

- Abnormal variants: sickle (see Section 26 Handbook 2), Hb C, Hb E, Hb D, etc.
- Defective synthesis: thalassaemias.
- Beta-thalassaemia major (autosomal recessive).

Thalassaemias

Introduction:

Thalassaemia is an umbrella term for a range of defects in the alpha and beta globin genes. The location and extent of the defect reflects the change in the amount of product made. Some defects are minor, and some are quite extensive with almost no alpha or beta chains produced. This is unlike sickle cell disease and the associated abnormal haemoglobins e.g. HbC which arise from a single point mutation.

Alpha Thalassaemias

There are 4 alpha globin genes

It is very common to miss one (alpha plus thalassaemia heterozygote) or miss two, one from each allele (alpha plus thalassaemia homozygote). Patients have a lower MCH, lower MCV and may have a mildly reduced haemoglobin. Haemoglobinopathy screen is normal though sometimes HbA2 is reduced. These people are well though it is important not to confuse this with iron deficiency (which is common) as unrestricted iron supplementation in someone who is not deficient can lead to iron overload. You can confirm diagnosis with genetics though this is not usually offered.

As HbF is comprised of alpha (and gamma) globin, a foetus needs at least one functioning alpha gene to survive pregnancy. Missing two alpha genes from the same allele is known as alpha zero heterozygote and this is much rarer and is usually seen in people from southeast asia and southern Europe e.g., Cyprus.

If two people with alpha zero heterozygosity make a baby, then there is a ¼ chance of the baby having no functioning alpha genes. This is known as alpha thalassaemia major and leads to profound foetal hydrops and foetal death. Patients can be transfused in utero and then brought up as a transfusion dependent thalassaemia patient, but this is rarely undertaken. Antenatal screening programmes should identify alpha zero heterozygotes through their blood count and ethnic origin and should be offered partner testing.

HbH disease is when three of the four genes are non-functioning/absent. Patients have a low MCV and MCH and haemoglobin 80-100g/l. Blood film can be very abnormal with HbH inclusions. There is an HbH peak on the haemoglobinopathy screen e.g., HPLC. People are surprisingly healthy though may need transfusion support at times of additional stress e.g., pregnancy. The constant spring genotype can give a more thalassaemia intermedia phenotype and results in organomegaly and transfusion dependency.

Beta thalassaemias:

Background

- There are over 200 thalassaemia mutations
- Each person has two beta globin genes, and they are used to form adult haemoglobin known as HbA. They are not used to form part of fetal haemoglobin, so children do not become symptomatic until the HbF to HbA is switched over post natally.
- Severe mutations result in absent globin (β^0)

- With milder mutations some globin is produced (β^+)
- Note haemoglobin E is an abnormal haemoglobin that combined with a beta thalassaemia mutation produces a thalassaemia type syndrome
- The gene is less widespread than alpha thalassaemia: southern Europe, middle east, South and South East Asian mostly

Beta thalassaemia trait:

- One of the two beta globin genes is faulty
- Diagnosis is made when MCV and MCH reduced and HbA2 is raised – we don't routinely genetic test
 - Often have a mild anaemia – e.g., Hb 90-130g/l
 - The patient will need genetic counselling,
 - Autosomal recessive carrier parents, $\frac{1}{4}$ chance of an affected child

Beta thalassaemia (two genes affected)

- ◆ When both genes are affected then the person will need clinical care and follow up.
- ◆ It very much depends on how much haemoglobin the patient can make as to how badly affected they are.
- ◆ If the person has two milder mutations for example, they may not be transfusion dependent – this is known as non transfusion dependent thalassaemia (NTDT) - this used to be known as thalassaemia intermedia
- ◆ If the person has more severe mutations, they may require regular transfusions – this is known as transfusion dependent thalassaemia (TDT) – this used to be known as thalassaemia major.

Diagnosis:

- FBC: low MCV and MCH, often high red cell count
- Haemoglobinopathy screen depends on what is being made and when it is being tested – all the patients will have significant HbF at birth
 - Beta thalassaemia trait, raised HbA2 (need to wait until 18 months of age)
 - HbE beta zero thalassaemia: HbE only (though HbF present if test early)
 - Homozygous beta zero thalassaemia HbF only
 - Compound heterozygote Beta zero beta plus thalassaemia, mostly HbF some HbA
- Genetics of beta globin gene allows you to predict transfusion dependency and clinical course
- Genetics of alpha globin gene: if alpha genes reduced then this improves clinical phenotype (less imbalance) if increased then worsens it
- Genetics of xmn polymorphism allows production of HbF beyond the neonatal period (can be significant production allowing for transfusion independency)

Screening:

Linked antenatal and newborn screening programmes for thalassaemia are the cornerstone of good patient care. Carrier parents can be identified so they may choose, should they so wish and if the facility exists locally, to have prenatal diagnosis. Also, the heel prick test will identify babies at birth so treatment can be instituted to prevent morbidity and mortality. For sickle cell this work mostly surrounds early uptake of penicillin and entry into clinical care. For thalassaemia this is more about identifying children so that anaemia and growth can be kept under review, so they do not present critically anaemic at emergency department at several months of age or die before diagnosis – which is an all-too-common scenario in resource poor countries.

Clinical Presentation

Beta thalassaemia patients can present in several ways. The most common ways to present are with severe anaemia which can be quite profound for example less than 20 g/L of

haemoglobin or with features of extra medullary haematopoiesis such as hepatosplenomegaly. In countries where access to healthcare is poor patients may present with severe features of extra medullary haematopoiesis such as:

- frontal skull bone bossing
- facial changes
- stunted growth
- cord compression
- severe hepatosplenomegaly

Patients who are identified as newborns due to screening or family history are fed into clinical care. That way they are monitored so that treatment is instituted prior to anaemia becoming too severe or before there is significant extramedullary haematopoiesis.

Clinical features of thalassaemia arise from

- Anaemia
 - Difficulty feeding
 - Loss of appetite
 - Failure to thrive
 - Breathlessness, tachycardia
 - Lethargy
 - It is often these that lead to the decision to transfuse.
 - Anaemia often results in a loss of appetite and both the decreased calorific intake, and the catabolic burden of excess erythropoiesis will lead to a failure to thrive.
- Intramedullary expansion:
 - Pathological fractures
 - Facial changes and other bone deformities
 - Osteoporosis
- Extramedullary haematopoiesis
 - Hepatosplenomegaly (note splenectomy increases the risk of thrombosis significantly)
 - Lymphadenopathy
 - Masses which can cause obstruction e.g. cord compression
- Increased iron absorption
- Increased iron intake through blood transfusion
 - Heart_ arrhythmias and death
 - Liver: fibrosis, cirrhosis, and hepatocellular carcinoma
 - Gonadal axis: hypogonadotrophic hypogonadism – pubertal failure and infertility
 - Pancreas: diabetes and exocrine deficiency
 - Thyroid: hypothyroidism
 - Parathyroid: hypoparathyroidism

Essentially the health of the patient and the development of complications is dependent on their access to blood and chelation that are given in the right quantities. That is if you can transfuse someone with blood and then chelate well, they will be in excellent health with little or no complications. If, however, you only have some access to blood, then they will grow well in childhood, have pubertal failure, develop further endocrine problems, and die of a cardiac arrhythmia in early adulthood. What is a common scenario in resource poor countries is that patients have some but imperfect access to care so that they are alive, sometimes into ages over 30 years but eventually die of liver or cardiac disease having suffered many of the complications outlined below. It should be mentioned that even in resource rich countries, older patients (45 years+) will not have had good chelation in childhood (it was not available) and compliance is still an issue even when access to care is good, so morbidity in thalassaemia is not insignificant.

Transfusion in thalassaemia

a. Rationale

There are two reasons for transfusing in thalassaemia.

1. To increase the haemoglobin
 - If haemoglobin very low
 - If patient is symptomatic
 - If child not growing properly
2. To suppress endogenous erythropoiesis and thus the complications of extra medullary and excessive medullary haematopoiesis

Some patients may need a one off transfusion before becoming transfusion dependent e.g. if they have an infection or are due for surgery

b. Transfusion requirements

- ABO Rh (CcDEe) and Kell compatible blood negative for clinically significant antibodies
- Full red cell genotype/phenotype pre transfusion if available
- All the systems should be in place to make transfusion safe e.g., haemo-vigilance, donor screening, record keeping etc.
- Leuco-depleted packed cells if available
- Younger blood preferably <2/52 if available

c. Aim of transfusion

- To deliver a safe and effective transfusion regimen whilst minimising the burden of transfusion therapy on everyday life
- Good growth and development
- Good energy levels
- Sufficient suppression of intra and extramedullary haematopoiesis

d. Transfusion targets

- The aim is to keep the trough Hb>95g/l , (usually by having a peak of Hb 140/150g/l) though this may need to be increased e.g. significant extramedullary haematopoiesis
- To achieve this, commonly patients have a transfusion every 3-4 weeks

Chelation

- Chelators are medicines that remove iron from the body
- Examples of chelators include:
 - Desferrioxamine (desferal): taken as an infusion 5-7 days a week
 - Maximum average daily dose is 40mg/kg/d in growing children can be increased up to 60mg/kg/d once growth has stopped
 - Risk of audio and oto-toxicity that increases with increasing doses
 - Deferiprone (ferriprox): a tablet taken three times a day
 - Risk of fatal agranulocytosis
 - License includes regular monitoring of FBC
 - Deferasirox (EXJADE): a tablet taken once a day
 - Not licensed <2 years of age nor in renal dysfunction
 - Causes creatinine rise and proteinuria often
- Chelation is usually commenced when the ferritin reaches 1000ug/l though with improved safety and ease of chelation, patients are often started on chelation prior to this.

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 Section 19 and 65 Handbook 2.).

- Check the following at least four times yearly: serum ferritin (iron overload), liver function tests, bone, renal function, vitamin D. For those who are iron overloaded additional monitoring will be needed e.g., thyroid and parathyroid, glucose tolerance test, tests of hypothalamo-pituitary axis. Those on chelators need more frequent monitoring. Please see references for more information on monitoring and management in thalassaemia
- Undertake yearly screening for HBV, HCV and HIV infection.
- If HCV is positive, assess viraemia (serotype) if possible, perform a liver biopsy and give interferon with or without ribavirin to avoid cirrhosis and hepatoma.
- If HIV-positive, continue transfusions and give the latest available antiviral treatment.
- All blood donors should be tested for HCV and HIV.

Acquired haemolytic anaemia

Immune mediated

- Haemolytic transfusion reaction.
- Haemolytic disease of the newborn (see section 19 Neonatal Handbook)
- Hypersplenism.
- Secondary to infection: EBV, CMV, Mycoplasma, rarely HIV.
- Secondary to malignancies: lymphomas, leukaemias.
- Secondary to autoimmune diseases: SLE, rheumatoid arthritis.

Diagnosis

- Anaemia with increased reticulocytes.
- Splenomegaly.
- Positive direct Coombs' test.

Management

- Most secondary cases (70–80%) are transient, lasting about 3 months.
- Infants and older children may develop the chronic form.
- Treatment may not be needed if the symptoms are not severe.
- Transfusion may be necessary if there is severe haemolysis.
- Steroids: prednisolone 2mg/kg/day (up to 6mg/kg/day in severe cases) can be given if treatment is needed until the rate of haemolysis declines, and then stopped gradually. Further information can be found here ([How I treat autoimmune hemolytic anemia | Blood | American Society of Hematology \(ashpublications.org\)](#)) Accessed 8th April 2021

Malaria

See Section 31 Handbook 1.

Secondary to organ disease

Renal failure (see Section 47 Handbook 1).

Liver disease (see Section 49 Handbook 1).

Burns (See Section 86 Handbook 1)

Miscellaneous

- Chemicals and drugs.
- Toxins (e.g., Haemophilus influenzae type B, staphylococcal, streptococcal, clostridial).
Venoms (e.g., cobra, viper, rattlesnake, bee, wasp, yellow jacket).

Further reading

British Committee for Standards in Haematology <https://b-s-h.org.uk/guidelines>

Accessed 8th April 2021

Section 22. Mental health conditions in children

Introduction

Around 10–20% of all children have one or more mental or behavioural problems (World Health Report 2001). The rates are higher in urban areas and increase in adolescence. One in ten young people suffers from mental illness or symptoms of mental distress severe enough to cause some level of impairment, yet less than one in five receives the treatment that they need.

Prematurity, poor nutritional status, low birth weight, organic brain damage and physical handicap often bring about biological stressors. A 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/ incarceration of a family member/ a history of substance abuse. Protective factors include stable care, an adaptable and engaging personality, problem-solving abilities and a supportive network of family and friends.

The total disease burden of these disorders has not been estimated, and it is complex because many of these disorders can be precursors to much more disabling disorders during later life. Mental health disorders of childhood and adolescence are very costly to society in both human and financial terms.

Psychiatric disorders that arise 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 the parents. There is also intense emotional interaction with friends, which makes adolescents especially vulnerable to the effects of peer pressure, and issues of sexuality. Emotional disorders include anxiety states, depression, hysteria, and specific phobias.

Conduct disorders occur in about the same proportion and include conditions that range from oppositional defiant behaviour to persistent patterns of aggression and rule breaking. About 20% of the teenagers may present with a mixture of disorders.

The link between adverse family environmental factors and mental health disorders in children and adolescents is fundamental and must be explored as part of the assessment and treatment.

Excellent information and resources are available through the Royal College of Psychiatrists (UK) website: <https://www.rcpsych.ac.uk/mental-health/parents-and-young-people>

Acute psychiatric emergencies: suicide and deliberate self-harm

In well-resourced countries there has been a persistent rise in fatal suicide attempts, especially in young males. A history of substance abuse, conflict with the law and personal and mental illness are important factors. The possibility of abuse within (most likely) or outside the family must be at the forefront of a search for why this occurred, as there may be other children 'at risk'. The method of suicide depends on the means available. Males are more likely to use violent means than females. Overdoses of drugs or poison, hanging and immolation are common methods of suicide.

Suicide is extremely rare in pre-pubescent children, but the frequency rises sharply during the teenage period. Again, a search for evidence of abuse must be undertaken.

Deliberate self-harm is a non-fatal act in which a child or young person deliberately ingests noxious substances in excess of therapeutic doses or causes self-injury. It is best interpreted as a 'cry for help'. Again, the possibility of abuse must be considered in all cases.

Assessment and questions to be asked

- Are there any indicators or clinical signs of physical or sexual abuse?
- Have there been any previous attempts at suicide?
- Is there a risk of suicide or of a repeated attempt?
- Was a suicide note left?
- Was there pre-planning?
- How likely was the young person to be found?
- What was the method used?
- How lethal was the method used?
- Did the young person know how toxic the substance was?
- What quantity of the substance was taken?
- Was it impulsive in the context of a conflictual relationship?
- Was it to attract sympathy or seek attention (e.g., following a disciplinary crisis or the 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 and cope with difficulties?
- How effective and who are the social/parental supports, including adequacy of 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

- Undiagnosed and unmanaged abuse
- The risk of repetition is higher in the next 4 weeks after the attempt. It also increases if there is a history of previous self-harm attempts
- Male gender
- Lack of support, and easy access to a means of committing suicide (e.g., a firearm or drugs belonging to other family members in the home)
- Presence of depressive illness, with loss of sleep, appetite, depressed mood, agitation, and in particular continued suicidal ideas (hopelessness, inability to enjoy life, asking 'What's the point?')

Treatment

- Treatment of the medical consequences of self-harm is the priority (see Section 87 Handbook 1).
- Assessment of the child and their family should be undertaken when the child is free from the after-effects of the drug overdose/self-inflicted injury.
- It is important to take the young person's suicidal ideas seriously and not to expose them to sarcasm or ridicule in discussions. Assessment should include consideration of a mental illness and of the family and social circumstances of the young person and the context in which the self-harm occurred.
- Nothing predicts behaviour better than past behaviour. Those with a low risk of repetition can be offered support during subsequent crises, and arrangements made to assist the child and their family in developing coping strategies. A psychologist, social worker or trained psychiatric nurse (if available) can assist the family in this way.
- If there is abuse, the child must be protected from further harm by arranging the involvement of social services and the police (as appropriate in the setting).
- Children at high risk of death need a major input, although inpatient 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.
- The presence of mental illness merits specific treatment (psychological therapies and/or medication) and intervention (see below). Issues that triggered the self-harm should be addressed if possible. Relationship difficulties should be borne in mind.

Depressive Disorders

Depression is a recurring illness characterised by episodes of dysfunction. It is common and has a lifetime prevalence in adults of 15–20%. It has been reported in 1% of preschool children, 2% of school-age children and 5–8% of adolescents; girls are twice as likely to suffer from depression as boys. The incidence is rising, or else depression is being recognised more, with each successive generation. It is detected at a younger age and there has been a parallel increase in suicide in the paediatric age group.

Sadness, unhappiness, and misery are common childhood experiences (usually in reaction to adverse family circumstances), but when sadness is extreme in intensity and duration, it may be due to a depressive illness. Depression or depressive illness always needs urgent attention.

The presentation of depression varies with the age of the child. Infants and preschool children cannot express feelings of sadness in language. In this age group, depressive symptoms must be inferred from apathy, withdrawal from caregivers, delay or regression of developmental milestones, and failure to thrive that has no organic cause. School-aged children are cognitively able to internalise family conflict, criticism, or failure to achieve. They display low self-esteem and guilt, but depression is often mainly expressed in somatic complaints (headaches, stomach aches, disturbed sleep and appetite), anxiety (school phobia, excessive separation anxiety), irritability (temper tantrums and other behavioural problems) and academic decline.

Common symptoms in adolescents resemble adulthood depression, with more anger than sadness, hostility mainly towards family, sleep and appetite often normal, drug abuse, academic decline and suicide attempts. A depressed mood (dysphoria) is accompanied by loss of emotional involvement (withdrawal), feelings of guilt and unworthiness, 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 or occur despite an absence of adverse environmental causes, and functioning is impaired, a diagnosis of depressive illness can be considered. It is worth noting that around 40% of children with conduct (behaviour) disorders have associated mood disturbances, and that children presenting with depression may have other problems, such as anxiety, substance abuse and/or academic problems.

Assessment

Assessment should include the child or adolescent and their family or other people who know the young person well. This may be impossible for the teenager who has no family, or for the older teenager who refuses to have their 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, such as abuse, and in situations where the child's safety is at serious risk – for example, of suicide. Assess the degree of dysfunction and distress that the symptoms are causing the child and their family. Assessment requires a detailed history, mental state examination, play and

observation of the child–parent interaction. A full medical examination and neuropsychological testing to rule out neurological or learning disorders and to assess the child’s developmental capabilities should be undertaken. Psychological assessment tests such as a strength and difficulty questionnaire may be helpful.

Risk factors

- A family history of depression predisposes to depression, and the children of depressed parents are three times more likely to develop depression themselves. Early onset in the parents is associated with a higher risk for the children.
- Family and social environmental risk factors include family conflicts, rejection, lack of communication, lack of expression of love, poor family support systems, abuse (physical, emotional, or sexual), and parents who are excessively controlling.
- Adverse life events, such as the death of a parent or other loved one, parental divorce, exposure to suicide, relationship problems and academic failure can precipitate depression.
- Negative emotions such as low self-esteem, self-criticism, negative interpretation of life events and a feeling of lack of control can all contribute.
- The process of puberty can precipitate depression.

Issues in management

The diagnosis of a depressive illness (as opposed to transient sadness, which is very common) should be made only after careful history taking and information from the family, school and (if possible) close friends. **Most often ‘depression’ in children or adolescents is related to environmental factors.**

In addition, and in older adolescents, bipolar disorder may present for the first time, and a history of symptoms of hypomania should always be undertaken. If such symptoms are elicited, a daily symptom diary may be helpful. There are specific drug treatments for bipolar disorder which include mood stabilisers, and the support of an adult psychiatrist can be very helpful.

Medical conditions that can present with depressive illness must be excluded, using appropriate investigations, in particular vitamin or mineral deficiencies (full blood count), thyroid dysfunction (TSH levels), tuberculosis (chest X-ray) and HIV infection.

If possible, address any stressful factors in the child’s environment.

Management of sadness

The opportunity to discuss their difficulties with a sympathetic and helpful listener can itself be very useful to a depressed child or adolescent. The depressed child will tend to blame him- or herself, and there should be an attempt to enable the child to deal with issues without such negative feelings.

It is important to explore sensitively any factors in the child’s life that may have led to the episode of self-harm, and to put the problems right as far as possible, while recognising that some situations cannot be changed.

Work to help the young person to understand him- or herself, identify feelings, change maladaptive patterns of behaviour and improve relationships can be very helpful and will provide useful skills for them later on in their life.

Consider the mental health of other members of the family, which may be having a significant effect on the child or adolescent. Helping the parents may help the child. Postnatal depression occurs in about 10% of mothers and tends to recur with each pregnancy. Family therapy and support can also be very helpful.

Regular exercise (e.g., involvement in sport) can be very useful for some people with depression. It is also important to get enough sleep and to eat as healthy a diet as possible. Formal 'talking therapies' such as cognitive-behavioral therapy can be helpful, but they require specialist training and are not widely available, even in well-resourced countries.

Management of depressive illness

Cognitive Behavioural Therapy (CBT) is the mainstay for depressive illness in children.

It is vital that the child or adolescent is informed that their symptoms and the effects of the symptoms on their behaviour and educational function are not due to anything they have done or are doing wrong. The young person and their family need to learn how to distinguish between the normal range of feelings and those, including sadness, that suggest the onset or presence of the depressive disorder.

In the most severe forms of depressive illness, particularly in adolescents, antidepressants can be helpful, but in general their use is best avoided.

Antidepressant medication

When medication is needed Fluoxetine is the preferred antidepressant of choice in children and young people where the benefit outweighs the risk. It should only be offered in combination with psychological support.

The starting dose of fluoxetine for 5-18yr olds is 10mg daily. This can be increased to 20mg daily after 1 week if needed. There is little evidence for doses higher than 20mg daily.

There is an increased risk of suicide in children starting fluoxetine. This usually occurs in the first 4-5 days due to increased restlessness. This is particularly in those with suicidal thoughts already so are more likely to act on them. It is important to ensure their safety is maintained at home by a vigilant family member. Suicidal feelings must be explored routinely at onset and at follow-up.

Abrupt discontinuation of SSRIs may induce withdrawal symptoms, some of which mimic a relapse of a depressive episode (e.g., tiredness, irritability and severe somatic symptoms).

Once the patient has been free of symptoms and back to normal life for at least 8 weeks, fluoxetine should be continued for 6 months, then gradually reduced and stopped over a period of 6-12 weeks. The speed of reduction should be decided with the patient, taking into consideration any symptoms of withdrawal.

Tricyclic antidepressants have major side effects, including cardiovascular complications. They are also very dangerous in overdose. Therefore, they must not be used.

Monoamine oxidase inhibitors (MAOIs) must not be used because of their dangers when taken with certain foods.

Somatisation disorders

This refers to loss or alteration of physical functioning without organic cause. The child presents with physical symptoms which result in 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 post-pubertal female adolescents. They usually arise when the adolescent is facing a predicament that they cannot resolve. This may be related to academic, family, interpersonal, sexual, abuse, religion or societal issues.

As well as a thorough history and interview, the assessment should include a full physical examination to assess symptoms that do not correlate with known neurological pathways (e.g., a gait that is inconsistent and varying). It is important to keep an open mind about the possibility of a physical problem which has not yet manifested itself, and reconsider this regularly.

Pseudo-seizures may occur in adolescents who also have epileptic seizures, and it may be difficult to distinguish between them and provide the correct treatment. Involving a family member in record keeping and/or the use of a mobile phone video can help clarify.

Once a psychiatric disorder has been diagnosed, the 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. Whatever the cause, the disability and the impact on the young person's life are real and may be more difficult to manage than a physical illness with similar symptoms. This aspect of the disorder must be carefully explained to the family as well, in case they believe that the young person is faking an illness.

Drugs and alcohol: use and abuse

Drug use and availability have 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 drugs increases. Which drugs are illegal and which are socially acceptable vary between countries and among different groups of people in the same country, and the use of drugs by children and adolescents is influenced by this. A child's abuse of drugs is contextual – that is, it depends on societal norms, family history, etc. Parental criminality or substance abuse increases the risk. Reliable data on drug abuse in children are scarce and not validated. However, there is increasing acceptance that the rates of drug use are increasing, particularly in inner-city areas among children who are deprived (e.g., 'street children').

Volatile substance abuse is common in children, but seldom persists into adulthood. Solvents are easily available (e.g., nitrous oxide, butane gas, lighter fuel, paint thinners, aerosols, etc.), and are most commonly abused through a plastic bag or balloon to maximise the effects.

Stimulants such as cocaine and amphetamines are taken in powder form, intranasally or injected. They produce elevation of mood, energy, a reduction in appetite and hallucinations.

Drugs may be used for pleasure, or to remove (however briefly) the pain of daily life. Many drug users live in very difficult conditions and/or have mental health problems in addition to their drug habit. Prostitution is often associated with drug use, trapping (mainly women) in a vicious cycle. There is also a strong association of substance abuse with conduct disorder. In conduct disorders there is a repetitive and persistent pattern of behaviour in which societal norms or rules are violated (e.g., fighting, bullying, cruelty to people and animals, destruction of property, stealing and deceit). Many drug abusers will take up crime to pay for their drugs.

Assessment

It is important to establish the extent, frequency and severity of drug abuse or dependence. In addition, information needs to be elicited concerning behavioral patterns, social competency, educational functioning, peer relationships and psychiatric status.

Physical examination should include a check for fresh injection marks, old scars or the physical sequelae of drug use.

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, only a limited impact is made on alcohol or marijuana abuse, whereas heroin or cocaine treatment programmes are more successful in reducing the use of these drugs.

Methadone as part of a well-controlled and structured treatment system is the most common approach to managing long-term opiate dependence. However, it is rarely available in resource-limited settings. The initial dose for children over 15 years of age is 10–20 mg daily, increasing by 10 mg/day until there are no signs of withdrawal or toxicity (the usual dose is 40–60 mg/day). Opiates can give rise to nausea and vomiting. Withdrawal symptoms include restlessness, irritability, and increased bowel activity with abdominal pain. Methadone treatment 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 first diagnosed in late adolescence, although rarely the onset can be seen in childhood. Consider the diagnosis if two or more of the following symptoms are present for 1 month or longer:

- delusions: beliefs which are unshakeable
- hallucinations: 80% of affected children have auditory hallucinations; visual hallucinations are more likely to be due to an organic medical disorder such as a brain tumour or poisoning
- disorganised (incoherent) speech
- grossly disorganised or catatonic behaviour
- negative symptoms (flat affect).

The onset is usually insidious. Many children have pre-existing problems with social withdrawal, disruptive behaviour, developmental delay and language problems, and then go on to develop more florid symptoms such as hallucinations. Mood disorders may present with schizophrenic-like symptoms and making the diagnosis may be difficult.

Assessment

Any 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 young person's life (i.e. education, family and social functioning).

Before a definitive diagnosis of schizophrenia is made, organic medical conditions must be excluded by the following tests. However, it has to be accepted that in resource-limited settings many of these tests will not be available: blood tests for haemoglobin, indices such as MCV (to rule out vitamin B12 deficiency), thyroid function, liver and renal function, heavy metals such as lead, mercury and arsenic, HIV indicators, the Wassermann reaction for syphilis, urine test for toxicology, an EEG to help to rule out temporal lobe epilepsy, and a CT scan of the brain.

Management

Children and adolescents with schizophrenia present a challenge as they are seriously ill, and often their social and educational progress is seriously disrupted. Treatment is difficult, and all management should be under the supervision of a psychiatrist (if available). Treatments aim to reduce the frequency of relapses and disability.

It is important to work closely with the family. Negative symptoms such as blunting of emotions, impoverished thinking and lack of motivation are particularly distressing to relatives. Furthermore, highly expressed emotions and negative feelings increase the risk of

relapse. The family will need support and help to manage their child's symptoms. The techniques for reducing highly expressed emotions require specialist training.

Pharmacological treatment to control symptoms is the important initial management. Psycho-educational, 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 will quickly become apparent, but the medication must be administered daily, and the patient may not always be compliant. Depot neuroleptics provide a way of enhancing compliance.

Atypical antipsychotic drugs are now the treatment of choice (if available), as they have less extrapyramidal side effects. Risperidone can be given to children over 12 years of age, starting at 2 mg per day and increasing by 1 mg per week to a maximum daily dose of 8 mg. Side effects include postural hypotension, weight gain, hyperglycaemia, and mild extrapyramidal signs.

Standard antipsychotic drugs are more likely to be available in resource-limited settings. Standard antipsychotic treatment for acute schizophrenic symptoms is usually initiated with chlorpromazine. For children aged 12–18 years, start with oral treatment with 25 mg three times a day or 75 mg at night, and then gradually increase the doses until there is control of symptoms, usually achieved with a maximum dose of 100–300 mg daily. Premature changes in drug choice should be avoided, as the response time may be 30 days or more.

Poor response may be due to an inadequate dose or poor compliance.

Depot medication is suitable for long-term treatment (flupenthixol by deep IM injection into the outer buttock or lateral thigh with a test dose of 20 mg, then after 7 days 20–40 mg repeated 3- to 4-weekly according to the response. The usual maintenance dose is 50 mg every 4 weeks to 300 mg every 2 weeks. Side effects include extrapyramidal signs (Parkinsonism, dystonia, restlessness, and tardive dyskinesia), hypotension and less commonly neuroleptic malignant syndrome (hyperthermia, fluctuating consciousness, muscle rigidity and autonomic dysfunction).

Post-traumatic stress disorder

Introduction

Post-traumatic stress can occur after any natural (earth quakes, tsunamis etc) or man-made (wars, riots etc) disasters, following sexual/physical/ emotional abuse, domestic violence, separation from parents etc. Nearly all children and adolescents who have experienced catastrophic situations where there has been a perceived or true risk to life will initially display symptoms of psychological distress, including intrusive flashbacks of the stress event, nightmares, withdrawal and inability to concentrate, among others. These symptoms are a common and normal reaction to an abnormal event and occur in the first few days or weeks after an event and most resolve spontaneously. Most children and adolescents will regain normal functioning once their basic survival needs are met, safety and security have been restored, and developmental opportunities have been regained, within the social, family and community context.

Post-traumatic stress disorder (PTSD) is a well recognised now and describes ongoing and intrusive symptoms beyond a month after the event and occurs in around 25–35% of those exposed to traumatic events. Individual differences in response to trauma depend on the following:

- stressor severity and degree of exposure to the stressor
- exposure to previous traumatic events
- the child's perception of the event

- the child's appraisal of the threat to their survival, and the 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 impairment in educational functioning can all occur.

Diagnostic criteria

- The child has experienced an event that is outside the range of normal experience and that is (or perceived to be) life-threatening to them or to those close to them. 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, and a range of signs of physiological arousal occur, such as difficulty in sleeping, irritability, or poor concentration.

In younger children, repetitive play related to the trauma may be present. They may have frightening dreams that have no obvious content and may regress in their development.

Assessment

It must first be established that the child has experienced a traumatic event that preceded the onset of the symptoms. The traumatic event may not necessarily lead to development of PTSD. Instead, the child may develop acute stress disorder or sadness.

When assessing the child, the interviewer will need to take account of the child's maturation, and their verbal facility and functioning. Details of the traumatic event, the child's perception of the event, and their response immediately and later, should be evaluated.

The differential diagnoses include obsessive-compulsive disorder, schizophrenia, and 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.

Management of trauma-affected children

If possible, reuniting the child with their parents or other close relatives and restoring normal comforting routines is helpful. Some children will require more specialised interventions to address their suffering and help to restore their flow of development. Immediately after traumatic events, activities and opportunities that allow children to talk about or otherwise express painful experiences and feelings (e.g., by physical and artistic expression) are most beneficial if facilitated by people whom the children know and trust and have continued contact with.

The goals of psychological 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 help 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.

Trauma counselling should never be provided unless an appropriate and sustained follow-up mechanism is guaranteed.

The psychosocial well-being of adults, particularly parents and caregivers, has a direct impact on that of children, and should therefore be addressed through concurrent parent-focused psychosocial interventions. The participation of children, and adults, in decisions that

affect their lives has a positive effect on their mental health, empowers them, and helps them to regain control over their own lives.

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 such attacks is to explain the physiological features of panic to the child and their family, and to teach proper breathing techniques (namely to breathe slowly at a rate normal for the child's age).

Preventive intervention

The promotion of mental health through a healthy lifestyle brought about by health education and life skills training has the potential to equip a young person for their journey through life. There is material available in the public domain on life skills training (e.g., on the WHO/UNICEF websites) which has been used in many resource-limited and middle- income countries.

Autism Spectrum Disorders (ASD)

This is a group of disorders with similar features, although an individual child may not display all of them, and the severity may vary. Autism becomes evident before 3 years of age, but children with other conditions that form part of the autistic spectrum may present later (e.g., at school age). Autistic behaviour may occur as an isolated problem, or it may be a component of a number of childhood developmental disorders. It is important to consider these when assessing a child, in case intervention may help. Sensory deficits, particularly deafness, which may be hard to identify in a young child, are especially important in this respect. Children with extreme prematurity, hydrocephalus, metabolic disorders (e.g., phenylketonuria), hypothyroidism, fetal alcohol syndrome, tuberous sclerosis, neurofibromatosis, Down's syndrome and other chromosomal disorders may exhibit features of autism.

Autistic children characteristically have difficulties with the following:

- social interaction and reciprocity
- language development and communication skills
- imagination and play
- rigid thinking
- restrictive and repetitive stereotypical patterns of behaviour, activities, and interests.

Social interaction

Children with ASD may make little or no eye contact. They may not be able to share experiences or to understand the feelings of others, or recognise clues to their feelings from their behaviour (e.g., that people who are crying are sad).

Girls with ASD often have better social skills but struggle in the other areas. They are therefore less likely to be picked up and diagnosed but have significant challenges at school and beyond.

Language and communication

Young children with autism may not point to get attention or to show another person something. They may never develop any useful language, or they may have unusual speech with abnormal intonation, jumbled words, incomprehensible sounds, or repetition of the same words again and again (echolalia).

Cognitive function

Children with autism may have global cognitive impairment or general impairment but considerable skill in some areas (e.g., numbers, art).

Rigid thinking and ritualistic behaviour

Children with ASD often have very structured repetitive play (e.g. organising objects in a certain pattern). They may persist much longer than usual in putting things in their mouths, or they may hold on to objects, moving and feeling them in their hands, for long periods. Routine is very important, and they are often very upset by any disruption, which may lead to outbursts of temper.

Management

There is no cure yet for ASD. Management focuses on encouraging the child to learn as much as possible and to develop behaviour that helps him or her to live happily within the family and community. Support for the family in caring for these children, who can be very challenging, is essential. Education for the family and community about the difficulties of autistic children, so that their behaviour is not misinterpreted as naughtiness, or caregivers criticised inappropriately, is also very important. Healthcare professionals and teachers experienced in the care of children with autism can give a great deal of assistance to families.

Vigilance for other problems, especially with hearing or vision, which if undiagnosed will add to the child's difficulties, should be maintained.

For some children with autism who have severe aggressive behaviour and only under expert supervision consider risperidone:

- **Child over 5 years and up to 50 kg:** 250 micrograms once daily increased, if necessary, in 250 microgram steps on alternate days. Usual dose is 500 micrograms once daily, maximum 750 micrograms per day
- **Child over 5 years and over 50 kg:** 500 micrograms daily increased, if necessary, in 500 microgram steps on alternate days. Usual dose is 1mg once daily, maximum 1.5mg per day

Review effectiveness and side effects after 3–4 weeks; stop if no response at 6 weeks.

Asperger's syndrome

Asperger's syndrome is within the Autistic Spectrum Disorders, in that affected individuals also have difficulties with social interaction, especially in understanding the usual patterns of social behaviour of their community. They often develop very deep and detailed knowledge about subjects that interest them, and become experts who can contribute to society, but they may also become isolated if others do not share or understand their interests.

Asperger's syndrome is usually identified later than autism, often when children are at school, and are recognised as different from their peers. They may be bullied and very lonely, as they long to have friends and 'fit in', but do not have the social skills to enable them to do so.

There is no cure for Asperger's syndrome but teaching from as early an age as possible about appropriate behaviour can help these children. Education for their families and communities, so that they understand that the child has a condition which makes it hard for him or her to pick up social clues, and is not just being difficult, along with appreciation of any special talents, is essential. If it is acceptable to the child or adolescent, written information to give to people to explain what they find hard may be useful.

Attention deficit hyperactivity disorder (ADHD)

Children with ADHD characteristically have difficulties with the following:

- inattention
- hyperactivity
- impulsivity

These features must be present before the age of 6–7 years, evident in more than one situation (e.g., at home and at school) and interfere with the child's social or educational functioning.

These characteristics may persist into adult life, resulting in inattentive and disorganised or impulsive risk-taking behaviour.

Inattention

Children with ADHD cannot concentrate for very long, especially on tasks they have been given which have no immediate reward (e.g., schoolwork).

Hyperactivity

These children are always on the move. Young children with ADHD may run, jump, climb, make a lot of noise, and never settle to anything. School-age children and adolescents have difficulty sitting still, and may be constantly tapping their feet, wriggling, and fidgeting.

Girls with ADHD often have less hyperactivity and so more likely to go undiagnosed as do not disrupt but their inattention results in poor educational performance.

Impulsivity

Children with ADHD do not think before they act. They may have accidents or get into trouble for recklessness.

Other problems

Children with ADHD can be exhausting. They are often in trouble because they are so active and may have poor relationships with other children and adults, and low self-esteem. They may sleep badly, struggling to get to sleep or waking frequently, and be poor eaters because they cannot sit still for long enough to finish a meal.

Causes

There is no known cause of ADHD. Some of the contributory factors include the following:

- genetic: parents or siblings affected
- living conditions: ADHD is more common in children from disadvantaged backgrounds
- depression in the child's mother
- child abuse

These above factors may all interact.

In addition to occurring alone, the features of ADHD may be seen in young people with neurological conditions such as head injuries, fetal alcohol syndrome, encephalitis and meningitis, epilepsy, hypothyroidism, and some syndromes, such as fragile X syndrome, Williams' syndrome and tuberous sclerosis.

Comorbidities

Children with ADHD often have additional problems, such as mild cognitive impairment, delayed language development, poor coordination, reading difficulties and mood disorders.

Assessment

- Developmental history, especially any delay and when noted.
- Pregnancy and birth, including any exposure to drugs or alcohol.
- Family history of similar problems, maternal depression and social circumstances.
- Educational progress: both intelligent children and children with specific learning difficulties who are bored can be disruptive.
- The parents' expectations of the child and their response to his or her behaviour.
- Medical history and examination for neurological problems, including any medication being taken by the child that might affect his or her behaviour.
- Careful observation of the child in several settings, including home and school.

Management

- There is no cure for ADHD, but careful management can help these children and their families a great deal.
- Look for any health problems that might be contributing to the condition and could be treated (e.g., large tonsils and adenoids preventing undisturbed sleep).
- Explain to the child and their family that they have a disorder that affects their behaviour, and that they are not just a naughty child.
- The most useful management is behavioural.
- Drugs may be helpful in severe cases.
- Ensure child protection issues are identified if present

Behavioural management

Children with ADHD do best where there are as few distractions as possible around them, and where there are clear rules about the conduct that is expected, with praise or reproof given immediately if merited. When doing tasks such as schoolwork, they are best on their own or in a small group, sitting near to the person in charge. They often have low self-esteem. Giving praise when they do well, with frequent small rewards, is very helpful.

Drugs

The most widely used drugs for ADHD are stimulants such as methylphenidate (Ritalin), which can be given to children aged over 6 years in a dose of 5 mg once or twice daily, increasing by 2.5–5 mg weekly up to a maximum daily dose of 60 mg. If methylphenidate has not made any difference after 3 weeks at full dose it should be stopped.

Slow-release preparations are also available and can be given less frequently. If effect wears off in the evening (with rebound hyperactivity) a dose at bedtime may be appropriate (establish need with trial bedtime dose). Careful supervision is needed, and ideally children who require these drugs should be treated by a professional experienced in their use.

Additional reading

Valsamma Eapen, Philip Graham, Shoba Srinath. (2012) Where There Is No Child Psychiatrist- A mental healthcare manual. The Royal College of Psychiatrists.

Section 23. Eating disorders in children and adolescents

Background

An eating disorder means having an unhealthy relationship with food. The most common types of eating disorders are anorexia nervosa, bulimia nervosa and binge eating disorder. With anorexia nervosa the young person is not eating enough food or is exercising too much resulting in weight loss or insufficient weight gain as expected. With bulimia nervosa the young person goes through periods of bingeing (eating a much larger than usual amount of food in a short period of time) followed by trying to get rid of the calories, for example by purging (inducing self-vomiting) or abuse of medication such as laxatives or by over-exercising. With binge eating disorder, the young person also binges, often in secret, but there are no compensatory behaviours. It is estimated that 9% of people will have an eating disorder at some stage in their life of which the majority will be female. The onset of eating disorders is often in adolescence and particularly anorexia nervosa results in significant morbidity and mortality.

It is important to identify young people with an eating disorder at an early stage as prompt treatment improves outcome and prevents young person developing a protracted eating disorder. Preferably, the young person and family are supported by a team of professionals with expertise in management of eating disorders consisting of psychiatrist/psychologist or other mental health specialist, family doctor/paediatrician and dietician.

Assessment

As starting point for assessment, the SCOFF questions¹ can be helpful:

Do you make yourself **S**ick because you feel uncomfortably full?

Do you worry you have lost **C**ontrol" over how much you eat?

Have you recently lost more than **O**ne stone (14 pounds 6.3Kg in a 3-month period)?

Do you believe yourself to be **F**at when others say you are too thin?

Would you say that **F**ood dominates your life?

*One point for every "yes"; a score of ≥ 2 indicates a likely case of anorexia nervosa or bulimia

Explore current eating behaviours, asking if eating has become restrictive or changed. Does it seem appropriate considering young person's age and activity levels? Ask if there is restricting of fluid intake, excessive exercise, or compensatory behaviours such as vomiting or laxative abuse. Social withdrawal or comorbidity such as anxiety, low mood and Obsessive-Compulsive Disorder (OCD) are common and should also be explored.

Ask about physical symptoms including dizziness, fainting, and feeling cold. In girls, periods may have stopped, become irregular or lighter or not started at an age when this would have been expected.

Weight for height should be calculated to assess if the young person is underweight but as important is to get indication of weight loss: (Section 65). Particularly girls can present with appropriate weight for height but lose in a short period significant amounts of weight secondary to losing control of their eating and / or over exercising.

Check for physical signs of malnutrition including poor circulation, bradycardia, and low blood pressure. Ideally, ECG should be done to confirm normal sinus rhythm. Biochemical abnormalities can be noted with purging but are unusual with other eating disorders and normal biochemical parameters are not indicator of low risk.

Muscular weakness is a late sign and indicates high risk. This is assessed by asking the young person to get up from squat to standing or from lying down flat to sitting. If the young person needs to use their hands this test is positive.

Treatment

First step in treatment of a young person with an eating disorder is physical risk assessment with stabilisation as required. This may need to be in hospital, particularly when there are significant symptoms such as fainting, muscle weakness, bradycardia, or biochemical abnormalities. A hospital admission may also be required when, despite best efforts from the family and team, the young person is unable to change their eating pattern at home.

Management, at home or as an inpatient, starts with psycho education and establishing regular intake. The aim of psycho education is to inform the young person and family about the eating disorder, balanced nutrition, and effects of malnutrition. A standard meal plan should include three meals and three snacks (breakfast – snack – lunch – snack - evening meal – snack) and aiming for fluid intake of 1.5-2 L per day. The size of the meals or snacks should be decided by clinician or family members: The young person may not be able to make the right choices, going for the lower calorie options.

A useful starting point is that the young person should have same size portion as her mother or his father.

Refeeding syndrome: Initiation of feeding in high-risk patients may result in fluid retention and an electrolyte shift which is insulin mediated and called refeeding syndrome. It is rare but can result in hypophosphatemia, hypotension, oedema, and cardiac arrhythmias. It is associated with significant mortality. Those most at risk are when there has been rapid weight loss (more than 1 kg per week for the last couple of weeks or more than 15% of total weight in the last 3-6 months), when the young person has low weight for height (less than 80% median weight for height: Section 65) or there are biochemical abnormalities.

To reduce the risk of developing refeeding syndrome, those deemed to be at high risk should be started on half of their meal plan with gradual increase to full meal plan over a 5-to-7-day period. The young person should be weighed daily, preferably after morning toilet visit and before breakfast. Blood pressure and heart rate should be checked regularly, and biochemistry should be done daily at least for one week period.

Refeeding syndrome tends to develop in the 1st 5 days but can occur up to two weeks after feeding is initiated. Commonly, the 1st biochemical abnormality noted is a drop in phosphate levels. Management of refeeding syndrome is treating the biochemical abnormality, for example by giving oral phosphate supplementation, and with only further increase in the meal plan until all parameters have stabilised. **Please note**, the risk of under-feeding is equally detrimental, and it is not good practice to refeed very slowly in aim to avoid refeeding syndrome.

For eating disorders in young people, the family will play an important role in helping the person to recover. They will need to be educated on eating disorders, including that although the young person may have chosen to eat more healthily or do more exercise, this now no longer is a choice-of but an illness. The family needs to be shown what the meal plan includes and how to support mealtimes.

The young person needs to be educated on healthy eating, healthy body weight and topics such as self-esteem, body image concerns and social skills may need to be addressed. Underlying co-morbidities, such as depression, OCD, anxiety will need to be treated once weight is restored.

For bulimia nervosa, it furthermore requires educating on the adverse effects of attempting to control weight with, for example, vomiting. When there is binge eating disorder, explore triggers for binge eating and advise that avoiding feeling hungry by adhering to the standard meal plan will reduce risk of bingeing.

Most young people will achieve full recovery with timely, specialist treatment however some will experience relapse. For this reason, the last stage of treatment focuses on relapse prevention, i.e., resilience building and developing coping strategies in stressful situations.

Further reading:

The SCOFF questionnaire: assessment of a new screening tool for eating disorders
<https://www.bmj.com/content/319/7223/1467> Accessed 6.4.2021

Junior Marsipan: <https://www.rcpsych.ac.uk/docs/default-source/improving-care/better-mh-policy/college-reports/college-report-cr168.pdf> Accessed 6.4.2021

NICE guidance: <https://www.nice.org.uk/guidance/ng69/resources/eating-disorders-recognition-and-treatment-pdf-1837582159813> Accessed 6.4.2021

Section 24. Muscular dystrophy

Introduction

The muscular dystrophies are 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 2 years but will have very high serum creatine kinase levels. They usually present between 2 and 5 years of age with delayed walking, frequent falls, and difficulty in climbing stairs and in getting up from the floor. The weakness affects proximal more than distal muscles, and the pelvic girdle more than the shoulder girdle. The facial muscles are unaffected. Prominent calves and thighs are characteristic. With time, these children 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 and 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 leads, deep Q waves in left-sided leads, and inverted T waves in most patients.

Diagnosis

The serum creatine kinase activity is very high (100 times normal). The electromyogram (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 (PCR) 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 degrees may delay shortening of the tendo-achilles.

When walking is becoming difficult, the fitting of lightweight callipers and intensive physiotherapy may keep the child ambulant for a few more years. Once the child is 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 of age a wheelchair will be 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 child's quality of life (see Section 14 and Section 91 Handbook 1). Death occurs in the twenties from respiratory or cardiac failure.

Genetic counselling

An elevated creatine kinase activity (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 and 10 years of age, and ambulation is maintained beyond 15 years and often into adulthood. This disorder is caused by partial deficiency of dystrophin.

Limb girdle muscular dystrophies

These are rare and vary in severity. A severe form is prevalent in North Africa. It has a clinical picture similar to Duchenne muscular dystrophy but affects boys and girls and has more prominent involvement of the shoulder girdle muscles, with winging of the scapulae. 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. Genetic tests for diagnosis are available in specialised laboratories.

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 an open mouth. There is no ophthalmoplegia. There are several subgroups, some with eye or brain abnormalities and demyelination. Merosin is deficient in one subgroup. Inheritance is autosomal recessive, and the gene defect is known for most of them. The creatine kinase may be elevated or normal, and the diagnosis is made by muscle biopsy or genetic testing. Management is supportive. Congenital muscular dystrophy must be distinguished from other causes of hypotonia (i.e. other congenital myopathies, infantile spinal muscular atrophy and organic acidaemias).

Section 25. Neuropathies including Guillain–Barré syndrome

Neuropathies

Introduction

Neuropathies are diseases that affect the anterior horn cells and/or the peripheral nerves.

Anterior horn cell disease

The most common neuropathies are:

- poliomyelitis (see Section 43)
- spinal muscular atrophy.

Spinal muscular atrophy

This is a motor neuron disease of the spinal cord and brainstem, inherited as an autosomal-recessive disorder and associated with deletions of the survival motor neuron (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 the lower limbs more than the upper ones. The children are areflexic, and fasciculation of the tongue is diagnostic (observed with the tongue at rest in the mouth). There are three clinical subtypes based on severity.

1. 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.
2. 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 it is absent, these children can survive into adulthood.
3. Mild type (also known as Kugelberg–Welander type). The onset is later, and these children can walk, but they do so late and with difficulty. Respiratory and bulbar weakness is not usually present. A coarse tremor of the hands is frequently seen in this and the intermediate form. This is a useful sign for distinguishing this type from muscular dystrophy, with which it is often confused (see Table 25.1).

Diagnosis

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 (PCR). Blood (2–5 mL in an EDTA tube), or DNA extracted from it, can be sent by post to a laboratory that will perform the test and confirm the diagnosis within a few days.

TABLE 25.1 Comparison of spinal muscular atrophy (mild type) and Duchenne muscular dystrophy

Feature	Spinal muscular atrophy	Duchenne muscular dystrophy
Motor milestones	Delayed	Normal or delayed
Hypotonia	+++	+/-
Pseudohypertrophy	+/-	+++

Feature	Spinal muscular atrophy	Duchenne muscular dystrophy
Hand tremor	+++	–
Tongue fasciculation	+	–
IQ	Normal	Normal or low
ECG	Baseline tremor	R- and Q-wave changes
Creatine kinase	Normal or slightly raised ($\times 2$)	Very high ($\times 100$)
EMG	Chronic denervation	Myopathic
Muscle biopsy	Denervation	Dystrophic

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 callipers for as long as possible. If the child is confined to a wheelchair, a brace may be needed to control the scoliosis. Surgery (fusion of the spine) may be necessary. Children with symptoms of nocturnal hypoventilation (disturbed sleep, headaches, daytime drowsiness and poor concentration) may benefit from assisted non-invasive night-time ventilation with a nasal mask (see Section 91 Handbook 1).

Gene replacement therapy (Zolgensma) for pre-symptomatic and symptomatic cases of the severe infantile form of spinal muscular atrophy was recently approved in the USA. Many infants who received this treatment achieved motor milestones and survived beyond 2 years without respiratory support which they would otherwise not have achieved. The cost however is prohibitive (\$2million for a once off intravenous infusion)

Peripheral neuropathy

The two commonest causes of peripheral neuropathy in children are Guillain–Barré syndrome (see below) and hereditary motor and sensory neuropathy.

Hereditary motor and sensory neuropathy

This is the commonest chronic peripheral neuropathy in children. It is progressive, and 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 develops. 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 the finding of very low motor conduction velocities in both the child and one of the parents, indicating demyelination. Type II is similar, but rare, and shows axonal rather than demyelinating changes in nerve conduction studies. There are no treatments for these diseases other than special boots and ankle orthoses to stabilise the ankle.

Other peripheral neuropathies

These include leukodystrophies (where peripheral nerve demyelination occurs as part of CNS demyelination), toxic neuropathy (due to glue or benzene sniffing, lead, or drugs) and diphtheria (see Section 19 Handbook 1).

Guillain–Barré syndrome

Introduction

This is the commonest peripheral neuropathy seen in childhood. It is a demyelinating neuropathy induced by an autoimmune process that is precipitated by a preceding viral or other infection. It has a peak incidence at around 8 to 9 years of age in well-resourced countries and 3 to 4 years of age in resource-limited ones, possibly due to overcrowding. Rarely, an acute axonal form occurs, especially in some countries, such as China.

Clinical features

The onset is usually acute. There is often a history of a preceding upper respiratory or gastrointestinal tract infection and insidious sensory symptoms (e.g., muscle tenderness, occasionally an unsteady gait, and frequent falls). The weakness starts in the lower limbs and ascends to affect the trunk, upper limbs, and the 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 frequently absent. Sensory loss is minimal and of the 'glove-and-stocking' distribution. Ophthalmoplegia, papilloedema and bladder involvement rarely occur. 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

Guillain–Barré syndrome may evolve into chronic inflammatory demyelinating polyradiculoneuropathy. This disease is similar to Guillain–Barré syndrome, consists of progressive or relapsing motor and sensory dysfunction, and lasts at least 2 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 of Guillain–Barré syndrome

The diagnosis is confirmed by an abnormal nerve conduction stimulation, but a high CSF protein level and almost normal cell count are very suggestive. These findings are usually present after the first week of onset. Other causes of acute flaccid paralysis such as poliomyelitis in endemic countries need to be considered (see Table 25.2.).

Management of Guillain–Barré syndrome

Supportive care is the cornerstone of successful management of the acute patient. Of greatest concern is respiratory failure due to paralysis of the diaphragm (the muscle that is most important for breathing). **Intubation** may be needed if there is evidence of impending respiratory failure. The following steps in management should be taken:

- Admit the child to hospital to monitor for impending respiratory and bulbar paralysis and autonomic dysfunction.
- Measure the respiratory rate, heart rate and blood pressure, perform pulse oximetry and, if possible, measure vital capacity (or peak flow), and check airway protection frequently. Blood gas analysis may be helpful (ideally transcutaneous or expired carbon dioxide levels)
- If the vital capacity is less than 50% of normal for age **and/or there is significant respiratory failure with hypoxaemia and hypercapnia**, ventilate the child if possible.
- If bulbar and respiratory paralysis occurs, airway protection, tube feeding and ventilatory support will be necessary. Airway protection can be achieved by intubation or tracheostomy.
- Plasma exchange (also called plasmapheresis) and high-dose immunoglobulin therapy may lessen the severity of the illness and accelerate recovery in some patients but are not widely available. These two treatments are equally effective, and a combination of the two is not significantly better than either alone. However, immunoglobulin is easier to administer.
- Children who require ventilation can be given high-dose human immune globulin if available (0.4 grams/kg IV over 6 hours daily for 5 days). This can be repeated if there is no response, if deterioration occurs or if there is a relapse.

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, and around 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 level of > 800 mg/litre in the first week of the disease. The prognosis is generally better in children than in adults

TABLE 25.2 Other causes and features of acute flaccid paralysis

Cause	Features
<i>Spinal Cord</i>	
Poliomyelitis	Preceding fever, headache and meningeal irritation, 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
<i>Neuropathies</i>	
Guillain–Barré syndrome	Symmetric, areflexia, ascending weakness
Diphtheria	Preceding history of diphtheria 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 levels Resolves quickly once tick has been removed
<i>Metabolic</i>	
Acute intermittent porphyria Hereditary tyrosinaemia	Family history, other symptoms
<i>Muscle</i>	
Myasthenia gravis (rare but treatable)	Fluctuating weakness that is worsened by activity and better after rest Tensilon test is positive
Acute viral myositis	Tender muscles, limp, fever, elevated muscle enzymes (creatine phosphokinase or aldolase)
<i>Other causes</i>	
Organophosphate poisoning	History of exposure, excessive salivation, twitching of muscles, meiosis, tachycardia

Section 26. Sickle-cell disease

Introduction

Sickle cell disease (SCD) was first reported in medical literature in 1910 by James B. Herrick, an American doctor who described it as a peculiar elongated and sickle shaped red blood corpuscle in a patient with severe anaemia. Since this discovery, research has improved the knowledge about sickle cell disease tremendously. SCD is an inherited lifelong illness of the red blood cell and characterized by changes in shape, consistency, and adhesiveness of the red blood cells from a round, smooth, soft disk shape into a crescent moon or sickle shape.

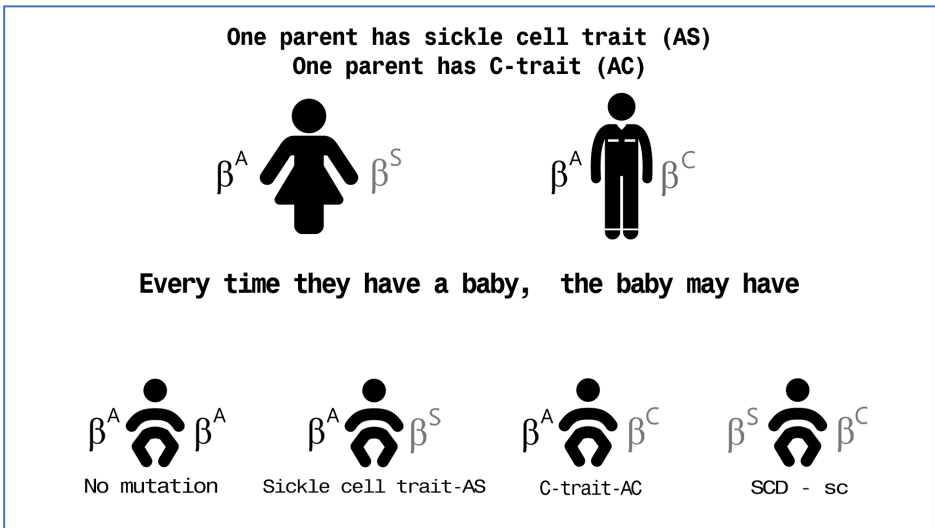
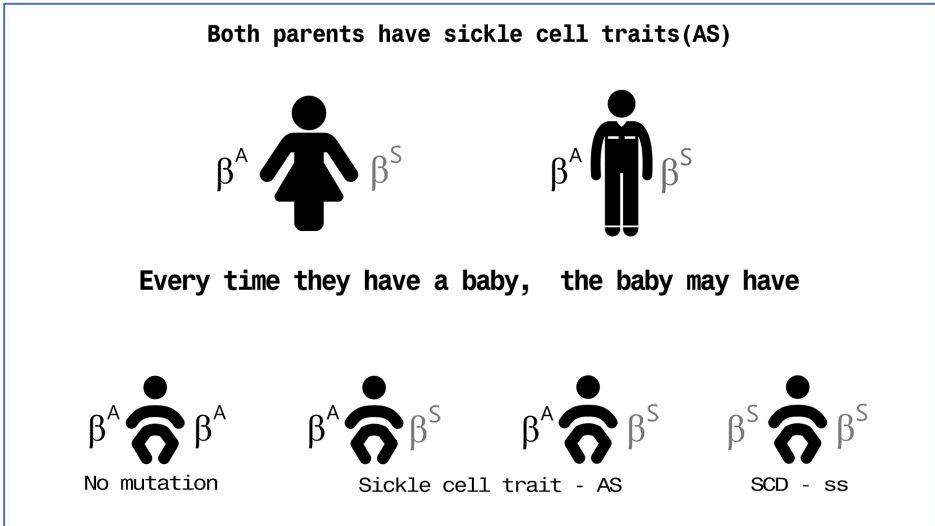
SCD affects millions of people throughout the world and is particularly common in Sub Saharan Africa (SSA), Mediterranean, Middle East and India. It can also be found in the Caribbean, North America and in Western Europe. Worldwide estimates indicate that about 20-25million people are affected by SCD with approximately 60% living in SSA. The global number of newborn infants with SCD is estimated to increase to about 400,000/year by 2050 with 85% expected to be born in Sub Saharan Africa.

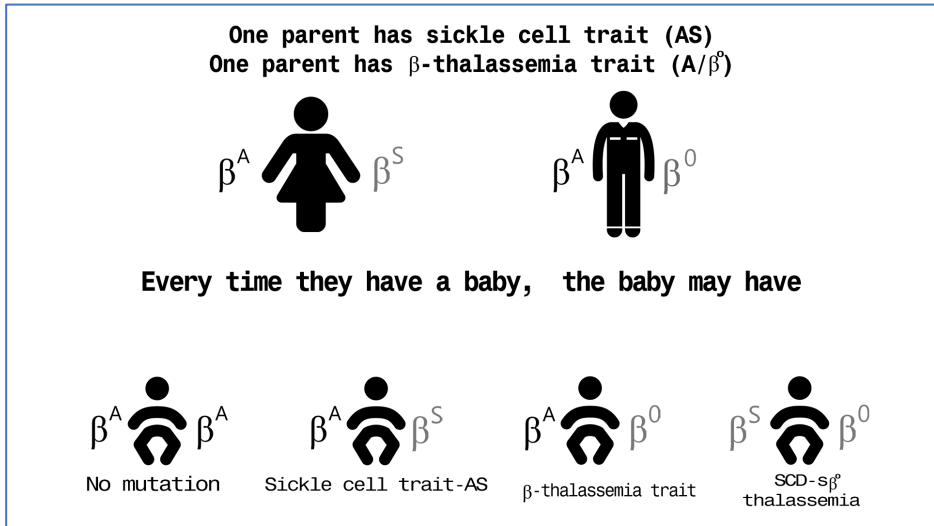
Genetic basis Sickle-cell disease is an autosomal recessively inherited disorder of haemoglobin synthesis which implies that both copies of the gene in each cell have the mutation. The genetic abnormality underlying the HbS gene is a point mutation at position 6 on chromosome 11 resulting in the substitution of valine for glutamic acid on the beta-globin chain. Those affected inherit two copies of the altered beta-globin gene and are therefore homozygous for HbS (HbSS) and also called sickle cell anaemia.

Alternatively, a single HbS may be inherited with another beta-chain mutation-Hb C, which occurs as a result of the substitution of glutamic acid with lysine on position 6 of the beta globin chain (HbSC). Beta globin gene mutations can also lead to beta thalassaemia. The gene mutation results in reduced(β^+) or no production of the beta globin subunits(β^0). Beta thalassaemia can also be inherited with HbS resulting in HbS β^{++} or HbS β^0 , disease. Other types of SCD that can occur include HbSD, HbSHPFH and HbSE.

A child who inherits two abnormal haemoglobin genes that are capable (in combination) of giving the sickle phenotype, will have their disease detectable on blood tests at birth, though symptoms usually evolve once the HbF recedes. A child whose parents both have the HbS trait has a 25% chance of receiving two HbS genes and developing the disease, a 50% chance of also having HbS trait, and 25% chance of being unaffected. These probabilities occur with each pregnancy that the couple will have. Most patients with HbS trait lead completely normal healthy lives.

Figure 26.1 Inheritance of Sickle Cell Disease





The HbS mutation is common. It is estimated that up to 5% of the world population are healthy carriers of sickle gene, and this can rise to 25% in West Africans. Although the HbS mutation is most common in Africa, it occurs widely across many groups. It is estimated that each year 300 000 children are born with homozygous sickle-cell disease worldwide.

Prognosis

In well-resourced countries, the life expectancy of individuals with sickle-cell disease has been continuously improving, and historic data suggest that it is now well beyond the sixth decade of life, with the overwhelming majority of children surviving into adulthood. As people age, organ damage accumulates resulting in chronic disease. Patients still largely die of their condition.

However, in resource-limited countries, sickle-cell disease is still associated with a very high mortality and morbidity, particularly during childhood. Sickle-cell disease remains a major cause of mortality in children under 5 years of age, with estimates as high as 50–90% in some rural areas of Africa. The major causes of death are infection, especially malaria and invasive pneumococcal infection, and severe anaemia, sometimes associated with malaria. For those who live with the condition, sickle-cell disease may be the cause of a great burden of suffering for those affected and their families.

Pathogenesis

The predominant clinical manifestations of sickle-cell disease are due to vaso-occlusion and chronic haemolysis, often in response to triggers such as illness, acidosis, vasoconstriction (cold), hypoxia or dehydration.

The presence of abnormal HbS leads to the production of a haemoglobin tetramer ($\alpha_2/\beta S_2$) that is poorly soluble when deoxygenated and polymerises readily into a rope-like fibre within the red blood cell. This leads to red cell distortion into the classic sickle shape, a reduction in red cell deformability, and increased red cell destruction through haemolysis, with consequent shortening of the red cell lifespan from 120 days to 10-20 days, and anaemia.

Vaso-occlusive episodes occur when blood vessels become occluded with sickle cells, causing pain, tissue oxygen deprivation, and progressive organ damage alongside altered cell adhesion and abnormal erythrocyte–endothelium interaction. In addition to vaso-occlusion, there is a chronic intravascular haemolysis leading to a compensated anaemia

Section 26. Sickle-cell disease [Dr. Sara Trompeter](#), [Dr. Vivian Paintsil](#), [Dr. Simon Parke](#) with functional nitric oxide (NO) dysregulation with chronic vascular endothelial damage. This means that polymerisation alone does not account for the pathophysiology of sickle-cell disease. Changes in red cell membrane structure and function, disordered cell volume control, and increased adherence to vascular endothelium also play an important role and much of the chronic organ damage is due to the vasculopathy that results.

Clinical presentations include the following:

In children, the most common presentation of sickle-cell disease is with an acute crisis, usually as a painful episode. More recently, as more countries adopt a newborn screening programme, children may be diagnosed with sickle-cell disease before their first crisis. Presentation includes the following:

- a painful vaso-occlusive crisis such as dactylitis (swelling of the hands and feet), acute vaso-occlusive pain crisis, priapism
- acute chest syndrome (ACS)
- stroke.
- severe anaemia
- infection and overwhelming sepsis

Newborn screening programmes

The goal of any newborn screening programme for sickle-cell disease is to identify affected children as early as possible and thus reduce the morbidity and mortality of sickle-cell disease, especially from bacterial infections, through the early introduction of antibiotic prophylaxis, immunization, education and access to specialist care. In well-resourced countries, the preferred option is the universal screening programme rather than selective screening of high-risk infants only.

Methodology of newborn screening

The methodology of screening can vary, but in principle involves the collection of a dried neonatal blood spot sample for transport and testing by thin-layer isoelectric focusing, or high-performance liquid chromatography (HPLC). Two tests using different techniques need to be performed for a diagnostic sample as there are over 200 different abnormal haemoglobins, some with overlapping results on a single technique.

Table 26.1 Results and patterns in the newborn period

Finding	Pattern (from most to least amount of haemoglobin)	Interpretation
HbF and HbA	FA	Normal baby or beta thalassaemia trait
HbF, HbA and HbS	FAS	Sickle-cell trait
HbF, HbS and HbA	FSA	Sickle-cell β+thalassaemia
HbF and HbS	FS	Sickle-cell disease (HbSS or HbS β0 thalassaemia)
HbF HbS and HbC	FSC	Sickle cell disease (HbSC)

Following the initial heel prick test, a second confirmatory test may be taken 1–2 weeks later for repeat testing by one of the diagnostic tests below by a reference laboratory. The tests used must have the capability to distinguish between HbF, HbS, HbA and HbC.

Diagnosis

- Historically, haemoglobin electrophoresis was the standard method for separating HbS from other haemoglobin variants and is relatively cheap though labour intensive and remains the commonest technique in resource poor laboratories
- High-performance liquid chromatography (HPLC) and column electrophoresis are precise and fully automated techniques for identification and quantification of haemoglobins and are the main diagnostic methods in well-resourced laboratories.
- Sickle solubility test. A positive sickle solubility test will detect the presence of HbS but will not identify whether the person is a carrier or has sickle-cell disease, cannot differentiate between a homozygote or compound heterozygote and also can be negative if the patient is heavily transfused. It is often used as a screening test or to confirm the findings of one of the two tests above.
- Isoelectric focusing is the technique often used in the newborn blood spot
- Mass spectrometry can be used but is not frequently used in this setting.
- Genetic tests can also be used and may well be the future as fast throughput cost efficient genotyping evolves.
- Point of Care test for Sickle Cell Disease are rapid, portable and inexpensive blood test with the added advantage of immediate feedback to the patient. It is thus helpful in emergency situations in resource constrained settings.
- Blood film: Examination of the peripheral blood shows sickled erythrocytes and then additional features depending on the genotype e.g. target cells and microcytosis in HbS β thalassaemia and target cells and crystals in HbSC

General principles of the management of an acute sickle crisis

Most children do not develop symptomatic disease in the first few months of life until adult haemoglobin production is established. At birth, fetal haemoglobin which is not affected by the sickle mutation, makes up 60 - 90% of the haemoglobin in the RBCs of the infants. The foetal haemoglobin does not rely on beta globin which can have the sickle mutation whereas adult haemoglobin does. The principles of managing any acute crisis are based on searching for, and actively treating, any precipitants (see Table 26.2.), good effective management of pain and monitoring for the evolution of acute complications. Any child presenting with an acute crisis should be considered at risk of sudden and life-threatening deterioration, and clinicians are advised to have an anticipatory approach. Crises can be unprovoked, but can be precipitated by Infection, dehydration, and extremes of temperature.

TABLE 26.2. Precipitants of sickle crises

Problem/precipitant	Approach
Fever and/or evidence of infection Child should be considered functionally asplenic and immunocompromised	Treatment dose of appropriate antibiotics
	Use of appropriate antimalarial drugs
	Use of antipyretic drugs
Dehydration	Rehydration
Extremes of temperature and cold	Warmth and rest

Clinicians should be alert to signs suggesting the possibility of a sudden acute deterioration during a crisis. The following trigger list may be helpful for identifying children at increased risk of sudden or rapid deterioration:

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- uncontrolled pain despite strong opiate analgesia
- increasing pallor, breathlessness, or exhaustion
- marked fever ($> 38^{\circ}\text{C}$)
- significant tachycardia, tachypnoea or hypotension
- chest pain with or without signs of consolidation
- A drop in SpO_2 of more than 4% or an SpO_2 below 94%
- abdominal pain with or without distension
- severe diarrhoea and vomiting
- sudden profound pallor with or without jaundice
- parents reporting a rapidly enlarging spleen
- any abnormal neurological signs, including painless loss of function of a limb, headache and seizures
- malaria.

TABLE 26.3. Management of an uncomplicated acute painful episode

Treatment	Comment
Antibiotics and antimalarial drugs (see later)	Any fever should prompt the search for infection and active treatment. Antibiotics should be administered empirically.
Hydration	Dehydration occurs readily in children with sickle-cell disease, due to impairment of renal concentrating power. Fluids should be given at 150% maintenance (orally, NG or IV) to achieve euvolaemia (normal circulating volume)
Analgesia	<p>Assess pain using an age-appropriate pain scale (see below).</p> <p>Use the appropriate pain scale to assess response to analgesia with the goal of minimal pain allowing successful mobilisation.</p> <p>Manage pain with prompt administration of the most appropriate choice and dose from the analgesic ladder.</p> <p>Take into account previous drugs and dosages given at home.</p> <p>Children in severe pain may need early use of opiates or paracetamol orally or IV. Do not use pethidine nor inhaled nitric oxide as repeated use can cause profound toxicity.</p> <p>Inadequately controlled pain leads to hypoventilation and acute chest syndrome.</p>
Severe anaemia	Consider transfusion if the haemoglobin concentration is very low (e.g. < 50 grams/L) or has fallen by > 20 grams/L from a known baseline level, or the child is clearly clinically compromised

Treatment	Comment
Oxygen and incentive spirometry	Provide oxygen if the saturations in air are below 94%. Falling saturations in air or a rising oxygen requirement should prompt re-evaluation and the search for an emerging complication of the crisis. Incentive spirometry improves lung expansion and decreases atelectasis and the risk of acute chest syndrome and should be used. Blowing a balloon and or similar can be a useful alternative.
Adjunctive treatments	<p>Constipation is common during a crisis due to a combination of immobility, the crisis itself and the opiate. Laxatives may be helpful.</p> <p>Anti-pruritics may be needed to counteract the pruritis from the opiates.</p> <p>Pharmaceutical thromboprophylaxis, e.g., with low molecular weight heparin, should be used in peripubertal and children who are pregnant and also considered in anyone who has an indwelling central venous catheter during a crisis. Venous thromboembolic disease is common in sickle cell disease and particularly so in a crisis or when septic.</p>

The acute painful sickle episode

- This is also referred to as a painful or vaso-occlusive crisis and is the most common presentation of sickle- cell disease in childhood, resulting from blockage of small vessels. The mainstay of treatment is effective and prompt pain control (see Section 9 Handbook 1), alongside management of any precipitants.
- Approximately 40% of children with sickle-cell disease who are not on disease modifying therapy, will have an episode of 'hand-foot syndrome' or dactylitis during early childhood, and this number rises to 50% of children under 2 years old who go on to develop symptomatic disease. Typically, children present with vaso-occlusion and infarction of the metacarpals or metatarsals, which is evident as an overlying soft tissue reaction with swelling, redness and marked tenderness affecting either one or all of the hands and feet.
- By later childhood the most common sites of bony sickle-related pain include the long bones, thighs, hips, spine, ribs, shoulders and upper humerus, as well as the bones of the cranium, joints and muscles.

Pain can be described in the following 5 ways (see Section 9 Handbook 1)

Level 1. Mild,

Level 2. Causing significant discomfort,

Level 3. Causing significant distress

Level 4. Is so severe that it can be described as horrible

Level 5. Is so very severe that it can be described as excruciating or the worst possible pain that the child could imagine.

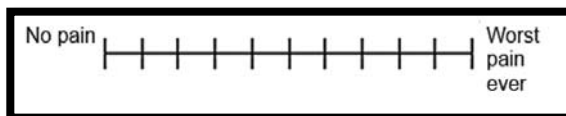


FIGURE 26.2 Visual analogue scale.

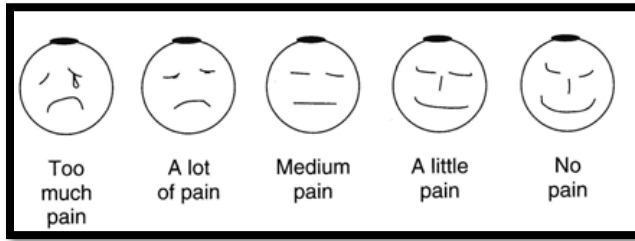


FIGURE 26.3 Faces scale.

Hydration and fluids in sickle-cell disease

- Dehydration occurs readily in children with sickle-cell disease, due to impairment of renal concentrating power.
- Diarrhoea and vomiting are thus of particular concern.
- Fluids can be delivered by the oral, nasogastric or IV route (or a combination of these) and titrated against clinical response. Children should be hydrated to euvolaemia.

Infection

Infection is a common precipitating factor in painful or other types of sickle crises. All children with sickle-cell disease (regardless of type) should be considered to be immunocompromised.

Bacteria

Patients with sickle-cell disease are immunodeficient due to functional asplenia. Functional asplenia occurs in the majority, irrespective of spleen size in sickle-cell disease, well before the age of 6 months in the majority of patients. Clinicians should therefore consider all patients to have increased susceptibility to infection, particularly with the encapsulated organisms listed below, all of which can cause life-threatening sepsis:

- *Pneumococcus*
- *Salmonella species*
- *Haemophilus*.
- *Meningococcus*
- *E-coli*

Sickle cell disease patients normally have increased absorption of iron due to ineffective erythropoiesis and chronic anaemia. This situation promotes bacterial growth. Also, recurrent vaso-occlusive episodes can lead to ischaemic and necrotic tissues which may harbour microorganisms and also make it hard for the immune cells and antibiotics to get to the site of infection. Any suspected bacterial infection should be managed with prompt institution of IV antibiotics to cover these organisms. Suggested choices are listed in Table 26.4 (note that these may vary according to region and local sensitivities). Persistent localised bone pain, swelling or fever should raise suspicion of osteomyelitis, which may require surgical treatment, and 6 weeks of antibiotic therapy

TABLE 26.4 Antibiotic choices in sickle-cell crises

Drug	Rationale and comment
Co-amoxiclav (Augmentin)	Good activity against <i>Pneumococcus</i> <i>Haemophilus</i> resistance is low Suitable for use with clarithromycin for pneumonia Does not mask <i>Salmonella</i> osteomyelitis

Drug	Rationale and comment
Clarithromycin	Good activity against Haemophilus Pneumococcal resistance is low Suitable for use with augmentin for pneumonia Does not mask Salmonella
Cefuroxime	Suitable for severe pneumonia with or without clarithromycin Masks Salmonella
Ceftriaxone and other third-generation cephalosporins	For suspected sepsis First-line treatment for suspected osteomyelitis (with clindamycin) Second-line treatment for Yersinia if there is glucose-6-phosphate dehydrogenase (G6PD) deficiency
Ciprofloxacin	For use in patients on desferrioxamine with suspected Yersinia infection Stop iron chelation if suspected

Specific infections

Osteomyelitis

Persistent localised bone pain, swelling or fever should raise suspicion of osteomyelitis, which may require surgical treatment, and at least 6 weeks of antibiotic therapy; if not longer.

This infection can be very difficult to distinguish from vaso-occlusive bone pain, which is commonly associated with localised swelling and joint effusions. Osteomyelitis should be considered in any child with persistent and localised pain who is systemically unwell.

The diagnosis of osteomyelitis in sickle-cell disease is more likely in the presence of:

- swinging pyrexia (fevers may not be persistent)
- severe systemic upset
- unusual swelling or pain
- positive blood cultures
- very raised CRP often > 150

Few if any investigations are absolutely conclusive in making the diagnosis. Bone biopsy can be diagnostic but is rarely performed. Treatment is complex and may involve surgical intervention (rare) and a prolonged course of IV antibiotics (6 weeks or more). The oral route can be used to complete a course of antibiotics once the child is systemically well (that is fevers have settled) and the tissue penetration of the antibiotic sufficient and any tests such as CRP have returned to normal. Antibiotic choices are broad, but may include the following:

- first line: IV ceftriaxone and clindamycin (consider flucloxacillin if no clindamycin available).
- second line: IV clindamycin.
- alternatives: meropenem, imipenem or ciprofloxacin.

Malaria

Although it is thought that sickle-cell trait (HbAS) may offer some protection against severe complicated malaria, including cerebral malaria and severe anaemia related to malaria in children, malarial infection in sickle-cell disease can be rapidly fatal and requires prompt recognition and urgent treatment. Although children with sickle-cell disease are not at greater risk of contracting malaria infection, once infected they have a higher mortality, especially

Section 26. Sickle-cell disease [Dr. Sara Trompeter](#), [Dr. Vivian Paintsil](#), [Dr. Simon Parke](#) related to severe anaemia. In addition to drug treatment, transfusion may be required. It should be noted that first generation children in non-malarial countries will not have the acquired immunity of their parents and need to adopt stringent anti-malarial precautions when travelling to malarial endemic countries. Prevention should be emphasised (see Section 31 Handbook 1).

Meningitis

Bacterial meningitis is more common in children with sickle-cell disease than in unaffected children, especially in the youngest age groups. The most frequent infecting organism is pneumococcus. Clinicians should maintain a high index of suspicion for this complication and treat it empirically. (Section 67 Handbook 1).

Gastroenteritis/diarrhoea

Severe diarrhoea may precipitate sickling and crisis, including stroke. Hydration must therefore be maintained vigorously using orally or NG given Oral Rehydration Salts (ORS) or IV fluid where necessary. Education relating to hand hygiene, clean water and prompt treatment should be given.

Children who are systemically unwell with a diarrhoeal illness may also be at higher risk of sepsis related to Gramnegative infection and may require IV antibiotic treatment in addition to vigorous rehydration under such circumstances. Children with diarrhoea, and those with iron loading, who are also on the iron chelation medication desferrioxamine are at high risk of Yersinia or Klebsiella infection, and require prompt treatment with ciprofloxacin, alongside discontinuation of the desferrioxamine until they recover.

Viral infection

Children with sickle-cell disease are at particular risk of profound anaemia secondary to parvovirus B19 infection, which may trigger an aplastic crisis.

Children with sickle-cell disease should also be protected from blood-borne viral infection, specifically HIV, HTLV, hepatitis C, and hepatitis B infection. Routine immunisation against HBV must be undertaken in view of the probability that a child with sickle-cell disease may at some stage be a recipient of blood products or be started on a long-term transfusion programme. Immunisation against influenza, and routine paediatric immunisations as per local protocol, should also be provided.

Data available on the impact of COVID-19 on children with sickle cell disease at the time of writing (March 2021), seems to be in parallel with what is found in the general population. That is, that this is largely a disease that is mild or un-noticed in the paediatric cohort. Once the data from the immunisation studies in childhood become available it may well be that children with sickle cell disease are recommended the immunisation in line with the recommendations made for the adult sickle cohort.

Severe anaemia in sickle-cell disease

Children with sickle-cell disease are known to have a compensated anaemia but are also at risk of events that may precipitate a sudden and potentially fatal drop in their haemoglobin levels. The main conditions to consider are as follows:

- acute sequestration events
- aplastic crisis
- infection with malaria.

Acute sequestration events

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Sequestration events are characterised by pooling of red cells in an organ, most commonly the spleen, lungs and liver, and are associated with a sudden and potentially life-threatening fall in haemoglobin level, with shock and collapse alongside rapid (and often painful) expansion of the organ affected.

Sequestration events are often precipitated by infection or sepsis that requires vigorous antibiotic treatment. There is a high mortality. Any child who appears to be deteriorating during an acute painful crisis should be re-examined to exclude undiagnosed sequestration. Parents should be taught to examine their child's abdomen so that they may identify the need for transfusion

Treatment includes administration of antibiotics to manage any precipitating infection, and blood transfusion in children with cardiovascular compromise, or who have a haemoglobin level of < 50 g/l, or where there has been a sharp fall in haemoglobin level by > 20g/l or if the child is compromised.

Urgent blood transfusion in children with sickle-cell disease is not uncommon but does carry some risks. Clinicians should be cautious about over-transfusing beyond a target of 80 grams/L (usually a maximum of 20 mL/kg) or at a higher rate than 5 mL/kg/hour (though this depends on whether this is whole blood or packed cells), due to the risks of hyper-viscosity associated with a sudden increase in haematocrit. However, some patients have a higher baseline Hb, especially those with HbSC genotypes, and may tolerate a higher haemoglobin, though transfusion beyond Hb115g/l is rarely recommended. Low sickle cell percentages reduce viscosity and thus higher Hb are safer if the sickle percentage is low. It should be noted that in sequestration, some of the blood in the organ may eventually re-enter the circulation and raise the haemoglobin further, so not over-transfusing is important.

Aplastic crisis

Transient red cell aplasia caused by parvovirus B19 (with an associated reticulocytopenia) can lead to a sudden severe worsening of the patient's anaemia. Ask about any recent viral prodromal illness, but classical erythema infectiosum ('slapped cheek syndrome') is uncommon. Second infections with parvovirus are extremely rare, as immunity to parvovirus is lifelong in the immunocompetent. Review other family members with sickle-cell disease because they too may be infected with parvovirus.

The differential diagnosis of a sudden fall in haemoglobin level includes sequestration crisis, and therefore abdominal palpation is mandatory in any acutely anaemic child to exclude this diagnosis.

Treatment includes use of blood transfusion in children who are cardiovascular compromised, if the haemoglobin level is < 50g/l, or if there has been a sharp fall in haemoglobin level by > 20 g/l.

Acute chest syndrome (ACS)

This is a major cause of morbidity and mortality in sickle-cell disease. It is strictly defined by evidence of new pulmonary infiltrates involving at least one complete lung segment consistent with the presence of alveolar consolidation but excluding atelectasis. Characteristically, children present with fever and pain and then develop the more characteristic features of hypoxia and crackles on auscultation. Cough, tachypnoea, and wheezing are often present. **It is important to recognise that patients can be in the process of developing ACS and be severely ill before these strict criteria are met and a drop in saturation of 4% on room air or SpO₂ < 94% should alert the clinical team to the possibility of chest syndrome.** Signs of lung consolidation, usually bilateral, generally

Section 26. Sickle-cell disease [Dr. Sara Trompeter](#), [Dr. Vivian Paintsil](#), [Dr. Simon Parke](#) start at the bases, but may be unilateral and impossible to distinguish from infection which may co-exist. ACS usually does not produce an air bronchogram.

Acute sickle chest syndrome is likely to be multifactorial in origin, with infection, thrombosis of pulmonary arteries and fat embolism all resulting in potentially similar clinical patterns.

Management of ACS

- Anticipatory clinical approach.
- Effective analgesia and incentive spirometry to prevent basal atelectasis.
- Careful observations, including regular pulse oximetry (measured on AIR).
- Chest X-ray: upper lobe consolidation without basal changes suggests pneumonia rather than ACS.
- Start dual IV antibiotics: treat pneumonia aggressively as it is often clinically indistinguishable.
- Oxygen to keep saturations >94%.
- Hydration.
- Blood transfusion: a small top up transfusion preferably with packed cells may be sufficient in milder cases and where the haemoglobin is low enough to permit it (given that haemoglobin should not rise by >40g/l in one day and nor higher than 110g/l). However, often an exchange transfusion may be needed as simple transfusion may not sufficiently decrease the sickle percentage given these limits.
- Nasal CPAP if available and saturations falling to the low 90s in air.
- May require non-invasive ventilation, though in bad cases invasive ventilation may be needed and there are some cases where ECMO has been used.
- There is no role for diuretics.

Neurological involvement in sickle-cell disease

Sickle-cell disease is associated with several central nervous system complications and events, as outlined below. The most significant event is stroke, mainly infarction, in early childhood. However, in later teens cerebral haemorrhage is commoner usually on the background of MoyaMoya secondary to cerebrovascular disease. The treatment approach is outlined in the next section.

Neurological complications of sickle-cell disease

- Infection: meningitis and malaria.
- Stroke: ischaemic stroke, subarachnoid haemorrhage and transient ischaemic attacks (TIAs).
- Silent infarcts.
- Convulsions.
- Neurocognitive decline particularly of executive function and frequent in those with silent cerebral infarcts.
- Posterior Reversible Leuco-encephalopathy syndrome (PRES) – often precipitated by use of high dose steroids and over-hydration during a crisis.

Stroke in sickle-cell disease

Stroke is a potentially devastating complication of sickle-cell disease, most commonly occurring in (but not limited to) individuals with homozygous disease (HbSS). The most common event is infarctive stroke, but haemorrhagic stroke can also occur with increasing frequency as children progress towards adulthood. Stroke can occur in any age group but is most common in children under 10 years.

Predictive factors for stroke include a history of transient ischaemic attacks, a recent episode of acute chest syndrome, hypertension, or a low haemoglobin F percentage and/or low

Section 26. Sickle-cell disease [Dr. Sara Trompeter](#), [Dr. Vivian Paintsil](#), [Dr. Simon Parke](#) baseline haemoglobin levels. The greatest predictor of stroke is a raised transcranial doppler (TCD) velocity >200cm/s (non-imaging) with a 10% risk of stroke per patient within the year and where commencement of regular transfusion to keep HbS<30% following such a result gives a 90% risk reduction in stroke events. This has led to TCD screening programmes (annually from 2-16 years) to identify children with raised velocities and commencing risk reduction treatment with regular transfusion to keep HbS <30%. Any child with sickle-cell disease can have a stroke (even if they are apparently not 'high risk').

Precipitating factors for stroke include a recent history of fever, infection, dehydration and acute chest syndrome. However, some children will have a stroke without any identifiable precipitating event or risk factor.

Symptoms and signs of stroke can be broad and range from the 'classic' presentation of a focal neurological deficit such as a hemiplegia (painless loss of function) to behavioural changes, severe headache, altered consciousness, convulsions, or coma.

Historic data, prior to the use of transcranial dopplers and routine use of hydroxycarbide, from the USA suggest that 11% of children with sickle-cell disease have an overt stroke and 40% silent infarcts. More recent data from well-resourced countries speculate that this figure is coming down with the advent of transcranial Doppler (TCD) screening to identify children at high risk of stroke and the aggressive use of regular long-term blood transfusion programmes as a primary prevention strategy as well as early commencement of hydroxycarbamide.

Treatment of acute stroke

Prompt treatment of an ischaemic stroke can potentially arrest a stroke in evolution.

Children with a suspected stroke require:

- rehydration with fluids
- antibiotic treatment of any suspected infection, including malaria or meningitis
- treatment of any convulsions (see Sections 69 and 70 Handbook 1)
- exchange transfusion to reduce the circulating sickle percentage as rapidly as possible to less than 30%.
- There are no prospective studies of the use of thrombolysis, but it has been used, predominantly in adulthood.

In the absence of accessible exchange transfusion, it may be reasonable to consider a cautious top-up blood transfusion to maximise oxygen-carrying capacity and reduce the HbS percentage through a dilutional effect. Extreme care must be taken to avoid over-transfusion and the risk associated with increasing blood viscosity thus further contributing to the stroke. In either situation, the haemoglobin should not be increased by >40g/l in a day or beyond 100g/l.

Most children make a good motor recovery from an initial stroke but may be left with intellectual defects, particularly of executive function. If untreated, most of these children will suffer a second cerebrovascular accident, usually within 2–3 years of the first episode, as a result of which many of them will die and most will be seriously impaired. Transient ischaemic attacks may presage a more major event.

Secondary prevention of stroke

Because of the risk of a subsequent stroke, all children should be considered for the long-term transfusion programme to reduce their recurrence risk (although the risk is never fully eradicated). Most children require a top-up transfusion every 4 weeks for life, and this is a heavy burden for patients and their families.

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The treatment goals of secondary prevention of stroke using the transfusion programme are as follows:

- to reduce and then maintain the pre-transfusion HbS% at below 30%
- to monitor and treat iron overload.

Note that there is no role for co-administration of desferrioxamine during transfusions.

Risks of the long-term transfusion programme include the following:

- transmission of blood borne viral infection
- allo-immunisation to foreign red cell antigens
- iron overload.

Some patients have undergone cerebral revascularization post stroke.

In well-resourced settings, some children may be able to receive alternatives to long-term top-up transfusions as outlined above. These alternatives include the use of manual or automated **exchange transfusions** to maintain a low HbS% without incurring iron overload states. Automated exchange, depending how the parameters are set, can be delivered without incurring iron overload. It is also delivered half as often though more blood is needed per episode. These children may be able to go for longer periods between blood transfusions, although the risk of exposure to blood does not change.

Recent studies have shown that after 18 months of primary prevention of stroke with regular transfusion, in those with a normal brain MRI/MRA, that transfer to hydroxycarbamide is not inferior. However, hydroxycarbamide for secondary prevention of stroke was inferior to continued transfusion and is not recommended where transfusion is available. Use of hydroxycarbamide for primary prevention of stroke in those with raised transcranial dopplers cannot be recommended where regular transfusion is available but can be considered where it is not available.

Prevention programmes

Iron overload and transfusions

Children who are exposed to multiple and regular blood transfusions are likely to develop iron overload. The most widely available iron-chelating agent is desferrioxamine (Desferal), which is administered as a subcutaneous dose around 20–40 mg/kg average daily dose over slow subcutaneous infusion (8–12 hours) for 5–7 nights per week. Many children become non-compliant with this regimen, and newer medications are available as outlined below.

TABLE 26.5 Drug treatments to reduce iron loading (iron chelation)

Drug	Advantages	Disadvantages
Desferrioxamine	Well-understood safety profile through long-term use Cheap	Relatively poor iron chelation properties Poor patient compliance Risk of Yersinia infection Needs refrigeration. Audiological and ophthalmological damage usually at higher doses and should be monitored.
Deferiprone	Oral administration Effective chelation agent	Given three times daily. Requires close monitoring due to risk of sudden neutropenia and risk of overwhelming infection

Drug	Advantages	Disadvantages
Deferasirox (Exjade)	Oral once-daily administration Well tolerated Highly effective iron chelation	Cannot be used if renal dysfunction. Requires monitoring

Prevention of infection

Prevention of infection is the mainstay of reducing mortality and morbidity in sickle-cell disease.

- All children should receive immunisation against Pneumococcus, Haemophilus influenzae, meningococcal ACWY, B. influenza and hepatitis B, in addition to any standard immunisation schedule. COVID-19 immunisation has been recommended for adults and may be recommended for children when results from the paediatric trials become available.
- Pneumococcal immunisation should be as broad as possible, including pneumococcal conjugate vaccine **and** Pneumovax. Pneumovax should be given from the age of 2 years, every 5 years for life.
- All children should receive prophylactic penicillin V (erythromycin or clarithromycin can be used as an alternative):
 age up to 1 year: penicillin 62.5 mg twice a day
 age up to 5 years: penicillin 125 mg twice a day
 age 5 years or over: penicillin 250 mg twice a day into adulthood.
- All children should be protected from malaria infection (see Section 31 Handbook 1).
- Families should be counselled about prevention, risks and signs of infection, so that they can seek prompt treatment.

Prevention of crises

- Good attention to health, diet and fitness to optimize baseline health.
- Maintain good fluid intake, especially during gastroenteritis or other infections.
- Folic acid:
 - a. age up to 1 year: 1 mg daily
 - b. age up to 5 years: 2.5 mg daily
 - c. age 5 years or over: 5 mg daily into adulthood.
- Families should be taught how to feel their child’s abdomen to identify the onset of a sequestration crisis.
- **Hydroxyurea (hydroxycarbamide) is** now recommended in many national guidelines to be used routinely from infancy, even in children who are asymptomatic. It is universally recommended for the HbSS and HbSBeta zero genotypes and discussed with those of other genotypes. Blood tests are needed to monitor for cytopenias (neutrophils <1 and platelets <80) and are usually performed 3-monthly once on a stable dose or 2-3 weeks post a change in dose.

Children are usually started on 20mg/kg/day and the dose can be incremented by 5mg/kg/day to a maximum tolerated dose which is generally 30-35mg/kg/day.

Hydroxycarbamide should be stopped 3 months before trying for a family as its use in pregnancy is not well known though is the subject of an ongoing clinical trial.

Males can be offered sperm analysis and saving where this exists.

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There is ample evidence that use of hydroxycarbamide does NOT increase risk of cancer in those taking it. It may raise baseline haemoglobin levels by promoting fetal haemoglobin (HbF) production. This may reduce the frequency and severity of crises in children. However, it can suppress the bone marrow and should be used with caution and only where facilities for monitoring blood counts exist and the dose can be monitored carefully. Families should be consented regarding its use and children assented in adolescence.

- Voxelator and other agents: there are other agents in various stages of clinical trials, some of which have already been licensed in adulthood at the time of writing. Most of these are being trialed for the prevention of painful crises and sometimes in conjunction with hydroxycarbamide. It is likely that over time, different combinations of medications will be recommended for the prevention of crises.
- Regular blood transfusion to keep HbS<30% can be used to prevent crises where it is available and usually is offered for stroke prevention or where hydroxycarbamide is insufficient, contraindicated or not tolerated.

Splenectomy in sickle-cell disease

Splenectomy is not routinely undertaken in children with sickle-cell disease, although it does have a role in improving hypersplenism and in those who have had an episode of life-threatening or repeated episodes of sequestration.

Splenectomy may also be indicated in children who have had an episode of life-threatening splenic sequestration.

Perioperative care in sickle cell disease

Peri-operative complications are higher for patients with sickle cell disease than for the general surgical population, but outcomes can be improved with careful peri-operative care and the involvement of specialist teams.

There should be ideally a nominated lead paediatrician or paediatric haematologist who is responsible for deciding the peri-operative transfusion plan, with support of the specialist centre where relevant. The surgical team booking the patient for surgery should ensure the relevant teams are aware of the patient and their sickle cell diagnosis. For elective surgery, patients should be reviewed in a pre-assessment clinic and for all surgery the cases discussed with the paediatrician/paediatric haematologist.

Other actions that improve perioperative care are the involvement of an acute pain team; scheduling early on the operating list to avoid prolonged starvation; ensuring good hydration pre-operatively. Patients are at increased risk of sickle complications (acute chest syndrome, pain, acute renal insufficiency, or stroke), sepsis and venous thrombo-embolism in the peri-operative period. The majority of complications occur postoperatively, and there should be a low threshold to admit patients to high dependency or intensive care. Patients require meticulous peri-operative care to avoid factors that may precipitate sickling such as dehydration, hypoxia, acidosis, hypothermia, and pain.

Routine surgery should be avoided if the patient is febrile or having a painful crisis. All patients require a full blood count, urea and electrolytes and group and full red cell antibody screen prior to surgery.

Pre-operative transfusion is considered based on the sickle genotype, the age of the patient, comorbidities, and the risk of surgery. Transfusion may be given as a simple 'top-up' or as a partial or full exchange transfusion to reduce the HbS% to a target level. The target Hb is kept < 110 g/l and should not be increased by more than 40 g/l in a single transfusion episode. Patients undergoing high-risk surgery or those with significant comorbidity are likely

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For low-risk surgery a simple top-up transfusion pre-operatively to a target of 100 g/l is usually recommended. If the Hb is ≥ 90 g/l and the risk of surgery is low, then patients do not universally need transfusion. Blood should be available on site for patients undergoing surgery so it can be given without delay should complications arise.

Growth

Growth failure and delayed puberty are common in children with sickle-cell disease, not on disease modifying therapy, especially in those with hypersplenism or who have had multiple acute sickle crises. Weight tends to be affected more than height, and malnutrition is a major factor in determining whether children achieve their full growth potential. Puberty may be delayed because of hypersplenism or malnutrition because of the hyper-metabolic state and inadequate nutrition. Dietary advice, treatment of any chronic infections and possibly splenectomy (if hypersplenism is present) may be helpful. Occasionally, children may benefit from temporary use of the monthly transfusion programme to assist them into puberty.

Priapism

Priapism is a serious but under-reported complication of sickle-cell disease. If untreated, it can lead to fibrosis of the corpus cavernosa and impotence, a risk which appears to be lower in pre-pubertal boys. The frequency or duration of an episode predicts the overall outcome. Therefore, prompt recognition and management are essential.

Patients typically present with an erect painful penis, which may be precipitated by a painful sickle crisis, fever, dehydration, use of recreational drugs, or sexual activity.

Acute fulminant priapism is characterised by a prolonged and sustained episode, more than 4 hours in duration. In stuttering priapism, episodes are repetitive and may be individually brief. Patients may have a combination of both events.

Treatment of acute priapism includes the initial use of warm baths, exercise, hydration and gentle sedation while preparing for a more definitive intervention. However, if this does not help prompt attendance for clinical care is needed. Subsequent definitive treatment choices include aspiration of blood from the corpus cavernosum followed by surgical washout using saline (irrigation) or adrenergic agonists, which can be performed under conscious sedation. The goal is rapid detumescence within 4–12 hours of the procedure. Ideally, treatment should start within 2 hours of an episode. After 12 hours the patient may require surgical intervention to achieve detumescence. Exchange transfusion (the target haemoglobin concentration is approximately 110 g/L, with a haematocrit no higher than 0.4) may be required.

Fulminant episodes are often preceded by bouts of stuttering priapism, Bladder emptying, gentle exercise, warm baths and analgesia may help abort an attack. Oral etilefrine, pseudoephedrine or cyproterone acetate are often used for secondary prevention.

Other problems

- Around 30% of SS children suffer from **sleep-related upper airways obstruction** with consequent hypoxaemia. Nocturnal hypoxaemia has been increasingly identified as a risk factor for acute chest syndrome (and possibly an independent risk factor for stroke) in children with sickle-cell disease and marked improvement can occur after adenotonsillectomy. Treatment is as indicated for other children with upper airways obstruction (see Section 32 Handbook 1).
- Chronic pain resulting from damage caused by acute vaso-occlusive crises occurs, and other pain secondary to the haemolytic process can occur.

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- 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 flucloxacillin, wound cleaning and protection together with rest and elevation of the leg are helpful. Compression stockings may also be of benefit.
- Children develop a renal tubular concentrating defect by the age of 2 years. Enuresis is very common. During adolescence, proteinuria, the nephrotic syndrome or chronic renal failure may develop.
- 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.
- Gallstones are common, due to pigment from haemolysis. They may be asymptomatic though if there are complications then cholecystectomy may be recommended following recovery from the acute event. Cholecystitis is treated with antibiotics. ERCP may be needed for stone extraction. Treatment is surgical. Antibiotic treatment of cholecystitis with amoxicillin and metronidazole may be required.

Blood transfusion in sickle cell disease

People with sickle cell disease should receive ABO Rh CcDEe and Kell compatible sickle negative blood that is negative for known clinically significant red cell antibodies. Blood should be obtained from screened un-renumerated donors. Blood should be screened for HIV, HTLV, Hepatitis B and C. The recipient's ABO and Rh group should be determined pre transfusion. Where it is possible a full red cell pheno/genotype should be performed at diagnosis. Leucodepletion is preferable. Patients should be consented as to the risks and benefits of blood transfusion and should be told if they have an antibody (and given a card with this information). There are international standards regarding blood donation, component quality, infusion and storage e.g., WHO and these should be adhered to. The full details of these are beyond the scope of this chapter.

When giving a blood transfusion in sickle cell disease the haemoglobin should not increase beyond 40g/l in a day or >110g/l generally speaking, though there may be exceptions to this. For stroke prevention or treatment or when patients are very unwell, attempts should be made to decrease the HbS<30%. Patients should be counselled as to the risk of blood transfusion and alerted as to how a transfusion reaction may present (lethargy, pallor, jaundice, dark urine) and to seek help if that occurs. Clinical teams looking after patients with sickle cell disease should be aware of their requirements, inform the laboratory, and have the training and ability to detect and manage a transfusion reaction.

Additional support

The impact of chronic disease on the child and the family should not be underestimated. Children may need psychological support and/or educational support.

Blood tests can be stressful, and the use of local anaesthetic cream or cold spray is often helpful. Distraction techniques can be useful for procedures that are difficult or stressful.

Support, engagement and education of the family and teenagers is key to good patient care and supports transition into adult services. Liaison with other teams such as education, primary care and social care is often needed.

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Section 27. Acute Skin Disorders

Introduction

In resource-limited countries, skin disease is dominated by bacterial infections such as impetigo and parasitic conditions such as scabies and pediculosis. It is often poorly managed and may incur a significant 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 will have 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 person to person, usually by direct contact. The adult female burrows a tunnel into the stratum corneum or outer skin layer, producing eggs which hatch into larvae within 3–4 days. 'Outbreaks' in communities may follow a cyclical pattern, with peaks of incidence occurring every 4–7 years but in highly endemic areas infection rates of over 10% are constant. Infection usually reflects overcrowding in households and transmission through contact with infected individuals, including infants.

Clinical presentation

The main sites of infection include fingers, wrists, elbows, ankles, genitals and buttocks; the face and head may be affected in babies, but these sites are seldom involved in older children. Important clues include the following:

itching in several members of the same household
lesions in characteristic sites, particularly the lateral borders of the fingers
papules, pustules and sinuous tracks or burrows (5–10 mm).

In onchocerciasis (see Section 42 Handbook 2), itching is also common, but lesions are seldom found on the fingers.

Diagnosis

This is often based on typical signs including burrows, distribution and presence of others affected in the same household. Mites can be extracted from their burrows with a sterile needle and examined microscopically.

Complications

Secondary bacterial (streptococcal) infection is common (see below). In severely immunocompromised individuals (e.g. those with AIDS) a crusted form of scabies, with- out severe itching but with large numbers of mites, may occur.



Figure 27.1 Scabies on and between the fingers

TABLE 27.1 Topical treatment of scabies

Anti-scabies preparation	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	Initial application, followed by a second one 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% Crotamiton cream	1–2 weeks of treatment	Not very effective, although it can reduce itching

Treatment

The cheapest treatment options are sulphur based. However, they are slow to take effect, and require daily applications for 7–14 days. Permethrin is the most rapidly active but also the most expensive option.

All potentially affected areas are treated, including the soles of the feet and, in babies, the scalp.

Failure of anti-scabetic agents often occurs because there is no place where individuals can apply these treatments in privacy. Treat all members of the household, including those without itching.

Clothes should be cleaned or changed after the first treatment. Ivermectin (2 doses of 200 microgram/kg orally a week apart) is highly effective as a mass treatment in communities where the prevalence is higher than 10%; as well as for crusted scabies but is not suitable

for children under 5 years who should receive a single dose of 150 micrograms/kg. No food should be taken for 2 hours before or after the dose.

Community-based treatments with ivermectin have been very successful in eliminating scabies.

Impetigo

The term **pyoderma** is used to describe a range of superficial bacterial infections that include impetigo, folliculitis, abscesses (furunculosis) or secondary bacterial infection (e.g., of scabies). Impetigo is a form of pyogenic infection that involves the epidermis and is caused by Group A streptococci or *Staphylococcus aureus*. It is not possible to separate the two infections clinically.

Ecthyma occurs when impetigo penetrates deeper, to affect the dermis and causes ulceration.

Clinical presentation

Impetigo presents with oozing and yellowish crusted plaques, often on exposed sites such as the face. These plaques may be infectious and may be transmitted to other parts of the body and to other children. Secondary infection of scabies with Streptococci may occur; papules become pustular and there may be surrounding impetiginised crusts on scabetic burrows. Boils (furuncles) are also common and are always caused by *Staphylococcus aureus*. Lesions are large tender fluctuant masses with surrounding inflammation. They may occur in other members of the same household.



Figure 27.2 Bullous impetigo with blisters on upper lip

Complications

A serious complication of streptococcal impetigo or secondary infection of scabies is glomerulonephritis, which follows infection by nephritogenic strains. In tropical environments, post-streptococcal glomerulonephritis more often follows skin infection rather than throat infection.

Management

Impetigo is transmissible, and treatment should include other contacts with lesions. Cover both *Staphylococcus 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 27.2). The choice of medication is influenced by cost, extent of disease and type of lesions.

Most *Staphylococcus aureus* strains, even in remote communities, are resistant to both penicillin and tetracycline. Most topical azole antifungal agents (e.g. clotrimazole, miconazole), apart from ketoconazole, have activity against Gram-positive bacteria. Boils are best managed by incision and drainage.

TABLE 27.2 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 effect, with clearance in 3–5 days	Expensive
Mupirocin	Topical	For localised infections. Rapid effect, with clearance in 3–7 days	Moderate
Fucidin	Topical	As for mupirocin	Moderate
Clioquinol	Topical	Slow to take effect (7–14 days). May stain skin; irritant	Cheap
Potassium permanganate (alternatives are chlorhexidine and povidone iodine)	Topical	Simple to use, stains skin. Slow to take effect (7–14 days)	Cheap

Tropical ulcer (tropical phagedenic ulcer)

A variety of ulcers occur in children and teenagers but are seldom seen in non-tropical environments. They are found patchily distributed in endemic foci throughout Africa, India and the West Pacific and are associated with humid regions.

Tropical ulcers are considered to result from several different causes such as synergistic bacterial infection, of which one anaerobic organism is usually *Fusobacterium ulcerans*; *Haemophilus ducreyi* also causes skin ulcers in children. It is possible that there are other causes of tropical ulcers, and it is important to distinguish all these from yaws (Section 53)

The initial lesion may be a soft papule with surrounding hyper-pigmentation overlying an area of skin necrosis. This develops over at least 1 week, and when the overlying skin sloughs a regular and deep ulcer, 3–10 cm in diameter, is revealed. Other ulcers appear de novo as round or oval ulcers with granulating bases

Complications

With proper care and early treatment, the area will heal. About 5–10% of lesions may progress to chronic ulceration.

Management

The objective of treatment is to allow rapid healing without secondary infection.

Regimen

- Dilute antiseptic (e.g., potassium permanganate solution), or 0.9% saline for cleansing the ulcer and surrounding skin. Daily dressings where necessary
- A single dose of oral azithromycin (30 mg/kg) or oral metronidazole (7.5 mg/kg every 8 hours for 5 days). 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 38).

Superficial fungal infections

Common childhood fungal infections are scalp ringworm or tinea capitis and oropharyngeal candidiasis.

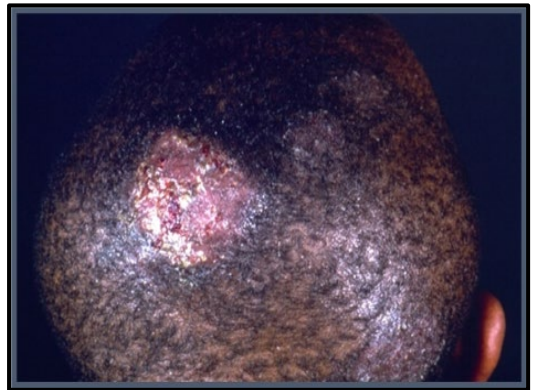
Tinea infections are caused by dermatophyte fungi, which are adapted to survive on the outer layer of the skin, the stratum corneum, or structures such as hair or nails derived from it. Dermatophyte infections are caused by genera of fungi, mainly *Trichophyton*, *Microsporum* and *Epidermophyton*, which are acquired by spread from soil, animal or human sources (geophilic, zoophilic or anthropophilic infections, respectively). By convention they are referred to by the term tinea followed by the appropriate Latin word for the site affected – for example, tinea pedis (feet), tinea corporis (body) or tinea capitis (scalp).

Figure 27.3 Tinea capitis

Tinea capitis is often endemic in rural or urban areas of resource-limited countries and inner-city areas of industrialised countries. Prevalence rates may reach over 20% in some communities.

The main signs of infection are as follows:

- scaling
- hair loss: this may be diffuse or in localised patches; scalp hairs in affected areas may break at scalp level or a few millimetres above the skin
- itching: this is variable.



The key to the diagnosis is the presence of broken hairs. Confirmation is by culture of scrapings taken from the scalp surface with a sterilised scalpel or sterile scalp brushes. The 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 the surface of inflamed lesions.

Management

Culture of fungus can distinguish whether infection is from a human or animal source, i.e., zoophilic species



(*Microsporum canis*, from cats and dogs; *Trichophyton verrucosum*, from cattle) or anthropophilic species (*Trichophyton violaceum*, *T. tonsurans*, *T. soudanense* or *Microsporum audouinii*). The presence of infections in close contacts (e.g., schoolmates or family) may signal child-to-child spread and alert schools to other infected children. In resource-limited countries, mass treatments have a low priority because of health resources and because cases usually self-heal. Children with severe symptoms (e.g., kerions, favus or widespread hair loss) should be treated.

- Whitfield's ointment or imidazole antifungal agents (e.g., clotrimazole) are generally ineffective in scalp ringworm.
- The treatment of choice is griseofulvin, which is available in oral tablet or solution form, 10 mg/kg once daily after food (max 500mg per dose and up to 20 mg/kg (max 1gram per dose) in refractory infections. Single-dose treatments with a 1-gram immediate dose, sometimes repeated after 1 month, have been successful for mass treatment of infected classes in school.
- Terbinafine child 10–20 kg 62.5 mg; child 20–40 kg 125 mg; child > 40 kg 250 mg all once daily for 4 weeks in tinea capitis is an alternative.
- 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 the epidermis and dermis. In childhood the commonest form of eczema is atopic dermatitis. The latter is uncommon in rural areas of resource-limited countries and appears to be associated with urban environments and increased affluence.

Clinical presentation

Severe itching and a scaling rash affect the skin flexures (e.g., the elbows, behind the knees, the neck). Scratching may be very severe, and sufficient to disturb sleep.

Management principles

- **Moisturise the 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 only intermittently, relying on emollients for long-term management.
- **Treat complications.** These are secondary bacterial infections, usually *Staphylococcus aureus*. An oral antibiotic (e.g., cloxacillin or flucloxacillin 12.5–25 mg/kg four times a day) in acute flare-up of eczema may produce a good response.
- Acute herpes simplex (eczema herpeticum) is a serious complication: apply aciclovir cream five times a day for 5–10 days (until healed) or aciclovir orally if severe, 20 mg/kg four times a day for 5–7 days.
- Secondary contact dermatitis may include allergy to topical medicaments such as lanolin and corticosteroids.

Atopic eczema ranges from a mild skin rash to a severe condition that 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 (e.g., the National Eczema Society in the UK) which provide support and advice to patients and their families. <https://eczema.org/> Accessed April 9 2021

Hypopigmentation and hyperpigmentation disorders

These are often secondary to other inflammatory processes which should be treated. There are no effective, inexpensive 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 (e.g. clotrimazole, miconazole) is effective.

Further reading

Regular updates on the management of skin disease in resource-limited environments are available in the Community Skin Health, which can be accessed without charge on <https://ilds.org/our-foundation/community-skin-health-journal/> Accessed April 9 2021

Section 28. Upper gastroenterological disorders

Introduction

Upper gastrointestinal disorders are not common complaints in the population presenting to hospitals in resource-limited 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 the oesophagus due to a foreign body, strictures due to caustic soda poisoning, haematemesis due to peptic ulcer or portal hypertension, and volvulus due to malrotation.

In resource-limited communities, particularly where facilities for upper gastrointestinal paediatric endoscopy are available, similar symptoms to those that occur in well-resourced countries present. These include recurrent abdominal pain, epigastric and substernal pain, recurrent/ persistent vomiting, dyspepsia and water-brash/heartburn.

Gastro-oesophageal reflux

TABLE 28.1 Gastro-oesophageal reflux

Symptoms	Complications
Vomiting (regurgitation)	In Infants
Water-brash/heartburn	Apnoea
Nausea	Life-threatening event
Epigastric/retrosternal pain	All ages
	Failure to thrive*
	Aspiration pneumonia
	Haematemesis
	Anaemia
	Oesophageal stricture

*Unusual in normal infants but particularly common in children with cerebral palsy.

Gastro-oesophageal reflux (GOR) is a normal physiological condition in infants, children, and adults. If GOR is associated with complications (as below) then it is termed gastro-oesophageal reflux disease (GORD). GORD is common in children with cerebral palsy.

Note: Sandifer–Sutcliffe syndrome is dystonic posturing associated with GOR.

Diagnosis

Often no investigations are needed, and a diagnosis can be made by taking a good clinical history. The following investigations are helpful if they are needed and available:

- **Barium swallow:** This is often the only diagnostic facility available in resource-limited countries. It is a much less sensitive method for diagnosing reflux than pH monitoring but will detect associated or other conditions such as oesophageal stricture, hiatus hernia, diaphragmatic hernia and malrotation.
- Endoscopy and biopsy (particularly looking for oesophagitis).
- pH Monitoring: this grades the frequency and duration of exposure of the lower oesophagus to acid (pH < 4.0).

Management

- Simple GOR in the thriving child: reassurance is all that is needed.

- Excessive regurgitation causing failure to thrive in an infant, or mild symptoms of oesophagitis: treatment by thickening feeds with Carobel (Cow & Gate) or an alginate preparation (e.g. Gaviscon) can be tried.
- Moderate to severe **GOR with oesophagitis**: the proton pump inhibitor, omeprazole:

Omeprazole.

Child 1 month–1 year 700 micrograms/kg once daily, increased, if necessary, to 3 mg/kg once daily (max. per dose 20 mg).

For Child 2–17 years (body weight 10–19 kg) 10 mg once daily, increased, if necessary, to 20 mg once daily, in severe ulcerating reflux oesophagitis, maximum 12 weeks at higher dose.

For Child 2–17 years (body weight 20 kg and above) 20 mg once daily, increased, if necessary, to 40 mg once daily, in severe ulcerating reflux oesophagitis, maximum 12 weeks at higher dose. <https://bnfc.nice.org.uk/drug/omeprazole.html#indicationsAndDoses>

- Motility stimulants such as domperidone (250 mcg/kg three times daily, increased up to 400 mcg/kg if needed. may be effective, particularly in children with cerebral palsy. However, proof of their efficacy is lacking. <https://bnfc.nice.org.uk/drug/domperidone.html#indicationsAndDoses>

- **Surgery**: Nissen fundoplication could be considered if, despite medical management, there was severe oesophagitis, failure to thrive or aspiration pneumonia. It is sometimes required in children with cerebral palsy and GOR.

Helicobacter pylori

Helicobacter pylori is a ubiquitous bacterium that commonly infects the stomach (especially the antrum) of children in resource-limited countries from an early age. Child-to-child transmission is important. In developed countries up to 40–60% of adults are infected, probably mainly during childhood. Conditions associated with *H. pylori* include the following:

- **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

Testing for *H. pylori* should only be undertaken if the child has symptoms of ulcer dyspepsia. Diagnostic tests (outlined below) are rarely available as routine in resource-limited countries.

- Faecal antigen testing: main test used if available, this is sensitive and specific in both children and adults.
- Serology: this is good for epidemiological studies but has reduced sensitivity in children under 7 years of age.
- Urea breath test (13C-U BT): this is sensitive and specific, especially in children over 6 years of age but requires complex equipment.
- Faecal antigen testing: this is sensitive and specific in both children and adults.

Endoscopy: histological demonstration and culture of *H. pylori*.

Management

Selection of optimal antibacterial agents is difficult because of the development of resistance.

Suggested treatment plan: Omeprazole plus metronidazole plus either amoxicillin or clarithromycin

Doses:

Omeprazole

For Child 1–5 years 1–2 mg/kg once daily (max. per dose 40 mg).

For Child 6–11 years 1–2 mg/kg once daily (max. per dose 40 mg).

For Child 12–17 years 40 mg once daily.

PLUS

Metronidazole

For Child 1–5 years 100 mg twice daily.

For Child 6–11 years 200 mg twice daily.

For Child 12–17 years 400 mg twice daily.

PLUS EITHER

Amoxicillin

For Child 1–5 years 250 mg twice daily.

For Child 6–11 years 500 mg twice daily.

For Child 12–17 years 1 g twice daily.

OR

Clarithromycin

For Child 1–5 year 7.5 mg/kg twice daily (max. per dose 500 mg)

For Child 6–11 years 7.5 mg/kg twice daily (max. per dose 500 mg).

For Child 12–17 years 500 mg twice daily.

<https://bnfc.nice.org.uk/drug/omeprazole.html#indicationsAndDoses>

<https://bnfc.nice.org.uk/drug/metronidazole.html#indicationsAndDoses>

<https://bnfc.nice.org.uk/drug/amoxicillin.html#indicationsAndDoses>

<https://bnfc.nice.org.uk/drug/clarithromycin.html#indicationsAndDoses>

Accessed 28th April 2021

Treatment should be continued for 1–2 weeks. Strict compliance in order to avoid the development of resistance is imperative.

Duodenal ulcer

Duodenal ulcers are uncommon in children, but can be life-threatening due to haematemesis, melaena and perforation.

There is often a family history. Common symptoms include epigastric pain that typically:

- is worsened by fasting
- is improved by eating or antacids
- causes **nocturnal waking**.

Diagnosis

- **Endoscopy, including biopsy for *H. pylori*, is the optimal method.**
- Barium swallow: this is less sensitive for 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*.
- See above for treatment regime.

Section 29. Malabsorption, including coeliac disease

Malabsorption

Malabsorption is an abnormality in absorption of food nutrients from the gastrointestinal tract.

Common causes of malabsorption and resultant failure to thrive in resource-limited countries include recurrent respiratory infection, 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 in respectively (Sections 56 & 62 Handbook 1). Only a limited response to nutritional support is expected in HIV infection, depending generally on the stage of disease and the response to antiretroviral (ARV) drugs (Section 36).

Types of malabsorption

- **Selective:** as seen in lactose intolerance.
- **Partial:** as observed in Crohn's disease and HIV infection.
- **Total:** as seen in coeliac disease.

Pathophysiology

The gastrointestinal tract functions to digest and absorb nutrients (fat, carbohydrate, protein, and fibre), micronutrients (vitamins and trace minerals), water and electrolytes. This is dependent on the proper processing of food by mechanical (chewing and gastric churning) and enzymatic (gastric, pancreatic, biliary, or intestinal) means. The final products of digestion are then absorbed through the intestinal epithelial cells.

Malabsorption constitutes the pathological breakdown of the normal physiological sequence of digestion (i.e. intraluminal process), absorption (i.e. mucosal process) and transport (post-mucosal events) of nutrients.

Clinical features

Symptoms and signs can be intestinal or extra-intestinal, and include the following:

- diarrhoea/steatorrhoea: watery, diurnal, and nocturnal, bulky, frequent stools
- bloating
- flatulence
- abdominal discomfort/cramping abdominal pain
- growth retardation
- weight loss
- failure to thrive
- delayed puberty
- swelling or oedema from loss of protein
- anaemia (vitamin B₁₂, folic acid and iron deficiency)
- fatigue
- weakness
- muscle cramp
- osteomalacia and osteoporosis
- bleeding tendencies.

Diagnosis

Investigation is guided by symptoms and signs. Since a range of different conditions can produce malabsorption, it is necessary to look for each of these specifically. Tests are also needed to detect the systemic effects of deficiency of the malabsorbed nutrients (e.g. anaemia with vitamin B₁₂ malabsorption).

Investigations may include the following:

- full blood count and blood film
- C-reactive protein and erythrocyte sedimentation rate

- serum albumin
- serum iron, ferritin, and total iron-binding capacity (TIBC)
- serum folic acid
- serum cholesterol or triglyceride
- serum calcium, phosphate and alkaline phosphatase
- prothrombin time and activated partial thromboplastin time
- blood chemistry (electrolytes, glucose, HCO₃, urea and creatinine)
- serum zinc levels
- stool studies, including cultures.

Serological studies

The following specific tests are carried out to determine the underlying cause:

- IgA anti-transglutaminase antibodies
- IgA anti-endomysial antibodies.

Radiological studies

Barium meal and follow-through.
 Barium enema.
 CT of the abdomen.

Specialised tests (if available)

- Biopsy of small bowel.
 - Colonoscopy can be helpful in colonic and ileal disease.
 - Endoscopic retrograde cholangiopancreatography (ERCP) will show pancreatic and biliary structural abnormalities.
 - Glucose hydrogen breath test for bacterial overgrowth.
 - lactose hydrogen breath test for lactose intolerance
- Magnetic resonance cholangiopancreatography (MRCP).

TABLE 29.1 Common causes of malabsorption

Common causes of malabsorption	
Due to infective agents	Intestinal tuberculosis
	HIV-related malabsorption
	Tropical sprue
	Traveller’s diarrhea
	Parasites, such as Giardia lamblia, fish tapeworm, roundworm, hookworm (Ancylostoma duodenale and Necator americanus)
Due to structural defects	Blind loops
	Inflammatory bowel diseases (e.g. Crohn’s disease)
	Intestinal hurry from surgical procedures (e.g. post-gastrectomy, gastro-jejunostomy)
	Fistulae, diverticulae and strictures
	Short bowel syndrome
Due to mucosal abnormality	Coeliac disease
	Cow’s milk intolerance
	Soya milk intolerance

Common causes of malabsorption	
Due to enzyme deficiencies	Lactose intolerance (constitutional, secondary or rarely congenital)
	Sucrose intolerance
Due to digestive failure	Pancreatic insufficiencies
	Cystic fibrosis
	Chronic pancreatitis
	Bile salt malabsorption
	Terminal ileal disease
	Obstructive jaundice
	Bacterial overgrowth
Due to systemic diseases	Coeliac disease
	Hypothyroidism and hyperthyroidism
	Addison's disease
	Diabetes mellitus
	Hyperparathyroidism and hypoparathyroidism
	Malnutrition

Management

Treatment is directed largely towards management of the underlying cause. In severe nutritional deficiency, hospital admission may be required for total parenteral nutrition (TPN). Subsequently, advice and support from a dietitian is vital.

Coeliac disease

Coeliac disease is an autoimmune disorder of the small intestine in genetically predisposed people of all ages from middle infancy onwards. It is caused by a reaction to gliadin, a gluten protein found in wheat and similar cereals. Therefore, it is common among populations whose diet contains substantial amounts of wheat. Apart from people of European origin, in whom it commonly manifests, it is also frequently seen in North Africa, the Middle East, and the north of the Indian subcontinent where wheat is a staple diet. Other populations at increased risk for coeliac disease include children with Down's syndrome and Turner syndrome, type 1 diabetes, and autoimmune thyroid disease, including both hyperthyroidism and hypothyroidism.

Pathophysiology

Upon exposure to gliadin, the enzyme tissue transglutaminase (tTG) modifies the immune system to cross-react with the small-bowel villous lining, causing an inflammatory reaction. This leads to villous atrophy, which interferes with the absorption of nutrients, minerals and the fat-soluble vitamins A, D, E and K.

Coeliac disease appears to be polyfactorial. Almost all people with coeliac disease have either the HLA-DQ2 or the HLA-DQ8 allele. However, additional factors are needed for coeliac disease to manifest besides the HLA risk alleles. Furthermore, around 5% of those people who do develop coeliac disease may not have typical HLA-DQ2 or HLA-DQ8 alleles.

Clinical features

Clinical features may range from severe to almost non-existent. Severe coeliac disease in young children leads to the characteristic symptoms of pale, loose and greasy stools

(steatorrhea) with weight loss or failure to gain weight. Adolescents and older children with milder coeliac disease may have symptoms that are much more subtle and occur in other organs rather than in the bowel itself.

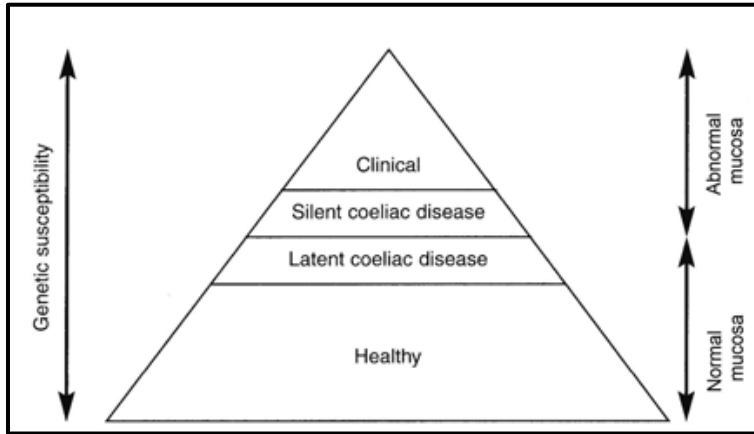


FIGURE 29.1 The spectrum of coeliac disease. Latent coeliac disease: positive anti-endomysial antibodies, normal small bowel, but risk of developing disease

TABLE 29.2 Clinical features of coeliac disease

Under 2 years of age	Over 2 years of age
Steatorrhea	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

Diagnosis

The diagnosis of coeliac disease is based on two types of testing.

Serological blood tests

It is important that gluten has not been excluded from the diet when performing these tests as they will return to normal.

These are the first-line investigation and include the following:

- IgA anti-tissue transglutaminase (tTG) antibodies: this test is reported to have a high sensitivity (99%) and specificity (over 90%) for identifying coeliac disease. Therefore, it should be done first. It is also an easier test to perform. An equivocal result on tTG testing should be followed by antibodies to endomysium.

- IgA anti-endomysial antibodies: this test has a sensitivity and specificity of 90% and 99%, respectively, for detecting coeliac disease.

It is important that the total serum IgA level is also checked, as coeliac patients with IgA deficiency may be unable to produce the antibodies on which these tests depend ('false-negative'). In such patients, IgG antibodies against transglutaminase (IgG-tTG) or IgG anti-gliadin antibodies (IgG-AGA) may be helpful in reaching a diagnosis

A tTG-IgA serum titre of x10 the upper limit of normal and positive endomysial antibodies is sufficient to make the diagnosis of Coeliac Disease without a biopsy.

Dudeno-jejunal biopsies

Because of the implications of a diagnosis of coeliac disease, guidelines recommend that a positive serological test may still be followed by a biopsy. Similarly, a negative serology may still be followed by a recommendation for a biopsy if clinical suspicion remains high.

Tissue biopsy is still considered the gold standard in the diagnosis of coeliac disease. For this purpose, biopsies can be obtained using metal capsules attached to a suction device. The capsule is swallowed and allowed to pass into the small intestine. After X-ray verification of its position, suction is applied to collect part of the intestinal wall inside the capsule. Commonly used capsule systems are the Watson capsule and the Crosby–Kugler capsule. This method has now been largely replaced by fibre-optic endoscopy, which carries a higher sensitivity and a lower frequency of errors.

TABLE 29.3 Investigations for malabsorption

General	Specific
Full blood count plus film Serum iron and ferritin Folate (red blood cell) levels Vitamin B12 Serum albumin Hydrogen breath test Serum calcium, phosphate and alkaline phosphatase Serum T4 and TSH	Immunoglobulins IgA and IgG IgA tTG antibodies IgA anti-endomysial antibodies IgA anti-gliadin (AGA) Small bowel biopsy: Villous atrophy on biopsy Hyperplasia of crypts on biopsy Increased inflammatory cells on biopsy

There are several ways in which these tests can be used to assist in diagnosing coeliac disease. However, all tests become invalid if the patient is already taking a gluten-free diet. Intestinal damage begins to heal within weeks of gluten being removed from the diet, and antibody levels decline over a period of months. In such cases, it may be necessary to perform a re-challenge with gluten-containing food over 2–6 weeks before repeating the investigations. A histology compatible with coeliac disease on a gluten-containing diet, followed by a clinical improvement (i.e., gain in weight and height and resolution of symptoms) once the gluten is removed from the diet is often enough to establish the diagnosis. Most guidelines do not recommend a repeat biopsy unless there is no improvement in the symptoms on the gluten-free diet. In some cases, a deliberate gluten challenge, followed by biopsy, may be conducted to confirm, or refute the diagnosis. A normal biopsy and normal serology after the challenge indicates that the diagnosis may have been incorrect. In resource-limited countries where facilities for biopsies may not exist, the same model can be used with serological tests. A positive serological test on a gluten-containing diet will revert to normal with clinical improvement once the patient is on a gluten-free diet.

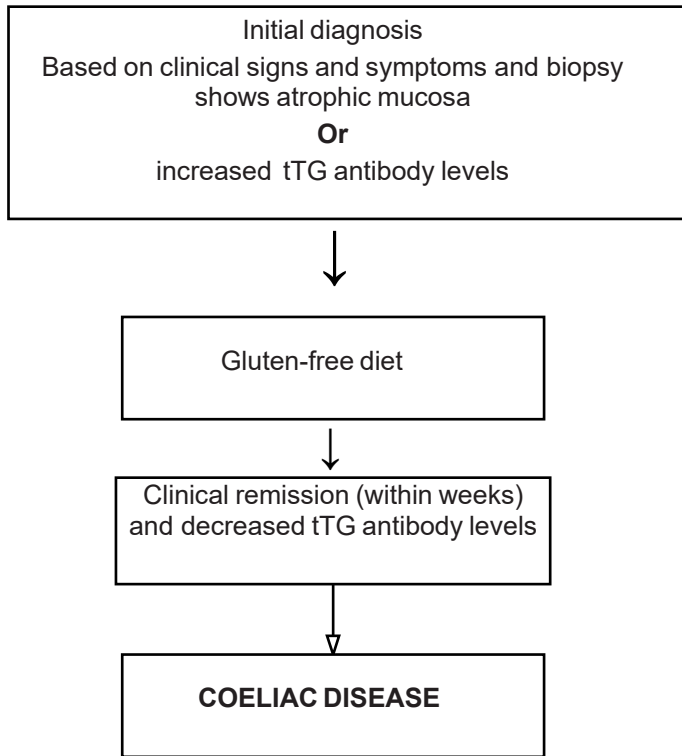


FIGURE 29.2 Diagnosis of coeliac disease

Treatment

At present, the only effective treatment is a lifelong gluten-free diet. Strict compliance allows the intestines to heal, with resolution of symptoms in most cases. Early intervention and good compliance can eliminate the heightened risk of intestinal cancer and in some cases sterility. Dietitian input can be helpful in ensuring that the patient is aware of which foods contain gluten, which foods are safe, and how to have a balanced diet despite the limitations. The diet can be cumbersome, and failure to comply may cause relapse.

The commonly implicated cereals include wheat (and its subspecies, such as spelt, semolina and durum), barley, rye, and oats. Other cereals, such as maize (corn), millet, sorghum, teff and rice, are safe for patients to consume. Similarly, non-cereal foods such as fruits, vegetables and foods derived from animal sources (i.e., milk, fish, poultry and other meat) can be used.

Further reading

European Society for Pediatric Gastroenterology, Hepatology, and Nutrition Guidelines for the Diagnosis of Coeliac Disease 2020: <https://pubmed.ncbi.nlm.nih.gov/31568151/>
Accessed 8th April 2021

Section 30. American Trypanosomiasis (Chagas Disease)

Introduction

- American trypanosomiasis is potentially life-threatening and is caused by the protozoan parasite, *Trypanosoma cruzi*.
- An estimated 6-7 million people are infected worldwide, mostly in Latin America, where it is endemic.
- In 2008 it killed more than 10 000 people.
- It is increasingly being detected in the USA, Canada, many European and some Western Pacific countries.
- In Latin America, *T. cruzi* is **mainly transmitted by the infected faeces of blood-sucking triatomine bugs.**
 - These bugs typically live in the cracks of poorly constructed homes in rural or suburban areas.
 - They become active at night when they feed on human blood by biting an exposed area of skin such as the face, where the bug defecates close to the bite.
 - The parasites enter the body when the person unwittingly smears the bug faeces into the bite, the eyes, the mouth, or any break in the skin.
- *T. cruzi* **can also be transmitted in the following ways:**
 - Via food and drinks contaminated with the parasite through, for example, contact with triatomine bug faeces. Oral ingestion is associated with more severe disease.
 - By blood transfusions from infected donors
 - By transmission from an infected mother to her newborn during pregnancy or childbirth.

Clinical management

Signs and symptoms

The disease presents in two phases.

- The initial **acute phase** lasts for about 1-2 months after infection.
- In less than 50% of people bitten by a triatomine bug, the characteristic first visible signs can be an inflamed skin lesion (Chagoma) or a purplish swelling of the lids of one eye (Romana's sign).
- During the acute phase, a high number of parasites circulate in the blood.
- In most cases, symptoms are absent or mild, but can include fever, headache, enlarged lymph glands, pallor, muscle pain, difficulty in breathing, swelling and abdominal or chest pain.
- Severe acute disease occurs in < 5% of patients and includes acute myocarditis, pericardial effusion, and meningoencephalitis.
- An asymptomatic 'indeterminate' phase follows the acute phase. This may be life-long or progress to chronic disease after 10 years or more.
- In the **chronic phase**, pathology is caused by parasites congregated in the heart and gastrointestinal tract.
- Between 14 – 45% of patients with chronic infection develop cardiovascular disorders (arrhythmias, heart failure, emboli) and up to 10 – 21 % develop gastrointestinal complications (motility disorders and dilatation of parts of the gastrointestinal tract, commonly the oesophagus or colon), neurological or mixed pathology.

Investigations

- Acute and congenital infection: Giemsa-stained blood films to identify trypanosomes.
- Indeterminate and chronic phase: Serology (ELISA, IFA, immunoblot); PCR; xenodiagnosis.

Treatment

Benznidazole and nifurtimox are both almost 100% effective in curing the disease if given soon after infection. However, the efficacy of both diminishes the longer a person has been infected. Treatment is also indicated for those in whom the infection has been reactivated (e.g. due to immunosuppression), for infants with congenital infection, and for selected patients during the indeterminate phase, including women of child-bearing age who intend to conceive at in the future. The potential benefits of medication in preventing or delaying the disease should be weighed against the long duration of treatment (up to 2 months) and possible adverse reactions (occurring in up to 40% of treated patients).

- Benznidazole and nifurtimox should not be taken by pregnant women or by people with kidney or liver failure.
- Nifurtimox is also contraindicated in people with a history of neurological or psychiatric disorders.
- In addition, specific treatment for cardiac or gastrointestinal manifestations may be required.

Benznidazole 100 mg tablets

Acute or indeterminate phase:

- Newborn infant:
 - 5 – 7 mg/kg daily divided in 2 doses (or once daily for low-birthweight babies).
 - Treat for 60 days.
- Infant or child < 12 years
 - 10 mg/kg daily divided in 2 doses for 60 days.
- Child > 12 years
 - 5 – 10 mg/kg daily divided in 2 doses for 60 days.

Indeterminate phase:

- Infant or child
 - 5 – 7.5 mg/kg daily divided in 2 doses for 60 – 90 days.

Nifurtimox Tablets 30, 120 and 250 mg

Acute or indeterminate phase (given after meals)

- Newborn infant:
 - 10 – 15 mg/kg daily divided in 3 doses (or once daily for low-birthweight babies).
 - Treat for 60 days.
- Infant or child < 12 years
 - 15 mg/kg daily divided in 3 doses for 60 days.
- Child > 12 years
 - 10 - 15 mg/kg daily divided in 3 doses for 60 days.

Indeterminate phase:

- Infant or child
 - 8 – 10 mg/kg daily divided in 3 doses for 60 – 90 days.

Vector control and prevention

- There is no vaccine for Chagas disease.
- Vector control is the most effective method of preventing this disease in Latin America.
- Blood screening is necessary to prevent infection through transfusion/transplantation.

The WHO recommends:

1. Insecticide spraying of houses and surrounding areas

Section 30. American Trypanosomiasis [Dr. Tim O'Dempsey](#)

2. House improvements to prevent vector infestation
3. Personal preventive measures such as bed nets
4. Good hygiene practices in food preparation, transportation, storage, and consumption
5. Screening of blood donors
6. Screening of newborn infants from infected mothers, and of siblings of infected children to provide early diagnosis and treatment.

Reference

[World Health Organization \(2019\) Guidelines for the diagnosis and treatment of Chagas disease](#) (accessed 04/03/2021)

Section 31. Buruli ulcer

Introduction

Buruli ulcer is a highly destructive ulcerating condition caused by *Mycobacterium ulcerans*, which ranks third among mycobacterial infections affecting immunocompetent humans.

Any part of the body may be affected, particularly areas exposed to minor trauma.

Management is a combination of medical treatment to eradicate the infective agent and surgery to excise the infected tissue.

Background and epidemiology

The disease occurs in several parts of Africa, Papua New Guinea, the Americas, South East Asia, Japan, China and Australia. The organism is found in soil or stagnant water and predominantly affects children in whom infection usually occurs following a minor penetrating injury.

Intercurrent helminthic infections may also predispose to ulceration. HIV infection, and other immunodeficiency states, can exacerbate Buruli ulcer and lead to severe complications.

Clinical features

A non-ulcerative lesion usually precedes ulceration. Four non-ulcerative presentations are recognised:

Papule: painless, may be itchy, non-tender intradermal lesion

Nodule: painless firm lesion 1–2 cm diameter in the subcutaneous tissue, usually attached to the skin

Plaque: painless well-demarcated elevated dry indurated lesion > 2 cm in diameter

Oedematous: diffuse extensive non-pitting swelling, ill-defined margin, firm, usually painful, with or without colour change over affected skin.

Subsequently an ulcer forms with central necrosis and often spreads very rapidly in all directions.

Characteristic features

- The ulcer is usually painless (hence delay in healthcare-seeking behaviour).
- Edge of ulcer is deeply undermined.
- Satellite ulcers often communicate with the original ulcer by subcutaneous tunnels.
- Skin between adjacent ulcers is often unattached to the underlying tissues.
- The extent of damage is always greater than it appears from the surface.
- Regional adenitis and systemic symptoms are unusual (and if present suggest primary or secondary bacterial infection).
- Erosion of underlying tissue may involve nerves, blood vessels and bone (in up to 15% of cases).

Complications

These include the following:

- tetanus
- osteomyelitis
- scarring
- ankylosis
- contractures.

Around 25% of patients develop long-term complications that may include amputation or loss of sight.

Differential diagnosis

1. *Papule*: granuloma annulare, herpes, insect bites, Leishmaniasis, acne, pityriasis, psoriasis.
2. *Nodule*: boil, cyst, Leishmaniasis, lipoma, lymphadenitis, mycosis, onchocerciasis.
3. *Plaque*: cellulitis, haematoma, insect bites, leishmaniasis, leprosy, mycosis, psoriasis.
4. *Oedema*: actinomycosis, cellulitis, Elephantiasis, necrotising fasciitis, onchocerciasis, osteomyelitis.
5. *Ulcer*: cutaneous diphtheria, guinea worm, Leishmaniasis, necrotising fasciitis, neurogenic ulcer, tropical ulcer, tuberculosis, sickle-cell disease, squamous-cell carcinoma, syphilis, venous ulcer, cutaneous amoebiasis, yaws.

Investigations

- Slough from ulcer usually contains numerous acid-fast bacilli on Ziehl–Neelsen stain (sensitivity 40%).
- Culture unhelpful (sensitivity 20–60%, time consuming (8 weeks), expensive, frequently gives false-positive results).
- Biopsy and histopathology (sensitivity is 90%).
- Polymerase chain reaction (PCR) is increasingly used in diagnosis (it is rapid, only taking 2 days, and has a sensitivity of more than 95%). Recently, a highly sensitive dry reagent-based PCR assay has been developed that is better suited for use in most endemic countries.

Management

The current recommendation is for combined medical and surgical management.

- 1 For small early lesions (e.g., nodules, papules, plaques, ulcers no more than 5 cm in diameter). Current standard recommendation - if immediate excision and suturing is possible, start antibiotics at least 24 hours before surgery and continue for 4 weeks. Otherwise, treat all lesions in this category with antibiotics for 8 weeks.
- 2 Non-ulcerative and ulcerative plaque and oedematous form: large ulcerative lesions (> 5 cm in diameter): lesions in the head and neck region, particularly the face: treat with antibiotics for at least 4 weeks, then surgery (if necessary), followed by another 4 weeks of antibiotics.
- 3 Disseminated/mixed forms (e.g., osteitis, osteomyelitis, joint involvement): treat with antibiotics for at least 1 week before surgery and continue for a total of 8 weeks.

Necrotic ulcers should be excised with care to remove all affected tissue by extending the margin into healthy tissue. Excision is followed by primary closure or split-skin grafting. Reconstructive surgery and physiotherapy may be required for patients with contractures and other permanent disabilities and disfigurements.

Antibiotics

Antibiotic combination treatment, by reducing ulcer size, makes larger ulcers more amenable to surgery and grafting.

Recommended treatment: Oral rifampicin (10 mg/kg once daily) plus clarithromycin (immediate-release preparation 7.5 mg/kg twice daily or extended-release preparation 15 mg/kg once daily) for 8 weeks.

Alternative treatment is with a combination of oral rifampicin (10 mg/kg once daily) and oral moxifloxacin (10 mg/kg once daily, maximum 400 mg).

Recent development: Philipps et al published a recent Randomised Controlled Trial showing that early, limited solitary lesions including ulcers (10cm or less in diameter) can

be treated with 8 weeks of rifampicin 10 mg/kg plus oral clarithromycin 15 mg/kg extended release once daily, with 96% resolution rate. The previous standard of treatment, which included streptomycin injections was no better than the above oral treatment. No patients in this study needed surgical treatment.

Prevention and public health aspects

- Long trousers and other mechanical barriers.
- BCG offers some protection.

The *Global Buruli Ulcer Initiative*, launched by the WHO in 1998, advocates the following:

1. Health education and staff training in the communities most affected
2. Development of educational materials adapted to the needs of the countries
3. Community-based surveillance system to increase early detection and referral for treatment in collaboration with diseases such as leprosy and Guinea worm
4. Assessment of local health services and resources currently available for the diagnosis and treatment of Buruli ulcer in endemic areas
5. Strengthening of the capacity of health systems in endemic areas by upgrading surgical facilities and improving laboratories
6. Rehabilitation of those already deformed by the disease.

Further reading Phillips RO et al. Rifampicin and clarithromycin (extended release) versus rifampicin and streptomycin for limited Buruli ulcer lesions: a randomised, open-label, non-inferiority phase 3 trial. *Lancet* 2020; 395; P1259-1267

[https://www.thelancet.com/pdfs/journals/lancet/PIIS0140-6736\(20\)30478-5.pdf](https://www.thelancet.com/pdfs/journals/lancet/PIIS0140-6736(20)30478-5.pdf)

(accessed 04/03/2021)

[World Health Organization \(2012\). Treatment of Mycobacterium ulcerans disease \(Buruli ulcer\): guidance for health workers.](#) (accessed 04/03/2021)

Section 32. Taeniasis/Cysticercosis

Introduction

Three tapeworms can infect humans but only *Taenia Solium* causes major health problems (cysticercosis). It occurs in Africa, Asia and Latin America largely in poor communities where it is calculated to be the leading cause of deaths from food-borne diseases and according to WHO results in around 2.8 million disability-adjusted life years (DALYs).

T. solium infection

This results from the eating of larval cysts (cysticerci) in undercooked infected pork (resulting in intestinal tapeworm), or ingesting eggs from contaminated water or food (resulting in larval infection of tissues, known as 'cysticercosis'). Patients harbouring the adult tapeworm in their gut excrete the tapeworm eggs (and sometimes segments) in their faeces. Defaecation away from toilets contaminates the environment including water supplies. Living in close proximity to roaming pigs is the major risk factor for propagating transmission. Person to person transmission of eggs may occur via contaminated hands, for example when preparing food. Auto-infection may occur when eggs from the anus are transferred to the mouth via contaminated fingers. Rarely, auto-infection may occur because of vomiting causing regurgitation of eggs from the intestine into the stomach.

Symptoms

T. solium produces only mild and non-specific symptoms (abdominal pain, nausea, diarrhoea, or constipation) when the tapeworm becomes fully installed in the bowel; usually 8 weeks after ingesting meat containing cysticerci. If untreated, intestinal tapeworms can persist for up to 25 years

The larvae (cysticerci) may develop within the muscles, skin, eyes and the central nervous system.

Cysts may be visible or palpable as subcutaneous nodules or may be evident on imaging of tissues.

The most dangerous consequence of *T. solium* infection relates to the larvae (cysticerci) invading the brain (called neurocysticercosis). Symptoms and signs include severe headaches, blindness, epileptic seizures, and death. Neurocysticercosis is the commonest preventable cause of epilepsy worldwide. Where the disease is endemic, it can cause 30 to 70% of all epilepsy. Stigma from the infection has been attributed to witchcraft.

Investigation

Faecal screening tests such as Kato-Katz that are used for other diseases (e.g., soil-transmitted helminths), can also be used to identify *Taenia* eggs and hence areas in which the parasite may be endemic. Faecal ELISA and PCR techniques have also been developed.

A point of care test for the diagnosis of *T. Solium* is urgently needed for children and their families. Similar tests that can be used on pigs could also be valuable in tracing and tracking cases.

Prevention and Treatment

Public health interventions including the role of veterinary health, human health and environmental activities are vital. Protocols for mapping (identifying the endemic or high-risk areas where *T. solium* is endemic) have been developed by WHO. Partnerships between WHO, The World Organisation for Animal health (OIE) and the Food and Agricultural organisation of the UN (FAO) are vital.

The following approaches to prevention have been proposed:

1. Core rapid impact interventions in pigs: treatment of taeniasis in pigs with Oxfendazole and vaccination with TSOL18 vaccine
2. Core rapid impact interventions in infected humans
3. Community health education, including hygiene and food safety plus improved sanitation and the ending of open defaecation.
4. Improved pig husbandry - no free-roaming pigs; and improved meat inspection and processing of meat products.

Individual treatment of infected children and their families with T. Solium

Intestinal Tapeworm of Taenia Solium

Praziquantel (one dose can be effective)

Child 4 to 17 years 5 to 10 mg/Kg for 1 dose taken after a light breakfast (tablets of 150mg, 500mg and 600 mg are available).

Albendazole

Child 4 to 17 years 400 mg twice daily for 3 consecutive days.

Niclosamide

Adults and children over 6 years one dose of 2 grams orally

Children aged 2 to 6 years one dose of 1 gram orally

Treatment of neurocysticercosis

There is controversy about the risks / benefits of antiparasitic treatment. The consensus is that treatment will benefit some patients with neurocysticercosis. A recent meta-analysis indicated that treatment with cysticidal drugs improved resolution of enhancing lesions and cysts, lowered risk of recurrence of seizures in patients with enhancing lesions and reduced the rate of generalized seizures in patients with viable cysts.

Therefore, a full course antihelminthic therapy is now recommended for patients with active parenchymal neurocysticercosis. Ocular involvement should be excluded prior to commencing antihelminthic therapy.

Patients who have active parenchymal neurocysticercosis should be managed as follows:

- Patients with 1-2 brain enhancing lesions: albendazole 15mg/kg/day (maximum 1200 mg / day) in two divided doses for 10 days.
- Patients with > 2 cysts: albendazole (as above) plus praziquantel 50 mg/kg/day in three divided doses for 10 days.

Repeated courses may be necessary. Cimetidine may be used to increase the levels of both albendazole and praziquantel. Prednisolone 1 mg/kg/day or dexamethasone 100 to 200 microgram/kg/day should be given before and during antiparasitic treatment to reduce inflammation around damaged cysts. Steroids are also needed for management of occasional flare-ups of inflammation and cerebral oedema that may occur as cysts spontaneously degenerate.

Inactive parenchymal cysts do not require antiparasitic treatment. Seizures usually respond to first-line anticonvulsant drugs (see Sections 69 and 70 in Handbook 1).

Surgical intervention, for example shunting, may be necessary for obstructive hydrocephalus and intracranial hypertension. If available, neuro-endoscopic extraction is now recommended for intraventricular cysts.

Extra-parenchymal cysts, depending on number, size and location, may require repeated courses of treatment with combinations of antiparasitic drugs, steroids and, possibly, surgery.

Caution when prescribing anti-helminthics in areas with a high prevalence of neurocysticercosis.

Problems may occur in cases of undiagnosed neurocysticercosis when prescribed albendazole or praziquantel for example in community helminth / schistosomiasis control programmes. Neurological symptoms may be precipitated by a single dose of either of these drugs. As routine screening is unlikely to be possible, patients should be advised to seek medical advice urgently if they develop neurological symptoms (usually within a week of treatment).

Section 33. Dracunculiasis (Guinea-Worm Disease)

Introduction

Guinea-worm disease is transmitted exclusively by drinking stagnant water contaminated with tiny water fleas (Cyclops species) that carry infective guinea-worm larvae. Once ingested, the larvae mature into worms, growing up to 1 metre in length. Humans were believed to be the only known reservoirs for the disease. However, recently infection has also been described in dogs, cats, and baboons.

About 1 year after infection, a very painful blister forms, in 90% of cases on the lower leg, and the female worm begins to emerge accompanied by a burning sensation. To soothe the burning pain, patients often immerse the affected area in water. The worm then releases thousands of larvae into the water, contaminating the water and bringing the life cycle full circle.

Epidemiology

- The main source of infection is stagnant water sources such as ponds and sometimes shallow or step wells.
 - 'Man-made' ponds are the main source of transmission.
- Only 5 African countries (Angola, Chad, Ethiopia, Mali, and South Sudan) remain endemic with the majority of cases in South Sudan.
- Guinea-worm disease is seasonal, occurring with two broad patterns found in endemic areas of Africa, depending on climatic factors.
 - In the Sahelian zone, transmission generally occurs in the rainy season (from May to August).
 - In the humid savanna and forest zone, the peak occurs in the dry season (from September to January).
- There are thousands of village volunteers in the remaining endemic countries who are trained to find new cases, take care of them and report them to the area supervisor.

Clinical effects

- Once a new case is identified, the wound must be disinfected and bandaged to help to prevent secondary infection.
 - The worm should be gently pulled out a few inches every day until all of it has been removed. The usual technique is to gently wind the worm onto a matchstick or similar.
 - Oral mebendazole or metronidazole may facilitate extraction.
 - Antitetanus toxoid vaccine should be offered if non-immune.
- Many patients are unable to leave their beds for a month after the emergence of the worm.
- Guinea-worm disease is not fatal, but infected people cannot work or attend school for months.
- Since the peak transmission period often coincides with the agricultural season, fields are left untended and food production declines.
- In Mali, guinea-worm disease is called 'the disease of the empty granary'.
- As adults lie sick, older children must take on the household chores and miss months of schooling.
- Younger children may miss vital vaccinations.

Prevention

- Effective surveillance to detect all cases within 24 hours of worm emergence.
- Ensure access to safe drinking water and convert unsafe sources to safe ones.
- Construction of copings around well heads or installation of boreholes with hand pumps.

- There must be regular and systematic filtering of drinking water derived from ponds and shallow unprotected wells, or from surface water.
- Fine meshed cloth or, better still, a filter made from a 0.15mm nylon mesh, is all that is needed to filter out the Cyclops species from the drinking water.
- Treatment of unsafe water sources with Temephos to kill the Cyclops species.
- Health education and social mobilisation to encourage affected communities to adopt healthy behaviour with regard to use of drinking water.
- In the case of infected dogs, tether the dog to prevent access to ponds or water sources until all worms are expelled or removed.

Section 34. Fascioliasis (Liver Fluke Infections)

Introduction

- This disease is caused by *Fasciola hepatica* and *Fasciola gigantica* and occurs in sheep- and cattle-rearing areas worldwide, especially South America.
- Freshwater snails act as the intermediate amplifying hosts, liberating free-swimming cercariae which encyst as metacercariae on water plants.
- Humans are infected following ingestion of metacercarial cysts on raw aquatic plants (e.g., watercress) or from contaminated water.
- Following ingestion, the larvae emerge in the duodenum, penetrate the intestinal wall, migrate via the peritoneal cavity to the liver, penetrate the liver capsule, and after 3 - 4 months mature into adults in the bile ducts.

Clinical features

- Infections may be asymptomatic.
- Acute presentations occur 6 - 12 weeks after infection.
- Fluke migration may be associated with fever, malaise, abdominal pain, weight loss, urticaria, cough and wheeze.
- In chronic presentations, symptoms may be minimal or may be due to recurrent cholangitis, intermittent biliary obstruction, or anaemia.
- Ectopic flukes may cause granuloma or abscess formation in various organs, and also present as migrating skin nodules.

Investigations

- Eosinophilia is common.
- Liver ultrasound is often normal.
- CT of the liver (if available) may reveal hypodense lesions.
- Serology may be helpful in established *F. hepatica* infections but is less reliable for *F. gigantica*. Fasciola excretory–secretory (FES) antigen detection in faeces is available for *F. hepatica*.
- In established infections, eggs may be found in faeces.

Treatment

1. Triclabendazole is the drug of choice for *F. hepatica* and *F. gigantica* infections.
2. One dose of 10 mg/kg taken with food is usually effective but should be repeated after 12 hours in severe infections.
3. Expulsion of dead or damaged flukes may cause biliary colic 3 - 7 days after treatment; the colic responds well to antispasmodics. Triclabendazole resistance has been reported in Ireland, the UK and Australia.
4. Bithional, 30–50 mg/kg/day in three divided doses on alternate days for 10–15 days was the preferred treatment previously. Side effects include mild gastrointestinal upset and pruritus.
5. Nitazoxanide may be effective.
6. Praziquantel is unreliable in the treatment of fascioliasis.

Prevention and control

- Avoid potentially contaminated watercress and other aquatic plants.
- Treat herbivores.
- Undertake snail control.

Further reading

[Triclabendazole in the treatment of human fascioliasis: a review, Transactions of The Royal Society of Tropical Medicine and Hygiene, Volume 113, Issue 12, December 2019, Pages 797–804](#) Accessed 9th April 2021

Section 35. Lymphatic Filariasis

Introduction

This painful and profoundly disfiguring disease is usually acquired in childhood. The disease is caused by three species of thread-like nematode worms, known as filariae, namely *Wuchereria bancrofti*, *Brugia malayi* and *Brugia timori*. Around 90% of infections are caused by *Wuchereria bancrofti* and most of the remainder by *Brugia malayi*. About 120 million people are affected worldwide (of whom 60% live in South-East Asia and 30% live in Africa).

Life cycle of filariae

1. Filariae are transmitted by mosquitoes.
2. When a mosquito with infective-stage larvae takes a blood meal, the parasites are deposited through the person's skin, from which they enter the body.
3. These larvae then migrate to the lymphatic vessels and develop into adult worms over a period of 6 - 12 months, causing damage to and dilatation of the lymphatic vessels.
4. The adult filariae live for several years in the human host.
5. During this time, they produce millions of immature microfilariae that circulate in the peripheral blood and are ingested by mosquitoes that bite the infected human.
6. The larval forms further develop inside the mosquito before becoming infectious to humans.
7. Thus, a cycle of transmission is established.
8. Threadlike adult worms of *Wuchereria bancrofti* live in the lymphatics (groin, scrotum, arm).
9. Male worms are about 3 - 4 cm in length, and female worms 8 - 10 cm.
10. The male and female worms together form 'nests' in the lymphatic system.
11. Females release thousands of microfilariae into the peripheral blood periodically every day, synchronising with the biting habits of the predominant local mosquito vector. Nocturnal periodicity is commonest, except in some Polynesian islands where microfilariae are more numerous by day.
12. *Brugia malayi* has two main forms: the nocturnal periodic form in swampy areas from India to Korea and Japan, and the nocturnal sub-periodic form in the damp forests of South-East Asia.
13. The parasites of *B. malayi* are transmitted by various species of the genus *Mansonia*, and in some areas anopheline mosquitoes are responsible for transmitting infection. Brugian parasites are confined to areas of East and South Asia, notably India, Indonesia, Malaysia, and the Philippines.
14. An estimated 120 million people in tropical and sub-tropical areas are infected, of whom almost 25 million men have genital disease (most commonly hydrocoele) and almost 15 million, mostly women, have lymphoedema or elephantiasis of the leg.

Diagnosis

- Eosinophilia is common in the acute stages.
- Examination of thick smears of 20 - 60 microlitres of blood from a fingertip or filtration of 1 mL of intravenous blood and examination of the filtrate can reveal the microfilariae provided that the concentration is high (> 100 microfilariae/mL).
- Concentration techniques can improve sensitivity (e.g., Nuclepore filtration). Samples should be appropriately timed (usually between 22.00 and 02.00 hours for *W. bancrofti*).
- A variety of more sensitive diagnostic techniques are now available, including complement fixation tests for circulating *W. bancrofti* antigen (e.g., an ELISA 'TropBio-test') and a rapid finger-prick immunochromatographic card test (Amrad ICT, Binax).
- The rapid ICT has a high sensitivity and specificity and is currently the preferred diagnostic test for *W. bancrofti*.
 - It is also used for monitoring the success of mass drug programmes.

- The test requires 100 microlitres of finger-prick blood drawn at any time, day or night.

Clinical features

The majority of infected people are asymptomatic, but virtually all have subclinical lymphatic damage, and up to 40% have kidney damage, with proteinuria and haematuria. Inflammatory episodes associated with lymphatic filariasis involve:

- Responses to the parasite itself
 - The effects of secondary bacterial infection
 - Sometimes inflammatory mediators associated with endosymbiotic bacteria (Wolbachia).
- Endosymbiotic bacteria infect most species of filarial nematodes that are pathogenic to humans (a notable exception being *Loa loa*) and contribute to the damage done by the filaria.
 - Further characterisation of the Wolbachia–nematode relationship might allow the development of new therapeutic approaches to these parasitic diseases.
 - Acute episodes of local inflammation involving the skin, lymph nodes and lymphatic vessels often accompany chronic lymphoedema or elephantiasis (see below).
 - Some of these episodes are caused by the body's immune response to the parasite, but many are the result of bacterial skin infections, linked to the partial loss of the body's normal defences because of underlying lymphatic damage.
 - In chronic disease, careful cleansing is extremely helpful in healing the affected areas and in both slowing and possibly reversing much of the damage that has already occurred.

Acute symptoms may recur several times a year in three forms:

1. Acute filarial fever without lymphadenitis.
2. Acute filarial lymphangitis (AFL) follows the death of an adult worm, causing an inflammatory nodule or cord with lymphangitis spreading away from the affected node. This is usually mild but may develop into an abscess.
3. Acute dermatolymphangioadenitis (ADLA) resembles cellulitis or erysipelas and is often associated with secondary bacterial infection and impaired lymphatic flow, ascending lymphangitis, and limb oedema. ADLA is more common than AFL and is an important cause of lymphoedema and elephantiasis.

Chronic lymphatic filariasis may develop over months or years even without a history of acute symptoms. Lymphatic obstruction eventually leads to elephantiasis, most commonly affecting the legs, scrotum, arms and breast. Recurrent secondary bacterial skin infections (often streptococcal) cause acute pain and fever and may be complicated by acute glomerulonephritis.

Other presentations of lymphatic filariasis include:

- Hydrocoele, usually unilateral
- Swelling of the scrotum
- Acute epididymitis
- Funiculitis (inflammation of the spermatic cord)
- Monoarthritis
- Glomerulonephritis
- Chyluria, chylous diarrhoea, chylous ascites (due to rupture of dilated lymphatics). (Malabsorption of fat-soluble vitamins may complicate chylous diarrhoea.)

Brugian filariasis is usually less severe than Bancroftian filariasis.

- The most severe symptoms generally appear in adults, and in males more often than in females.
- In endemic communities, around 10 - 50% of men suffer genital damage (hydrocoele and elephantiasis of the penis and scrotum).
- Elephantiasis of the entire leg or arm, the vulva and the breast may affect up to 10% of adults.
- In endemic areas, chronic and acute manifestations of filariasis tend to develop more often and sooner in refugees or newcomers than in local populations.
- Lymphoedema may develop within 6 months, and elephantiasis as soon as 1 year after arrival.

Tropical pulmonary eosinophilia (TPE)

- A hypersensitivity response to microfilariae in the lungs can develop in some patients, causing cough and wheeze, especially at night.
- There may also be an enlarged liver, spleen and lymph nodes.
- Chest X-ray may show diffuse miliary shadows.
- Untreated TPE may progress to irreversible lung fibrosis.
- The condition is usually associated with high eosinophilia and high microfilaria titres. Microfilariae are usually absent from peripheral blood, but the rapid antigen test is usually positive.

Treatment and control of filariasis

Several antihelminthic agents are effective, although care must be taken in the choice of antihelminthic depending on the risk of co-infection with onchocerciasis and/or *Loa loa*.

Mass drug administration is an important strategy in community control. The treatment and control options are summarised in Table 35.1.

TABLE 35.1 Recommended treatment strategies for mass drug distribution, individual drug administration, and morbidity control and treatment of lymphatic filariasis

Mass drug administration		Individual drug administration	Morbidity control and treatment
Africa	Rest of world		
		DEC (with or without ALB) 6 mg/kg single dose ¹ OR DEC 12-day course of 6 mg/kg per day in two or three divided doses OR Doxycycline 200 mg/day for 4 weeks followed by one dose of IVM OR Alternative treatment for <i>Wuchereria bancrofti</i> (if risk of Loa Loa co-infection): Doxycycline 200 mg/day for 30 days plus ALB 400 mg/day for final 7 days.	 <i>Lymphoedema:</i> <ul style="list-style-type: none"> • Hygiene, physiotherapy, • Doxycycline 200 mg/day for 6 weeks <i>Hydrocoele:</i> <ul style="list-style-type: none"> • Surgical hydrocoelectomy, • Doxycycline 200 mg/day for 6 weeks <i>Tropical pulmonary eosinophilia:</i> <ul style="list-style-type: none"> • Doxycycline 200 mg/day for 4 weeks followed by one dose IVM
IVM + ALB for at least 5 years	DEC + ALB for at least 5 years		

Based on Taylor M et al. (2010) Lancet, 376, 1175–85.

¹ If the patient continues to live in an endemic area or is less than 8 years of age (contraindication of doxycycline).

- ALB = Albendazole; DEC = diethylcarbamazine (omit if there is onchocerciasis co-infection or a risk of serious adverse events with Loa loa); IVM = ivermectin (omit if there is a risk of serious adverse events with Loa loa).

- Doxycycline: the doses above are suitable for children aged ≥8 years and weighing > 45 kg. Children aged ≥8 years but weighing < 45 kg should receive 4.4 mg/kg/day. Doxycycline should not be used for children < 8 years.

Warning

DEC should be avoided in areas endemic for onchocerciasis because of the risk of provoking a Mazzotti reaction or encephalopathy. Mazzotti reactions can be life-threatening and are characterized by fever, urticaria, swollen and tender lymph nodes, tachycardia, hypotension, arthralgias, edema and abdominal pain that occurs within 7 days of treatment of filariasis.

DEC and IVM should be avoided / used with caution in areas endemic for Loa loa because of the risk of provoking encephalopathy in individuals with Loa loa microfilaraemia > 2500 Mf/ml.

Treating individuals with filariasis

- Most problems result from bacterial and fungal ‘superinfection’ of tissues, linked to compromised lymphatic function caused by earlier filarial infection.
- Antibiotics against streptococcal and other bacterial infections are important
- Surgical procedures are available to correct hydrocoele.
- Because secondary bacterial infections play an important role in precipitating acute adeno-lymphangitis episodes and progression of lymphoedema, simple hygiene (either

alone or in combination with antibiotic treatment) plays an important role in preventing episodes of acute disease and in the management of lymphoedema.

- Daily washing of affected limbs with soap and safe water to prevent secondary infection, combined with simple exercises, elevation of the limb, and treatment of cracks and entry points, provides significant relief from acute episodes and slows progression of the disease.

Treating endemic communities who have filariasis (see Table 35.2)

- The goal is to eliminate microfilariae from the blood of infected individuals in order to interrupt the cycle of transmission by mosquitoes.
- A single dose of diethylcarbamazine citrate (DEC) has the same long-term (1-year) effect in decreasing levels of micro-filaraemia as the formerly recommended 12-day regimen of DEC.
- More importantly, the use of single doses of two drugs administered together (optimally albendazole with DEC or ivermectin) is 99% effective in removing microfilariae from the blood for a full year after treatment.
- There are new data from Papua New Guinea showing the effectiveness of the combination of Ivermectin + Albendazole + DEC (“IDA”)

Table 35.2. Summary of MDA for elimination of LF in areas co-endemic with onchocerciasis and Loa Loa

Area endemic for:			Mass Drug Administration
Lymphatic Filariasis	Onchocerciasis	Loa Loa	
+	+/-	+	ALB 400 mg alone twice yearly
+	+	-	IVM 200 µg/kg + ALB 400 mg yearly*
+	-	-	IVM 200 µg/kg + DEC 6 mg/kg + ALB 400 mg yearly
* Biannually if IVM is already delivered biannually to eliminate onchocerciasis. Note: ALB = albendazole; IVM = ivermectin; DEC = diethyl carbamazine.			

Vector control

- Avoidance of mosquito bites through personal protection measures or community-level vector control.
- If possible, malaria and lymphatic filariasis vector control should be integrated.
- Periodic testing for infection and initiation of the above treatment is essential.

[World Health Organization \(2017\) Guideline: alternative mass drug administration regimens to eliminate lymphatic filariasis](#)

Section 36. Human Immunodeficiency Virus

Introduction

The human immunodeficiency virus (HIV) epidemic has spread to all corners of the world, affecting millions of infants, children, and adults. It is the most common cause of acquired immune deficiency in children. The timely and early diagnosis of neonates, infants and children with HIV is vital. However, the healthcare system of resource-limited countries has its own strengths and limitations.

Pitfalls in accurate testing, poor access to health, and high financial costs are a few of the many factors that hamper efforts to limit the spread of diseases such as HIV. In addition, the management of HIV is complex and intricate. Many factors, including proper assessment and indication, counselling, availability and choice of antiretroviral drugs, toxicity, monitoring, financial burden, and social and psychosocial support must be addressed for HIV care to be successful.

Education, evaluation, building expertise, establishing HIV referral centres with diagnostic testing (including virological testing), ensuring availability of antiretroviral drugs, monitoring and follow-up are some of the key elements necessary for programmes to have adequate impact.

Epidemiology

There are two major strains of the human immunodeficiency virus:

- HIV1
 - HIV1 is the more pathogenic and is responsible for the global epidemic.
- HIV2.
 - HIV2 is largely confined to West Africa. This subsection 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 untreated infected individuals.
- HIV/AIDS is now one of the leading causes of death in children.
- Mother-to-child transmission results in approximately 400 children becoming infected with HIV each day worldwide. There were 1.8 million children under 15 years old living with HIV in 2019.
- Around 97% of the world's new HIV infections occur in people living in low- and middle-income countries. About 92% of children living with HIV are from sub-Saharan Africa.
- The number of new infections in children under 15 years in 2019 were estimated to be 150,000 worldwide.
- Deaths from AIDS have decreased by 60% between 2004 and 2019 with increasing access to antiretroviral therapy.
- Around 95% of the world's HIV-infected children have been from resource-limited countries. About 90% have been from sub-Saharan Africa, but the prevalence elsewhere 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 via sexual transmission among 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 without treatment ranging from 17% to 24%. Breastfeeding roughly doubles the risk of transmission. In breastfed cohorts from resource-limited 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 institution of measures to reduce transmission.

In almost all countries, antiretroviral therapy for mothers and infants, has reduced transmission rates to less than 2%.

- Without prenatal counselling and screening, management begins only when the child becomes symptomatic, with subsequent identification of the HIV infection in the mother.
- Treatment may not be successful if presentation is at an advanced stage of immunosuppression.
- In all societies, even those with a high prevalence, HIV is a potentially stigmatising condition, and the mother or both parents may be reluctant to undergo testing. Confidentiality is essential.
- Even if a child born to an infected mother is uninfected, he or she will inevitably be affected.
- Approximately 13.8 million children were estimated to have lost one or both parents to AIDS in 2019 (World Bank data).
 - 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

- Before the advent of highly active antiretroviral therapy (HAART), infant mortality doubled and mortality in children aged 1–5 years increased from 8 to 20 per 1000 in Harare between 1990 and 1996.
- However, even in resource-limited countries, some children may be symptom-free into the second decade of life. There is no upper age limit at which it is appropriate to test for HIV if the mother does not have a negative test after last breastfeeding her child.
- Data from large long-term prospective perinatally recruited cohort studies are limited in resource-limited countries.
- 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 Cote d'Ivoire and clinical studies in Malawi and South Africa showed that *Pneumocystis jiroveci* pneumonia (PCP) is a frequent cause of death in children under 15 months of age.
- Malignancy is a relatively rare AIDS-defining illness in children, compared with HIV-infected adults. However, substantial increases in Kaposi's sarcoma in children have been reported from East and Central Africa. Co-infection with the human herpes virus (HHV8) is a crucial aetiological factor. Kaposi's sarcoma typically presents with large non-tender firm mobile lymph nodes in the head and neck region, and there may be skin lesions and pulmonary disorders. Median survival in one series was only 3 months.

Diagnostic issues

Diagnosing HIV infection clinically in young children can be difficult so always have a low threshold for testing at every opportunity.

Early infant diagnosis of HIV:

1. Exposed infants should be tested at 6 weeks or as soon as possible thereafter.
2. Infant blood samples are sent as dried blood spots to a laboratory that has the required equipment for HIV PCR testing.
3. This laboratory may be close by but could also be far from the site.
4. The results then need to be sent back to sites and returned to caregivers in a timely manner.
5. Finally, infants who have tested positive must be started on ART.
6. In areas of high HIV prevalence, WHO Integrated *Management of the Child* recommends that, when assessing sick children aged 2 months to 5 years, healthcare workers ask about a history of:
 - a. Pneumonia,

- b. Persistent diarrhoea,
- c. Ear discharge or,
- d. Very low weight, and,
 - i. Look for oral thrush,
 - ii. Parotid enlargement and,
 - iii. Generalised persistent lymphadenopathy.
- e. If there are two or more of the above, HIV infection should be suspected, and an HIV antibody test performed.

TABLE 36.1 Signs and symptoms for use in endemic areas with limited access to diagnostic laboratories

Signs or illness specific to HIV infection	Signs or illness uncommon in HIV-negative children	Signs common in both HIV-positive and ill non-HIV-infected children
Pneumocystis pneumonia	Molluscum contagiosum with multiple lesions	Persistent diarrhoea (> 14 days)
Oesophageal candidiasis	Oral thrush (especially after the neonatal period) without antibiotic treatment and lasting > 1 month or recurrent	Failure to thrive
Herpes zoster	Generalised pruritic dermatitis	Persistent cough > 1 month
Lymphoid interstitial pneumonia	Recurrent severe infections (three or more per year)	Generalised lymphadenopathy
Kaposi's sarcoma	Persistent and/or recurrent fever lasting > 1 week	Hepatosplenomegaly
Chronic parotid enlargement	Neurological dysfunction (progressive neurological impairment, delayed development, intellectual impairment, hypertonia)	Chronic otitis media
Recto-vaginal fistula (rare)	Failure to thrive in a fully breastfed infant < 6 months of age	Moderate or severe malnutrition

Clinical features

- The symptoms and signs are often 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.
 - After the neonatal period, the presence of oral thrush without antibiotic treatment, or lasting over 30 days despite treatment, or recurring, or extending beyond the tongue, is highly suggestive of HIV infection.

- Also typical is extension to the back of the throat, which indicates oesophageal candidiasis.
- Chronic parotitis, the presence of unilateral or bilateral parotid swelling (just in front of the ear) for 14 or more days, with or without associated pain or fever or shingles, is highly suggestive of HIV infection.
- Shingles is unusual in healthy children. Herpes zoster ophthalmicus (i.e. shingles around one eye) is said to have greater than 95% PPV for HIV infection in African children.
- Geographical variation in patterns of disease must be recognised. *Penicillium marneffeii* infection, an opportunistic fungal disease that presents 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.

There are many clinical signs or conditions that are quite specific to HIV infection, which should be strongly suspected if these conditions are present (see Table 36.1). Some of these features are listed below.

Signs or conditions that are very specific to HIV-infected children:

- *Pneumocystis pneumonia* (PCP).
- Oesophageal candidiasis.
- Lymphoid interstitial pneumonia (LIP).
- Kaposi's sarcoma.

Signs that may indicate possible HIV infection:

- Recurrent infection: three or more severe episodes of a bacterial infection (e.g., pneumonia, meningitis, sepsis, cellulitis) in the past 12 months.
- Oral thrush: after the neonatal period, the presence of oral thrush in the absence of antibiotic treatment, or lasting over 30 days despite treatment, or recurring, or extending beyond the tongue.
- Chronic parotitis: the presence of unilateral or bilateral parotid swelling for 14 or more days.
- Generalised lymphadenopathy: the presence of enlarged lymph nodes in two or more non-inguinal regions without any apparent underlying cause.
- Hepatomegaly with no apparent cause.
- Persistent and/or recurrent fever.
- Neurological dysfunction: progressive neurological impairment, microcephaly, developmental delay, hypertonia, encephalopathy.
- Herpes zoster.
- HIV dermatitis: typical skin rashes include erythematous papular rashes, extensive fungal infections of the skin, scalp, and nails, and extensive molluscum contagiosum.
- Chronic suppurative lung disease.

Signs that are common in HIV-infected and non-HIV- infected children:

- Chronic otitis media.
- Persistent diarrhoea.
- Moderate or severe malnutrition.

Counselling and testing

If there are reasons to suspect HIV infection, and the child's HIV status is not known, the family should be offered diagnostic testing for HIV.

Counselling used to be more complex when there was no treatment available. There is now no question that it is in the child's best interest to be tested so that treatment can be given to prolong life if they are HIV-positive. The lessons of obtaining informed consent still apply, just as with any other important investigation. If the mother has not already been tested, a positive result in a child is most likely to mean that the mother is infected, too. However, if the mother is not present when the child presents, the onus of responsibility of the paediatrician is to the child, and testing should not be delayed.

The additional consideration with testing for HIV is the stigma associated with the diagnosis. When HIV meant inevitable death, there was enormous fear of the diagnosis. The potentially better prognosis has been hard to accept when so many present too late. Stigma is also associated with the fact that it is a sexually transmitted disease. This raises issues of where the infection was acquired and contact tracing. The fear of domestic violence and social ostracization sometimes creates reluctance to allow children to be tested.

The process of counselling starts with always ensuring that the test is done with consent. The person giving consent should be the carer at the time when the test is indicated. If the child is of an age at which they can be responsible for taking their own medicines, they can give their own consent for testing. There are no surrogate tests for HIV, such as lymphocyte counts; the appropriate test according to what is available should always be done. If there is delay in getting a result it may be necessary to start appropriate treatment – for example, for suspected PCP pneumonia with IV co-trimoxazole.

If there is refusal to allow testing, the test cannot be carried out. If the test is positive, the family need to have confidence in the healthcare professionals to ensure adherence to treatment. If staff at the first referral level have not been trained, assistance should be sought from other sources, such as local community AIDS support organizations. A time limit should be set to prevent repeated procrastination and the risk of death from opportunistic infection.

HIV counselling should take account of the child as part of a family. This should include the psychological implications of HIV infection for the child, mother, father and other family members. Counselling should stress that, although cure is currently not possible, there is much that can be done to improve the quality and duration of the child's life. Antiretroviral treatment (ART) is available and greatly improves survival and the quality of life of the child and the parents. Counselling should make it clear that the hospital staff want to help, and that the mother should not be frightened of going to a health centre or hospital early in an illness, even if this is only to ask questions.

HIV is talked about much more openly now than was the case at the start of the epidemic, and testing is seen as an expected part of routine healthcare. The request for testing should not be built up as a major event but included as part of the diagnostic work-up along with malaria, TB and other investigations.

Indications for HIV counselling

HIV testing is indicated in the following situations.

Child with unknown HIV status presenting with clinical signs of HIV infection and/or risk factors (e.g., a mother or sibling with HIV/AIDS):

1. Take advice from local people experienced in offering testing, so that any advice given is consistent with what the mother will receive from professional counsellors at a later stage.
2. Where available, arrange an HIV test, according to national guidelines, to confirm the clinical diagnosis, alert the mother to HIV-related problems, and discuss prevention of future mother-to-child transmission.

Note the following:

In countries with generalised HIV epidemics, routine healthcare provider-initiated testing and counselling (PITC) is recommended for all children seen in paediatric health services (World Health Organization, 2007).

Child known to be HIV-infected but responding poorly to treatment or needing further investigations.

Discuss the following:

1. The parents' understanding of HIV infection
2. Management of current problems
3. The role of ART and adherence to regular drug administration
4. The need to refer to a higher level, if necessary
5. Support from community-based groups (if available).

Child known to be HIV-infected who has responded well to treatment and is to be discharged (or referred to a community-based care programme).

Discuss the following:

1. The reason for referral to a community-based care programme, if appropriate
2. Follow-up care
3. Risk factors for future illness
4. Immunisation and HIV
5. Adherence and ART treatment support.

Laboratory diagnosis

The definitive diagnosis of HIV requires laboratory confirmatory testing.

The HIV antibody test is commonly used as a screening test. However, in the neonatal and infantile period, the antibody test is not recommended. This is because the maternal HIV antibodies readily cross the placenta and persist in the neonate for up to 18 months. Also, all screening tests should be confirmed by a second test. The following are some of the tests used for laboratory diagnosis of HIV in children:

Antibody tests:

- HIV IgG antibody tests.
- Rapid Test.

Virological tests:

- HIV DNA polymerase chain reaction (PCR).
- HIV RNA PCR.
- HIV culture.

P24 antigen assay:

- Direct.
- Acid hydrolysis.

RNA or DNA-based assays are the most reliable for diagnosis and are recommended for diagnosis in infants. However, the cost and availability of tests may be an issue in resource-limited countries.

The simplest laboratory test is an HIV antibody test, usually done by enzyme-linked immunosorbent assay (ELISA). However, even this may not be affordable or available in many settings.

The WHO recommends the use of a presumptive clinical diagnosis of severe HIV disease pending results of virologic testing if:

- An infant's HIV exposure is confirmed by antibody testing and if either:
- Clinical stage 3 or 4 or AIDS-indicator condition(s) are present or

- The child has two or more of the following:
 - Oral thrush,
 - Severe pneumonia,
 - Severe sepsis.

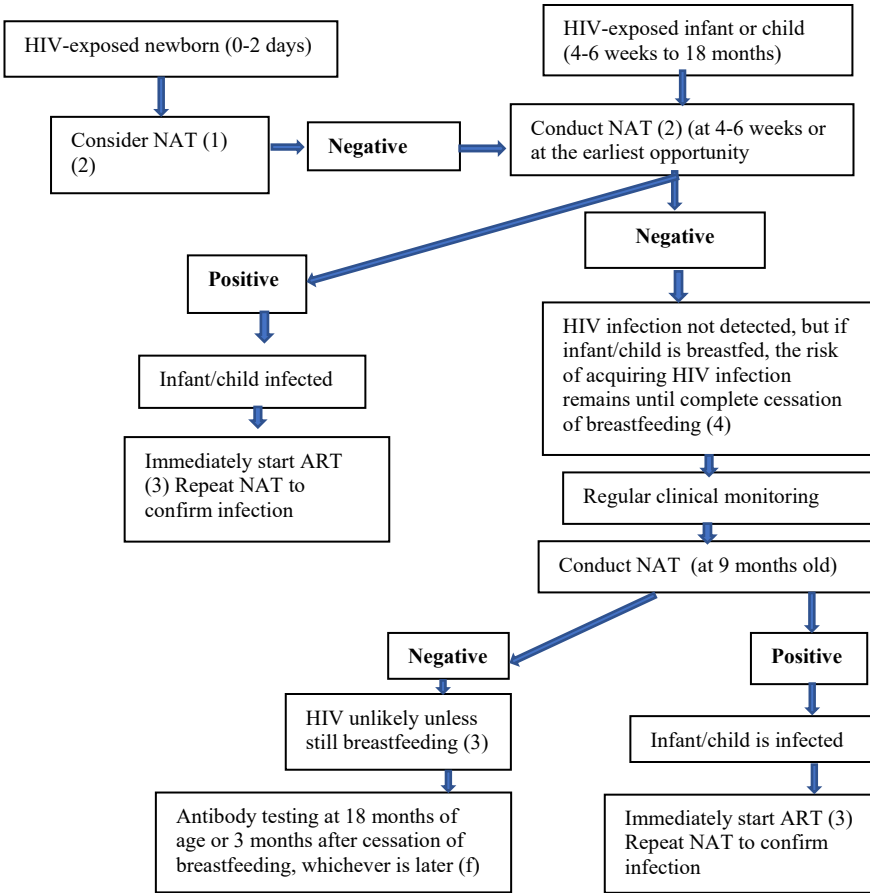
A presumptive diagnosis of AIDS can also be made in an antibody-positive infant if CD4 percentages are below 20% or other factors are present, including recent HIV-related maternal death or advanced HIV disease in the mother.

Infants acquire maternal HIV IgG trans-placentally, and this 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.

Additional diagnostic challenges arise if the child is still breastfeeding or has been breastfed.

Although HIV infection cannot be ruled out until 18 months for some children, many children will have lost HIV antibodies between 9 and 18 months of age. The importance of this is that a rapid antibody test can be done if the mother is unwilling or unavailable to be tested herself. If the antibody test is negative and the child has not been breastfed in the last 6 weeks, there is then no need for further testing unless there is ongoing exposure (e.g., through breastfeeding). If the antibody is positive, this does not mean that the child is infected, but if other signs of immune deficiency are present it may warrant empirical treatment according to WHO guidelines pending PCR testing.

FIGURE 36.1 WHO Algorithm for diagnosing HIV infection in infants and children less than 18 months of age. NAT = Nucleic Acid Test



Legend to Figure 36.1 above

- (1) Based on the 2016 WHO consolidated ARV guidelines, addition of NAT at birth to the existing testing algorithm can be considered.
- (2) POC NAT can be used to diagnose HIV infection and to confirm positive results.
- (3) Start ART, without delay. At the same time, retest to confirm infection. As maternal treatment is scaled up and mother-to-child transmission rates decrease, false-positive results are expected to increase; retesting after first positive NAT is hence important to avoid unnecessary treatment, particularly in settings with lower transmission rates. If the second test is negative a third NAT should be performed before interrupting ART.
- (4) For children who were never breastfed, additional testing following a negative NAT at 4-6 weeks is included in this algorithm to account for potential false-negative NAT results.
- (5) The risk of HIV transmission remains as long as breastfeeding continues. If the nine-month test is conducted earlier than three months after cessation of breastfeeding, infection acquired in the last days of breastfeeding may be missed. Retesting at 18 months or three months after cessation of breastfeeding (whichever is later) should be carried out for final assessment of HIV status.
- (6) If breastfeeding extends beyond 18 months, the final diagnosis of HIV status can only be assessed at the end of breastfeeding. If breastfeeding ends before 18 months, final diagnosis of HIV status with antibody testing can only be assessed at 18 months old. Antibody testing should be undertaken at least three months after cessation of

breastfeeding (to allow for development of HIV antibodies). For infants younger than 18 months of age, NAT should be performed to confirm infection. If the infant is older than 18 months, negative antibody testing confirms that the child is uninfected; positive antibody testing confirms that the infant is infected.

Many children born to HIV-infected mothers may die before this age, and a diagnosis of HIV infection may be presumptive, dependent on signs and symptoms. Thus, based on age, the clinical, serological, and virological tests and status of breastfeeding will determine the diagnosis of a child undergoing evaluation (see Table 36.2).

TABLE 36.2 Proposed methods for diagnosing HIV in children (born to mothers identified as HIV-positive or with unknown HIV status) in resource-limited settings*

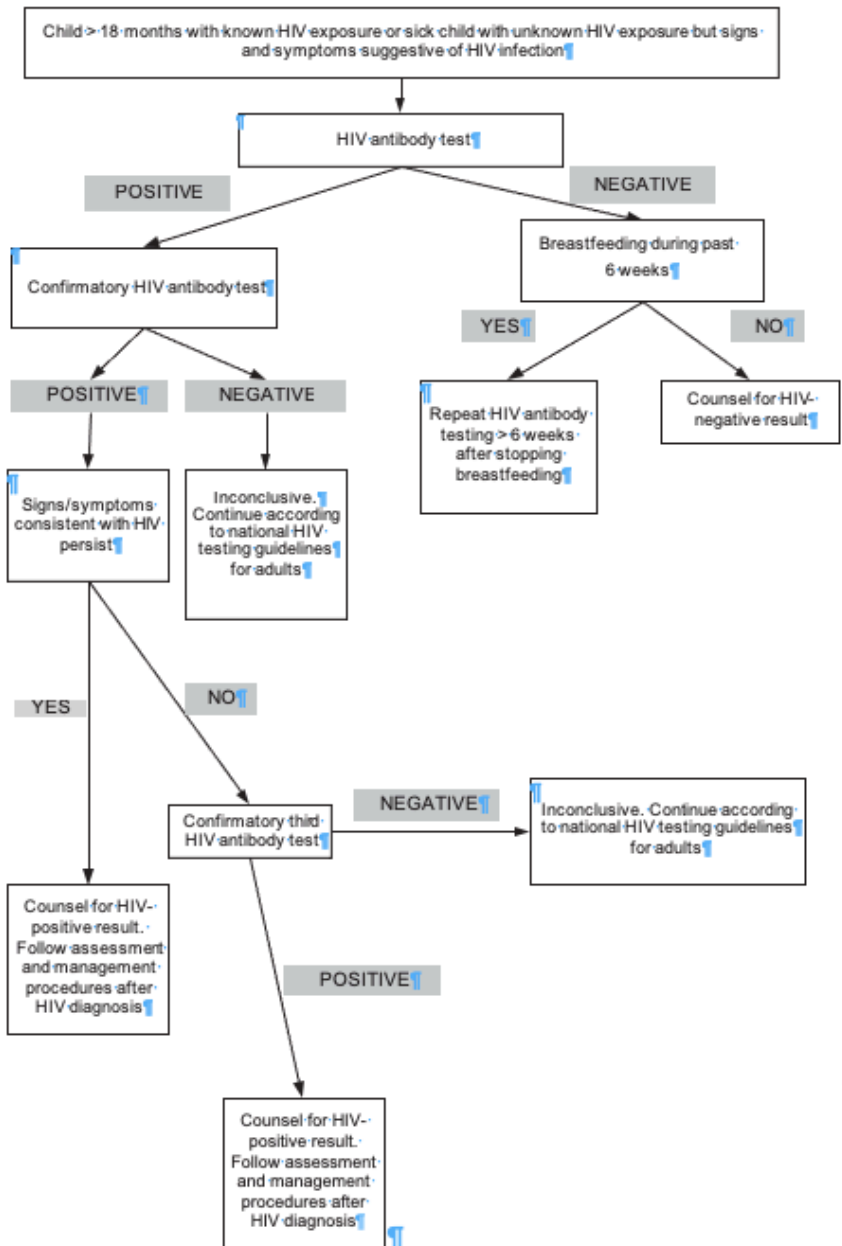
Diagnostic method	Age of child		
	< 12 months	12–18 months	> 18 months
Clinical staging	Yes	Yes	Yes
Serological (antibody)	May be helpful†	Yes‡	Yes
Virological	Yes	Yes, if serology is positive	No, serology is definitive

*If the child is breastfeeding, a negative diagnostic test, either serological or virological, would have to be repeated 6 weeks after cessation of all breastfeeding.

† A positive antibody test in a child under 12 months of age defines the exposure status of the child and may be helpful when the mother's HIV status is unknown.

‡ By the age of 12 months, HIV antibody-positive testing can be considered indicative of probable HIV infection and should be confirmed by a second antibody test after 18 months.

FIGURE 36.2 Algorithm for diagnosing HIV infection in infants and children aged 18 months or older.



Notes to Figure 36.2 above.

1. A definitive diagnosis of HIV infection in children aged ≥ 18 months can be made with antibody testing. HIV testing procedures for children aged ≥ 18 months follow the national HIV testing guidelines for adults. Virological testing can be used to diagnose HIV infection at any age.

2. One positive HIV antibody test (rapid test or ELISA) should be confirmed by a second HIV antibody test (rapid test or ELISA) using an assay that relies on a different antigen or has different operating characteristics. In low-HIV-prevalence settings, a third confirmatory test may be required.
3. Children who are breastfed have an ongoing risk of acquiring HIV infection. Therefore, HIV infection can be excluded only after stopping breastfeeding for more than 6 weeks.

HIV antibody test (ELISA or rapid tests)

Rapid tests are widely available and are safe, effective, sensitive, and reliable for diagnosing HIV infection in children above the age of 18 months. For those under 18 months, HIV antibody tests are a sensitive reliable way to exclude HIV infection in non-breastfeeding children. For those children under 18 months, confirm all positive HIV antibody tests by virological tests as soon as possible (see below). Where this is not possible, repeat antibody testing at 18 months.

Rapid HIV tests can be used to exclude HIV infection in a child presenting with malnutrition or other serious clinical events in areas with high HIV prevalence.

To confirm the diagnosis, it is necessary to use assays that detect the virus itself or viral components. Such tests include antigen detection tests, viral culture, amplification techniques and HIV-specific IgA tests.

Virological testing

Virological testing for HIV-specific RNA or DNA is the most reliable method of diagnosing HIV infection in children under 18 months of age. This requires sending a blood sample to a specialised laboratory that can perform this test, and these are becoming increasingly available in large centres in many countries. It is relatively inexpensive, easy to standardise, and can be done using dried blood spots. In infants and children undergoing virological testing, the following assays (and respective specimen types) are potentially available:

- HIV DNA on whole blood specimens or dried blood spots (DBS)
- HIV RNA on plasma or DBS
- Ultrasensitive p24 antigen (Up24 Ag) on plasma or DBS.

The new ultrasensitive p24 assay is as accurate as PCR virology, significantly less expensive, and less resource demanding when used to diagnose HIV. It is particularly valuable in infants under 12 months old.

A test at birth will only detect in-utero infection, whereas most infection occurs at delivery. Infected infants can suffer life-threatening infections in the first weeks of life. Where there is a high risk of infection (e.g., a mother who seroconverts in pregnancy, has a low CD4 count or other genital lesions and has had no antiretrovirals), testing should be done at 2 weeks of age.

Where the risk is low (e.g., a mother who has been on HAART throughout pregnancy), the test can be delayed until 6 weeks of age. At the first DTP immunisation of all infants, the maternal HIV status should be checked from records or rapid testing. If it is positive or unavailable, the child should be tested. All HIV-exposed infants should have HIV virological testing at 4–6 weeks of age or at the earliest opportunity thereafter.

One virological test that is positive at 4–8 weeks is sufficient to diagnose infection in a young infant. If the young infant is still breastfeeding, and the DNA virological test is negative, it needs to be repeated 6 weeks after the complete cessation of breastfeeding to confirm that the child is not HIV infected.

Infants with signs or symptoms suggestive of HIV infection must undergo HIV serological testing and, if this is positive, virological testing. In breastfeeding infants or children, it is strongly recommended that breastfeeding is not discontinued to perform any kind of diagnostic HIV test.

In sick infants in whom HIV infection is being considered as an underlying cause of symptoms and signs, and virological testing result is pending, HIV serological testing and the use of a clinical algorithm for presumptive clinical diagnosis of HIV infection are strongly recommended.

In infants with an initial positive virological test result, it is strongly recommended that ART be started without delay, and, at the same time, a second specimen be collected to confirm the initial positive virological test result. Do not delay ART. In infected infants, immediate initiation of ART saves lives, and commencement of ART should not be delayed while waiting for the results of the confirmatory test. Results from the CHER trial suggests initiation of ART before 12 weeks of age results in a 75% reduction in early mortality. (Cotton MF, Violari A, Otwombe K, et al. (2013)

Early time-limited antiretroviral therapy versus deferred therapy in South African infants infected with HIV: results from children with HIV early antiretroviral (CHER) randomised trial. *Lancet* 382: 1555–63.)

Test results from virological testing in infants should be returned to the clinic and the child and their mother or carer as soon as possible, but at the very latest within 4 weeks of specimen collection. Positive test results should be fast-tracked to the mother–baby pair as soon as possible to enable prompt initiation of ART.

All infants with unknown or uncertain HIV exposure who are being seen in healthcare facilities at or around the time of birth or at the first postnatal visit (usually 4–6 weeks), or at another child health visit, should have their HIV exposure status ascertained.

Clinically well, HIV-exposed infants should undergo HIV serological testing at around 9 months of age (or at the time of the last immunisation visit). Those who have positive serological assays at 9 months should have a virological test to identify whether they need ART.

Recently the WHO Technical Reference Group for Paediatric HIV/ART and Care made the following key recommendations regarding when and how to test for HIV in children:

Infants known to have been exposed to HIV should have a virological test (HIV nucleic acid test) at 4–6 weeks of age, or at the earliest opportunity for infants seen after 4–6 weeks.

Urgent HIV testing is recommended for any infant presenting to healthcare facilities with signs, symptoms or medical conditions that could indicate HIV.

All infants should have their HIV exposure status established at their first contact with the healthcare system, ideally before 6 weeks of age.

Infants under 6 weeks of age, of unknown HIV exposure status and in settings where local or national antenatal HIV seroprevalence is greater than 1%, should be offered maternal or infant HIV antibody testing and counselling to establish their exposure status.

Other laboratory tests

A low CD4 count or CD4:CD8 ratio suggests HIV infection but requires specialised equipment. A low total lymphocyte count is a much less expensive though less specific surrogate marker of HIV infection and immunosuppression.

HIV infection can also cause anaemia or thrombocytopaenia. It is appropriate to test for HIV in children who present with low platelet counts.

Lack of thymic shadow on chest X-ray is a feature of advanced disease, but is clearly not specific, as the thymus tends to shrink in volume in response to a variety of acute infections in childhood.

Assessment of HIV-infected and HIV- exposed children

Any child with an illness compatible with HIV infection should be properly evaluated irrespective of HIV exposure. This includes neonates, infants and children with perinatal exposure, and those with specific signs and symptoms suggestive of HIV infection, chronic or unexplained illness, or known exposure during childhood. Figure 36.3 shows a useful algorithm that can be used by paediatricians and other clinical care providers for the initial evaluation and management of children with known exposure to HIV, or sick children with symptoms suggestive of HIV infection but unknown history of exposure.

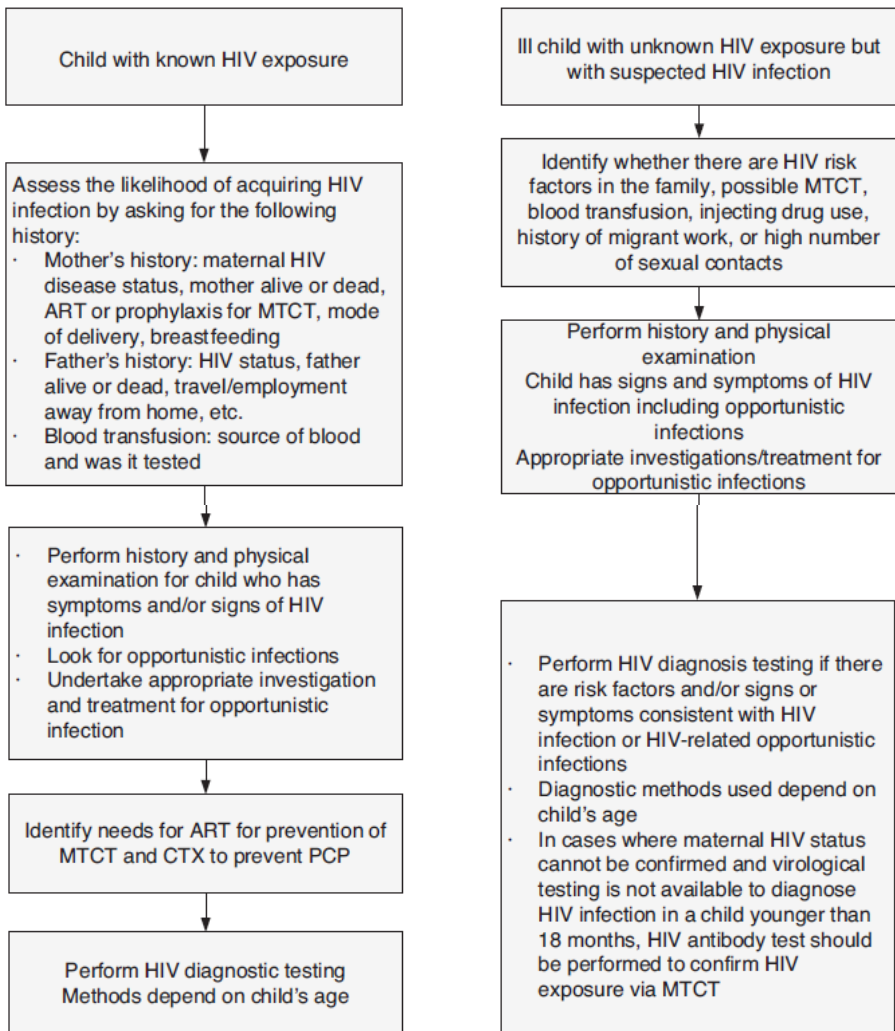


FIGURE 36.3 Initial assessment of a child with known HIV exposure or a sick child with unknown HIV exposure but with suspected HIV infection. MTCT: mother-to-child transmission; ART: Antiretroviral Therapy; PCP: Pneumocystis Pneumonia. CTX = Cotrimoxazole

Notes to Figure 36.3 above

1. An expert in the management of children with HIV should be consulted wherever this is feasible.
2. If HIV is suspected, compassionate counselling before HIV testing should be arranged.
3. Maternal advanced HIV disease and low CD4 are risk factors for HIV transmission.
4. Successful treatment with ART in mothers reduces the risk of transmission.
5. An infant remains at risk of acquiring HIV for as long as he or she is breastfed.

Perinatally acquired HIV infection

HIV1 transmission occurs more frequently than that of HIV2. It occurs in late pregnancy, during delivery and through breastfeeding, and transmission is more likely if any of the following factors are present:

- Advanced maternal HIV disease
- Premature labour
- Prolonged rupture of membranes
- Contact with maternal blood
- In the first twin
- Maternal genital infection.

Prevention of mother-to-child transmission (PMTCT) of HIV and infant feeding in the context of HIV

HIV transmission may occur during pregnancy, labour and delivery, or through breastfeeding. The best way to prevent transmission is to prevent sexually acquired HIV infection, especially in pregnant women, and to prevent unintended pregnancies in HIV-positive women. If an HIV-infected woman becomes pregnant, she should be provided with services including antiretroviral drugs, safe obstetric practices, and infant feeding counselling and support (see Section B15 MCAI Handbook Of Hospital Care For Maternal Emergencies Including Major Trauma https://12dec1c3-e3f4-e784-7f3a-6e7903610962.filesusr.com/ugd/dd2ba4_3f90a11b6ec844cb9969c7c75e7f9bab.pdf Accessed 29.03.2021).

The key recommendations of the 2018 WHO guidance on ARV drugs for treatment of pregnant women and prevention of HIV in infants are as follows:

As early as possible, provide ART for all HIV-positive pregnant women both to benefit the health of the mother and to prevent HIV transmission to her child during pregnancy and breastfeeding.

Start lifelong ART for all pregnant women, regardless of symptoms. HIV-positive pregnant women in need of treatment for their own health (i.e. as soon as the eligibility criteria are met) should start ART irrespective of gestational age, and should continue with it throughout pregnancy, delivery, during breastfeeding and thereafter.

The recommended first-line regimens for pregnant women are as follows:

- TDF + 3TC (or FTC) + DTG or
- TDF + 3TC (or FTC) + EFV.

The infant is given NVP or AZT starting as soon as possible after birth (aim for less than 6 hours postpartum) and continued for 4 – 6 weeks. (See Neonatal Handbook Section 13)

In women who are not immunocompromised; Antiretroviral (ARV) prophylaxis is indicated for HIV-positive pregnant women with relatively strong immune systems who do not need ART for their own health. This will reduce the risk of HIV transmission from mother to child.

All pregnant and breastfeeding women infected with HIV should initiate ART as lifelong treatment.

Breastfeeding

The global availability of ART means that there is enough evidence for the WHO to recommend breastfeeding for mothers with HIV.

Even with ART there is still a small risk of HIV transmission, particularly if there is any interruption to treatment, either in supply or absorption (due to diarrhoea or vomiting). HIV can be transmitted through breast milk at any point during lactation, so the rate of infection in breastfed infants increases with duration of breastfeeding.

In many countries, public health services where there is poor access to clean drinking water and alternatives to breastfeeding have not been able to adequately support and provide safe replacement feeding. HIV-positive mothers have faced the dilemma of whether to give their babies all the benefits of breastfeeding but expose them to the risk of HIV infection or avoid all breastfeeding and increase the risk of their baby's death from diarrhoea and malnutrition. The effectiveness of ART in reducing transmission through breastfeeding has resulted in two major changes in 2012 from previous guidelines:

1. National health authorities should decide whether health services will principally counsel and support HIV-positive mothers to either:
 - a. breastfeed and receive ARV interventions or
 - b. avoid all breastfeeding, as the strategy that is most likely to give infants the greatest chance of HIV-free survival.
2. In settings where national authorities recommend that HIV-positive mothers should breastfeed, and provide ARVs to prevent transmission, mothers should exclusively breastfeed their infants for the first 6 months of life, introducing appropriate complementary foods thereafter, and should continue breastfeeding for the first 12 months of life.

Mothers who are known to be HIV infected and who decide to stop breastfeeding at any time should stop gradually over 1 month. Stopping breastfeeding abruptly is not advisable (World Health Organization, 2016).

These new guidelines have great potential to improve the mother's own health and to reduce the mother-to-child HIV transmission risk to 5% or lower in a breastfeeding population in the absence of any interventions and with continued breastfeeding. With ART the UNAIDS is aiming for 95% coverage of services for eliminating vertical transmission worldwide by 2025.

Where a decision has been made to continue breast-feeding because the child is already infected, infant feeding options should be discussed for future pregnancies. This should be carried out by a trained and experienced counsellor.

If a child is known to be HIV-infected and is being breastfed, encourage the mother to continue breastfeeding if living in a resource-limited country, as there is usually a high risk of gastroenteritis in such regions.

If the mother is known to be HIV-positive and the child's HIV status is unknown, the mother should be counselled about the benefits of breastfeeding as well as the risk of HIV transmission through breastfeeding. If replacement feeding is acceptable, feasible, affordable, sustainable, and safe, avoidance of further breastfeeding is recommended.

Otherwise, exclusive breastfeeding should be practised if the child is less than 6 months of age, and breastfeeding should be discontinued as soon as these conditions are in place.

Infants born to HIV-positive mothers who have escaped perinatal infection have a lower risk of acquiring HIV if they are not breastfed. However, their risk of death may be increased if they are not breastfed in situations where there is no regular access to nutritionally adequate, safely prepared breast milk substitutes, and there is a high risk of gastroenteritis.

Counselling should be provided by a trained and experienced counsellor. Take advice from local people experienced in counselling so that any advice given is consistent with what the mother will receive from professional counsellors at a later stage.

If the mother decides to use breast milk substitutes, counsel her about their correct use and demonstrate their safe preparation.

Management of the child with a suspected or proven HIV infection

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 disease affects the whole family, and the child must be treated in the context of the needs of all of the family.

Currently there are far more questions than evidence-based answers; published data on many management issues in the context of resource-limited 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 empirical trials of therapy.

A low threshold for antibiotic use is appropriate, but may exacerbate diarrhoea and candidiasis, and may only be effective if given IV or IM, in the presence of diarrhoea and malabsorption.

Most infections in HIV-positive children are caused by the same pathogens as in HIV-negative children, although they may be more frequent, more severe and occur repeatedly. There is recent evidence that *Staphylococcus aureus* may be more invasive in children with HIV.

Clinical staging of HIV infection

In a child with diagnosed or highly suspected HIV infection, a clinical staging system helps to identify the degree of damage to the immune system and to plan treatment and care options. The clinical stages identify a progressive sequence from least to most severe, such that the higher the clinical stage the poorer the prognosis. For classification purposes, once a stage 3 clinical condition has occurred, the child's prognosis will probably remain that of stage 3, and will not improve to that of stage 2, even with resolution of the original condition, or the appearance of a new stage 2 clinical event. ART with good adherence dramatically improves the prognosis.

The clinical staging events can also be used to identify the response to ARV treatment if there is no easy or affordable access to viral load or CD4 testing.

TABLE 36.3 WHO paediatric clinical staging system for use in children under 13 years with confirmed laboratory evidence of HIV infection (HIV antibody where age is > 18 months, DNA or RNA virological testing where age is < 18 months)

Stage	Symptoms and signs
Stage 1	<ul style="list-style-type: none"> • Asymptomatic • Persistent generalised lymphadenopathy (PGL)
Stage 2	<ul style="list-style-type: none"> • Unexplained persistent hepatosplenomegaly • Papular pruritic eruptions • Fungal nail infections • Lineal gingival erythema (LGE) • Extensive wart virus infection • Extensive molluscum infection (> 5% of body area) • Recurrent oral ulcerations (two or more episodes in 6 months) • Parotid enlargement • Herpes zoster • Recurrent or chronic upper respiratory tract infections (otitis media, otorrhoea, sinusitis, two or more episodes in any 6-month period)
Stage 3	<ul style="list-style-type: none"> • Unexplained moderate malnutrition not responding to standard therapy • Unexplained persistent diarrhoea (for > 14 days) • Unexplained persistent fever (intermittent or constant, for > 1 month) • Oral candidiasis (outside the neonatal period) • Oral hairy leukoplakia • Pulmonary tuberculosis¹ • Severe recurrent presumed bacterial pneumonia (two or more episodes in 6 months) • Acute necrotising ulcerative gingivitis or periodontitis • Lymphoid interstitial pneumonia (LIP) • Unexplained anaemia (< 8 grams/dL), neutropenia (< 500/mm³) or thrombocytopenia (< 30, 000/mm³) for > 1 month

Stage	Symptoms and signs
Stage 4	<ul style="list-style-type: none"> • Unexplained severe wasting or severe malnutrition not responding to standard therapy • Pneumocystis pneumonia • Recurrent severe presumed bacterial infections (two or more episodes within 1 year, e.g., empyema, pyomyositis, bone or joint infection, or meningitis, but excluding pneumonia) • Chronic orolabial or cutaneous herpes simplex infection (for > 1 month) • Disseminated or extrapulmonary tuberculosis • Kaposi's sarcoma • Oesophageal candidiasis • Symptomatic HIV seropositive infant • < 18 months of age with two or more of the following: oral thrush, with or without severe pneumonia, with or without failure to thrive, with or without severe sepsis² • CMV retinitis • CNS toxoplasmosis • Any disseminated endemic mycosis, including cryptococcal meningitis (e.g., extrapulmonary cryptococcosis, histoplasmosis, coccidiomycosis, penicilliosis) • Cryptosporidiosis or isosporiasis (with diarrhoea for > 1 month) • Cytomegalovirus infection (onset at age 1 month in an organ other than liver, spleen, or lymph nodes) • Disseminated mycobacterial disease other than tuberculous • Candida of trachea, bronchi, or lungs • Acquired HIV-related recto-vesical fistula • Cerebral or B-cell non-Hodgkin's lymphoma • Progressive multifocal leukoencephalopathy (PML) • HIV encephalopathy • HIV-related cardiomyopathy • HIV-related nephropathy

¹ TB may occur at any CD4 count, and CD4% should be considered where available.

² Presumptive diagnosis of stage 4 disease in seropositive children under 18 months of age requires confirmation with HIV virological tests, or with HIV antibody test if over 18 months of age.

Antiretroviral therapy (ART)

Antiretroviral (ARV) drugs are becoming more widely available and have revolutionised the care of children with HIV/AIDS. ARV drugs are not a cure for HIV, but they have dramatically reduced mortality and morbidity, and improved the quality and length of life. The WHO recommends that in resource-limited settings, HIV-infected adults and children should start ARV therapy using simplified standardised treatment guidelines.

Resistance to single or dual agents is quick to emerge, so single-drug regimens are contraindicated. Indeed at least three drugs are the recommended minimum standard for all settings. Although new ARV drugs are coming on to the market, frequently these are not available for use in children, due to lack of suitable formulations or dosage data, or their high costs.

As children with HIV are often part of a household that includes an adult with HIV, ideally access to treatment and ARV drugs needs to be ensured for other family members, and where possible similar drug regimens should be used. Fixed-dose combinations are

increasingly available and are preferred as they promote and support treatment adherence, as well as reducing the cost of treatment. Existing tablets often cannot be divided into lower dosages for children (under 10 kg), so syrups or solutions and suspensions are needed.

The underlying principles of ART and the choice of first-line ART in children are largely the same as for adults. However, it is also important to consider the following:

1. Availability of a suitable formulation that can be taken in appropriate doses
2. Simplicity of the dosage schedule
3. Taste/palatability and thus compliance in young children
4. The ART regimen that the parent(s) or carers are or will be taking.

Suitable formulations for children are not available for some ARVs (particularly the protease inhibitor class of drugs).

Antiretroviral drugs

Antiretroviral drugs fall into four main classes, namely nucleoside analogue reverse transcriptase inhibitors (NRTIs), non-nucleoside reverse transcriptase inhibitors (NNRTIs), protease inhibitors (PIs) and integrase inhibitors (INSTI) (see Table 32.4). Triple therapy is the standard of care.

The WHO currently recommends that first-line regimens should be based upon two nucleoside analogue reverse transcriptase inhibitors (NRTIs) plus one integrase inhibitors (INSTI). Alternative regimes include non-nucleoside drug (NNRTI) if INSTIs not available. Protease inhibitors are usually recommended as part of second-line regimens in most resource-limited settings.

Efavirenz (EFV) is the NNRTI of choice in children who are on rifampicin, if treatment needs to start before anti-tuberculous therapy is completed.

For drug dosages and regimens, see Appendix to this section.

Calculation of drug dosages

Drug doses are given per kg for some drugs and per m² surface area of the child for others. A table giving the equivalent weights of various surface area values is provided in Section 66 of this textbook, to aid dosage calculation. In general, children metabolise PI and NNRTI drugs faster than adults, and require higher than adult equivalent doses to achieve appropriate drug levels. Drug doses have to be increased as the child grows, otherwise there is a risk of under-dosage and development of resistance.

Formulations

Liquid formulations may not be readily available, are more expensive, and may have a reduced shelf-life. As the child gets older, the amount of syrup that needs to be taken becomes quite considerable. Therefore, in patients over 10 kg in weight, it is preferable to give parts of scored tablets or combination preparations (see **Appendix** to this Section).

TABLE 36.4 Classes of antiretroviral drugs recommended for use in children in resource-limited settings

Nucleoside analogue reverse transcriptase inhibitors (NRTIs)		
Zidovudine	ZDV/AZT	180–240 mg/m ² twice daily
Lamivudine	3TC	4 mg/kg twice daily up to a maximum of 150 mg twice daily
Abacavir	ABC	8 mg/kg/dose given twice daily up to 300 mg twice daily

Non-nucleoside reverse transcriptase inhibitors (NNRTIs)		
Nevirapine	NVP	160–200 mg/m ² up to a maximum of 200 mg twice daily
Efavirenz	EFV	15 mg/kg/day up to 600 mg once daily
Etravirine	ETV	200 mg twice daily for adolescents
Protease inhibitors (PIs)		
Lopinavir/ ritonavir	LPV/r	230–350 mg/m ² twice daily
Darunavir	DRV	10–20 mg/kg twice daily
Atazanavir	ATV	7 mg/kg once daily
Ritonavir	RTV	Given as a 'booster' with another PI
Integrase inhibitors		
Raltegravir	RAL	400 mg twice daily for adolescents
Dolutegravir	DTG	50 mg >20 kg

Table 36.5 First line ART regimens

Population	Preferred first-line regimen	Alternative first-line regimen	Special circumstances
Adults and adolescents	TDF+3TC(or FTC)+DTG	TDF+3TC+EFV 400mg	TDF+3TC (or FTC) + EFV 600mg AZT+3TC+EFV 600mg TDF+3TC (or FTC) + PI/r TDF+3TC (or FTC) +RAL TAF+3TC (or FTC) +DTG ABC +3TC +DTG
Children	ABC + 3TC+DTG	ABC+3TC+LPV ABC+3TC+RAL TAF+3TC (orFTC) + DTG	ABC+3TC3EFV (or NVP) AZT+3TC+EFV (or NVP) AZT + 3TC +LPV/r (or RAL)
Neonates	AZT+3TC+RAL	AZT+3TC+NVP	AZT+3TC+LPV/r

For dosing tables see: [World Health Organization \(2018\) Updated recommendations on first-line and second-line antiretroviral regimens and post-exposure prophylaxis and recommendations on early infant diagnosis of HIV: interim guidelines. Supplement to the 2016 consolidated guidelines on the use of antiretroviral drugs for treating and preventing HIV infection.](#) (Accessed 8th March 2021)

When to start ART

About 20% of HIV-infected infants in developing countries progress to AIDS or death by 12 months of age (with a substantial contribution from PCP infections in infants under 6 months of age who are not receiving co-trimoxazole treatment).

ART should be initiated in all children infected with HIV, regardless of WHO clinical stage or CD4 count.

Infants and children with specific conditions

For children or adolescents with severe anaemia (< 7.5 g/ dL) or severe neutropenia (< 0.5/mm³), avoid AZT.

For adolescents over 12 years of age with hepatitis B, the preferred regimen is tenofovir (TDF) + emtricitabine (FTC) or 3TC + EFV.

Side effects of antiretroviral therapy and monitoring

The response to antiretroviral treatment and the side effects of treatment both need to be monitored. Where CD4 cell count or viral load monitoring is available, this should be done every 3 to 6 months and can provide information on the success or failure of the response to treatment, and therefore guide changes to treatment. Where this is not possible, clinical parameters, including clinical staging events, need to be used (see Table 36.6).

Monitoring the response after ARV initiation

- After ARV initiation or a change in ARVs see the child at 2 and 4 weeks after the start or change.
- All children should be seen if there are any problems that concern the caregiver, or inter-current illness.

Long-term follow-up

- A clinician should see the child at least every 3 months.
- A non-clinician (ideally the provider of ARV medication, such as a pharmacist, who would assess adherence and provide adherence counselling) should see the child monthly.
- The child should be seen more frequently, preferably by a clinician, if clinically unstable.

Monitoring the response (see Appendix to this section)

At entry into care and at initiation of ART, and then at regular intervals and as required by symptoms, monitor the following:

- Weight and height (monthly)
- Neurodevelopment (monthly)
- Adherence (monthly)
- CD4 (%) if available (then every 3 to 6 months)
- Viral load if available (every 3 to 6 months)
- Baseline haemoglobin or haematocrit (if on ZDV/AZT), full chemistry (renal function, liver enzymes, especially ALT for liver toxicity) and lipids (if available)
- Symptom-related determination: haemoglobin or haematocrit or full blood count, ALT.

General long-term side effects of antiretroviral therapy include lipodystrophy. The specific side effects of individual antiretroviral drugs are summarised in the **Appendix** to this Section.

When to change treatment

Drugs need to be substituted for others when there is:

- Treatment-limiting toxicity, such as:
 - Stevens–Johnson syndrome (SJS)
 - Severe liver toxicity
 - Severe haematological findings
- Drug interaction (e.g. tuberculosis treatment with rifampicin interfering with NVP or PI)
- Potential lack of adherence by the patient if they cannot tolerate the regimen.

TABLE 36.6 Clinical and CD4 definition of ARV treatment failure in children (after 6 months or more of ARV)

Clinical criteria	CD4 criteria
Lack of or decline in growth among children with an initial growth response to ARV	Return of CD4% if aged < 6 years (% or count if aged ≥6 years) to pre-therapy baseline or below, without other cause
Loss of neurodevelopmental milestones or onset of encephalopathy	≥50% fall from peak CD4% if aged < 6 years (% or count if aged ≥6 years), without other aetiology
New or recurrent WHO clinical Stage 4 conditions	

First-line regimen treatment failure, when to switch regimens

- A switch to a second-line regimen is recommended when:
 - Clinical failure is recognised and/or
 - Immunological failure is recognised and/or
 - virological failure is recognised.
- Clinical failure is defined as the appearance or reappearance of WHO clinical stage 3 or stage 4 events after at least 24 weeks on ART in a treatment-adherent child. It is important to exclude TB as a cause of clinical failure, especially when there is poor growth.
- Immunological failure is defined as developing or returning to the following age-related immunological thresholds after at least 24 weeks on ART, in a treatment-adherent child:
 - CD4 count of < 200 cells/mm³ or CD4+% < 10% for a child between 2 and 4 years of age
 - CD4 count of < 100 cells/mm³ for a child aged 5 years or older.
- Virological failure is defined as a persistent viral load above 5000 RNA copies/mL, after at least 24 weeks on ART, in a treatment-adherent child.

Principles

Virological failure is due to resistance mutations acquired either at the time of infection or because of poor adherence. Even a resistance test before starting treatment (not widely available) will not always show archived mutations (not in most of the virus tested). Acquired resistance is more likely if there is initial improvement (ideally documented virologically). A history of missed doses is usually not given readily, but it is essential that support to ensure 100% adherence is put in place before second-line treatment is started.

In the absence of routine CD4 or viral load assays, judgements should be made about treatment failure based on:

- Clinical progression
- CD4 decline as defined in Table 36.6.

Generally, patients should have received 6 months or more of ARV therapy, and adherence problems must be ruled out where possible before considering treatment failure and switching ARV regimens.

If an apparent deterioration is due to the immune reconstitution inflammatory syndrome (IRIS), this is not a reason for switching therapy. IRIS usually starts within weeks or the first few months after starting ART in children who have very low CD4 counts (< 15%). The most common initiator is TB which has been latent, but the symptoms of other opportunistic

infections can develop as the immune recovery enables a response. Treatment is of the infection, and ART should be continued.

Immune reconstitution inflammatory syndrome (IRIS)

It is important to differentiate IRIS clinically from treatment failure, because the symptoms may be similar. IRIS can be confused with several other clinical events that are also observed in children with advanced HIV disease, such as opportunistic infections, ARV-related toxicity, or HIV disease clinical progression.

What is IRIS?

IRIS is an exaggerated immune response to antigens or organisms. The related organisms could be mycobacteria (e.g., *Mycobacterium tuberculosis*, non-tuberculosis mycobacteria), viruses (e.g. herpes zoster, herpes simplex) or fungi (e.g. *Cryptococcus neoformans*).

Who is at risk of developing IRIS?

It usually occurs in a child with low baseline CD4 or WHO clinical stage 3 or 4 before initiation of ART. The incidence rate of IRIS could be as high as 15 – 25%.

When does IRIS develop?

It usually occurs during the first 6 months after initiation of ART, although it commonly manifests during the first month. During the initial period of ART, antiretroviral drugs cause a rapid decline in HIV viral load and a rapid rise in CD4, so a brisk immune response to antigen is developed.

What are the common manifestations of IRIS?

There are two types of IRIS:

‘Worsening type’:

- Clinical worsening of a previously treated opportunistic infection. For example:
 - Worsening of respiratory symptoms and/or chest X-ray finding in a child with previously treated pulmonary tuberculosis
 - Severe headache in a child with previously treated cryptococcal meningitis.

‘Unmasking type’:

- Unmasking of a previously subclinical infection with exaggerated inflammatory response. For example:
 - Suppurative lymphadenitis from *Mycobacterium*
 - Infection
 - Development of an abscess at the BCG vaccination site.

How should IRIS be managed?

1. ARVs should be continued.
2. For the ‘unmasking type’, the appropriate anti-infective agents are needed.
3. In most cases, the symptoms of IRIS resolve after a few weeks.
 - a. However, some reactions can be severe or life-threatening, requiring a short course of steroid treatment e.g.:
 - i. IRIS from pulmonary tuberculosis with acute respiratory distress syndrome (ARDS),
 - ii. IRIS from *M. avium* complex infection with high-grade fever and severe abdominal pain,
 - iii. IRIS from cryptococcal meningitis with a severe increase in intracranial pressure).

Second-line treatment regimens in the event of treatment failure

After failure of a first line PI or NNRTI-based regimen, DTG plus two NRTIs are recommended for second-line ART.

Third-line ART

Third-line regimens should include new drugs with minimal risk of cross-resistance to previously used regimens, such as integrase inhibitors and second-generation NNRTIs and PIs.

Patients on a failing second-line regimen with no new ARV options should continue with a tolerated regimen.

Strategies that balance the benefits and risks for children need to be explored when second-line treatment fails.

For older children and adolescents who have more therapeutic options available to them, constructing third-line ARV regimens with novel drugs used to treat adults, such as ETV, DRV and RAL, may be possible.

Children on a second-line regimen that is failing with no new ARV drug options should continue with a tolerated regimen. If ART is stopped, opportunistic infections still need to be prevented, symptoms relieved and pain managed.

Nutritional care and failure to thrive (see Appendix to this Section)

Nutrition is a long-term concern in all HIV-infected children. 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 to be evaluated.
- Regular vitamin A as per WHO guidelines.
- Supplementary feeding if possible (aim for 150 kcal/ kg/day).
- Exclude or treat *Candida*.
- Exclude or treat enteric infection.
- Consider zinc deficiency (see Section 55 Handbook 1).
- Consider fever.
- Consider depression.
- Consider pain.

Clinical management of nutrition

1. HIV-infected children should be assessed routinely for nutritional status, including weight and height, at scheduled visits, particularly after the initiation of ART.
2. HIV-infected children on or off ART who are symptomatic, who have conditions requiring increased energy (e.g. TB, chronic lung disease, chronic oral infections, malignancies) or who have weight loss or evidence of poor growth, should be provided with 25 – 30% additional energy.
3. HIV-infected children who are severely malnourished should be managed as per the guidelines for uninfected children and provided with 50 – 100% additional energy.
4. HIV-infected children should receive one recommended daily allowance of micronutrients daily. If this cannot be ensured through the diet, or there is evidence of deficiency, supplementation should be given.
5. HIV-infected infants and children should receive high dose vitamin A supplementation every 6 months between 6 and 59 months of age, as per the guidelines for uninfected children (see Section 55 Handbook 1).
6. HIV-infected children who have diarrhoea should receive zinc supplementation as part of management, as per the guidelines for uninfected children.
7. For infants and young children who are known to be HIV infected, mothers are strongly encouraged to exclusively breastfeed for 6 months and to continue breastfeeding as per recommendations for the general population (i.e. up to 2 years of age and beyond).

Respiratory disorders in children with HIV infection

Symptoms include cough, shortness of breath, fever, sweats, and cyanosis.

The aetiology of acute respiratory infections is similar to that of community-acquired infections in immunocompetent children (*Mycobacterium tuberculosis*, *Pneumococcus*, *Haemophilus influenzae*, *Staphylococcus aureus*, *Mycoplasma pneumonia*) (see Handbook 1 Sections, 25 and 29, and This handbook Section 51). However, children with HIV may require more prolonged courses of treatment.

Studies on the aetiology of pneumonia among HIV-infected children in resource-limited countries have identified *Streptococcus pneumoniae*, *Staphylococcus aureus*, *Haemophilus influenzae* and *Klebsiella* species as the major bacterial pathogens in HIV-infected children. HIV-negative children are affected by the same pathogens, although at lower rates. The post-mortem studies showed similar results, except that *H. influenzae* was slightly less prominent. *M. tuberculosis* was prevalent regardless of HIV status, reflecting its significance in resource-limited countries. From the limited data available, RSV and parainfluenza appear to be the most prevalent viral causes of pneumonia.

In cases of failed treatment, consider using a second- line antibiotic.

Treatment of recurrent infections is the same, regardless of the number of recurrences.

Specific HIV-related causes of infection and illness

***Pneumocystis jiroveci* (formerly *carinii*) pneumonia (PCP)**

- PCP should be suspected, and anti-pneumocystis therapy considered in any HIV-positive infant with severe pneumonia.
- Severe generalised pneumonia usually includes ventilation/perfusion mismatch and severe hypoxaemia.
- High fever is uncommon compared with bacterial pneumonia.
- PCP is most likely to develop in a child whose HIV infection occurred in the previous 12 months (the peak time is 4 – 6 months), or over 12 months if they have a low CD4 count and are not on co-trimoxazole prophylaxis.
- There is an absent or low-grade fever, non-productive cough and difficulty breathing.
- Signs include severe respiratory distress (tachypnoea, chest indrawing), which is disproportionate to findings on auscultation (usually normal breath sounds or only a few crackles).
- If an oxygen saturation monitor is available, check at rest and, if normal, again after exercise. The latter may show hypoxia or, if there is a severe infection, cyanosis.
- There may be a history of a poor response to 48 hours of first-line antibiotics, and elevated levels of lactate dehydrogenase.
- PCP is often the first clinical indicator of HIV infection and is a WHO clinical stage 4 criterion.
- Clinical and radiological signs are not diagnostic. However, a clear chest or diffuse chest signs on auscultation are typical with PCP infection, as is the presence of diffuse infiltrates and areas of hyperinflation rather than focal signs on a chest X ray.
- Induced sputum and nasopharyngeal aspiration are useful for obtaining sputum for examination. Nasopharyngeal aspirate has a low sensitivity with conventional staining techniques and requires PCR. Induced sputum techniques greatly increase the diagnostic yield.
- Beware the risk of infection being transmitted to operators, especially of multiple drug-resistant tuberculosis, for example: Bronchoalveolar lavage may provide a diagnosis if adequate resources are available.

Treatment of PCP

Severe disease (severe respiratory distress, severe hypoxia):

1. Treat with co-trimoxazole 60–90 mg/kg IV 12-hourly for a minimum of 7 days, followed by oral drugs in the same doses for another 2 weeks (IV if there is severe nausea).
2. In addition, give high-dose dexamethasone for the first 5 days (150 micrograms/kg/dose 6-hourly for 4 days) or prednisolone 0.5 mg/kg 12-hourly for 5 days, then 0.25 mg/kg 12-hourly for 5 days, then 0.25 mg/kg daily for 5 days.
3. The response usually occurs after more than 5 – 7 days of appropriate high-dose therapy.

Less severe disease:

1. Treat with oral co-trimoxazole 30 mg/kg 6-hourly for 21 days (trimethoprim (TMP) 5 mg/kg; sulfamethoxazole (SMX) 25 mg/kg).
2. If the child has a severe drug reaction, change to pentamidine (4 mg/kg once a day by IV infusion) for 3 weeks, or trimethoprim 5 mg/kg/dose orally 6-hourly and dapsone 100 mg/kg once a day for 21 days.
3. Continue co-trimoxazole prophylaxis (see below) on recovery and ensure that ART is being given.

Co-trimoxazole prophylaxis

Co-trimoxazole prophylaxis has been shown to be very effective in HIV-infected infants and children in reducing mortality and the likelihood of PCP as a cause of severe pneumonia.

PCP is now unusual in countries where prophylaxis is routine.

Co-trimoxazole also protects against common bacterial infections, toxoplasmosis, and malaria.

Who should be given co-trimoxazole?

- All HIV-exposed children (children born to HIV-infected mothers) from 4–6 weeks of age, whether or not they are part of a prevention of mother-to-child transmission (PMTCT) programme.
- Any child under 5 years old identified as HIV-infected, regardless of CD4 count.
- Any child over 5 years old with a CD4 count of less than 25%.
- Any child with a history of PCP.
- See Appendix Tables 36.12 and 36.13 for management.

For how long should co-trimoxazole be given?

HIV-exposed children:

- For the first year, or until HIV infection has been definitively ruled out and the mother is no longer breastfeeding.

HIV-infected children:

- Indefinitely where ARV treatment is not yet available.
- Where ARV treatment is being given, co-trimoxazole may only be stopped once clinical or immunological indicators confirm restoration of the immune system for 6 months or more (also see below).
- Based on current evidence, it is not yet clear whether co-trimoxazole continues to provide protection after immune restoration is achieved.
- If there is a history of PCP pneumonia, continue indefinitely.

Under what circumstances should co-trimoxazole be discontinued?

- If severe cutaneous reactions such as Stevens–Johnson syndrome occur with co-trimoxazole or other sulpha drugs, or if there is renal and/or hepatic insufficiency or severe haematological toxicity (severe anaemia or pancytopenia).
- It is contraindicated in children with glucose-6-phosphate dehydrogenase (G6PD) deficiency.
- In an HIV-exposed child, only after HIV infection has confidently been excluded:

- For a non-breastfeeding child under 18 months of age, this is by negative DNA or RNA virological HIV testing.
- For a breastfed HIV-exposed child under 18 months of age, negative virological testing is only reliable if performed 6 weeks after cessation of breastfeeding.
- For a breastfed HIV-exposed child over 18 months of age, negative HIV antibody testing 6 weeks after stopping breastfeeding.
- In an HIV-infected child:
 - If the child is on ARV therapy, co-trimoxazole can be stopped only when evidence of immune restoration has been obtained. Continuing co-trimoxazole may continue to provide benefit even after the child has clinically improved.
 - If ARV therapy is not available, co-trimoxazole should not be discontinued.

What doses of co-trimoxazole should be used?

- Recommended dosages of 6–8 mg/kg TMP once daily should be used.
- For children under 6 months of age, give 2.5 mL of suspension (40/200 mg in 5 mL) or 1 paediatric tablet (or ¼ adult tablet, 20 mg TMP/100 mg SMX: tablets can be crushed).
- For children aged 6 months to 5 years, give 5 mL of suspension or 2 paediatric tablets (or ½ adult tablet).
- For children aged 6–14 years, give 10 mL of suspension or 1 adult tablet.
- For children over 14 years, give 2 adult tablets.
- Use weight-band dosages rather than body-surface- area doses.
- If the child is allergic to co-trimoxazole, dapsone is the best alternative.
- Give dapsone after 4 weeks of age in an oral dose of 2 mg/kg/24 hours once daily.
- If the patient is G6PD-positive, consider giving pentamidine or atovaquone.

What follow-up is required?

Co-trimoxazole prophylaxis should be a routine part of the care of HIV-infected children and must be assessed for tolerance and adherence at all regular clinic visits or follow-up visits by healthcare workers and/or other members of the multidisciplinary care team.

It is suggested that initial clinic follow-up in children takes place monthly, and then every 3 months if co-trimoxazole is well tolerated.

Lymphocytic interstitial pneumonitis (LIP)

LIP is a non-infectious pulmonary disorder caused by white cell infiltration into alveolae. It is most common in children over 2 years old, and is a clinical stage criterion which is an indication for starting ART.

LIP is common in children (it occurs in at least 40% of children with perinatal HIV), but rare in adults (it occurs in about 3% of adults with HIV). Various studies in Africa have documented a 30–40% prevalence of LIP in HIV-infected children, and up to 60% prevalence in those with chronic lung disease. LIP is often mistaken for pulmonary TB (miliary) because of the chronic cough and the miliary-like pattern on chest X-ray.

Pathogenesis: Possible explanations for LIP include a co-infection of the lungs by HIV and Epstein–Barr virus (EBV), leading to immune stimulation with lymphoid infiltration and chronic inflammation.

The child is often asymptomatic in the early stages, but may later have a mild persistent cough, with or without difficulty in breathing, bilateral parotid swelling, persistent generalised lymphadenopathy, poor growth, hepatomegaly, and other signs of heart failure (tender hepatomegaly, bilateral pitting pedal oedema, loud second heart sound and finger clubbing). Chest auscultation may be normal, or there may be widespread crackles. It may produce severe ventilatory perfusion mismatch with hypoxaemia but may be asymptomatic.

There is an increased risk of lower respiratory tract infection, including bronchiectasis. It is also associated with parotid, adenoid and tonsillar enlargement (and may produce sleep-related upper airway obstruction; see Section 32 in Handbook 1).

LIP may be mistaken for miliary TB, but the child is systematically too well.

Suspect LIP if the chest X-ray shows a bilateral reticulonodular interstitial pattern that is prominent in the lower lobes, nodules less than 5 mm in diameter, a single patchy alveolar opacity, hyperinflation, or isolated bullae. It must be distinguished from pulmonary tuberculosis and bilateral hilar adenopathy (see Figure 36.4). Chest X-ray diffuse infiltrations and hilar lymphadenopathy persisting for more than 2 months despite antibiotic treatment are also a clue.

X-ray appearances are often more severe than the clinical features.



FIGURE 36.4 Chest X ray showing lymphocytic interstitial pneumonia (LIP): typical is hilar lymphadenopathy and lace-like infiltrates.

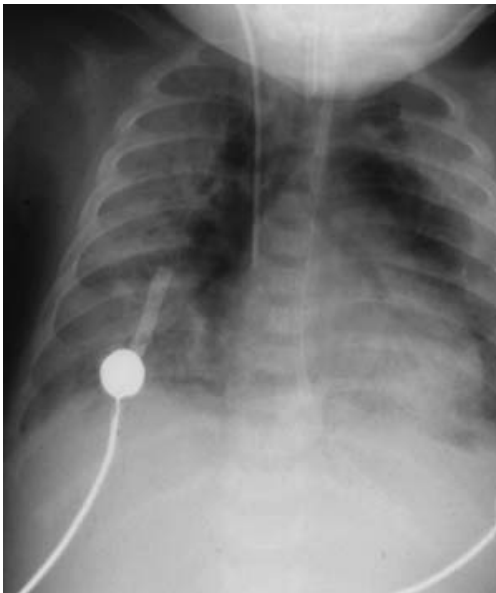


FIGURE 36.5 Pneumocystis jiroveci pneumonia (PCP): typical is a ground glass appearance.

Treatment of LIP

1. Give oxygen therapy during episodes of hypoxia.
2. Give a trial of antibiotic treatment for bacterial pneumonia before starting treatment with prednisolone.

3. Start treatment with steroids only if there are chest X-ray findings suggesting lymphocytic interstitial pneumonitis, plus any of the following signs:
 - a. Fast or difficult breathing
 - b. Cyanosis
 - c. Pulse oximetry reading of oxygen saturation < 90% (normal value is 94 or higher 93%).
4. Bronchodilators (e.g., salbutamol) are of benefit where wheezing is a problem.
5. For moderate symptoms give oral prednisone, 1–2 mg/kg daily for 3 days, and for more severe symptoms for up to 4 weeks. Then slowly decrease the dose over 2–4 weeks depending on the treatment response. If there is no response by 4 months, slowly taper the dose to stop over a further 2 months.
6. Only start steroid treatment if it is possible to complete the full treatment course (which may take several months depending on the resolution of signs of hypoxia), as partial treatment is not effective and could be harmful.
7. Beware of reactivation of TB.

Tuberculosis (see also Section 51)

In a child with suspected or proven HIV infection, it is important always to consider the co-diagnosis of tuberculosis, a diagnosis which is often difficult.

- Early in HIV infection, when immunity is not impaired, the signs of tuberculosis are similar to those in a child without HIV infection.
- Pulmonary tuberculosis is still the commonest form of tuberculosis, even in HIV-infected children.
- As HIV infection progresses and immunity declines, dissemination of tuberculosis becomes more common. Tuberculous meningitis, miliary tuberculosis and widespread tuberculous lymphadenopathy occur.
- All children with HIV should be screened for TB.
- Avoid, if practicable, children with HIV being in contact with a TB-infected person.

Isoniazid preventive therapy (IPT)

1. All HIV-infected infants and children who are exposed to TB through household contacts, but show no evidence of active disease, should begin isoniazid preventive therapy (IPT).
2. Children who are infected with HIV (living with HIV) who have either poor weight gain, fever, cough, or a contact with TB should be evaluated for active TB. If TB is excluded, give IPT.
3. Children living with HIV (over 12 months of age, and including those previously treated for TB), who are not likely to have active TB, and who are not known to be exposed to TB, should receive 6 months of IPT as part of a comprehensive package of HIV care.
4. Infants living with HIV, who have been exposed to TB but are evaluated as not having active TB, should receive IPT as part of a comprehensive package of HIV care.
5. The recommended dose of isoniazid (INH) for preventive therapy in HIV co-infection with TB is 10 mg/kg daily for 6 months (maximum 300 mg/day).
6. See the child monthly and give a 1-month supply of isoniazid at each visit.

Investigations

A tuberculin skin test (TST, Mantoux) is unreliable in HIV and should not be used. Since a definitive diagnosis of TB in children is difficult, there could be clinical features that are very suggestive of TB, leading to a high index of suspicion. In such cases a negative TST should not prevent you from starting anti-TB treatment.

Furthermore, several people develop TB infection when they come into contact with the TB pathogens, but they do not go on to develop signs and symptoms of TB disease, because their immune systems control the infection.

When the immune systems break down, such individuals develop signs and symptoms suggestive of TB disease. A TST can be positive in either state, and without signs and symptoms would be suggestive of TB infection and not TB disease.

- Chest X-ray: this may be normal, or it may show non-specific infiltrates, hilar or paratracheal lymphadenopathy, persistent opacities after an antibiotic trial, or a miliary pattern.
- Microscopy (alcohol and acid-fast bacilli, AAFB), Ziehl–Neelsen (ZN) stain) and culture:
 - This is the most important investigation.
 - Sputum which may need to be induced by saline nebuliser (in an isolation room with staff wearing a fine-particle (FP3) mask), and,
 - Gastric aspirate (if the child is coughing, take this in the early morning before they have had anything to eat or drink).
 - Collect at least three specimens.

Treatment of infants and children diagnosed with TB and HIV

Treat TB in HIV-infected children with the same anti-TB drug regimen as for non-HIV-infected children with TB.

Thiacetazone is associated with a high risk of severe, and sometimes fatal, skin reactions in HIV-infected children and must not be given. These reactions can start with itching, but progress to severe reactions.

Recommended ART regimens for children who need TB treatment

Recommended regimens for children and adolescents initiating ART while on TB treatment is ABC or AZT, 3TC and DTG

1. TB should be treated with standard regimes, the emphasis being on achieving high adherence rates.
2. The development of multi-drug-resistant TB is a very real threat if compliance is poor.
3. Directly observed therapy (DOT) may be the best approach.
4. Any child with active TB disease and HIV infection should begin TB treatment immediately, and start ART as soon as tolerated in the first 8 weeks of TB therapy, irrespective of CD4 count and clinical stage.
5. For all HIV-infected children, anti-TB therapy should be started immediately upon the diagnosis of TB, and ART should continue.

Bronchiectasis (see Section 18 here and Handbook 1 Section 39)

Suspect bronchiectasis if there is:

- A persistent cough productive of copious sputum (in vomit in young children) or,
 - Haemoptysis associated with fever, anorexia and failure to thrive.
 - There may be clubbing and localised coarse crackles on auscultation.
- Obtain a chest X-ray
- Acquire sputum for Gram stain and culture as well as AAFB; differential diagnoses include TB and LIP.
- Treatment consists of:
 - Physiotherapy with postural drainage,
 - Bronchodilators and,
 - Antibiotics may be required for 2 weeks.
 - Intravenous amoxicillin (50 mg/kg 6-hourly) and
 - Gentamicin (7.5 mg/kg daily)

Cytomegalovirus (CMV) infection

CMV can present with:

- Pneumonia with fever,
- Dry cough,
- Respiratory distress and,
- Hypoxia.

CMV also causes:

- Oesophagitis and Gastroenteritis presenting with nausea,
- Difficulty swallowing,
- Diarrhoea and vomiting.

CMV retinitis is often asymptomatic or may cause:

- Blurred vision,
 - Strabismus and,
 - Ultimately blindness.
 - The fundi show white perivascular infiltrates and haemorrhages, reduced acuity and field defects.
-
- A chest X-ray may show diffuse interstitial infiltrates.
 - Oesophageal endoscopy may show linear, localised or punctate ulcers.
 - Biopsies show typical inclusion cells.

Treatment is with Ganciclovir 6 mg/kg 12-hourly for 14 days.

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 and oesophageal problems

Oral candidiasis

This is the most common form of fungal infection and the most common orofacial manifestation encountered in HIV-infected children. It progresses to involve the oesophagus in 20% of cases and denotes significantly impaired T-cell function.

It presents as white plaques on mucosa that are difficult to remove, loss of taste, pain on swallowing, reluctance to eat, increased salivation and crying during feeds.

Treatment

1. Nystatin 100 000 IU/mL oral suspension, 1–2 mL four to six times a day for 7 days or
2. Local gentian violet 0.5% aqueous solution twice daily for 7 days (dissolve one teaspoonful (5 mL) of crystals in 1 litre of water, filter off the residue, and use within 7 days) or
3. Clotrimazole 1%, miconazole 2% gel, or amphotericin B suspension/lozenges three times daily or
4. Fluconazole 3–6 mg/kg on the first day, then 3 mg/kg (maximum 100 mg) daily for 1–2 weeks. If there is rare resistance to fluconazole, give ketoconazole oral tablets, 3.3–6.6 mg daily.

Oesophageal candidiasis

Oesophageal candidiasis is a stage 4 clinical feature indicating profound immune impairment (advanced HIV disease). The only clinical symptom may be reluctance to feed. It presents as difficulty or pain while vomiting or swallowing, reluctance to take food, excessive salivation, or crying during feeding. The condition may occur with or without evidence of oral candida.

If oral candida is not found, give a trial of treatment with fluconazole (3–6 mg/kg once a day). Exclude other causes of painful swallowing (e.g., cytomegalovirus, herpes simplex, lymphoma and, rarely, Kaposi's sarcoma), if necessary, by referral to a larger hospital where appropriate testing is possible.

Treatment

- 1 Give oral fluconazole, 3–6 mg/kg once a day for 7 days, except if the child has active liver disease.
- 2 Give amphotericin B, 0.5–1 mg/kg/dose once a day) by IV infusion for 10–14 days to children with liver disease and in cases where there is a lack of response to oral therapy, inability to tolerate oral medications, or the risk of disseminated candidiasis (e.g., in a child with leukopenia).

Viral oesophagitis

Herpes simplex virus (HSV)

Herpes simplex virus (HSV) infection may either be primary (herpetic gingivostomatitis) or secondary (herpes labialis). The prevalence of oral HSV infection ranges from 10% to 35% in adults and children with HIV infection. The presence of HSV infection for more than 1 month constitutes an AIDS-defining condition.

Clinical appearance

HSV infection appears as a crop of vesicles, usually localised on the keratinised mucosa (hard palate, gingiva) and/ or the vermillion borders of the lips and perioral skin. The vesicles rupture and form irregular painful ulcers. They may interfere with mastication and swallowing, resulting in decreased oral intake and dehydration.

Systemic therapy with antiviral agents is recommended. The treatment is more effective if it is instituted in the prodromal stage of infection. Treat with:

- Aciclovir 20–40 mg/ kg orally or IV four times daily for 7 days (maximum single dose is 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 an H₂-antagonist, such as omeprazole (see Section 28)

Omeprazole

Aged < 2 years 700 micrograms/kg to 3 mg/kg once daily up to a maximum of 20 mg.

Aged > 2 years, body weight 10–20 kg, give 10 mg once daily up to maximum 20 mg once daily.

Aged > 2 years, body weight > 20 kg, give 20 mg once daily up to a maximum of 40 mg once daily.

Idiopathic aphthous ulcers:

- If possible, these need to be differentiated from HSV by viral culture.
- Pay attention to oral hygiene.
- Thalidomide is useful if they are severe.

Severe periodontal and gingival disease (cancrum oris)

Periodontal (gum) disease is common among HIV-infected patients. It is characterised by bleeding gums, bad breath, pain or discomfort, mobile teeth, and sometimes sores. Its reported prevalence ranges widely, from 0% to 50%.

If left untreated, HIV-associated periodontal disease may progress to life-threatening infections, such as Ludwig's angina and noma (cancrum oris).

Noma is a gangrenous condition that primarily affects children. It is a multifactorial disease. The most important risk factors are poverty, chronic malnutrition, poor oral hygiene and severe immunosuppression. Although it is considered to be a preventable disease, noma has a case-fatality rate of 70 – 90% if left untreated.

Treatment of cancrum oris

- Benzylpenicillin 50 mg/kg 4-hourly or,
- Amoxicillin 40 – 60 mg/kg IV 8-hourly.
- Change to oral antibiotics once the child is able to swallow (usually after 24 – 48 hours).
- Provide materials for and education on dental hygiene.

Rarely, malignancy (Kaposi's sarcoma or non-Hodgkin's lymphoma) or oral hairy leukoplakia (white lacy markings on the sides of the tongue associated with Epstein-Barr virus infection; no treatment required) occur. Visceral Kaposi's sarcoma may present with persistent diarrhoea, intestinal obstruction, and abdominal pain.

Persistent diarrhoea (see Section 62 Handbook 1)

Case management should start with management of dehydration with oral rehydration solution. Dysentery (loose stools with blood) should be managed in the same way as for non-HIV-infected children (e.g., for *Shigella* infection). Concur with the local prevalence of treatable infections. Giardiasis, cryptosporidiosis, microsporidiosis, *Shigella*, *Salmonella*, *Campylobacter*, enteropathogenic *E. coli* and *Yersinia* may each contribute to gastrointestinal dysfunction.

HIV itself may cause an enteropathy, and in highly immuno-suppressed 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. All antiretroviral drugs (except AZT) can cause diarrhoea, particularly ritonavir.

Chronic or recurrent diarrhoea (see Section 62 Handbook 1)

Normal endemic pathogens may be responsible, such as rotavirus, *Giardia lamblia*, *Campylobacter jejuni* (see Section 29 and Section 62 Handbook 1), salmonellae (typhoid and non-typhoid), *E. coli*, *Shigella*, *Entamoeba histolytica* and *Strongyloides stercoralis*.

Investigations

- Fresh stool microscopy and culture: *Giardia*, *Entamoeba*.
- Ova and cysts: helminths.
- ZN stain: cryptosporidia, cyclospora.
- pH and reducing substances: lactose intolerance.
- CD4 count < 50: CMV, mycobacterium avium intracellulare
- CD4 count < 100: cryptosporidium, microsporidiosis.

- Look for signs of vitamin deficiencies (see Section 55 Handbook 1)
 - Vitamin A: night blindness, dry eyes, Bitot's spots (on conjunctivae).
 - Vitamin D: rickets (wide wrist, double malleoli, bowed legs, rachitic rosary, Harrison's sulcus).
 - Vitamin E: dry rough skin.
 - Vitamin K: ecchymosis, purpura.

Opportunistic infections such as those listed below may be responsible:

- Bacterial: atypical mycobacterial infections, such as
- 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, Candida. If severe, treat with fluconazole 3 mg/kg once daily.

Treatment

1. Diarrhoea may be secondary to antibiotics, either by direct effects or through Clostridium difficile.
 - a. Stop antibiotics as soon as possible.
 - b. Give live yoghurt with or without oral vancomycin.
 - c. If lactose intolerance is present, give lactose-free feeds.
2. Treat dysentery with:
 - a. Ciprofloxacin 15 mg/kg 12-hourly for 3 days, or,
 - b. Ceftriaxone 50 mg/kg once a day for 2–5 days, plus,
 - c. Metronidazole 7.5 mg/kg 8-hourly for 7 days.
3. Nutritional management includes calorie replacement, which may need to be nasogastric, but always encourage eating.
4. Ensure that additional food is not just recommended but actually given.
Vitamin A, multivitamins, and zinc (10–20 mg once a day for 10–14 days) supplementation may be of benefit (see Section 55 Handbook 1).

Prevention

1. Use hygienic practices during food preparation
2. Always use clean water
3. Avoid bird and animal faeces
4. Avoid swimming in fresh water, and,
5. Avoid reptiles (salmonellae).

Abdominal pain

This is most frequently related to infections, but occasionally is caused by tumours (non-Hodgkin's lymphoma and Kaposi's sarcoma).

Malabsorption

HIV can be directly associated with an enteropathy. Lactase deficiency and other brush-border defects can also be responsible. Consider trial of a lactose-free diet.

Central nervous system disorders

Many HIV-related CNS diseases have been described.

Primary CNS infection by HIV is quite common, as it is a neurotropic virus. Various abnormalities of the central nervous system (CNS) and peripheral nervous system (PNS) are associated with HIV and AIDS. These abnormalities may be attributable to the following causes:

- HIV infection,
- Complications related to immunosuppression,
- Neurotoxic effects of antiretroviral treatments, and,
- Other systemic complications of HIV that affect brain function.

Neurological disorders in people with HIV infection include peripheral neuropathies (nerve disorders that affect the limbs or feet and hands), myelopathy (disorders of the spinal cord), focal cerebral mass lesions (brain tumours such as CNS lymphoma), CNS complications of opportunistic infections, vascular (blood vessel) abnormalities, seizures and encephalopathies.

Developmental delays and regression are also important CNS-related problems in HIV-infected children.

The neurological manifestations of HIV infection include the following:

- Progressive or static encephalopathy
- Seizures
- Strokes
- HIV myopathy
- HIV myelopathy
- Peripheral neuropathy
- Psychiatric manifestations
- Sleep problems.

The effect of HIV on the brain ranges from severe effects, found in more than 50% of patients dying of AIDS at post-mortem, to the much more common and milder effects on the developing brain in children, which result in mild learning difficulties. It is particularly important to recognise this so that appropriate support can be given (e.g. reminding the patient to take ART).

TABLE 36.7 Comparison of features of major CNS mass lesions in HIV

Clinical			Neuroimaging (if available)		
Disease	Timing	Fever	No. of lesions	Type of lesions	Location of lesions
Cerebral toxoplasmosis	Acute onset of symptom	Common	Multiple	Enhancing spherical rings; mass effect	Basal ganglia
Primary CNS lymphoma	Insidious onset of symptom	Usually absent	One or few	Irregular shape, weakly enhancing, mass effect	Periventricular, periependymal, corpus callosum
Tuberculoma	Insidious onset of symptom	Common	One or few	Discrete lesions, significant surrounding oedema, mass effect	Supratentorial in adults, infratentorial in children (at the base of the brain near the cerebellum)
Cryptococcoma cryptococcal meningitis	Acute onset of symptom	Common	Variable	Mass lesions, dilated perivascular spaces, oedema	Basal ganglia

Care of HIV-infected children with CNS involvement requires a thorough evaluation and stepwise therapy according to the underlying aetiology. A multidisciplinary approach is usually needed for appropriate care.

Specific neurological problems

HIV encephalopathy

1. Rapid onset or chronic and relapsing forms.
2. Hypertonic (spastic) diplegia and expressive language delay.
3. Acquired microcephaly with developmental regression (loss of skills).
4. White-matter disease predominates.
5. It does not cause seizures, and therefore seizures need to be fully investigated for another pathology.

Encephalitis

Toxoplasma gondii

- Prevention:
 - Avoid cats and cat faeces
 - Avoid raw uncooked or partially cooked food
 - Can be acquired congenitally.
- Diagnosis:
 - CT/MRI of the brain, and serology (if available).
- Treatment:
 - Co-trimoxazole 60 mg/kg orally (IV if there is severe nausea) 12-hourly for 2 weeks.
 - Then give lifelong prophylaxis:
 - Sulfadiazine 85–120 mg/kg/day in two doses
 - Pyrimethamine 1 mg/kg/day (max mum 25 mg) and,
 - Folic acid 5 mg every 3 days.

JC virus (papovavirus)

The JC virus is associated with progressive multifocal leukoencephalopathy, a disease characterised by altered mental status, limb weakness, or both.

Patients may also exhibit personality changes with frequent emotional outbursts.

There is no treatment for this illness, but strong antiretroviral medications (if available) can sometimes improve the symptoms.

Fungal lesion

This is rare.

Diffuse CMV encephalitis

Treat with ganciclovir 5 mg/kg orally or IV 12-hourly for 14–21 days.

Malaria

See Section 31 Handbook 1.

TABLE 36.8 Neurological manifestations of paediatric HIV infection

Abnormality	Clinical findings	Diagnostic studies (if available)
Focal cerebral mass lesions	Headache, Nausea/vomiting, Motor deficits (usually asymmetrical), Discoordination, Visual changes, Altered mental status	CT/MRI: enhancing lesions Lumbar puncture: CSF may reveal abnormal cytology or Epstein–Barr virus via PCR Brain biopsy: sometimes needed to confirm diagnoses
Myelopathy	Gait disturbances, Lower-extremity weakness/ spasticity incontinence, Sensory abnormalities, Abnormal lower-extremity reflexes	CT/MRI on mass lesions seen; nerve-root thickening may be present CSF: polymorphonuclear pleocytosis

Abnormality	Clinical findings	Diagnostic studies (if available)
Myopathy	Muscle weakness, Muscle soreness, Weight loss	EMG: irritative myopathy Muscle biopsy: inflammation, degeneration
Opportunistic infections	Headache, Nausea/vomiting, Fever, Seizures, Altered mental status, Malaise	CT/MRI: multiple enhancing lesions (toxoplasmosis), periventricular and meningeal abnormalities (CMV)
Peripheral neuropathy	Distal symmetrical neuropathy Distal numbness/pain, Paraesthesia, Stocking/glove sensory loss, Decreased ankle reflexes	EMG: distal axonopathy
	Inflammatory demyelinating polyneuropathy Progressive weakness Paraesthesias Areflexia Mild sensory loss	EMG: demyelination
Peripheral neuropathy (cont.)	Progressive polyradiculopathy Lower-extremity weakness Paraesthesia's Urinary incontinence and retention Diminished reflexes	EMG: polyradiculopathy Serum: increased creatine kinase
Progressive encephalopathy	Fine and gross motor deficits (usually symmetrical), Abnormal tone, Neurodevelopmental delay, Microcephaly, Altered mental status	CT/MRI: brain atrophy, white-matter abnormalities

Abnormality	Clinical findings	Diagnostic studies (if available)
Seizures	Focal or generalised seizures, Post-ictal stare (fatigue and confusion following seizure)	EEG: abnormal patterns CT/MRI and CSF studies: mass lesions may be seen via imaging, and CSF may be positive for pathogens and abnormal cells if aetiology is infectious or neoplastic Lumbar puncture: may reveal infection
Strokes (cerebrovascular accidents)	Rapid onset of focal neurological signs, Seizures, Altered mental status	CT/MRI: extent of bleeding seen (in ischaemic strokes, CT may not show changes during the first 2 – 1 hours): contributing factors such as CNS neoplasms may be identified Lumbar puncture: with subarachnoid haemorrhages, blood will be present in the CSF

Meningitis

Bacterial meningitis

This has the usual spectrum of pathogens, such as Pneumococcus, Haemophilus influenzae, Meningococcus and Mycobacterium TB. (see Section 67 Handbook 1).

Viral meningitis/encephalitis

See Section 68 Handbook 1.

Cryptococcosis and other fungi

- Prevention:
 - Avoid bird faeces.
- Clinical features:
 - Chronic onset,
 - Headache (common),
 - Fever,
 - Meningism (usually but not always present), and,
 - There may be a change in mental state.
- Diagnosis:
 - Based on staining of CSF sample with Indian ink.
- Treatment:
 - Fluconazole 6–12 mg/kg orally or,

If there is severe nausea, IV once daily for 14 days.

There is a high relapse rate, therefore give prophylactic fluconazole 3 – 6 mg/kg/day or amphotericin IV 0.5 – 1.5 mg/kg/day for 14 days followed by oral fluconazole for 8 weeks.

Syphilis

Treat with benzylpenicillin IV 15 mg/kg every 6 hours for 10-15 days (20% of cases may have a systemic febrile response to penicillin).

Tuberculosis

See Section 51

Cerebral abscess (see Section 73 Handbook 1)

Acute bacterial or tuberculosis.

TABLE 36.9 Care guidelines for children with neurological manifestations of paediatric HIV infection

Abnormality	Care guidelines
Focal cerebral mass lesions	Assess for signs of increased intracranial pressure, fever, focal neurological signs and behavioural changes Administer chemotherapy or antibiotics as needed Provide support to family and education regarding specific medications needed by patients
Myelopathy	Assess for pain, muscle weakness, lower-extremity weakness, incontinence, and spasticity Administer HAART to reverse immune suppression Administer muscle relaxants as needed Provide physical therapy for weakened muscles and to maintain range of motion Teach exercises to the family so that they can help the patient at home
Myopathy	Assess for pain, muscle weakness and range of motion Consider discontinuing medications that may be contributing to the condition Administer corticosteroids and pain medications as needed Provide physical therapy for weakened muscles and to maintain range of motion Teach exercises to the family so that they can help the patient at home
Opportunistic infections	Assess for signs of increased intracranial pressure, fever, focal neurological signs and behavioural changes Administer appropriate medication based on the suspected or confirmed pathogen: Toxoplasmosis: pyrimethamine, sulfadiazine, clindamycin Cryptococcosis: fluconazole, flucytosine, amphotericin B Herpes simplex: aciclovir Cytomegalovirus: ganciclovir, foscarnet
Peripheral neuropathy	Assess for numbness, paraesthesias, pain and weakness Administer analgesics, tricyclic antidepressants, anticonvulsants and steroids as needed Provide support to the family and education regarding progression of symptoms

Abnormality	Care guidelines
Progressive encephalopathy	Assess for progressive motor dysfunction, and failure to reach or loss of age-appropriate milestones Administer antiretroviral medications and muscle relaxants as needed Assist with ambulation and activities of daily living Provide information to the family regarding progression of symptoms
Seizures	Assess for seizure activity Protect the patient from injury during seizure activity Monitor respiratory status: suction airway and administer oxygen as needed Administer anticonvulsant medications as needed Provide support to the family during seizures Educate the patient and the family about long-term use of anticonvulsant medications and seizure precautions (e.g. patients with seizures should never swim alone or climb to high places from which they could fall during a seizure)
Strokes	Intensive care, including neurosurgical intervention, is often needed immediately after a stroke occurs Look for contributing factors, such as low platelet levels, which may be correctable Assess for seizures Assist with ambulation and activities of daily living Provide physical therapy as needed Provide support and education to the family regarding the long-term prognosis

Skin disorders (see Section 27)

Cutaneous lesions are often the first manifestation of HIV noted by patients and healthcare professionals. These can be due to infectious or non-infectious causes.

Viral, bacterial and fungal infections have been very frequently reported in HIV-infected children. These usually tend to be more severe and resistant to therapy.

Common skin diseases may present with unusual skin lesions such as Norwegian scabies and disseminated, confluent and large lesions of molluscum contagiosum (see Table 36.10).

Seroconversion rash

Maculopapular erythematous rash (very rarely observed in infants).

TABLE 36.10 Common infectious and non-infectious skin lesions in paediatric HIV

Infectious disorders and lesions	Non-infectious disorders and lesions
Viral infections: <ul style="list-style-type: none"> • Herpes simplex, herpes zoster • Molluscum contagiosum • CMV • Warts Fungal infections: <ul style="list-style-type: none"> • Candida • Tinea onychomycosis Bacterial infections: <ul style="list-style-type: none"> • Impetigo • Scabies 	<ul style="list-style-type: none"> – Seborrhoeic dermatitis, – Atopic dermatitis – General dermatitis – Nutritional deficiency – Eczema – Psoriasis – Drug eruptions – Vasculitis – Alopecia

Viral infections

Varicella

Chickenpox: can be very severe (affecting the lungs and brain) or even fatal.
 Herpes zoster: can involve single or multiple dermatomes and may affect the eyes.

Treatment:

1. IV aciclovir (poorly absorbed by the oral route):
 - a. Age < 3 months: 10 mg/kg 8-hourly.
 - b. Age > 3 months: 20 mg/kg 8-hourly.
2. Valacyclovir is the prodrug of aciclovir and achieves better blood levels orally and is an alternative to IV aciclovir (if available).
3. VZIG within 96 hours of contact (if available).

HSV 1 and 2

Infection appears as a crop of localised vesicles. It may affect the lips, mouth and anogenital areas (rare in children unless sexually abused). The vesicles rupture and form irregular painful ulcers. They may interfere with mastication and swallowing, resulting in decreased oral intake and dehydration. May be recurrent and severe.

Treat with oral aciclovir, 20 mg/kg four times daily for 5 – 7 days (maximum single dose 800 mg).

Molluscum contagiosum

Umbilicated papular lesions.

Treat with ARVs (no other measures are effective). If neglected, giant lesions can result which require surgical excision.

Measles

Prevent by immunisation (see below and Section 15 Handbook 1).

- 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), and ARVs are the only effective treatment.

Bacterial infections

Impetigo and furunculitis due to *Staphylococcus aureus* are common. Treat with flucloxacillin, 12.5–25 mg/kg four times daily orally, or a first-generation cephalosporin such as **cephradine**.

Fungal infection

Fungal infection is common and involves the feet, hands and groin.

- Treat with topical imidazole (e.g. miconazole 2% twice daily until healed) or terbinafine cream. If severe, widespread or for nail infections, use itraconazole or terbinafine.
- Treatment will be needed for 4–6 weeks.
- Antifungal drugs commonly used in paediatric patients include the following:

Itraconazole:

- Children aged 1 – 12 years:
 - Course ('pulse') of 5 mg/kg (maximum 200 mg) daily for 7 days, with subsequent courses repeated after 21-day intervals.
 - Fingernails need two courses
 - Toenails need three courses.
- For children aged 12 – 18 years:
 - Either 200 mg once daily for 3 months or course ('pulse') of 200 mg twice daily for 7 days, with,
 - Subsequent courses repeated after 21-day intervals.
 - Fingernails need two courses
 - Toenails need three courses.

Terbinafine: children aged over 1 year:

- Body weight 10 – 20 kg, 62.5 mg once daily.
- Body weight 20 – 40 kg, 125 mg once daily.
- Body weight over 40 kg, 250 mg once daily, for 6 weeks to 3 months.

Fluconazole: 6 mg/kg weekly.

Griseofulvin: 20 – 25 mg/kg/day (micro-size formulation) or 10 – 15 mg/kg/day (ultra-micro-size formulation) for 6 – 12 weeks.

Seborrhoeic dermatitis and pityriasis versicolor

Seborrhoeic dermatitis occurs in up to 85% of adults and children with HIV infection. It may be an early sign of HIV. It is caused by the yeast *Malassezia furfur*.

It is characterised by thick yellow hypopigmented scaly macules occurring on the scalp but also on the face or in the diaper (nappy) area. Older children may also have involvement of the nasolabial folds, the skin behind the ears, and the eyebrows.

Treatment

1. Selenium-based or ketoconazole shampoo
2. Topical coal tar or antifungal creams
3. Aqueous cream
4. UVB light therapy or salicylic acid
5. To decrease inflammation, 1% hydrocortisone cream can be applied to the affected area (except for the face) three times per day
 - a. Parents should be instructed to use 1% hydrocortisone cream sparingly in the diaper area
6. If the condition is severe, give oral fluconazole 3 mg/ kg once daily.

Non-specific pruritic papular rash

This is a common and severe problem in children with HIV infection. In a previously untested child, it should be an indication for HIV testing. In a child who is known to have HIV, the CD4 count should be checked, and ARV started if appropriate.

Treatment

1. Bathe in a skin antiseptic wash (e.g. dilute chlorhexidine solution)
2. Antihistamines: give chlorpheniramine:
 - a. Age < 1 year: 1 mg twice daily
 - b. Age 1–5 years: 1–2 mg three times daily
 - c. Age 6–12 years: 2–4 mg three times daily
 - d. Age > 12 years: 4 mg three times daily
3. Aqueous cream and calamine lotion may be of benefit.

Drug side effects

- Drug eruptions can occur in patients who are receiving treatment for HIV infection. These can be severe (e.g. erythema multiforme, Stevens–Johnson syndrome, toxic epidermal necrolysis).
- Drug side effects are most common with co-trimoxazole, sulfadiazine, anti-tuberculous drugs (e.g. thiacetazone, which is contraindicated in HIV infection), penicillin, cephalosporins and dapsone.
- Drug eruptions usually appear as pink to erythematous papules that run together and create a blotchy appearance, but may include elevated patches (hives), mucous membrane ulceration, scaling and light sensitivity.
- NRTIs (nevirapine and efavirenz) have been associated with pruritic maculopapular eruptions. Most eruptions are mild, and the medication can be continued with eventual spontaneous resolution of the eruption.
- To promote comfort, the patient can be given oral antihistamine such as diphenhydramine hydrochloride 1 mg/kg
- every 6 hours. In more severe cases, the eruptions resolve when the medication is discontinued.
- Most drug eruptions are mild and resolve after the causative medication is discontinued.

Infestations

Scabies *Sarcoptes scabiei* (see Section 27) may present as in children without HIV infection. It is characterised by pruritic papular lesions that most commonly occur in the webs of the fingers and toes, the folds of the wrist, the antecubital area and the axilla. Infants may also have lesions on the palms and soles of the feet. Scrapings observed under a microscope may reveal the mite, eggs or faeces. HIV-infected patients with advanced disease can experience a variant of scabies called Norwegian scabies. This type of scabies is characterised by generalised scaling and enlarged crusted plaques.

Treatment

Consists of an application of topical benzyl benzoate lotion, 25%, which is left on the skin to dry and repeated the next day. After a patient is treated for scabies, the family should be advised to wash all clothing and bedclothes in hot water and iron them to kill any mites that may be living in the cloth.

Malignancy

Consider Kaposi's sarcoma in children presenting with nodular skin lesions, diffuse lymphadenopathy and lesions on the palate and conjunctiva with peri-orbital bruising. The diagnosis is usually clinical but can be confirmed by a needle biopsy of skin lesions or a lymph node biopsy. Suspect Kaposi's sarcoma also in children with persistent diarrhoea, weight loss, intestinal obstruction, abdominal pain or a large pleural effusion. Consider referral to a larger hospital for management.

Eye involvement (see Section 20)

- Malignancy (e.g. non-Hodgkin's lymphoma, Kaposi's sarcoma).
- HIV retinopathy: this is a microangiopathy with soft exudates. It is asymptomatic and does not require treatment. It needs to be differentiated from tuberculosis.

- Cytomegalovirus retinitis: this 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: this may produce corneal ulceration and retinal necrosis.
- Toxoplasmosis: this usually causes CNS disease and reactivation disease and may cause visual problems or blindness.
 - Treatment includes pyrimethamine, 1–2 mg/ kg/day orally for 2 days, then 1 mg/kg/day orally for 2 months, then 1 mg/kg/day orally for 3 days a week (maximum 50 mg).
 - Secondary prophylaxis may also be given to prevent reactivation.

Prophylaxis for any opportunistic infections consists of the following:

- Primary prophylaxis: giving medication to prevent infection that has not yet occurred.
- Secondary prophylaxis: giving medication to prevent recurrence of an infection after an episode.
- Prophylaxis can often be stopped after sustained immune reconstitution secondary to ART, but not in all cases.

Mycobacterium avium complex (MAC)

This produces a systemic infection with fever, chronic diarrhoea, abdominal pain, chronic malabsorption, generalised lymphadenopathy and obstructive jaundice (from lymph node enlargement around the porta hepatis).

Treatment

1. Clarithromycin 7.5 mg/kg twice daily IV or orally or azithromycin 10 mg/kg once daily and ciprofloxacin and rifabutin.
2. Consider prophylaxis with the above drugs if CD4 cell counts are persistently < 50/mm³ despite antiretroviral therapy, as the risk of MAC is high.
3. The opportunistic infection guidelines recommend that all HIV-infected individuals with CD4 counts of < 50 cells/ mm³ should have primary prophylaxis against disseminated MAC initiated.
4. Prior to initiating prophylaxis, patients should be evaluated for active MAC infection by clinical assessment.

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 blood transfusion for malaria-associated anaemia.
- Have a low threshold for diagnosing septicaemia and meningitis and giving powerful empirical antibiotics if severe sepsis is suspected.
- Consider tuberculosis and non-Hodgkin's lymphoma.

Immunisation

Early immunisations can help HIV-infected children who are more likely to acquire diseases that are preventable by immunisation because of their compromised immune system. Appropriate immunisations vary according to geographical location.

Routine immunisations appear to be generally safe for children with HIV infection without fever. Although immune responses may be suboptimal in some HIV-infected children, because of the severe nature of infections and associated mortality, routine immunisation of all children with HIV exposure or confirmed HIV infection is recommended with few exceptions.

Immunisations should generally follow the Expanded Programme on Immunisation (EPI) scheme. The current EPI schedule includes DTP, OPV, hepatitis B, Haemophilus influenzae type B vaccine (Hib) and measles vaccine. The difference in HIV-infected children is an extra dose of measles vaccine to be given at the age of 6 months. BCG and yellow fever vaccines 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, as 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.

Because most HIV-positive children have an effective immune response in the first year of life, EPI should be started as early as possible after the recommended age of vaccination.

There are theoretical risks associated with 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 the WHO for children in resource-limited 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. The WHO recommends giving an extra dose of measles vaccine at 6 months, as well as the standard dose at 9 months, to HIV-infected children. It is important to be aware of measles in the differential diagnosis of any child with fever or pneumonia and HIV, as a typical morbilliform rash and standard symptoms may not be present.

Other non-EPI vaccines are encouraged and recommended, especially in children whose immune systems have recovered. These more expensive but strongly recommended vaccines include MMR, pneumococcal conjugate vaccine, hepatitis A, typhoid and varicella vaccines. Diseases caused by these organisms have a greater propensity to cause severe life-threatening infections in HIV-infected children.

Terminal care of children dying from HIV infection See also Section 7.

Despite the increased availability and effectiveness of ARVs, death is still a possible outcome of HIV/AIDS. Each year, millions of children lose one or both parents to AIDS. Although relatives often go to heroic lengths to provide orphans with food, shelter and housing, often the children's psychosocial needs are overlooked, and these young people are not given full recognition or support after their loss. This is usually due to the belief that children are too young to understand what is happening or are better off not dwelling on their loss. Consequently, they are not properly supported in their time of mourning. Local groups for the support of families with HIV infection are essential, and ideally should be funded by local government.

Give end-of-life (terminal) care if:

1. The child has had a progressively worsening illness
2. Everything possible has been done to treat the presenting illness.

Keep up to date on how to contact local community-based home care programmes and HIV/AIDS counselling groups. Find out whether the carers are receiving support from these groups. If not, discuss the family's attitude towards these groups and the possibility of linking the family with them.

Pain control for children with HIV (See Section 9 Handbook 1.)

Need for referral

Often the facilities or expertise that are needed will not be available at the health centre or hospital to which a child with suspected or confirmed HIV has come for treatment.

If the child is not suffering from a life-threatening condition that requires urgent treatment, and referral can be arranged, it is advisable to refer the child to a paediatric infectious disease specialist or HIV treatment centre for the following:

- HIV testing with pre and post-test counselling
- Further investigations to confirm the diagnosis
- Evaluation of immunological status and the need to initiate ART
- Management of complicated HIV-related conditions and infections
- Evaluation of possible treatment failure
- Second-line treatment if there has been little or no response to treatment
- HIV medication-related toxicities
- HIV-related expert counselling.

Summary

The major practical focus should be on prevention of childhood HIV infection. This means implementing effective strategies for reduction of mother-to-infant transmission, such as prenatal screening of mothers and administration of ARV drugs for mother and baby.

Unfortunately, establishing the infrastructure that is required to implement effective interventions is lagging far behind the scientific advances in this field. Surmounting the sense of hopelessness among health-care 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.

Limiting the use of blood transfusions and ensuring that the blood supply is safe, and preventing sexual transmission among adolescents, are vital public health issues.

Positive education is required to encourage testing in the knowledge that there is now accessible safe treatment to keep children alive so that they can have a full healthy life. The key to successful treatment is 100% adherence. Do not start outpatient treatment until the child's carer and ideally all the family have expressed a commitment to treatment. Choose ART regimes which are simple and ensure that there is no problem with swallowing.

Predict growth for dosing so that the child is never under-dosed.

Frequent review is necessary to re-emphasise the importance of adherence and education of young people.

It is essential that resource-limited countries are permitted by multinational drug companies to develop low-cost and effective forms of HAART, without being limited by international patent regulations.

Without question, health system strengthening is essential if the advent of ARVs for all is to be adequately managed.

This is a very optimistic time in the field of paediatric HIV, with the potential to aim for eradication of mother-to-child transmission, and to provide successful treatment.

Appendix

TABLE 36.11 Monitoring children on ART

Item	Before or at ART initiation	Month (M) 1	M2	M3	M4	M5	M6	Every 6-12 months	Symptom-directed
Clinical evaluation: History and physical examination (including neurodevelopment)	X	X	X	X	X	X	X	X	X
Weight and height	X	X	X	X	X	X	X	X	
Calculation of ART dose ¹	X	X	X	X	X	X	X	X	
Concomitant medications ²	X	X	X	X	X	X	X	X	
Check ART adherence ³		X	X	X	X	X	X	X	
Haemoglobin and white blood cell count ⁴	X								X
Full chemistry									X
CD4% or count ⁶	X							X	X
HIV viral load measurement	7								X

⁶If signs of clinical progression of disease are seen, the CD4 count should be done earlier.

The child should be seen again within 1 week of starting ART to resolve any problems. If the child has missed a visit, attempts should be made to call or visit the child's home. In addition to these suggested appointments, caregivers should be encouraged to bring the child in if he or she is sick, especially during the first few months of ART when the child may experience ART side effects and intolerance.

1. Children may show rapid weight and height gain after ART, in addition to expected normal growth. Therefore, the ART dose should be recalculated at every visit. Underdosing of ART can lead to the development of resistance.
2. Concomitant drugs should be asked for at every visit to ensure that the child is on appropriate CTX dosing (if indicated) and is not taking drugs that have potential interactions with ART.
3. ART adherence can be assessed by asking questions about missed doses and the times when the child takes ART. Performing a pill count is time consuming, but may give a more accurate indication of adherence, if done correctly.
4. Haemoglobin (Hb) and white blood cell count (WBC) monitoring may be considered in children on ZDV at 1, 2 and 3 months.
5. Full chemistry includes but is not restricted to liver enzymes, renal function, glucose, lipids, amylase, lipase and serum electrolytes. Monitoring depends on symptoms and regimens. Regular liver enzyme monitoring during the first 3 months of treatment may be considered for certain children using nevirapine-based regimens, in particular for adolescent girls with a CD4 count of > 250 cells/mm³, and for infants and children who are co-infected with hepatitis B or hepatitis C virus, or other hepatic disease.
6. TLC is not suitable for monitoring of therapy; therefore, it cannot be a substitute for CD4. On the other hand, whereas both CD4 and viral load are used for routine monitoring of therapy, CD4 is less preferred to viral load.
7. Viral load is the preferred method of monitoring therapy. It should be done every 6 to 12 months

Treatment of PCP infection**TABLE 36.12** Starting co-trimoxazole (CTX) prophylaxis for Pneumocystis jiroveci pneumonia (PCP)

HIV-exposed infants and children	All ages groups
CTX prophylaxis is universally indicated, starting at 4 - 6 weeks after birth, and maintained until cessation of risk of HIV transmission and exclusion of HIV infection	CTX prophylaxis is indicated regardless of CD4 percentage or clinical status

Patient information: It needs to be explained to patients that although CTX does not cure HIV, regular dosing is essential for protection of children from infections that are more common or more likely to occur in HIV infection. CTX does not replace the need for antiretroviral therapy.

TABLE 36.13 Dosing for PCP: once-daily CTX dosing

Weight	Suspension: 40 mg TMP + 200 mg SMX/5 mL	Tablets (SS): 80 mg TMP/400 mg SMX	Tablets (DS): 160 mg TMP/800 mg SMX
1–4 kg	2.5 mL	–	–
5–8 kg	5 mL	½ tablet	–
9–16 kg	10 mL	1 tablet	½ tab
17–50 kg	20 mL	2 tablets	1 tablet
> 50 kg	20 mL	2 tablets	1 tablet

Summary of nutritional recommendations and support for HIV-infected children

- Regular growth monitoring.
- Safe infant feeding advice (the emphasis is on an exclusive infant feeding option). Substitute feeds if they are acceptable, affordable, feasible, sustainable and safe, otherwise exclusive breastfeeding, and early weaning. Avoid all mixed feeding.
- Dietary counselling for asymptomatic children to increase energy intake by 10% compared with HIV-uninfected children.
- Dietary counselling for symptomatic children to increase energy intake by 20 - 30% compared with HIV-uninfected children.
- Counselling on the importance of a balanced diet, including affordable choices from all food groups (micronutrient requirement of 1 RDA for age).
- Counselling on high-energy affordable food options for children with growth failure.
- Counselling on the use of clean water and hygienic food preparation.
- Vitamin A supplementation to prevent vitamin A deficiency in children aged 6 to 59 months with dosing schedule as follows:
 - Children aged 6–11 months: 100 000 IU (30 mg) once every 6 months.
 - Children aged 12–59 months: 200 000 IU (60 mg) once every 6 months.
- Zinc supplementation during diarrhoeal episodes: 10 mg once daily for 10 days in children older than 6 months if weight is ≤10 kg, and 20 mg if weight is > 10 kg.
- Assessment and management for underlying HIV- associated illnesses.
- Assessment for need to initiate ART.
- Referral to outreach service providers for food assistance, if needed.

Additional Tables

[Table 1 Simplified dosing of child friendly fixed dose solid formulations for twice daily dosing in infants and children 4 weeks of age and older. P.391](#)

[Table 2 Simplified dosing of child friendly fixed dose solid formulations for once daily dosing in infants and children 4 weeks of age and older. P.392](#)

[Table 3 Simplified dosing of child friendly solid and oral liquid formulations for twice daily dosing in infants and children 4 weeks of age and older. P.393](#)

[Table 4 Drug dosing of liquid formulations in infants less than 4 weeks of age. P.394](#)

[Table 5. Simplified dosing of isoniazid and co-trimoxazole prophylaxis for infants and children who are at least 4 weeks of age. P.394](#)

All of the above tables are found in the WHO publication *Consolidated guidelines on the use of antiretroviral drugs for treating and preventing HIV infection*.

More information on ART drug treatments below

TABLE 36.14 Nucleoside analogue reverse transcriptase inhibitors (NRTIs)

Name of drug	Formulation	Age	Age (weight), dose and dose frequency	Other comments
Zidovudine (ZDV) AZT	Syrup: 10 mg/mL Capsules: 100 mg, 250 mg Tablet: 300 mg	All ages	< 4 weeks: 4 mg/kg/dose twice daily 4 weeks to 13 years: 180–240 mg/m ² /dose twice daily Maximum dose: ≥13 years: 300 mg/dose twice daily	Large volume of syrup is not well tolerated in older children, Syrup needs to be stored in glass jars and is light sensitive Can give with food Doses of 600 mg/m ² /dose per day required for HIV encephalopathy Capsule can be opened, and its contents dispersed, or tablet crushed, and its contents mixed with a small amount of water or food and immediately taken (solution is stable at room temperature) Do not use with d4T (stavudine) (antagonistic antiretroviral effect)
Lamivudine (3TC)	Oral solution: 10 mg/ mL Tablet: 150 mg	All ages	< 30 days: 2 mg/kg/dose twice daily ≥30 days or < 60 kg: 4mg/kg/dose twice daily Maximum dose: 60 kg: 150 mg/dose	Well tolerated Can give with food Store solution at room temperature (use within 1 month of opening) Tablet can be crushed, and its contents mixed with a small amount of water or food and immediately taken

Name of drug	Formulation	Age	Age (weight), dose and dose frequency	Other comments
Fixed-dose combination of ZDV plus 3TC	No liquid formulation available Tablet: 300 mg ZDV plus 150 mg 3TC	Teens and adults	Maximum dose: 13 years old or weight > 60 kg: 1 tablet per dose twice daily (should not be given if weight is < 30 kg)	Ideally, tablet should not be split Tablet can be crushed, and its contents mixed with a small amount of water or food and immediately taken At weights of < 30 kg, ZDV and 3TC cannot be dosed accurately in tablet form
Abacavir (ABC)	Oral solution: 20 mg/ mL Tablet: 300 mg	Over 3 months	< 16 years or < 37.5 kg: 8 mg/kg/dose twice daily Maximum dose: > 16 years or ≥37.5 kg: 300 mg/dose twice daily	Can give with food Tablet can be crushed, and its contents mixed with a small amount of water or food and immediately ingested Warn parents about hypersensitivity reaction. ABC should be stopped permanently if a hypersensitivity reaction occurs

Table 36.15 Non-nucleoside reverse transcriptase inhibitors (NNRTIs)

Name of drug	Formulation	Age	Age (weight), dose and dose frequency	Other comments
Nevirapine (NVP)	Oral suspension: 10 mg/mL Tablet: 200 mg	All ages	15–30 days: 5 mg/kg/dose once daily for 2 weeks, then 120 mg/m ² /dose twice daily for 2 weeks, then 200 mg/m ² / dose twice daily 30 days to 13 years: 120 mg/m ² /dose once daily for 2 weeks, then 120–200 mg/ m ² /dose twice daily Maximum dose: > 13 yrs: 200 mg/dose once daily for first 2 weeks, then 200 mg/dose twice daily	If rifampicin co-administration, avoid use Store suspension at room temperature, but it must be shaken well Can give with food Tablets are scored and can be divided into two equal halves to give a 100 mg dose; can be crushed and combined with a small amount of water or food and immediately administered Warn parents about rash

Name of drug	Formulation	Age	Age (weight), dose and dose frequency	Other comments
				Do not dose escalate if rash occurs (if mild or moderate rash, hold drug; when rash has cleared, restart dosing from beginning of dose escalation; if severe rash, discontinue drug)
Efavirenz (EFV)	Syrup: 30 mg/mL (note that syrup requires higher doses than capsules; see dosing chart) Capsules: 50 mg, 100 mg, 200 mg	Only for children over 3 years of age or who weigh > 10 kg	Capsule (liquid) dose: 10–15 kg: 200 mg (270 mg = mL) once daily 15–19 kg: 250 mg (300 mg = 10 mL) once daily 20–24 kg: 300 mg (360 mg = 12 mL) once daily 25–32 kg: 350 mg (450 mg = 15 mL) once daily 33–39 kg: 400 mg (510 mg = 17 mL) once daily Maximum dose: ≥40 kg: 600 mg once daily	Capsules may be opened and added to food, but have a very peppery taste; however, can mix with sweet foods or jam to disguise taste. Can give with food (but avoid giving after high-fat meals which increase absorption by 50%) Best given at bedtime, especially in the first 2 weeks, to reduce CNS side effects

TABLE 36.16 Protease Inhibitors (PIs)

Name of drug	Formulation	Age	Age (weight), dose and dose frequency	Other comments
Lopinavir/ritonavir (LPV/r)	Oral solution: 80 mg/ mL lopinavir plus 20 mg/mL ritonavir Note that oral solution contains 42% alcohol Capsules: 133.3 mg lopinavir plus 33.3 mg ritonavir	6 months or older	6 months to 13 years: 225 mg/m ² LPV and 57.5 mg/m ² ritonavir twice daily OR Weight-based dosing as follows: 7–15 kg: 12 mg/kg LPV and 3 mg/kg ritonavir twice daily 16–40 kg: 10 mg/kg lopinavir and 5 mg/kg ritonavir twice daily	Preferably oral solution and capsules should be refrigerated. However, they can be stored at room temperature up to 25°C (77°F) for 2 months; At temperatures >25°C (77°F) the drug degrades more rapidly Liquid formulation has low volume but bitter taste

Name of drug	Formulation	Age	Age (weight), dose and dose frequency	Other comments
			<p><i>Maximum dose:</i> >40 kg: 400 mg LPV and 100 mg ritonavir (3 capsules or 5 mL) twice daily</p>	<p>Capsules are large</p> <p>Capsules should not be crushed or opened, but must be swallowed whole</p> <p>Should be taken with food</p>
Darunavir plus Ritonavir (RTV)	75 mg (white), 150 mg (white), 400 mg (light orange), 600 mg (orange)	3 years and older	<p>Protease inhibitor (PI) experienced</p> <p>3–6 years: <i>10–11 kg:</i> 200 mg twice a day + RTV 32 mg twice a day</p> <p><i>11–12 kg:</i> 220 mg twice daily + RTV 32 mg twice a day</p> <p><i>12–13 kg:</i> 240 mg twice a day + RTV 40 mg twice a day</p> <p><i>13–14 kg:</i> 260 mg twice a day + RTV 40 mg twice a day</p> <p><i>14–15 kg:</i> 280 mg twice a day + RTV 48 mg twice a day</p> <p>> 6 years: <i>15–30 kg:</i> 375 mg twice a day + RTV 50 mg twice a day</p>	DRV and RTV levels reduce in combination

Name of drug	Formulation	Age	Age (weight), dose and dose frequency	Other comments
			31–40 kg: 450 mg twice a day + RTV 60 mg twice a day > 40 kg: 600 mg twice a day + RTV 100 mg twice a day	

Table 36.17 Integrase inhibitors

Name of drug	Formulation	Age	Age (weight), dose and dose frequency	Other comments
Raltegravir	400 mg tablets (pink)	> 6 years	> 25 kg: 400 mg twice a day	With or without food Avoid indigestion remedies
Dolutegravir	50mg	1 month and older (depending on availability of appropriate formulations)	>20 kg: 50mg once a day	-

TABLE 36.18 Side effects of ARVs

Drug		Side effects	Comments
Nucleoside analogue reverse transcriptase inhibitors (NRTIs)			
Lamivudine	3TC	Headache, nausea, abdominal pain, diarrhoea, fatigue, pancreatitis	Well tolerated, can be crushed
Zidovudine	AZT	Headache, anaemia, neutropenia, nausea, hepatitis, neuropathy, nail pigmentation	Do not use with d4T (antagonistic ARV effect)
Abacavir	ABC	Hypersensitivity reaction, with fever, mucositis and rash; this is rare, but if it occurs stop the drug	Tablets can be crushed
Non-nucleoside reverse transcriptase inhibitors (NNRTIs)			
Efavirenz	EFV	Vivid dreams, sleepiness, rash, mood changes, hypercholesterolaemia	Take at night, avoid taking with fatty food

Drug		Side effects	Comments
Nevirapine	NVP	Rash (Stevens–Johnson syndrome), liver toxicity (check liver function tests at 2, 4 and 8 weeks)	When given with rifampicin, increase NVP dose by 30% or avoid use
Protease inhibitors (PIs)			
Lopinavir/ritonavir*	LPV/r NFV SQV	Diarrhoea, nausea, vomiting, headache Diarrhoea, vomiting, rash Diarrhoea, abdominal discomfort	Liquid: bitter taste. Take with food Take within 2 hours of food
Darunavir	DRV	Rash, nausea, diarrhoea, headache	
Atazanavir	ATV	Rash, nausea, jaundice, headache	Avoid antacids
Ritonavir*	RTV	Rash, nausea, diarrhoea, peri-oral paraesthesia, flushing, hepatitis	Liquid: bitter taste
Integrase inhibitor			
Raltegravir		Nausea, dizziness, insomnia, rash, pancreatitis, elevated ALT, AST, GGT	Avoid indigestion remedies
Dolutegravir	DTG	Weight gain and metabolic abnormalities (particularly glucose control) #	

*Requires cold storage and cold chain for transport.

References to whole section on HIV

PMTCT

[World Health Organization \(2018\) Updated recommendations on first-line and second-line antiretroviral regimens and post-exposure prophylaxis and recommendations on early infant diagnosis of HIV](#)

World Health Organization (2016) [Consolidated guidelines on the use of antiretroviral drugs for treating and preventing HIV infection](#) (Accessed 04/03/2021)

[World Health Organization \(2019\) HIV molecular diagnostics toolkit to improve access to viral load testing and infant diagnosis](#) (Accessed 04/03/2021)

[World Health Organization \(2016\) Updates on HIV and infant feeding](#) (Accessed 04/03/2021)

Paediatric HIV

World Health Organization (2015) Recommendation on 36 months isoniazid preventive therapy to adults and adolescents living with HIV in resource-constrained and high TB- and HIV-prevalence settings – 2015 update

[World Health Organization \(2018\) Updated recommendations on first-line and second-line antiretroviral regimens and post-exposure prophylaxis and recommendations on early infant](#)

[diagnosis of HIV: interim guidelines. Supplement to the 2016 consolidated guidelines on the use of antiretroviral drugs for treating and preventing HIV infection.](#) (Accessed 04/03/2021)

[World Health Organization \(2019\) Update of recommendations on first- and second-line antiretroviral regimens.](#) (Accessed 04/03/2021)

Section 37. Hydatid Disease

Introduction

The adult stage of the tapeworm *Echinococcus granulosus* lives in the gut of dogs and certain other carnivores. The usual intermediate hosts are herbivores. Humans may become an accidental intermediate host for the cystic stage of the parasite following ingestion of eggs in dog faeces contaminating the fingers, food or water. Because of the slow rate of growth of hydatid cysts, symptoms from infection in childhood often present in adulthood. Many cysts remain asymptomatic, eventually calcify and become sterile.

Epidemiology

The disease is widespread in sheep-farming countries and wherever there is intimate contact between humans and dogs or other canids, and where dogs scavenge dead animals or offal. There is a high incidence in the Turkana region of Kenya.

Clinical features

- Cysts may occur in virtually any organ.
- Many cysts are asymptomatic but may be palpable if they are large or superficial.
- Cyst rupture may cause anaphylaxis and/or spread by 'seeding' of scolices (heads of immature worms) resulting in 'daughter' cysts.
- Abdomen:
 - Palpable mass: liver (60% of all cysts), spleen, other intra-abdominal cysts.
 - Communication with the biliary tract: cholangitis, rigors, jaundice.
 - Abdominal pain.
 - Rupture from trauma.
 - Cyst rupture may cause anaphylaxis and/or spread by 'seeding' of daughter scolices (heads of immature worms).
 -
- 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. The presence of a separated membrane or daughter cysts makes the diagnosis highly likely. The condition needs to be differentiated from simple hepatic cysts.

1. Plain X-ray for lung or bone cysts. CT or MRI (if available) is also useful (e.g. for brain cysts).
2. Eosinophilia is present in <15% of cases. This may be due to cyst leakage or rupture.
3. Serology: specific IgG ELISA AgB (antigen-B-rich fraction) (if available) is most sensitive. Serology lacks sensitivity for extra-hepatic cysts (note that false-positive results are obtained in cysticercosis).
4. A urine antigen detection test appears promising.

Treatment

Calcified cysts require no treatment.

Medical treatment

- Albendazole is useful for patients with inoperable, widespread or numerous cysts, and

for patients unfit for surgery.

- Continuous treatment is now recommended (for up to 2 years). Its duration depends on the lesion's response.
 - The dose is 7.5 mg/kg orally twice daily.
 - The maximum dose is 400 mg twice daily.
- The absorption of albendazole is enhanced if it is taken with fatty meals.
- Albendazole plus praziquantel has greater protoscolicidal activity. The combination is successful for inoperable spinal, pelvic, abdominal, thoracic, or hepatic hydatid, and as an adjunct to surgery.
- Anthelmintics may reduce the need for surgery in uncomplicated pulmonary cysts.
- Patients undergoing surgery or PAIR (see below) should receive pre-operative albendazole (for 1 - 3 months) with or without praziquantel.

Percutaneous aspiration under ultrasound control

Puncture, aspiration, injection, re-aspiration (PAIR):

- The patient should be on albendazole for at least 4 weeks prior to PAIR.
- Following initial aspiration of the cyst, hypertonic saline is injected into the cyst and re-aspirated after 20 minutes.
- Percutaneous aspiration combined with an 8-week course of albendazole is more effective than either treatment alone.
- Laparoscopic treatment of liver and spleen hydatid is also effective.
- Contraindications to PAIR include cysts in the CNS or heart, and cysts communicating with the biliary tree, abdominal cavity, urinary tract or bronchi.

Surgery

Surgical removal is standard treatment if the lesion is accessible but is unsuitable for PAIR. The procedure is as follows:

1. The patient should be on albendazole for at least 4 weeks prior to surgery.
2. Pack around the cyst and avoid spillage of the cyst contents (there is a risk of anaphylaxis and seeding).
3. Drain the cyst, replace fluid with hypertonic saline, drain again, and then remove the cyst capsule.

High rates of recurrence and of surgical complications are recorded in inexperienced hands.

It is important to avoid hypertonic saline entering the bile ducts, as this may cause sclerosing cholangitis.

Prevention

- Ensure disposal of infected herbivore carcasses and offal.
- Treat dogs with praziquantel.
- Maintain strict hygiene and protect food and water from contamination.

Further reading

[World Health Organization \(2017\) Meeting of the WHO Informal Working Group on Echinococcosis \(WHO-IWGE\)](https://www.who.int/publications/i/item/WHO-HTM-NTD-NZD-2017.01)

<https://www.who.int/publications/i/item/WHO-HTM-NTD-NZD-2017.01>

Section 38. Leishmaniasis

Introduction

Leishmaniasis is caused by *Leishmania*, protozoa parasites whose reservoir is in animals, including rodents and dogs, and in some areas (e.g. India) in humans. The vector is the female sandfly.

There are three main clinical types of disease are:

1. Cutaneous (CL)
2. Mucocutaneous (MCL)
3. Visceral leishmaniasis (VL) or kala-azar.

Parasite and life cycle

About 30 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 or by PCR.

In animals and humans, *Leishmania* lives in macrophages in the reticulo-endothelial system in the form of amastigotes (*Leishman-Donovan* bodies). When taken up by the biting sandfly the parasite 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 her blood meal at night by feeding on animals, and also on humans if they are living or working in the vicinity.

Epidemiology

The Old World comprises Africa, Asia and Europe (collectively known as Afro-Eurasia), plus the surrounding islands. The term is used in contrast to the 'New World' (i.e. the Americas and sometimes Oceania).

Old world CL and VL are found in the Mediterranean basin, the Middle East, Sudan, South Sudan, Ethiopia, Kenya, Afghanistan, the Indian subcontinent, and southern regions of the former Soviet Union, and China. Where HIV infection and VL coexist, there are major problems in the treatment of VL. Drug resistance in VL is a serious concern in India and Sudan. Bihar State has 90% of VL in India and 45% of world cases.

In the New World, CL and MCL are the main forms of infection. VL occurs mainly in North-East Brazil.

Currently, leishmaniasis occurs in four continents and is considered to be endemic in 88 countries, 72 of which are resource-limited:

- 90% of all visceral leishmaniasis cases occur in Bangladesh, Brazil, India, Nepal and South Sudan
- 90% of mucocutaneous leishmaniasis cases occur in Bolivia, Brazil and Peru
- 90% of cutaneous leishmaniasis cases occur in Afghanistan, Brazil, Iran, Peru, Saudi Arabia and Syria.

Leishmaniasis is a disease of poverty associated with malnutrition, displacement, poor housing and migration of non-immune people to endemic areas. It is linked with deforestation and urbanisation.

Immunology

- A strong cell-mediated immune (CMI) response is required for control of 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 in VL is usually due to a secondary infection (e.g. respiratory tract or gut infection).

Cutaneous and mucocutaneous leishmaniasis

Cutaneous leishmaniasis

The species responsible are *L. tropica*, *L. major*, *L. aethiopica* in the Old World, and *L. mexicana* and *L. amazonensis* in the New World.

Single or multiple nodules develop on exposed areas, especially the face or extremities, and usually ulcerate. Lesions may be itchy or painful, and secondary bacterial infection may occur. Most heal spontaneously within months to a year or so, leaving scars.

Sporotrichoid-like nodular lymphangitis may occur in which there is thickening of the lymphatic channels draining the primary lesion with nodules at intervals along the path. Regional adenopathy may occur. Koebner phenomena and 'seeding' at sites of skin trauma, also may occur.

Mucocutaneous leishmaniasis

Most cases are caused by the *Viannia* subgenus, particularly *L. (V.) braziliensis*, *L. (V.) panamensis* and *L. (V.) guyanensis*.

The onset is usually a few years after the original cutaneous lesion has healed. Amastigotes spread from the skin to the naso-oropharyngeal mucosa. Initially there may be symptoms of chronic nasal congestion. Destructive lesions with chronic ulceration follow and secondary bacterial infections occur. Gradually, the nasal septum, other nasal cartilaginous structures and palate may be destroyed. Rarely, destructive lesions may occur in the urogenital region. In endemic areas, the risk of mucosal disease following a primary cutaneous lesion is around 1- 10% within 1-5 years, although there are reports of incidence rates of up to 25%. Recently, the presence of endosymbiotic *Leishmania* RNA virus (LRV) has been identified as an important virulence factor in *Viannia* sub-genus species in the New World (LRV-1) and in *L. aethiopica* in the Old World (LRV-2). Infections associated with LRV are likely to be more severe and more difficult to treat.

Diagnosis of CL and MCL

Diagnosis is usually made by biopsy of the edge of the ulcer or other lesion. The specimen is divided for:

- an impression smear (touch preparation) on a microscope slide that is then fixed with methanol and stained with Giemsa;
- histopathology (less sensitive than impression smear);
- polymerase chain reaction (PCR). PCR is particularly useful as a relatively rapid way of distinguishing *Viannia* from non-*Viannia* sub-genus infections.

Other techniques that are sometimes used include needle aspirates and dermal scrapings.

- Serology is generally unhelpful but is more likely to be positive in MCL than in CL. Culture, isoenzyme and DNA sequencing techniques are available only in specialist centres. A new microcapillary culture technique using a monophasic medium is also used in some centres. An interferon-gamma release assay has been developed for epidemiological studies. If available, detection of LRV may be useful to guide management and prognosis.

Management

Most CL lesions are self-limiting.

Before starting treatment, the following issues should be considered:

- the number, size, evolution and persistence of lesions;
- the location of lesion(s) (e.g. on the face);
- whether the patient has, or is at risk of, MCL;
- whether the patient is immunocompromised; and
- other features (e.g. the presence of nodular lymphangitis).

Treatment of cutaneous leishmaniasis

Cosmetically unimportant lesions caused by non-destructive and non-metastasizing species usually heal spontaneously and therefore may not require active treatment.

Choosing the most appropriate treatment for Cutaneous Leishmaniasis

The key questions are 'who needs systemic treatment' and 'for how long'?

It is useful to classify clinical presentations as 'simple' or 'complex' based on the following criteria:

"Complex": > 2-3 lesions; > 40 mm diameter; lymphatic / lymph node spread; cosmetic problems; functional problems; failure to respond to treatment as a "simple" lesion.

Local, topical, and physical treatments

Various local, physical, and topical therapies are sometimes used, including:

- heat treatment or cryotherapy;
- photodynamic therapy;
- topical amphotericin B (*L. major*);
- intralesional antimony therapy; and
- paromomycin ointment.

Treatment with paromomycin may result initially in increased ulceration, so it is best avoided for lesions on the face.

Oral Treatment

The following oral agents can be used for treating relatively benign cosmetically unimportant lesions.

1. *Ketoconazole* — modest activity against *L. mexicana*, *L. (V.) panamensis* and possibly *L. major*.
2. *Itraconazole* — better tolerated than ketoconazole but may be less effective against the *Viannia* subgenus and *L. major*.
3. *Fluconazole* – variable effectiveness against *L. major*. High dose fluconazole has been reported to be effective against *L. [V.] braziliensis* in a limited number of patients in Brazil. Treatment with 5 mg/kg/day achieved a cure rate of 75% with a mean time to healing of 7.5 weeks, whereas 8 mg/kg/day achieved 100% cure within 4-5 weeks.
4. *Miltefosine* — now recommended as the treatment of choice for New World CL due to *Viannia* sub-genus species. Miltefosine has also been used successfully in the treatment of Old-World CL and in the treatment of CL in immunosuppressed patients. Dose: 2.5 mg/kg/day PO × 28 days (max dose: 150 mg/day) in two or three divided doses daily.

Parenteral treatment

Pentavalent antimony therapy (SbV) (i.v. or i.m.) is probably still the best option if optimal effectiveness is important.

Studies on *L. panamensis* in Colombia, *L. braziliensis* in Guatemala, and *L. tropica* in the USA, showed no significant difference in outcome when treatment duration with SbV 20 mg/kg/day was reduced from 20 days to 10 days.

Liposomal amphotericin B (LAMB) is sometimes used to treat CL. Although, less effective than conventional amphotericin B, it is usually preferred because it is less toxic. Efficacy appears to be good in treatment of *L. infantum* infections and poor in treatment of infections due to *Viannia* sub-genus species.

Short course pentamidine has been shown to be effective in Colombia where disease is predominantly caused by the *Viannia* subgenus.

MCL treatment is of greater importance. Adequate systemic treatment of cutaneous lesions is assumed (but not proven) to decrease the risk of mucosal disease. MCL is harder to treat than cutaneous lesions and becomes increasingly so as it progresses. Treatment with SbV 20mg/kg iv/im for 28 days achieves cure rates of about 75% for mild disease and 10–63% for more advanced disease.

Alternatives to SbV are miltefosine or amphotericin B. Concomitant corticosteroids are indicated if respiratory compromise develops. A combination of oral pentoxifylline (a TNF- α inhibitor) plus SbV for 30 days reduced the relapse rate and accelerated cure in comparison with SbV alone among patients with refractory MCL in Brazil.

Table 38.1 Treatment of CL

(Adapted from Showler AJ, Boggild AK. Cutaneous leishmaniasis in travellers: a focus on epidemiology and treatment in 2015. *Curr Infect Dis Rep.* 2015 Jul;17(7):489. doi: 10.1007/s11908-015-0489-2. PMID: 26031962.)

Treatment	Curative efficacy
Local therapy Cryotherapy + intra-lesional antimony 0.2–5 mL every 3–7 days \times 5–8 treatments, or until healed	OWCL: <i>L. major</i> , <i>L. tropica</i> : 89–100 %; <i>L. donovani</i> : 100 % NWCL: <i>L. (V.) braziliensis</i> (ILSb alone): 70–80 %
Topical 15 % paromomycin/12 % methylbenzethonium ointment, OR 15 % paromomycin/0.5 % gentamicin twice daily \times 10–20 days	OWCL: <i>L. major</i> : 41–82 % NWCL: <i>L. (V.) panamensis</i> : 79–90 %; <i>L. (V.) braziliensis</i> , <i>L. mexicana</i> : 91 %; <i>L. (Viannia) spp.</i> mixed: 79 %
Thermotherapy Single treatment 50 °C for 30–60 min	OWCL: <i>L. major</i> : 48–82 %; <i>L. tropica</i> : 54–98 % NWCL: <i>L. mexicana</i> : 90–95 %; <i>L. (V.) panamensis</i> : 58 %; <i>L. (V.) braziliensis</i> : 58–64 %
Systemic therapy Pentavalent antimonials (sodium stibogluconate, meglumine antimoniate) 20 mg Sb/kg/day IV/IM \times 10–20 days	OWCL: <i>L. major</i> : 54 %–81 (81 % with adjunctive pentoxifylline); <i>L. tropica</i> *: 41–53 %; <i>L. aethiopica</i> : 85 %. NWCL: Most species: 70–96 %; <i>L. (V.) guyanensis</i> : 55–90 %, region specific
Miltefosine 2.5 mg/kg/day PO \times 28 days (max dose: 150 mg/day) Formulation: 50 mg capsules 30–44 kg: 50 mg Twice daily, \geq 45 kg: 50 mg TID OR	OWCL: <i>L. major</i> : 81–88 % NWCL: <i>L. (V.) panamensis</i> : 60–94 %; <i>L. (V.) braziliensis</i> : 33–88 %; <i>L.</i>

Treatment	Curative efficacy
Liposomal amphotericin B 3 mg/kg IV daily ×5–7 doses (preferred) OR Amphotericin B deoxycholate: 0.5–1.0 mg/kg IV every other day ×20–30 days (alternative)	(V.) guyanensis: 71 %; L. mexicana: 60 % 80–90 % in non-randomized studies for both OWCL and NWCL
Fluconazole 200–600 mg PO daily ×4–6 weeks	OWCL: L. major: 400 mg 81 %; 200 mg 44–79 % NWCL L. (V.) braziliensis: 8 mg/kg/day: 100 %; 6.5 mg/kg/day: 93 %; 5 mg/kg/day: 75 %
Pentamidine 2–3 mg/kg IV/IM daily or every other day ×4–7 doses	NWCL: L. (V.) panamensis, L. (V.) guyanensis: >90 % ; lower efficacy 60 % reported in some recent studies L. (V.) braziliensis: 35–86 %

* Pentavalent antimonials 20 mg/kg per day for 15 days plus oral allopurinol 20 mg/kg for 30 days to treat LR caused by *L. tropica*.

Visceral leishmaniasis (kala azar)

Epidemics occur in situations of famine, complex emergencies and mass movements of populations. It has a high fatality rate if untreated. It is estimated that there are > 200,000 new cases every year globally, of which more than 60% occur in Northern India (Bihar).

Of the new cases reported to WHO, 95% occur in 10 countries: Brazil, China, Ethiopia, India, Iraq, Kenya, Nepal, Somalia, South Sudan, and Sudan. The species responsible are *L. donovani* and *L. infantum* in the Old World, and *L. chagasi* in the New World.

The major presenting features include the triad of prolonged fever, anaemia and moderate to marked splenomegaly. In the 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, toxæmia, and mild splenomegaly. Pancytopenia is the main laboratory finding.

Post-Kala Azar Dermal Leishmaniasis (PKDL) may occur within 6 months to 2 years following treatment of VL. Macules and papules initially appear around the mouth then gradually spread over the face and sometimes over the trunk and limbs. Hypopigmented macules may resemble vitiligo. Nodules may develop resembling lepromatous leprosy. The papules and nodules are usually packed with amastigotes and patients with this condition, which may persist for > 20 years, may act as an important reservoir of infection particularly in the Indian sub-continent, where active case detection and treatment is a key transmission control measure.

Diagnosis

- In children, the diagnosis is usually confirmed by demonstrating amastigotes on bone-marrow or splenic aspirate.
- Splenic aspirates have a higher sensitivity, and this procedure is safe in skilled hands so long as the platelet count is above 40×10^9 /litre and coagulation is normal. **Splenic aspirate should not be performed if the differential diagnosis under consideration includes portal hypertension, vascular anomalies, or hydatid cyst** (Section 37).
- If there is lymphadenopathy, diagnosis may be attempted by fine-needle aspiration or biopsy.

- Serological antibody tests such as ELISA have a high sensitivity. The fast agglutination-screening test (FAST) is a rapid (< 3 hour) test that detects antibody in serum or filter paper blood-spot samples. The rK39 test is a commercially available immunochromatographic strip that uses recombinant leishmanial antigen.
- Serology may be positive in asymptomatic individuals living in endemic regions and may remain positive for years following successful treatment.
- Serology is unreliable in immunocompromised patients and is positive in only about 50% of patients with *Leishmania* and HIV co-infection.
- In *Leishmania* and HIV coinfecting patients, amastigotes may be detectable in the peripheral blood buffy coat in 50%.
- Polymerase chain reaction (PCR): sensitivity is 82 – 100% for bone marrow or splenic aspirate and 72 – 100% for peripheral blood. Promising real time PCR assays are under development.
- Recombinase polymerase amplification (RPA) is emerging as an alternative to conventional PCR. A *Leishmania donovani* RPA assay has been developed as a field-based diagnostic test for use in low resource settings.
- Loop-mediated isothermal amplification. The LAMP: Loopamp *Leishmania* detection kit (Eiken Chemical, Japan) uses peripheral blood for diagnosis of VL and PKDL. It is rapid, simple and has a high sensitivity and specificity.
- Lateral flow immunochromatographic RDT. The leishmaniasis IgG1 RDT (Coris Bioconcept, Belgium) is a rapid diagnostic test that detects anti-*Leishmania* IgG1 as a potential marker of relapse post-treatment. Zero or low levels correlate with cure, whereas raised IgG1 levels are indicative of treatment failure or relapse.
- Urine ELISA *Leishmania* antigen detection ELISA (InBios International, Seattle, USA) and *Leishmania* antigen ELISA (visceral leishmaniasis ELISA) (Kalon Biologicals, UK). These tests detect urinary *Leishmania* antigens in patients with VL and demonstrate clearance after successful treatment. Studies in Asia and Africa have demonstrated a high sensitivity and specificity.

Differential diagnosis

- Differential diagnosis of hepatosplenomegaly, anaemia and pancytopenia includes a wide range of bacterial and viral infectious diseases, hyper-reactive malaria splenomegaly (tropical splenomegaly) syndrome, tuberculosis, portal hypertension, leukaemia, lymphoma, and various auto-immune and metabolic diseases.
- In acute-onset disease, malaria, disseminated tuberculosis, typhoid, brucellosis, African trypanosomiasis, relapsing fever, other acute febrile illnesses and leukaemia should be considered.
- HIV infection greatly increases the risk of visceral leishmaniasis, and thus co-infection is common in some regions.

TABLE 38.2 Clinical features of visceral leishmaniasis

Clinical features of visceral leishmaniasis
Incubation period: 2–4 months (weeks to two years)
Fever: intermittent at first
Anaemia: bone-marrow depression, hypersplenism
Splenomegaly: progressive enlargement
Hepatomegaly
Weight loss
Epistaxis: haemorrhage from other sites may occur in advanced disease

Clinical features of visceral leishmaniasis
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

TABLE 38.3 Clinical pathology of visceral leishmaniasis

Haemoglobin: low; normochromic, normocytic film
White blood cells: low, 2–3 × 10 ⁹ /litre Eosinophils low
Platelets: low, < 100 × 10 ⁹ /litre
Reticulocytes: low
Serum albumin: low
Serum globulin: elevated
Liver transaminases and serum bilirubin: normal

Management of visceral leishmaniasis

1. Consider HIV and other possible co-infections such as malaria, respiratory and gut infections, and tuberculosis.
2. Blood transfusion for anaemia is seldom required, as the child has usually adapted to the low haemoglobin level.
3. Give haematinics and vitamin supplements during nutritional rehabilitation and convalescence.

Choice of treatment will depend on availability, cost, feasibility, contraindications, potential side-effects and the possibility of resistance. Drug resistance is common in the Bihar state of India.

Combination treatment is increasingly recommended to mitigate against the development of resistance, shorten the duration (and possibly cost) of treatment, and reduce the likelihood of relapse.

Pentavalent antimonial drugs (SbV)

Sodium stibogluconate (Pentostam) and meglumine antimonate (Glucantime) are most used as first-line treatment in East Africa. The usual dose is 20mg SbV/kg/day by slow intravenous infusion (the manufacturers of Pentostam recommend a minimum of 5 minutes) or intramuscularly for 20–40 days, depending on the geographical region. Side-effects include arthralgia, nausea, abdominal pain and pancreatitis. Cardiotoxicity tends to occur with high-dose regimens, particularly with prolonged use, and includes ST segment inversion, prolongation of the QTc interval and fatal arrhythmias. Toxicity, particularly pancreatitis, is increased in HIV-positive patients. Furthermore, SbV have been shown to stimulate HIV-1 replication *in vitro*. Mortality during treatment is four times higher with SbV than with miltefosine.

In East Africa a combination of sodium stibogluconate plus paromomycin for 17 days is now recommended in preference to the standard 30-day course of sodium stibogluconate (see

below).

Aminosidine (paromomycin)

Aminosidine (paromomycin) at doses in the range 15–20mg/kg/day i.m. may be used alone for 21 days or combined with SbV or pentamidine, allowing a shorter duration of treatment.

Combination treatment of SbV 20 mg/kg (im / iv infusion) with paromomycin 15 mg/kg/day im for 17 days is now recommended as first-line treatment for VL in East Africa.

Amphotericin B

Conventional amphotericin B (AmB) is sometimes used in regions with high levels of resistance to SbV, such as Bihar, India. It is usually administered by slow intravenous infusion in 5% dextrose over 4–6hours, commencing at 0.1mg/kg/day and gradually increasing to 1mg/kg/day until a *total* dose of 20mg/kg has been given. Side-effects include anaphylaxis, fever, chills, bone pain and thrombophlebitis. Hypokalaemia, renal impairment, and anaemia may also occur.

Liposomal amphotericin B (LAMB)

LAMB is currently regarded as first-line treatment in Europe and the Americas. Although comparatively expensive, preferential pricing has now made LAMB available for first line treatment of VL in LMICs.

LAMB is the least harmful option for treatment of VL in pregnancy.

The standard regimen for immunocompetent patients is an infusion of 3-5mg/kg/day for 6 to 10 days up to a total dose of 30 mg/kg. However, a recent study in India showed that administration of a single infusion (5mg/kg) or five daily infusions of 1mg/kg cured 92% of patients. A single dose of LAMB plus a one-week course of miltefosine has been shown to have a 98% cure rate at six-month follow-up in India.

A single 10 mg/kg infusion of LAMB has been used successfully to treat individuals with VL in the case detection and management strategy for kala-azar elimination Asia.

Miltefosine

Originally developed as an oral antineoplastic agent, miltefosine is the first highly effective oral treatment for VL. The recommended dose is 2.5mg/kg in divided doses daily for 28 days (maximum dose recommended is 150 mg; usual dose recommended in adults is 50 mg twice daily if weight 30 – 45 kg; 50 mg three times daily if weight > 45 kg). Cure rates of around 95% have been achieved in India and Northern Ethiopia in HIV negative patients, falling to 78% in HIV co-infected patients. Relapse rates are also higher in HIV co-infected patients. Nevertheless, daily miltefosine may have a role in preventing relapses in HIV co-infected patients. Side-effects include gastrointestinal upset, but this is rarely severe.

However, miltefosine is abortifacient and teratogenic and may also reduce male fertility. Miltefosine has a long half-life (2–3 weeks) and a narrow therapeutic index, thus increasing the opportunity for the development of resistance. Combination treatment should reduce this risk.

Pentamidine

Pentamidine 4mg/kg deep IM. on alternate days for 5–25 weeks has been used as second-line treatment for VL. However, widespread resistance and toxicity (sudden hypotension following injection, acute hypoglycaemia, renal impairment, arrhythmias and irreversible insulin-dependent diabetes in more than 10% of patients treated) have curtailed use. Pentamidine may still have a role in preventing relapses in patients co-infected with HIV. Lower dose combination with allopurinol reduces toxicity.

Treatment of VL in HIV co-infected patients

East Africa and India (*L. donovani*)

Either:

LAMB 3–5 mg/kg infusion daily or intermittently for ten doses (days 1–5, 10, 17, 24, 31, and 38) up to a total dose of 40 mg/kg

Or

Combination treatment: LAMB (30 mg/kg total dose: intravenous infusion 5 mg/kg on days 1, 3, 5, 7, 9, and 11) plus oral miltefosine for 28 days.

Europe and South America (*L. infantum / chagasi*)

LAMB Infused at a dose of 3–5 mg/kg daily or intermittently for ten doses (days 1–5, 10, 17, 24, 31, and 38) up to a total dose of 40 mg/kg

The relapse rate is high in VL-HIV coinfecting individuals, even following commencement of effective ART. Therefore, secondary prophylaxis is recommended (e.g. LAMB 3 mg/kg every 21 days) at least until the CD4 count has reached and remained > 200 cells/microL for six months and there is no evidence of VL. However, relapses may occur even when the CD4 count is > 350 cells/microL. Thus, some practitioners recommend continuing secondary prophylaxis.

Follow-up and prognosis

1. Splenomegaly slowly regresses but may take a year or more to completely resolve.
2. Prolonged follow-up (at least 1 year) is necessary to detect relapse.

Prevention and control

Prevention is similar to that of malaria and includes insect repellents and the use of fine-mesh bed nets impregnated with permethrin. Control includes spraying of sandfly resting sites and human dwellings, destruction of animal reservoirs, case finding and treatment of cases.

References

[World Health Organization \(2010\) Control of Leishmaniasis](#) .

Showler AJ, Boggild AK. Cutaneous leishmaniasis in travellers: a focus on epidemiology and treatment in 2015. *Curr Infect Dis Rep.* 2015 Jul;17(7):489. doi: 10.1007/s11908-015-0489-2. PMID: 26031962

Section 39. Leprosy

Leprosy is caused by a Mycobacterium which favours cooled exposed skin and mucosa or incisor teeth. Impaired natural immunity largely determines the ineffectiveness of processes of elimination and is encouraged by poor nutrition and overcrowding. The affected skin with few bacteria (paucibacillary) is hypopigmented and has reduced sensation.

Multibacillary infected tissue presents with many different patterns and usually severely infected cutaneous nerves.

For two decades leprosy has been declared eliminated, but globally there are still about 200,000 cases per annum. Most of these are in India and Brazil and the prevalence is highest in children. In this small residue the trend for children to have multi-bacillary leprosy and with disability at the time of diagnosis is higher than before elimination.

Differential diagnosis

Vitiligo is totally de-pigmented, whiter than leprosy and usually symmetrical. There is no sensory loss. It is long lasting.

Pityriasis alba is very common, mild, dry eczema, usually symmetrical on both cheeks and the extensor surface of both limbs. It varies over days or weeks and responds to moisturising ointments or hydrocortisone.

Pityriasis versicolor is a common infection of the skin from *Malassezia Furfur* producing depigmentation and fine scaling especially of the upper trunk. The organism and the slight inflammation it causes, accounts for a dull red to brown discolouration of white skin. Pigmented skin loses pigment due to exfoliation. It responds to selenium sulphide shampoo, Whitfield's (benzoic acid and salicylic acid) ointment or ketoconazole, plus sun exposure for rapid re-pigmentation.

Post-inflammatory depigmentation is preceded by undisputed injury such as a burn, chickenpox, fungal infection or psoriasis. There may be loss of normal skin texture as in a scar.

Reactions

Reactions are immunological responses to Mycobacterium leprae or its antigen. There are two types:

- Erythema nodosum-like with multiple, tender, symmetrical red lumps anywhere in the skin due to immune complexes and accompanied by fever and malaise. It often responds to rest and non-steroidal anti-inflammatory drugs, but persistent reactions will need oral steroids. There is usually a history of prior diagnosis of leprosy.
- The other type of reaction is focused on a previous plaque or infected nerve. There is redness, swelling and tenderness. It is destructive of nerves. An early prescription of an initially high dose of prednisolone is necessary (1 mg/ kg/day). Complete withdrawal of steroids should only occur after several weeks if nerve destruction is to be avoided

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, 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 bacterium.

A daily regimen for 1 year of three drugs is necessary for more widespread multibacillary disease. Lepromatous leprosy is subject to reaction even after 1 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.

For children younger than 10 years the dose must be adjusted according to body weight.

- Rifampicin 10 mg/kg once a month
- Clofazimine 100 mg once a month,
- Dapsone 2 mg/kg daily

WHO-recommended treatment for paucibacillary leprosy in children (10–14 years):

- Once a month for six months rifampicin 450 mg supervised. No clofazimine
Plus once a day for six months: one tablet of dapsone 50 mg

Full course: six blister packs over 6 months.

WHO-recommended treatment for multibacillary leprosy in children (10-14 years)

- Once a month on day 1: rifampicin 450mg, clofazamine 150mg, dapsone 50mg.
- Day 2 to the end of the month clofazamine 50mg every other day
- Day 2 to the end of month Dapsone 50mg every day

WHO-recommended treatment for multibacillary leprosy in children (15 years/adult):

Once a month: On day 1, rifampicin 600 mg supervised

plus

Once a day: on day 2 and every day to end of month, clofazimine 50 mg, plus dapsone 100 mg daily.

Full course: 12 blister packs over 12 months.

Children may be more troubled by the haemolytic side effect of dapsone and are less tolerant to rifampicin.

New drug regimens include ofloxacin, minocycline and clarithromycin. Several experimental and clinical studies have demonstrated that these drugs either alone or in combination with other anti-leprosy 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 over 12 years).

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. Support and counselling are necessary for the patient along with education for family and community, or else the cured patient may still not be acceptable to either family or community.

Further reading

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[World Health Organization \(2019\) Global leprosy \(Hansen disease\) update](#) (accessed 04/03/2021)

[World Health Organization \(2018\) Guidelines for the diagnosis, treatment and prevention of leprosy.](#)
(accessed 04/03/2021)

Section 40. Lyme Disease

Introduction

This disease is caused by the bacterium *Borrelia*. *Borrelia Burgdorferi* is the main cause in North America, whereas *Borrelia Afzelii* and *Borrelia Garinii* cause most European cases. The prevalence of Lyme disease in sub-Saharan Africa is presently unknown, but cases have been reported. The abundance of hosts and tick vectors would support the presence of this infection in Africa where it is probably grossly under-diagnosed.

Transmission

Lyme disease is transmitted to humans from a natural reservoir among rodents by ticks that feed on both rodents and other animals, such as deer. Tick bites often go unnoticed because of the small size of the tick in its nymphal stage, as well as tick secretions that prevent the host from feeling any itch or pain from the bite. However, transmission is quite rare, with only about 1% of recognised tick bites resulting in Lyme disease. This may be because an infected tick must be attached for at least a day for transmission to occur.

Days to weeks following the tick bite, the spirochetes spread via the bloodstream to joints, heart, nervous system, and distant skin sites, where their presence gives rise to the variety of symptoms of disseminated disease.

If untreated, the bacteria may persist in the body for months or even years, despite the production of antibodies against *Borrelia* by the immune system.

Diagnosis

Lyme disease is diagnosed clinically based on symptoms, objective physical findings (such as erythema migrans (EM), facial palsy or arthritis) or a history of possible exposure to infected ticks, as well as serological blood tests. The EM rash is not always a bull's-eye (see below) (i.e., it can be red all the way across). When making a diagnosis of Lyme disease, healthcare providers should consider other diseases that may cause similar illness. Not all patients infected with Lyme disease will develop the characteristic bull's-eye rash, and many may not recall a tick bite but do not diagnose Lyme disease in people without symptoms even if they have had a tick bite.

Signs and symptoms

Many of the symptoms are not specific to Lyme disease. The incubation period from tick bite to the onset of symptoms is usually 1–2 weeks, but can be much shorter (days), or much longer (months).

Early localised infection

The classic sign of early local infection with Lyme disease is a circular, outwardly expanding rash called erythema chronicum migrans (also erythema migrans or EM), which usually occurs at the site of the tick bite 3–30 days after the bite, typically 1–4 weeks. The rash is red, and may be warm, but is generally painless, not itchy or hot. Classically, the innermost portion remains dark red and becomes thicker and firmer; the outer edge remains red; and the portion in between clears, giving the appearance of a bull's-eye. EM is thought to occur in about 80% of infected patients. Patients can also experience flu-like symptoms, such as headache, muscle soreness, fever and sweats, lymphadenopathy, neck pain or stiffness, fatigue and malaise. Lyme disease can progress to later stages even in patients who do not develop a rash.

Early disseminated infection

Within days to weeks after the onset of local infection, the *Borrelia* bacteria begin to spread through the bloodstream. EM may develop at sites across the body that bear no relation to the original tick bite. Other discrete symptoms include migrating pain in muscles, joints, and tendons, and heart palpitations and dizziness.

Various acute neurological problems appear in 10 - 15% of untreated patients. These include facial palsy, arthritis, and meningitis. Radiculoneuritis causes shooting pains that may interfere with sleep, as well as abnormal skin sensations. Mild encephalitis may lead to memory loss, sleep disturbances, or mood changes.

The disease may also have cardiac manifestations including cardiac arrhythmias.

Late disseminated infection

After several months, untreated or inadequately treated patients may go on to develop severe and chronic symptoms that affect many parts of the body, including the brain, nerves, eyes, joints and heart. Many disabling symptoms can occur.

Chronic encephalomyelitis, which may be progressive, can involve cognitive impairment, weakness in the legs, awkward gait, facial palsy, bladder problems, vertigo, and back pain.

In rare cases untreated Lyme disease may cause frank psychosis, which has been misdiagnosed as schizophrenia or bipolar disorder. Panic attacks and anxiety can occur; there may also be delusional behaviour, including somatoform delusions, sometimes accompanied by a depersonalisation or derealisation syndrome, where the patients begin to feel detached from themselves or from reality. Lyme arthritis usually affects the knees.

Treatment

In most cases, the infection and its symptoms are eliminated by antibiotics, especially if the illness is treated early. Delayed or inadequate treatment can lead to the more serious symptoms, which can be disabling and difficult to treat.

The antibiotics of choice for early infections are given orally:

- In children over 8 years:
 - Doxycycline, 5 mg/kg/day in two divided doses (maximum of 100 mg per dose) on day 1 followed by 2.5 mg/kg daily in 1 or 2 divided doses for 21 days or for severe infections 5 mg/kg daily for 21 days
- In younger children (less than 8 years):
 - Amoxicillin 30 mg/ kg 3 times per day for 21 days
 - Second alternative: azithromycin 10 mg/kg daily for 17 days
 - Doxycycline should not be given in children who are pregnant, instead use amoxicillin 250–500 mg three times daily for pregnant girls.
 - If early infection is severe, ceftriaxone 50 mg/kg IV/IM once daily can be given at any age.

Late-diagnosed central nervous system or cardiac Lyme disease is treated with oral as above or intravenous antibiotics for a minimum of 4 weeks, frequently ceftriaxone 50–75 mg/kg once a day IV.

Reference

National Institute for Health and Care Excellence (2018) Lyme disease <https://pubmed.ncbi.nlm.nih.gov/31194312/> Accessed 9th April 2021

Section 41. Mumps

Introduction

Mumps is a systemic disease characterised by swelling of one or more of the salivary glands, usually the parotid glands. It is caused by a virus of the paramyxovirus family (which also includes measles and parainfluenza). The virus is spread by airborne droplets through the respiratory tract, mouth and possibly the conjunctivae and urine, and is present in saliva, CSF, blood, and urine.

Other viruses and bacteria (cytomegalovirus, parainfluenza virus types 1 and 3, influenza A virus, coxsackieviruses and other enteroviruses, human immunodeficiency virus (HIV), *Staphylococcus aureus* and non-tuberculous *Mycobacterium*) may also cause parotitis.

Clinical presentation

- The 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 and usually neither serious nor recognised clinically. 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

- Cervical adenitis,
- pyogenic parotitis,
- Recurrent parotitis,
- Tumours of the parotid
- Tooth infections.
- Mumps orchitis can mimic hernias, tumours, haematomas, epididymo-orchitis and testicular torsion.

Complications

- Oophoritis,
 - Mastitis
 - Pancreatitis
 - Nephritis
 - Myocarditis
 - Thyroiditis
 - Labyrinthine disturbance
 - Painful swelling of the lacrimal glands
 - Optic neuritis
 - Uveokeratitis
 - Rapid loss of vision
 - Arthritis
 - Jaundice
 - Pneumonia
 - Thrombocytopenia
- Transient or permanent unilateral nerve deafness has been reported.
 - Infection during pregnancy very rarely causes disease of the fetus (e.g. aqueductal stenosis, hydrocephalus).

Management

Symptomatic treatment includes analgesics, fluids, and scrotal support for orchitis. Antibiotics are usually not warranted for the uncomplicated disease, but each complication should be treated on its own merits with antibiotics (in the case of pneumonia or wherever a secondary bacterial infection is suspected), or with appropriate local treatment and monitoring. The value of corticosteroids for orchitis is not established.

Prevention

Measles, mumps, rubella (MMR) immunisation is routine in well-resourced countries and has reduced mumps by over 90%. The recommended two-dose vaccine schedule has an effectiveness of approximately 90% (range 88–95%).

<https://www.who.int/publications/i/item/surveillance-standards-for-vaccine-preventable-diseases-2nd-edition> (accessed 04/04/2021)

Section 42. Onchocerciasis

Introduction

Onchocerciasis is caused by the filarial worm *Onchocerca volvulus* and is an important cause of blindness and skin disease in tropical Africa and Yemen. Although previously endemic in parts of Latin America, only one focus now remains in the Yanomani area of South America, spanning parts of Venezuela and Brazil. It is transmitted by the bite of blackflies (*Simulium* species).

Epidemiology

This infection mainly affects people living or working near fast-flowing rivers (*Simulium* breeding sites) but may be more widely distributed by flies carried on winds.

Pathology

- Adult worms evade the host immune response and cause few symptoms.
- The main problems are the result of immunological reactions to dying and dead microfilariae and their endosymbiotic bacteria (*Wolbachia*), which release bacterial mediators that trigger the innate immune system.
- In addition, activated eosinophils release cellular proteins that cause connective tissue damage.
- Onchocerciasis may increase the risk of HIV-1 seroconversion.
- Treatment of onchocerciasis is associated with reduced HIV-1 viral replication.
- Onchodermatitis is more severe in HIV-positive patients.

Clinical features

- The incubation period is usually 15 – 18 months.
- Infected patients may be asymptomatic.
- Palpable firm painless subcutaneous nodules (intertwined adult worms), several centimetres in diameter, may be most obvious over bony prominences.

Skin disease

A variety of different skin manifestations are seen, usually with a significant degree of overlap:

- Acute papular onchodermatitis (APOD):
 - An intensely itchy papular rash, sometimes with local oedema.
- Chronic papular onchodermatitis (CPOD):
 - Larger pruritic (itchy) hyperpigmented papules.
- Lichenified onchodermatitis (LOD):
 - Discrete or confluent pruritic hyperpigmented papulonodular plaques, often with lymphadenopathy.

Severe itching may give rise to excoriation and secondary bacterial infection. Healing is associated with progressive hyperpigmentation, blackening, and thickening of the skin. Unrelenting itching may result in chronic sleep disturbance, poor concentration, and depression.

Heavy infections in childhood can impair growth. After some years, skin atrophy and depigmentation give a wrinkled prematurely aged appearance (presbydermia). Patchy depigmentation, especially of the legs, results in a 'leopard-skin' appearance.

Inguinal or femoral lymphadenopathy may give rise to the so-called 'hanging groin' appearance.

Eye disease

Early symptoms include itching, redness and excess lacrimation. Late disease leads to varying degrees of loss of vision, and eventually to blindness.

Anterior eye disease

- Punctate keratitis due to death of microfilariae in the cornea may appear as a reversible 'snow-flake' opacity.
- Pannus forms as blood vessels invade the cornea from the sides and below. The pannus may cover the pupil (sclerosing keratitis) and cause blindness.
- Iritis leads to a loss of the pigment frill and to synechiae that cause a deformed, often pear-shaped pupil. Secondary cataracts occasionally result.

Posterior eye disease

- Chorioretinitis with pigmentary changes.
- Optic atrophy.
- 'Tunnel vision' and various other forms of visual loss may become evident in young adults.

Epilepsy

Recently onchocerciasis has been associated with epilepsy and neuroendocrine disorders. Presentations include tonic-clonic seizures and an unusual 'nodding syndrome' due to atonic neck seizures. Hypopituitarism has also been described. The incidence of these disorders falls with successful onchocerciasis control.

Diagnosis

1. Skin snips in saline examined under the microscope for microfilariae.
2. Skin-snip microscopy is less sensitive than newer biochemical methods, including skin-snip PCR, ELISAs, EIAs, and antigen detection.
3. Rapid diagnostic tests. A new luciferase immunoprecipitation systems (LIPS) assay has 100% sensitivity and specificity for *O. volvulus* using a rapid 15-minute format (QLIPS).
4. Biochemical methods: Recent advances include a serum antibody test card using recombinant antigen to detect *O. volvulus*-specific IgG4 in finger-prick whole-blood specimens, a triple-antigen indirect ELISA rapid-format card test, and a highly sensitive and specific urine antigen dipstick test.
5. A highly sensitive and specific urine antigen dipstick test has recently been developed.
6. Slit-lamp examination for microfilariae in the anterior chamber of the eye.
7. Surgery:
 - a. Subcutaneous nodules can be removed to demonstrate adult worms or aspirated with a needle to look for microfilariae.
8. DEC patch test: Diethylcarbamazine (DEC), although no longer recommended for the treatment of onchocerciasis because of the risk of provoking a Mazzotti reaction (see below), may be used in the following manner in patients with repeatedly negative skin snips, where other diagnostic techniques are unavailable.
 - a. A 1-cm square of filter paper soaked in a solution of DEC is applied to the skin of the patient.
 - b. If positive, this will provoke intense localised itching and inflammation at the site of application.
 - c. DEC patch testing of children aged 3 - 5 years is advocated as an effective low-cost method for monitoring the endemicity and transmission of onchocerciasis in Africa.
9. Warning: A DEC patch test may precipitate a full-blown Mazzotti reaction.
 - a. This consists of microfilaria death resulting in an intensely itchy papular rash, may be accompanied by fever, limb oedema, hypotension and worsening of eye damage, and may be fatal.
 - b. It is commonly associated with the use of oral DEC and is rarely caused by ivermectin.

Treatment

- i. Ivermectin kills microfilariae by immobilising them so that they are carried away via the lymphatics.
- ii. Warning: Ivermectin may precipitate meningoencephalitis or renal failure in patients who have Loa loa with a high microfilaraemia (> 2500 microfilariae/mL).
- iii. It is therefore important to exclude Loa loa if there is any possibility of co-infection, before giving ivermectin.
- iv. Doxycycline kills the endosymbiotic Wolbachia, resulting in the slow, less pathogenic death of the microfilariae.
 - a. The drug also blocks worm embryogenesis and has a significant macrofilaricidal effect.
 - b. Contraindications to doxycycline include age less than 9 years, pregnancy and breastfeeding.

Treatment of individual patients

- i. Provided that the patient does not have a high Loa loa microfilaraemia, and ivermectin (or doxycycline) is not otherwise contraindicated, the following options are available:
- ii. If the patient will continue to live in an endemic area or is less than 9 years old and weighs more than 15 kg, give ivermectin 150 micrograms/kg every 3–6 months.
- iii. If interruption of worm embryogenesis and cessation of microfilariae production is desired, give doxycycline 200 mg/day for 4 weeks, or 100 mg/day for 6 weeks, followed by one dose of ivermectin after 4 - 6 months (children aged > 9 years).
- iv. If a strong macrofilaricidal effect is desired, give doxycycline 200 mg/day for 6 weeks, followed by one dose of ivermectin after 4 - 6 months.

Patients with onchocerciasis who do have a high Loa loa microfilaraemia may be treated with doxycycline 200 mg daily for 6 weeks, unless contraindicated. If they are under 9 years of age, in which case doxycycline is contraindicated, Loa loa microfilaraemia must be reduced by treatment with albendazole prior to treatment with ivermectin.

Surgical removal of head nodules (nodulectomy) was advised in the past in an attempt to reduce the likelihood of eye disease. There is no guarantee that this will eliminate the risk of eye disease, because not all nodules are evident, and the remaining nodules continue to produce microfilariae. Improved drug treatment has reduced the justification for nodulectomy.

Control

- There has been rapid progress in the past 30 years, largely due to successful international public–private partnerships, sustained funding for regional programmes, and technical advances.
- Initial efforts in vector control using the organophosphate larvicide Temephos proved inadequate.
- A major breakthrough came with Merck's donation of Ivermectin. Thereafter larviciding was abandoned in favour of regular mass drug treatment.

The African Programme for Onchocerciasis Control (APOC)

(APOC) is a Community-Directed Treatment with Ivermectin (CDTI) programme that aims to treat over 90 million people annually in 19 countries, protecting an at-risk population of 115 million, and should prevent over 40 000 cases of blindness every year. In 2009 the programme objective switched from 'control' to 'elimination' of onchocerciasis.

The Expanded Special Project for Elimination of Neglected Tropical Diseases (ESPEN) has incorporated onchocerciasis into control / elimination programmes for other neglected tropical diseases, including lymphatic filariasis, soil transmitted helminths, schistosomiasis and trachoma.

High-risk foci of Loa loa are currently excluded from community ivermectin programmes.

The Onchocerciasis Elimination Programme for the Americas (OEPA) adopts a similar approach to APOC, except that ivermectin is administered twice a year until transmission has been interrupted. By the end of 2017, transmission of the infection, judged by surveys following WHO guidelines, had been interrupted or eliminated in 11 of the 13 endemic foci in the Americas. The main remaining foci are in the Yanomani area, where MDA has been scaled up to 4 times a year. The aim is to eliminate onchocerciasis in the Americas by 2022.

Recent Development

Moxidectin, a more potent relative of ivermectin, is currently undergoing clinical trials. Moxidectin is microfilaricidal and also sterilizes or kills the adult worms. Moxidectin has a half-life 20-43 days, compared to < 1 day for ivermectin. Programmatically, moxidectin has two main advantages over ivermectin:

1. annual treatment could be as effective as biannual treatment with Ivermectin;
2. the effect on transmission will not depend so much on the time of distribution relative to peak transmission season.

Therefore, moxidectin could interrupt transmission within about 6 annual treatment rounds. However, given the similarity in mode of action, it is unlikely to be of use in the event of ivermectin resistance.

Further reading

[NTD Modelling Consortium Onchocerciasis Group \(2019\) The World Health Organization 2030 goals for onchocerciasis: Insights and perspectives from mathematical modelling: NTD Modelling Consortium Onchocerciasis Group. *Gates Open Res.* 2019;3:1545.](#)

Section 43. Poliomyelitis

Introduction

Poliomyelitis is caused by polioviruses type 1, 2 and 3, which are ingested and then multiply in the tonsils and Peyer's patches of the gut. In most cases, infection is contained at this point and the child is asymptomatic.

Due to both vertical and mass vaccination campaigns, the number of reported cases has fallen by 99% since 1988. Wild poliovirus is now only found in Afghanistan and Pakistan.

Diagnosis

CSF initially shows neutrophil predominance, but after 5 - 7 days is mainly lymphocytic. CSF protein levels are normal or slightly elevated. CSF glucose levels are normal. Virus can be isolated from throat and stool for up to 3 months after onset. The differential diagnosis includes other causes of acute flaccid paralysis (see Section 25).

Severity

Minor illness

This is associated with viraemia and non-specific symptoms, such as nausea, vomiting, abdominal pain and sore throat.

Major illness

Non-paralytic poliomyelitis

- This occurs in a minority of symptomatic children.
- Incubation period is 10 - 14 days and symptoms include:
 - Fever
 - Headache
 - Two to five days later; signs of meningeal irritation with severe pain and stiffness of neck, back and limbs.

Paralytic poliomyelitis

- Paralysis occurs within the first 2 days of major illness.
- It can affect any muscles, but particularly large ones and those of the lower limbs.
- Asymmetrical paralysis, flaccid muscles and absent tendon reflexes are characteristic. There is intact sensation. Paralysis is maximal within 3 - 5 days of onset, and rarely extends once the temperature has settled.
- In bulbar form, the involvement of cranial nerve nuclei and vital centres in the brainstem results in paralysis of the facial, pharyngeal, laryngeal and tongue muscles, causing swallowing difficulties, aspiration, and respiratory failure.
- Hypertension may occur, as well as transient bladder paralysis.

Prognosis

- This depends on the extent of paralysis and the quality of care during the acute phase.
- Early identification of and intervention for respiratory and bulbar paralysis will reduce mortality to 5 - 10%.
- With appropriate physiotherapy, improvement in the 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 or teeth during the incubation period increases the risk of bulbar paralysis.

Management

Acute phase

1. Absolute bed rest is mandatory. Avoid intramuscular injections and exercise.

2. Analgesics should be given for severe pain.
3. Keep paralysed muscles in a neutral position to prevent contractures.
4. Gentle passive exercises and warm compresses should be used to help to relieve pain.
5. Active exercises are introduced a few days after the temperature has settled.
6. Respiratory paralysis requires ventilatory support (if available) (see Sections 13, 14) and 91 in Handbook 1.
7. Bulbar paralysis requires nasogastric tube feeding and, to protect the airway, may require tracheostomy.

Convalescent phase

1. Aim to improve motor function, prevent deformities, and generally reintegrate the child into society.
2. Encourage active participation by the parents in the rehabilitation process.
3. The educational and emotional needs of the child must not be neglected.
4. The services of an orthopaedic surgeon and an orthoptist may be required.
5. See Sections 5 and 58 in this handbook) for further information on the long-term care of children with a disability.

Prevention

Immunisation.

Reference

[World Health Organization \(2018\) Surveillance Standards for Vaccine-Preventable Diseases, Second Edition](#) (accessed 04/03/2021)

https://www.who.int/immunization/monitoring_surveillance/burden/vpd/WHO_Surveillance_VaccinePreventable_18_Polio_R2.pdf?ua=1

Accessed 10th April 2021

Section 44. Rabies management and prevention after animal bites

Rabies encephalitis

- Transmission of rabies to humans is via mammal bites and saliva-contaminated scratches with viral entry through mucosae and broken skin. Domestic dogs are the major reservoir and vector of human rabies throughout most of Africa, Asia, and parts of Latin America, but cats, foxes, bats, jackals, wolves, mongooses, and domestic mammals may also transmit the infection. Infected mothers may deliver healthy babies.
- The first symptom is often itching near the healed bite site. Then symptoms of either furious or paralytic rabies develop.
- Furious rabies is characterised by agitation, hyperexcitability and hydrophobia, which is due to spasms of the inspiratory muscles, accompanied by an inexplicable feeling of terror.
- Spasms occur on attempting to drink water or from a draught of air.
- Flaccid paralysis without hydrophobia occurs in some patients, but this paralytic form is rarely recognised. It is likely to be misdiagnosed as another encephalitis or cerebral malaria.
- Furious rabies encephalomyelitis is usually fatal within a few days and paralytic rabies within 30 days. A few documented partially vaccinated survivors in Asia have devastating neurological sequelae.
- No antiviral or other specific treatment has proved successful.

Management

1. Treat established rabies with active compassionate palliative care (see Section 7).
2. Sedatives (e.g., IV or intra-rectal diazepam, midazolam) and strong analgesics (e.g., morphine) may be given to control distressing symptoms and relieve pain and terror.
3. An IV infusion assuages the feeling of thirst.
4. Relatives and staff should wear gloves when handling the child, their vomitus, or their saliva.
5. Close attendants are at risk of exposure to the virus, but there is no documented case of transmission of rabies to a carer.
6. However, anti-rabies vaccine if available, it should be offered to them, but rabies immunoglobulin (RIG) is not needed.

Rabies prophylaxis

Estimating the risk of exposure to rabies

Intact skin is a barrier against the virus.

- a. Is there a bite wound with broken skin?
- b. Have mucous membranes or an existing skin lesion been contaminated by virus in the animal's saliva?
- c. How did the animal behave?
- d. 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.
- e. Is rabies known to occur in the biting animal species?

Regularly vaccinated animals are unlikely to be rabid, but vaccinated dogs or cats can transmit the infection.

Try to have the animal's brain examined for rabies. If this is not possible, the animal may be kept under safe observation, but post-exposure treatment must not be delayed. If the cat or dog is still healthy after 10 days, vaccine treatment of the patient can be stopped.

Post-exposure treatment

- Table 44.1 lists the criteria for initiating treatment.

- The aim is to chemically kill or neutralise the rabies virus at the wound site before it can enter a nerve ending and travel to the brain.
- All three parts of post-exposure therapy are always urgent.
- *Wound care* is important for all bites, irrespective of the rabies risk.
- *Rabies vaccine* induces neutralising antibody (active immunisation). Even in previously vaccinated people, booster vaccination is essential.
- *Rabies immunoglobulin* (RIG) (passive immunization) provides immediate antibody locally at the wound site, until the vaccine-induced antibody appears.

Wound care

- a. Scrub and flush the lesion repeatedly and energetically with soap or detergent and water.
- b. Remove any foreign material.
- c. Local analgesia may be necessary.
- d. Apply povidone iodine (or 70% ethyl alcohol, but this is painful).
- e. Do not suture the wound, or at least delay suturing.
- f. Give tetanus immunisation if appropriate.
- g. Only use oral antibiotic if there is evidence of bacterial wound infection.

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 prequalified by WHO include the following:

- Purified chick embryo cell vaccine (PCEC) (Rabipur®, RabAvert®) (1.0 mL/vial).
- Purified vero cell vaccine (PVRV) (Verorab®) (0.5 mL/ ampoule).

These vaccines are interchangeable, so a different vaccine can be used during a course of treatment. The side effects are mild local or non-specific generalised symptoms. Transient maculopapular or urticarial rashes are occasionally seen.

Other vaccines

Tissue culture rabies vaccines not listed above should be used according to the manufacturer's instructions.

Selected PEP post-exposure regimens

Regimens for patients who have NOT previously completed a course of rabies vaccine:

- *Four vial IM methods:*

Give whole vial IM (deltoid) on days 0, 3, 7 and 21-28 (4 visits)

OR

Give 2 vials IM on day 0 and one on days 7 and 21 (3 visits)

OR

- *Economical three visit ID method:* (0.1 or 0.2mL/site*)

Day 0: divide whole vial* between four ID sites (deltoids and suprascapular or thighs)

Day 7: use half vial ID divided between two sites

Day 28: give ID dose (0.1 or 0.2mL*) at 1 site

Post-exposure vaccine booster for those who have had at least 2 doses of rabies vaccine previously (RIG not needed)

- *IM method:* Give whole vial IM on days 0 and 3

OR

- *Single day ID method*

Day 0: divide whole vial* between four ID sites (deltoids and suprascapular or thighs) (0.1 or 0.2mL per site*)

***The intradermal dose is 0.1 mL** per site for vaccines containing 0.5 mL/ampoule (e.g., Verorab®), and 0.2 mL per site for vaccines containing 1 mL/ampoule (e.g. Rabipur®).

Practical points

- i. Intradermal injections should raise a papule (See Handbook 1 Section 89 Figure 89.5).
- ii. If ampoules are shared, use a new sterile needle and syringe for each patient with strict aseptic precautions.
- iii. If there is difficulty in injecting 0.2 mL intradermally, withdraw the needle and inject the remainder at an adjacent site.
- iv. Do not waste vaccine. Vials shared between patients must be stored at 5°C and used on the same day.
- v. If few patients are treated, on day 0 consider asking the patient to bring relatives and friends for pre-exposure vaccine at the next visit.
- vi. The timing of the final dose can be varied for economy.

Rabies immunoglobulin (RIG)

If not previously vaccinated, passive immunisation with RIG is needed to accompany vaccine following major exposure to a suspect rabid animal (see Table 44.1).

- Dosage:
 - Equine RIG (40 IU/kg) or human RIG (20 IU/kg)
- Infiltrate the whole dose or as much as possible into and around the wounds. If the volume is too small to inject all wounds, dilute the RIG with a little 0.9% saline.
- If it is not anatomically possible to use the whole dose locally (e.g., on a finger), inject any remainder IM, eg. thigh but not the gluteal region. If supplies are limited consider injecting wounds only, *except* for those with severe exposure: with multiple bites, deep wounds, or bites to the head, neck, or hands; patients with severe immunodeficiency; if the biting animal has confirmed or probable rabies; or where bites, scratches or mucous membrane exposure is caused by a bat.
- If RIG is not available immediately, it should be given up to 7 days after the first dose of vaccine. After that it is no longer needed.
- In very rare cases anaphylaxis may occur. Skin tests do not predict reactions and should not be used.

Anaphylaxis treatment (see also Section 36 Handbook 1)

Adrenaline (epinephrine) intramuscular treatment is essential.

Dosage:

- Age < 6 years: 150 micrograms or 0.15 mL of 1:1000 (1 mg/mL).
- Age 6–12 years: 300 micrograms or 0.3 mL of 1:1000.
- Age > 12 years: 500 micrograms or 0.5 mL of 1:1000.
- The dose can be repeated at 5-minute intervals if necessary.

In addition, if available give the following:

Chlorpheniramine maleate IM or by slow IV injection

Dosage:

- Age 6 months to 6 years: 2.5 mg.
- Age 6–12 years: 5 mg.
- Age > 12 years: 10 mg.

Hydrocortisone sodium succinate by slow IV injection or IM

Dosage:

- Age 1–5 years: 50 mg.
- Age 6–12 years: 100 mg.
- Age > 12 years: 200 mg.

Pre-exposure treatment

Pre-exposure vaccination is the best means of rabies prophylaxis. No one who has had pre-exposure treatment and a post-exposure booster injection is known to have died of rabies.

Indications for pre-exposure rabies prophylaxis

- People working with dogs, bats or other wild mammals should be immunised.
- Anyone in an area where dog rabies is enzootic is at risk of infection, especially children. Ideally, rabies should be included as part of the routine Expanded Programme on Immunisation (EPI).
- Pre-exposure vaccine should be given whenever it is affordable to residents of dog rabies areas and should be strongly encouraged if RIG may not be available locally.

Pre-exposure rabies vaccine regimens

IM three visits: One vial IM days 0, 7 and 21-28

OR

ID three visits: 0.1 ml any vaccine ID on days 0, 7 and once during 21-28

Economical if aseptic vial sharing is possible

OR

ID two visits: 2 site ID 0.1 ml/site on days 0, 7 (deltoids)

- Having had one or two doses is still an advantage if the individual is exposed to rabies in the future, especially if RIG may not be available.
- Patients on chloroquine, steroids or other immunosuppressive drugs should have IM not intradermal injections for pre-exposure treatment.
- People who have been vaccinated should keep a record of their immunisations.
- Routine booster doses are only recommended for people at high occupational risk of exposure.
- If contact with a rabid animal occurs, post-exposure booster vaccine treatment is still urgently required.

TABLE 44.1 Recommended criteria for post-exposure treatment

Type of exposure	Criteria	Action
No exposure*	Touching animals, or being licked on intact skin	No treatment
Minor exposure, WHO Category II	Nibbling (tooth contact) with uncovered skin or minor scratches or abrasions without bleeding	Start vaccine immediately

Type of exposure	Criteria	Action
Major exposure, WHO Category III	Single or multiple bites or scratches that break the skin, or licking on broken skin, or licking or saliva on mucosae, or physical contact with bats	Immediate vaccine and, if not previously vaccinated, give RIG.
Severe exposure, WHO Category IV	Bites on the head, neck or hands, or multiple bites	Immediate vaccine with RIG

For all cases:

- Stop treatment if the dog or cat remains healthy for 10 days.
- Stop treatment if the animal's brain is shown to be negative for rabies by appropriate investigation.

**The confusing term 'WHO Category I' should be avoided, as misunderstanding leads to unnecessary treatment.*

Summary

- The only treatment for rabies encephalitis is palliative care.
- Exposure to rabies can be prevented by education about the dangers of animal contact, the need for vaccination of pets.
- Post-exposure prophylaxis is urgent.
- After possible contact with the virus, immediately clean wounds thoroughly with soap and water, then attend a clinic for vaccine.
- If rabies vaccine is unaffordable or in short supply use robust economical ID regimens that are suitable for use globally.
- Pre-exposure vaccination should be encouraged, especially for children and if RIG is not available locally.

Further reading

World Health Organization (2018) Expert Consultation on Rabies. 3rd Report. Technical Report Series 1012 <http://apps.who.int/iris/bitstream/handle/10665/272364/9789241210218-eng.pdf?ua=1> (accessed March 2021)

https://assets.publishing.service.gov.uk/government/uploads/system/uploads/attachment_data/file/723607/GreenBook_chapter_27_rabies.pdf (accessed March 2021)

Section 45. Schistosomiasis

Introduction

Schistosomiasis occurs in areas of the world where there is a 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 from humans in stool or urine into freshwater containing snails, *Bulinus* (*S. haematobium*), *Biomphalaria* (*S. mansoni*) and *Oncomelania* (*S. japonicum*). Miracidia hatch from the eggs, penetrate the snail, and replicate into cercariae (larval forms) which are then released into the water.

The cercaria penetrates the skin (or pharyngeal mucosa) of humans, loses its tail and becomes a schistosomula, which is then 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 about 1 cm in length in the liver.

Adult males and females copulate and migrate in pairs to their preferred egg-laying sites, *S. haematobium* to the vesical veins and pelvic plexus, and *S. mansoni* to the superior and inferior mesenteric veins.

Female flukes produce eggs daily throughout their average 3- to 4-year 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 (genitourinary system - *S. haematobium*; bowel and liver – other species). Eggs may also be transported to other organs causing serious pathology (e.g., lungs -> pulmonary hypertension, CNS -> transverse myelitis).

Pathogenesis

Pathogenesis can be divided into four stages.

- Dermatitis.
 - An itchy papular rash 'swimmers itch' lasting one to two 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.
- Katayama fever (2–8 weeks).
 - A 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.
 - It is a self-limiting disease.
- Established disease (usually after 2 months).
 - A T-cell delayed-hypersensitivity response to eggs deposited in tissue results in granuloma formation.
 - If the worm load is reduced by drug therapy at this stage, granulomata may resolve, leaving little disease.
- Fibrotic complications.
 - Repeated infections without treatment eventually result in fibrosis, for

- example of the ureter and bladder (*S. haematobium*) and liver (*S. mansoni*).
- There is little response to drug therapy at this stage.

TABLE 45.1 Schistosomiasis: geographical areas (the commonest species and areas are shown in bold type)

Schistosoma species	Disease	Area
S. haematobium	Genitourinary tract	Africa, Middle East
S. mansoni	Intestines, liver	Africa, Middle East, South America
<i>S. intercalatum</i> <i>S. guineensis</i>	Intestines, liver	Central and West Africa, uncommon
S. japonicum	Intestines, liver	China, Indonesia, Philippines
<i>S. mekongi</i>	Intestines, liver	Laos, Kampuchea, small number of foci

Epidemiology

- Schistosomiasis affects at least 240 million people worldwide, and more than 700 million people live in endemic areas.
- Schistosomiasis is associated with communities living near swamps, rivers, irrigation canals and rice fields, who have poor hygiene and sanitary facilities and lack a 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 45.2 Symptoms and complications of *S. haematobium* and *S. mansoni*

Initial stage	<i>S. haematobium</i>	<i>S. mansoni</i>	Comments
Swimmers’ itch	<ul style="list-style-type: none"> • Terminal haematuria 	<ul style="list-style-type: none"> • Bloody diarrhoea • Anaemia 	<i>S. japonicum</i> is similar to <i>S. mansoni</i>
Katayama fever	<ul style="list-style-type: none"> • Obstructive uropathy • Calcification of bladder and lower ureters • Bladder calculi • Bladder cancer • Infertility 	<ul style="list-style-type: none"> • Hepatic fibrosis • Portal hypertension • Ascites • Colonic polyposis • Nephropathy 	Hepatic fibrosis is most often seen with <i>S. mansoni</i> Katayama fever is more severe with <i>S. japonicum</i>

S. haematobium

- This causes urinary schistosomiasis.
- Terminal haematuria, there may be dysuria.
- In a minority of children, frequent untreated infections eventually lead to structural disorder of the 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

- This causes intestinal schistosomiasis along with other species, namely *S. intercalatum*, *S. japonicum* and *S. mekongi*, *S. intercalatum* and *S. guineensis*.
- Bloody diarrhoea. In long-standing cases there is severe iron-deficient anaemia, and even heart failure (due to anaemia).
- Protein-losing enteropathy with hypoalbuminaemia may result from colonic granulomatous disease and polyps.
- The left lobe of the liver is affected more than the right lobe. Liver function is usually well preserved. Ascites may occur as the disease progresses.
- Ultrasonography is useful for grading the degree of peri-portal fibrosis and in differential diagnosis from other liver diseases.
- Marked splenomegaly due to portal hypertension results in pancytopenia.
- Haematemesis from oesophageal varices influences the prognosis and may be fatal.
- Acute and long-term management of oesophageal varices requires endoscopy and decisions regarding sclerotherapy (see Section 49 Handbook 1 on liver disease).
- Nephropathy due to immune complex disease may manifest with microscopic haematuria and proteinuria or nephrotic syndrome. Nephrotic syndrome associated with schistosomiasis-salmonella co-infection usually responds to treatment with praziquantel and an appropriate antibiotic. Nephrotic syndrome may have a poor prognosis, especially if associated with amyloid disease (see Section 46 Handbook 1).

Complications common to S. haematobium and S. mansoni:

- Spinal cord myelopathy (less common with *S. haematobium*).
- Brain granulomata (more common with *S. japonicum*, less common with *S. haematobium*).
- Pulmonary hypertension (less common with *S. haematobium*).

Salmonella co-infection

- *Schistosoma* worms may harbour *Salmonella* species, including *S. Typhi*, which cannot be eradicated until the schistosomiasis is treated. This phenomenon occurs in both *S. haematobium* and *S. mansoni* infections.
- *Salmonella* may cause a reversible nephritis in *S. mansoni* infection.

Diagnosis

Microscopy of urine or faeces

S. haematobium

- An end-stream midday 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 that 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. Contra-indicated if there is portal hypertension.

Serology

- Serology is of little value for diagnosis in indigenous patients but may be useful in the non-immune (e.g. tourists to an endemic area).
- Antigen tests are being developed.
- PCR of urine or faeces may be useful.

Treatment

Praziquantel is effective against all human *Schistosoma* species, and is the only widely available drug treatment in endemic regions. Treatment at least three times in childhood usually prevents adult disease.

- Praziquantel is given at a dose of 40 to 60 mg/kg in two divided doses given 4 - 6 hours apart on one day.
 - It can also be given as a single dose of 40 to 60 mg/kg.
 - For heavy *S. mansoni* infection and for *S. japonicum* infection, 60 mg/ kg is advised, given in two doses 4 - 6 hours apart. This higher dose is also increasingly recommended for *S. haematobium* infections.
 - Repeat urine or stool examination should be done at 3 - 4 months.
- Praziquantel is safe during pregnancy.
- The safety of praziquantel in children under 4 years of age has not been established, but this drug can be used to treat individually infected children.
- Side effects include dizziness, drowsiness, skin reactions, fever, headache and vomiting.

Prevention

Control of schistosomiasis is very difficult.

Measures include:

- Regular mass treatment of communities and improvement in water supply, sanitation and hygiene.
- Mollusciciding (use of chemicals to kill the snails) is usually impractical and too expensive for general use.

[World Health Organization \(2015\) Female genital schistosomiasis: a pocket atlas for clinical health-care professionals.](https://apps.who.int/iris/bitstream/handle/10665/180863/9789241509299_eng.pdf?sequence=1) (accessed 04/03/2021)

https://apps.who.int/iris/bitstream/handle/10665/180863/9789241509299_eng.pdf?sequence=1

Section 46. Scrub Typhus

Epidemiology

- Geographical distribution:
 - Asia, Australia and Pacific Islands.
- Agent:
 - *Orientia tsutsugamushi* (*Rickettsia tsutsugamushi*).
- Hosts:
 - Rodents are reservoir hosts, and humans are accidental hosts.
 - The most affected age group is 5–14 years, and the disease is 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 is 5 - 18 days.
- Abrupt onset of fever, severe headache, myalgia, cough, suffused conjunctivae, dark red papular or maculopapular rash (5 - 7 days after fever) on the trunk, arms, and thighs.
- Eschar (19 - 28% in children, 46 - 82% in adults) may be seen at the site of the mite bite, especially in the perineum, axilla, or trouser-belt region. Eschar is a firmly adherent black scab, 3 - 6 mm in diameter, with a raised red margin.
- There is regional or generalised lymphadenopathy, hepatomegaly and sometimes a 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 2 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 (PCR).
- Weil–Felix test titres of 1:160 (or a fourfold rise after 2–4 weeks) occur in only 50% of cases.
- More sensitive serological tests are the indirect immune-peroxidase test and the indirect immunofluorescent tests.
- For individuals living in endemic areas the positive titre is $\geq 1:400$ or a fourfold rise in acute and convalescent sera. The positive titre indicating infection may be lower in non-endogenous children. PCR on the eschar material is more sensitive than on the blood.
- 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 (e.g., 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

The drug of choice is:

- 1 Doxycycline orally 2.2 mg/kg initially followed by,
 - a. 2.2 mg/kg 12 hours later, then,
 - b. 1.1 mg/kg every 12 hours until the patient is afebrile for 2–3 days, or
 - c. Continue treatment for 5–7 days.

Alternative drugs are:

- 2 Tetracycline 250 mg orally four times a day for 7 days (in children over 8 years) or,
 - 3 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. If this happens, repeat the dose of antibiotic.
 - Tetracycline should not be given to oliguric patients. Doxycycline is safe in renal impairment.
 - Rifampicin and azithromycin have been used successfully in areas where the rickettsia is resistant to conventional treatment.
 - In severe cases, the risk of dying outweighs the risk of tooth discoloration from doxycycline or tetracycline.
 - Remember that antimicrobial agents only suppress infection. Cure depends on host immunity.
 - Treatment should not be withheld pending laboratory confirmation for a clinically suspected infection.

Reference

[European Centre for Disease Prevention and Control \(2020\) Facts about epidemic louse-borne typhus](#) (accessed 04/03/2021)

Section 47. Sexually Transmitted Infections

Introduction

Anogenital infections in childhood are most acquired through sexual contact or abuse but may also 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.

Diagnosis

The diagnosis of sexually transmitted disease is considered in the following circumstances:

1. A history of recent sexual abuse
2. The isolation of sexually transmitted organisms in cases without obvious trauma leading to a diagnosis of chronic sexual abuse
3. Specific syndromes and diseases usually transmitted by the sexual route in adults
4. Congenital syphilis or perinatally acquired chlamydia, or
5. Gonorrhoea transmitted from the mother in utero or postnatally
6. HIV infection not acquired perinatally, through transfusion or another known mechanism.

There are more than 30 different infections that may be spread by the sexual route. These range from the classic sexually transmitted diseases (e.g., syphilis, gonorrhoea), through conditions that are mainly sexually transmitted (e.g. genital herpes, human papillomavirus), to those infections that can also be transmitted by sexual means (e.g. hepatitis B and C).

Sexual abuse

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 such as gonorrhoea, syphilis or chlamydia are usually manifested soon after the assault, with 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

1. Management of the sexual abuse (see Section 2)
2. Local management of injuries, including tetanus toxoid if applicable
3. Bacteriological swabs for gonococcus and chlamydia (or nucleic acid amplification test (NAAT)) and if blisters or ulcers swab for herpes simplex PCR and syphilis PCR
4. Serological tests for syphilis, hepatitis B and HIV, repeated 6 weeks later
5. Prophylactic broad-spectrum antibiotics:
Ceftriaxone 50 mg/kg IM as a single dose (maximum dose 500mg)
plus
Azithromycin 1gram orally in a single dose in children weighing >45kg
or
Erythromycin 20-40 mg/kg/day in three divided doses for 7 days in children weighing <45kg
6. Post-exposure hepatitis B vaccination if not previously vaccinated; follow-up doses at 1-2 and 4-6 months after the first dose

7. In males aged 9-21 years and females aged 9-26 years, initiate HPV vaccine series.; follow up at 1-2months and 4-6months ***

*** Rationale: Child sexual-assault survivors are at increased risk for future unsafe sexual practices that have been linked to higher risk of HPV acquisition and are more likely to engage in these behaviours at an earlier age. HPV vaccine will not protect against progression of infection already acquired or promote clearance of the infection but will protect against vaccine types not yet acquired.

8. Pregnancy prophylaxis in post pubertal girls
9. Assessment of the risk of HIV transmission and prophylaxis if indicated.

Children are at higher risk because episodes of assault are often multiple and mucosal trauma is likely. Factors that should be assessed include the following:

- Assailant's HIV status or likelihood of having HIV
- Time elapsed since incident (< 72 hours)
- Exposure characteristics
- Possible benefits and risks associated with post- exposure prophylaxis (PEP).

Post Exposure Prophylaxis (PEP) is generally well tolerated in children. The choice of antiretroviral drugs will depend on local availability and policy. An example is a combination of zidovudine, lamivudine, and lopinavir/ritonavir, or tenofovir with emtricitabine and raltegravir. Follow-up and appropriate treatment of identified infection (see below) should be undertaken. (see Section 36 for more on HIV).

The presence of a sexually transmissible infection in a child alerting 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 aged around 2–10 years, 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.

However, Herpesvirus type 2, *Trichomonas vaginalis*, *Mycoplasma* species and bacterial vaginosis are not so commonly acquired because of sexually transmitted infection in this age group.

Although 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 adolescent children

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 or RPR) and HIV (ELISA) should be offered with appropriate counselling in all patients.
- Prevention of new infection by education about safe sex practices and condom use.
- Motivation to engage in health-seeking behaviour.

Genital ulcers and lymphadenitis

The infections presenting with genital ulcers with or without inguinal adenopathy and bubos are most often acquired because of voluntary or involuntary sexual activity but may occur because 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, lymphogranuloma venereum or granuloma inguinale.

Genital herpes

Characteristics are painful vesicular or shallow ulcerative lesions on the genitals. Grouped or single lesions occur on a thin erythematous base but with generally uninfamed intervening epithelium. These regress spontaneously but may recur.

Oral aciclovir 200 mg five times daily for 5 days 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, immunofluorescent antibody stains or PCR. At this stage, serological tests for syphilis are usually still negative.

The treatment in children over 12 years consists of **benzathine benzylpenicillin**, 50 000 U/kg IM as a single dose (50 000 U = 37.5 mg). Benzathine benzylpenicillin must not be given IV.

Chancroid

The aetiological agent of chancroid is the bacteria *Haemophilus ducreyi*. Painful papules form pustules which erode to form ulcers on the genitals. These 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 the treatment of children over 12 years of age, the following are satisfactory: **azithromycin** 1 gram orally as a single dose, **OR ceftriaxone** 250 mg IM as a single dose, **OR erythromycin** base 500 mg orally four times daily for 7 days.

Lymphogranuloma venereum

Patients with lymphogranuloma venereum (LGV) commonly present with unilateral tender inguinal and/or femoral lymphadenopathy. Genital ulcers are usually less obvious and have often disappeared by the time of presentation. LGV is caused by *Chlamydia trachomatis*.

Treatment for children over 12 years of age is with **doxycycline** 100 mg orally twice daily or **erythromycin** 500 mg orally four times daily and should be continued for 21 days.

Granuloma inguinale

Klebsiella granulomatis is the cause of this uncommon ulcerative disease. The lesions are painless and slowly progressive. Subcutaneous granulomas, which may ulcerate, often occur on the genitals and perineum, but regional lymphadenopathy is absent.

Treatment in older children is with **azithromycin** 1 g orally once per week or 500 mg daily for at least 3 weeks and until all lesions have completely healed.

Alternatively, doxycycline, erythromycin, ciprofloxacin or trimethoprim-sulfamethoxazole can be used.

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 non-specific.

The organisms responsible for this mode of presentation include *Neisseria gonorrhoeae*, *Chlamydia trachomatis*, *Trichomonas*, *Candida* species, *Gardnerella vaginalis* and *Ureaplasma* species.

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.

Recommended treatment for children over 12 years of age includes:

- Ceftriaxone 250 mg IM as a single dose, or cefixime 400 mg orally in a single dose, against *Neisseria gonorrhoeae*.
- Azithromycin, 1 gram orally as a single dose, or
- Doxycycline 100 mg orally twice daily for 7 days
- Alternatively, Erythromycin 40–50 mg/kg/day given as 4 divided doses 6 hourly for 14 days for children under 12 years should be added for *Chlamydia*.
- If *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 can be treated with a short course (3 days) of topical azoles such as clotrimazole, miconazole or butoconazole cream. An alternative is treatment with local nystatin (100 000 U/mL three to four times daily), but this is less effective.

Acute balanoposthitis

Inflammation of the glans and prepuce can have many 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 may 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, STDs are also 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 over 12 years is azithromycin 1 gram 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 (HPV) 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.

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.
 - Other precautions to avoid the development of complications include limiting the application to less than 0.5 mL of podophyllin and an area of over 10 cm² of warts per session.
- The preparation should be washed off 1–4 hours after application to reduce local irritation.
- The process can be repeated in 7 days.
- Other treatment modalities include cryotherapy, surgical excision, curettage or electrocautery.
- An alternative is not to treat, and to await possible spontaneous resolution.

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.

Two HPV vaccines are now available. They offer protection against HPV types that cause a large percentage of carcinomas as well as genital warts.

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 outpatient treatment typically includes a third-generation cephalosporin like ceftriaxone plus doxycycline. Metronidazole may be added to treat bacterial vaginosis which frequently accompanies PID.

In severe cases, or where there is no response to the above treatment within 72 hours, intravenous broad-spectrum antibiotics including an aminoglycoside and clindamycin should be given.

[WHO guidelines for the treatment of genital herpes simplex virus \(accessed 04/03/2021\)](#)

[WHO guidelines for the treatment of Neisseria gonorrhoeae \(accessed/04/03/2021\)](#)

[WHO guidelines for the treatment of Chlamydia trachomatis \(accessed 04/03/2021\)](#)

[WHO guidelines for the treatment of Treponema pallidum \(syphilis\) \(accessed 04/03/2021\)](#)

Section 48. Strongyloidiasis

Introduction

This parasite affects 100–200 million people worldwide, and occurs in warm, wet, tropical and subtropical regions where sanitation is poor.

Strongyloides stercoralis is the main species infecting humans.

However, *Strongyloides fülleborni*, which is principally a parasite of primates, also occurs in humans in Africa and Papua New Guinea.

Human infection is due to percutaneous penetration of filariform larvae in contaminated soil. Filariform larvae travel via the lungs to the small intestine, where they develop into adults and penetrate the duodenal and jejunal mucosa. Fertilised females produce eggs which hatch in the intestinal mucosa and release the first stage rhabditiform larvae, which are excreted in faeces. In favourable conditions, the rhabditiform larvae transform into infectious filariform larvae within 48 hours and remain viable in the soil for weeks.

An important feature of *Strongyloides* is auto-infection. This occurs when rhabditiform larvae transform into infectious dwarf filariform larvae in the gut lumen and penetrate the mucosa or the peri-anal skin. Infection may persist for decades without further exposure.

Person-to-person transmission may also occur.

Clinical features

Initial skin penetration may cause itching and urticaria. Migration through the lungs may cause cough, wheeze and evidence of pneumonitis. Invasion of the small bowel may cause abdominal pain, vomiting, malabsorption, and paralytic ileus. Chronic infection is often asymptomatic, but may cause intermittent abdominal pain, diarrhoea and urticaria. Malabsorption and a protein-losing enteropathy may occur. A transient, intensely itchy serpiginous (snake like) rash, known as '*larva currens*' or '*creeping eruption*', may appear on the trunk, buttocks or elsewhere. Episodic pneumonitis and, more rarely, a reactive arthritis may occur.

Strongyloides hyper-infection syndrome

One of the major dangers associated with *Strongyloides* infection occurs because of massive autoinfection. Risk factors include immunosuppression induced by various drugs, including corticosteroids, or associated with diseases such as malignancies (particularly leukaemia and lymphoma), severe malnutrition and severe infections, including advanced AIDS and human T-cell leukaemia virus type 1 (HTLV-1).

Hyper-infection syndrome may present with severe diarrhoea, often with blood in the stool. Bowel inflammation with micro-perforations may give rise to paralytic ileus, peritonitis, and Gram-negative septicaemia. Proliferation and dissemination of larvae and enteropathogens may cause widespread pathology, including endocarditis, pneumonitis, and meningitis.

All patients with a history of possible exposure to *Strongyloides* should be screened before being treated with any drugs that cause immunosuppression. Those at significant risk should be treated empirically even if investigations are negative.

Investigations

- Eggs are rarely found in the stool, and larvae may be difficult to identify. Stool culture (e.g., on charcoal or agar) is recommended.
- Faecal PCR, if available, may be useful.

- Larvae may be seen in duodenal aspirates or using the string capsule technique (Enterotest).
- Larvae may also be found in sputum, CSF, and urine in hyper-infection syndrome.
- Serology is useful for immune-competent patients who are not normally resident in an endemic area. However, interpretation of a positive test may be a problem due to cross-reactions with filarial antigens.
- Eosinophilia is common in immune-competent patients but may be absent in hyper-infection syndrome.

Treatment

- Ivermectin is the drug of choice for children over 5 years old or weighing more than 15 kg.
- An oral dose of 200 micrograms/kg/day for 2 days gives excellent results.
- There is evidence that ivermectin at this dose is also safe in children < 5years and weighing < 15 kg.
- Ivermectin should be used with caution in areas endemic for Loa loa because of the risk of provoking encephalopathy in individuals with Loa loa microfilaraemia > 2500 Mf/ml.
- Albendazole 400 mg every 12 hours for 7 days may also be effective and can be used in children over 2 years of age.
- Hyper-infection syndrome can be very difficult to manage.
- There may be problems with administration or absorption of oral medication, and no IV or IM preparations of ivermectin or albendazole are licensed for use in humans.
- However, parenteral ivermectin, available as a veterinary preparation, has been administered subcutaneously in the successful treatment of Strongyloides hyper-infection.
- Patients with hyper-infection syndrome also require treatment for Gram-negative septicaemia.

Prevention and control

- Improve hygiene and sanitation.
- Wear shoes.
- Avoid contact with contaminated soil.

https://www.who.int/selection_medicines/committees/expert/22/applications/s6.6_ivermectin.pdf

Section 49. Syphilis

Introduction

Syphilis is a dangerous bacterial infection caused by *Treponema pallidum* which, when it occurs in pregnancy, can cause early fetal death, stillbirth, preterm birth, neonatal death, or congenital infection. Mother-to-child transmission is a major problem, especially in resource-limited countries.

Congenital syphilis may be acquired from an infected mother via trans-placental transmission of *Treponema pallidum* at any time during pregnancy. If the mother receives adequate treatment, ideally before the second trimester, the risk of adverse outcome to the fetus is minimal.

Clinical signs in infants may include any of the following:

- low birth weight with a heavy placenta
- palms and soles showing a red rash, grey patches, blisters, or skin peeling
- abdominal distension due to large liver and spleen
- jaundice
- anaemia
- some low-birth-weight infants with syphilis show signs of severe sepsis, with lethargy, respiratory distress, skin petechiae or other signs of bleeding.

Investigation

No newborn infant should be discharged from hospital without determination of the mother's serologic status for syphilis at least once during pregnancy, and also at delivery in communities and populations in which the risk of infection with congenital syphilis is high.

If you suspect syphilis, perform a venereal disease research laboratory (VDRL), rapid plasmin reagent (RPR) or rapid syphilis test on the infant's serum (not cord blood). Interpretation of the serological results in the neonate can be difficult, as maternal IgG antibodies are transferred across the placenta. A non-treponemal serological titre that is fourfold higher than the mother's titre is definitely significant, although a lower titre does not exclude congenital syphilis. As well as a careful examination of the infant, an x-ray of long bones (if available) may help with the diagnosis. Periostitis, metaphysitis and erosions of long bones are the commonest findings.

Because of the diagnostic difficulty, and the fact that infants may be asymptomatic, assessing the adequacy of maternal treatment is very important.

Treatment

All newborn infants of mothers with syphilis should be investigated and treated. Adequate treatment for the mother is 3 doses of benzathine penicillin, given at least 4 weeks before delivery.

Infants should be treated for congenital syphilis if they have proven, or probable, disease demonstrated by one or more of the following:

- physical, laboratory or X-ray evidence of active disease
- a reactive result on maternal or infant VDRL testing where the mother has had no treatment, or inadequate treatment, or has had a non-penicillin antibiotic, even if the infant is asymptomatic.

Parenteral benzyl penicillin remains the preferred drug for treatment of an infant with any signs of congenital syphilis. The dose is 100,000-150,000U/kg/day given

intravenously as 50,000 U/dose (37.5 mg) 12 hourly for the first 7 days and then 8 hourly for 3 days (total 10 days).

An alternative is procaine penicillin 50 000 units/kg or 50 mg/kg as a single dose by deep IM injection daily for 10 days. Ensure that this is not injected into a vein.

Asymptomatic neonates born to VDRL-positive or RPR-positive women, who have been adequately treated, should receive 37.5 mg/kg (50 000 units/kg) of benzathine benzyl penicillin as a single IM dose into the anterolateral thigh whether or not their mothers were treated during pregnancy.

Routine CSF examination is not required. Ensure that the needle is not in a vein when this drug is given, by drawing back and ensuring that no blood is in the needle, as it can cause cardiac arrest and severe CNS damage if given IV.

Early congenital syphilis generally responds well to penicillin. Recovery may be slow in seriously ill infants with extensive skin, mucous membrane, bone, or visceral involvement.

If the patient is allergic to penicillin (this is unusual), give ceftriaxone IM/IV once daily for 10 days. The dose for a neonate aged up to 15 days is 50mg/kg once daily; 15-28days 75-100mg/kg once daily, or give erythromycin, 7.5–12.5 mg/kg orally, four times a day for 14 days but erythromycin is less effective.

Where congenital syphilis was treated or suspected, the baby should be followed up. Non-treponemal test titres should decline over 6 months. If titres remain high at 6-12 months, the infant should be re-evaluated. Always treat both the mother and partner for syphilis and check for other sexually transmitted infection.

Section 50. Trachoma

Introduction

Trachoma is the most common infectious cause of blindness worldwide. It is caused by *Chlamydia trachomatis*, certain serotypes of which preferentially infect the conjunctival epithelium.

The organism is transmitted from person to person by direct contact, fomites (objects capable of carrying infectious organisms), and eye-seeking flies. Disease clusters in families; the greatest risk factor for infection is sharing a bedroom with an active case.

Repeated episodes of infection over many years cause an accumulation of scar tissue in the tarsal plate and tarsal conjunctivae of the upper lids. Contraction of the scar may produce trichiasis and/or entropion, and the resulting corneal abrasion by in-turned lashes leads to corneal scarring. This eventually causes blindness. In paediatric practice in endemic areas, active trachoma is seen frequently.

Blinding complications may start to appear in the second and third decades of life.

Clinical features

These are best presented using the framework of the WHO simplified clinical grading system. Examination for trachoma involves inspection of the lashes and cornea, followed by eversion of the upper eyelids to examine the tarsal conjunctivae (see Figure 50.1).

A $\times 2.5$ 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—follicular (TF):

The presence of five or more follicles at least 0.5 mm in diameter in the central part of the upper tarsal conjunctiva.

Follicles appear as white or yellow-grey semi-transparent patches or swellings beneath the conjunctiva.

Fewer than five follicles, or follicles at the nasal or temporal margin, may be normal.

Trachomatous inflammation—intense

Pronounced inflammatory thickening of the upper tarsal conjunctiva obscuring more than half of the normal deep tarsal blood vessels.

TF and TI are both forms of 'active trachoma'; they are associated with infection with *Chlamydia trachomatis*, although not all infected individuals exhibit these signs, and not all individuals with these signs are infected. Patients with active trachoma may be asymptomatic or complain of irritable red eyes.

Trachomatous scarring (TS):

The presence of easily visible scars in the tarsal conjunctiva.

Scars appear as white bands, lines, or sheets. TS is the result of repeated cycles of inflammation and resolution over many years and itself is virtually asymptomatic, although scarring of eyelid glands may produce symptoms of dry eye.

Trachomatous trichiasis (TT):

At least one eyelash from the upper eyelid rubs on the eyeball, or there is evidence of recent removal of in-turned eyelashes from the upper eyelid. TT is intensely irritating to the sufferer, and they may choose to pull out their eyelashes in an attempt to reduce the discomfort.

There may be discharge from superadded bacterial infection of the abraded cornea. Except in hyperendemic areas, it is unusual to observe TT in children.

Corneal opacity (CO):

Easily visible corneal opacity over the pupil, so dense that at least part of the pupil margin is blurred when viewed through the opacity. Such corneal opacities cause significant visual impairment.

It is important to remember that these grades are not mutually exclusive. A patient with active trachoma (TF and/or TI) may also show signs of the late complications of the disease.

There are other signs of trachoma that are not included in the simplified grading scheme:

- Papillae are often visible in individuals with active trachoma 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.
- Fibrovascular connective tissue may grow inwards from the limbus to invade the anterior layers of the superior cornea in response to infection. The ingrowth is known as pannus. The new blood vessels may persist after resolution of infection.
- Sometimes follicles are found under the bulbar conjunctiva at the limbus as well as deep to the tarsal conjunctiva. Scarring of limbal follicles may subsequently leave small depressions known as Herbert's pits.

Treatment

For active trachoma (TF and/or TI), antibiotics are required.

- Topical tetracycline eye ointment 1% is effective when applied to both eyes twice daily for 6 weeks.
- A single dose of oral azithromycin (20 mg/kg, up to a maximum dose of 1 gram) is just as effective, is better tolerated than topical tetracycline, and can be directly observed, so is associated with higher compliance rates.
- Trichiasis or entropion requires surgical management to restore the margin of the eyelid to its normal position, so that contact between the lashes and globe is interrupted.
 - Bilamellar tarsal rotation and posterior lamellar tarsal rotation (Trabut) are the procedures currently recommended by the World Health Organization; they are performed under local anaesthetic and can be undertaken at the village level by trained ophthalmic nurses or ophthalmic assistants.
- Corneal opacity can theoretically 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 may be high.

The identification of signs of trachoma in an individual should prompt screening of other members of that individual's community. Antibiotic treatment of individuals presenting to healthcare facilities is unlikely to have any impact on the incidence of blindness from trachoma in the communities from which those individuals come.

Comprehensive community-based management of trachoma is necessary wherever the prevalence of disease is high.

FIGURE 50.1

- (a) Normal tarsal conjunctiva (×2 magnification). The dotted line shows the area to be examined.
(b) Trachomatous inflammation – follicular (TF).
(c) Trachomatous inflammation – follicular and intense (TF + TI).

Prevention

Blindness from trachoma is preventable.

The acronym SAFE has been adopted by the WHO and partners to encapsulate the recommended approach to controlling trachoma. It comprises:

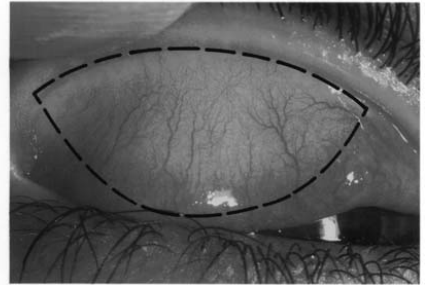
- **S**urgery for trichiasis
- **A**ntibiotics to clear infection, and
- **F**acial cleanliness, and
- **E**nvironmental improvement (provision of water and acceptable means for disposal of human faeces) to reduce transmission.

Surgery should be offered to all individuals with trichiasis. The ‘A’, ‘F’ and ‘E’ components should be implemented district-wide wherever the prevalence of TF in 1- to 9-year-olds is 5% or higher. WHO and partners are using the ‘SAFE’ strategy with the target of achieving global elimination of trachoma as a public health problem by the year 2030.

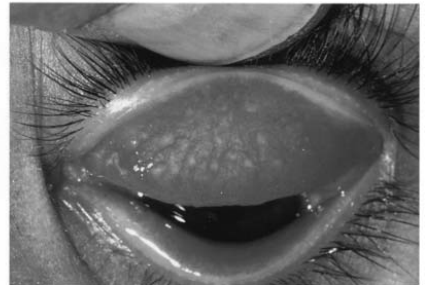
References

[WHO | Trachoma simplified grading card \(accessed 04/03/2021\)](#)

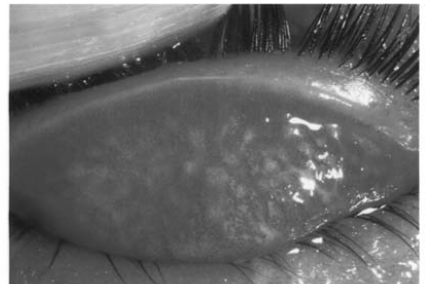
WHO Strategic and Technical Advisory Group on Neglected Tropical Diseases. Design parameters for population-based trachoma prevalence surveys (WHO/HTM/NTD/PCT/2018.07). Geneva: World Health Organization; 2018.



a Normal tarsal conjunctiva (x 2 magnification). The dotted line shows the area to be examined.



b Trachomatous inflammation – follicular (TF).



c Trachomatous inflammation – follicular and intense (TF + TI).

Section 51. Tuberculosis

Introduction

The global incidence of tuberculosis (TB) has been falling very slowly since the Moscow Declaration to End TB from the WHO conference in 2017. Children aged <15 years old account for 12% of TB cases. 49% of people with TB face catastrophic costs. About 8.2% of TB cases occur among people living with HIV. South-East Asia accounted for 44% of notified cases and Africa accounted for another 25%. In 2019 close to half a million people developed drug resistant TB.

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. Multi-drug resistance is a major concern.

Epidemiology

- In low-income countries, the risk of developing infection is up to 2.5% per annum.
- The age group at highest risk of developing disease is 0–5 years, with risk up to 30–40% (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 non-infectious, except for older children and adolescents with cavitary TB.
- Children with untreated tuberculosis contribute to the pool of adults with reactivated disease.

Factors that predispose tuberculosis-infected children to develop systemic disease:

- Age under 5 years, and especially under 1 year.
- Household contact with smear-positive disease.
- Malnutrition.
- Tuberculosis infection in previous 2 years.
- Immunosuppression, especially HIV infection.

Latent TB

Clinically assess children of contacts with TB. If no features of active TB recommend treatment for all children <5 years old or children with HIV who have household contact with TB (if no contraindication such as active hepatitis or peripheral neuropathy). Children without HIV or >5 years old, in contact with bacterially proven TB, perform a tuberculin skin test.

Tuberculin skin test

The tuberculin skin test (TST), also called the Mantoux test, is useful for screening contacts. The TST is less useful for diagnosing active TB because a negative result does not exclude TB. If TB is clinically suspected, efforts should be made to collect diagnostic specimens, exclude other causes, and then treat if TB is the most likely diagnosis (do not treat as a diagnostic test).

1. Use either 5 TU of tuberculin (PPD-S) or the 2 TU of tuberculin (PPD RT 23).
2. Inject tuberculin (PPD-S) intradermally into the upper third of the flexor surface of the forearm with a 1-mL syringe and a short bevel gauge 25–27 needle producing a wheal of at least 5 mm.
3. Read the transverse diameter of induration at 48–72 hours.
 - a. Regard induration of <10 mm as negative and likely to be associated with environmental mycobacteria, and 10 mm or more as indicative of infection with *Mycobacteria tuberculosis*.

In resource-limited countries where BCG is given at birth, most children will have a negative tuberculin test by 10 years of age, and thus an induration of 10 mm in children this age or 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, with these conditions an induration of >5 mm may be indicative of tuberculosis. Remember that a negative tuberculin does not exclude tuberculosis, and additional work-up may be warranted in any suspected child.

Tuberculin skin test interpretation must be undertaken bearing in mind the age, epidemiology and underlying illness, if any.

Serology

Antibody tests are inconsistent and imprecise. They do not improve outcomes for patients and should not be used. The WHO recently gave guidelines recommending against the use of serology tests for TB. It is the first negative recommendation to be made by the WHO, describing it as 'inaccurate and useless', after 'overwhelming' evidence that suggested it produced an 'unacceptable level' of false-positive or false-negative results.

Pathogenesis

Inhalation of the tubercle bacillus into an alveolus establishes the primary (Ghon) focus. In the 4 to 8-week period before the cell-mediated immune (CMI) response develops, there is spread to regional lymph nodes, and small numbers of bacilli disseminate throughout the body in the lympho-haematogenous system.

Certain organs favour survival of tubercle bacilli, including regional nodes, epiphyseal lines of bones, cerebral cortex, renal parenchyma and apical regions of the lungs (Simon focus).

Establishment of an adequate CMI response (which coincides with the appearance of sensitisation to tuberculin) in most cases results in control or eradication of proliferating tubercle bacilli at these sites.

The 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 lympho-bronchial disease, with local compression or erosion of the bronchi.

Extrathoracic disease is due to local spread of disease at metastatic sites (e.g. 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.

The risk of developing symptomatic disease following primary tuberculosis is highest (around 50%) in the first 1 to 2 years after infection and the rest in the individual's lifetime.

Tuberculosis in adolescence

This 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 extra-thoracic dissemination is uncommon.

Clinical features

In well-resourced countries, the majority of children with respiratory tuberculosis are asymptomatic and are picked up through contact tracing and will generally have early primary disease.

In resource-limited countries, only children with symptomatic disease present, and they are therefore only ‘the tip of the iceberg’.

The following are some of the key features of tuberculosis in children:

- Fever, cough, anorexia, weight loss, wheezing, night sweats and malaise are common.

Extrapulmonary disease may involve other tissues and organs, such as the central nervous system, lymph nodes and gastrointestinal tract.

Findings can include lung findings (dull resonance) or involvement of other organs in extrapulmonary tuberculosis, such as hepatosplenomegaly, lymphadenopathy, mass, etc. (see Table 51.1).

TABLE 51.1 Typical features of common forms of extrapulmonary TB in children

Type of extrapulmonary TB	Key clinical findings
TB lymphadenitis (most common)	Enlargement and swelling of lymph nodes
Pleural/pericardial TB	Cough and shortness of breath
TB meningitis	Headache, vomiting, fever, neck stiffness, seizures, confusion and coma
Miliary TB	Very sick, respiratory distress, hepatosplenomegaly, diffuse lymphadenopathy
Gastrointestinal TB	Abdominal pain, diarrhoea, mass or ascites
Spinal TB	Backache with or without loss of function in lower limbs
TB arthritis	Pain and swelling of joints (usually mono-arthritis)

HIV and tuberculosis

Children living with HIV who have poor weight gain, fever or current cough, or contact history with a TB case, may have TB and should be evaluated for TB and other conditions. If the evaluation shows no TB, children should be offered isoniazid preventive therapy (IPT) regardless of their age.

Children living with HIV who do not have poor weight gain, fever or current cough are unlikely to have active TB. Children living with HIV who are more than 12 months of age and who are unlikely to have active TB on symptom - based screening and have no contact with a TB case should receive 6 months of IPT (10 mg/kg/day, maximum 300 mg/ day) as part of a comprehensive package of HIV prevention and care services.

Regarding children living with HIV who are less than 12 months of age, only those who have contact with a TB case and who are evaluated for TB (using investigations) should receive 6 months of IPT if the evaluation shows no TB disease.

All children living with HIV after successful completion of treatment for TB disease should receive isoniazid for an additional 6 months.

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 jirovecii* pneumonia before contracting tuberculous infection.

In older children, there is difficulty in diagnosis due to the following reasons:

- The tuberculin reaction is positive in only 20% of cases.
- There is confusion with HIV-related respiratory disorders, including:
 - Lymphocytic interstitial pneumonitis (LIP),
 - Superimposed viral/bacterial infections,
 - Chronic interstitial pneumonitis,
 - Kaposi's sarcoma and bronchiectasis.
- There is often a lack of facilities for culturing *M. tuberculosis*.
- Atypical clinical and radiological features of TB are much more common in children with HIV with more severe and complicated disease.
- HIV/tuberculosis co-infected children are more likely to develop disseminated tuberculosis and meningitis, have cavitary pulmonary disease and may have a poor response to treatment and a higher mortality if not started on ART.

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.

Standard 6-month chemotherapy is given in uncomplicated pulmonary tuberculosis.

Respiratory tuberculosis

Most respiratory tuberculosis results from complications of lympho-bronchial 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, other immunosuppressed states and/or malnutrition.

Pericarditis

Tuberculosis should be considered in all cases of pericarditis. *M. tuberculosis* may be cultured from a pericardial tap in over 50% of the cases.

Lymph node disease

This may result from a focus in the upper lung fields or from haematogenous spread.

Diagnosis may be made by biopsy or fine-needle aspiration.

Swelling and softening of nodes may continue for months after treatment has been completed.

In well-resourced 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

This is commonest in young children and in those who are immunosuppressed, usually occurring within 3–12 months of primary infection.

Chest X-ray (except in the early stages) will demonstrate a 'snowstorm' or miliary appearance.

Meningitis is a common complication. Therefore, a lumbar puncture should be performed in all cases.

The WHO advises 6 months of anti-TB chemotherapy unless TB meningitis.

Meningitis

This is commonest in children under 5 years, and often occurs within 6 months of infection. The onset is usually insidious, and the diagnosis is often delayed. Late diagnosis is invariably complicated by neurological dysfunction or death.

Prolonged fever, irritability, headache, vomiting, mental status changes, visual symptoms, focal neurological deficits or cranial nerve palsies, and seizure are some of the common presentations in children with tuberculous meningitis.

CSF: cell count is usually less than 500/mm³ and mainly lymphocytic, but polymorpho-neutrophils may be prominent early on, which may cause confusion with partially treated bacterial meningitis. Protein levels are usually raised (0.8–4 grams/litre) and glucose levels are low. However, on admission CSF values may be within normal limits and lumbar puncture must be repeated if there is any doubt.

Brain imaging, such as CT or MRI (if available), should be undertaken at diagnosis and at 3 - 4 months, and at any time when there is neurological deterioration, to detect complications such as hydrocephalus and tuberculomata.

Management

A four-drug regimen in the upper range of drug doses is recommended for 2 months, followed by a two-drug regimen for 10 months in uncomplicated tuberculosis meningitis. It consists of the following four drugs given for first 2 months:

1. H: isoniazid 10 (7-15) mg/kg once daily orally, or by IM or slow IV injection; (maximum 300 mg daily) plus
2. R: rifampicin 15 (10–20 mg/kg once daily orally or by IV infusion over 2–3 hours; (maximum 600 mg daily) plus
3. Z: pyrazinamide 35 (30-40 mg/kg once daily orally; (maximum 2 grams daily) plus
4. E: ethambutol 20 (15-25) mg/kg once daily orally (maximum 1.5 grams daily).

Thereafter, isoniazid plus rifampicin alone is continued for 10 months. The WHO also now advises 12 months of therapy, although shorter regimens have been shown to be adequate in some studies.

Corticosteroids must be given in all cases with initiation of therapy.

Dexamethasone 0.6 mg/kg/day in two divided doses or prednisolone 2 - 4 mg/kg/day is given for 4 weeks and tapered over 2 weeks for a total duration of 6 weeks.

A ventriculoperitoneal shunt may be required for obstructive hydrocephalus (if available).

Bone and joints

These are frequently missed in the early stages because of a low index of suspicion.

The spine is affected in 50% of the cases, followed by knee, hip and ankle. The most serious complication is spinal compression.

The diagnosis is made by histology, Ziehl–Neelsen (ZN) stain and mycobacterial culture of tissue that may be positive, and if in doubt specimens should be sent for polymerase chain reaction (PCR).

The WHO advises the standard 12 months of anti-TB chemotherapy, similar to that for TB meningitis.

Abdominal tuberculosis

This may present with ascites, abdominal nodes or masses, or diarrhoea with or without abdominal pain, or as gastrointestinal obstruction.

The diagnosis is usually made on bacteriological examination of ascitic fluid or a biopsy. The standard three- to four-drug regimen is used for therapy for a total of 6–9 months in uncomplicated cases.

Ultrasound and CT or MRI (if available) may be required in evaluation and to detect any complications.

Perinatal tuberculosis

Congenital tuberculosis is rare but should always be considered in sick neonates or 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 10 mg/kg once daily for 3 - 6 months.

Once the mother and infant are both on appropriate treatment, the infant may breastfeed unless the mother has multi-drug-resistant TB. A tuberculin test and chest X-ray is then performed on the infant.

If they are negative, BCG is given; if it is positive, full investigations for tuberculosis are undertaken.

If no evidence of disease is detected, isoniazid is continued for another 3 - 4 months. If tuberculosis is suspected, full treatment with 4 drugs is given at standard doses (see Table 51.2 and Table 51.3 on management).

Danger signs for TB

- Suspicion of tuberculous meningitis.
- Extensive pulmonary or miliary TB.
- TB in an infant or a child with HIV.
- Symptoms and signs such as seizures, coma, severe respiratory distress, gastrointestinal obstruction, or severe malnutrition.

Diagnosis of TB

Diagnosis depends on eliciting key points that may increase the yield of TB cases. A high index of suspicion in a child who has prolonged or unexplained illness should warrant investigation for TB.

Sputum or gastric aspirate for acid-fast bacilli (AFB) stain and culture should always be attempted. Standard methods for diagnosis are the tuberculin test and a chest X-ray. Even in resource-limited countries, every effort should be made to obtain a diagnostic specimen from gastric aspiration or sputum induction (see below). In poor communities the tuberculin test is often negative (or unavailable) and the chest X-ray might not be available, easy to interpret or have films of good enough quality.

Many children are often over-diagnosed, especially in areas with high HIV prevalence.

TB infection is diagnosed using the tuberculin skin test. It is considered positive if there is Mantoux induration of ≥ 10 mm in children

Interferon-gamma release assays (IGRA) detect latent and active infection but cannot differentiate between the two. They may be positive in some cases of HIV infection and malnutrition when the tuberculin test is negative, but in these circumstances, there is also a higher rate of false-negative IGRA results.

Key features suggestive of pulmonary TB

Three or more of the following should strongly suggest a diagnosis of TB:

- Chronic symptoms suggestive of TB (prolonged fever, cough, night sweats weight loss)
- Physical signs suggestive of TB (chronic lymphadenopathy, abdominal mass, gibbus or monoarthritis)
- A positive tuberculin skin test (induration > 10 mm)
- Chest X-ray suggestive of TB (hilar adenopathy, cavitation, pleural effusion, infiltrate; see below for pictures).

Investigations

1. Tuberculin test > 10 mm or > 5 mm in malnutrition or HIV.
2. Chest X-ray: lymphadenopathy, collapse/consolidation with or without persistent opacity, cavitation, miliary appearance.
3. Histology: lymph node or other tissue biopsy.
4. Smear/culture: gastric aspirate, induced sputum, nasopharyngeal aspirate, laryngeal swab, bronchoscopy, or body fluids.
5. Ultrasound: chest, abdomen, lymph nodes, pericardium and brain.
6. CT or MRI (if available).
7. HIV antibody tests.

Except in adolescents with cavitory disease, most tuberculosis in children is paucibacillary (low number of mycobacteria). Young children cannot expectorate.

Tuberculosis may be evident on chest X-ray, especially in older children.

Gastric aspiration should be undertaken in the early morning while the child is lying down. Ziehl–Neelsen (ZN) staining of gastric aspirate is positive in only about 10% of children with advanced pulmonary tuberculosis, and culture is positive, under optimal conditions, in only 30–50% of cases.

Alternative methods are sputum induction using nebulised 3% hypertonic saline, nasopharyngeal aspiration and laryngeal swabs. None of these has a sensitivity of more than

25–30%. Sputum induction requires a nebuliser and appropriate equipment and must be undertaken in a room with adequate ventilation.

The polymerase chain reaction (PCR) on histological specimens may be useful if the ZN stain is negative. In CSF it has similar sensitivity (around 50%) to culture. It is reserved for special cases where an urgent diagnosis is required.

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 for 7–10 days and the chest X-ray repeated after 2 weeks or so. If there is no improvement or deterioration, a full course of anti-tuberculosis chemotherapy may be given, and progress carefully monitored to document the response.

If the tuberculin test is negative initially it should be repeated after 3 months, when the patient's immune system has normalised, and it may become reactive at that time.

Increase in weight (measured daily or weekly) and loss of fever (measured twice daily) indicate a response to treatment. If treatment is given for suspected rather than proven tuberculosis, and no resolution or improvement in symptoms occurs within 4 weeks, this suggests that tuberculosis is unlikely. However, the course should still be completed, and an alternative diagnosis sought, such as drug-resistant tuberculosis, fungal infection or malignancy.

Xpert MTB/RIF test

The Xpert MTB/RIF is a test for rapid diagnosis of TB and drug-resistant TB. It is a TB-specific automated, cartridge-based nucleic amplification assay, and it detects *Mycobacterium tuberculosis*, as well as mutations conferring resistance to rifampicin, directly from sputum in an assay that provides results within 100 minutes.

Results from field demonstration studies found that a single Xpert MTB/RIF test can detect TB in 99% of patients with smear-positive pulmonary TB and more than 80% of patients with smear-negative pulmonary TB. The co-existence of HIV does not significantly affect the performance of Xpert MTB/RIF.

Furthermore, Xpert MTB/RIF can detect rifampicin resistance with 95.1% sensitivity and exclude resistance with 98.4% specificity. The WHO endorsed the Xpert MTB/RIF assay in December 2010. It should be used as the initial test in individuals with suspected multi-drug-resistant TB (MDR-TB) or HIV/TB. It may be used as a follow-on test to microscopy where MDR-TB and/or HIV is of lesser concern, especially in smear-negative specimens.

It is effective in children where sputum may need to be obtained by induction via nasopharyngeal aspirate after salbutamol and then saline nebuliser.

Management of TB in children

Except for CNS and osteo-articular disease (see below), both pulmonary and extra-pulmonary tuberculosis may be treated with standard 6-month chemotherapy.

The standard treatment regimen for all patients with drug-susceptible, uncomplicated TB is made up of an intensive phase lasting 2 months and a continuation phase lasting 4 months.

During the intensive phase 4 drugs (isoniazid, rifampicin, pyrazinamide, and ethambutol) are used to rapidly kill the tubercle bacilli. Infectious patients become less infectious within approximately 10 - 14 days of starting treatment and symptoms abate. In the continuation phase, 2 drugs (isoniazid, rifampicin) are used, over a period of 4 months.

For non-HIV-infected children with a low risk of isoniazid resistance, ethambutol can be omitted. Ethambutol should not be given in a dose higher than 20 mg/kg/day to children under 5 years, as they may be unable to report visual disturbance associated with optic neuritis.

TABLE 51.2 Regimens for treatment of uncomplicated susceptible pulmonary tuberculosis

Regimen	Total Duration
Standard daily	
Isoniazid, rifampicin, pyrazinamide ethambutol* for 2 months, then isoniazid, rifampicin for 4 months†	6 months

* In HIV-uninfected children with a low risk of isoniazid resistance, ethambutol can be omitted.

† For central nervous system and osteo-articular disease, the continuation phase should be 10 months (total duration 12 months).

- Thiacetazone is no longer used as a first-line drug. **Thiacetazone may cause severe reactions in HIV-infected patients.**
- Presently DOTS (directly observed treatment, short course) is not generally practised for children, as it is assumed that parents will supervise treatment, but where DOTS is practised in the community it may be appropriate to include children.
- *WHO Recommendation:* In all patients with drug-susceptible pulmonary TB, the use of thrice-weekly dosing is not recommended in both the intensive and continuation phases of therapy, and daily dosing remains the recommended dosing frequency (Conditional recommendation, very low certainty in the evidence).
[WHO | Guidelines for treatment of drug-susceptible tuberculosis and patient care \(2017 update\)](#) (accessed 04/03/2021)
- Our advice is that in low resource settings, all children with TB should be treated with daily regimes (for dosage see Table 51.3).
- For central nervous system and osteo-articular disease, the continuation phase should be 10 months (total duration 12 months).
- 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 (e.g. nausea, vomiting, malaise or liver tenderness).
 - Viral hepatitis (especially hepatitis A) should be considered if jaundice occurs. Adjunct treatment with corticosteroids in meningitis is indicated at initiation of therapy (see above) and may enhance resolution of disease in lympho-bronchial disease, pericarditis, pleural effusion and severe miliary disease with alveolar capillary block.
- Prednisolone 1.5–2.0 mg/kg/day is given for 2–3 weeks and then tailed off over 2 weeks (see treatment of meningitis).

Follow-up

All children who are started on anti-tuberculous therapy must be followed closely, preferably every month. Clinical, radiologic and mycobacteriologic improvement and adverse effects of drugs must be monitored.

In children, weight gain and resolution of signs and symptoms are indicators of a good response to treatment. Routine laboratory tests such as liver function tests and X-rays are rarely needed in children.

Those children with severe disease, poor response, unusual presentations or suspected resistant TB must be referred to an expert.

TABLE 51.3 Daily dosage schedule for anti-tuberculous drugs and side effects

Drug	Children: once daily dose	Children: three times weekly dose given once daily	Side effects
Isoniazid daily	10 mg/kg range 7–15 mg/kg. Max. 300mg.	20–40 mg/kg Max. 900 mg	Hepatic enzyme elevation, hepatitis, peripheral neuropathy, hypersensitivity
Rifampicin	15 mg/kg range 10–20 mg/kg. Max. 600mg.	10–20 mg/kg Max. 600 mg	Orange discoloration of urine and secretions (and contact lenses), nausea, vomiting, hepatitis, febrile reactions, thrombocytopenia
Pyrazinamide	35 mg/kg range 30–40 mg/kg. max 2 g	50–70 mg/kg Max. 3 grams	Hepatotoxicity, hyperuricaemia, gastrointestinal upset, arthralgia, skin rash
Ethambutol	20 mg/kg range 15–25 mg/kg. max 2.5 g	25–30 mg/ kg	Optic neuritis, skin rash

Higher range of isoniazid applies to young children and is recommended by WHO 2010. Use mean dosage and round up rather than round down when prescribing except when prescribing ethambutol.

Fixed dose combinations

The fixed dose combinations come from the **World Health Organization (2017) [Statement on the use of child-friendly fixed-dose combinations for the treatment of TB in children](#)** (accessed 04/03/2021)

Table 51.4 Dispersible tablets:

RHZ 75/50/150

RH 75/50

Weight kg	Number of tablets	
	Intensive phase: RHZ 75/50/150	Continuation phase: RH 75/50
4-7	1	1
8-11	2	2
12-15	3	3
16-24	4	4
>25	Adult dosage	Adult dosage

Ethambutol should be added in the intensive phase for children with extensive disease or living in settings where the prevalence of HIV or of isoniazid resistance is high.

Isoniazid resistant TB

In patients with confirmed rifampicin-susceptible and isoniazid-resistant tuberculosis, treatment with rifampicin, ethambutol, pyrazinamide and levofloxacin is recommended for a duration of 6 months

Multi-drug-resistant TB (MDR-TB) and extreme-drug-resistant TB (XDR-TB)

- Rapid drug susceptibility testing of isoniazid and rifampicin should be done at the time of diagnosis (if available).
- After treatment is started for MDR-TB, further sputum (induced or gastric aspirate) should be obtained monthly to ensure successful treatment.
- An expert in the management of paediatric TB must be involved in choosing the optimal regimen for a child with drug-resistant TB.
- Fluoroquinolones may be used in treatment of MDR-TB in children.
- Theoretical concerns about cartilage damage from early trials in young dogs have not been evident in children, and these are far outweighed by the benefits in treatment of TB.
- Later-generation fluoroquinolones (see below) are more effective than earlier ones.
- In children with HIV and MDR-TB, antiretrovirals should be started as soon as possible following initiation of anti-tuberculous therapy, irrespective of CD4 count.
- The preferred regimen is zidovudine, lamivudine and efavirenz, but if already on antiretrovirals, continue the same regimen. Co-trimoxazole should be added for pneumocystis prophylaxis.
- Treatment should be ambulatory rather than in hospital as much as possible.

Groups of second-line anti-tuberculosis agents

Second-line parenteral agent (injectable anti-tuberculosis drugs)

Kanamycin (Km): 15–30 mg/kg/day (maximum 1000 mg).

Capreomycin (Cm): 15–30 mg/kg/day (maximum 1000 mg).

Streptomycin (S) : 15–20 mg/kg/day (maximum 1000 mg).

Fluoroquinolones

Levofloxacin (Lfx): 15–25 mg/kg/day (maximum 1000 mg).

Moxifloxacin (Mfx): 7.5–10 mg/kg/day (maximum 400 mg).

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Ofloxacin (Ofx): 15–20 mg/kg/day (maximum 800 mg).

Oral bacteriostatic second-line anti-tuberculosis drugs

Ethionamide (Eto): 15–20 mg/kg/day (maximum 1000 mg).

Cycloserine (Cs): 10–20 mg/kg/day (maximum 1000 mg).

Terizidone (Trd), 10–20 mg/kg/day (maximum 1000 mg).

p-aminosalicylic acid (PAS), 150 mg/kg/day (maximum 8 grams (PASER)).

For regimes see: [WHO consolidated guidelines on drug-resistant tuberculosis treatment](#)
Accessed 8th March 2021

Prevention of TB

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 (10 mg/kg/ day, maximum dose 300 microgram) alone for 6 months or in high-incidence countries, isoniazid and rifampicin for 3 months.

Rifapentine and isoniazid weekly for 3 months may be offered as an alternative to 6 months of isoniazid monotherapy as preventive treatment for both adults and children in countries with a high TB incidence.

If the contact is isoniazid resistant, rifampicin for 3-4 months can be used alone.

HIV-infected infants and children exposed to tuberculosis infection but without active disease should receive isoniazid prophylaxis as described above. The WHO advises that HIV-infected children over 1 year old who may have tuberculosis, even in the absence of exposure to tuberculosis, should receive a routine course of isoniazid for 6 months. There must be facilities for investigation of tuberculosis and regular follow-up.

Neonatal BCG may reduce the risk of tuberculosis meningitis and disseminated disease by 60–80%, especially in children under 5 years of age. However, it has a limited efficacy against pulmonary disease. Because of the increased risk of disseminated BCG infection in HIV-infected infants, the WHO advises that all infants known to be HIV-infected should not receive BCG. However, this has practical implications in resource-limited countries where PCR is not usually available to detect HIV infection in infants under 18 months of age. BCG should be given to neonates born to mothers with HIV.

Further reading

World Health Organization (2020) Global tuberculosis report 2020

World Health Organization (2018) Latent tuberculosis infection: updated and consolidated guidelines for programmatic management. <https://www.who.int/tb/publications/2018/latent-tuberculosis-infection/en/>

Accessed 8th March 2021

World Health Organization (2017) [Statement on the use of child-friendly fixed-dose combinations for the treatment of TB in children](#) (Accessed 04/03/2021)

World Health Organization (2020) Consolidated guidelines on tuberculosis. Module 4: treatment - drug-resistant tuberculosis treatment.

<https://www.who.int/publications/i/item/9789240007048> Accessed 8th March 2021

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World Health Organization (2014) Guidance for National Tuberculosis Programmes on the management of Tuberculosis in Children, Second edition.

www.who.int/tb/publications/childtb_guidelines/en/ (Accessed_04/03/2021)

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Section 52. Helminth Infections: Worms

Introduction

In low-resource countries, children presenting to medical facilities may harbour intestinal helminthiasis (worms) or their juvenile forms (larvae) in other organs. Often this situation may exist without the presence of any signs or symptoms. In such situations, ill health or the risk of serious complications is directly related to the number of parasites in a child; although children bearing heavy loads of parasites are in a minority. These patients will often present for other reasons, without their heavy worm infections being recognised.

Although mass deworming has been widely implemented to reduce the burden of helminthic infection and disease, recent major systematic reviews have concluded that there is little or no evidence that mass deworming improves child health outcomes, school performance, or cognitive development.

Parasitology of worms

There are several important groups of helminth infections:

Cestodes:

Beef tapeworm (*Taenia saginata*) and pig tapeworm (*Taenia solium*) (*Taeniasis/cysticercosis*) See Section 32)

Nematodes:

Roundworms (*Ascaris* species)

- *Ascaris lumbricoides* is the commonest human roundworm.
- Adult worms are whitish pink, several millimetres (mm) wide and up to 30 centimetres (cm) long, which may live for years in the small intestine.
- They are often seen in stool or vomit.
- Transmission is by ingestion of embryonated eggs from soil.
- Adult worms shed their eggs in faeces.
- *Ascaris* pneumonitis accompanied by eosinophilia, known as Löffler's syndrome, is associated with migration of larvae through the lungs. Symptoms usually resolve within 10 days.
- Most established infections are asymptomatic.
- The ill effects of adult ascaris include lactose intolerance and malabsorption of vitamins and micronutrients. Heavy infections may cause intestinal obstruction, volvulus or perforation and peritonitis. Obstruction of ducts may also occur.

Hookworms (Ancylostoma duodenale, Necator americanus)

- Both adult forms are hair-like, about 1 cm long, with cutting plates at the mouth end. *Ancylostoma* is generally the more virulent pathogen.
- Both species occur widely.
- They have invasive larvae. Skin penetration is the commonest mode of infection. Less commonly, ingestion of embryonated eggs may give rise to infection.
- Sustained blood loss in the small intestine leads to an accumulating risk of anaemia and hypo-albuminaemia.

Whipworms (Trichurius species)

- *Trichuris trichiura* commonly known as the whipworm can be up to 4 cm long with thickness of a hair except at the tail end which is wider.
- Transmission is similar to *Ascaris*, but maturation occurs only in the gut without tissue invasion beyond the mucosa.
- Most infections are asymptomatic.
- Heavy infestations may resemble inflammatory bowel disease.
- Severe trichuris dysentery syndrome often leads to rectal prolapse.

Threadworms (Enterobius species).

- *Enterobius vermicularis* is an intestinal helminth which is spread in the form of embryonated eggs by personal contact among children and within families.
- It is largely harmless but can cause intense nocturnal perianal itching and, sometimes secondary infection.

Strongyloides stercoralis

- See Section 48.

Trematodes or flukes, which include blood flukes (e.g. schistosomiasis) (see Section 45) and biliary tract, lung and gut flukes.

Investigations

Stool microscopy for eggs is usually adequate for diagnosing established infection.

- The Kato-Katz concentration technique can also be used.
- FLOTAC, a recently developed method for detecting helminth ova by centrifugation of stool samples in a flotation solution, is more sensitive than routine microscopy or Kato-Katz and is also useful for diagnosing intestinal protozoal infections.
- Multiplex PCR assays are now available, which simultaneously detect hookworm, *ascaris lumbricoides*, *strongyloides* and *trichuris*.

Treatment

1. The broad-spectrum anti-helminthic's, 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 enterobiasis.
2. Ivermectin is recommended for strongyloidiasis (See Section 48 Handbook 2).

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 reasonably palatable. The 500 mg tablet is useful for mass campaigns against *Trichuris* or hookworm. It is not approved for use in children under 2 years of age, but clinical judgement should be used in a symptomatic child. It is considered unsafe in pregnancy or lactation.

Threadworms and pinworms

Oral dose:

- Children from 6 months up to 10 kg body weight:
 - Give 50 mg as a single dose; repeat after 2 weeks.
- Children over 1 year of age or more than 10 kg body weight:
 - Give 100 mg as a single dose; repeat after 2 weeks.

Whipworms, roundworms and hookworms

Oral dose:

- Children from 6 months up to 10 kg body weight:
 - Give 50 mg twice daily for 3 days.
- Children over 1 year of age or more than 10 kg body weight:
 - Give 100 mg twice daily for 3 days.

Albendazole

This drug is closely related to mebendazole, with similar pharmacokinetics. It has superior efficacy to mebendazole in systemically invasive conditions and is more effective against

migrating larvae. It is available as 200 mg tablets or 200 mg/5 mL liquid. Cautions are as for mebendazole, noting its greater systemic absorption.

Hookworms, roundworms, pinworms, threadworms, whipworms

Oral dose:

- Children aged 12 months to 2 years:
 - Give 200 mg as a single dose.
- Children over 2 years or 10 kg:
 - Give 400 mg as a single dose before food. Treatment may be repeated in 3 weeks.
- Severe infections may require initial treatment for three days.

[World Health Organization \(2020\) 2030 targets for soil-transmitted helminthiases control programmes \(accessed 04/03/2021\)](#)

Section 53. Yaws

Introduction

Yaws is caused by the bacterium *Treponema pallidum*; sub-species *pertenue*. It is closely related to the bacterium that causes syphilis, but this disease is not sexually transmitted.

Yaws mainly affects children in rural tropical areas, such as the Caribbean Islands, Latin America, West Africa, India, and South-East Asia. Yaws is transmitted by direct contact with the skin sores of infected people.

Symptoms

About 2 - 4 weeks after infection, the child develops a sore called a 'mother yaw' where bacteria entered the skin.

- The sore is reddish and looks like a berry. It is usually painless but does cause itching.
- These sores may last for months.
- More sores may appear shortly before or after the mother yaw heals as the person scratches or spreads the bacteria from the mother yaw to uninfected skin. Eventually the skin sores heal.
- Some patients develop destructive ulcerations of the nasopharynx, palate, and nose (termed gangosa), painful skeletal deformities, especially in the legs (termed saber shins), and other soft-tissue changes (gummas, inflammatory cell infiltration).
- In the advanced stage, sores on the skin and bones can lead to severe disfigurement and disability.

Signs and tests

A sample from a skin sore is examined using a dark-field microscope. There is no blood test for yaws. However, the blood test for syphilis is usually positive in children with yaws because the bacteria that cause these two conditions are closely related.

Treatment

Recently a single dose of oral azithromycin (30 mg/kg) has been shown to be as effective as a single IM injection of benzathine benzylpenicillin 50 000 units/kg (37.5 mg/kg) with less risk of a dangerous anaphylactic response and the need for needles.

If the child vomits within 30 minutes of the oral dose of azithromycin, a repeat dose should be given. Benzathine benzylpenicillin must not be given IV.

Anyone who lives in the same house with someone who is infected should be examined for yaws and treated if they are infected. Skin lesions may take several months to heal. By its late stage, yaws may have already caused damage to the skin and bones. It may not be fully reversible, even with treatment for the infection.

Note: *H. ducreyi* has also been shown to be a cause of yaws-like lesions in both Africa and the Western Pacific. Azithromycin, as above, can be effective in treating this bacterial infection.

[World Health Organization Health Topics Yaws](#) (accessed 04/03/2021)

Section 54. Blood transfusion

Introduction

Blood or blood products should be transfused only when needed to save life or to prevent major morbidity.

- The risk of transmission of infection is a major concern in countries with limited resources and poorly organised blood transfusion services.
- Blood must be stored safely, or a bank of adequately screened donors must be available 24 hours a day, especially for obstetric emergencies or major trauma.
- In emergency situations, relatives accompanying patients are often asked to donate blood if compatible. Unlike stored blood it is warm and contains active clotting factors.
- When giving a blood transfusion, care must be taken to ensure that the blood is compatible with that of the recipient, is infection free and the transfusion is monitored by someone who is able to recognize any adverse reactions.

Clinical situations that require blood transfusion

Normal haemoglobin (Hb) levels (after the neonatal period) are around 129 g/L (129 g/L). Children with severe anaemia have Hb levels of 50 g/L or less. An Hb level of 50 g/L is widely accepted as the level at which transfusion might be indicated, and less than 40 g/L if there is severe malnutrition. *Note:* g/L divided by 10 = g/dL.

The WHO defines anaemia as any Hb level below 110 g/L. However, in children who are pregnant normal haemodilution means that a cut-off value of less than 100 g/L is more appropriate. In children who are pregnant, transfusion may be considered at an Hb level of 60–70 g/L. However, Hb concentration should not be the only factor when deciding to transfuse.

In addition to Hb level, the following factors must be considered:

1. Heart rate. If it is rapid, this will support the decision to transfuse.
2. Respiration rate. If it is rapid, this will support the decision to transfuse.
3. Is the patient already in circulatory collapse (shock) due to bleeding or is there visible massive haemorrhage? If so **the need for transfusion is very urgent.**

Some patients will not show any of these features, and it might then be justifiable to use haematinics (i.e., iron and folic acid). Some patients may show the above features and have an Hb level higher than 50 g/L. It will also be necessary to transfuse patients if their symptoms are caused, or significantly worsened, by anaemia (e.g., heart failure).

Causes of anaemia in neonates and young infants

- Hypovolaemic shock can result from acute blood loss, as for example in premature separation of the placenta or fetomaternal haemorrhage, twin-to-twin transfusion, and other causes of fetal or neonatal haemorrhage.
- Neonates may lose a considerable blood volume because of sampling for laboratory tests. Therefore, samples should be minimised.
- Reduce the need for transfusion in neonates by providing adequate antenatal care, to reduce the risks of premature delivery and when possible, prevent nutritional anaemia in the mother.
- Encourage breastfeeding.
- Ensure that there is early provision of vitamin K prophylaxis, iron, vitamins, and other haematinics, especially in premature babies.

After birth, the haemoglobin level drops to less than 100 g/L in term infants at 8–12 weeks of age, but in premature infants it can drop to 70–100 g/L even earlier, at 6 weeks. (Oxygen delivery is well maintained because of rising levels of haemoglobin A, which releases oxygen more freely than haemoglobin F, which is found in the fetus.)

Causes of anaemia in older infants and children

These include the following:

- surgery
- haematological malignancies
- malaria
- sickle-cell disease
- congenital haemolytic anaemias (thalassaemia, glucose- 6-phosphate dehydrogenase deficiency)
- burns
- major trauma.
- malnutrition (see Section 56 Handbook 1).

Causes of anaemia in the child who is pregnant

These include the following:

- obstetric emergencies such as antepartum and post- partum haemorrhage
- severe anaemia that is untreated or unresponsive to haematinics
- major trauma.

Transfusion policies and guidelines

In hypovolaemic shock due to haemorrhage, always give a blood transfusion as rapidly as possible.

In other causes of hypovolaemic shock crystalloid infusions (R/L or 0.9% saline) or 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 anaemia-induced heart failure.
- When possible, provide malaria prophylaxis, particularly in pregnant children (see Sections 31 Handbook 1). Early treatment of clinical malaria reduces the profound haemolysis that is a major reason for transfusion in endemic areas.

Anaemia due to malaria responds to treatment with antimalarial drugs and folic acid.

In situations **where blood transfusion is unavailable or potentially unsafe**, the following recommendations have been made:

- Transfusion is not necessary if the Hb level is more than 50 g/L. and there is no evidence of heart failure
- Transfusion may be necessary if the Hb level is more than 50 g/L and there is incipient cardiorespiratory distress (air hunger, hypotension, tachycardia, and oedema).
- Transfusion may be necessary if the Hb level is less than 40 g/L and complicated by malaria or bacterial infection, even without incipient cardiac failure.
- Transfusion may be necessary if the Hb level is less than 30 g/L, with no apparent complications.

In situations **where blood transfusion is safe and available**, recommendations for its use are as follows.

Neonates and infants less than 4 months old

- Blood loss of more than 15% over 2 days.
- Haemoglobin level of less than 70 g/L with clinical manifestations of anaemia.

Infants and children aged 4 months or older

- Acute blood loss that is unresponsive to crystalloid and colloid infusions.
- Intra-operative blood loss of more than 15% of total blood volume and post-operative haemoglobin level of less than 80 g/L with clinical symptoms.
- Haemoglobin level of less than 110 g/L with severe pulmonary disease.
- Acute haemolysis with haemoglobin level of less than 80 g/L with signs of anaemia.
- When possible, provide malaria prophylaxis, particularly in young children and in those who are pregnant. Early treatment of clinical malaria reduces the profound haemolysis that is a major reason for transfusion in endemic areas. Anaemia due to malaria responds to treatment with antimalarial drugs and folic acid.

Blood transfusion is not required for sickle-cell disease in the steady state. It may be indicated in severe anaemia with incipient or established cardiac failure, acute splenic enlargement, sequestration crisis with rapidly falling haemoglobin levels, aplastic crisis, acute chest syndrome, stroke, and sometimes as exchange transfusion for severe priapism (see Section 26 on sickle-cell disease).

National programmes for thalassaemia (see Section 21) and other congenital haemolytic disorders, such as glucose-6-phosphate dehydrogenase deficiency, help to reduce transfusion requirements.

In situations where blood transfusion is safe and available, recommendations for its use are as follows.

Red-cell-free components

- Fresh frozen plasma (FFP) is only recommended when a specific blood clotting defect has been identified. In the absence of specific testing, consider administering FFP to a patient with signs of disseminated intravascular coagulation who is acutely unwell, as it may be lifesaving.
- Freeze-dried plasma is available, and its advantages include a long shelf life and the lack of need for refrigeration.
- Platelets are prepared from fresh blood using a special, simple centrifugation method, and the remaining blood can be given back to the donor. Once extracted by this method, platelets can last for up to 5 days at room temperature (around 23°C). Platelets should not be stored in a refrigerator. Transfused platelets survive only briefly in the body, and repeated infusion may be required for active bleeding, or before essential procedures such as a lumbar puncture.

Blood donation and provision

1. Most transfusions are required and given as an emergency procedure.
2. In low resource situations where fresh frozen plasma or platelets are never available, **fresh blood transfusion from a live donor is the most effective.**
3. Ideally blood for storage is obtained by routine whole blood collection from an established panel of blood donors with quality standards for testing, processing and distribution.
4. Safe storage of blood for transfusion is enhanced by the following measures:
 - collection of blood from repeat regular donors screened using a standard health-check questionnaire, and who are found negative for all markers for transfusion-transmissible infection
 - collection in a multi-pack 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 pre-transfusion tests.
5. Group O rhesus-negative packs facilitate transfusion across the ABO barrier. They must be checked for high-titre anti-A or anti-B by a suitable antiglobulin method.
 6. Establish a routine procedure for collection, testing and processing which should cover routine and emergency transfusions.
 7. In an emergency where a newborn infant is shocked, take a blood sample from the baby for blood grouping and cross-matching then take a blood sample of 30ml directly from the mother's vein and infuse 10mL/Kg IV into the baby over 5 minutes. Alternatively, if available, give 10ml/Kg of virus screened O Rh-negative blood. Fresh maternal blood is best because it is warm and contains clotting factors and does not require testing for viruses.

Pre-transfusion testing

Minimum acceptable tests on blood prior to transfusion

1. ABO and Rhesus D grouping.
 2. Screening for hepatitis B antigen, antibodies to HIV-1 and -2, hepatitis C virus antigen and syphilis.
 3. 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.

The inclusion of control A, B, O, Rh D-positive and Rh D-negative cells in the procedure is part of good laboratory practice and should be part of the testing method.

- If possible, two methods should be used for grouping, to ensure reliability
- The most suitable method for compatibility is the anti-human globulin technique at 37°C for 1 hour. Agglutination should be read before and after the addition of the anti-human globulin reagent.

Blood groups

There are four major blood groups: A, B, AB and O. To avoid ABO incompatibility, the blood group of both the donor and the receiver must be known. Blood can only be donated in the direction of the arrows shown in Figure 54.1.

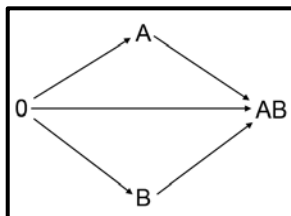


Figure 54.1 Safe transfusion of ABO blood groups.

For ABO typing:

- 1 Donors with blood group O can donate to patients (receivers) with blood group A, B, AB or O.
- 2 Donors with blood group A can donate to patients with blood group A or AB.
- 3 Donors with blood group B can donate to patients with blood group B or AB.
- 4 Donors with blood group AB can donate only to patients with blood group AB.

For Rhesus typing

Blood is also categorised according to its rhesus status. Therefore:

- Rhesus-negative donors can give to Rhesus-positive and Rhesus-negative patients
- Rhesus-positive donors can only give to Rhesus-positive patients

Persons with blood group O Rhesus negative are known as universal donors that is they can donate to people with any ABO and Rhesus blood group

Persons with blood group AB Rhesus positive are known as the universal recipient and can receive blood from people with any blood group

If the blood group is unknown and blood is required before a crossmatch can be performed, give O-Rhesus-negative blood if this is available.

The importance of live donor fresh blood in low resource settings

Fresh blood contains platelets and clotting factors and is particularly valuable in most emergency settings where blood transfusion is needed. This is especially the case with acute severe haemorrhage and DIC.

When undertaking resuscitation, Control bleeding ABC (CABC), potential live donors should be identified as soon as possible.

Exchange transfusion

This is used for haemolytic disease of the newborn with severe anaemia and/or severe hyperbilirubinaemia (see Neonatal Handbook for details). 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 are prepared by removing approximately 100 mL of plasma to create a haematocrit of 0.5–0.6.

Patients with sickle-cell anaemia and acute chest syndrome or impending cerebrovascular episodes may benefit from exchange transfusion (see Section 26).

Blood should be fresh (less than 5 days old) and screened for HbS if it is issued for sickle-cell disease.

Bedside transfusion

A child's body contains 80 mL of blood for every kg of body weight. For example, a 3-year-old weighing 12 kg will have 960 mL of blood in their body.

A pregnant child's body contains 100 mL of blood for every kg of body weight.

Venous access for bedside transfusion should be chosen with no smaller than a 22- to 24-gauge vascular catheter, and a much larger one in older and pregnant children.



Figure 54.2 Drip counter infusion monitor (SN-1500H from Sino MDT Ltd)

Blood is usually cleaned and filtered in the lab, so when transfusing it to a patient the only filter that needs to be used is the usual on-line filter in a standard giving set (170 – 200 microns).

Blood should be given using an accurate measurement of rate and time. **Close observation of the patient is needed during transfusion, especially in the first 15-30 minutes in case of transfusion reactions. Ideally use an infusion droplet monitor (see Figure 54.2).**

Always check the suitability of the IV giving set for giving a blood transfusion.

Blood transfusion for severe anaemia where there is heart failure

1. Give a high concentration of oxygen, bed rest and sit the patient upright
2. Consider transfusion with packed cells if the haemoglobin concentration is less than 50g/L (with IV furosemide of 40mg for each unit of packed cells). If blood cannot be centrifuged, let the bag hang until the cells have settled. Infuse the cells slowly and dispose of the remaining serum.
3. Partial exchange transfusion may be helpful. Use a cannula in a large vein in the antecubital fossa, withdraw 20 mL of the patient's anaemic blood and infuse 40 mL of new blood (ideally packed red blood cells) over 5 minutes and repeat 5–10 times.

Blood transfusion reactions

Blood transfusion can be lifesaving and provides great clinical benefit to many patients, but, like any treatment potential benefit should outweigh risks, which include the following:

- immunological complications
- errors and 'wrong blood' episodes
- infections (bacterial and viral).

Causes and management of acute complications of transfusion

1. Acute haemolytic transfusion reaction

- Due to ABO incompatibility. The patient has anti-A or anti-B antibodies that destroy incompatible infused red blood cells. This may lead to disseminated intra-vascular coagulation (DIC) and acute renal failure.
- Infusion of ABO-incompatible blood is almost always a result of errors in labelling sample tubes and/or request forms, or inadequate checks at the time of transfusion. When red cells are mistakenly administered, there is about a 1 in 3 risk of ABO incompatibility and a 10% risk of mortality, with the most severe reaction seen in a group O individual receiving group A red cells.
- Non-ABO red cell antibody haemolytic reactions (for example those involving anti-rhesus Rh D alloantibodies) tend to be less severe.

Management

1. Stop blood, replace giving set, and keep IV open with 0.9% saline
2. Insert bladder catheter and monitor urine output
3. Give fluids to maintain urine output >30ml per hour
4. If urine output <30 ml per hour despite fluid challenges, give frusemide
5. Check compatibility label of blood corresponds with patient ID
6. Inform lab staff of reaction
7. Take blood for blood cultures – alternative diagnosis may be an infected blood unit.

2. Infective shock

- Bacterial contamination of blood can be fatal.
- Rapid onset of sepsis tachycardia, low pulse pressure, hypotension, rigors, and collapse follows the transfusion.
- Usually occurs during transfusion of first 100ml of blood

Management

- Stop transfusion and change giving set
- Treat sepsis. Give IV fluids and IV antibiotics – e.g., ceftriaxone plus gentamicin

3. Transfusion-related acute lung injury (TRALI)

- TRALI is a form of acute respiratory distress due to donor plasma containing antibodies against the patient's leucocytes. The donor is usually a multiparous woman.
- During, or soon after infusion, a non-productive cough, breathlessness, hypoxia and frothy sputum develops. Fever and rigors may be present.

- Chest X-ray, if available, shows multiple perihilar nodules with infiltration of the lower lung fields.

Management

- High flow oxygen
- **Do not give diuretics which worsen the situation**

4. Fluid overload

- This occurs when too much blood is transfused or is transfused too quickly, leading to pulmonary oedema and acute respiratory failure.
- Hypoxaemia, tachypnoea, raised jugular venous pressure, enlarged liver and basal pulmonary crepitations are signs of resulting pulmonary oedema
- Patients at particularly high risk are those with severe or chronic anaemia, or severe malnutrition, and who have symptoms of cardiac failure or normal blood volumes (i.e. who are not bleeding) prior to transfusion.

Patients at risk should receive packed cells rather than whole blood via slow transfusion, with diuretics if required.

Management (see Section 42 Handbook 1)

- **Sit patient upright**
- Call for an anaesthetist
- Do not give any more IV fluid until fluid overload has gone
- Give **Furosemide** as follows:

Neonate: 0.5 to 1 mg/kg by slow IV injection

Child 1 month to 11 years: 0.5 to 1 mg/kg by slow IV injection (max per dose 40 mg) and repeat dose of 0.5 to 1mg/Kg if inadequate response (max per dose 40 mg)

Child 12-17 years: 20 to 40 mg by slow IV injection and repeat if needed.

- Consider **morphine** (try and ensure anaesthetist is present) as follows:

Child 6 months to 11 years: 100 microgram/kg IV over at least 5 minutes

Child 12 to 17 years: 5 mg IV over at least 5 minutes

5. Non-haemolytic febrile reactions to transfusion of platelets and red cells

- Fevers (more than 1.5°C above baseline) and rigors may develop during transfusion due to the patient's antibodies to transfused white cells.
- This type of reaction affects 1–2% of patients.
- Children who have received multiple previous transfusions are most at risk. Such reactions are unpleasant but not life-threatening. Usually, symptoms develop towards the end of a transfusion or in the subsequent 2 hours.

Management

Most febrile reactions can be managed by slowing or stopping the transfusion and giving paracetamol.

5. Allergic reaction or anaphylaxis

Allergic reactions occur when patients have antibodies that react with proteins in transfused blood components. Anaphylaxis occurs when an individual has previously been sensitised to an allergen present in a blood transfusion, and subsequently, on re-exposure, releases immunoglobulin IgE or IgG antibodies.

Anaphylaxis See Section 36 Handbook 1

Presentation

Symptoms or signs may occur after only 5–10 mL of transfusion of incompatible blood, so patients should be observed very closely at the start of each blood unit transfused.

Symptoms

These include the following:

- a feeling of apprehension or that 'something is wrong'
- flushing
- chills
- pain at the venepuncture site
- muscle aches
- nausea
- pain in the abdomen, loins or chest
- shortness of breath.

Signs

These include the following:

- fever (a rise in temperature of 1.5°C or more) and rigors
- urticaria
- hypotension or hypertension
- tachycardia
- flushing and swelling of the face
- respiratory distress including wheeze or stridor
- oozing from wounds or puncture sites
- haemoglobinaemia (outside red corpuscles in the plasma)
- haemoglobinuria (in the urine)

For treatment of **anaphylaxis**, see Section 36 Handbook 1. **Adrenaline intramuscularly is the key.**

SUMMARY: Management of an acute transfusion reaction

1. Where the only feature is a rise in temperature of less than 1.5°C from baseline, or urticaria, recheck that the correct blood is being transfused, give paracetamol and antihistamine, reset the transfusion at a slower rate and observe the patient more frequently.
2. Although fever or rigors are not uncommon in response to a transfusion and may represent a non-haemolytic febrile reaction, they may also be the first sign of a severe adverse reaction.
3. Where the reaction is more severe:
Stop the transfusion and call a doctor/senior clinician urgently to review the patient. Vital signs (temperature, blood pressure, pulse, respiratory rate and oxygen saturation levels) and respiratory status (dyspnoea, tachypnoea, wheeze and cyanosis) should be checked and recorded. Look for signs of heart failure (basal lung crepitations and enlarged liver).
Check the patient's identity and recheck against details on the blood unit and compatibility label or tag.
4. Initial management if ABO incompatibility is suspected is as follows:
 - Take down the blood bag and the giving set with blood in it.
 - Keep the IV line open with Ringer-lactate /Hartmann's solution or 0.9% saline.
 - Give oxygen and fluid support.
 - Monitor urine output, usually following catheterisation, and maintain it at more > 30 mL/hour in pregnancy, giving Furosemide if it falls below this.
 - Consider inotropic support if hypotension is prolonged.
 - Treat DIC by giving fresh donor blood fully matched to the recipient.
 - Inform the hospital transfusion department immediately.

5. If another haemolytic reaction or bacterial infection of blood unit is suspected:
 - Send haematological and microbiological samples for investigations outlined above.
 - General supportive management is as for ABO incompatibility.
 - Start broad-spectrum IV antibiotics if bacterial infection is considered likely.
6. If anaphylaxis or severe allergic reaction is suspected: follow the anaphylaxis protocols (see Section 36 Handbook 1)
7. If TRALI is suspected:
 - Give high-concentration oxygen, IV fluids and inotropes (as for acute respiratory distress syndrome).
 - Assisted ventilation may be urgently required; discuss this with an anaesthetist.
 - TRALI improves within 2–4 days in over 80% of cases if there is adequate management and respiratory support.
8. If fluid overload is suspected:
 - sit the patient upright
 - stop blood transfusion and do not give any IV fluids
 - give IV furosemide (see doses above)
 - give a high concentration of oxygen
 - if anaesthetist is present give morphine (see doses above)

Delayed complications of transfusion

Delayed haemolysis of transfused red cells

In those who have previously been immunised to a red cell antigen during pregnancy or by transfusion, the level of antibody to the blood group antigen may be so low as to be undetectable in the pre-transfusion sample.

However, after transfusion of red cells bearing that antigen, a rapid secondary immune response raises the antibody level dramatically, leading to the rapid destruction of transfused cells.

At 5–10 days post-transfusion, patients present with fever, falling haemoglobin levels (or an unexpectedly poor rise in haemoglobin levels), jaundice and haemoglobinuria. A rise in bilirubin levels and positive direct antiglobulin test (DAT) will also be present.

Development of antibodies to red cells in the patient's plasma (alloimmunisation)

Transfusion of red cells of a different phenotype to that of the patient will cause allo-immunisation (for example the development of anti-RhD in RhD-negative patients who have received RhD-positive cells). This is particularly dangerous if a pregnant child later receives a red cell transfusion and can cause haemolytic disease of the newborn (HDN).

Iron overload

Each unit of blood contains 250 mg of iron, and those receiving repeated red cell transfusion over a long period of time may develop iron accumulation in cardiac and liver tissues.

Chelation therapy with desferrioxamine can be used to minimise iron accumulation in those most at risk.

Infection

Viral infections. The risk of becoming infected with HIV, hepatitis B or hepatitis C from transfusion is now small in situation where there are safe and reliable blood transfusion systems. However, since there is always the potential for unrecognised or unknown infection to be spread via transfusion, **all** non-essential transfusions should be avoided.

Bacterial infections. Blood must be always stored at the correct temperature (at 2–6°C for up to 35 days if using citrate- phosphate-dextrose adenine anticoagulant *or* up to 21 days if using citrate-phosphate-double dextrose). This means that electrical power for the blood transfusion storage fridge must not stop and if it does an alarm must indicate to laboratory staff that blood can rapidly become infected if temperature exceeds 6 degrees C for more than a very short time. Ideally each blood bag should be labelled with a temperature-sensitive strip that changes colour when the correct temperature for storage has been exceeded for a clinically significant period of time.

Improving patient safety

Reducing transfusion errors

- Introduce robust hospital transfusion protocols.
- Provide training for all staff involved in blood administration/taking samples for cross-matching.
- An understanding of transfusion medicine should be a core curricular component for all doctors/paediatric clinicians in training.
- Improved information technology, such as use of a unique barcode on the patient's wristband/blood sample and prepared blood, is important.
- **Appoint specialist transfusion practitioners where possible.**

Reducing unnecessary transfusion

- Transfusion risks related to the use of allogeneic blood can be eliminated using autologous blood (whereby patients collect and store their own blood for use in planned surgery). However, this practice is not risk-free.
- Ensure that blood products are only used when the patient is judged more likely to benefit from than be harmed by a transfusion.
- Ensure that electrical power to blood storage refrigerators is constant and have measures to detect failures in power supply.
- Always record in the patient's notes the indication for giving blood.
- Adopt procedures such as checking for and correcting anaemia prior to planned surgery, stopping anticoagulants and antiplatelet drugs before surgery, minimising the amount of blood taken for laboratory samples, and using a simple protocol to guide when haemoglobin should be checked and when red cells should be transfused.
- Accept a lower haemoglobin concentration as a trigger for transfusion.
- Accept a lower post-transfusion target haemoglobin level.

Section 55. Fractures in Children

Introduction

As any parent knows, all children are susceptible to injury. However, children in resource-limited countries are probably more at risk than their developed-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 re-modelling with continued skeletal growth greater degrees of mal union 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 or weight-bear
- Bony crepitus at the fracture site
- Consider the possibility of child abuse (see Section 2) if the fracture appears inconsistent with the history given or with the child's developmental status.

Neurovascular Compromise

The limb distal to the fracture should be examined as routine including:

- Peripheral Pulses
- Capillary refill time
- Skin integrity over fracture
- Loss of sensation

Any evidence of neurovascular compromise or high risk to skin integrity should result in urgent action to reduce the fracture into normal anatomical position. Analgesia should be given and the fracture roughly realigned and splinted. More accurate realignment can be performed in hospital. Following realignment of the fracture the limb should be re-examined.

Open fractures

Open fractures occur where the fracture site communicates with a laceration or break in the skin relating to it. There is potential for the introduction of contaminants and resultant infection. Often open fractures are the result of a greater degree of violence than is the case for closed fractures.

Open fractures are graded according to the Gustilo classification:

Grade 1: Skin wound of < 1 cm with minimal soft-tissue injury

Grade 2: Skin wound of > 1 cm, with moderate soft tissue injury

Grade 3: These wounds typically involve a far greater degree of violence and energy transfer.

This is further subdivided into:

- 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 therefore a regional or free flap may be necessary.
- C. Any open fracture with an associated major vascular injury requiring repair for limb salvage.

Treatment

- Treatment is dictated by the extent of soft-tissue injury as reflected in the above grading system.
- Intravenous co-amoxiclav (30mg/kg 8 hourly over 2 months age) should be given as soon as possible.
- The initial priority is a thorough debridement and copious irrigation of the fracture site in order to reduce the burden of contamination and lower the risk 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.
- Safer options are plaster application (with or without windowing to expose the wound) or external fixation.
- It is often useful for a photograph of an open fracture to be taken by the initial assessor (perhaps on a mobile phone) so that the wound can remain covered until the patient is in the operating theatre.
- It helps to prevent frequent opening of the dressings and infection.

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. If 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 include the following:

- A hard-woody swollen extremity
- Severe pain on passive movement
- Tingling or burning sensations (paraesthesia) in the skin
- Pain out of proportion to the severity of the fracture and not relieved by splinting or analgesia
- Numbness or paralysis (loss of movement) and absent distal pulses are late signs.

Although it is possible to monitor intra-compartment 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 there are four muscular compartments separated by strong fascia:

- The lateral compartment containing the peroneal muscles
- The anterior compartment containing the dorsiflexor muscles of the ankle and toes
- The superficial posterior compartment containing the gastrocnemius and soleus muscles
- 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 antero-lateral longitudinal incision. A single postero-medial 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 to permit swelling and prevent the build-up of pressure within the compartment.

X-rays

X-rays are the most useful and specific diagnostic modality. Where possible, two orthogonal X-rays (at 90 degrees 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 include the following:

- Transverse: at 90 degrees to the long axis of the 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.
 - Greenstick fractures are only seen in paediatric fractures, due to the flexible nature of paediatric bone; this implies intact periosteum along the opposite side to the 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 occur in about 15 - 20% of major long bone fractures and 34% of hand fractures in childhood. There are five grades, with increasingly poor prognosis for fracture outcome with increasing grade because of an increasing degree of damage to the growth plate. This will lead to limb shortening as the child grows.

1. **Fracture across the epiphyseal line, not extending into the epiphysis or metaphysis.** This occurs when the growth plate is very thick, and thus tends to be seen in young children. Healing is rapid and complications are rare.
2. **Fracture across the epiphyseal line extending into the metaphysis, but not into the epiphysis.** This usually occurs in children over the age of 10 years. Healing is usually rapid and there is rarely growth disturbance.
3. **Fracture extending completely across the epiphyseal line and into the epiphysis.** This type of fracture can occur at any age and is associated with a poor prognosis.
4. **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 it has a poorer prognosis.
5. **Crush injury to the growth plate.** This is caused by severe axial loading during a fall from a height. Inevitably there is partial destruction of the epiphyseal plate, and thus a considerable risk of growth disturbance.

Treatment of fractures

- A. Reduce the fracture (if displaced).
- B. Hold the fracture while bony healing occurs.
- C. 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 to prevent secondary arthrosis.

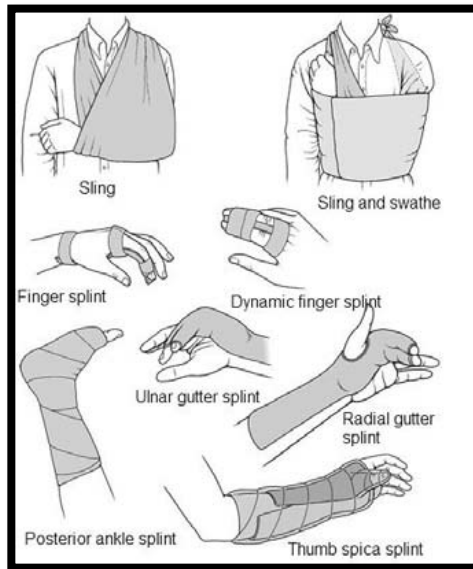
Children will often be unable to tolerate reduction under local anaesthesia. General anaesthetic will usually be required.

Once reduced, the fracture needs to be held in position while bony union occurs. During reduction of fractures, particularly in the lower limb, rotation of the limb should be checked clinically and compared with the opposite limb. X-rays, although useful for judging angulation, length, and translation, are not very helpful for judging rotation.

Splinting

- Splinting of a fracture involves immobilising the fracture, thereby preventing relative motion of the bone ends.
- In the acute phase, this will help to relieve the pain associated with the fracture. In the longer term, the fracture stability conferred by the splint will help to promote bony union.
- The commonest form of splintage uses plaster of Paris bandages (see plaster craft below).
- If these are not available preformed, then it is possible to make them from crepe 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, splints can often be fashioned from locally available materials.
- One example of this is the use of strips of bamboo and bandaging.
- The splint should be applied with the limb in the position of function.
- Then if stiffness does occur the limb will still have some use.
- For the elbow, this position is 90 degrees of flexion.
- For the ankle, a position of neutral plantar/dorsiflexion (the sole of the foot at 90 degrees to the lower leg) is preferred.

FIGURE 55.1 Slings and splints for childhood fractures. From the Merck Manual Home Health Handbook, edited by Robert Porter.



Plaster craft

- Before starting to apply a plaster, all the necessary 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 time bubbles should have stopped rising 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 one by about two-thirds.
- Twists and turns in the plaster should be avoided, as these can constrict the limb.
- Once the plaster bandage has been applied, the limb should be held still until the plaster sets.
- If proprietary plaster of Paris bandages is 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 on to an unrolled bandage that is just damp (so 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'.
- These details can include the date of the fracture, the date when the 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 and X-rays can easily be mislaid or lost.
- Wedging of the plaster can be useful for improving reduction of the fracture.

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 be 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 on the limb by means of a bandage (usually adhesive) applied around the limb.
- Balanced traction
 - 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
 - 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 in which these methods are used in the upper limb. One example is temporary skin traction for a supracondylar fracture of the humerus.

External fixation

- This involves 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 to prevent the build-up of crust, infection of the track and secondary osteomyelitis.
- The fixator can remain in position until bony union occurs or be replaced by 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 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 K-wires-type needles is possible.

Internal fixation

- This involves the use of screws, plates, and other types of metalwork to rigidly hold the reduction of a fracture.
- Although 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 splinting, 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, a constant supply of fixation devices needs to be available.
- The surgeon should be trained in the application of the device and in the surgical approach necessary for it.
- The fixation devices and tools should be sterile and the level of asepsis in the operating theatre must be high.
- In some cases, intra-operative X-ray guidance (fluoroscopy) is necessary for accurate fixation.
- Intramedullary methods of fixation, popular in well- resourced countries for the fixation of adult long bone fractures, are rarely appropriate in 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, the age of the child, the operative resources available and the training of the surgeon involved.

Ongoing fracture care

Once reduction and stabilisation of the fracture have occurred, ongoing 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 that secondary displacement has not occurred. The overall duration of immobilisation required 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 time period during 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 all indicate 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 1 week per year of age, but this can be shortened by transfer into a plaster hip spica once fracture stability permits.

- *Age 0 - 2 years and weight under 12 kg:*
 - Gallows traction, thighs in 45-degree flexion and hips 30-degree abduction.
 - Limb length inequality is seldom a problem as fracture does not shorten excessively.
 - Shortening of up to 1.5 cm and angulation of up to 30 degrees is acceptable.
 - Early spica casting is often possible.
 - This fracture is associated with non-accidental injury in 50 - 80% of cases at this age.
- *Age 2 - 10 years:*
 - Skin traction, either in the 90/90 position (hip and knee flexed to 90 degrees) 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 the position is acceptable.
 - With skin traction the weight used should not exceed 5 kg, but with skeletal traction up to 10% of body weight can be applied.
- *Age 10 - 15 years:*
 - Skeletal traction, either in 90/90 position or straight.
 - There is a much greater risk of shortening in this group, and less potential for subsequent growth acceleration and length equalisation.
- Above the age of 15 years
 - Children can be treated as 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 degrees 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, to ensure that the length of the fractured limb is the same length as the uninjured side.
- Acceptable degrees of shortening are 5 - 10 mm in the 0 - 5 years age group but aim for none in any older age group.
- Acceptable axial alignment is less than 10 degrees of recurvatum (where the apex of the fracture site points posteriorly) and less than 5 degrees 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.
- Un-displaced fractures in children aged 1 - 5 years can often be treated in below-knee casts or even below-knee plaster cylinders.

Distal humerus

- Supracondylar fractures of the humerus have the highest rate of complications and some of the poorest results of treatment of all paediatric fractures.
- They are also difficult to diagnose without an X-ray.
- The peak incidence is at the age of 6 or 7 years.
- The vast majority (98%) are extension type, featuring a posteriorly displaced distal part. Only 2% are flexion type, resulting from a fall on to 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, closed reduction should be attempted.
- If this fails to restore pulses, immediate open reduction and surgical exploration of the brachial artery should be performed.
- In other displaced fractures with palpable distal pulses, closed reduction should be attempted, possibly combined with percutaneous pin fixation for unstable fractures.
- Acceptable reductions will have no more than 4 degrees of varus as determined by Baumann's angle on the antero-posterior radiograph.
- Additionally, the axis of the capitellum should be at 45 degrees to the humeral shaft.
- If an acceptable position is not obtained, this may be an indication for open reduction and percutaneous K-wire fixation.
- Alternatively, the limb can be placed on traction in extension.
- As the swelling subsides, it will become easier to affect 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 degrees will assist in maintaining the reduction of extension-type fractures, but care should be taken to ensure that distal pulses are maintained.
- Ideally, X-rays should be taken on a weekly basis to ensure that reduction is maintained.
- The plaster cast can be completed once swelling has resolved, and percutaneous wires can be removed after 3 weeks.
- The typical duration of immobilisation necessary for union is 4 - 5 weeks in the 0 - 5 years age group and 6 - 7 weeks in the 5 - 10 years age group.

Forearm fractures

- Both types of 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 radio-ulnar joints needs to be determined.

- Be aware of the possibility of a Monteggia fracture, which consists of dislocation of the radial head along with fracture of the ulna.
- 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 years, acceptable reduction can be defined as anything less than 15 degrees of displacement and 45 degrees of malrotation.
- Above 9 years, at least bayonet apposition is required with less than 30 degrees of malrotation, less than 10 degrees angulation if the fracture is proximal or less than 15 degrees 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 2 years of skeletal growth remaining.
- Following reduction, an assessment of forearm supination and pronation should be undertaken to ensure that there is no block.
- The arm should be immobilised in an above-elbow cast with the elbow flexed to 90 degrees.
- Opinion varies as to the position of the wrist in the cast.
- Some surgeons place 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 1- and 2-week intervals following manipulation to ensure that secondary displacement has not occurred.
- If displacement does occur, re-manipulation 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 (intra-operative fluoroscopy is required).

Distal radial ('wrist') fractures

Children's distal radial fractures are usually the result of a fall on the outstretched hand and are rarely intra-articular.

Common types include the following:

– *Galeazzi fracture*

(isolated fracture of the distal radius) implies associated disruption of the distal radio-ulnar joint.

– *Physeal fracture*

pattern of injury described by the Salter-Harris classification)

– *Torus*

(buckling of the cortex on the compression side of the fracture without angulation).

– *Greenstick fracture* (incomplete fracture).

In children these distal radial fractures can almost always be treated with closed reduction and plaster immobilisation.

The reduction manoeuvre is to hyperextend the wrist, followed by traction and 'hinging' 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 2 years of skeletal growth remain.

Check X-rays should be taken at 1- and 2-weeks post-reduction to exclude secondary displacement.

The duration of immobilisation required depends upon the fracture configuration and the age of the child but is typically 3 - 5 weeks.

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.

Further reading

2020 American Heart Association guidelines for cardiopulmonary resuscitation and emergency cardiovascular care

<https://www.ahajournals.org/doi/epub/10.1161/CIR.0000000000000901>

Section 56. Gunshot Wounds (see Major Trauma in Handbook 1 Sections 77 and 79)

Introduction

Although the end of the Cold War led to a reduction in the risk of conflict in Europe, numerous conflicts continue to rage in the developing 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 following issues are relevant to the mechanism of wounding:

- When a bullet impacts on tissue it will impart some of its kinetic energy to that tissue.
- 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 as follows:

$$\text{Kinetic Energy} = \frac{1}{2} m (V1^2 - V2^2)$$

where m is the mass of the projectile, V1 is the velocity on entering the tissues and V2 is the velocity on exiting.

The degree to which the projectile's velocity is attenuated while 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 used bullets are similar, and thus the velocity of the projectile largely defines the injury potential. In this regard, gunshot wounds can largely be divided into three categories depending on the nature of the weapon used.

Handguns

- The commonest types of handguns feature a bullet with a diameter of 9 mm and a muzzle velocity of around 1000 feet/second.
- Only a small temporary cavity is formed, and the injury is essentially confined to the bullet track.
- Provided that 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 range from 1 mm ('birdshot') to 10 mm ('buckshot').
- Once fired, the pellets disperse in a cone-shaped pattern.
- The degree and rapidity of dispersion are 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 that will hit a particular target as the range increases.
- The above factors lead to this 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 7.62 mm in diameter that leaves the weapon at a speed of around 3000 feet/second.
- Rifling of the barrel sets the bullet spinning, which, combined with the increased velocity, leads to greater accuracy at long range.
- Rather than following 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 larger temporary cavity than is seen in low-velocity munitions.
- The sub-atmospheric 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 situ.
- 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 made by such a bullet being very much larger than the entry wound.
- 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. Therefore, a high-velocity bullet may transit lung causing only contusion, whereas transiting solid organs causes gross disruption.

Bullet type

Bullets are designed to either maim or kill. Military style pointed bullets are designed to maim and injure rather than kill. The bullet at high velocity may pass through more than one individual. An injured soldier on the battlefield takes up much more resource than one that has been killed as they will require evacuation.

Bullets used by police forces etc are more designed to kill the individual to remove the threat to others. They have a flatter design and cause more extensive damage.

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 gunshot wounds to the head, chest and abdomen have a high rate of on-scene mortality.

Protocols for treating gunshot wounds have been adopted and publicized 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 (compressing exsanguinating haemorrhage, airway, breathing, circulation), is the priority (see Sections 77 and 79 Handbook 1).

The degree to which fluid resuscitation should be carried out has been controversial. An initial bolus of 10 mL/kg in a child or 500 mL in pregnancy of Ringer-lactate or Hartmann's should be given and the response to this initial fluid challenge assessed. The concern is to avoid restarting massive bleeding again from disrupting a just-clotting wound by increasing peripheral perfusion. So, until the patient can be in a position to have any torn vessels managed, i.e. be in an operating theatre with competent staff, and receive a blood transfusion, crystalloid fluid management remains the minimum that keeps vital organs perfused.

1. Give analgesia as required (usually IV morphine) (see Section 9 Handbook 1).
2. Apply dressings to the open wounds.
3. Undertake emergency splintage of fractures.
4. Antibiotics: the ICRC recommend IV benzylpenicillin at a dose appropriate to the size of the child (usually 50 mg/kg IV 6-hourly) and in pregnancy 600–1200 mg IV 6-hourly.
5. Give tetanus toxoid and anti-tetanus serum.
6. Appropriate radiographs of the injured areas should be taken.

Wound Assessment

Before proceeding to surgical treatment, the following aspects of the wound need to be assessed:

- From the history, the nature of the weapon used (if known)
- The site of the entrance wound (and exit wound if present)
- The sizes of the entrance and exit wounds
- Cavity formation
- The anatomical structures that may have been transited
- Distal perfusion
- Presence of fractures
- Degree of contamination

Wound Debridement and Management

- 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 debridement should be a planned procedure with prior consideration given to the position of the patient and the type of anaesthesia required.

- For limb wounds, a pneumatic tourniquet should be used where possible to reduce blood loss.
- Skin incision 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, so 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 forceps 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, if 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 debrided wound should be washed with copious quantities of saline and then a dry bulky sterile dressing applied.
- Some low-energy-transfer (low-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 debridement 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 4 days (12.5 mg/ kg four times daily).
- Monitoring of the patient for signs of sepsis; check their tetanus status.
- 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 5 days, but most recent 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 5 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.
- Breaches 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 more than 85% chance of bowel or major organ transit. Exploratory laparotomy is therefore virtually mandatory.

Conclusion

Gunshot wounds from any type of weapon represent a severe injury. Some understanding of ballistics can help in the assessment of these injuries. 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.

Section 57. Landmine Injuries

Patterns of Injury

Injuries caused by stepping on to a buried blast mine or improvised explosive device (IED): traumatic amputation of the detonating limb, with fragment and minor blast damage to the other leg (most common injury).

Injuries caused by fragmentation landmine or IED:

- Widespread fragment injury to the limbs and trunk.

Injuries caused by close-proximity detonation of a landmine or IED in the hand or close to the face:

- Amputation of the hand or 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 lie 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. Recently IEDs have been placed next to roads and pathways, causing similar injuries.

Specific Problems in Children

Children sustain a higher level of injury per gram of explosive than adults, because of their smaller body mass. A small antipersonnel mine of approximately 30 grams, which would normally require a below-knee amputation in an adult, may result in an above-knee amputation in a child.

Children are susceptible to close-proximity detonation injuries, because of their tendency to pick up and play with objects that they find.

Treatment

- Initial surgical management follows the basic principles of resuscitation (see Sections 77 and 79 Handbook 1).
- In injury caused by stepping on a buried blast mine or IED, airway maintenance is not usually a problem, as the child is frequently conscious.
- As with all injured children, fear and bewilderment due to pain and the unfamiliar surroundings can be distressing for all involved.
- 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 anti-tetanus toxoid should be administered in all cases.
- Anaesthesia can be achieved using a ketamine infusion (see Section 9 Handbook 1 for pain relief).
- 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 peri-operative blood loss, which is proportionally greater in children than in 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 the medial vastus above the knee. Leave generous skin flaps, as the muscle in the stump will swell considerably post-operatively.
- 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 5 days post-operatively.
- 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 5 days post-operatively).
- Do not take the dressing down on the ward unless the patient manifests signs of systemic toxicity (i.e., fever, tachycardia, foul-smelling dressing).
- Give blood only if the haemoglobin level falls to less than 8 grams/dL.
- Give IV benzylpenicillin for 48 hours (50 mg/kg 6-hourly), then orally for a further 3 days (12.5 mg/kg four times daily).
- Give appropriate tetanus prophylaxis (see Section 26 Handbook 1).
- Inspect the wound at 5 days. If the tissue is healthy and not infected, close with interrupted non-absorbable sutures over a drain. Leave the sutures in for 3 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.

Section 58. Care of children and young people with a spinal cord injury

Introduction

The acute and immediate management of children and adolescents with a traumatically injured spine in the context of major injury is described in Sections 79 and 80 Handbook 1. Much of the important advice given in this section is adapted from the following excellent book, which is essential reading for all healthcare workers in resource-limited settings:

Werner D (1987) *Disabled Village Children: a guide for community health workers, rehabilitation workers and families*. Palo Alto, CA: Hesperian Foundation.

Mechanisms

- The following conditions predispose to spinal injuries: achondroplasia, Klippel–Feil syndrome, Down's syndrome, and juvenile rheumatoid arthritis.
- Injuries can occur during birth and from abuse.
- Most of the injuries are caused by road traffic accidents, sports, falls from trees or donkeys, bullet wounds and diving accidents.
- Non-traumatic causes include transverse myelitis, epidural abscess, tuberculosis of the spine, neuroblastoma (see Section 15), astrocytoma, eosinophilic granulomata, lipoma, teratoma and aneurysmal bone cysts).

Diagnosis

- In the conscious patient, localised tenderness in the spine, and impairment or loss of sensation, voluntary motor power and reflexes can help to 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.
- Around 10–20% of injuries are in more than one site. Therefore, radiological investigations (X-ray, CT or MRI scan if available) of the whole spine are necessary.
- Other associated injuries are common, and loss of sensation may delay their diagnosis.

Level of the injury

The magnitude of the area of the body that is affected will depend on the level of the injury. The higher the injury is, the greater the area of the body that is affected.

In paraplegia:

- there is loss of controlled movement and feeling in the legs
 - the hips and part of the trunk may be affected (the higher the injury, the greater the area of the body that is affected)
- there may be partial or complete loss of urinary and bowel control
- there may be spasticity (muscle spasms) or hypotonia in the legs.

In Quadriplegia:

- In addition to the features of paraplegia mentioned above, there is loss of controlled movement and feeling in the arms
- The diaphragm and other muscles of respiration may be affected (the higher the injury, the greater the respiratory muscle involvement).

Complete and incomplete injuries

When the spinal cord is damaged so completely that no nerve messages get through, the injury is said to be 'complete'. Feeling and controlled movement below the level of the injury are completely and permanently lost. If the injury is 'incomplete', some feeling or feeling as well as movement may remain. Alternatively, feeling and controlled movement may return (partly or entirely) little by little over a period of several months. In incomplete injuries, one side may have less feeling and movement than the other.

X-rays often do not show how complete a spinal cord injury is. Sometimes the backbone (spinal column) may be badly broken, yet the spinal cord damage may be minor. And sometimes (especially in children) the X-ray may show no damage to the backbone, yet the spinal cord injury may be severe or complete. Often only time will tell how complete the injury is.

Examination of and documentation of the patient with a spinal cord injury

The level and severity of spinal cord injury is assessed according to the International Standards of Neurological Classification of Spinal Cord Injury (ISNCSCI), which is currently a universal classification tool developed by American Spinal Injuries Association (ASIA). This involves examination of sensory and muscle power to determine the level of injury and also to identify whether the injury is complete or incomplete.

In children, performing neurological examination to identify the level and severity of spinal cord injury can be quite difficult. It requires time and may have to be done in stages. Correlation with findings of radiological imaging may be a helpful hint towards identification of level of spinal cord injury.

Neurological examination of spinal cord injury in adolescent children is similar to adults, but in very young children, some of the information may have to be obtained by observation by team members. Examination is likely to become particularly difficult in children below 6 years of age, children with learning difficulties, head injuries and injury before toilet training for the following reasons:

1. inability of the child to cry aloud,
2. an abnormal breathing pattern,
3. absence of or weakness of spontaneous movement in the lower limbs compared to the upper limbs in the absence of injury to the limbs,
4. inability to bend but not straighten the elbow or move the wrist up against gravity are likely to indicate the presence of a spinal cord injury.

Learning resources for neurological examination and classification are available- one such resource is the Pediatric Standards Training Program, WeeStep, from American Spinal Injuries Association (ASIA) e-Learning Centre. <https://asia-spinalinjury.org/learning/>
Accessed 4th April 2021

Neurological deterioration

This may be caused by:

- further mechanical damage and/or further non-mechanical damage to neural tissue during treatment
- hypoxia, hypotension and sepsis that develop due to poor management of the multisystem malfunction.

Acute management of spinal cord injuries: overall approach

For acute ABC management in the context of major traumatic injury, see Sections 77, 79 and 80 in Handbook 1.

- Aim to prevent complications related to multisystem dysfunction by good ABC resuscitation.
- Aim 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, and the risk of complications is high.
- 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 6 hours for 48 hours.
- 'Rehabilitation' should begin in parallel with the medical treatment as soon as possible.
- Arrange early counselling and psychological support for the child, their parents and family members.
- Start physiotherapeutic procedures to prevent contractures of paralysed muscles, chest infections and pressure sores.
- Train all systems of the body to function as safely and with as near normal convenience as possible.
- Aim for psychosocial and physical reintegration of the child in the community without significant loss of education.
- Ensure a teaching programme for the child and/or their parents aimed at minimising the development of complications (medical, physical and psychological) in the medium and long term.

Offer lifelong regular hospital assessment and treatment, if necessary, to maintain health and rehabilitation.

Acute spinal injury

- Keep the spine in a neutral position (with pillow arrangements). For cervical spine injury, immobilise with sandbags at the side of the head for about 6 weeks (see Section 94 Handbook 1), followed by bracing for 6 to 8 weeks. In children under 6 years of age, the sagittal diameter of the skull exceeds that of the chest, forcing the neck into flexion. A cut-out should be made in the board or the mattress to recess the occiput.
- Children with an unstable fracture of the spine but with intact neurology can be adequately braced in a Minerva cast for cervical spine injuries and a body cast for thoracolumbar injuries. Alternatively, or later, surgery can be undertaken, but only in a specialist centre (if available).
- 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 quadriplegic patient 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 their body 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. Neurogenic shock which is caused by an acute spinal cord injury is not to be confused with hypovolemic shock caused by blood loss.
- Care is needed with IV hydration, as circulatory overload and pulmonary oedema can easily occur. Consider positioning and vasopressor medication.
- 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 a significant drop in blood pressure which may affect spinal cord blood flow and adversely affect neurological recovery.
- Following the 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). Any would be painful condition below the level of spinal cord injury can cause this. The commonest causes are urinary retention and constipation. Treat this condition by placing the patient in the upright position (usually sitting) and if they are over 12 years of age 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.

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 that no major chest injury is present.
- Encourage deep-breathing exercises and postural drainage, assist coughing and monitor oxygen saturation.

Gastrointestinal system

- In the acute phase after a spinal cord injury, all patients are at risk of developing paralytic ileus. The resulting abdominal distension can embarrass the diaphragm and further impede diaphragmatic breathing. Avoid oral intake in the first 48–72 hours following injury and until bowel sounds are audible.
- The risk of gastrointestinal bleeding from stress ulcers is high. Therefore, administer PPI inhibitors such as oral omeprazole (child 1 to 11 years 1-2 mg/Kg once daily (maximum per dose 40mg) and (Child 12-17 years 40 mg once daily) or antacids, for the first 3 to 4 weeks following injury.
- A regular bowel regime consisting of suppositories at fixed and regular intervals not exceeding 24 hours should be instituted initially by a nurse or parent, and later by the child (see below for details).

Hypercalcaemia

- This occurs in 10–20% of children, especially in quadriplegia and complete spinal cord injuries. The onset is insidious 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 treatment.

Management of nutrition

What food should be given?

If the child is malnourished, give them 200 kcal/kg/day (see Section 56 Handbook 1). The daily number of kilocalories should be divided by the number of meals given during the day, usually four meals per day.

F100 can be used to correct malnutrition (see Section 56 Handbook 1)

Commercial F100

This special milk is prepared in a sachet. All that the family has to do is open the packet and dilute the contents in 2 litres of water.

Home-made F100

When commercial milk is not available, F100 can be prepared from the recipe shown in Section 56, Table 56.7 in Handbook 1.

The basic diet is composed of F100 meals. However, when the patient is gaining weight quickly other foods can be introduced. For example, the usual food eaten in the area can be used, but this should be enriched with the addition of oil and vitamin and mineral mix, and sometimes dried skimmed milk.

Example of calculation:

A child who weighs 20 kg should receive $200 \text{ kcal} \times 20 = 4000 \text{ kcal}$ per day.

They will receive 4 meals per day, therefore 1000 kcal per meal.

Doses of supplemental nutritional aids

These are as follows (see Sections 55 and 56 Handbook 1):

- Zinc: 2 mg/kg/day, or for children over 5 years of age, 40 mg once a day (of the elemental formula).
- Vitamin C: 45 mg/day.
- Iron: one ferrous sulphate tablet of 200 mg once weekly for children over 5 years of age.

TABLE 58.1 Recipe for preparing 1 litre of high-energy food

Food item	Quantity
Dried skimmed milk (DSM) or oiled full cream milk	80 grams (900 mL)
Vegetable oil	60 grams (20 mL)
Sugar	50 grams
Water (boiled)	Add water to make 1 litre of preparation
CMV (minerals and vitamin mix)*	20 mL (should be added after the water)

*The CMV should be added when the preparation of milk is ready. Whisk to prevent the oil from separating. This keeps for 12 hours.

Subsequent nursing and medical management and education of the child and their family

Early questions that a child with spinal cord injury and their family may ask

Will my child always remain paralysed?

This will depend on how much the spinal cord has been damaged. If paralysis below the level of the injury is not complete (for example, if the child has some feeling and control of movement in their feet) there is a better chance of some improvement.

Usually, the greatest improvement occurs in the first months. The more time that goes by without improvement, the less likely it is that any major improvement in feeling, or movement will occur.

After 1 year, the paralysis that remains is likely to be permanent. As gently as you can, help both the child and their parents to accept this fact by reassuring them that with the appropriate support education and training the child can grow to live a long, enjoyable, productive, dignified life. It is important that they learn to live with the paralysis as best they can, rather than waiting for it to get better or going from clinic to clinic in search of a cure.

Immediately after a spinal cord injury, the paralysed limbs are in 'spinal shock', and are hypotonic. Within a few days or weeks, the legs may begin to stiffen, especially when the

hips or back are straightened. Also, when it is moved or touched, a leg may begin to 'jump' (a rapid series of jerks, or 'clonus').

This stiffening and jerking is an automatic reflex called 'spasticity'. It is not controlled by the child's mind, and often happens where spinal cord damage is complete. It is not a sign that the child has begun to feel where they are touched or is recovering control of movement. Some children with spinal cord injury develop spasticity, while others do not.

If the spinal cord injury is above the level of the top edge of the hipbone (above the second lumbar vertebra), spasticity will be likely. If the injury is below this level, muscle spasms will not develop.

Severe spasticity often makes moving and control more difficult. However, the child may learn to use both the reflex jerks and spastic stiffness to help them to do things. For example:

- When the child wants to lift their foot, they hit the thigh, triggering the jerks that lift the leg.
- In lower back injuries, the spasticity or stiffness of the legs may actually help the child to stand supported for short periods.

Will my child be able to walk?

This will mainly depend on how high or low in the back the injury is and how severe the cord damage or the damage to the nerves is.

If the child's injury is in the lower back and if their arms are strong and they are not overweight, there is a chance that they may learn to walk with crutches and braces. However, they will probably still need a wheelchair to go long distances.

It is best not to place too much emphasis on learning to walk. Many children who do learn to walk find it so slow and tiring that they prefer to use a wheelchair.

It probably makes sense to give most paraplegic children a chance to try walking.

However, do not make the child feel guilty if they prefer a wheelchair. Let the child decide which is the easiest way for them to move about.

For independent living, other skills are more important than walking, and the family and child should place greater emphasis on these skills, such as dressing, bathing, getting in and out of bed, and toileting. Self-care in toileting is especially important and is made more difficult because of the child's lack of bladder and bowel control.

What are the prospects for my child's future?

The likelihood of a child with paraplegia leading a reasonably normal life are good, provided that three major medical risks are avoided:

1. skin problems (pressure sores)
2. recurrent urinary tract infections
3. contractures (shortening of muscles, causing deformities); these are not life-threatening, but they can make moving about and doing things much more difficult.

The child is helped to become more self-reliant:

1. home training and encouragement to master basic self-help skills such as moving about, dressing and toileting
2. education: learning of skills that make keeping a household, helping other people, and earning a living more achievable.

It is more difficult for children with quadriplegia to lead a normal life because they are more dependent on physical assistance.

In well-resourced countries, many children with paraplegia manage to lead full rich lives, earn their own living, get married, and play an important role in the community. With effort and organisation, the same potential for leading a normal life can exist in all countries.

Can anything be done about loss of bladder and bowel control?

Yes, it certainly can. Although normal control rarely returns completely, the child can often learn to be independent in their toilet, and to stay clean and dry (except for occasional accidents). Often, they will need to learn to use a urinary catheter and learn to bring down a bowel movement with a finger or suppository (see below).

What about sexual relationships and having children?

Many people with spinal cord injuries marry or have fulfilling sexual relationships. Women with spinal cord injuries can become pregnant and have babies. Men are likely to require medical assistance to father their own children.

Helping the child and their family to adjust to and accept the injury

Perhaps the biggest problem is that one day the child is physically active and able, and the next they are suddenly paralysed and (at first) unable to do much for themselves.

They have lost all feeling and control in part of their body, which feels like a 'dead weight'. This is very hard for both the child and their family to accept. Both have enormous and partially justified fear and uncertainty about the future. The child may become deeply depressed, or angry and uncooperative. They may refuse even to sit in a wheelchair because this means accepting that they are unable to walk. There are no easy ways to address the child's fear and depression, but here are some suggestions that families have found helpful.

Recognise that the child's fear, depression, and anger are natural responses, and that with love, understanding and encouragement they will gradually overcome them. Be honest with the child about their disability. Do **not** tell them '*We will find a cure for you*' or '*Soon you will get well and be able to walk again.*' Very probably this is untrue. Misleading the child in this way only makes it more difficult for them to accept their disability and begin to shape a new life. Also, as the promised '*cure*' fails to materialise, the child will become more uncertain, distrustful, and afraid. In the end, it will be much easier if you gently tell them the truth.

Provide opportunities to keep the child's mind active by playing, working, exploring, and learning through stories, games, and studies. But at the same time respect and be supportive of the child when they feel sad and frightened. Let the child cry, comfort them when they do so, but do not tell them not to cry. Crying helps to relieve fear and tension.

Start the child with exercises, activities, and relearning to use their hands and body as soon as possible. Start with what the child can do and build on that.

Try to arrange for the child to watch, talk with and get to know other people with spinal cord injury.

Invite the child's friends to come and visit, play with him or her, and let the child know that they are eager for the day he or she will be back in school.

Encourage the child to do as much for him- or her- self as possible, even if it takes a little longer.

As far as possible **avoid the use of 'tranquillizers** or other inappropriate medication. The child needs an alert mind and the ability to move actively all day.

To prevent or reduce the harmful effects of the complications of spinal cord injury; special precautions need to be taken early and continued throughout life.

Early care

Early care following spinal cord injury is best provided in hospital in a specialist spinal injury centre, if available. Family members should stay with the child to make sure that he or she is kept clean and turned regularly, so that bed sores and pneumonia are avoided. Busy hospital staff with little experience of treating spinal cord injuries sometimes allow severe bed sores to develop, which may be life-threatening for the child.

The damage that has already been done to the spinal cord cannot be corrected with surgery or medicine.

Preventing pressure sores (bed sores)

When sensation has been lost, pressure sores can easily form on the skin over bony areas, especially on the hips and bottom. The time of greatest risk of sores developing is in the first weeks after the injury. This is because the child must stay very still and has not yet learned to move or turn over their body. Prevention of pressure ulcers, also known as pressure sores, is extremely important, and needs understanding and continuous care, both by the child and by those caring for them.

Early prevention of pressure sores

- Lie the child on a soft mattress or a thick firm foam rubber pad.
- Place pillows and pads to keep the pressure off bony areas.
- Change their position (turn over from front to back and side to side) every 2 to 3 hours. To avoid pressure sores, lying on the abdomen is the best position.
- Keep the skin and bedclothes clean and dry.
- Give the child healthy food rich in vitamins, iron and protein.
- Move and exercise the child a lot to promote healthy flow of the blood.
- Check their skin daily for the earliest signs of pressure sores and keep all pressure off areas where sores might be developing until the skin is healthy again.

Avoiding contractures

In the first weeks following a spinal cord injury, when the child is in a lying position, joint contractures (muscle shortening) can easily develop, especially in the feet and elbows. Pillows and pads should be placed to keep the feet supported, the elbows straight, and the hands in a good position. Gentle range-of-motion exercises of the feet, hands and arms should begin as early as possible, taking care not to move the back until the injury has healed.

Movement and exercise

Do range-of-motion exercises for about 10 minutes for each leg. In the first weeks, do the exercises twice a day; later, once a day may be enough. If any signs of contracture develop, spend more time and effort on those parts of the body. From the start, exercises should be both passive (someone else moving the child's body parts) and, whenever possible, active (the child moving them).

Range-of-motion exercises should begin with great care the day after the spine is injured. The exercises will help to improve the flow of blood (which reduces the likelihood of bed sores), prevent contractures, and build the strength of the muscles that still work. Range-of-motion exercises should be continued throughout life, when possible as a part of day-to-day activity.

Cautions

- Until any breaks or tears in the spine have healed (this takes 6 weeks or more), exercise must be very gentle and limited, with smooth motions and no jerking.
- Especially at first, take great care that exercises do not move the position of the back and neck (depending on the site of injury). Start with the feet, ankles, hands, wrists and elbows.
- If exercises trigger severe muscle spasms or jerking, do not do them until the break in the spine has healed.
- Do not use force in trying to get the full range of motion, as joints can easily be damaged.
- Try to keep the full range of motion of all parts of the body but work most with those joints that are likely to develop contractures, especially paralysed parts that tend to hang in one position, such as the feet and joints that are kept straight or bent by spasticity or by muscle imbalance.

Maintaining a healthy position

The position that the body is in during the day and night is also important to prevent contractures.

Contractures that cause 'tiptoeing' of the feet can develop easily, especially when there is spasticity. Keep the feet in a supported position flexed at 90 degrees to the lower leg, not in the extended position, for as much of the time as possible when lying down and when sitting.

Teach the child to check that their feet are in a good position. Even for the child who may never walk, maintaining the feet in a flexed position makes moving from chair to bed, toilet or bath easier.

Another common problem for children with spasticity is that the knees pull together and in time contractures prevent the legs from separating. To prevent this, when the child lies on their side, they should learn to place a pillow between the legs, and to keep it there most of the time.

A common problem with wheelchair users is that they slump forward. In time this can deform the spine. In a wheelchair with a straight-up back a person with spinal cord injury slumps like this in order to balance. A chair can be designed (or adapted) so that it tilts back. This provides balance for a better position.

A special cushion also helps to prevent the child's bottom from sliding forward (and also helps to prevent pressure sores). If possible, use a cushion made of 'micropore' foam rubber (foam containing very tiny air bubbles). Rubber-coated coconut fibre also works well.

Early physical development

The goal is for the child to become as independent as possible in doing what they want and need to do. However, even before the skills of daily living are relearned, the child needs to learn to protect the body where functions that used to be automatic have been lost. The protective functions that may be lost or changed include the following:

- adjustment of the blood pressure to changes in body position
- sensation (including pain) that protects the body from injuries (for example bed sores)
- the sense of body position and ability to keep balance
- muscle strength and coordination.

A sudden fall in blood pressure in the brain when the person rises from lying to sitting, or from sitting to standing, can cause dizziness or fainting. This is a common problem in spinal cord injury because the blood pressure adjustment mechanism is partly lost. The body can be helped to gradually readapt, but precautions are needed. (These same precautions are the same for anyone who has been kept lying down for a long time.) Before beginning to sit, raise the head of the bed – a little more and a little

longer each day. If the child starts to feel dizzy or faint when sitting, tilt their back and lift their feet. Lifting exercises help the body to relearn to adjust blood pressure, and also prevent pressure sores and strengthen the arms.

The loss of sensation in parts of the body can lead to pressure sores and other injuries, such as burns and cuts. This is because the body no longer feels pain, so does not warn the child to change position or move away from danger. It is important that the child learns to protect him- or herself by changing positions often and avoiding injuries. This includes the following:

- learning to roll over
- turning at least every 4 hours when lying or sleeping
- lifting from a sitting position every 15 minutes
- washing daily
- examining the whole body every day for signs of injuries or sores
- learning to protect him- or herself from burns and other injuries.

Keeping clean is very important for people with reduced sensation, especially if they lack bladder and bowel control. Take care to bathe them daily. Wash and dry the genitals, the bottom, and between the legs as soon as possible each time they get wet or dirty. If redness, rash, or sores develop, wash more often and keep the sore area dry. Keep the legs spread open and exposed to the air. When they must be covered, use soft absorbent cotton cloth.

For treatment of specific fungal, yeast and bacterial infections of the skin, see Section 27.

Loss of the sense of body position affects a person's sense of balance, as does loss of muscle control. The child needs to develop new ways to sense the position of their body and keep their balance.

Start with the child sitting on a bench, if possible, in front of a mirror, and help them to progress through the following stages:

- 1 Place both hands on the bench.
- 2 Place both hands on the knees.
- 3 Lift one arm sideways, forward, and back.

After doing this in front of a mirror, ask the child to do it without the mirror.

As the child begins to develop better balance, start doing different movements with first one and then both arms, such as lifting weights or playing ball.

Some children may experience so much difficulty with balance that they have to start in a wheelchair or a chair with a high back and arm supports.

Redeveloping muscle strength and coordination.

All muscles that still work need to be as strong as possible to make up for those that are paralysed. Even imagining movements helps to re-educate the brain about body posture. The most important muscles are those around the shoulders, arms, and stomach.

Self-care

With the help and encouragement of family, friends and rehabilitation workers, the child can learn to become as independent as possible in meeting their basic needs, including moving about, eating, bathing, dressing, toileting, and in time other skills for daily living.

Progress toward self-care, especially at first, may be slow and frustrating. The child will need a great deal of understanding and encouragement. To make activities easier both for the child and for their helper, it is important **that they avoid becoming overweight**.

Useful methods and techniques have been devised for helping to relearn basic skills. Much depends on determination, imagination, and common sense. Start with movements like rolling over and sitting up in bed.

Keeping active

Many of the 'complications' of spinal cord injury occur because the person spends a lot of time either lying down or sitting. To stay healthy, the body needs to keep active. Lack of movement and activity causes poor flow of the blood. This can lead to pressure sores, swollen feet, painful or dangerous blood clots (thrombosis), especially in the legs, increasing weakness of bones (osteoporosis) with the risk of fracture, bladder or kidney stones, increased risk of urinary tract infections, and general physical weakness and poor health.

It is important for both body and mind that people with a spinal cord injury keep physically active. Children should be allowed to do as much for themselves as they can, including pushing their own wheelchair, bathing, transferring, washing their clothes, helping to clean the house, and helping with work.

Active participation in games and sports can also be encouraged. Swimming, basketball, and archery can be done well using only the upper body.

To keep the leg bones growing well and to prevent them from becoming weak and breaking easily, even children who may always have to use a wheelchair should, if possible, stand for a short time every day. This can be done by strapping the child to a 'standing board', or by making some kind of standing frame. Standing also helps to prevent constipation.

Management of bowel movements

When a person's spinal cord is damaged, they almost always lose control over when they will open their bowels. This makes it difficult to stay clean, which can be inconvenient and embarrassing. Although they can never regain complete control over the muscles that hold in or push out the stool, they can learn to help the stool come out, with assistance, at certain times of day. This kind of 'bowel programme' can greatly increase the person's self-confidence and freedom to take part in school, work, and social activities.

People with spinal cord damage often have problems with constipation. Some constipation can be an advantage when a person lacks bowel control. However, sometimes it can lead to serious problems, such as impaction or dysreflexia. It is therefore important to prevent serious constipation by adopting the following measures:

- Drink plenty of water.
- Eat foods that are high in fibre (for examples bran, wholegrain cereals, fruits, vegetables, cassava, beans, nuts).
- Stick to a scheduled bowel programme.
- Keep active.

Planning a bowel opening programme

Any bowel programme will work better if the child:

- does the programme every day (or every other day) and at the same time, even if they have had an accidental bowel movement shortly before, or have diarrhoea
- does the programme at the same time of day as they normally had bowel movements before their injury

- performs the task after a meal; often the bowels move best after a meal or a hot drink
- if possible, performs the task on a toilet or pot; the bowels work better in a sitting position than when lying down
- is patient; the bowels sometimes take days or even weeks to change their pattern.

Types of bowel programme in spinal injury

Different people require different types of bowel programme, depending on whether their bowels are 'automatic', 'flaccid' or 'pull back'.

- Automatic bowel usually occurs in people who have muscle spasms in their legs, and an 'automatic bladder'. The muscle or 'sphincter' in the anus remains shut until there is a stimulation of the bowel to make it open, so that the stool can come out. An automatic bowel will 'move' in response to a suppository or stimulation by a finger.
- Flaccid bowel usually occurs in children with low spinal cord damage who have limp (not spastic) legs and bladder. The sphincter muscle in the anus is also limp, so the person tends to 'ooze' or 'dribble' faeces. A limp bowel does not respond to finger stimulation.
- A bowel that pulls back is neither automatic nor limp. When you insert a finger in the anus, the stool moves back up instead of coming out.

Management of an automatic bowel

1. Start with a suppository (glycerine) prior to digital evacuation of the bowel. Glycerine suppository sizes are as follows: for an infant, 1 gram; for a child aged < 12 years, 2 grams; for a child aged > 12 years, 4 grams. With a finger covered with a glove or plastic bag, and then vegetable oil or Vaseline, push the suppository about 2 cm (1 inch) up the anus. Do not push it into the stool but push it against the wall of the bowel. It is possible to try this same activity without a suppository; often finger stimulation alone is enough to stimulate a bowel action.
2. Wait for 5–10 minutes. Then help the child to sit on a toilet or pot. If they cannot sit, have them lie on their left side (on top of toilet paper or newspaper).
3. Put an oiled finger into the anus for a distance of about 2 cm. Gently move the finger in circles for about 1 minute, until the anus relaxes, and the stool pushes out.
4. Repeat the finger action three or four times, or until no more stool is felt.
5. Clean the bottom and anus well and wash your hands.

Management of a flaccid bowel

Since the bowel does not push, the stool must be taken out with a finger. This is best done after each meal, or at least once a day.

- If possible, the child should be sitting on a toilet or pot or lying on their left side.
- With a gloved and oiled finger, remove as much stool as you can.
- Since a limp bowel tends to ooze stool, the child should be given foods that make the stool firm or slightly constipated (do not give the child stool-loosening foods).

Management of a bowel that 'pulls back'

For this kind of bowel, the bowel programmes described above do not usually work. Finger stimulation makes the bowel act in the opposite direction and pull the stool back in. The child will have 'accidents' during the day. Often it is more effective to first put some anaesthetic jelly (e.g., lidocaine) up the anus. If you cannot obtain the jelly, you can mix some liquid injectable lidocaine with Vaseline or any other jelly. Wait for several minutes, and then proceed to the automatic bowel management.

Other important issues regarding bowel problems

- Children can almost always learn to do their own 'bowel programme'.
- Do not use enemas or strong laxatives regularly. They stretch the bowel, injure its muscles, and make following a regular programme more difficult. A mild laxative (senna or

Dulcolax; see Section 17) may be taken occasionally when needed. However, drinking more liquid and eating food high in fibre is usually sufficient.

- If there is bright red blood in the stool, a blood vessel was probably torn during the management described above. Be more gentle! If there is dark old blood and the stools are black and tar-like, and the child is generally unwell, the parents must seek hospital advice urgently.
- A small amount of liquid stool (diarrhoea) may be a sign of 'impaction', which is a ball of hard stool stuck in the bowel. Only liquid stool can leak around it. Do not give medicine that is used to stop diarrhoea, as this could make the impaction worse. Try to remove the stool with a finger or use stronger laxatives on a temporary basis (see Section 17).

A bowel management programme may at first seem difficult and messy and is initially very embarrassing for the child. However, it soon becomes a habit. It is very important both for the child's health and for their social well-being. Do it regularly at the same hour of the day, and do not miss a day.

Constipation is almost always a potential problem, and can cause haemorrhoids, anal fissures, and mucosal tears. For management of an acute episode, see Section 16. If constipation is regularly a problem, consider giving regular senna tablets (7.5 mg sennoside): aged 6–12 years, 1–2 tablets once daily; for 12–18 years, 2–4 tablets once daily or liquid (7.5 mg sennoside in 5 mL); for children under one year of age, 2.5–5 mL once daily; and for children over 6 years of age, 5–10 mL or 1–2 tablets once daily.

Locomotor system problems

- There is a 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.

Management of spasticity. Excess spasticity should be first regarded as a symptom of one or more complications or, indeed, a would-be painful pathology that cannot be manifested as pain or discomfort because of the sensory impairment/loss. Constipation, urinary infection, skin damage, contractures of muscles, fractures or infections and intra-abdominal pathology should be whenever possible prevented and excluded to minimise the level of spasticity. When children are well managed, and their parents are educated in the condition and how to prevent complications, spasticity is not a common problem.

The administration of anti-spasticity medication should be regarded as temporary until the pathology is treated following which it should be withdrawn gradually. Baclofen is the most helpful drug and given in the following doses:

Child 1 month to 7 years: Initially 300 micrograms/Kg daily in 4 divided doses, gradually increased at weekly intervals until satisfactory response: maintenance 0.75 to 2 mg/kg daily in divided doses. Review treatment if no benefit within 6 weeks of achieving maximum dose: maximum is 40 mg per day.

Child 8 to 17 years: Initially 300 micrograms/Kg daily in 4 divided doses, gradually increased at weekly intervals until satisfactory response: maintenance 0.75 to 2 mg/kg daily in divided doses. Review treatment if no benefit within 6 weeks of achieving maximum dose: maximum is 60 mg per day

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 to 3 years, 4-hourly gentle suprapubic pressure will empty the bladder.
- Children above the age of 3 years are best managed with intermittent catheterisation until effective reflex micturition occurs and the residual urine is consistently below 60mL.
- Children with lower motor neuron lesions are likely to require intermittent catheterisation for the rest of their life. Initially this should be done by an attendant or parent. However, with teaching and training, a child with good hand function can learn to do clean intermittent self-catheterisation. Intermittent catheterisation is the safest method of bladder drainage.
- The use of indwelling urethral catheters is not recommended after the first 48–72 hours but may sometimes be appropriate (see below).
- Antibiotics should be reserved for urinary tract infections with systemic manifestations.

Most children with spinal cord injury do not have normal bladder control. This can be inconvenient, embarrassing, and cause social and emotional difficulties. In addition, the loss of control can cause skin problems and dangerous urinary tract infections. For these reasons, it is important to learn ways to stay clean, dry and healthy. Most of the methods are not difficult, so children should be able to do this themselves, and this in turn will help them to feel more self-reliant.

The main goals of urine system management are as follows:

- to prevent urinary infection
- to promote self-care in staying as dry as possible.

Prevention of urinary tract infections is extremely important. Infections of the urinary system (bladder and kidneys) are very common in spinal cord injury and are one of the main causes of early death. Therefore, any method that is used for self-care or staying dry must also help to prevent urinary tract infections. Make every effort to prevent infection from entering the bladder. Keeping clean is essential. It is also important to empty the bladder regularly and as completely as possible. If some urine remains in the bladder, bacteria will grow in it and cause infection.

The ideal method of urinary control empties the bladder completely and in a clean, regular, easy and self-reliant way.

Types of bladder problems

Automatic bladder: A child with paralysis whose legs have 'reflex spasms' (uncontrolled stiffening or jerking) may have reflex spasms in their bladder. As the bladder fills with urine, the walls of the bladder stretch and cause a reflex spasm. As the bladder squeezes, the muscles that hold back the urine relax, letting the urine flow out. This is called an 'automatic bladder' because it empties automatically when it gets full.

Flaccid bladder: When a child's paralysed legs are limp (due to lower motor neuron damage) and do not have spasms, usually the bladder is also limp or flaccid. No matter how much urine fills the bladder, it will not squeeze to empty. The bladder stretches until it cannot hold any more urine. The urine then begins to drip out and overflow incontinence develops. The bladder does not completely empty, and because some urine remains in the bladder, there is an increased likelihood of infection.

The simplest methods of bladder management work well with an automatic bladder but do not work with a limp bladder. Therefore, it is important to try to establish which type of bladder the child has.

For the first few days or weeks after the spinal cord injury occurred, the bladder is almost always flaccid. Urine either drips out or does not come out at all. Then, as the 'spinal shock' wears off, people with higher back injuries (above the second lumbar vertebra) usually develop an automatic bladder. In people with lower back injuries the bladder usually remains flaccid.

During the first weeks after the spinal cord injury, usually a Foley catheter is kept in the bladder all the time. However, after about 2 weeks it is a good idea to test how the bladder works by removing the catheter and trying one of the methods described below. If the child is often wet, try another method for that type of bladder.

Methods for managing the automatic bladder

Triggering programme: This method usually causes the bladder-emptying reflex to work when the person is ready to pass urine. It can be done using a urinal, toilet, potty or jar. This is the first method to try, because nothing is put into the bladder. It is easy, so a child can do it unaided.

- 1 Tap the lower belly (over the bladder) firmly with your hand for about 1 minute. Stop and wait for the urine to flow.
- 2 Tap again. Repeat several times until no more urine flows.

If possible, once a week after triggering, use an in-out catheter to see how much urine is left. If there is less than a cupful (150 mL), continue the triggering programme. If there is more than a cupful on several occasions, the bladder is not emptying well enough, and another method should be tried (see below).

Periodic use of a catheter: This method allows the bladder to be emptied completely before it becomes too full. Sometimes it can be used to prepare the body for triggering. Put a clean or sterile standard catheter into the bladder every 4–6 hours to empty the urine, and then remove the catheter. If the child drinks more liquid than usual, put in the catheter more frequently to keep the bladder from stretching too much.

To reduce the risk of urinary tract infections, regular frequent use of the catheter is more important than using a sterile catheter. It is a mistake to stop using the catheter only because you have not had an opportunity to boil it (for example when travelling, or at school). Just wash out the catheter with clean drinkable water after use and keep it in a clean jar or towel. Do not go too long without catheterising, and do not stop catheterising altogether. It is important not to leave a large amount of urine in the bladder.

How to insert a catheter

Healthcare workers and parents can easily be taught to put in a catheter. With a little practice, children with paraplegia can also learn to do this. A mirror can help girls to see the perineal area.

The best catheter size is usually #8 or #10 for a small child, and #14 or #16 for a large child.

Vigilance about cleanliness (that is boiling the catheter and wearing gloves) is important when using a fixed (Foley) catheter. However, for periodic use of a regular (in and straight out) catheter, a clean rather than sterile technique is more practicable (and therefore may be

safer). Wash the catheter well with clean water after each use and keep it in a clean container. Wash your hands well before using it.

The procedure for insertion of a catheter is as follows:

- 1 If possible, boil the catheter for 15 minutes, or at least wash it well and keep it clean.
- 2 Bathe the child well (at least daily). Wash well under the foreskin or between the vaginal lips and the surrounding areas.
- 3 Wash your hands with soap. After washing, touch only things that are sterile or very clean.
- 4 Put very clean cloths or towels under and around the area.
- 5 Put on sterile gloves or rub your hands well with alcohol or surgical soap.
- 6 Cover the catheter with a lubricant (slippery cream) such as KY Jelly that dissolves in water (do not use oil or Vaseline).
- 7 Pull back the foreskin or open the vaginal lips.
- 8 Holding the foreskin back or the vaginal lips open, gently put the catheter into the urethra. Twist it as necessary, **but do not force it**. Hold the penis straight at this angle.
- 9 Push the catheter in until urine starts to flow out, then push it in 3 cm further.
- 10 If using a regular catheter, each time the child passes urine they should tighten their stomach muscles or gently massage the lower abdomen to empty all urine. Then take out the catheter, wash it well, boil it, and store it in a clean jar or towel.

To avoid introducing infections when using a catheter, it is important to be very clean and to use only a catheter that is sterile, boiled or very clean.

Using a fixed (Foley) catheter:

With this method, the catheter is left in all the time to drain the urine from the bladder continuously. A Foley catheter is often used immediately after injury, and in some cases for many months or years. The catheter connects to a collection bag that can be attached to the leg and worn under the clothes. The catheter should be changed using a sterile technique once weekly or more frequently if there is a urinary tract infection (see below).

In many areas this is the easiest method because other supplies are difficult to obtain. However, a Foley catheter can cause many problems, including the following:

- bacteria entering the bladder, causing a high risk of infection
- continuous bladder irritation, which can cause bladder stones to form.
- Urethral ulcer that may develop into fistula (a channel of communication between the urethra that breaks through the skin)

If you have tried other methods unsuccessfully or no other equipment is available, a Foley catheter may be the only option. To prevent complications from occurring it is very important that the Foley catheter is used carefully:

- Always wash your hands thoroughly before touching the catheter.
- Clean the skin around the catheter with soap and water at least twice a day, and after each bowel movement.
- Do not disconnect the collection bag except to empty and wash it. Wash out the collection bag with soap or diluted bleach and water once a day.
- If the catheter must be clamped, use a sterile plug, never a glass ampoule (small bottle), as this may break and cause injury.
- Keep the collection bag below the level of the bladder to keep the urine from flowing back into the bladder via gravity.
- Tape the catheter to the leg when the child is in a wheelchair.
- Check regularly to make sure that the urine is emptying, and that the catheter is not blocked. Make sure that there are no sharp bends or folds in the tubing.
- When turning, lifting or moving the child, remember to move the bag, too. Do not let it pull at the catheter or stay under the child.

- If the catheter becomes blocked, take it out, squirt boiled water through it, and put it back. Alternatively, use a new catheter. Use a sterile or very clean syringe.

Condom catheter for male children:

This is a practical method for male children and adolescents who cannot control their urine flow. It can be used in combination with triggering, to avoid accidental wetting.

A condom catheter is a thin rubber bag that fits over the penis. It has a tube that connects to a collection bag. Condom catheters are available in different sizes. If they are too costly or not available, a regular condom can be attached to the collection tube with a rubber band or tape. Alternatively, a thin, very clean plastic bag or the finger of a rubber glove (or a 'finger cot') can be used.

To hold the condom on the penis, stretchy adhesive tape can be used.

Important precautions for condom catheter use include the following:

- Ensure that it is not too tight, otherwise it could stop the blood flow and seriously harm the penis. Avoid the use of non-stretch tape.
- If the penis has erections, try to put on the condom when it is erect.
- Remove the condom once a day and wash the penis well.
- If possible, remove the condom at night, and use a bottle or urinal to catch the urine.
- Check the condom and penis often, to ensure that everything is all right.
- If the penis becomes injured, swollen or looks sore, remove the condom until it is healthy again.

Methods for the limp bladder

If the person's bladder is flaccid, it never empties by reflex. The bladder will constantly contain some urine unless an effective emptying method is used.

Girls can use a Foley catheter. This is often the simplest method, but it can lead to urinary tract infections. Alternatively, try an 'intermittent' (in-and-out) programme, using a regular catheter every 4–6 hours. If there is leaking in between catheter times, use diapers, rags or a thick sanitary pad to catch the urine. Change them often and wash the skin often to protect the skin and prevent sores. Boys can use an intermittent catheter every 4–6 hours.

Other suggestions for the flaccid bladder

- The push method: Strain to push the urine out by tightening the abdominal muscles. This method is recommended by many professionals, but it can cause problems. If the muscles do not relax to let the urine flow out, pushing on the bladder can force urine back into the kidneys, causing kidney infection and damage. Therefore, the push method should only be used if the urine flows out easily with gentle pressure, or if no other method is possible.
- However, it is best to also use a regular catheter at least three times a day. This is because the bladder may not have emptied completely, which makes infection more likely.

Management of urinary tract infections (see Section 46 Handbook 1)

Children with spinal cord injury have a high risk of urinary tract infections, for the reasons discussed above. Long-term or untreated infections and kidney problems are a common cause of early death. Preventive measures are essential, but even when precautions are taken, some urinary tract infections are still likely to occur. Therefore, it is very important to recognise the signs and provide effective treatment.

Clinical signs

When a person who has normal sensation has a urinary tract infection, pain is felt when they pass urine or when they pass urine more frequently, including at night. The person with spinal cord damage may not feel this pain or be able to have frequency or nocturia, and therefore has to use other signs to know when they have an infection. The child may learn to recognise certain unpleasant non-specific feelings or may only know that they do not feel as well as usual. Parents and healthcare workers should learn to listen to the child and be aware of changes in behaviour or other signs that might mean that an infection is probably present.

Possible signs of a urinary tract infection include the following:

- cloudy urine, possibly with mucus, pus or blood specks
- dark or red urine
- strong-smelling or bad-smelling urine
- increased bladder spasms (cramps)
- increased wetting or changes in bladder function
- pain in the back or loins
- body aches
- general discomfort
- increased muscle spasms
- fever
- dysreflexia (headache, 'goose-bumps' when sweating, high blood pressure).

Treatment of urinary infection

At the first signs of infection, the child should drink even more water than usual.

Antibiotics may also be necessary. However, avoid frequent use of antibiotics because they may become less effective as bacterial resistance develops. If the child has had urinary infections before, they can start with the last medicine that was effective (for details of antibiotic treatment, see Section 46 Handbook 1).

If a medicine seems to help, continue taking it for at least 1 week, or for 4 days after the last clinical signs have disappeared. Do not change from one medicine to another unless the medicine is not working or causes serious side effects.

Prevention of urinary tract infections in patients with spinal injury

- Drink plenty of liquid (for normal daily fluid intake that should always be maintained, see Section 46 Handbook 1), with higher intake if there is a high ambient temperature. An intake of at least 2 litres (eight 250-mL glasses) a day is required for a teenager.
- Eat apples, grapes or cranberries, or drink juice made from these fruits, or take vitamin C tablets to make the urine more acidic. It is more difficult for bacteria to grow in acidic urine. (Note: the fruit and juice of oranges, lemons and other citrus fruits do not have this effect, and in fact make the urine less acid.)
- Keep hands, catheter, and collection bags very clean before, during and after the child's bladder programme.
- Encourage the child not to lie in bed all day, but to remain active.
- Do not clamp the Foley catheter or plug it with anything unless absolutely necessary, in which case always use a sterile plug.
- Adhere strictly to the bladder programme, and do not allow urine to remain in the bladder.
- Ensure that the catheter does not become bent or twisted so that the flow of urine is blocked.
- If you are using an in-out catheter, put it in regularly, at least every 4 to 6 hours. For prevention of infections, frequency of catheter use is even more important than

cleanliness. It is safer to put in the catheter without boiling it, than not to put it in. If infections are common, catheterise more often.

Sexuality and fertility

- Discuss the situation with sensitivity as soon as the child reaches early adolescence.
- Advice about contraception is necessary for girls, as fertility is not affected, regardless of the level and severity of the spinal cord injury.
- Boys with spinal cord damage above the level of the hips are likely to have reflexogenic but not psychogenic erections. Male fertility is significantly affected. However, male adolescents should be reassured that the results of assisted fertility (if available) are good.

Psychosocial integration, education, vocational training, and employment. Continuing education, vocational training and employment must be pursued as the child grows older.

Skin problems

Sensory impairment or loss, impairment of vasomotor regulation of skin blood flow associated with paralysis, double incontinence, possible anaemia and urinary tract 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 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 parents and the child in self-care, hygiene and the provision of adequate seating can all assist.

Pressure sores

Pressure sores, or 'bed sores', form over bony parts of the body when a person lies or sits on that part of the body for too long without moving. Where the skin is pressed against the bed or chair, the blood vessels are squeezed shut so that the blood cannot transport air to the skin and underlying tissue. If too much time passes without the person moving or rolling over, the skin and underlying tissue in that spot may become injured or die. First, a red or dark patch appears. Then, if the pressure continues, an open sore can form. The sore may start on the skin and work inwards, or it may start at a deep level, near the bone, and gradually work its way to the surface.

Risk factors for pressure sores

When a healthy person lies or sits in one position for a long time, it begins to feel uncomfortable, or even painful, so they move or roll over, and the formation of pressure sores is prevented. The people who are most likely to develop pressure sores are those who are unconscious or who have no sensation in parts of their body, and who therefore do not feel the warnings of pain or discomfort when their body is being damaged. This includes people with spinal cord injury.

Commonest sites of pressure sores

Pressure sores can form over any bony area. The sites where they are most likely to develop are shown in Figure 58.1.

Risks and complications associated with pressure sores

If pressure sores are not very carefully managed, they can become large and deep. Because they contain dead skin and tissue, they can easily become infected. If a sore reaches the bone, which often happens, the bone can also become infected. Bone infections

can be very difficult to cure, may last for years, and may keep recurring even after the original pressure sore has healed.

Infections in deep pressure sores often spread to the blood and then affect the whole body, causing fever and general illness, including bacteremia and septicaemia.

Incidence of pressure sores

Although pressure sores are preventable in patients who have lost sensation and movement in parts of their body, pressure sores are unfortunately very common following a spinal cord injury. Most people with spinal cord injuries in developed countries, and nearly all people with such injuries in resource-limited countries, develop pressure sores. Often the sores start to develop in hospital shortly after the injury, due to inadequate nursing care. Therefore, it is important that the families of patients with spinal cord injuries, and those patients themselves, learn as early as possible about the prevention and early treatment of pressure sores, and put this knowledge into practice.

Prevention of pressure sores

It is important that both the child and their family are taught about the risk of pressure sores developing, and how to prevent them. The following actions are important:

Avoid staying in the same position for very long. When lying down, turn from side to side or from front to back at least every 2 hours (or up to 4 hours if padding and cushioning are adequate). When sitting, lift the body up and change position every 10 to 15 minutes.

Use thick, soft padding, pillows or other forms of cushion arranged so as to protect bony areas of the body.

Use soft, clean dry bed sheets, and try to avoid them wrinkling. Change the bedding or clothing every day and also each time it becomes wet or soiled. A child who stays wet will develop pressure sores, especially if the wetness is caused by urine.

Bathe the child daily. Dry the skin well by patting it, not rubbing it. It is probably best not to use body creams or oils, or talc, except on the hands and feet to prevent cracking, as these products soften the skin and make it weaker. Never use heat-producing oils, lotions, or alcohol.

Examine the whole body carefully every day, checking in particular those areas where pressure sores are most likely to develop. If any redness or darkness is present, take extra care to prevent all pressure over this area until the skin returns to normal.

Good nutrition is important for preventing pressure sores. Make sure that the child gets enough to eat (but do not let them become overweight). Give them plenty of fruits, vegetables, and protein-containing foods (beans, lentils, eggs, meat, fish and milk products). If the child looks pale, check for signs of anaemia (see Section 57 Handbook 1), and make sure that they are given iron-rich foods (meat, eggs and dark green leafy vegetables) or take iron tablets (ferrous sulphate), as well as foods rich in vitamin C (oranges, lemons, tomatoes, etc.).

As far as possible, the child should learn to examine their own body for pressure sores every day, and eventually learn to take responsibility for all the necessary preventive measures themselves.

Other precautions

- To avoid pressure sores or other injuries developing on feet that have lost sensation, use well-fitted, well-padded sandals or shoes.
- Changing positions is important. When a child has recently had a spinal cord injury, they must be turned regularly, taking great care not to bend their back. Using a sheet under the body can help with turning.
- As the child gets stronger, hang loops and provide other aids, if necessary, so that they can learn to turn themselves.

At first it is important that the person turns, or is turned, at least every 2 hours day and night. Later, if there are no signs of pressure sores, the time between turns can gradually be lengthened to 4 hours. To avoid the child (or the person turning them) sleeping through the night without turning, an alarm clock can be very helpful.

When the child begins to sit or to use a wheelchair, there is a new and serious danger of pressure sores developing. The child must now get into the habit of taking the pressure off their bottom at least every 30 minutes. If their arms are strong enough, the child can lift up their whole body and hold it up for a minute or two. This allows the blood to circulate in the bottom.

If the chair has no arm rests, or if they can be removed, the child can lie sideways over a pillow on a high bed. They can rest for 15 to 30 minutes like this.

To prevent pressure sores, it is essential that the person who has lost sensation lies and sits on a soft surface that reduces pressure on bony areas. It is best for them to lie on a flat surface with a thick spongy mattress. A thick foam rubber mattress often works well. However, some foam is so spongy that it sinks completely under a person's weight, so that the bony area is not protected from the hard board underneath. A firm sponge with very small air bubbles (microcell rubber) works well but is expensive.

A 'waterbed' (a bag-like mattress filled with water) or air mattress also works well.

In some countries, an excellent mattress material is made from rubber-coated coconut fibre. Urine can be washed out of the material by pouring water through it. Because this material is costly, a rehabilitation programme in Bangladesh has adopted the practice of cutting a square out of a cheap mattress and replacing it with a square of the coconut fibre sponge.

Careful placement of pillows, pads or soft folded blankets can also help to prevent pressure sores. Such measures are especially important in the first weeks or months after a spinal cord injury, when the person must lie flat and be moved as little as possible. Pillows should be placed to avoid pressure on bony areas, and to keep the person in a position that is healthy and that helps to prevent contractures.

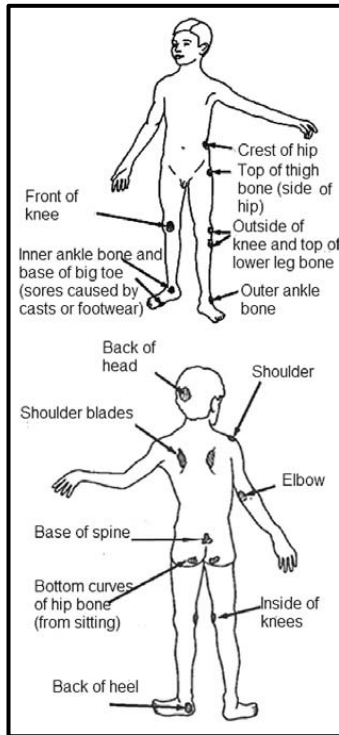


FIGURE 58.1 Sites where pressure sores are most likely to develop, with the most high-risk areas (in the hip region) labelled in type.

Chair and wheelchair cushions

For the child who has lost sensation in their bottom, the type of seat cushion used is very important, especially if the paralysis makes it difficult for them to lift up or change positions. All patients with spinal cord injury should use a good cushion. Sitting directly on a canvas seat or a poorly padded wooden seat will cause pressure sores.

Good cushions can be made of 'microcel' rubber, which is fairly firm. It works best if it is cut and shaped to reduce pressure on bony areas.

A useful low-cost way to make a fitted cushion is to build a base out of many layers of thick cardboard glued together, and then cover it with a 2 or 3 cm thick layer of sponge rubber.

Wet the cardboard and sit on it wet for 2 hours, so that it moulds to the shape of the bottom. Then let it dry and varnish it. If you do not feel normally in the seating area you will need to check the skin in the seating area every 30 minutes and dry it as the wetness and pressure will increase the risk of skin damage.

Before making a specially fitted cushion, you can make a 'mould' of the patient's bottom by having them sit in a shallow container of soft clay, mud or plaster. Note the bony hollows and form the seat to fit them.

Air cushions made from bicycle tyre inner tubes are excellent for the prevention of pressure sores, and for bathing on a hard surface. Use one, two or more tubes, depending on the size of the tube and the size of the child. Bind loops of the tubes together

with thin straps of inner tube. Then pump in enough air to ensure that the whole of the child's bottom is held up by air. (This idea was suggested by wheelchair rider-builders at Tahanang Walang Hagdanan (House With No Stairs), Quezon City, Philippines.)

Treatment of established pressure sores

Be alert for the first signs of a pressure sore by examining the whole body every day. Teach the child to do this using a mirror. If early signs of a pressure sore (redness, darkness, swelling or open skin) are observed, change body positions, and use padding to protect that area from pressure. For larger areas (such as the bones near the base of the spine), you can try using a small (motor scooter) inner tube to keep the weight off the sore area. Put a towel over the tube to soak up sweat, as sweaty skin against the rubber can also cause sores.

Warning: For small areas such as the heels, never use a ring or 'doughnut' of cloth to keep the weight off the sore, as this can cut off the blood supply to the skin inside the ring and make the sore worse.

If a pressure sore has already formed:

- Keep the pressure off the sore area completely and continuously.
- Keep the area completely clean. Wash it gently with clean or boiled water twice a day. Do not use alcohol, iodine, or other strong antiseptics.
- Make sure that the child has a healthy diet. If a large amount of liquid is lost from the sore, a lot of protein and iron will be lost with it. These must be replaced to allow quicker healing. The child should also take iron tablets if signs of anaemia are present, and they should eat foods rich in protein (beans, lentils, eggs, meat, fish, and milk products).
- Do not rub or massage areas where pressure sores might be forming, as this could tear weakened tissue and make the sore inside larger.

If the sore is deep and contains dead tissue within it:

- Clean the sore three times a day.
- Each time you clean the sore, try to scrape and pick out more of the dead rotten tissue. Often you will find that the sore is much larger inside than you first thought. It may go deep under the edges of the skin. Little by little remove the dead tissue until you come to healthy red flesh (or bone).
- Each time you have cleaned out the dead tissue, wash the sore out well with soapy water. Use liquid surgical soap if possible. Then rinse with clean (boiled and cooled) water. A syringe without a needle can help with irrigation.

If the sore is infected (signs of this include pus, foul smell, a swollen hot red area around the sore, or the presence of fevers and chills):

- Clean out the sore three times a day as described above.
- If possible, take the person to a clinical laboratory where a sample from the sore can be removed and cultured to find out what organisms are causing the infection and what is the most appropriate medication to treat it.
- If this is not possible, try treating the patient with penicillin, cloxacillin or flucloxacillin.

If the sore does not get better, or if liquid or pus keeps draining from a deep hole, the bone may be infected, so tell the parents to take their child to the hospital.

Honey and sugar

Once a pressure sore is free of dead tissue, filling it two to three times a day with honey or sugar helps to prevent infection and speeds up healing. This treatment, which was used by the ancient Egyptians and was recently rediscovered by modern doctors, works remarkably well. It is now being used in some hospitals in the UK and the USA.

To make it easier to fill the sore, mix honey with ordinary sugar until it forms a thick paste. This can easily be pressed deep into the sore. Then cover the sore with a thick gauze bandage.

It is important to clean out and refill the sore at least twice a day. If the honey or sugar becomes too diluted with liquid from the sore, it will feed the bacteria rather than kill them.

The amount of honey that is needed on the wound depends on the amount of fluid that is being produced by the pressure sore. If there is a lot of fluid, it will dilute the honey and make it less effective. The frequency of dressing changes required will depend on how rapidly the honey is being diluted. If there is no exudate, dressings need to be changed twice weekly to maintain the antibacterial properties of the honey as it enters the pressure sore. If the sore is producing a lot of fluid, the dressing will need to be changed twice a day.

To achieve the best results the honey should be applied to a dressing (cotton plus cellulose) which can absorb this prior to application. If applied directly to the wound, the honey tends to run off and be less effective. Honey will not soak easily into absorbent dressings. Soaking is helped by warming the honey to body temperature and/or adding 1 part of water to 20 parts of honey to make the honey more fluid. If the pressure sore is producing a lot of fluid, the absorbent dressing can be secured in place using cling film taped over it to help to keep the honey on the wound. Alginate dressings impregnated with honey are a good alternative to cotton/cellulose dressings, as the alginate is converted into a honey-containing soft gel. Any holes in the wound need to be filled with honey in addition to using a honey-impregnated dressing. As infection may be present in the tissues underlying the edges of the pressure sore, honey dressings need to extend beyond the inflamed area surrounding the sore.

Maintaining spinal alignment:

As the child grows, the spine may deform and likely to require a seating assessment and special support to stop the deformity increasing.

Further reading

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Active physiological conservative management in traumatic spinal cord injuries – an evidence-based approach – *Trauma Journal* 27th March 2017

<http://journals.sagepub.com/eprint/V9qda2SDWRT7fEMyTtqF>

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Accessed 29.03.2021

Section 59. Emergency Trauma Radiology

Introduction

Essential initial trauma films to screen for major injuries include the following:

- Lateral cervical spine radiograph
- Chest X-ray
- Pelvic X-ray

These should only be taken after immediately life-threatening injuries have been identified and treated (resuscitation).

The ABCD approach to X-ray interpretation is as follows:

- A. Adequacy, Alignment and Apparatus
- B. Bones
- C. Cartilage and soft tissues
- D. Disc spaces (in the spine), Diaphragm (in the chest)

First, all X-ray films should be checked for adequacy.

- Do they include all of the part that needs imaging?
- Is the film a proper antero-posterior view or is it at an angle?
- If the film is not of reasonable quality, interpretation is difficult and may be faulty.

Cervical spine

The cervical spine should be immobilised (see Section 94 Handbook 1) before any radiology. The standard film is a lateral radiograph, which may be supplemented by an AP (lower cervical spine and odontoid peg views) if appropriate.

Bony injury is not the primary focus in spinal injury. The main concern is to delineate actual or potential injury to the cord, as any unstable fracture, if inadequately immobilised, may lead to progressive cord damage.

A normal lateral cervical X-ray film may be falsely reassuring. The plain film only shows the position of the bones at the time when the film was taken and gives no idea of the magnitude of flexion and extension forces applied to the spine at the time of injury. The cord may be injured even in a child without any apparent radiographic abnormality. This phenomenon is known as SCIWORA (see below).

Unlike adult spine injuries, most paediatric cervical spine injuries occur either through the discs and ligaments, at the cranio-vertebral junction (C1, C2 and C3), or at C7/T1. The relatively large head of the child, moving on a flexible neck with weaker muscles, leads to injury in the higher cervical vertebrae.

Children show three patterns of spinal injury:

1. Subluxation or dislocation without fracture
2. Fracture with or without subluxation or dislocation
3. Spinal cord injury without radiographic abnormality (SCIWORA).

The last of these, SCIWORA, is said to have occurred when radiographic films are completely normal in the presence of significant cord injury. If the film is normal in a conscious child with clinical symptoms (such as pain, loss of function or paraesthesia in a limb), neck protection measures should be continued. In an unconscious child at high risk, a cord injury cannot be excluded until the patient is awake and has been assessed clinically, even in the presence of a normal cervical spine film. Adequate spinal precautions should be continued until the child is well enough to be assessed clinically.

The most common site of a 'missed' spinal injury is where a flexible part of the spine meets the fixed part. In the neck these are the cervico-cranial junction and the cervico-thoracic junction.

The whole spine should be viewed from the lower clivus down to the upper body of T1 vertebra.

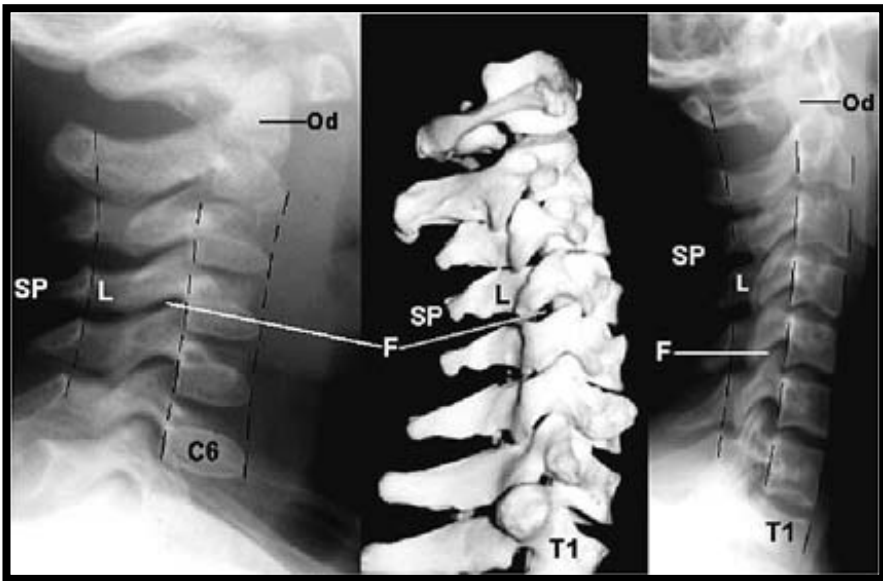
Alignment

When studying a cervical spine X-ray, look for the four lines shown in Figure 59.2. These lines should be uninterrupted. If there is a 'step' in any line, the spinal cord is at risk. The cervical immobilisation must be continued and an orthopaedic opinion sought.

The four lines are as follows:

1. Anterior vertebral line
2. Posterior vertebral line (anterior wall of the spinal canal)
3. Facet line
4. Spino-laminar line (posterior wall of the spinal canal).

FIGURE 59.1 X-ray of the lateral cervical spine, showing three of the lines that are indicated in Figure 59.2. Od, odontoid; F, facet joint; L, lamina; SP, spinous process spaces.



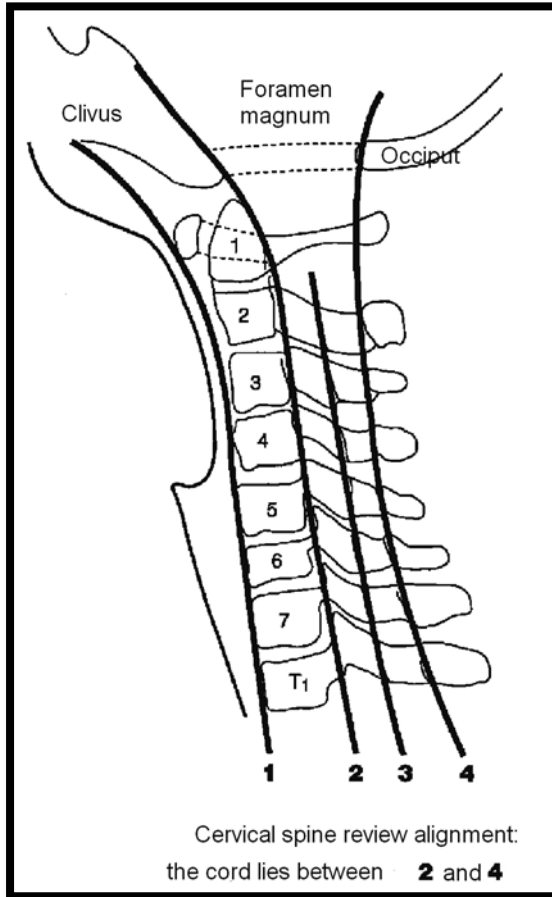


FIGURE 59.2 Lines to examine on X-ray of the lateral cervical spine.

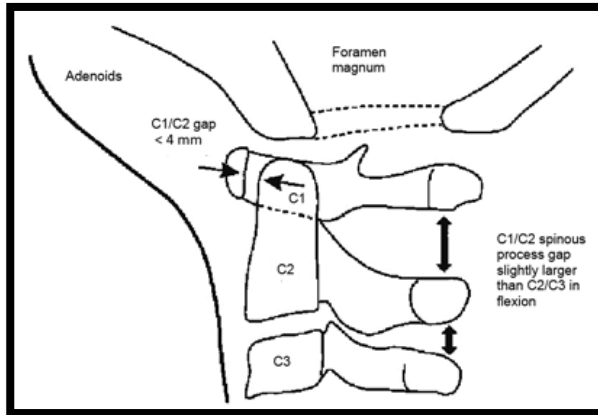
Figure 59.2 shows an actual cervical spine X-ray with three of the lines delineated and the odontoid, a facet joint, a spinous process, and a lamina identified. The gaps between the adjacent spinous processes and between each facet joint should be similar. Again, any discrepancy is suggestive of a potentially unstable spine. The spinal cord lies in the canal between the posterior vertebral line (2) and the spino-laminar (4) line.

Bones

The outline of each vertebra should be reviewed in turn. Fracture lines going through the cortex, vertebral bodies, laminae or spinous processes should be sought.

The spaces between the facet joints and the gaps between adjacent spinous processes should be similar. The joint between the odontoid peg and the anterior arch of the atlas should be 1 - 4 mm in a child (see Figure 59.3).

FIGURE 59.3 C1/C2 anatomy in the older child.



The orientation of the odontoid peg should always be perpendicular to the body of C2.

Cartilage and Soft Tissues

Abnormal widening of the pre-vertebral soft tissues may indicate a haematoma due to cervical spine injury. However, there may be a significant spinal injury with normal soft tissues. Thus, the absence of soft-tissue swelling does not exclude major bony or ligamentous injury. When a child is intubated, it is difficult to assess pre-vertebral soft-tissue swelling. Small children have large adenoids, which are seen as well-demarcated soft tissue swelling at the base of the clivus.

Acceptable soft-tissue thicknesses are as follows:

- Above the larynx: less than one-third of the vertebral body width
- Below the larynx: not more than one vertebral body width.

Below the level of the larynx, the pre-vertebral soft tissues become progressively narrower towards the cervico-thoracic junction (see Figure 59.4). If the pre-vertebral soft tissues are wider at C7 than at the C5 level, this suggests trauma at the C7/T1 level.



FIGURE 59.4 Lateral cervical spine showing soft tissues.

Any soft tissue swelling outside these limits should be regarded as abnormal, and neck protection measures maintained until a further clinical opinion can be obtained. In small children the soft tissues may appear abnormally wide if the film is taken with the infant lying in flexion. If in doubt, maintain the neck protection and ask for advice.

Discs

The height of the vertebral disc should be compared from C2/C3 to C7/T1. The discs should all be of similar height, as shown earlier in Figure 59.1. Any significant discrepancy suggests a crush fracture of the vertebrae (usually caused by a fall from a height). Flexion and extension cervical spine films should never be performed in the acute trauma situation.

Chest X-ray

Adequacy and alignment

Adequacy can be assessed by evaluating both radiographic penetration and the depth of the patient's inspiration. The film should just show the disc spaces of the lower thoracic vertebrae through the heart shadow. At least five anterior rib ends should be seen above the diaphragm on the right side. If the film is taken in expiration, it may mimic a chest infection. Films are difficult to take in young children, as they are unable to 'hold their breath' on command, so the radiographer has to try to take the picture at the moment of full inspiration.

Alignment can be assessed by ensuring that the medial ends of both clavicles are equally spaced about the spinous processes of the upper thoracic vertebrae. Abnormal rotation may create an apparent mediastinal shift. The trachea should be equally spaced between the clavicles.

Apparatus

Check the position of any apparatus, including the following:

- Tracheal tube
- Central venous lines
- Chest drains

Misplacement of the endotracheal tube (ETT) into a bronchus should be evident clinically but may be seen on a chest film if you look for it. Do this first when reviewing any chest X-ray on an intubated patient. Ventilation of only one lung will lead to hypoxia in a compromised patient.

The ideal position for an ETT is below the clavicles and at least 1 cm above the carina. To find the carina, identify the slope of the right and left main bronchi. The carina is where the two lines meet in the midline.

Bones

Look at each rib in detail. This can be done by tracing out the upper and lower borders of the ribs from the posterior costochondral joint to the point where they join the anterior costal cartilage at the mid-clavicular line. The individual internal bone patterns can then be assessed.

The ribs in children are soft and pliable, and only break when subjected to considerable force. Even greater force is required to fracture the first rib or to break multiple ribs. Consequently, the presence of these fractures should stimulate you to look for other sites of injury both inside and outside the chest. Fractures in children's rib bones are hard to see while fresh unless there is displacement. Diagnosis is often made a week or so later if an X-ray is taken then, when the calcifying new callus is seen.

Finish assessing the bones by inspecting the visible vertebrae and the clavicles, scapulae and proximal humeri. Thoracic spine injuries may be overlooked on a chest radiograph. Abnormal flattening of the vertebral bodies, widening of the disc spaces, or gaps between the spinous processes or pedicles may be seen. On the antero-posterior views, increased vertical or

horizontal distances between the pedicles or spinous processes indicate an unstable fracture, as shown in Figure 59.5.

If there are rib fractures in the first three ribs, these may be associated with major spinal trauma and great vessel injury.

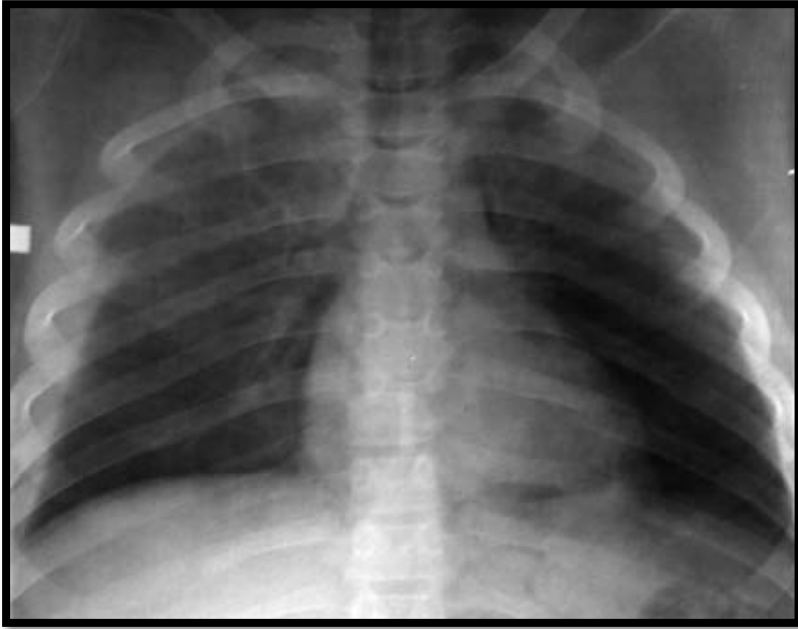


FIGURE 59.5 Vertical fracture of the thoracic spine.

Cartilage and Soft Tissues

Lungs

In a well-centred film, the lungs should appear equally black on both sides. Compare the left and right lungs in the upper third, middle third and lower third of the chest.

Check that the lungs go all the way out to the rib cage (i.e. that there is no pleural effusion or pneumothorax). A lung that is black on one side may be due to a pneumo-thorax or air trapping. A lung that is white on one side may be due to collapse, pulmonary haemorrhage, contusion or effusion (including haemothorax).

On the supine film, blood or fluid lies posteriorly, giving a generalised greyness to the lung, rather than the typical meniscus sign seen on the erect film. At the apex of each lung, an effusion displacing the lung downward may indicate spinal injury or major vessel damage.

A suspected tension pneumothorax should be treated clinically in the emergency situation, without confirmatory X-ray.

On a supine film, the air in a simple pneumothorax rises anteriorly and may only be evident from an abnormal black-ness or 'sharpness' of the diaphragm or cardiac border. The standard appearances of a pneumothorax, where there is a sharp lung edge and the vessels fail to extend to the rib cage and the lung edges, may not occur in the supine film.

The Heart

The cardiac outline should lie one-third to the right of the midline and two-thirds to the left of the midline. If the film is not rotated, which should be checked, mediastinal shift is due to the heart being either pushed from one side or pulled from the other. For example, mediastinal shift to the left may be due to a pneumothorax, air trapping or effusion on the right side, or collapse of the left lung.

All emergency major trauma X-rays are taken in the supine position because of the seriousness of the patient's condition, often using portable X-ray machines. The X-ray tube is near to the patient and the heart is anterior with the film posterior. The heart in this situation appears abnormally magnified (widened), and the cardiothoracic ratio is difficult to assess on supine AP films.

The mediastinal cardiac outline should be clear on both sides. Any loss of definition suggests consolidation (de-aeration) of adjacent lungs. A 'globular' shape to the heart may suggest a pericardial effusion. Tamponade is managed clinically. A cardiac ultrasound scan is useful in equivocal cases.

The upper mediastinum

In the teenager the mediastinum should appear as narrow as in an adult. In children under the age of 18 months, the normal thymus is large, causing a confusing and often 'sail-shaped' upper mediastinal shadow. A normal thymus may touch the right chest wall, left chest wall, left diaphragm, or right diaphragm, making it very difficult to exclude mediastinal pathology. Fortunately, mediastinal widening due to aortic dissection or spinal trauma is very rare in small children.

In the older child involved in trauma, mediastinal widening may mean aortic dissection, or major vessel or spinal injury. Ultrasound scanning will be helpful (if available).

Diaphragms

The cardio-phrenic and costo-phrenic angles should be clear on both sides. The diaphragms should be clearly defined on both sides, and the left diaphragm should be clearly visible behind the heart. Loss of definition of the left diaphragm behind the heart suggests left lower lobe collapse, an abnormal hump suggests diaphragmatic rupture, and an elevated diaphragm suggests effusion, lung collapse or nerve palsy.

At the end of the systematic ABCD review of the X-ray, check again in the key areas shown in the following list:

- Behind the heart: Left lower lobe consolidation or collapse.
- Apices: For effusions, pneumothorax, rib fractures and collapse or consolidation.
- Costophrenic and cardio-phrenic angles: Fluid or pneumothorax.
- Horizontal fissure: Fluid or elevation (upper lobe collapse).
- Trachea for foreign body (and endotracheal tube)).

Pelvic X-ray

A single, antero-posterior pelvic view is sufficient.

Adequacy and alignment

It is very important to have the pelvic film positioned as a true antero-posterior (AP) view, as rotation causes interpretation problems. In a true AP film, the tip of the sacrum will be aligned with the symphysis pubis.

The whole of the pelvis from the top of the iliac crests to the ischial tuberosities and both hip joints should be seen. The femoral necks shown to the level of the trochanters should be included.

Bones

The pelvis is composed of the sacrum, innominate bones (iliac wings), ischium and pubic bones. These come together to form a Y-shaped cartilage in the floor of the acetabulum. In young children, the joint between the ischium and the pubis (ischiopubic synchondrosis) is commonly seen and may simulate a fracture.

FIGURE 59.6 Normal pelvis in a young child.



The pelvis is reviewed as a number of rings on the two-dimensional film. These include the pelvic brim, the two obturator rings and both acetabular fossae. The rings should appear smooth and symmetrical in a well-centred film (see Figure 59.6, which shows a normal child pelvis). The femoral necks must be checked for fracture.

Figure 59.7 shows a pelvis with multiple fractures, at major risk of serious pelvic bleeding as large vessels are torn with the force shown by the widespread fractures.



FIGURE 59.7 Multiple pelvic fractures.

Cartilage and soft tissues

Minor rotation, hip flexion or rotation will distort the fat plane and make assessment of soft-tissue displacement difficult. Abnormal widening of the obturator fat pad may indicate a pelvic side wall haematoma.

The paediatric pelvis is held together by cartilage. Separation through the cartilage of the sacro-iliac joint, the symphysis pubis or the 'Y' cartilage of the acetabular floor may occur without apparent bony injury. Comparison of both hips and sacro-iliac joints on a well-centred film may show this. On a well-centred film, the distance between the femoral head and the floor of the acetabulum 'crescent' should be symmetrical – it is abnormal in effusion or dislocation of the hip joint.

Section 60. Tracheal intubation

Aims

- to secure the airway
- to protect the airway
- to facilitate prolonged and intra-operative ventilation
- for tracheo-bronchial toilet
- in the application of high airway pressures and positive end-expiratory pressure (PEEP)
- during cardiopulmonary resuscitation to improve ventilation and allow uninterrupted chest compressions.

Choice of tube

An uncuffed tube is often recommended in children who weigh less than 25 kg, as the larynx is narrowest below the glottis at the circular non-distensible cricoid ring, and inexperienced use of the cuffed tube may cause damage at that point, although the cuffed tube gives better airway protection. The choice ultimately depends on the experience of the practitioner (see also Section 13).

The correctly sized tube is one that passes easily through the glottis and subglottic area with a small air leak detectable at 20 cm H₂O (sustained gentle positive pressure).

Size of tube

The correct size of tube is:

- one that can just fit into the nostril or
- in preterm neonates, 2.5–3.5 mm internal diameter or
- in full-term neonates, 3.0–4.0 mm internal diameter or
- in infants after the neonatal period, 3.5–4.5 mm internal diameter or
- in children over 1 year:
 - the internal diameter (in mm) is $\text{age}/4 + 4$
 - the length of tube (in cm) is $\text{age}/2$ plus 12 for an oral tube, and $\text{age}/2$ plus 15 for a nasal tube.

Aids to intubation

- Laryngoscope: blade (straight for neonates and infants because of long, floppy epiglottis, curved for older children), bulb and handle.

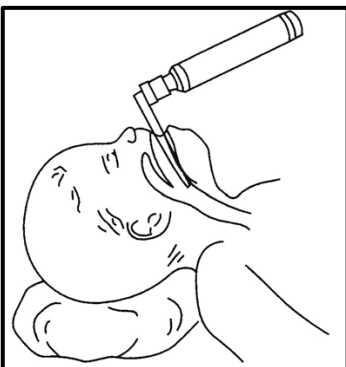


FIGURE 60.1 Straight-blade laryngoscope, suitable for infants.

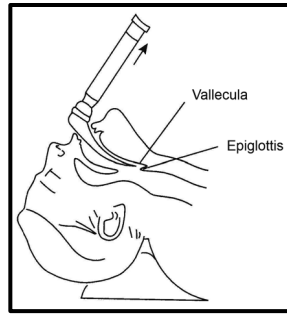


FIGURE 60.2 Curved-blade laryngoscope, suitable for children.

- Magill's forceps.
- Introducer (not further than the end of the tube itself).
- Syringe (for cuffed tube).
- Gum elastic bougie (over which the tube can pass).
- Suction apparatus must be immediately available.

Predicting difficulty with intubation

1. Difficulty in opening mouth.
2. Reduced neck mobility.
3. Laryngeal/pharyngeal lesions.
4. Congenital: Pierre-Robin syndrome, mucopolysaccharidoses.
5. Acquired: burns, trauma.

If on viewing the child's face from the side, the chin is unusually small (micrognathia), the intubation will be difficult, and senior help is required (but see below).

Complications

1. Displacement: oesophageal, endobronchial, out of larynx!
2. Obstruction: kinking, secretions.
3. Trauma: lips to larynx.
4. Hypertensive response.
5. Spasm: laryngeal, pharyngeal.
6. Aspiration: gastric contents.

Procedure

Prepare and check the equipment.

1. Choose an appropriate tube size, with one size above and one size below it available.
2. Get the tape ready to fix the tube.
3. Suction must be available.
4. Induce anaesthesia and give a muscle relaxant unless the patient is completely obtunded.
5. Do not attempt the procedure in a semi-conscious child.

Position the child.

1. Children over 3–4 years of age: the 'sniffing morning air' position (head extended on the shoulders and flexed at the neck).
2. Children under 3 years of age (especially neonates and infants): a neutral position (large occiput).

3. Keep the child in a neutral position with in-line immobilisation in the case of unstable cervical spine (e.g. trauma, Down's syndrome).

Oxygenate the child using a face mask and reservoir (if patient is breathing) or bag and mask ventilation to provide high flow oxygen.

1. Introduce the laryngoscope into the right side of the mouth.
2. Sweep the tongue to the left.
3. Advance the blade until the epiglottis is seen.
4. Curved blade: advance the blade anterior to the epiglottis. Lift the epiglottis forward by moving the blade away from your own body.
5. Straight blade: advance the blade beneath the epiglottis, into the oesophagus. Pull back, and the glottis will 'flop' into view.

Recognise the glottis.

- Insert the endotracheal tube gently through the vocal cords.
- Stop at a predetermined length.

Confirm the correct placement.

- The chest moves up and down with ventilation.
- Listen to breath sounds in the axillae and anterior chest wall.
- Confirm that there are no breath sounds in the stomach.
- Oxygen saturations do not go down.
- Carbon dioxide is measured from expired gases (if available: ideal).

Secure the tube.

Secure with tape around the tracheal tube and on to the patient's face (see below).

Nasal intubation

Although oral intubation is quicker and more reliable in an emergency, for prolonged ventilation nasal intubation is preferable, if a skilled operator is available, as the tracheal tube is more securely fixed. The technique is similar, but with the additional use of the Magill's forceps to grasp and guide the tracheal tube as it emerges into the posterior pharynx downward into the trachea through the vocal cords.

Fixation of endotracheal tubes

Two people should be available to do this, one of whom should hold the tube at all times.

Cut two strips of sticky zinc oxide tape (see below); they should reach from just in front of the ear across the cheek and above the upper lip to the opposite ear.

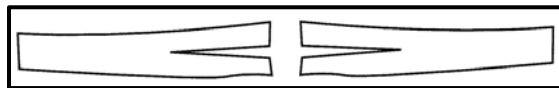


FIGURE 60.3 Tape for tracheal tube fixation.

1. If available, apply some benzoin tincture to the cheeks, above the upper lip and under the chin, which will make the tape stick well.
2. Make sure that the endotracheal tube is clean and that no old tape is left on it.
3. Start with the broad end of the tape and stick this on to the cheek. Then wrap one of the thinner ends carefully around the tube. It is useful if it is still possible to see the endotracheal tube marking at the lips.
4. Tape the other half across the philtrum to the cheek.

5. The second tape starts on the other cheek, and the thinner half is stuck across the chin, while the other half is also wrapped around the tube (see below).

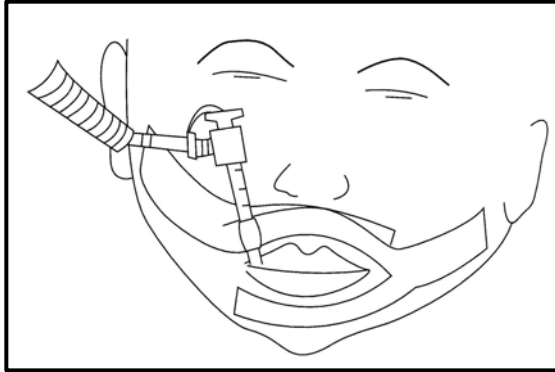


FIGURE 60.4 Taped tracheal tube.

Section 61. Microscopy of urine

Introduction

Urinary tract infections (UTIs) are common in children. Although many of these infections are not serious, some of them cause kidney damage and lead to scarring. 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 it at once.

Urine microscopy is the only way to diagnose UTIs immediately and reliably.

In a patient with a UTI the urine contains: one species of bacterium at a concentration of at least 100 000/mL and an excess of white blood cells.

Bacterial numbers

Most children with a UTI have in the range of 10–1000 million bacteria/mL. In fact, 100 000/mL is a very small number of bacteria. 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 including invasive procedures.

White blood cells

Children frequently have extra urinary white blood cells without a UTI.

- Around 10% of febrile children have hundreds of extra white blood cells.
- Girls void some urine into the vagina, so vaginal white blood cells are readily washed into the urine (as are vaginal epithelial cells, which are seen in the urine of most girls after puberty).

Children with UTIs often have no excess of white blood cells.

- White blood cells do not last long in urine, especially if it is alkaline, so it must be examined soon after collection.
- Ill infants may be unable to mount a white blood cell response.

Therefore, white blood cells alone are an unreliable and potentially misleading sign.

How to count bacteria

Laboratory culture

This is the most widely used method, and the traditional approach. It remains acceptable, but, if you use it, you will:

- have to accept that some positive reports will be false
- have to wait at least 48 hours for the result. In real life, 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 within 3 days
- 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
- miss the occasional UTI caused by anaerobes.

Advantages of urine microscopy

If you use this method, you can:

- discard sterile urines, and reassure the child's family at once

- repeat a contaminated urine sample at once
- treat children with UTIs immediately
- diagnose anaerobic UTIs as easily as aerobic ones
- save time and money because it is quicker and cheaper than urine culture.

Choice of microscope

With an ordinary light microscope, bacteria are only easy to see after they have been stained.

Phase-contrast microscopes 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 to do urine microscopy

1. You can microscope fresh urine on a slide with a counting chamber. There is no need to stain or spin the urine.
2. The slide has two chambers, each of which has a grid etched on to the glass surface. In certain clinical situations, such as examination of peritoneal dialysis fluid for suspected peritonitis, the grid can be used to make accurate counts of the concentrations of elements present.
3. Usually, this degree of accuracy is unnecessary. However, the grid is always useful because it confirms that the microscope is 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.
4. 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 (see Figure 61.1).

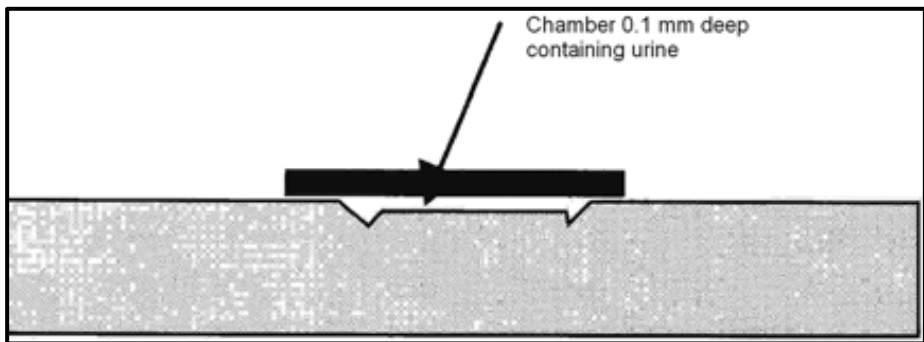


FIGURE 61.1 Side view of grid slide with coverslip in place.

5. Test the urine with a dipstick (to check for blood, protein and glucose). Then touch the tip of the dipstick on the slide so that a small amount of urine is drawn into the chamber by capillary action.

Bacteria

- Most bacteria that cause UTIs are bacilli (rod-shaped).
- They are easy to identify, as they look like straight lines, usually about 3 mm long.

- Mostly they remain still, or just move slightly, like a shimmer. This movement is caused by Brownian motion (which occurs when they are hit by water molecules) and is not due to them swimming.
- Rarely will you see moving bacteria.

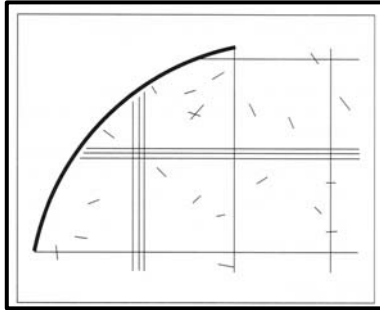


FIGURE 61.2 Rod-shaped bacteria.

Infections also sometimes occur with streptococci, which are bacteria that resemble strings of beads. There are always some strings that are four or more cocci long. If you think that you can see 'cocci' individually, or in clumps, these are in fact phosphate crystals. If they appear to be moving, this is just the result of Brownian motion.

White blood cells

These are round, and between 3 and 5 mm in diameter. All white blood cells have a 'granular' appearance to their cytoplasm. In the case of the larger ones, you can often make out the individual granules shimmering and moving within the cell, and the nucleus (which is lobed in neutrophils).

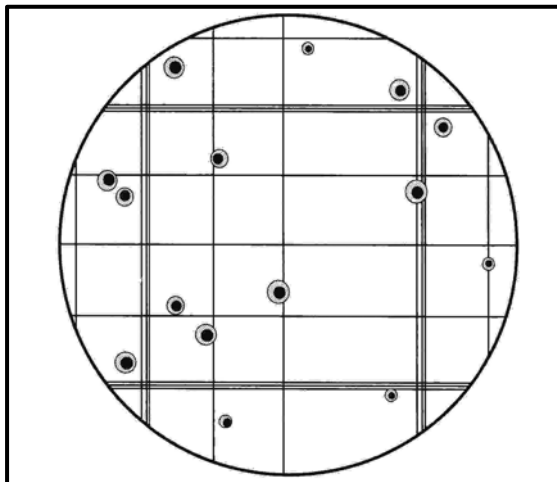


FIGURE 61.3 White blood cells.

Red blood cells

These are smaller than white blood cells, and do not have any content or granular appearance.

If the red cells are present because of trauma (for example after an injury or post-surgery) or a UTI, they will either look just like red cells in the blood (i.e. biconcave discs), or they will all appear slightly shrunken and wrinkled, or slightly swollen. The important thing is that they all look the same.

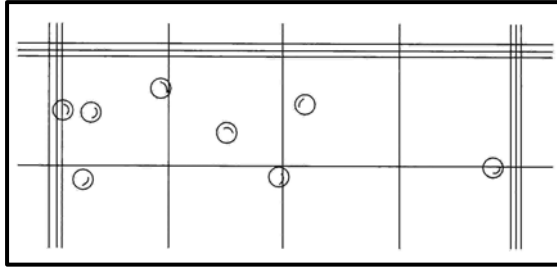


FIGURE 61.4 Red blood cells.

If the red cells are in the urine because of kidney inflammation (glomerulonephritis), they are usually smaller, but they 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.

Epithelial cells

These are very large flat cells with an easily visible round nucleus. They are from the vagina and are only seen in the urine of older girls, in which they are common. If large numbers of epithelial cells are present this suggests particularly heavy vaginal contamination.

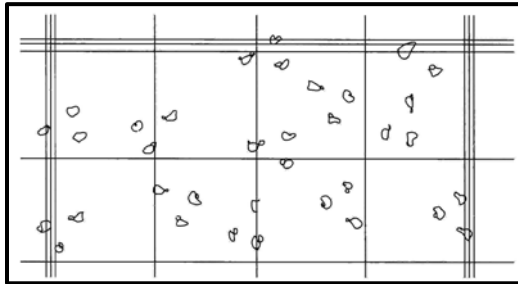


FIGURE 61.5 Glomerular red blood cells.

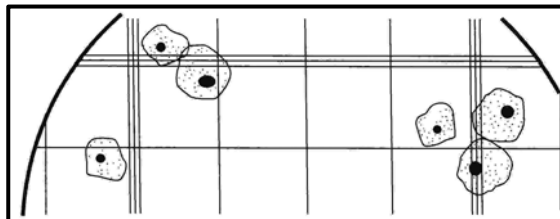


FIGURE 61.6 Epithelial cells.

Casts

These indicate kidney inflammation (glomerulonephritis). Casts consist of abnormal kidney tubule contents that have solidified and have retained the shape of the tubule as they passed into the urine.

Pure protein casts look glass-like and are described as hyaline. Those consisting of debris (for example, dead tubule cells in acute tubular necrosis) are called granular casts. Some casts are composed of red or white cells. Many casts consist of a mixture of these.

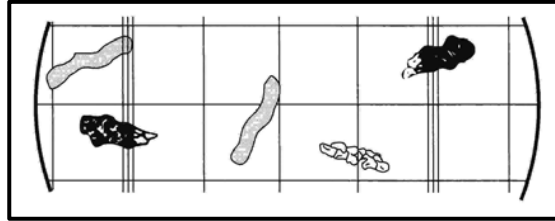


FIGURE 61.7 Casts.

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.

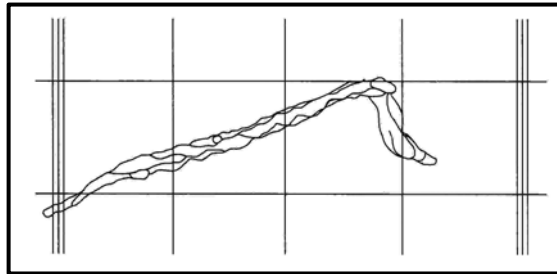


FIGURE 61.8 Debris.

Crystals

Urine samples often contain obvious crystals, whose shape allows their chemical origin to be identified. However, this is rarely of clinical significance.

The commonest 'crystals' in fact look more like small black dots, either singly, or in clumps (and even in casts). They move slightly (or 'shimmer') as a result of Brownian motion and can be mistaken by the unwary for small round bacteria (cocci).

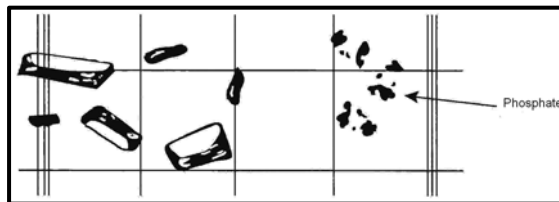


FIGURE 61.9 Crystals.

Diagnosing urinary tract infections (UTIs)

UTIs are primarily diagnosed by looking for bacteria.

Infected urine

About 99% of urine infections are caused by rod-shaped bacteria known as bacilli.

In most UTIs, every field you view will have many bacteria (in some cases thousands), and they all look the same. Therefore, when you see many bacteria in fresh urine, all with the same appearance, you can be sure that the child has a UTI.

If you see at least one rod, but less than 10 rods in the centre of the grid (square 5), you have to consider the possibility of contamination, so collect another sample to see whether the finding persists (and think about vaginal lactobacilli; see below).

What to do if you find a positive microscopy

You can start treatment immediately with an appropriate antibiotic. In addition, send the urine for culture with direct sensitivities.

The laboratory will grow the bacteria to confirm which species they are, and to test their sensitivity to a range of different antibiotics. Without direct sensitivity testing this takes 2 days, but with it you will usually obtain the result the next day.

Sterile urine

Most urine samples will be sterile. If you see no bacteria or cells, check by looking at five 'size-A' squares (i.e., about five fields).

If you see nothing in that area, then you can be certain that the urine is not infected.

Even if you can see other elements, if there are no bacteria, it is not a UTI.

Remember that you will see white blood cells in the urine of many children with fever (e.g. due to tonsillitis or pneumonia). Also remember that many girls have white blood cells in their urine from the vagina (and often epithelial cells, too).

Contaminated urine

If you see any of the following, collect a repeat sample, as the first sample is likely to have been contaminated:

- more than one shape of bacterium
- some bacteria, but also a large amount of debris (for example cotton fibres or many epithelial cells)
- many bacteria in a urine sample that was collected several hours ago, or from a nappy that had been on the baby for several hours.

If necessary, you need to go on collecting repeat urine samples until one is either definitely sterile or definitely infected.

Vaginal contamination

Girls 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 (see Figure 61.6).

Also, in many older girls, lactobacilli are washed 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, ask the lab either to Gram stain them or to culture them. Unlike the bacteria that cause UTIs, lactobacilli 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 on the clinical notes. This is important because negative urine samples will be discarded, and this will be the only record of the test.

A typical format is as follows:

URINE PHASE CONTRAST MICROSCOPY

Name: Date:

MICROSCOPY – Bacteria: WBC: RBC:
..... Casts, etc.:

STICKS – Protein: Blood: Glucose:
Other:

ACTION – (tick one of the three options)

1. Urine not infected: sample discarded
2. Urine contaminated: sample repeated
3. UTI: urine sent for culture and direct sensitivities, and antibiotics started

SIGN and PRINT NAME:

Counting what you see

- For most clinical purposes it is not necessary to count the exact concentration of cells or bacteria that you see and estimates such as 'many' or 'few' are enough.
- Sometimes it is helpful to quantify the findings more carefully (for example, to monitor the numbers of casts in a child with glomerulonephritis).
- Occasionally it is essential to count the exact numbers (for example, the number of white blood cells is critical for the diagnosis and treatment of peritonitis in children on peritoneal dialysis from a dialysis sample).

Calculate all the counts per microlitre (μL). Count at least 10 of each element of interest. The number and size of the squares you need to count will therefore depend on the concentration of the elements in the urine.

Figure 61.10 shows the etched counting grid for microscopy.

- The central square ('3') is 1×1 mm.
- With the coverslip on, the chamber is 0.1 mm deep, so the central square has a volume of $0.1 \mu\text{L}$.
- Therefore, the whole grid of nine similar squares has a total volume of $0.9 \mu\text{L}$.
- Note that 1 microlitre is one-thousandth of a mL.
- Therefore, a count of 100,000 bacteria/mL is equivalent to $100/\mu\text{L}$, so a 'significant' culture in a urinary tract infection would mean at least 100 bacteria/ μL , or 10 bacteria in the central square of the grid.

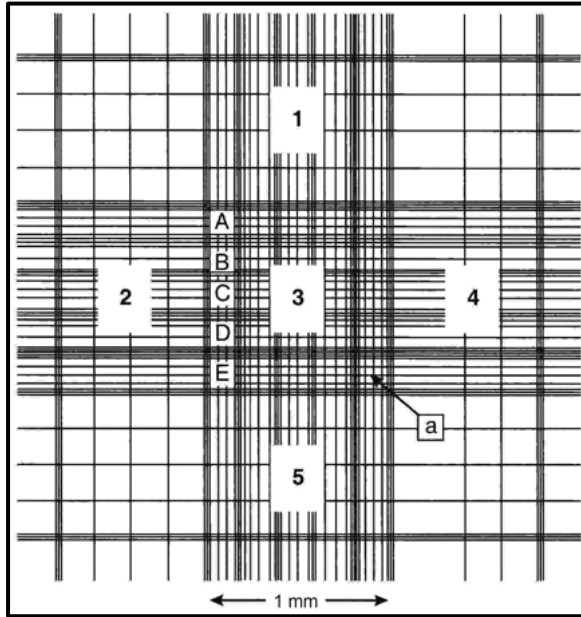


FIGURE 61.10 Counting grid for microscopy.

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 square 5, and multiply by 10.

Frequent elements

Count all those in five smaller squares (e.g. squares 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 and multiply by 4000.

Section 62. Chest Physiotherapy

Therapy for bronchiectasis, cystic fibrosis, and other conditions with excess airway secretions as well as post operatively.

Postural drainage

This is positioning to allow drainage by gravity from lung segments to central airways.

For infants, use a maximum of five positions in 10 minutes, progressing in older children to two to three positions in up to 30 minutes.

Positions for postural drainage of secretions

Upper lobe

Apical segments: sitting (1).

Posterior segments: prone, one pillow below the affected side (2).

Anterior segment: supine (3).

Middle lobe/lingual

Chest tipped 15 degrees below the horizontal, lying supine, with a pillow supporting the ipsilateral hip and shoulder (4).

Lower lobe

Apical segments: prone (5).

Anterior basal: chest tipped 20 degrees below the horizontal, lying supine (6).

Lateral basal: chest tipped 20 degrees below the horizontal, lying on the unaffected side (7).

Posterior basal: chest tipped 20 degrees below the horizontal, lying on the unaffected side (8).

Figure 62.1 shows all of these positions in sequence.

Equipment

Carer's lap (in the case of an infant), otherwise bean bags, pillows or a tilted bed.

Adjuncts to postural drainage

The following may be combined with postural drainage:

- chest clapping: done over the 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 withhold, and relaxed expiration. Huff – that is, forced expiration at mid to low lung volumes with the glottis open (as if misting glass), cough to clear secretions, and repeat the cycle until the chest is clear.

Note that where bronchoconstriction is an issue:

- 1 Increase the amount of time spent doing tidal volume breathing.
- 2 Omit percussion.
- 3 Increase tidal volume breathing and omit percussion.

Consider the use of inhaled bronchodilators (e.g. salbutamol 200–500 micrograms inhaled through a spacer) (see Section 35 Handbook 1).

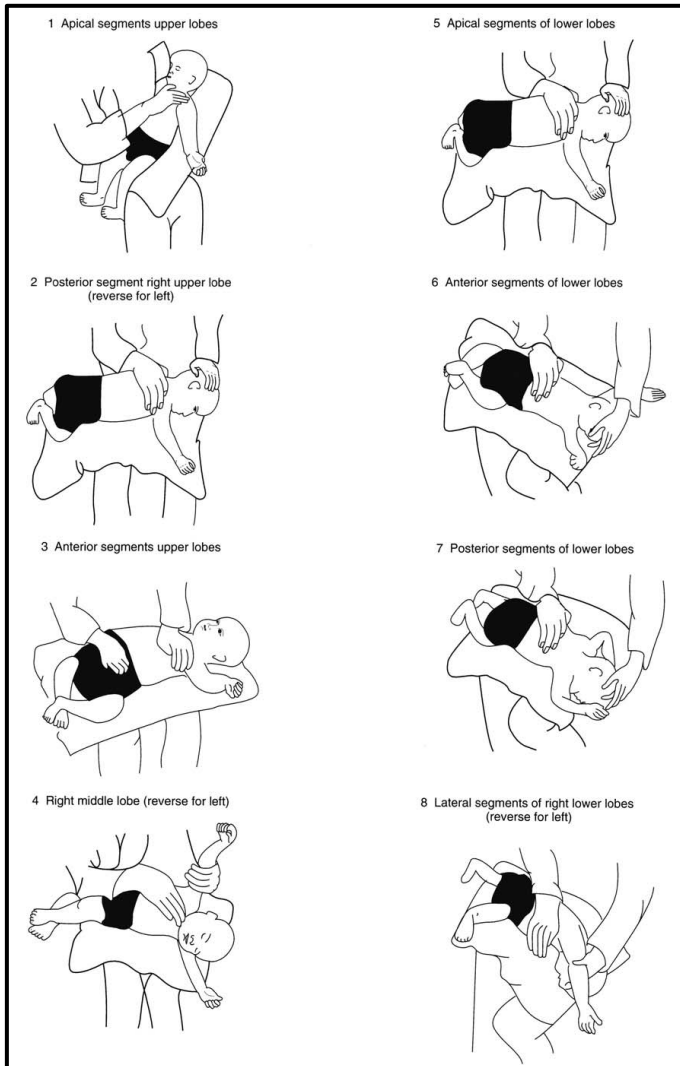


FIGURE 62.1 *Positions for postural drainage.*

Relative contraindications to chest physiotherapy

These include the following:

- raised intracranial pressure
- severe hypertension
- after abdominal surgery
- after major haemoptysis
- pulmonary oedema
- surgical emphysema
- after treatment of tension pneumothorax
- cardiac arrhythmias
- gastro-oesophageal reflux (only omit postures with upper body dependent).

Patient positioning

To maximise ventilation–perfusion matching (for example, in pneumonia, asthma, pneumothorax) in self-ventilating patients, position with the better ventilated lung uppermost.

In severely breathless patients, use sitting with a forward lean, or the recovery position.

Use pillows to raise and support the chest if the patient cannot tolerate lying flat.

Section 63. WHO treatment plans A, B and C for acute diarrhoea 2005

WHO Treatment Plan A: home therapy to prevent dehydration and malnutrition

Children with no signs of dehydration need extra fluids and salt to replace their losses of water and electrolytes due to diarrhoea. If these are not given, signs of dehydration may develop.

Mothers should be taught how to prevent dehydration at home by giving the child more fluid than usual, how to prevent malnutrition by continuing to feed the child, and why these actions are important. They should also know what signs indicate that the child should be taken to a health worker. These steps are summarised in the four rules of Treatment Plan A.

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:

Fluids that normally contain salt, such as:

- ORS solution
- salted drinks (e.g. salted rice water or a salted yoghurt drink)
- vegetable or chicken soup with salt.

Teaching mothers to add salt (about 3 g/L) to an unsalted drink or soup during diarrhoea is also possible but requires a sustained educational effort.

A home-made solution containing 3 g/L of table salt (one level teaspoon) and 18g/l of common sugar (sucrose) is effective but is not generally recommended because the recipe is often forgotten, the ingredients may not be available or too little may be given.

Fluids that do not contain salt, such as:

- plain clean water
- water in which a cereal has been cooked (e.g. unsalted rice water)
- unsalted soup
- yoghurt drinks without salt
- green coconut water
- weak tea (unsweetened)
- unsweetened fresh fruit juice.

Unsuitable fluids

A few fluids are potentially dangerous and should be avoided during diarrhoea. Especially important are drinks sweetened with sugar, which can cause osmotic diarrhoea and hypernatraemia. Some examples are:

- commercial carbonated beverages
- commercial fruit juices
- sweetened tea.

Other fluids to avoid are those with stimulant, diuretic or purgative effects, for example

- coffee
- some medicinal teas or infusions.

How much fluid to give

The general rule is to give as much fluid as the child wants until the 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 up to 10 years: 100–200 mL (a half to one large cup)
- older children and including those who are pregnant: as much fluid as they want.

Rule 2: Give supplemental zinc (10–20 mg) to the child every day for 10 to 14 days

Zinc can be given as a syrup or as dispersible tablets; whichever formulation is available and affordable. By giving zinc as soon as diarrhoea starts, the duration and severity of the episode as well as the risk of dehydration will be reduced. By continuing zinc supplementation for 10–14 days, the zinc lost during diarrhoea is fully replaced and the risk of the child having new episodes of diarrhoea in the following 2 to 3 months is reduced.

Rule 3: Continue to feed the child, to prevent malnutrition

The infant's usual diet 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. These children should be encouraged to resume normal feeding as soon as possible.

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 six 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 (if fluids other than breastmilk are given, use a cup, not a bottle). This approach usually takes about 1 week. If possible, infants 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 old 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, 5–10 mL of vegetable oil should be added to each serving of cereal. (Most staple foods do not provide enough calories per unit weight for infants and young children. This is improved by adding some vegetable oil.) 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 2 weeks. If the child is malnourished, extra meals should be given until the child has regained normal weight for height.

Rule 4: Take the child to a healthcare worker if there are signs of dehydration or other problems

The mother should take her child to a healthcare 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
- does not get better in 3 days.

WHO Treatment Plan B: oral rehydration therapy for children with some dehydration

Children with some dehydration should receive oral rehydration therapy with ORS in a healthcare facility following the treatment plan described below.

Children with some dehydration should also receive zinc supplementation as described above.

TABLE 63.1 Guidelines for treating children with some dehydration: approximate amount of ORS to give in the first 4 hours

Age	< 4 months	4–11 months	12–23 months	2–4 years	5–14 years	15 years or older
Weight (kg)	< 5	5–7.9	8–10.9	11–15.9	16–29.9	30 kg or more
Volume (mL)	200–400	400–600	600–800	800–1200	1200–2200	2200–4000

How much ORS is needed?

Use Table 63.1 to estimate the amount of ORS needed for rehydration. If the child’s weight is known, this should be used to determine the approximate amount of solution needed. The amount may also be estimated by multiplying the child’s weight in kg by 75 mL. 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 the child wants more than the estimated amount of ORS, and there are no signs of over-hydration, give more.*

Oedematous (puffy) eyelids are a sign of over-hydration. They may be a sign of chronic malnutrition. If this occurs, stop giving ORS, but give breast milk or plain water, and food. Do not give a diuretic. When the oedema has gone, resume giving ORS or home fluids according to Treatment Plan A.

How to give ORS

A family member should be taught to prepare and give ORS. 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, as 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 again, but more slowly (e.g. a spoonful every 2–3 minutes).

Monitoring the progress of oral rehydration therapy

Check the child from time to time during rehydration to ensure that ORS is being taken satisfactorily and that signs of dehydration are not worsening. If at any time the child develops signs of severe dehydration, switch to WHO Treatment Plan C.

After 4 hours, reassess the child fully, following guidelines in Table 61.2 Handbook 1. Then decide what treatment to give next:

- If signs of severe dehydration have appeared, intravenous (IV) therapy should be started following Treatment Plan C. This is very unusual, however, occurring only in children who drink ORS poorly and pass large watery stools frequently during the rehydration period.
- If the child still has signs indicating some dehydration, continue oral rehydration therapy by repeating Treatment Plan B. At the same time start to offer food, milk and other fluids, as described in Treatment Plan A (see above) and continue to reassess the child frequently.

Table 63.2 Signs of Dehydration (From Section 61 Handbook 1)

	Symptoms and signs present	Treatment
No dehydration	None Increased thirst	<ul style="list-style-type: none"> • Treat at home with extra fluids. WHO Treatment Plan A • Breastfeeding or standard diet must continue • Warn mother about danger signs of some or severe dehydration and when to return • Zinc supplements
Some dehydration (5–9% fluid deficit)	Two or more of the following <ul style="list-style-type: none"> • Restless and irritable • Sunken eyes • Drinks eagerly/thirsty • Loss of skin turgor; tents when pinched and goes back slowly • Any one additional sign of severe dehydration below 	Treat with WHO Treatment Plan B in hospital for at least 24 hours (if feasible) Give ORS or ReSoMal if there is malnutrition Breastfeeding or standard feeding to continue

	Symptoms and signs present	Treatment
Severe dehydration (10% or greater)	<p>Two or more of the following</p> <ul style="list-style-type: none"> • Prostration • Sunken eyes • Loss of skin turgor; tents when pinched and goes back slowly (≥ 3 seconds) • Not able to drink or drinks poorly <p>In addition, may show:</p> <ul style="list-style-type: none"> • rapid deep breathing from acidosis • Lack of urine output 	<p>Treat with WHO Treatment Plan C</p> <ul style="list-style-type: none"> • Rapid IV rehydration, giving ORS while IV cannula is put in place • Test for and treat any hypoglycaemia • Breastfeeding or standard feeding as soon as possible • Zinc supplements
Shock	<p>As above with:</p> <ul style="list-style-type: none"> • High and increasing heart rate; weak pulse volume • Poor skin circulation time (cool and poorly perfused extremities) with prolonged capillary refill time (> 3 seconds) • Low or even unmeasurable blood pressure • Reduced conscious level or coma 	<ul style="list-style-type: none"> • Urgent IV or intra-osseous access • Urgent IV/intra-osseous fluid bolus of 10 mL/kg Ringer-lactate/ Hartmann's solution or 0.9% saline • Repeat 10 mL/kg boluses if remains shocked, up to a total of 40 mL/kg, then beware of fluid overload • Then rehydrate more slowly • Use NG or oral ORS/breast milk as soon as tolerated

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 and food following Treatment Plan A. Give the mother enough ORS sachets for 2 days. Also teach her the signs that mean she should bring her child back.

- Use the patient's age only when you do not know their 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 is shown above, give more.
- Encourage the mother to continue breastfeeding her child.
- For infants under 6 months who are not breast fed, if using the old WHO ORS solution containing 90 mmol/L of sodium also give 100–200 mL clean water during this period.

However, if using the new reduced (low) osmolality ORS solution containing 75 mmol/L of sodium, this is not necessary.

Note: During the initial stages of therapy, while still dehydrated, children who are pregnant can consume up to 750 mL per hour, if necessary, and children up to 20 mL/kg body weight/hour.

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 for as long as the infant wants, even during oral rehydration.
- **Non-breastfed infants under 6 months of age:** if using the old WHO ORS solution containing 90 mmol/L of sodium also give 100–200 mL clean water during this period. However, if using the new reduced (low) osmolality ORS solution containing 75 mmol/L of sodium, this is not necessary.
- **Older children:** throughout rehydration and maintenance therapy, offer as much plain boiled water to drink as they wish, in addition to ORS.

If oral rehydration therapy must be interrupted

If the mother and child must leave hospital before rehydration with ORS 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 4-hour treatment and to continue oral rehydration for two more days, as shown in Treatment Plan A.
- Show her how to prepare ORS solution.
- Teach her the four rules in Treatment Plan A for treating her child at home.

When oral rehydration fails

With the previous ORS, signs of dehydration would persist or reappear in about 5% of children. With the new reduced (low) osmolality ORS it is estimated that such treatment 'failures' will be reduced to 3% or less. The usual causes for these 'failures' are:

- continuing rapid stool loss (more than 15–20 mL/kg/ hour), as occurs in some children with cholera
- insufficient intake of ORS due to fatigue or lethargy
- frequent severe vomiting.

Such children should be given ORS by nasogastric (NG) tube or Ringer Lactate Solution intravenously (IV) (75 mL/ kg in four hours) usually in hospital. After confirming that the signs of dehydration have improved, it is usually possible to resume ORT successfully.

Rarely, oral rehydration therapy should not be given. This is true for children with:

- abdominal distension with paralytic ileus, usually caused by opiate drugs (e.g. 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).

In these situations, rehydration should be given IV until the diarrhoea subsides; nasogastric therapy should *not* be used.

Giving zinc

Begin to give supplemental zinc, as in Treatment plan A, as soon as the child is able to eat, following the four-hour rehydration period.

Giving food

Except for breast milk, food should not be given during the initial 4-hour rehydration period. However, children who are continued on Treatment Plan B for longer than 4 hours should be given some food every 3–4 hours as described in Treatment Plan A. All children older than 6 months of age should be given some food before being sent home. This helps to emphasise to mothers the importance of continued feeding during diarrhoea.

WHO Treatment Plan C: intravenous rehydration therapy for patients with severe dehydration

The preferred treatment for children with severe dehydration is initial rapid intravenous rehydration following Treatment Plan C. If possible, the child should be admitted to hospital. Guidelines for rehydration based on weight of the child are given in Table 61.1 Handbook 1 and based on age in Table 63.3 below.

Children who can drink, even poorly, should be given ORS by mouth until the IV drip is running. In addition, all children should receive some ORS solution (about 5 mL/kg/ hr) when they can drink without difficulty, which is usually within 3–4 hours for infants and 1–2 hours for older patients. This provides additional base and potassium which may not be adequately supplied by the IV fluid.

Monitoring the progress of intravenous rehydration

Patients should be reassessed every 15–30 minutes until a strong radial pulse is present. 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 as in Table 61.1 Handbook 1 and Table 63.2 above.

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 (able to drink) but still shows signs of some dehydration, **discontinue** the intravenous infusion and give ORS for 4 hours, as specified in Treatment Plan B.
- If there are *no signs of dehydration*, follow Treatment Plan A. If possible, observe the child for at least six hours before discharge while the mother gives the child ORS, to confirm that she is able to maintain the child's hydration. Remember that the child will require therapy with ORS 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 two days and teach her the signs that mean she should bring her child back.

What to do if intravenous therapy is not available?

- If IV 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 and show her how to give it to her child during the journey.
- If IV therapy is not available nearby, healthcare workers who have been trained can give ORS by NG 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 should be given more slowly until the abdomen becomes less distended.
- If NG treatment is not possible but the child can drink, ORS 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 this case, give ORS more slowly until the vomiting subsides.
- Children receiving NG 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 IV therapy.
- If neither NG nor oral therapy is possible, the child should be taken immediately to the nearest facility where IV or NG therapy is available.

TABLE 63.3 WHO Guidelines for intravenous treatment of children with severe dehydration

Start IV fluids immediately. If the patient can drink give ORS by mouth until the drip is set up. Give 100 mL/kg of Ringers Lactate Solution divided as follows		
Age	First give 30 mL/kg in:	Then give 70 mL/kg in:
Infants under 12 months	1 hour	5 hours
Older	30 minutes	Over 2.5 hours

Reassess the patient every 1–2 hours. If hydration is not improving, give the IV drip more rapidly. After six hours (infants) or three hours (older patients), evaluate the patient using the assessment chart. Then choose the appropriate Treatment Plan (A, B or C) to continue treatment

If Ringers Lactate Solution is not available, normal (0.9%) saline may be used

Repeat once if radial pulse is still very weak or not detectable

Further reading

World Health Organization (2005) The Treatment of Diarrhoea: a manual for physicians and other senior health workers, 4th revision. . <http://whqlibdoc.who.int/publications/2005/9241593180.pdf> Accessed 29 March 2021

World Health Organization and UNICEF (2013) Ending Preventable Child Deaths from Pneumonia and Diarrhoea by 2025: the integrated Global Action Plan for Pneumonia and Diarrhoea (GAPPD). https://www.who.int/woman_child_accountability/news/gappd_2013/en/ Accessed 29 March 2021

Section 64. Normal oxygen saturation levels SpO₂ at sea level and at altitude in healthy children [Prof. David Southall](#)

Section 64. Oxygen saturation levels in healthy pre-term and full-term newborn infants, older infants when healthy and during respiratory infections and in healthy older children who are pregnant AND Oxygen saturation at different altitudes.

Since the 1990s, pulse oximetry has been used as a non-invasive way of assessing hypoxaemia. It is particularly of value in identifying respiratory compromise before it is recognised clinically. However, although studies of “normal” oxygen saturations measured by pulse oximetry (SpO₂) are consistent between publications, advice on values to be aimed for when treating patients with respiratory failure vary widely (see Section 2 Handbook 1). The following Table 64.1 summarises the results of studies aimed at determining SpO₂ values in healthy babies and children and pregnant women (including children) at different ages,

Table 64.1 Data on health babies, children and pregnant women at different ages

Research group	Methods	No. studied	Ages at data collection	SpO ₂ values
National Heart and Lung Institute, Royal Brompton Hospital 1991-1993	Tape recordings overnight of Nellcor 200 oximeter in beat-to-beat mode (no averaging), all individual light plethysmographs, breathing movements to identify regular breathing patterns associated with sleep. Recordings 11.5 to 12.3 hours manually analysed after printing.	16 (3 studies each at different ages)	6 weeks, 3 months and 1 year	Baseline 97.3% or higher Increased 99.6% to 99.9% from 6 weeks to 3 months
		67 healthy full-term infants at home	29 to 54 days Median 39 days	Median baseline SpO ₂ during regular breathing 99.8% (range 97.0 to 100%)
		160 preterm on discharge from hospital. All preterm at 32.8 weeks gestation at birth (SD 2.5)	Recordings at gestation 36.6±2.1 SD Recordings at ages 26.3 ±24.0 SD days of life.	On discharge baseline SpO ₂ 99.6% (SD 0.5) Range 88.7 to 100% 4 babies out of 160 had SpO ₂ < 96% (88.8%, 92.1%, 92.7%, and 93.2%) Three of these had required additional inspired oxygen for > 28 days but none were considered to be clinically hypoxaemic at time of recordings.
		110 of the 160 above preterm infants at home	Gestation 42.9±3.3 weeks	Significantly higher baseline values (5 th percentile = 97.9%)

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Research group	Methods	No. studied	Ages at data collection	SpO ₂ values
		around 6 weeks later	At ages 70.3 ± 29.4 SD days of life Median age difference initial and follow up recordings 37 days range 19 to 110 days (IQR 32-49 days) later	For the 6 infants with values < 5% baseline at discharge, including the one with baseline 88.7%, later follow up recordings were made. The lowest value of baseline found was 98.4%
		19 who had graduated from SCBU /NICU Underwent 2 recordings: one during upper (n = 12) or lower (n = 7) respiratory infection	Median age at recording 76 days Range 23-181 days GA at birth was 32 wk (range 26-41 wk) Birth weight 1800 g (range 825 to 3780 g). 16 had been born preterm	In 15 infants, baseline SpO ₂ was 97% or more during infection. In 4 infants (3 of whom had bronchiolitis) their baselines during infection were SpO ₂ 93.2%, 84.3%, 93.4%, and 95.5%. All 4 had baseline >98% prior to their illness.
	Mean duration of recordings 7.7 hours, SD 1.7 hrs	19 from above study when free of infection: 14 recordings prior to and 5 after infection		All 19 infants when infection free had baseline SpO ₂ of 97% or higher.
	Mean duration of recording 12.3 hours, SD 1.4 hrs	55 preterm infants from 1 of 4 SCBUs with no signs of respiratory distress and had been median GA	55 infants Median age at study 1 day (range 1 to 7 days) 36 of the 55 infants had shown no clinical	Median baseline SpO ₂ during regular breathing was 99.4% (range 90.7 to 100%; 5 th percentile 95.5%) 49 infants had baseline SpO ₂ of 97% or higher

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Research group	Methods	No. studied	Ages at data collection	SpO ₂ values
		<p>at birth 35 weeks (range 30 to 36 weeks) Birth weights ranged 1082 to 4090 g, median 2140 g. All breathing room air at 24 hours of age. Those enrolled had GAs and birth weights similar to those not enrolled.</p>	<p>signs of respiratory distress after birth and had not received oxygen or respiratory support. 19 had some degree of respiratory distress during the first 24 hrs of life. No infants were receiving oxygen or any form of respiratory support during their recordings.</p>	<p>One infant had baseline of 90.7% when studied age 2 days. He was born at 34 weeks GA and had received 30% oxygen for 18 hrs after birth but was then considered well. His respiratory rate was high at 77 bpm.</p>
National Heart and Lung Institute, Royal Brompton Hospital 1991-1993	Median duration of recording was 21.5 hours (range 4.5 to 23.4 hours)	<p>90 infants all being born at term (37 wks. GA or higher) 71 born following unassisted vaginal delivery, 9 by assisted vaginal delivery and 10 by Caesarean section (4 as an emergency). 8 infants received bag and mask ventilation for failure to breathe at birth</p>	Median age at commencement of recording was 1 hour	<p>Median baseline 98.3% (range 88.7 to 100 %) Results at 4 hr intervals were as follows: 0-4 hrs baseline 99.0% (range 88.4 to 100%) 4 to 8 hrs baseline 98.4% (range 87.6 to 100%) 8 – 12 hours 98.6% (range 78.1 to 100%) 12-16 hrs baseline 98.1% (range 79.6% to 100%) 16 – 20 hrs 98.3% (range 81.2% to 100%) 20-24 hrs baseline 97.8% (range 88.7%-100%)</p>

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Research group	Methods	No. studied	Ages at data collection	SpO ₂ values
				<p>Levels in babies receiving bag and mask resuscitation at birth were significantly lower during the 24 hr. period (P < 0.03)</p> <p>7 recordings had low baseline SpO₂ (median 91.5% and SpO₂ was 80% or less in a proportion of regular breathing episodes: the lowest SpO₂ value within an episode of regular breathing was median 81.7% (range 78.1% to 88.6%). This subgroup of 7 infants included 5 were showing respiratory distress. Two received bag and mask ventilation and 2 were delivered by Caesarean section. Only 1 of these 7 had a normal vaginal delivery.</p>
National Heart and Lung Institute, Royal Brompton Hospital 1991-1993	Mean duration of recording 9.8 hours (SD 1.9 hours)	<p>70 healthy children. Baseline SpO₂ median 99.5% range 95.8 to 100%</p> <p>5th percentile 96.6%. similar to that in full-term infants at around 6 weeks of age</p>	Mean age 8 years (range 2-16 years)	<p>Baseline SpO₂ median 99.5% range 95.8 to 100% 5th percentile 96.6%. similar to that in full-term infants at around 6 weeks of age</p> <p>Baseline SpO₂ decreased with increasing age r = -0.3 P < 0.01</p>

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Research group	Methods	No. studied	Ages at data collection	SpO ₂ values
		Baseline SpO ₂ decreased with increasing age $r = -0.3$ $P < 0.01$ 45 were healthy white volunteers and 25 had black skin and sickle cell disease		
Green et. al	Blue toothed enabled pulse oximeter (WristOx2 3150)	1041 pregnant subjects 16 years or older without significant comorbidities although 145 had BMIs of 30 or higher	Aged 31.5 years; SD 4.9 Gestation 12 to 40 weeks.	At 12 weeks median SpO ₂ was 98% (94-99% = 3 rd to 97 th percentile) At 40 weeks median 97% (93 to 99% = 3 rd to 97 th percentile)
Uliel S et.al Normal Polysomnographic Respiratory Values in Children and Adolescents CHEST 2004; 125:872–878)	Nellcor 200 pulse oximeter averaging mode 2-3 seconds plus pulse waveforms Also: polysomnography including EEG, electromyography, electrooculography, ECG, chest wall and abdomen motion, oral and nasal airflow, and	70 healthy children and adolescents	Age ranged from 1 to 15 years (mean 4.57 years \pm SD, 8.02).	The mean SpO ₂ was 97.2 \pm 0.8% (SD)

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Research group	Methods	No. studied	Ages at data collection	SpO ₂ values
	end-tidal PCO ₂ .			
Urschitz M et al Reference Values for Nocturnal Home Pulse Oximetry During Sleep in Primary School Children CHEST 2003; 123:96–101)	VitaGuard VG 300; Getemed AG; Teltow, Germany) with a new-generation oximeter module (Masimo SET, software version 3.0.2.1; 2- to 4-s moving averaging mode.	100 randomly selected school children. 10 recordings excluded because of insufficient recording time (<5 hours) Total reported = 90 54 girls and 36 boys	Mean age 9.3 years (SD 0.6)	SAT ₅₀ (that is the median SpO ₂ during which time there was artefact free recordings) was as follows: Mean 97.9% ± 0.8% SD Median 98% IQR 98-98% 5 th centile = 97% 2.5 th centile = 97% Range = 94-100%
Terrill PI, et al. Arch Dis Child 2015;100:18–23.	Full overnight polysomnography, including 2 s averaging pulse oximetry (Masimo Radical, software-build 4.1) pulse-oximeter with 2 s averaging and 1 Hz temporal resolution	34 healthy term infants (born by normal vaginal delivery or planned caesarean section at term (38–42 weeks) with normal birth weight (10th–90th percentile), 23 Apgar score >7 at 5 min and from a non-smoking household. Exclusion criteria included congenital or anatomical	2 weeks (days 5 to 20), 3, 6, 12 and 24 months of age No: =30, 25, 27, 26, 20, respectively). Median duration of recordings: Age 2 wk. = 550 min Age 3 mth. = 535 min Age 6 mth. = 499 min Age 12 mth = 499 min Age 24 mth = 486 min	2 weeks: Median SpO ₂ 98.0% Range 95-100% 3 months: Median SpO ₂ 99.0% Range 97-100% 6 months: Median SpO ₂ 98.0% Range 97-100% 12 months: Median SpO ₂ 99.0% Range 96-100% 24 months: Median SpO ₂ 99.0% Range 97-100%

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Research group	Methods	No. studied	Ages at data collection	SpO ₂ values
		abnormality; supplemental oxygen requirement >5 minutes following delivery		

An additional study measuring SpO₂ in school children attending an emergency room.

Masaru Kobayashi, MD et al Can a pulse oxygen saturation of 95-96% help predict further vital sign destabilisation in school-aged children? A retrospective observational study Medicine (2018) 97:25(e 11135)

Children who presented to a hospital emergency department had SpO₂ measured and compared with their other vital signs. It was found that if SpO₂ was 97-100%, their other vital signs were more likely to be normal than if their SpO₂ was 95-96%. Their conclusion was that pulse oximeter saturations of 95-96% may be classed as low in school-aged children.

Assessment of oxygenation above sea level

A systematic review in 2009 found an SpO₂ of 90% to be the 2.5th centile for a population of healthy children living at an altitude of approximately 2500 m above sea level. This decreased to 85% at an altitude of approximately 3200 m.

A more recent systematic review (2020) of oxygen saturations in healthy children aged between 1 month and 17 years living at altitudes of over 2500m above sea level found that SpO₂ becomes lower as altitude increases. The mean/median SpO₂ also increases with aging at high altitude. A significant gap in SpO₂ between waking and sleeping is seen in the first months of life, which narrows as the infant gets older. The range of oxygen saturations at high altitude is wider between individuals at younger ages, and more so during sleep. In this review the median saturations at altitudes of 2500m above sea level were 94-97% and at 5100m above sea level 80-81%.

TABLE 64.2 SpO₂ levels at different altitudes

Altitude	Location	No:	Age	SpO ₂ (%)	Author	Year
1610 m	Colorado	150	< 48 hours 3 months	95% CI, 88–97 Mean, 93 95% CI, 86–97 Mean, 92.2	Thilo <i>et al.</i>	1991
1670 m	Nairobi	87	7 days to 3 years	Range, 89.3–99.3 Mean, 95.7	Onyango <i>et al.</i>	1993
2640	Bogota	189	5 days to 2 years	Range, 84–100 Mean, 93.3	Lozano <i>et al.</i>	1992
2800	Colorado	72	3–670 days	Range, 88–97 Mean, 91.7	Nicholas <i>et al.</i>	1993
2810	Quito, Ecuador	1378	1 month-12years	median 94.5 2.5 centile: <5yrs 91% >7yrs 90%	Andrade <i>et al</i>	2020
3100	Colorado	14	6 hours to 4 months 1 week to 4 months	Range, 81–91 Mean, 80.6±5.3 Mean, 86.1±4.6	Niemeyer <i>et al.</i>	1993
3658	Tibet**	15	6 hours to 4 months	Immigrant, 76–90 Indigenous, 86–94	Niemeyer <i>et al.</i>	1995
3750	Peru	153	2 –60 months	Range, 81–97 Mean, 88.9	Reuland <i>et al.</i>	1991

** Ranges refer to those born to immigrant Chinese mothers and to those indigenous babies whose families had lived at that altitude for innumerable generations.

Section 64. Normal oxygen saturation levels SpO₂ at sea level and at altitude in healthy children [Prof. David Southall](#)

Ucros S. *et al* Oxygen Saturation in Childhood at High Altitude: A Systematic Review
High Altitude Medicine and Biology (2020);21:114-125

Andrade *et al* Pulse oximetry curves in healthy children living at moderate altitude: a cross-sectional study from the Ecuadorian Andes *BMC Pediatrics* (2020); 20:440

[Pulse oximetry curves in healthy children living at moderate altitude: a cross-sectional study from the Ecuadorian Andes \(nih.gov\)](#)

Section 65. Assessing nutrition, growth, and pubertal development

Assessing Nutritional Status:

This is done using a variety of anthropometric measures, indices and indicators which form case definitions for different types of malnutrition (see Section 56, Severe Malnutrition, handbook 1).

Whilst clinically useful and closely related to nutritional status, it is vital to note that all anthropometric measures are indirect and imperfect measures of nutrition. Each has advantages and disadvantages, and it is important not to confuse an anthropometry-based case definition with a 'gold standard' measure of nutritional status. What matters is the association with clinically important outcomes such as risk of mortality and morbidity. Factors like underlying body composition influence this association but are currently difficult to assess outside of research environments.

For practical, clinical and public health purposes, assessment of nutrition involves:

- **Anthropometric measures:** e.g. weight, length/height, mid upper-arm circumference (MUAC)
- **Anthropometric indices:** a combination of two measures or a measure and age e.g. weight-for-length; weight-for-age.
- **Anthropometric indicators:** an index is compared with a reference population. WHO growth standards are an internationally applicable reference population describing how healthy, breastfed children *should* grow in an optimal environment. These have been shown to be remarkably consistent across different countries. Z-scores refer to standard deviations (SDs) from the population median. Common anthropometric cut-offs are:

< -3 Z-scores (below 3 SD from WHO median) = severe deficit

-2 to -3 Z-score (between 2 to 3 SD from WHO median) = moderate deficit

Older charts using older growth references (e.g., NCHS) or other comparisons (e.g. % of median growth, percentiles) are sometimes seen but should ideally be replaced with up-to-date WHO-Growth Standard-based charts (see below). This allows for cross-programme and cross-country comparability.

Equipment

Weighing scales, length/height boards and MUAC tapes are all key pieces of equipment and should be sourced from reputable manufacturers to ensure quality, reliability, and durability.

- **Calibration check** should be regularly done using a standard unit:
 - weight* – e.g., a 10kg test weight. Scales should be checked *daily*
 - length* – a 1-meter wooden stick is appropriate as are *weekly* checks
 - circumference* – a tube of known circumference.
- **Make and model** of all equipment should be recorded in case of future comparability issues (e.g., MUAC tapes printed on materials of different thickness can result in 2mm differences: this can affect admission /non-admission decisions)
- **Cleaning** is important. Dirty equipment can be a source of cross-infection so should be regularly cleaned, especially in a health facility between each patient.

Length/height boards can be made locally by a good carpenter according to standard designs.

Weighing scales should ideally be:

- *Class III* (i.e., approved for use in healthcare settings for medication, treatment, and monitoring)
- *Digital*: these can be easier to read and are increasingly affordable. Spring-based scales can easily lose calibration as the spring stretches.
- *Sufficiently precise*: digital scales weighing to nearest 10g or 20g are now common.

They can help monitor smaller changes which are especially important for very small and sick patients. For example, they enable better monitoring during rehydration, which is critical for severely malnourished children.

Ensuring measurement quality

Quality of anthropometric measures is important and can directly influence whether or not a child gets specific medical and nutritional treatments, or not. As well as having high quality equipment, the following can help with high quality measures:

- **Staff training and supervision** staff taking measures should be well trained and supervised. There are standard protocols for assessing post-training performance and determining intra/inter-observer measurement differences. These should be used to identify any staff who are struggling and need further support training.
- **Ensuring children are calm and settled:** a child who is anxious and moving around lots is difficult to measure. Every paediatrician has their own calming techniques ranging from bubbles to song. These should be used.
- **Two-observer measurement system.** Used when developing the WHO Growth Standards study, the following system was used and is simple enough to replicated in many settings. Where not possible, extra attention must be paid to training and supervision (with senior staff for instance re-checking occasional measures to ensure that quality is maintained)
 - Two staff members independently measure a child and write down their results (taking care not to look at each other's measure/recording)
 - They then compare their measurements to see if they are within the below 'maximal allowable differences':
 - 0.7 cm for length/height (length is lying down, height is standing).
 - 0.5 cm for MUAC and for head circumference.
 - 0.2 cm for skinfold thickness
 - 100 g for weight
 - *If close enough*, as per above, then the final measure recorded is the average of the two. e.g., observer 1 measures MUAC 10.5 cm, observer 2 measures 11.0cm → final MUAC is 10.75cm
 - If outside of the above limits (e.g., observer 1 measures MUAC 10.5 but observer 2 measures 11,1) BOTH re-measure – since one or both must have made a mistake
 - This repeat continues until both measures are within the allowable difference. The final measure is again the average. Usually only 1- 2 repeats are needed.

Measuring Mid Upper Arm Circumference (MUAC)

MUAC is an important measure not only because it is simple, cheap, and quick but because it reliably identifies children at the highest risk of mortality. Mothers as well as health professionals can measure MUAC reliably and could be given tapes to monitor child progress once discharged home.

MUAC is mainly used for children aged 6 to 59 months, though:

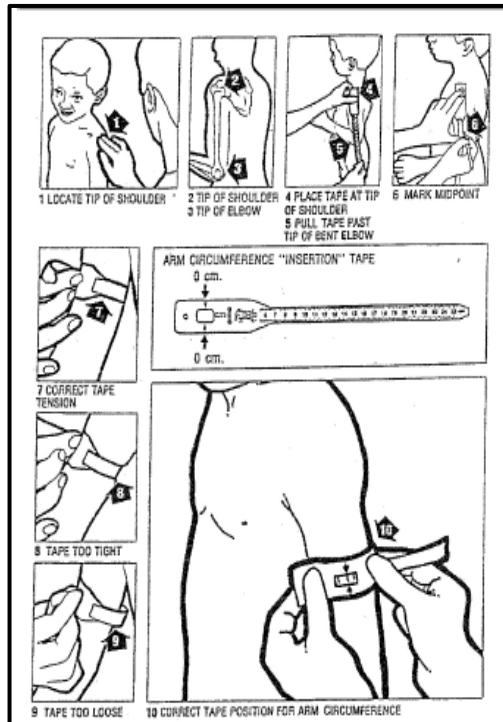
- There are also MUAC standards for older children, adolescents, and adults
- There is increasing evidence of utility for infants aged under 6 months and in older children (though cut-offs are yet to be agreed)

MUAC measurement involves:

- Exposing the left arm by removing any clothing
- With the arm flexed, finding the mid-point of the upper arm. Estimates are acceptable in a busy clinic but ideally measure from the: acromion process to olecranon tip. Use a make-up pen or similar to mark the mid-point exactly.
- For the main MUAC measurement, holding the arm straight and loose

- Loop the tape around and measuring. Correct tension is indicated by tape being flat against a child's arm with correct tension. It should be neither too tight (pinching the skin) nor too loose (a gap between tape and arm).

Figure 65.1. Measuring MUAC



Weight for length/height

This is an important measure for children with complicated severe acute malnutrition who need admission for inpatient-care.

Length or Height (and conversion factors)

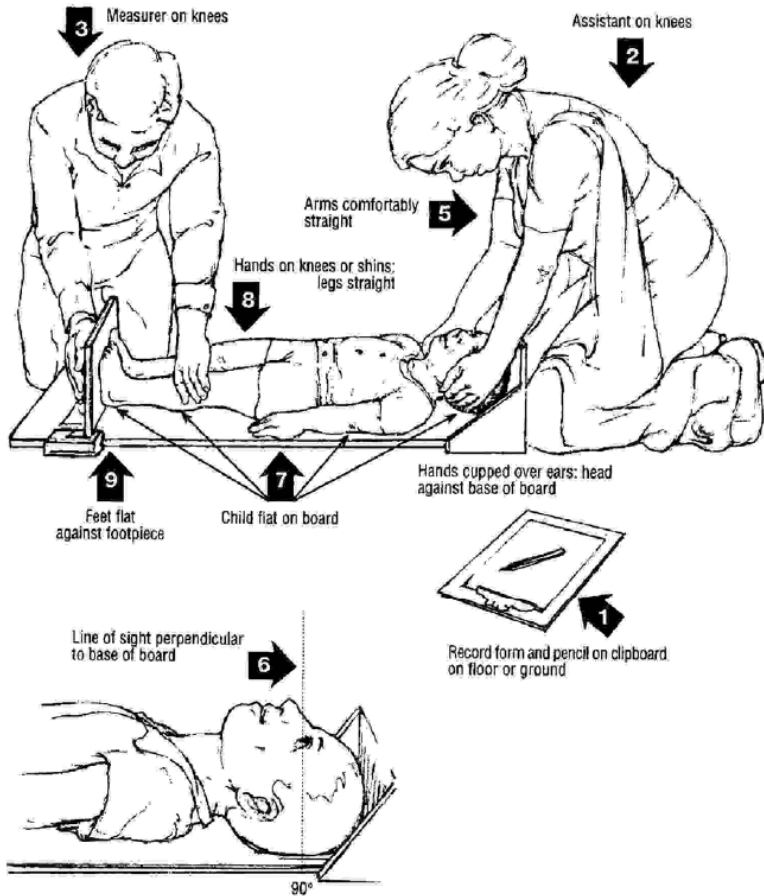
According to convention, length is measured for young children aged <2 years, and height for those ≥ 2 years. Most growth charts are based on this distinction.

Sometimes:

- < 87cm is taken as a proxy of <2 years where age is unknown (a seasonal calendar can also be used to try and establish the child's age).
- Length has to be measured for children >2 years (e.g. if unable to stand) or height is measured in a younger child (e.g. if unwilling to lie down). In such cases, use a **conversion factor of 0.7cm**. Height is less than length due to gravity-associated spinal compression. e.g.
 - If a child less than 2 years old is measured standing, **add 0.7cm** to convert to an equivalent length
 - If a child aged 2 years or more is measured lying down, **subtract 0.7cm** to convert to an equivalent height

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 Since the conversion factor is often forgotten in a busy clinical setting, **Notes should always record actual measurement PLUS whether length or height was taken** (hence the conversion factor can be applied afterwards, when interpreting the information)

Figure 65.2 Measuring length for children aged less than 2 years



Measuring length for children aged less than 2 years

Ideally, two people are needed to take this measurement, and the child should be supine on a flat surface.

The *first* person should:

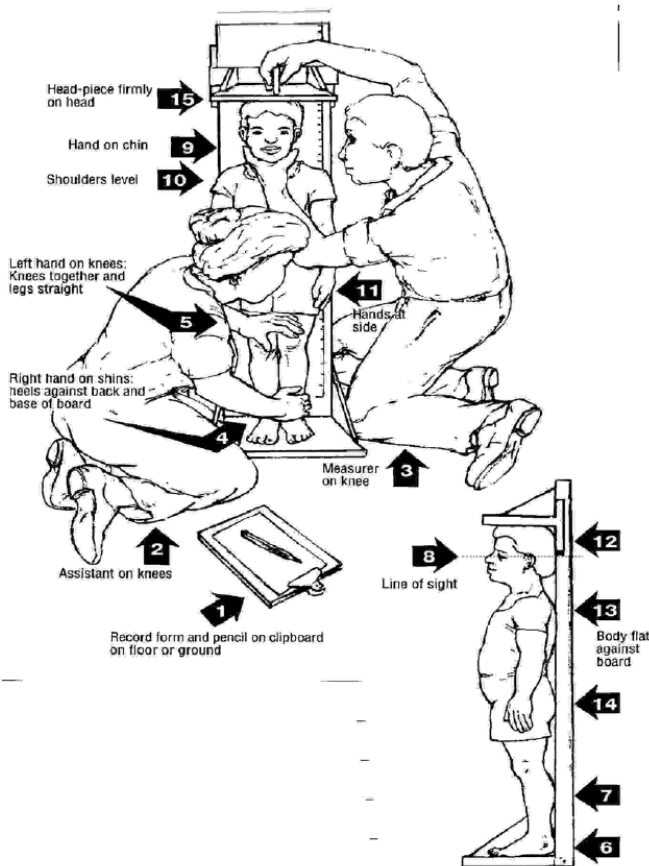
- assist in positioning the child face up on the measuring board, supporting the head and placing it against the headboard
- position the crown of the head against the headboard, compressing the hair
- check that the child lies straight along the centre line of the board and is not slanted and does not 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

- lie the child flat along the board
- place one hand on the shins above the ankles or on the knees and press down firmly, and with the other hand place the foot-piece firmly against the heels measure the length (to the nearest 0.1 cm) and record it immediately.

Figure 65.3 Measuring height for children aged 2 years and older



- This measurement should be taken without the child wearing shoes.
- The child should stand with their heels and back in contact with an upright wall.
- The head is held to look straight forward with the lower eye sockets in line with the ears. The nose must not be tilted upward.
- A weighted block at right angles to the wall is then lowered on to the head and a scale fixed to the wall is read.
- During measurement the child should be asked to stretch their neck to be as tall as possible, but their heels must not leave the ground. The measurer should help to stretch the neck by firm pressure upward under the mastoid processes.
- Measure the height immediately to within 0.1 cm.

Measuring weight at 2 years and younger

- Leave a cloth in the weighing pan to prevent chilling of 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 (ideally to the nearest 10 grams) and record immediately.

Measuring weight at 2 years and older Figure 65.4 below

- The child should be weighed naked or, if pants are worn, 0.1 kg should be subtracted from the weight measured.
- The bladder should be emptied before weighing.

Determining weight for length/height Z-score (SD score)

- Locate the row containing the child’s length in the central column of the table.
- 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.

Boys' weight (kg)					Length	Girls' weight (kg)				
-4 SD	-3 SD	-2 SD	-1 SD	Médian	(cm)	Médian	-1 SD	-2 SD	-3 SD	-4 SD
1.7	1.9	2.0	2.2	2.4	45	2.5	2.3	2.1	1.9	1.7
1.8	2.0	2.2	2.4	2.6	46	2.6	2.4	2.2	2.0	1.9
2.0	2.1	2.3	2.5	2.8	47	2.8	2.6	2.4	2.2	2.0
2.1	2.3	2.5	2.7	2.9	48	3.0	2.7	2.5	2.3	2.1
2.2	2.4	2.6	2.9	3.1	49	3.2	2.9	2.6	2.4	2.2
2.4	2.6	2.8	3.0	3.3	50	3.4	3.1	2.8	2.6	2.4
2.5	2.7	3.0	3.2	3.5	51	3.6	3.3	3.0	2.8	2.5
2.7	2.9	3.2	3.5	3.8	52	3.8	3.5	3.2	2.9	2.7
2.9	3.1	3.4	3.7	4.0	53	4.0	3.7	3.4	3.1	2.8
3.1	3.3	3.6	3.9	4.3	54	4.3	3.9	3.6	3.3	3.0
3.3	3.6	3.8	4.2	4.5	55	4.5	4.2	3.8	3.5	3.2
3.5	3.8	4.1	4.4	4.8	56	4.8	4.4	4.0	3.7	3.4
3.7	4.0	4.3	4.7	5.1	57	5.1	4.6	4.3	3.9	3.6
3.9	4.3	4.6	5.0	5.4	58	5.4	4.9	4.5	4.1	3.8
4.1	4.5	4.8	5.3	5.7	59	5.6	5.1	4.7	4.3	3.9
4.3	4.7	5.1	5.5	6.0	60	5.9	5.4	4.9	4.5	4.1
4.5	4.9	5.3	5.8	6.3	61	6.1	5.6	5.1	4.7	4.3
4.7	5.1	5.6	6.0	6.5	62	6.4	5.8	5.3	4.9	4.5
4.9	5.3	5.8	6.2	6.8	63	6.6	6.0	5.5	5.1	4.7
5.1	5.5	6.0	6.5	7.0	64	6.9	6.3	5.7	5.3	4.8
5.3	5.7	6.2	6.7	7.3	65	7.1	6.5	5.9	5.5	5.0
5.5	5.9	6.4	6.9	7.5	66	7.3	6.7	6.1	5.6	5.1
5.6	6.1	6.6	7.1	7.7	67	7.5	6.9	6.3	5.8	5.3
5.8	6.3	6.8	7.3	8.0	68	7.7	7.1	6.5	6.0	5.5
6.0	6.5	7.0	7.6	8.2	69	8.0	7.3	6.7	6.1	5.6
6.1	6.6	7.2	7.8	8.4	70	8.2	7.5	6.9	6.3	5.8
6.3	6.8	7.4	8.0	8.6	71	8.4	7.7	7.0	6.5	5.9
6.4	7.0	7.6	8.2	8.9	72	8.6	7.8	7.2	6.6	6.0
6.6	7.2	7.7	8.4	9.1	73	8.8	8.0	7.4	6.8	6.2
6.7	7.3	7.9	8.6	9.3	74	9.0	8.2	7.5	6.9	6.3
6.9	7.5	8.1	8.8	9.5	75	9.1	8.4	7.7	7.1	6.5
7.0	7.6	8.3	8.9	9.7	76	9.3	8.5	7.8	7.2	6.6
7.2	7.8	8.4	9.1	9.9	77	9.5	8.7	8.0	7.4	6.7
7.3	7.9	8.6	9.3	10.1	78	9.7	8.9	8.2	7.5	6.9
7.4	8.1	8.7	9.5	10.3	79	9.9	9.1	8.3	7.7	7.0
7.6	8.2	8.9	9.6	10.4	80	10.1	9.2	8.5	7.8	7.1
7.7	8.4	9.1	9.8	10.6	81	10.3	9.4	8.7	8.0	7.3
7.9	8.5	9.2	10.0	10.8	82	10.5	9.6	8.8	8.1	7.5
8.0	8.7	9.4	10.2	11.0	83	10.7	9.8	9.0	8.3	7.6
8.2	8.9	9.6	10.4	11.3	84	11.0	10.1	9.2	8.5	7.8
8.4	9.1	9.8	10.6	11.5	85	11.2	10.3	9.4	8.7	8.0
8.6	9.3	10.0	10.8	11.7	86	11.5	10.5	9.7	8.9	8.1

Figure 65.4 Weight-for-length reference chart (below 2 years of age).

SD = standard deviation score or Z-score. Based on World Health Organization data.

Example 1. Boy of length 61 cm and weight 5.2 kg; this child is just below -2 SD weight for length so is moderately wasted (has moderate acute malnutrition)

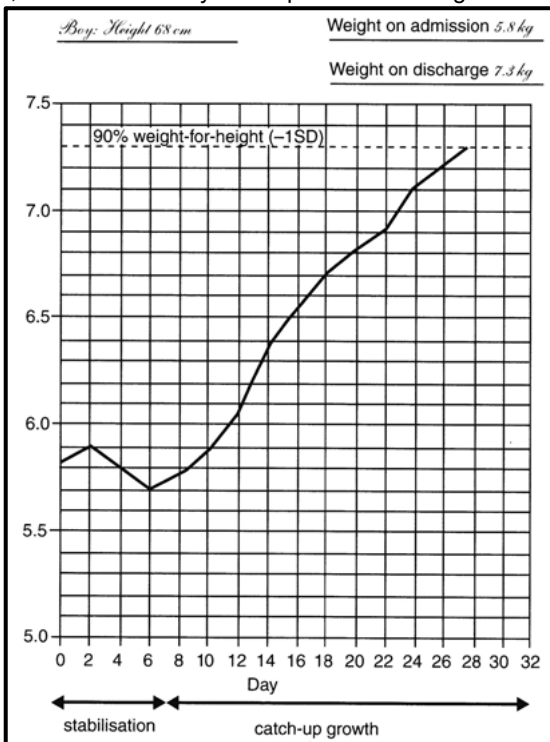
Example 2. Girl of length 67 cm and weight 4.3 kg; this child is less than -4 SD, so has a very low weight for length / is very wasted (has severe acute malnutrition)

Monitoring weight gain.

The example below is for weight gain over 3 days, but the same procedure can be applied to any interval.

- Subtract the child's weight (in grams) that was measured 3 days earlier from their current weight.
- Divide by three to calculate the average daily weight gain (grams/day).
- Divide by the child's average weight (in kg) to calculate the daily weight gain per Kg

Figure 65.5 shows a weight chart which has been used to monitor the weight gain of a severely malnourished child. It is also important in the monitoring of preterm births and newborn infants admitted to hospital. Failure to regain birth weight by 2 weeks of age is an important marker of risk. The horizontal 'x' axis represents the number of days after admission, while the vertical 'y' axis represents the weight of the child in kilograms (kg).



Notice that the weight in kilograms is stepped in 0.5 kg increments. In this example, the range has been written in from 5.0 to 7.5 kg to provide a suitable range for this individual child's expected growth.

For other children, fill in the starting weight at the appropriate level (e.g. 5 kg, 5.5 kg, 6 kg, etc., or 7 kg, 7.5 kg, 8 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 and thus shows the pattern of change more clearly.

Figure 65.6 shows 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.

Name: _____			Ward: _____			
Age: _____			Hospital number: _____			
Weight: _____			Date of admission: _____			
Date Feed: feeds ofmL eachmL 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)
Total:			Sub total			Total in 24 hours

Additional measurements for assessing nutritional status

Measures of body composition can add value to anthropometry alone but are mainly used in research settings. Note that most of these scales do not have the correct algorithms for internal calculations to produce results for young children Examples include:

Bioelectrical impedance (BIA): this measures a very weak current passing through the body and from this can calculate estimates of body fat and muscle mass. Some research suggests that BIA can differentiate malnourished children at high vs low risk of death that is not otherwise apparent using clinical signs/anthropometry alone. Scales which can measure BIA are rapidly dropping in price and may become more available in clinical settings in years to come.

Triceps skinfold thickness: Special skinfold calipers are used to measure the double layer of skin and subcutaneous fat when the skinfold is lifted. Triceps, subscapular and supra-iliac are common sites to measure skinfolds. Again, percentage of body far can be estimated from these measures.

Measurement of head circumference (See Figures 65.19 to 65.22)

Use a non-stretchable tape.
 Measure around the forehead above the eyebrows to the maximum occipital point.
 Measure twice for accuracy.

Measuring growth and development based on WHO data

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Individual measurements of weight and height/length can be plotted sequentially on charts to identify growth faltering or growth failure. See the charts below for commonly used height and weight for boys and girls (Figures 65.7 to 65.18).

A full range charts can be found on the WHO Website

- Child Growth Standards for children aged 0 to 5 years
<https://www.who.int/tools/child-growth-standards>
- Growth Reference Data for ages 5 to 19 years
<https://www.who.int/tools/growth-reference-data-for-5to19-years/indicators/height-for-age>
(note that BMI is used in preference to weight-for-height for older children)

Prematurity and Low Birth Weight (LBW)

Ideally anthropometric and growth assessment should take into account prematurity and/or LBW. These are often not known so this is not possible. Whilst adjusted charts are available, for most infants and children, standard charts are acceptable and serve well to identify most problems with growth.

BMI PERCENTILE FOR AGE-WEIGHT STATUS

- < 5th percentile - Underweight
- 5th–84th percentile - Normal weight
- 85th–94th percentile - At risk for overweight
- ≥ 95th percentile - Overweight

Formula for calculating BMI in kg/m² from weight and height

To calculate BMI from Metric values

$$\text{Weight (kg) divided by [Height (metres)]}^2$$

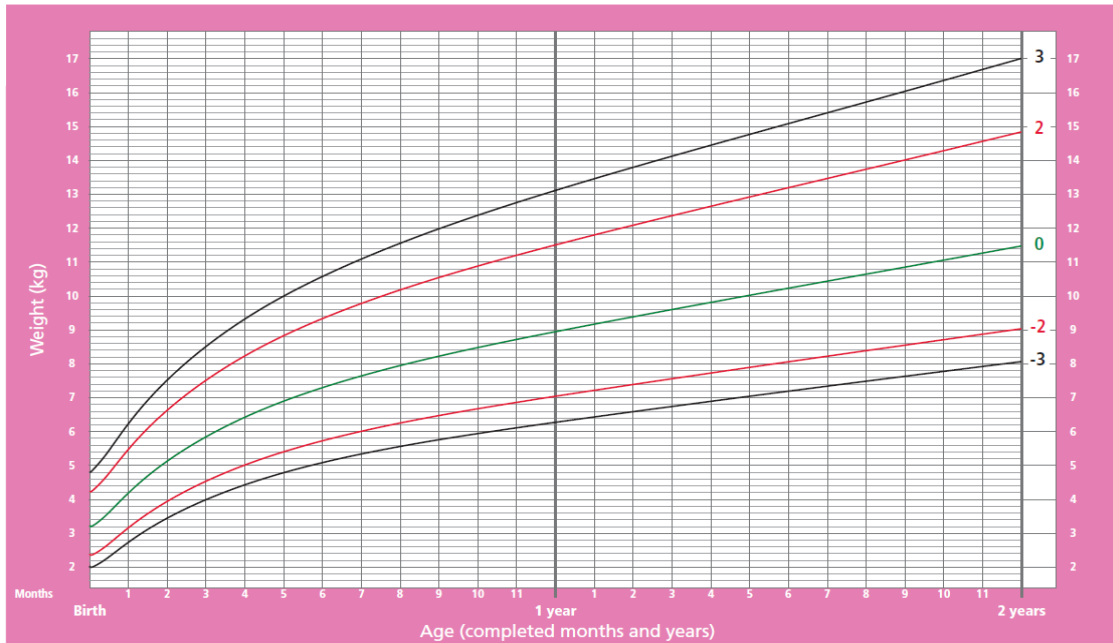
OR

To calculate BMI from Imperial values

$$703 \times \text{Weight (lbs) divided by [Height (inches)]}^2$$

Weight-for-age GIRLS

Birth to 2 years (z-scores)

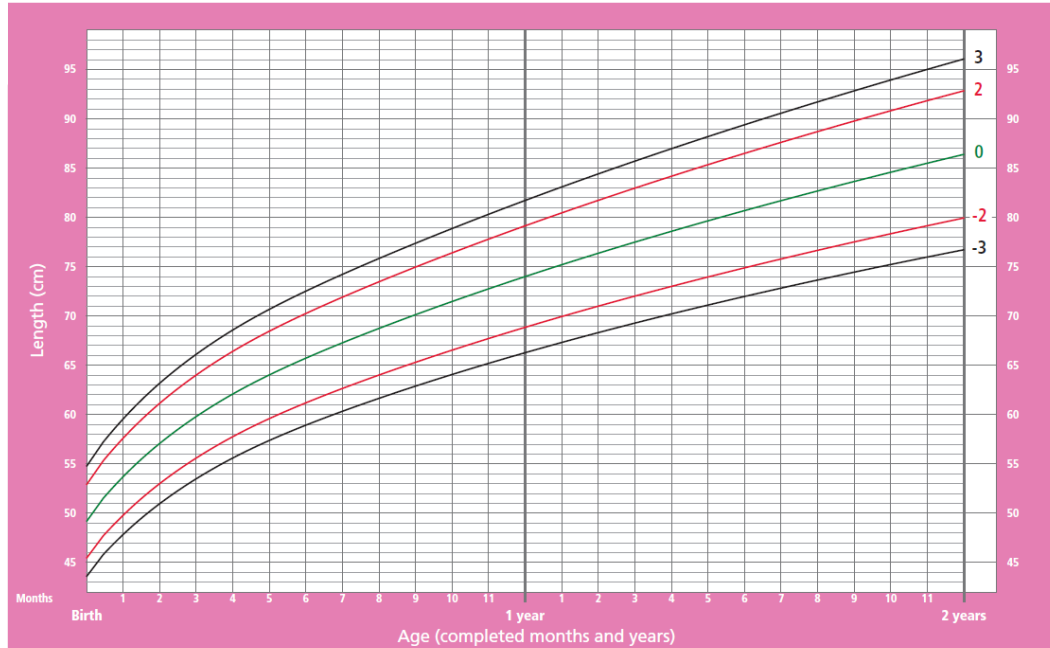


WHO Child Growth Standards

Figure 65.7 Weight-for-age chart for girls from birth to 2 years.

Length-for-age GIRLS

Birth to 2 years (z-scores)

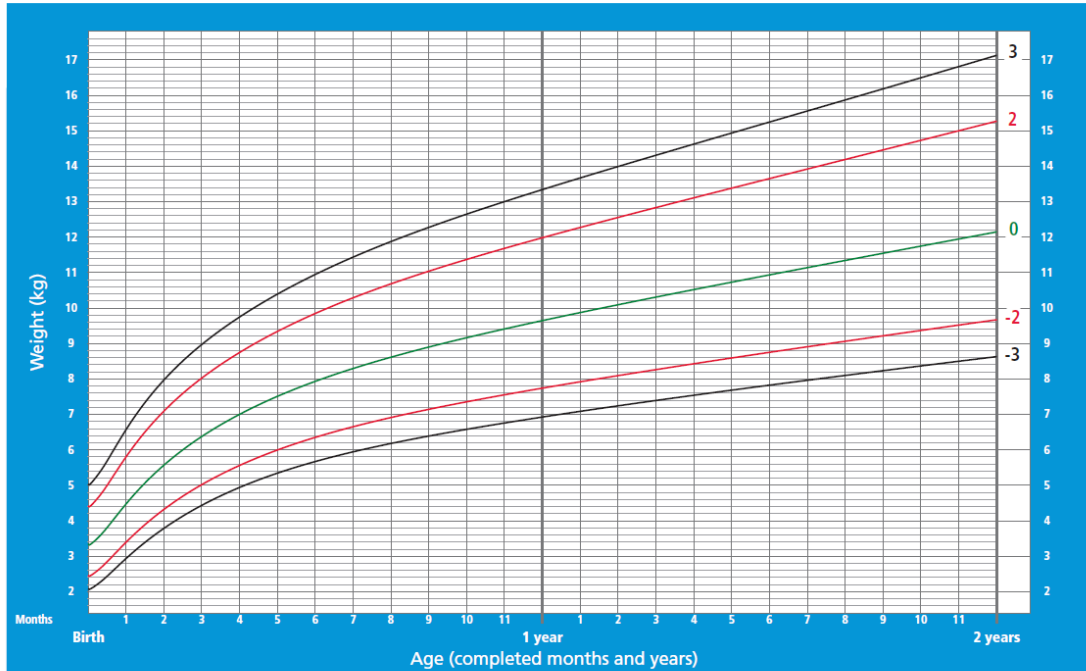


WHO Child Growth Standards

Figure 65.8 Length-for-age chart for girls from birth to 2 years.

Weight-for-age BOYS

Birth to 2 years (z-scores)

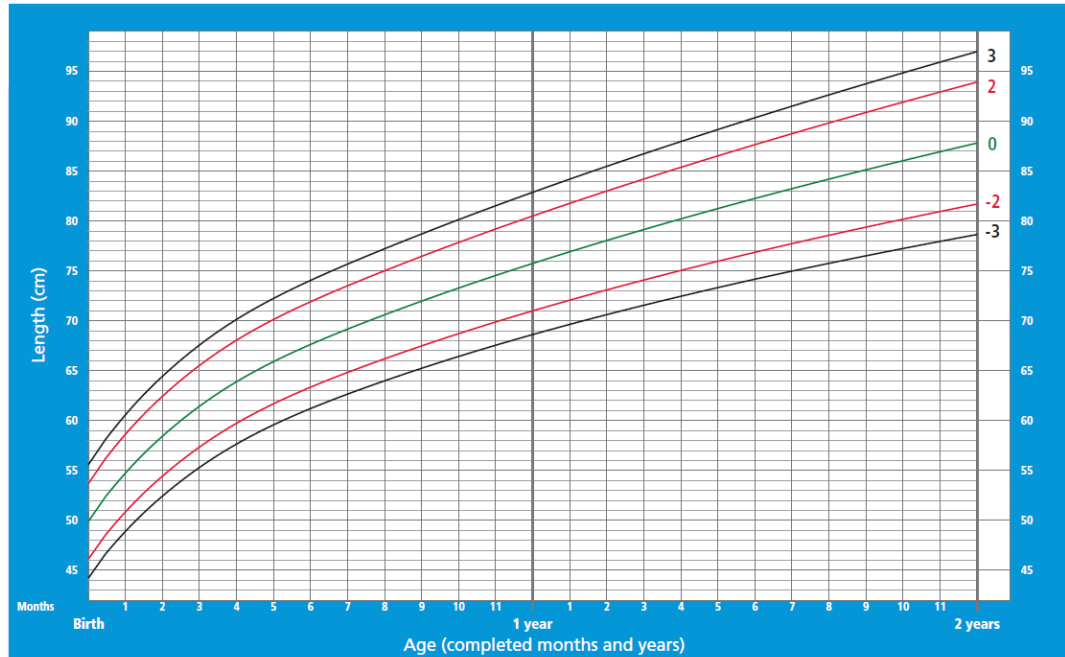


WHO Child Growth Standards

Figure 65.9 Weight -for-age chart for boys from birth to 2 years.

Length-for-age BOYS

Birth to 2 years (z-scores)

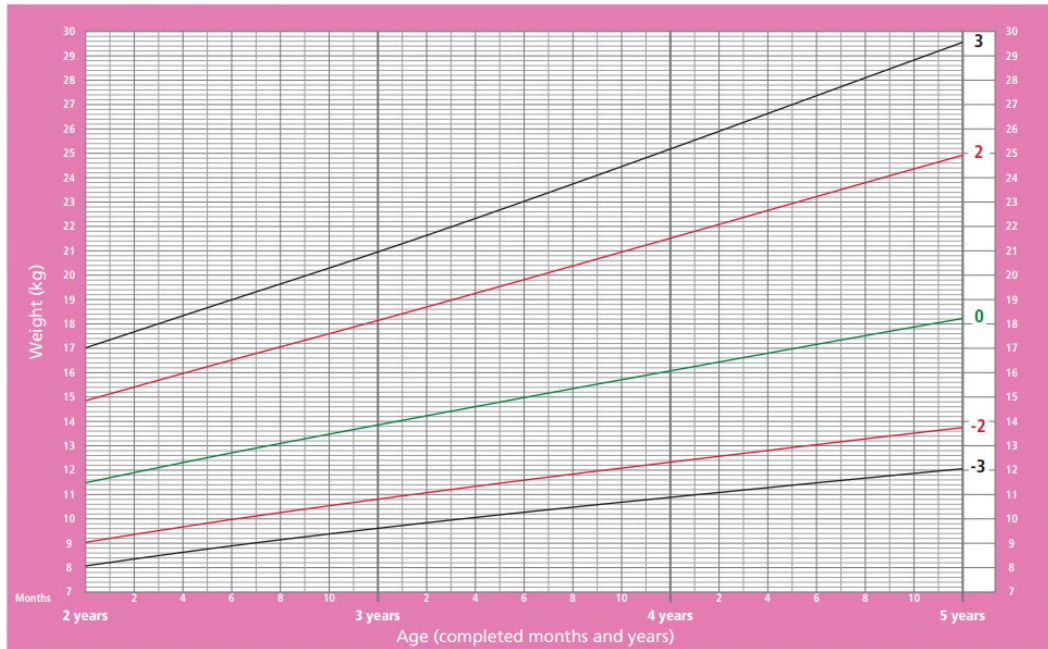


WHO Child Growth Standards

Figure 65.10 Length-for age chart for boys from birth to 2 years.

Weight-for-age GIRLS

2 to 5 years (z-scores)

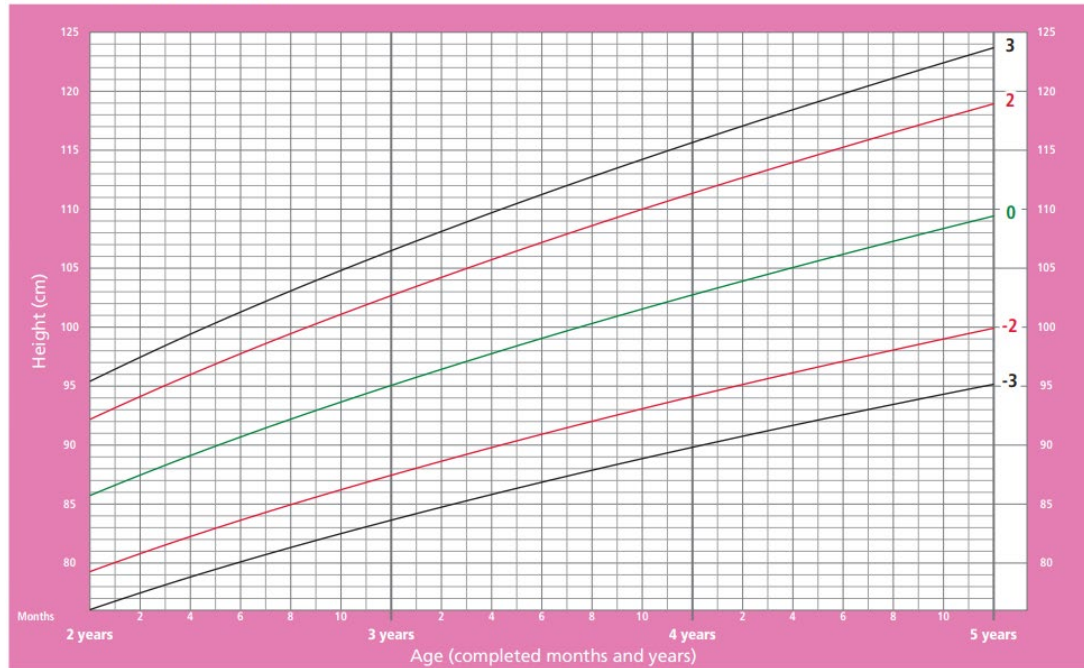


WHO Child Growth Standards

Figure 65.11 Weight-for-age chart for girls from 2 to 5 years. (Z scores)

Height-for-age GIRLS

2 to 5 years (z-scores)

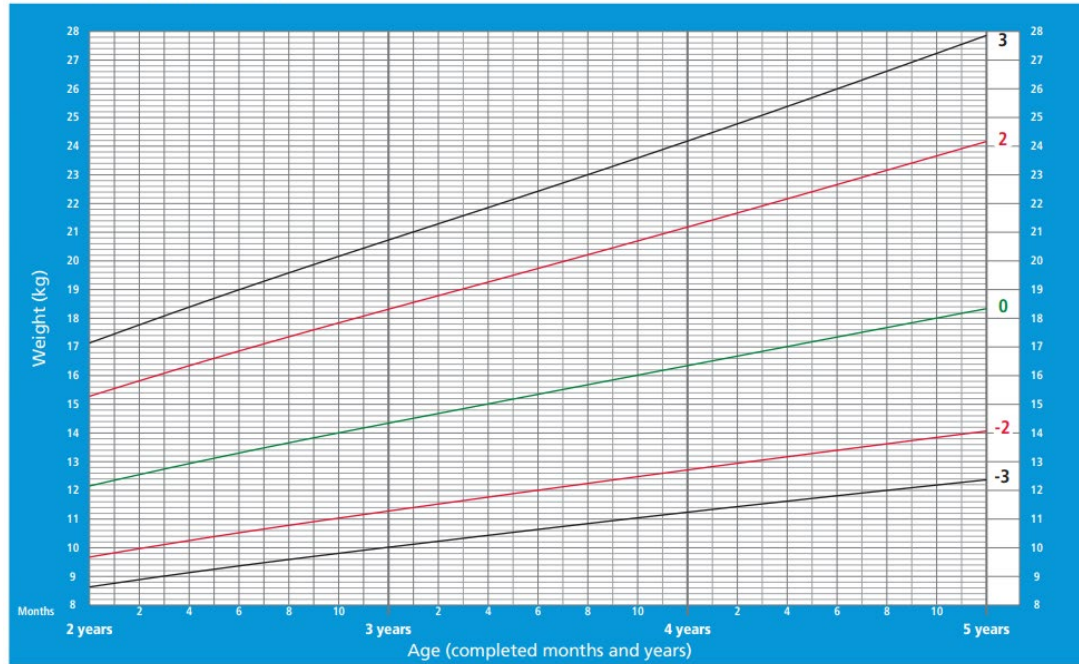


WHO Child Growth Standards

Figure 65.12 Height-for-age chart for girls from 2 to 5 years. (Z scores)

Weight-for-age BOYS

2 to 5 years (z-scores)

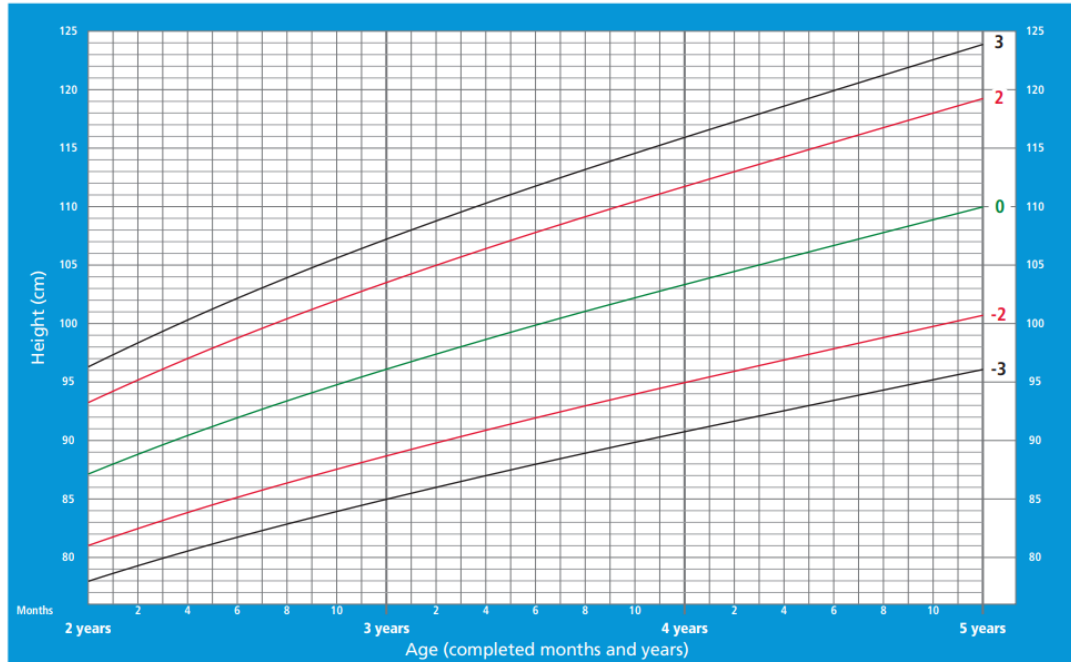


WHO Child Growth Standards

Figure 65.13 Weight-for-age chart for boys from 2 to 5 years. (Z scores)

Height-for-age BOYS

2 to 5 years (z-scores)

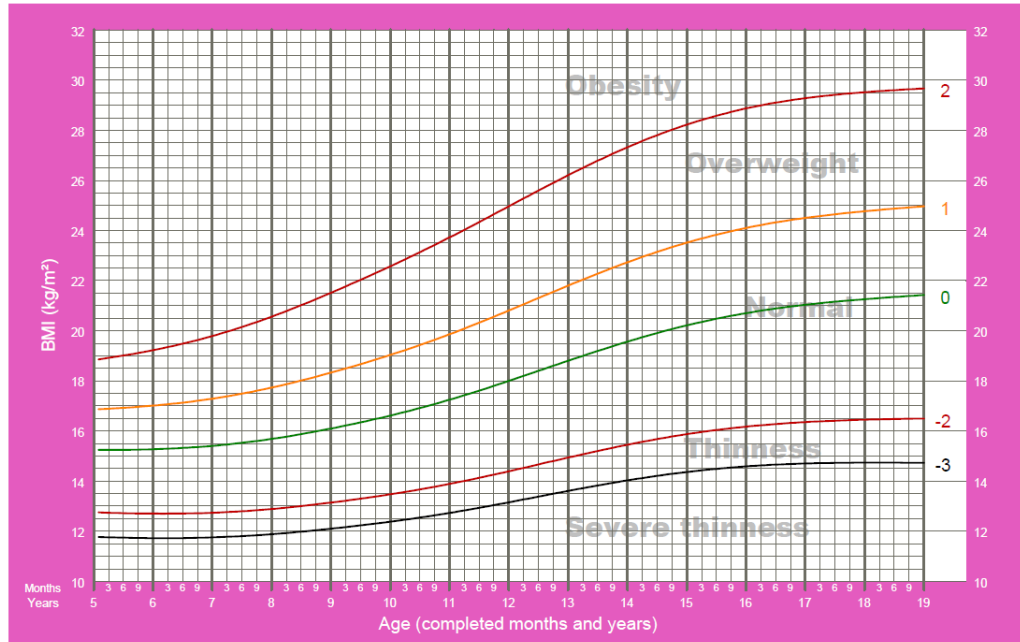


WHO Child Growth Standards

Figure 65.14 Height-for-age chart for boys from 2 to 5 years. (Z scores)

BMI-for-age GIRLS

5 to 19 years (z-scores)

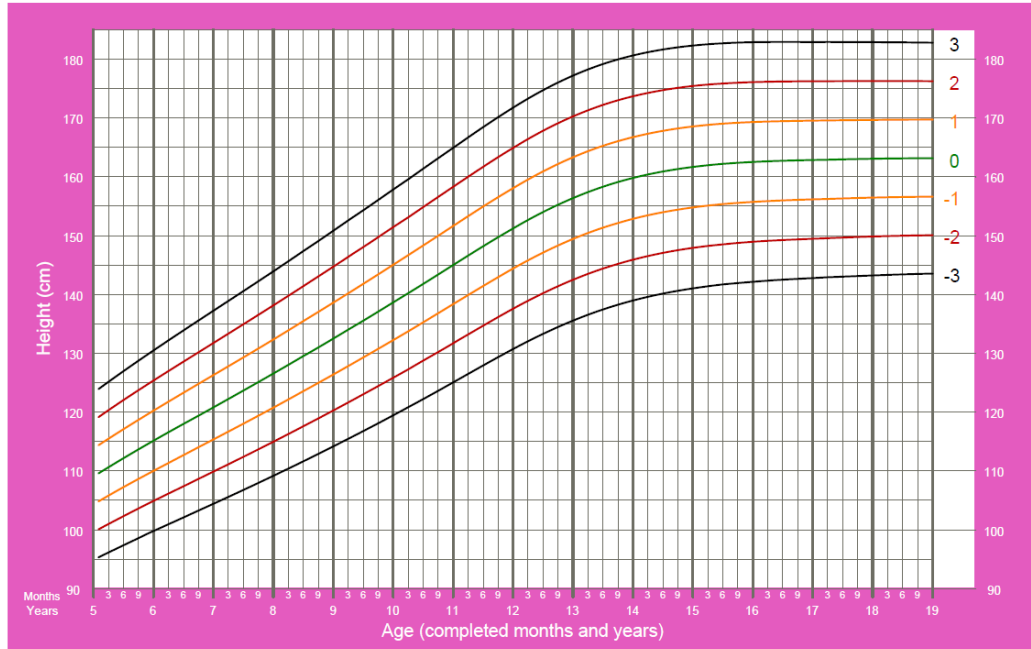


2007 WHO Reference

Figure 65.15 BMI-for-age for girls aged 5–19 years.

Height-for-age GIRLS

5 to 19 years (z-scores)

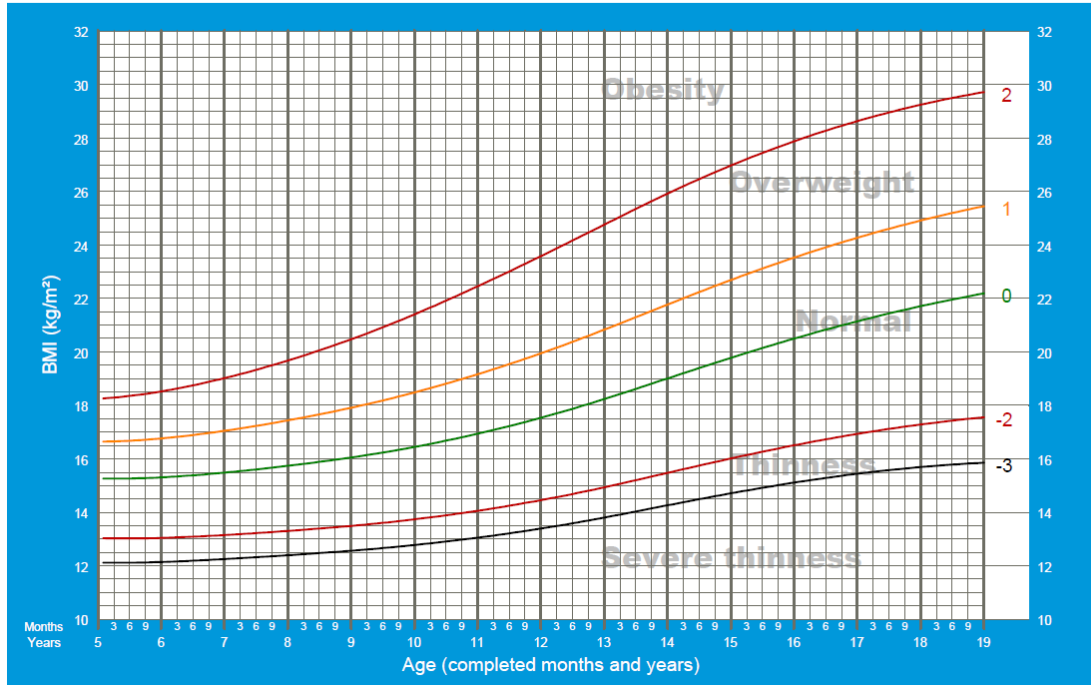


2007 WHO Reference

Figure 65.16 Height-for-age chart for girls aged 5 - 19 years

BMI-for-age BOYS

5 to 19 years (z-scores)

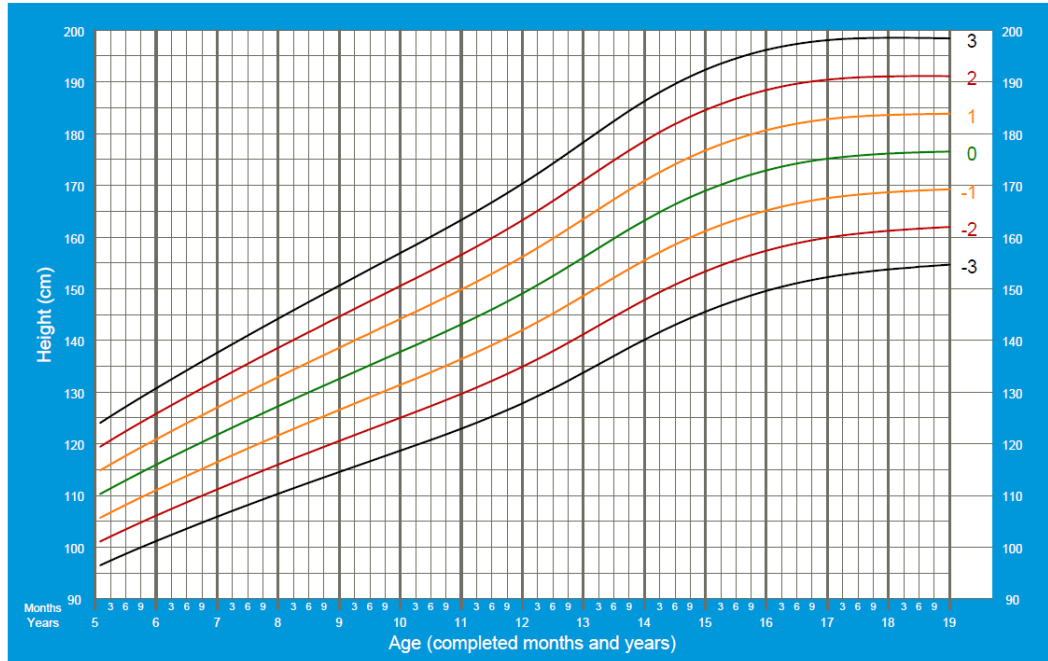


2007 WHO Reference

Figure 65.17 BMI for age chart for boys aged 5–19 years.

Height-for-age BOYS

5 to 19 years (z-scores)

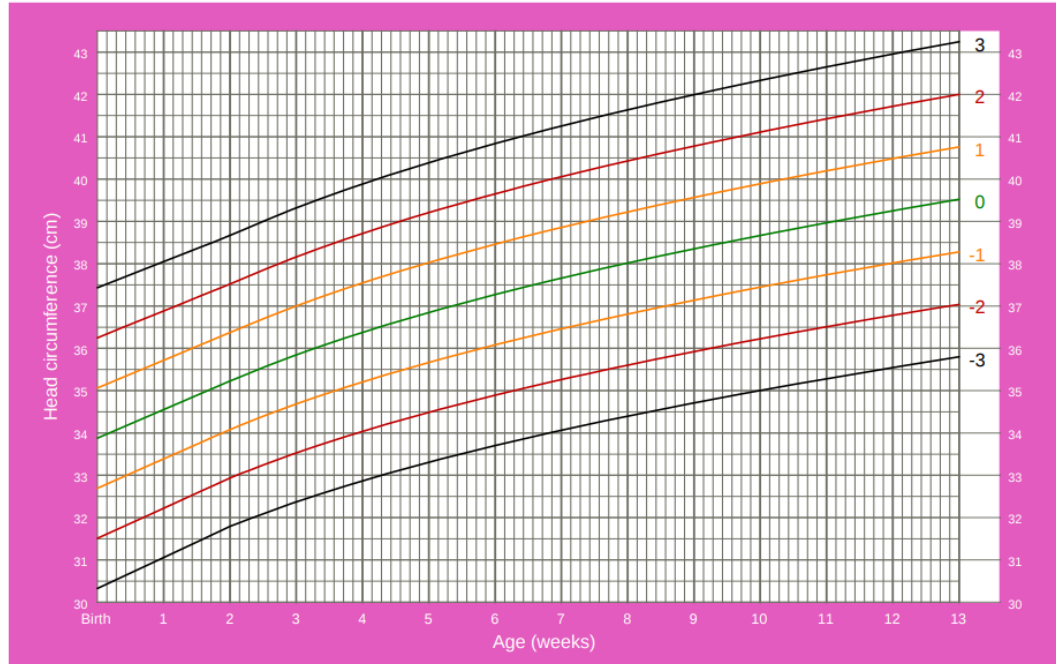


2007 WHO Reference

Figure 65.18 Height-for-age chart for boys aged 5–19 years.

Head circumference-for-age GIRLS

Birth to 13 weeks (z-scores)

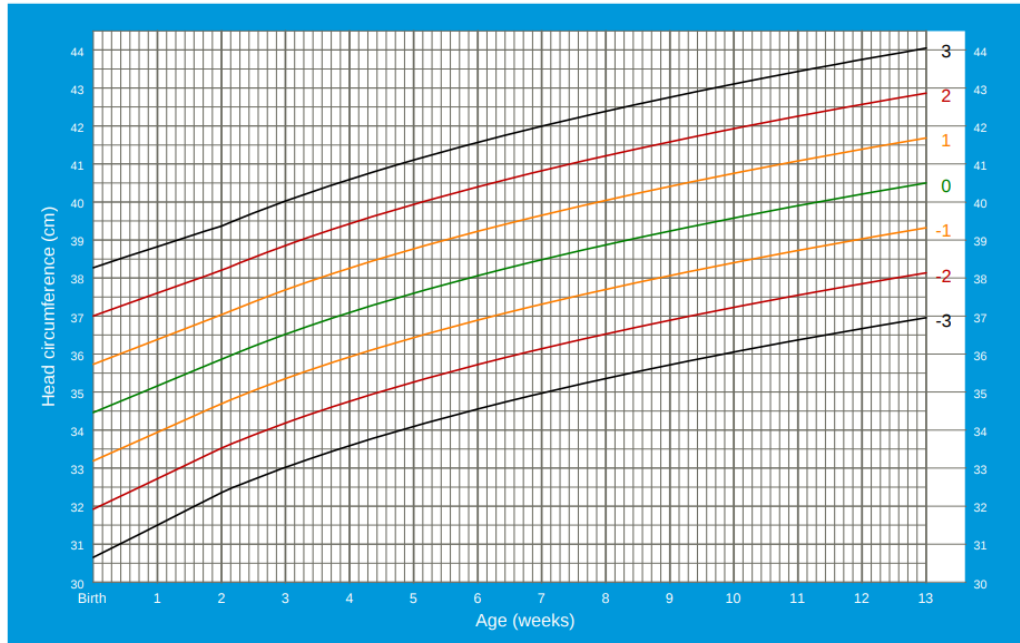


WHO Child Growth Standards

Figure 65.19 Girls chart- Head circumference for age: Birth to 13 weeks (z)

Head circumference-for-age BOYS

Birth to 13 weeks (z-scores)

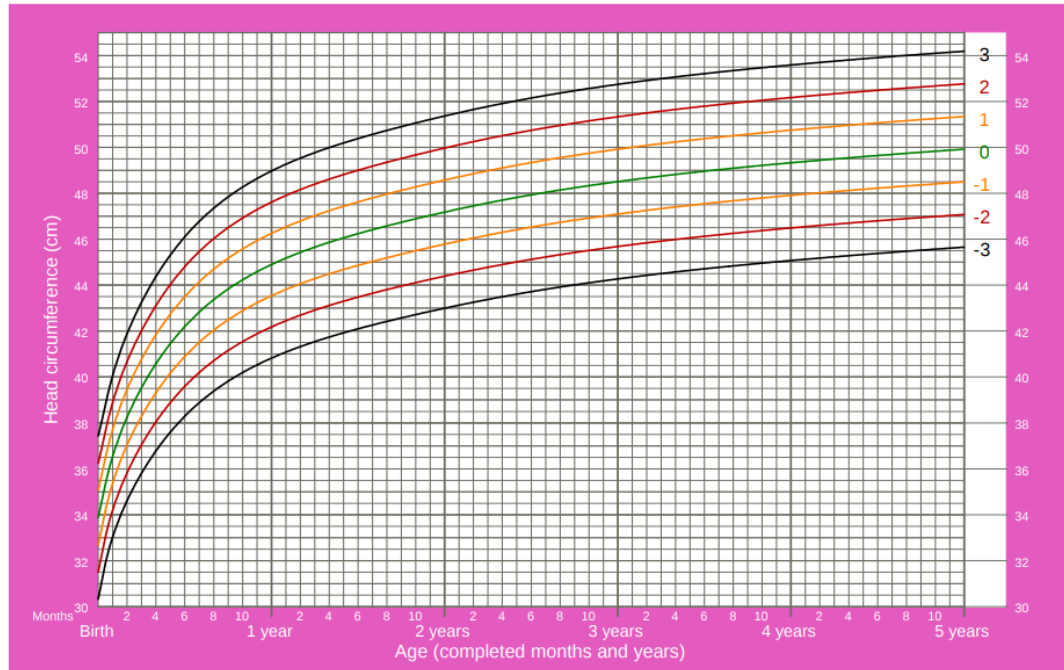


WHO Child Growth Standards

Figure 65.20 Boys chart- Head circumference for age: Birth to 13 weeks (z)

Head circumference-for-age GIRLS

Birth to 5 years (z-scores)

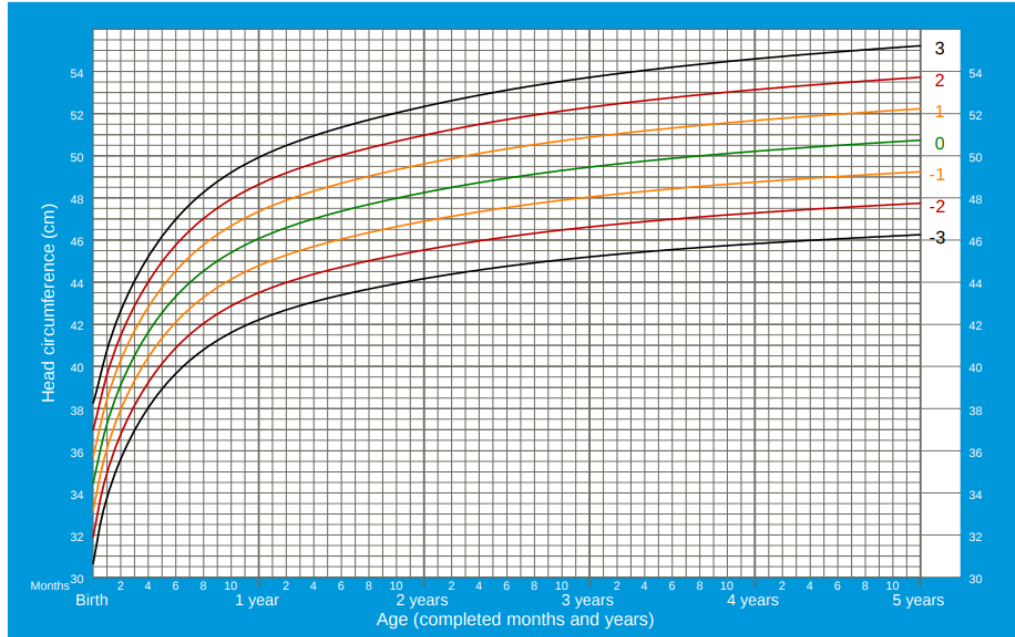


WHO Child Growth Standards

Figure 65.21 Girls chart- Head circumference for age: Birth to 5 years (z)

Head circumference-for-age BOYS

Birth to 5 years (z-scores)













WHO Child Growth Standards

Figure 65.22 Boys chart- Head circumference for age: Birth to 5 years (z)

Assessment of pubertal development

This is especially important when interpreting adolescent growth. Tanner staging is commonly used. Since examination can be difficult in some settings, it is worth noting that self-reported staging is possible using the below picture scale and has reasonable accuracy compared with expert assessment.

Girls The following should be recorded.

	Breast	Pubic Hair
Stage 1	Small nipples. No breast. 	No pubic hair. 
Stage 2	Breast and nipples have just started to grow. The areola has become larger. Breast tissue bud feels firm behind the nipple. 	Initial growth of long pubic hairs. These are straight, without curls, and of light color. 
Stage 3	Breast and nipples have grown additionally. The areola has become darker. The breast tissue bud is larger. 	The pubic hair is more widespread. The hair is darker, and curls may have appeared. 
Stage 4	Nipples and areolas are elevated and form an edge towards the breast. The breast has also grown a little larger. 	More dense hair growth with curls and dark hair. Still not entirely as an adult woman. 
Stage 5	Fully developed breast. Nipples are protruding, and the edge between areola and breast has disappeared. 	Adult hair growth. Dense, curly hair extending towards the inner thighs. 

Breast development

Stage 1. Pre-adolescent: elevation of papilla only.

Stage 2. Breast bud stage: elevation of breast and papilla as a small mound, and 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 papilla to form a secondary mound above the level of the breast.

Stage 5. Mature stage: projection of papilla 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 not further developed than that over the abdominal wall (i.e. there is no pubic hair).

Stage 2. Sparse growth of long, slightly pigmented downy hair, straight or slightly curled, chiefly along the labia.

Stage 3. Hair is considerably darker, coarser and more curled, and spreads sparsely over the junction of the pubes.











Stage 4. Hair is now adult in type, but the area covered is still considerably smaller than in the adult; there is no spread to the medial surface of the thighs.

Stage 5. Hair is adult in quantity and type, with distribution of the horizontal (or classically 'feminine') pattern

Document whether *axillary hair* is present.....YES/NO .

Document *age at menarche*.....years

Boys The following should be recorded.

	Genitals	Pubic Hair
Stage 1	No signs of puberty. Scrotum, testes, and penis as in childhood. 	No pubic hair. 
Stage 2	Initial growth of scrotum and testes. The skin on the scrotum has become redder, thinner, and more wrinkled. The penis may have grown a little in length. 	Few hairs around the root of the penis. The hairs are straight, without curls, and of light color. 
Stage 3	The penis has now grown in length. Scrotum and testes have grown. The skin of the scrotum has become darker and more wrinkled. 	Hairs are darker and curlier and still sparse, mostly located at the penis root. 
Stage 4	The penis has grown in both length and width. The head of the penis has become larger. The scrotum and testes have grown. 	More dense, curly, and dark hair. The hair growth is reaching the inner thighs. 
Stage 5	Penis and scrotum as an adult. 	Pubic hair extends upwards to the umbilicus. It is dense and curly. 

Genital (penis) development

- Stage 1.** Pre-adolescent: the testes, scrotum and penis are of about the same size and proportion as in early childhood.
- Stage 2.** Enlargement of the scrotum and testes. The skin of the scrotum reddens and changes in texture. There is little or no enlargement of the penis at this stage.
- Stage 3.** Enlargement of the penis, which initially is mainly an increase in length. Further growth of the testes and scrotum.
- Stage 4.** Further enlargement of the penis, with an increase in breadth and development of the glands. The testes and scrotum are larger, and the scrotal skin is darkened.
- Stage 5.** Genitals are adult in size and shape.

Pubic hair

- Stage 1.** Pre-adolescent: the vellus over the pubes is not further developed than that over the abdominal wall (i.e. there is 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.** Hair is considerably darker, coarser and more curled, and spreads sparsely over the junction of the pubes.
- Stage 4.** Hair is now adult in type, but the area covered is still considerably smaller than in the adult; there is no spread to the medial surface of the thighs.
- Stage 5.** Hair is adult in quantity and type, with spread to the medial surface of the thighs but not up the linea alba or elsewhere above the base of the inverse triangle. Document whether axillary hair is present.....YES/NO

Testicular volume

The approximate volume at each genital stage is shown below.

- 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.

Source: The standard illustrations in Tanner JM (1962) *Growth at Adolescence*, 2nd edn. Oxford: Blackwell Scientific Publications.

Section 66. Estimating body surface area and WHO/UNICEF child growth standards

In paediatrics, body surface area is commonly used to calculate some 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 for 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 under-prescribing in smaller children. Examples of drugs for which it should be used are most cancer chemotherapy agents, and corticosteroids.

Although 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 (see Table 66.1). This is not only quicker, but it also reduces the risk of making an error almost to zero.

TABLE 66.1 Surface area and weight

Weight (kg)	Surface area	Weight (kg)	Surface area	Weight (kg)	Surface area
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.	0.49	34	1.14	80	2.03
11.	0.53	36	1.19	90	2.19

Section 67. Integration of Traditional Birth Attendants (TBAs) in the hospital care of pregnant adolescent girls and their newborn babies



Figures 67.1 and 67.2 Training TBAs in the resuscitation of the non-breathing newborn infant using manikins

In poorly resourced countries, many people do not have access to healthcare practitioners who have had formal training in conditions such as the safe management of pregnancy and delivery for various reasons

- 1 In rural areas there are few public health facilities
- 2 The high cost of transport and treatment
- 3 Impassable roads

In rural areas, therefore, many pregnant women and adolescent girls receive care from Traditional Birth Attendants (TBAs).

WHO defines a TBA as “a person who assists a mother during childbirth and who initially acquired her skills by delivering babies herself or through apprenticeship to other traditional birth attendants.”

TBAs' skills are often passed on to younger generations in the family and other members of the community.

TBAs have different roles in different communities. In sub-Saharan Africa there are over 3000 communities with their own languages, culture and spiritual traditions.

In rural parts of Africa, 60-90% of births are attended by a TBA and not a skilled birth attendant (SBA). A SBA is defined by WHO as *individuals who have been specifically trained to care for women during pregnancy, birth and postnatal periods and include midwives, nurses and physicians.*

TBAs are highly respected and trusted in their communities, have proven birthing skills and help in the immediate care of the newborn.

Surveys involving women living in rural areas found that many women prefer to deliver in their villages with help from a TBA rather than having to go to a formal health facility.

Some of the reasons given were:

Cost

Many women cannot afford antenatal or intrapartum care provided by government health facilities or transport to reach facilities. TBAs are paid by the community, sometimes in kind.

Accessibility

TBAs are always available, even at night

Empathy

Women and adolescents in labour know the community TBAs, who understand local practices and encourage traditional birthing positions which SBAs may not know. In their own communities, family members are available for support

Some women who had delivered in formal health facilities commented that some of the staff were verbally and physically abusive.

Availability and cost of transport

Some TBAs examine the abdomen and assess cervical dilatation during labour, as well as listening to the fetal heart abdominally with a bamboo cane. If the TBA has concerns that labour is not progressing, she will often organise transport and accompany the woman to a facility that has basic (BEmOC) or comprehensive emergency obstetric care (CEmOC), providing support and a link between formal hospital facility and community.

Historically, TBAs have worked independently of public midwifery and obstetric services. However, as part of countries' programmes to reduce maternal mortality, there have been many suggestions regarding the role of TBAs.

- 1 training more SBAs has been considered and implemented as a way of reducing maternal mortality with the aim of excluding TBAs from any perinatal care. However, most SBAs are reluctant to work in rural areas and tend to stay in towns and cities or migrate to other countries where they are attracted by better pay and conditions.
- 2 Integrating TBAs into the country's health system and defining their roles in the commitment to reduce maternal and neonatal mortalities. Some TBAs have received some biomedical training in pregnancy and childbirth care and WHO define these as trained TBAs.

TBAs' concerns about these changes include losing income if they did not have a role in women's pregnancies and childbirth. Having no transport to carry women to hospital if complications developed.

In some settings these concerns have been considered and now the roles of TBAs in maternity care have been modified to provide a safer link between women, families and the formal public health system care. *These include:*

1. accompanying pregnant women to antenatal clinics and identifying women who should deliver in a health facility. This encourages professional relationships between TBAs and skilled birth attendants and involves the sharing of knowledge and skills.
2. access to skilled care in labour if needed, with resources including transport to accompany women to a suitable facility able to provide basic or comprehensive obstetric and neonatal care
3. WHO have recommended the use of lay health workers, including training of TBAs to distribute: calcium supplementation in areas where needed, iron and folate supplements, intermittent treatment for malaria, vitamin A supplements if indicated.

There may also be a role for TBAs in giving contraceptive advice.

In some countries where integration has occurred, TBAs have been formally introduced into the healthcare system and paid by governments.

As part of the integration of TBAs into care of pregnant women and adolescent girls and newborn infants, some countries have provided training, *including*:

- The recognition of danger signs, for example severe pre-eclampsia, convulsions, and major haemorrhage which indicate when urgent referral to a Health Centre is needed.
- Encouraging attendance at antenatal clinics.
- Training in hygienic practices when caring for women in labour and newborn infants and supplying soap
- Accompanying women in labour to an appropriate health facility if needed and supporting women in labour.
- Giving misoprostol for PPH
- Resuscitation of the newly born infant who is not breathing
- Recognising the significance of high and low temperature in mothers and babies
- Recognising the danger of some traditional practices such as putting cow dung on the umbilical cord



After their recognition of danger signs, there are frequently logistical problems in getting pregnant patients to a facility with CEmOC facilities. These have to be overcome and the solutions depend on local conditions. All links between the community and emergency care must be reliable 24 hours a day to improve maternal mortality and reduce morbidity.

Complication of pregnancy or delivery diagnosed in community



Arrange transport – depends on fuel, reliability of vehicle



Poor roads – especially during rainy season



A suitable facility providing basic or comprehensive emergency obstetric or neonatal care – employing 24/7 adequate numbers of trained and skilled staff



Emergency drugs and equipment available for surgery such as Caesarean section

Strengthening Emergency Care

In the Gambia a training program was established by MCAI in collaboration with the Ministry of Health. The President of The Gambia provided a dedicated 4-wheel drive vehicle for transporting patients needing hospital treatment for complications of pregnancy or delivery from the community to the nearest CEmOC facility. Since there was almost universal mobile phone coverage in the country, TBAs were provided with a mobile phone and solar charger. A local telephone company provided free emergency calls to a driver allowing transport to formal health care facilities.

Most TBAs had received no school education and were illiterate, so pictorial manuals were used in training sessions https://12dec1c3-e3f4-e784-7f3a-6e7903610962.filesusr.com/ugd/dd2ba4_85862f867fd34f2da365c9ae373b9e59.pdf

To undertake some of the lifesaving activities listed above, TBAs were provided with the following equipment and trained how to use it.

- 1 A self-inflating bag-valve-mask to assist breathing of newborn at birth if needed.
- 2 A thermometer to take the mother's and/or baby's temperature
- 3 A pictorial manual indicating danger signs in pregnancy and advice on when to arrange transfer to a facility providing CEmOC. https://12dec1c3-e3f4-e784-7f3a-6e7903610962.filesusr.com/ugd/dd2ba4_e9154b6f467c410470283d8a79ba662e.pdf

Conclusion

TBAs can form a vital link between community and health facility midwifery, having the trust of women and girls who are pregnant, and being able to recognise when delivery at a formal Health facility is needed. When linked with adequate communication links, transport, infrastructure and skilled health care workers, their involvement may help to reduce maternal mortality and morbidity.

Further reading

The Gambian program

Ramou Cole-Ceesay R , Meena Cherian , Alieu Sonko , Nestor Shivute , Mamady Cham , Mike Davis , Famara Fatty , Sue Wieteska , Momodou Baro , Diane Watson , Barbara Phillips , David Southall Strengthening the Emergency Healthcare system for mothers and children in The Gambia: a pilot programme description and interim evaluation *Reproductive Health* 2010, 7:21 <http://www.reproductive-health-journal.com/content/7/1/21>

<https://www.mcai.org.uk/strengthening-emergency-care-c1100>

https://12dec1c3-e3f4-e784-7f3a-6e7903610962.filesusr.com/ugd/dd2ba4_6edcd986bbf4e339d631367100009.pdf

https://12dec1c3-e3f4-e784-7f3a-6e7903610962.filesusr.com/ugd/dd2ba4_fc186cdb162148d4b09c78b5160f82d6.pdf

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SulaymanH, AdajiS. Integration of traditional birth attendants (TBAs) into the health sector for improving maternal health in Nigeria: a systematic review. *Sub-Saharan Afr J Med* 2019;6:55-62 https://www.ssajm.org/temp/Sub-SaharanAfrJMed6255-4919466_133954.pdf Accessed 4th April 2021

Aziato L and Omenyo CN Initiation of traditional birth attendants and their traditional and spiritual practices during pregnancy and childbirth in Ghana *BMC Pregnancy and Childbirth* (2018) 18:64 <https://bmcpregnancychildbirth.biomedcentral.com/track/pdf/10.1186/s12884-018-1691-7.pdf> Accessed 4th April 2021

Section 67. Integration of Traditional Birth Attendants into hospital care [Dr. Diane Watson](#), [Prof. David Southall](#)

Primus Che Chi and Henrik Urdal SAGE Open Medicine Volume 6: 1-9 The evolving role of traditional birth attendants in maternal health in post-conflict Africa: A qualitative study of Burundi and northern Uganda. Accessed 4th April 2021
<https://journals.sagepub.com/doi/pdf/10.1177/2050312117753631>

Section 68. Paediatric Advanced Warning Scores

Instructions for the use of the PAWS charts

Dr/senior nurse/paediatric clinician should be informed if:

- 1 one or more recordings in red
- 2 two or more recordings in orange
- 3 if you think the child is deteriorating even if no change in observations
- 4 Increase frequency of vital signs assessment to every 15 minutes

When informing staff to review patient use **SBAR format**

S - situation	I am (name), nurse on ward I am calling about..... I am concerned that..... (e.g., BP is low/high, temperature is low/high, paediatric early warning score is 2 reds, 2 oranges)
B - background	Patient (X) was admitted on (date) with Patient's condition has changed in the last (XX) mins The last set of observations were (XX)
A - assessment	I think the problem is (XXX) and I have given..... OR I am not sure what the problem is, but the patient is deteriorating OR I don't know what's wrong but I'm really worried
R - Recommendations	I need you to come to see the patient in the next (XX mins) What should I do in the meantime

Actions taken after senior health worker informed

Date/time	Reason for concerns	Senior responds (Y/N)	Advice given	Will come to review

Senior's notes after seeing child

Date/time	Assessment	Management	Any change in acceptable parameters

Section 68. Paediatric Advanced Warning Score **Dr Diane Watson, Dr. Alistair Morris, Prof. David Southall**

Paediatric Advanced Warning Score (PAWS) **age 0-1year**

Name:

Age:

Date/time																				
Temperature Degrees C	>39																			>39
	37.6-39																			37.6-39
	36-37.5																			36-37.5
	35-35.9																			35-35.9
	<35																			<35
Respiratory rate Breaths per minute	>59																			>59
	45-59																			46-59
	30-45																			30-45
	25-29																			25-29
	<25																			<25
SpO ₂ (%)	94-100																			94-100
	92-93																			92-93
	<92																			<92
Heart rate (beats per minute)	>170																			>170
	161-170																			161-170
	110-160																			110-160
	90-109																			90-109
	<90																			< 90
AVPU responds to responds to no response	Awake																			Awake
	Voice																			Voice
	Pain																			Pain
	U																			U

Section 68. Paediatric Advanced Warning Score **Dr Diane Watson, Dr. Alistair Morris, Prof. David Southall**

Paediatric Advanced Warning Score (PAWS) **age 1 to 2 years**

Name:

Age:

Date/time																				
Temperature Degrees C	>39																			>39
	37.6-39																			37.6-39
	36-37.5																			36-37.5
	35-35.9																			35-35.9
	<35																			<35
Respiratory rate Breaths per minute	>40																			>40
	36-40																			36-40
	25-35																			25-35
	20-24																			20-24
	<20																			<20
SpO ₂ (%)	94-100																			94-100
	92-93																			92-93
	<92																			<92
Heart rate (beats per minute)	>170																			>170
	151-170																			151-170
	100-150																			100-150
	85-99																			85-99
	<85																			<85
AVPU responds to responds to no response	Awake																			Awake
	Voice																			Voice
	Pain																			Pain
	U																			U

Section 68. Paediatric Advanced Warning Score **Dr Diane Watson, Dr. Alistair Morris, Prof. David Southall**

Paediatric Advanced Warning Score (PAWS) **age >2 to 5 years**

Name:

Age:

Date/time																				
Temperature Degrees C	>39.5																			>39.5
	37.6-39.4																			37.6-39.4
	36-37.5																			36-37.5
	35-35.9																			35-35.9
	<35																			<35
Respiratory rate	>40																			>40
Breaths per minute	31-40																			31-40
	25-30																			25-30
	20-24																			20-24
	<20																			<20
	SpO ₂ (%)	94-100																		
	92-93																			92-93
	<92																			<92
Heart rate (beats per minute)	>160																			>160
	141-160																			141-160
	95-140																			95-140
	80-94																			90-94
	<80																			<90
AVPU responds to responds to no response	Awake																			Awake
	Voice																			Voice
	Pain																			Pain
	U																			U

Paediatric Advanced Warning Score (PAWS) age 6 to 12 years

Name:

Age:

Date/time																		
Temperature Degrees C	>39.0																	>39.5
	37.6-39.0																	37.6-39.5
	36-37.5																	36-37.5
	35-35.9																	35-35.9
	<35																	<35
Respiratory rate Breaths per minute	>40																	>40
	26-40																	26-40
	20-25																	20-25
	15 - 19																	15-19
	<19																	<19
SpO ₂ (%)	94-100																	94-100
	92-93																	92-93
	<92																	<92
Heart rate (beats per minute)	>150																	>150
	121-150																	121-150
	80-120																	80-120
	70-79																	70-79
	<70																	<70
AVPU responds to responds to no response	Awake																	Awake
	Voice																	Voice
	Pain																	Pain
	U																	U
Systolic BP mm/Hg	90 - 110																	90 - 110
	75 - 89																	75 - 89
	<75																	<75

Paediatric Advanced Warning Score (PAWS) **age > 12 years**

Name:

Age:

Date/time																		
Temperature Degrees C	>39.0																	>39.5
	37.6-39.0																	37.6-39.5
	36-37.5																	36-37.5
	35-35.9																	35-35.9
	<35																	<35
Respiratory rate Breaths per minute	>25																	>25
	21-25																	21-25
	15-20																	15-20
	12-14																	12-14
	<12																	<12
SpO ₂ (%)	94-100																	94-100
	92-93																	92-93
	<92																	<92
Heart rate (beats per minute)	>110																	>110
	101 - 110																	101-110
	60-100																	60-100
	55-59																	55-59
	<55																	<55
AVPU responds to responds to no response	Awake																	Awake
	Voice																	Voice
	Pain																	Pain
	U																	U
Systolic BP mm/Hg	90 - 130																	90 - 130
	75 - 89																	75 - 89
	<75																	<75

Further reading

Ireland's Paediatric Early Warning System (PEWS) RCPI

[https://www.rcpi.ie/paediatric-early-warning-](https://www.rcpi.ie/paediatric-early-warning-system/#:~:text=The%20Paediatric%20Early%20Warning%20System,whom%20they%20have%20clinical%20concerns.)

[system/#:~:text=The%20Paediatric%20Early%20Warning%20System,whom%20they%20have%20clinical%20concerns.](https://www.rcpi.ie/paediatric-early-warning-system/#:~:text=The%20Paediatric%20Early%20Warning%20System,whom%20they%20have%20clinical%20concerns.) Accessed April 9th

2021
