PAEDIATRIC SURGERY

WORKBOOK

J H R Becker

EDITOR


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Preface

This book is designed to act as a guide to lecturers in paediatric surgery, preparing undergraduate students to have the level of knowledge in this field of that of a General Practitioner in practice. The text is not intended to be a complete guide to paediatric surgery; it is rather a guide to the outcomes that we in South Africa would like to achieve with our undergraduate students so that on qualifying, they are all on a par throughout the country. The new curriculum that has been developed at the different Medical Schools has resulted in the fragmentation of teaching in paediatric surgery, resulting in the possibility that certain aspects of paediatric surgery may be omitted so that there may be serious gaps in important knowledge.

This is a workbook; the text will never be identical to the style of the lecturer. Ample space has been provided for adding additional information included by the lecturer so that the end-result will be in line with the particular lecturer's preferences. The editor welcomes comments from all who use the book regarding any glaring omissions or commissions and any other suggestions so that any necessary alterations can be made in the next edition.

The editor and co-authors hope that this guide to paediatric surgery teaching in South Africa will achieve its goal by creating a good foundation in the principles of how to treat our children with paediatric surgical problems in an appropriate manner, thus improving the morbidity and mortality of all our patients.

JHR Becker

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This is indeed a composite work and much appreciation goes to all those who helped to make its publication possible.

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Physiological differences

1

JHR BECKER, GP HADLEY & SW MOORE

1.1 Introduction

There are certain physiological differences between adults and children that are of surgical importance.

The development of the human being begins as a two-celled organism during fertilisation. The growth and development of organs occurs in the uterus; all organs progress through developmental phases of 'maturity' and are not capable of sustaining life early in their development – therefore the necessity of the maternal placenta to sustain life *in utero*. A critical point is reached when the organs have developed to a stage of maturity that will allow survival independently of the maternal placenta and it is at this time that birth occurs. If the foetus is delivered prematurely, the organs will be insufficiently developed and not always capable of sustaining life outside of the maternal 'incubator'.

Even at birth, the organs are not yet sufficiently matured to be able to function at their full physiological potential.

The physiological reserves of the organs are therefore limited, but as a result of the continual growth of the newborn through the phases of baby, toddler, child, adolescent and young adult, the organs develop to their full potential. Development reaches its peak in the third decade of the human lifecycle and thereafter the process of aging takes over, which debilitates the body.

The organs of a newborn baby have an inherent reserve on which they draw during times of stress. Sadly, this reserve is limited and there is often rapid progression to organ failure. Conversely, if the stress factors are removed, the baby has enormous potential for growth and regeneration.

As in the adult, the organs are interdependent, which means that the failure of one organ system may lead to the failure of another system. The interdependence of organ systems is more notable in babies as a result of their physiological immaturity.

The growth of the organism has secondary benefits in that proteins are used as building blocks and fewer are catabolised, resulting in a lighter load of toxins on the immature kidney.

The child is a growing being with organs that are physiologically and anatomically immature and that must adapt to a dynamic world. The earliest great adaptation is the change from intra-uterine to extra-uterine life. A child can become deathly ill within minutes to hours as a result of the immaturity of his or her physiological systems and the lack of reserves. Development from a blocked nose to pre-terminal meningitis can occur within hours. Gastro-enteritis can develop into pre-terminal dehydration and shock within minutes.

The opposite of the above statements is also true, namely that a child receiving rapid satisfactory therapy can react very quickly and recover completely.

1.2 Basal metabolic rate

The child is oxygen-hungry and requires oxygen for energy, growth and normal daily activities. This requirement is expressed in terms of oxygen consumption: 5–8 ml O_2/kg/minute in comparison with that of the adult: 2–4 ml O_2/kg/minute (MacMahon 1984). The time span of maximum oxygen consumption is usually the first 18 months of life. The oxygen requirement diminishes progressively until puberty, at which stage adult values are achieved.

1.3 Temperature regulation

- The ratio of body surface area to mass in a baby is greater than in an adult. For example, 0,2 m^2 : 3,5 kg in comparison with 1,73 m^2 : 70 kg – hence the greater loss of heat in the child (MacMahon 1984).
- The vasomotor control of the surface blood vessels is poorly developed in the neonate (birth to 4 weeks). In addition, the neonate cannot shiver to generate energy. There is indeed brown fat present between the scapulae that can be activated by noradrenaline in stress situations, but this is a limited resource.
- The sweating mechanism is ineffective until the age of 3 months. The baby is also unable to manipulate any environmental factors: the baby cannot undress, open windows or switch on fans.
- Central temperature control is immature as a result of the lack of myelinisation in the central nervous system. For this reason it is not unusual to witness large fluctuations in the baby's temperature, with peak fluctuations of 3 °C.

Surgery therefore has the concept of a 'neutral thermal environment'. This is the ideal temperature at which the baby requires minimum energy for temperature regulation. For example:

- The neutral thermal environment for a naked 1 kg baby at 50% humidity is 34,5–35,5 °C.
- For a baby of 3 kg, it would be 31,5–34,5 °C.

The above principles are important for the transport of the paediatric patient and for creating an ideal environment in which the child can be operated on.

As a standard, there should be a neutral thermal environment created with environmental heaters, as well as heating of all instrumentation, draping, fluids and anaesthetic gases that will be in contact with the child.

1.4 Body fluids

The newborn baby's body consists of 85–90% body water, which decreases to approximately 80% by the third day. It is for this reason that the newborn should not be overloaded with administered intravenous fluid in the first 3–4 days of life. The anaesthetist should also pay special attention to this matter, because an overload of fluid may lead to a right-to-left shunting, with catastrophic consequences. The kidneys are also immature and cannot handle a concentration higher than 800 mmol/ℓ, in comparison with the adult's 1 400 mmol/ℓ. At birth, the glomerular filtration rate is 38 ml/min/1,73 m^2. This quickly increases to the adult value of 125 ml/min/1,73 m^2. The adult value is achieved by the age of 12–18 months. Therefore care should be taken not to overload the newborn with fluid and electrolytes.

1.5 Gastro-intestinal tract

Immaturity of the tract in the first 2 years of life leads to poor absorption of fluids from the large and small bowels. A baby's stool therefore has a relatively higher fluid content. As a result, a baby can develop pre-renal failure within 24 hours of the start of gastroenteritis.

A fluid loss of 400 ml in a baby of 4 kg is the equivalent of losing 10% of the body mass. Those who do not know this can easily confuse watery stools with urine.

1.6 Blood volume

A baby has approximately 80 ml/kg of blood in circulation, in comparison with an adult's 70 ml/kg (MacMahon 1984). The total blood volume of a 3,5 kg neonate is therefore approximately 300 ml and a loss of 30 ml would be the equivalent of losing 10% of the total blood volume. This volume of blood loss in an adult would be negligible.

1.7 Heart

It is general knowledge that the foetal circulation undergoes changes to form adult circulation shortly after birth. It is of importance to the surgeon because the foramen ovale has not yet closed and can open again under the following circumstances:

- Hypoxia and acidosis lower the systemic and left atrial pressures, and increase the pulmonary arterial and right atrial pressures with subsequent re-opening of the ductus arteriosis and foramen ovale with a right-to-left shunting.
- Fluid overloading gives rise to pulmonary hypertension and increased right atrial pressure with a consequent right-to-left shunting over the foramen ovale.

Heart rate

The average heart rate of a baby is 140–180 beats per minute. By implication, this means that what is a normal heart beat for a baby is a definite tachycardia for an adult and that a beat of 72 beats per minute, which is normal for an adult, is a dangerous bradycardia in a baby.

Myocardial contractility

The baby's myocardium is poorly developed and the only manner in which the baby can respond to fluid changes is to adapt the heart rate. Stroke volume adaptations are not yet possible (MacMahon 1984).

1.8 Respiration

Respiration is important to the surgeon because most operations that involve the abdomen will have a direct effect on respiration. The baby already begins to 'exercise' his or her organs *in utero* for use in later extra-uterine life. Examples are the swallowing of amniotic fluid and the passing of urine. The best recognised and most widely known of these exercises or acts is probably the foetal heartbeat. The foetus also 'breathes' while in the uterus. Amniotic fluid is 'inhaled' and 'exhaled'. The fluid in the lung alveoli is quite important for the development of the lungs in order to prepare them for extra-uterine life. Lung fluid can be analysed by means of amniocentesis for dipalmitoil lecithin acid, a product of Type I pneumocytes, to determine whether the lung is mature enough for extra-uterine life.

The chest cavity is almost barrel-shaped. This is obvious on a chest X-ray (CXR) of the neonate as the lower ribs run almost horizontally, as seen in the adult emphysema patient. Ventilation is therefore mainly diaphragmatic and one can understand that raised intra-abdominal pressure, regardless of the cause, leads to splinting

of the diaphragm. This, in turn, has a strong influence on breathing and oxygenation.

The structural support of the chest, i.e. the ribs, is still rather soft and if there are problems with breathing, the obvious recession of the ribs into the diaphragm and accessory muscles is an important clinical sign. In this situation, the effort of being able to breathe increases dramatically because the shape of the chest has to be distorted for sufficient oxygenation. These signs, in addition to an increase in respiratory rate of more than 60 breaths per minute, are an absolute indication for ventilatory support.

The neonate breathes at a rate of 30–40 breaths per minute, in comparison with the adult tempo of 10–18 per minute. The reason for this is the higher oxygen consumption of the neonate, as well as the higher basal metabolic rate (BMR). It is more energy-efficient to increase the rate of breathing rather than the tidal volume because the muscles are still immature and it requires greater effort to distort the chest.

The tidal volume of a baby is 7 ml/kg and this must be carefully controlled during mechanical ventilation. The tidal volume of a 3,5 kg baby is therefore 25 ml. If the 'dead space' of the ventilator is 25 ml or more, it is possible that the baby may not receive *any* ventilation. The difference between mechanical ventilation in an adult and a child is that the tubes used in paediatric ventilation must be as thin as possible.

1.9 Central nervous system

It is common knowledge that myelinisation of the central nervous system and cortical connections are poorly developed at birth. This is the reason why babies cannot walk, turn around, lift their heads or control their temperature. Development of the brain occurs exponentially in the first 6 months of life. For example, the circumference of the neonate's head doubles in the first 6 months of life and at 1 year it reaches half the size that it is going to be as an adult. This can be a crucial or detrimental time in which the infant could be poorly fed as a result of surgery. It is therefore of great importance that any baby who is not fed enterally for longer than 5 days be placed immediately on a balanced hyperalimentation that includes carbohydrates, proteins, fats, trace elements and vitamins.

1.10 Pain control

Due to the early deficiency of development in a baby's central nervous system, it is claimed that babies cannot experience the cortical interpretation of a pain stimulus. This is a dubious statement as a baby cries and struggles during the pain stimulus when, for example, being given an injection.

As it is the writer's belief that babies *do* experience pain, it is sug-

gested that one should be very circumspect in administering analgesics to an infant under the age of 1 year. The reasons why analgesics should be administered with caution are as follows:

- The blood-brain barrier (BBB) is extremely permeable under the age of 2 years. Analgesics such as opiates could easily cause respiratory depression, particularly if administered to babies less than 6 months of age.
- The liver metabolises the analgesic slowly as the enzyme systems are immature.
- The kidneys are not mature enough to excrete analgesics satisfactorily. The consequence is that the analgesic is present for much longer in the baby's circulation than it would be in the case of a child or an adult.

Principles in the administration of analgesia

- If there are no facilities for paediatric intensive care, then a baby under the age of one year should not receive analgesia.
- If there is an intensive care unit (ICU) available and analgesia really has to be administered, then only one-third of the normal dose per gram for babies may be prescribed.

■ ANALGESIA FOR THE BABY

Older than 1 year for lesser procedures

Prescribe Tilidine® (Valoron, Warner, S7) – one drop for each year of age, sublingually, 6-hourly.

Older than 1 year for major procedures (persistent monitoring is required)

Morphine® (morphine sulphate, Labethica, S7) – 0,2 mg/kg in 20 ml 5% dextrose and water as a constant infusion, administered over a period of 10 hours.

If the baby is less than 1 year old, on a ventilator and there is continuous monitoring, then this infusion may be prescribed.

■ ANALGESIA FOR LESS SERIOUS PAIN

Panado® syrup (Saphar Med paediatric, S2)
Stilpane® syrup (Lennon S2)

1.11 Transport of the sick child

GP HADLEY

Transportation of a child is a potential interruption of the continuum of care provided (Hadley and Mars 2001, Hadley 2002). Speed is rarely of the essence and there is invariably time to prepare the child for the move and to ensure proper communication with the

receiving unit. The simple mnemonic, TWO SIDES, is presented as an *aide memoire* so that no aspect of management is forgotten.

■ TUBE

A common cause of death during transportation, particularly but not exclusively of babies with intestinal obstruction, or children with an altered level of consciousness, is aspiration of vomitus. This can be prevented by keeping the stomach empty with a nasogastric tube. A nasogastric tube will, however, only keep the stomach empty if it remains in situ and unobstructed. This requires nursing supervision at all times. In the neonate keeping the stomach empty has additional benefits in that the excursion of the diaphragm is increased, breathing is more efficient and less tiring, blood is better oxygenated and the bowel less distended which in turn improves bowel perfusion. In children with frontal head trauma, an orogastric tube is preferable but fixation of the tube can be problematic.

■ WARMTH

Childen and babies are efficient radiators. They have a large surface area relative to their mass. It is also clear that many babies with surgical pathology are born prematurely, often on the basis of polyhydramnios, and they have little or no energy reserves. If they have to use up their energy reserves to keep warm, then the metabolic chaos of hypothermia will ensue rapidly.

An already warm baby can be kept warm by insulation. It should be obvious that insulating a cold baby will simply ensure that he or she remains cold. We prefer aluminium foil as the insulator of choice, but blankets will suffice. It is freely available and cheap. Since babies lose a lot of heat through their heads, the head should be included in any insulation wrap. The best way to warm a cold baby is to add warm air to the baby's immediate environment. Heaters only warm one side of the body and can cause thermal burns. However, if this is all that is available, it should be used with caution.

■ OXYGEN

Surgical pathology seldom gets worse in the presence of oxygen. The purpose of providing supplemental oxygen is to increase oxygen delivery to the brain and bowel.

■ STABILISATION

It is often noted that if journeys were good for babies, we would prescribe them. Unstable patients die during transport and time spent resuscitating a patient is not optional – it is a *sine qua non*. The need for extreme urgency in the transfer of a child or baby is rare and when it is thought to be necessary, e.g. major intra-abdominal bleeding, some intervention at the primary hospital is probably a more intelligent course of action than risking the almost

inevitable death during transfer.

A stable patient is normovolaemic when fluid loss is replaced with an appropriate fluid, usually Ringer's Lactate. Normovolaemia will be reflected by a warm, well-perfused skin, normal cerebral function and a urine output of 1 ml/kg/h in a normoglycaemic patient.

■ INTRAVENOUS FLUIDS

Fluid loss is virtually pathognomic of surgical pathology. Given the small vascular volumes of children of all ages, replenishment of the circulating volume is an urgent matter and must be performed on a continuous basis. This means that venous access is imperative. If difficulty is experienced, then intra-osseous fluids should be given early. It should be regarded as gross negligence to turn off a paediatric infusion. Intravenous fluid replacement is a life-saving intervention, and failure to do so for trivial reasons such as "I don't have a drip stand" is unforgiveable.

■ DOCUMENTATION

All children have histories, even the newly born. A referral letter detailing pregnancy and perinatal events should accompany all neonates. If the mother is unable to be transferred with her baby, it is important that she be given an outline of the suspected problem and that support be offered in what is for her a stressful situation. All radiographs, scans, biochemical results, etc. should accompany each referral. It is a waste of time and resources to repeat such tests on arrival.

■ ESCORT

Transporting a child, particularly a neonate, is probably the pinnacle of nursing achievement. It requires great skill under difficult circumstances. Unfortunately, this difficult task is frequently left to the most junior and least experienced member of the care team. Many transfers are best accomplished with a doctor in attendance. The mother is never a suitable escort. The most important concept is that care must be *continuous*. It is pointless at a cocktail party to drop your crystal glass but claim that you were looking after it carefully most of the time. The glass is still broken. So it is with babies. No credit can be given for caring competently most of the time if the baby vomits and aspirates during a period of inattention.

■ SPECIMENS

Any specimens taken from the patient should be included in the referral package. If the mother does not accompany her baby, then a specimen of her clotted blood should be sent with the baby. This allows a safer cross-match.

Land, sea or air?

In South Africa the road system allows access to most places. There

are exceptions, but by and large transport by ambulance is the most efficient and cheapest way of effecting transfers. The additional advantage is that an ambulance can stop at any point, or be diverted to an intermediate hospital if necessary.

Gas expands at altitude, and although the volume expansion may be trivial for an adult, the increase may threaten life in the case of a neonatal pneumothorax, congenital diaphragmatic hernia, closed-loop intestinal obstruction, pneumoperitoneum, etc. Air transport should not be considered until there has been discussion between the receiving hospital and the ambulance medical officer. Helicopters do not usually fly at altitudes that make such considerations vitally important. They do, however, take time to mobilise and are a very expensive alternative to standard ambulance transfer.

Fig. 1.1 Raised bath for security reasons and also for the comfort of the person who has to bath the children.

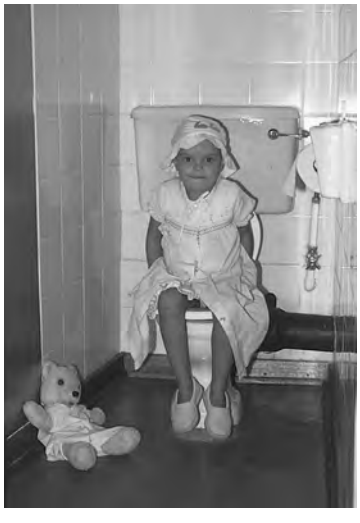

Fig. 1.2 Adapted small toilet seat to accommodate a smaller bottom.

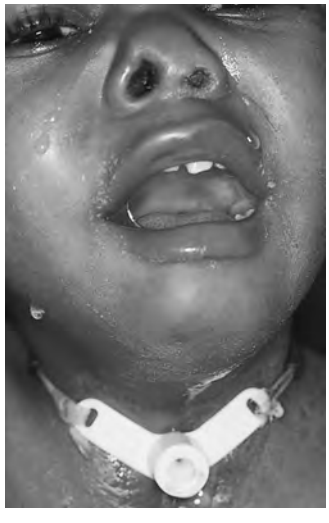

Fig. 1.3 Nasal orifice damage due to insertion of a naso-gastric tube.

Pre- and post-operative care

2

JHR BECKER & AS SHAIK

2.1 Pre-operative care

Surgery is only a part of the overall management of a patient, although it is an important part. Regarding the specifics of preparation for theatre, many of the principles of resuscitation and general management overlap. For the neonate with its limited reserves, adequate preparation is critical prior to exposing the child to the stress and trauma of the anaesthetic and surgery.

The general principles of the care include:

1. Transfer to a paediatric surgical unit
2. Evaluation and diagnosis
3. Continuing resuscitation
4. Anaesthetic involvement
5. Surgery
6. Post-operative care

Transfer to a paediatric surgical unit

It is not necessary to know what is wrong in order to start treatment. It is sufficient to know that something is in fact wrong. The mnemonic TWO SIDES is an aid to remembering simple principles that optimise the condition of the child during transfer. (See Section 1.11 – *Transport of the sick child.*)

Evaluation and diagnosis

Evaluate the general condition of the child in order to continue or alter the resuscitation as required. Accurate assessment of the physiological disorders and their correction will optimise prognosis.

Ventilation and haemodynamics are initially assessed and then investigations are conducted as indicated by the history and examination.

A diagnosis may be established by:

■ HISTORY

- Antenatal, peri-natal and post-natal (see relevant chapters)

■ EXAMINATION

- Directed to pathology (e.g. anal anomaly, but never forget to examine the whole baby)
- Ancillary
  - * associated anomalies (usually picked up during the full-body examination, which will influence decision-making and therapy)
    - – prematurity
    - – anatomical and functional anomalies
    - – associated factors that may preclude surgery, e.g. trisomy 18
  - * fitness for theatre
    - – recent or concurrent infections
    - – pulmonary or cardiac disease
    - – nutritional status
    - – metabolic status
    - – etc.

■ INVESTIGATIONS

- Directed by pathology
- Early neonatal period is a reflection of the mother's homeostasis
- Specific investigations include Hb, bilirubin, electrolytes, ABG (arterial blood gas), INR
- Radiology – a plain X-ray in the form of a babygram will identify the anomaly, as well as associated anomalies and complications of the pathology

■ CONSENT

Consent must be informed. This requires a detailed discussion of the following:

1. Pathology
2. Extent of surgery
3. Stomas
4. Blood and blood products
5. Likely outcome
6. Complications
7. Where the child will be cared for post-operatively
8. Prognosis

Protocols must be available to guide consent issues in the absence of a guardian. The mother is often recovering from a Caesarean section and may not have accompanied the child. Local policy should be followed in this case.

■ RESUSCITATION

1. Ensure warmth
2. Correct fluid replacement – replace losses with appropriate fluids
3. Nutrition – implement plan for correction of nutrient deficits pre- and post-operatively
4. Correct blood sugar and electrolytes
5. Monitor adequacy of resuscitation
   - Urine output
   - Skin colour
   - Level of consciousness
6. Monitor general parameters – pulse, temperature, SO_2, etc.

■ PROPHYLACTIC MANAGEMENT

Resuscitation and preparation for theatre may sometimes require additional procedures to optimise the patient. These may include:

- Replogle tube to prevent aspiration pneumonia (a Replogle tube is a multilumen suctioning tube to remove saliva and prevent choking)
- Relief of a pneumoperitoneum
- Ventilation
- Vitamin K
- Occasionally, administration of antibiotics

■ INVOLVEMENT OF ANAESTHETICS

The anaesthetist is an integral part of the team. He or she should be involved early in the management of the surgical patient, especially for emergencies and when the child is critically ill. Specifically, he or she needs to be involved in:

- Timing of surgery
- Specific requirements
- Primary role, e.g. in ventilation of a child with congenital diaphragmatic hernia

■ THEATRE BOOKING

The following need to be arranged for theatre:

- Time of surgery
- Operation to be performed
- Specific requirements regarding instruments, sutures, etc.
- Ancillary services such as screening and frozen section to be arranged timeously

■ TIMING OF SURGERY

The paediatric surgical patient presents with a variety of pathologies. The urgency of surgery in paediatric patients therefore varies. In general, neonatal pathology requires urgent correction. In the older child, surgery is more often an elective procedure. Surgery may be classified as:

- Urgent/emergencies
  - * True emergencies are rare and there is usually adequate time for resuscitation. However, in some conditions surgery is part of the resuscitation, e.g. a midgut volvulus.
  - * Emergencies are due to:
    - – Obstruction of bowel (midgut volvulus), obstruction of blood flow, e.g. torsion of testis, obstruction of air, e.g. tracheal foreign body.
    - – Fluid loss (blood or plasma)
    - – Hollow viscus perforations
    - – Infections
- Semi-elective
  - * These need to be performed on the next elective slate
  - * Some conditions include:
    - – Inguinal hernias in neonates and premature babies
    - – Soft tissue masses in children
- Elective
  - * Most operations in older children
  - * A risk profile must always be established prior to surgery

■ PRE-OPERATIVE STARVING

An empty stomach is only a pre-requisite for semi-elective and elective surgery. Although desirable for all cases, in surgical emergencies a period of pre-operative starving is not necessary. The following guidelines for pre-operative starvation may be used:

1. Clear fluids – 2 hours
2. Breast milk – 4 hours
3. Formula feeds/soft solids – 6 hours

■ PREMEDICATION

The choice of premedication is often made by the anaesthetist. It is essential that the timing of administration of the medication should be correct. This is only possible in elective cases. The medication chosen must be appropriate for the patient as well as the type of surgery, e.g. day-case surgery.

■ PARENTS AT INDUCTION

Often the presence of a parent at induction precludes the need for premedication. This should be encouraged. Adequate counselling of what to expect will make the induction less stressful for the child, parent and theatre staff.

2.2 Post-operative care

Preparation for the post-operative care of the child starts prior to surgery. Several factors need to be addressed to varying degrees which depend on the procedure performed. Some of the factors include:

1. Post-operative ward – ICU, high care, general ward
   * Arrange pre-operative care
   * Confirm availability of bed
   * Arrange for medical and nursing care
   * Inform parents
   * Arrange for ventilator or other special requirements
2. Monitoring
   * Continue intra-operative monitoring of respiration and ventilation
   * Haemodynamic parameters
   * Intake and output
3. Analgesia
   * Pain relief is essential
   * No role for intermittent intra-muscular analgesia
   * Regional anaesthesia, e.g. epidural
   * Intravenous infusions of morphine
   * Oral or rectal medication
4. Intravenous fluids
   * Remember post-operative fluid retention
   * Balance fluids using intake and output charts
   * Review fluids regularly
   * Maintenance and replacement fluids required
5. Nutrition
   * Pre-operative starvation and underlying malnutrition play a role
   * Predict period of post-operative starvation
   * Ensure adequate nutrition during post-operative stay
   * Enteral nutrition whenever possible
   * Supplementation with amino acids (Vamin)
   * Full TPN if necessary
6. Antibiotics
   * As necessary
   * Prophylactic or therapeutic
7. Physiotherapy
   * Chest physiotherapy – pain inhibits coughing and predisposes to chest infections
   * Mobility

8. Wound care
   * Leave dressings on unless:
   * Soiled
   * Smelly
   * Suspected wound sepsis
   * For planned wound infection
   * Counsel day cases or elective surgery cases on when dressing is to be removed and care of wound
9. Parental communication
   * Important to relay progress of child
   * Relieves anxiety and stress
   * Policies for contact to be in place
   * Communicate results as soon as available
   * Keep older patients well informed

Fig. 2.1 Intravenous infusion set with a Buratrol in line for safety purposes.

Fluids and electrolytes

3

JHR BECKER

The principles discussed in this chapter can be fruitfully applied to all patients, from babies to adults.

The child is a growing being. Growth means that building blocks are being laid down, which leads to increased size of the organism, in comparison with the adult in whom an ideal weight is the goal.

Growth is dependent on the intake of these building blocks, of which the most basic are water and electrolytes. These are followed in apparent importance by proteins (amino acids), carbohydrates (glucose), fats, vitamins and trace elements.

The most basic feeding requirements that need to be complied with over the short term are: adequate intake of water and electrolytes (Na, K, Cl) and a little glucose for calories.

As soon as it has been determined that the patient's enteral route will not be able to be used within 5 days, parenteral feeding with proteins, carbohydrates and fats must commence immediately. Maintenance must be continued but feeding (enteral or parenteral) should be commenced as soon as possible. No infant or child should be deprived of food for any period of time.

Fluid and electrolytes are the most general 'medications' administered daily in the hospital, but they are also those handled most poorly by the average doctor. Few known complications take place because of the ability to rely on the average patient's inherent adaptability to a 'hostile' environment. The reason why the complications of fluid misuse are not known is because they are seldom investigated.

The most important principle that should be taken into consideration with fluid and electrolyte therapy is: *Think in kg*.

It is unacceptable to speak of an 'adult dose' or a 'paediatric dose' because body mass can differ drastically. A woman of 50 kg is as adult as a man who weighs 90 kg. The same argument is also valid with a premature neonate, a baby and a toddler – not everyone has the same body mass and needs. The *Think in kg* principle is

also valid with the administration of other medications, e.g. antibiotics.

An average mass formula (when it is necessary to work this out quickly in one's head and a scale is not readily available) is:

- 5 months twice birth weight
- 1 year thrice birth weight
- 1–9 years (age + 4) × 2
- 8–12 years age × 3

The basic components that are necessary are:

- water
- Na
- K
- Cl
- glucose

As a starting point for the therapeutic decision about what and how much to give to the patient, two questions have to be answered:

- What are the individual needs of the patient?
- What is available in the trade to fulfil these needs?

3.1 Individual needs

- Basal needs
- Existing shortages
- Abnormal losses

Basal needs

- Water
- Electrolytes
- Trace elements
- Proteins (essential amino acids)
- Calories
  - fats (essential fatty acids)
  - carbohydrates
- Vitamins
  - water soluble
  - fat soluble

Water

Water is the single most important substance that the body requires daily – it is far too easy to give either too much or too little, with the result that oedema or dehydration occurs. For all patients on an infusion (of whatever nature) an input and output chart must be set up and adhered to. As with the pulse and temper-

ature chart, this chart must be studied several times a day and the necessary fluid adjustments must be made. It is absolutely senseless to make adjustments, e.g. the next day after reviewing the previous day's chart and to make them based on that information.

It is possible for everyone to make regular, accurately calculated adjustments in a orderly manner.

TABLE 3.1 Daily basal water needs

Patient's mass (kg)	ml of H_2O/kg/ 24 hours	ml of H_2O/kg/ hour
0–5	150*	6
5–10	125*	5
10–15	100	4
15–25	75	3
25–35	60	2,4
35–50	50	2
>50	45	1,8

* Infants after the neonatal period should not receive clear intravenous (IV) fluids in excess of their insensible and other losses (not more than 80 ml/kg)

Notes: Neurosurgery: 1 ml/kg/h
Renal failure patients: 7 ml/kg/day plus excretion

Fluid needs are lower in cases of:

- renal failure
- heart failure
- patent ductus arteriosus (PDA)
- cerebral oedema

Fluid needs are higher in cases of:

- phototherapy
- fever
- dysmature and post-mature babies

CAVEAT

Sometimes with respiratory distress IV fluids should be restricted.

Methods of fluid administration

- All babies with a birth mass of less than 1 500 g receive intravenous, potassium-free neonatalyte and thereafter potassium containing neonatalyte depending on kidney function. Oral fluid is commenced as soon as the baby has passed meconium and peristalsis is present.
- All babies with a mass of less than 1 500 g must be fed via a nasogastric tube.

- Babies with a birth mass of 1 500 to 2 000 g can be fed orally 4 to 6 hours post-partum.

If the swallowing mechanism is not yet well developed, then initially the baby can be fed via a nasogastric tube.

Babies with a birth mass of more than 2 000 g can begin with oral feeds within 4 to 6 hours after birth. If the baby is able to suckle well, then move directly to breastfeeding.

TABLE 3.2 Schedule for oral feeding

Birth mass (g)	Initial amount(ml)/ feed/kg	Increase in amount (ml)/feed/ kg/day	Number of feeds per day
< 1 000	2	1–2	8
1 001–1 250	3	2	8
1 251–1 500	4	3	8
1 501–2 000	5	3–5	8

The rest of the fluid needs are given intravenously.

NB: Before each feed the stomach content is aspirated. If there is poor passage, then the feed must be decreased to the previous day's amount. The rest of the daily fluid needs are given intravenously. When the baby can absorb 80 to 90 ml/kg/day, then the intravenous fluid administration can be ceased.

Type of feed

- The first feed is always a clear fluid – 5% dextrose.
- Larger babies can begin with milk feeds after the first clear fluid feed.
- Small premature babies and sick babies receive breast milk.
  - If the gut continuity is in question, sterile water should be given.
  - Clear fluid is not administered routinely to even the smallest babies in preference to breast milk.
  - Aspiration into the lungs of 5% dextrose is as deleterious as milk.
- Breast milk is the first choice for all babies, whether as breastfeeding or via nasogastric tube. The breast milk must be the baby's own mother's milk.
- Babies with a birth mass of less than 2 000 g can receive Prenan (Nestlé) if breast milk is not available.
- Babies with a birth mass of more than 2 000 g can receive S26 (Wyeth) or Nan 1 (Nestlé) if breast milk is not available.

Supplements

- Vidaylin (or Abedec drops) (0,6 ml/day) is added to the feeds of all babies (from the 14th day) when full oral feeds are tolerated.
- If there is an unsatisfactory mass increase after the baby has received the full volume of feed and supplements, and all other pathology has been excluded, then sunflower oil is added in the proportion of 2,5–5 ml/100 ml feed.
- Iron is added (from the 4th to the 6th week) when full oral feeds are tolerated.

Table 3.1 is one of the handiest tables that you will ever learn because if you know and understand it, then it can be successfully applied each day when you have to calculate the fluid requirements of your patients.

A baby needs to receive more fluid per kg of body mass than an adult because its ratio of body surface area to body mass is greater and a baby

- breathes faster
- has immature renal function
- has a large enterohepatic circulation of fluid – therefore large secretion of fluid takes place in the bowel which is then reabsorbed via the portal circulation. This is the reason for the danger of dehydration when a child vomits excessively and has diarrhoea.

Electrolytes

All electrolytes and trace elements are important, but for short-term management of fewer than 5 days, a person needs little more than Na, K and Cl. If parenteral feeding is required for longer than 5 days, then total parenteral feeding (containing everything) must be commenced as soon as possible, even if it is day 2.

TABLE 3.3 Short-term electrolyte needs (mmol/kg/day)

Electrolyte	Baby	Adult
Na	2–3	1–2
K	2	1
Cl	3–4	3

The newborn baby also requires calcium, magnesium and phosphate to prevent tetany and for this reason the neonatal fluid mixture, Neonatalyte®, contains these elements. In the older child and adults, these elements are not necessary in the short term.

As the basal needs of H_2O, Na, K, Cl are known, it is possible to calculate what fluid mixture will fulfil these requirements. The trade provides the electrolyte mixture in a sterile condition and conveniently packaged. The principles, with regard to the composition of the electrolyte fluid mixture, contained in the following table must therefore also be committed to memory.

TABLE 3.4 Constituents of electrolyte mixtures

Type of fluid	Na	K	Ca	Mg	Cl	HCO_3	Lactate	HPO_4	Dextrose	Osmol
0,9% NaCl	154				154					308
0,45% NaCl	77				77					154
5% Dextrose									50 g	
10% Dextrose									100 g	
Plasmalyte® B	130	4		1,5	109	28				273
Plasmalyte® L	131	5,4			107		29			273
Ringer's lactate®	131	5,4	2		111		29			279
Electrolyte No. 2	62	25		3	50		25	7	100 g	723
Maintelyte®	35	25		2,5	65				100 g	683
½ strength Darrows® with 5% dextrose	61	17			51		27		50 g	434
Neonatalyte® with 10% dextrose	20	15		0,5	21		20	3,75	100 g	670
5% NaCl	855				855					1 710
4,2% $NaHCO_3$	500					500				1 000
8,5% $NaHCO_3$	1 000					1 000				2 000
Paediatric maintenance fluid	35	12			47				55 g	372

Example

The maintenance fluid for a 9 kg baby will be as follows:

- H_2O 9×125 = 1 075 ml per day
- Na 9×2 = 18 mmol per day
- K 9×2 = 18 mmol per day
- Cl 9×3 = 27 mmol per day

The fluid solution that best suits this per litre is Neonatalyte®. The Maintelyte® that is used for adults will provide just too much Na and K.

One can extrapolate the same formula for the requirements of an adult man of 70 kg:

- H_2O $70 \times 1,8$ = 3 litres per day
- Na $70 \times 1,5$ = 105 mmol per day
- K 70×1 = 70 mmol per day
- Cl 70×2 = 140 mmol per day

The fluid solution that will fulfil these requirements is Maintelyte®.

NB: Ringer's lactate® and Plasmalyte® B are not maintenance fluids, but are isotonic to plasma. If they are administered as maintenance fluids, then the patient will become hypernatraemic and

hypokalaemic within only one day. An adult will, e.g., receive the following:

- H_2O 3 litres per 24 h – this is correct
- Na 390 mmol per 24 h – too much
- K 15 mmol per 24 h – too little

Existing shortages

When a patient has not eaten or drunk for a considerable time, is vomiting or suffering from diarrhoea, then the normal fluid balance is already disturbed. There is therefore a measure of dehydration and/or an electrolyte deficit that must be replenished. This water and electrolyte shortage must be replaced in addition to the maintenance fluids that have already been calculated.

Most patients will be isotonically dehydrated. The body therefore has a shortage of fluid that has the same composition as plasma and the fluid that is returned must be isotonic to plasma.

The best fluid mixtures that fulfil these requirements are:

- Plasmalyte® B or L
- Ringer's lactate®

If the patient's serum K is rather high due to dehydration, in other words there is pre-renal failure, then 0,45% of NaCl should preferably be administered until the patient excretes and the serum K returns to normal. Thereafter K-containing fluids, such as Plasmalyte® or Ringer's lactate®, can be switched to as necessary. If there are ongoing losses due to, e.g., diarrhoea, one usually converts to ½ Darrows® 5% dextrose in water when the intravascular volume has been redressed and the urine output is satisfactory. (NB: ½ Darrows® is not a maintenance solution.)

Depending on the level of dehydration, 5%, 10% or 15%, the calculated deficit can be administered as follows: one to two-thirds of the deficit over a period of 3 to 6 hours, and the rest during the following 18 hours. If the patient is in fact in a state of shock, the fluid mixture must be administered as a rapid bolus until the perfusion and blood pressure improve.

If large volumes of fluid are administered rapidly, it is necessary to monitor the patient's fluid therapy closely, e.g. by means of a:

- central venous pressure catheter
- bladder catheter

CAVEAT

- No child may ever receive fluid unless a Buratrol (Baxter) or Pedatrol (Baxter) is connected into the infusion line.
- No more fluid than is to be administered in one hour or per bolus may be placed in the Buratrol (Baxter) or Pedatrol (Baxter).
- The above are very important safety measures to prevent the child or baby from being too rapidly infused or receiving too much fluid, and then overhydrating and dying of heart failure or lung oedema.

- With the management of the patient in a state of shock, the bolus to be administered is measured out in the Buratrol (Baxter) and the patient is re-evaluated after each administration before the next bolus is rapidly given.
- Whenever fluid requirements are administered rapidly, the doctor remains in attendance to re-evaluate the patient regularly and to direct proceedings.

Determination of the level of dehydration, expressed as a percentage

- 5% (mild)
  - decreased skin turgor
  - dry mucosae
- 10% (moderate)
  - worsening of the above
  - sunken fontanelle
  - decreased ocular pressure
  - tachycardia
  - oliguria
- 15% (severe)
  - worsening of all of the above
  - hypotension
  - sleepiness

Determination of the fluid and electrolyte requirements for rehydration

■ WATER

Percentage of dehydration × kg, e.g.

- 5% = 50 ml/kg mass
- 10% = 100 ml/kg mass
- 15% = 150 ml/kg mass

A baby of 10 kg who is 10% dehydrated will therefore need 10 × 100 ml of fluid to replace the deficit.

The fluid is given over the following periods:

- one to two-thirds over 3–6 hours
- the rest over 18 hours

■ SODIUM

(140-sNa) × one-third of total body water = sNa deficit that must be administered over and above the daily requirements until the sNa is normal. This must be done gradually over a period of days. Sodium is an extra-cellular electrolyte; one-third of the total body water is extra-cellular and this represents the sodium deficit that needs to be replaced. If the sNa is very low, refer the patient to an

intensive care unit, because if the replenishing is done too rapidly, the patient can be brain damaged.

Acidosis

This condition usually clears up on its own as soon as the perfusion improves. Bicarbonate supplementation, if necessary, is calculated as follows:

$$\frac{(\text{Bicarbonate deficit} \times \text{kg})}{3} = \text{mmol/litre}$$

Half of this is administered over a period of 2 hours, after which the analysis is repeated and again half of the supplementation is given, if necessary, over the same period until the patient recovers.

1 ml of 8,5% solution $NaCO_3$ = 1 mmol

Deficits that occur as a result of blood loss

These patients must receive a blood transfusion. Whole blood is not always readily available during resuscitation and first it must be thoroughly cross-matched for compatibility and tested for HIV. While you are waiting for the blood and the patient needs fluid, a crystalloid (Plasmalyte® B or Ringer's lactate® at 20 ml/kg) is administered. As soon as the blood is available, it is given according to need as calculated according to the Hb or Hct determined. Blood is not given alone during resuscitation – it must be administered in a 3 : 1 ratio of crystalloid:colloid (blood). If blood alone is administered as a resuscitation fluid, the mortality is higher than when the above ratio of 3 : 1 is administered.

When a patient is not in a state of shock, the rule of simultaneous crystalloid and colloid administration does not apply. The blood is given slowly over a period of 6 to 8 hours. It is acceptable in this case to administer only packed cells.

When the pre-existing deficits in the patient have been dealt with and managed satisfactorily, apart from the previous important point (namely continuing maintenance fluid), the next important aspect is ongoing losses.

Ongoing losses

You must have knowledge of:

- the volume of fluid the patient is losing per time unit
- the composition of the fluid that is being lost

Armed with the above information, you can determine what is available to satisfy the need.

Ongoing losses imply:

- persistent vomiting or diarrhoea
- continuous stomach suctioning

- high- or low-output small bowel fistula
- pancreas fistula
- bile fistula cholesystostomy or a T-tube in the biliary ducts
- any other condition in which the patient is losing fluid, plasma or blood, irrespective of where in the body it originates – burns, etc.

TABLE 3.5 Estimated composition of different types of fluid

Fluid type	Na	K	Cl	HCO_3
Saliva	60	20	16	50
Stomach	60	10	90	–
Duodenum	105	5	100	10
Ileum	120	5	105	20
Bile	140	5	100	50
Pancreas	140	5	75	70
Diarrhoea	40	40	40	40

The volume must be measured precisely so that it can be replaced millilitre for millilitre. In the intensive care unit, abnormal losses are measured hourly and replaced over the following hour.

In the general ward where the ratio of nursing staff to patients is lower and the patients are not as sick, the abnormal losses can be measured 3–6 hourly and replaced over the following period of 3–6 hours.

For example, a patient who has lost 300 ml of fluid over the previous 6 hours will receive 50 ml of fluid per hour over the next 6 hours in addition to the maintenance fluid that he or she is already receiving.

Type of fluid

The type of fluid that is used depends on the nature of the fluid losses. Table 3.5 is a good guide, but if you have any doubts, then a specimen can be sent to the laboratory for analysis.

The following general rule can be used:

- 0,45% NaCl solution with 15 mmol K added for losses up to the pylorus
- Plasmalyte® B or L solution with HCO_3 added for losses from the duodenum, ileum, biliary tract and pancreas – see Table 3.5 for the correct bicarbonate requirements

NB: The maintenance fluid is calculated and set for a continuous infusion. The existing deficits, as well as the abnormal losses, are calculated and administered as an additional infusion. The two infusion volumes are calculated separately and each is administered in its own right, even if a single infusion cannula is used.

Example

A	* maintenance fluid as a constant infusion	ml/h
B	* existing deficit and/or ongoing losses	ml/h administered simultaneously according to need and seriousness of situation

3.2 Parenteral feeding

The aim of parenteral feeding is to fulfil all the patient's feeding requirements, namely:

- water
- electrolytes
- trace elements
- calories:
  - carbohydrates
  - fats, including essential fatty acids
- proteins, including essential amino acids
- vitamins, water-soluble and fat-soluble

Water

The values given in Table 3.1 are satisfactory.

Electrolytes

The basic electrolytes (as discussed) must be administered, plus calcium and magnesium, phosphate, etc.

Trace elements (e.g. Zn, I, Fe)

There are standardised mixtures available in the trade for children and adults.

Calories

The patient's calorie requirements are calculated as 0,8 calorie for each 1 ml of water that the patient needs. (Table 3.1 is therefore also of importance here.)

The energy in each calorie is subdivided and made up of:

- carbohydrate 60%
- fats 40%

Proteins

The protein requirement is calculated in g/kg/day as follows:

Baby and child 1,5 g/kg/day
Adult 1–1,5 g/kg/day

The protein requirements are not calculated as a source of calories. Amino acids are a very important source of structural building blocks for normal wound healing and recovery of a patient. If too much protein is given, it is excreted by the kidneys, but if too little is given, the body's own proteins (muscle) are used and healing is impaired. Amino acids must always be given with calories (carbohydrates and fats), otherwise the proteins are used as a source of energy and not as building blocks. The serum pre-albumin level is a reasonable monitor for determining whether anabolism is in fact being achieved from amino acids.

Parenteral feeding is very specialised: these patients must preferably be referred to a specialist unit where attention can be given to:

- Specialised venous access, centrally placed catheters, etc.
- Specialised mixtures that can be ordered, administered and monitored

3.3 Blood tests

These are needed if circumstances force you to give full parenteral nutrition. With the initiation of parenteral feeding, serum electrolytes must be checked daily so that the following day's needs can be calculated. You can change checking to twice a week as soon as the serum electrolytes have stabilised on the intravenous feeding regimen that is being adhered to.

NB: All formulas are merely guidelines for therapy and each patient's management must be individualised and adjusted because needs differ.

Parenteral feeding is calculated and administered within the volume limits of fluid that the patient can manage per time period of 24 hours (Table 3.1).

Baseline investigations on the first day

- S-electrolytes
- Liver function test
- Kidney function test
- Chest X-ray
- S-osmolality
- Full blood count and haemoglobin

Blood investigations that should be repeated twice a week

- Full blood count and haemoglobin
- Liver function test: total protein, albumin and enzymes
- Electrolytes: Na, K, Cl, HCO_3, calcium, magnesium, HPO_4
- Kidney function test: urea and creatinine
- S-osmolality

An approach to swellings in the child

4

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The questions that parents usually ask concerning a swelling are the following:

- Is it congenital or genetic and can other siblings also be affected?
- Is it malignant or benign?
- How will it affect my child or myself, cosmetically and functionally?

A few principles can be used as guidelines regarding the management of swellings.

In the examination of a mass, the following mnemonic (MacMahon 1984) is important: '**S**hould **T**he **C**hildren **F**ind **L**umps':

- S – site, size, shape, shift
- T – temperature, tenderness, transillumination, thrill
- C – colour, contour, consistency, cough impulse
- F – fluctuation, filling-emptying, flow-bruit
- L – lymph drainage, lumps elsewhere

The most important of all the above-mentioned points that must be examined is the first one – *site* (position). The anatomical site will lead you to the most probable diagnosis (Cockroft 1988):

- A mass in the flank in the region of the kidney will most likely be a renal tumour.
- A mass in the middle of the neck will most likely be a thyroglossal cyst.
- A mass or tumour in the groin will most likely be a lymph node or hernia.

Two types of masses will be discussed individually in more detail, namely:

- benign
- malignant

Abnormal lymph nodes (Zitelli 1981)

Palpable lymph nodes are common in children.

The *indications for further investigations* are:

- Larger than 2 cm in diameter
- Present for longer than 2 months
- Associated generalised lymphadenopathy
- Liver and splenic enlargement
- Organ enlargement or impairment for which no other good explanation can be found
- Matted lymph nodes
- No shrinkage on a course of antibiotics

The possibilities in the *differential diagnosis* are the following:

- Tuberculosis
- Metastasis
- Lymphoid hyperplasia due to immune stimulation
- Lympho-reticular disease, e.g. leukaemia or lymphoma

Vascular tumours

(See Chapter 18)

Haemangiomas, of which the *infantile haemangioma* is the most common, are distinguished from vascular malformations by their increased endothelial cell turnover, rapid post-natal growth and slow involution. They immunostain positively and reliably, for glucose transporter-protein 1 (GLUT 1) (Leon-Villapalos *et al*. 2005). Two other haemangiomas, which both present as existing vascular tumours at birth but which either rapidly involute (*RICH*), or do not involute at all (*NICH*) and do not routinely stain positively for this marker, are also recognised (Mulliken and Enjolras 2004).

It is *advisable* that all haemangiomas discovered are examined by a plastic surgeon as soon as possible after birth. Advice can then be given on early surgical intervention (e.g. excision and purse-string closure) (Mulliken *et al.* 2002), laser therapy or watchful inactivity. The latter option is usually exercised when a yellow-white, scar-like patch, also known as a '*herald spot*' in, or close to, the centre of the haemangioma is seen.

It is *mandatory* that the other vascular tumours, e.g. kaposiform haemangioendothelioma and tufted angiomas, which are associated with a life-threatening thrombocytopaenia and coagulopathy (also known as the *Kasabach-Merritt* syndrome), be referred on diagnosis to either a plastic surgeon or paediatrician for immediate pharmacological intervention (interferon or steroids) (Enjolras *et al.* 2000). This consumption coagulopathy does not occur with the common infantile haemangioma. Distinction between the various vascular tumours, when in doubt, is made on biopsy.

Vascular malformations

(See Chapter 18)

Vascular malformations, on the other hand, consist of mature endothelial cells and therefore do not involute and grow progressively with the child. They are classified by the predominant vessel type and subdivided into *high-flow* (arterial or arteriovenous) and *low-flow* (capillary, venous and lymphatic, or a mixture of the latter two) groups (Mulliken and Glowacki 1982).

One of these, the **lymphatic malformations** (Seashore *et al.* 1985), tend to be either *microcystic* (formerly known as lymphan-

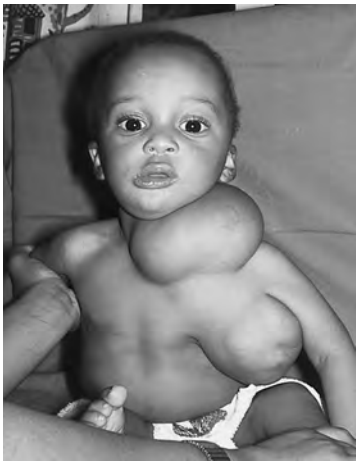


Fig. 4.1 Pre-operative macrocystic lymphangioma (cystic hygroma).

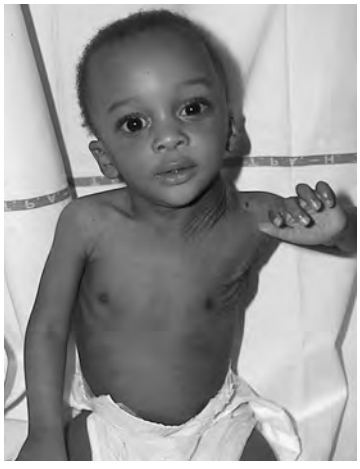


Fig. 4.2 Post-operative macrocystic lymphangioma (cystic hygroma).

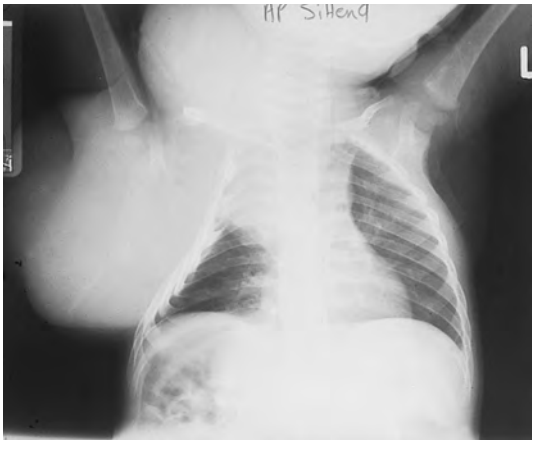


Fig. 4.3 Pre-operative chest X-ray of the baby with the macrocystic lymphangioma (cystic hygroma).

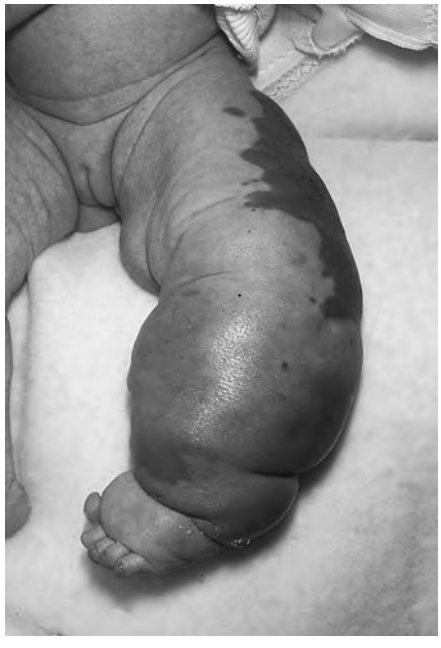


Fig. 4.4 Vascular malformation of the left leg.

gioma) or *macrocystic* (formerly known as cystic hygroma) or a combination of both, e.g. in the head and neck region where they may cause airway obstruction. Microcystic malformations appear diffuse and violate tissue planes in contrast to the more localised macrocystic malformations. Lymphoscintigraphy and MRI (magnetic resonance imaging) are most useful for diagnosis and for planning treatment, which could entail sclerotherapy alone, surgery, or perioperative sclerotherapy followed by surgical extirpation (Puig *et al.* 2003; Lee *et al.* 2005).

Sternomastoid 'tumour' (neonatal torticollis)

(See Chapter 21)

These neonates present with a torticollis (Latin *torti* = twisted and *colli* = neck) within the first few weeks after birth (rarely at birth). On examination, a firm mass may be palpable on the side of the tilt, in the sternocleidomastoid muscle. The aetiology is unknown but may have been caused by trauma sustained intrauterine or at birth.

The practitioner is, in conjunction with a paediatric or orthopaedic surgeon, obliged to:

- *Confirm* the diagnosis by excluding other (secondary) causes of torticollis:
  - congenital cervical anomalies (radiograph)
  - ocular disorders
  - cervical adenitis
  - acute fasciitis
- *Prevent* the condition from worsening as neglect could result in (Hollier *et al.* 2000):
  - plagiocephaly
  - hemiatrophy of the face
  - diplopia

Management includes physiotherapy and exercise of the affected sternomastoid muscle. It is recommended that the baby be carried on the arm in the 'anti-reflux position' with the tumour on the side of the forearm, because in doing so the affected sternocleidomastoid muscle is continuously stretched. Most (80–85%) of these tumours will resolve spontaneously within the first 18 months.

Only a small percentage of the tumours will later form a fibrotic band which should be released by means of a tenotomy, performed as either a subcutaneous or as an open procedure between the ages of 18 months to 2 years.

Post-operatively, a broad neck collar must be worn so that healing does not occur in a shortened position as these children tend to hold their necks in the position of previous shortening. The neck collar must not be of the soft type that is used for *neck* injuries, but rather a hard collar so that the child cannot move the neck at all.

Thyroglossal cyst

This is a unilocular *midline*, subplatysmal, clear, transilluminable cyst that originates somewhere along the embryonal course of the thyroid: from the foramen caecum in the tongue up to the pretracheal position of the thyroid. Clinically, it typically moves when the tongue is stuck out. Anatomically, the communicating tract runs between the cyst and the foramen caecum in close relation to the corpus of the hyoid bone, and therefore the middle segment of the hyoid bone must always be excised during an operation – the so-called *Sistrunk* operation.

Complications that could develop are usually infection of the cyst and abscess formation associated with chronicity, fistulae and scars. Once the diagnosis has been established the cyst should be removed, preferably by an experienced surgeon as a re-operation is fraught with difficulties.

Dermoid cyst

These development cysts are found where embryological migration occurred or at lines of fusion.

The cyst presents at birth or shortly thereafter as a firm, round mass, either on the lateral eyebrow region (so-called angular dermoid), the bridge of the nose, or in the neck or genital regions. An occasional familial link has been reported.

Epidermoid cysts (formerly known as sebaceous cysts), in contrast to dermoid cysts, are usually not present at birth, unless they form part of a syndrome.

Dermoid cysts contain mature ectodermal structures other than bone or nerve. They are filled with keratinous material (Braun-Falco *et al.* 2000). The presence of bone or nerve would favour a diagnosis of a *cystic teratoma*.

Should the cyst present in the midline, as is the case on the bridge of the nose, an intracranial extension should always be excluded.

Regarded by some as the second most common eyelid tumour, the angular dermoid can be removed either endoscopically or through an incision above, or just below, the eyebrow. Current opinion favours direct excision. In all instances, great care must be taken not to injure the frontal branch of the facial nerve, which would leave the child with a permanent eyebrow ptosis if transected.

Midline swellings located over the neural ridge

Swellings found on a line extending from the bridge of the nose, over the vertex of the skull, and down the spinal column up to the coccyx, may include:

- Those of *neuro-ectodermal* origin:
  - meningocoele
  - encephalocoele

  - neurofibroma
  - large-to-moderate sized pigmented naevi, particularly the hairy naevus
- Others:
  - lipoma
  - haemangioma

CAVEAT

Any swelling discovered in the midline should not be removed until intra-vertebral or cranial extension has been excluded by means of either radiographs, computed tomography scans or magnetic resonance scans.

4.2 Malignant tumours

A malignant tumour is an entity that grows. It does not come and go and it does not become larger and then shrink again. It is an abnormal growth and increase of cells.

Early diagnosis of cancer in children

Early diagnosis of this disease is possible when there is a strong suspicion. The general practitioner or paediatrician who sees the child regularly and regularly conducts a complete clinical examination will eventually be 'rewarded' with the early diagnosis of this disease. The prognosis is related directly to early diagnosis, as well as to speedy referral to a central unit where specialised services are available (even if it is only a lymph node biopsy in which touch preparations are necessary for T and B-cell immunocytochemistry).

Cardinal symptoms of cancer in children

- Unexplained fever
- Unexplained pain
- Pallor
- Abnormal bleeding: epistaxis, purpura, petechiae
- Weight loss
- Abnormal masses
- Changes in balance, gait or personality
- Absent redeye reflex or squint

■ FEVER

Differential diagnosis:

Bacterial origin	– responds to antibiotics
Viral origin	– of shorter duration
Neutropenic origin	– high fever, leukaemia
Persistent fever	– lymphoma
	– necrosis in neuroblastoma
	– histiocytosis X

■ PAIN

- Diffuse bone pain and arthralgia – In 15–23% of patients, leukaemia must be distinguished from rheumatoid arthritis and acute rheumatic fever.
- Focal bone pain – Bone tumours in particular are associated with swelling and tenderness. If the pain is around the knee joint or shoulder joint, particularly in teenagers, consider it as malignant until otherwise proven.
- Abdominal pain is usually a late sign of large intra-abdominal tumours.
- Headache is a classic sign of intracranial tumours, except in the small child with open sutures.
  - The older child with a raised intracranial pressure usually has repetitive early morning headaches and vomiting, which later become constant.
  - In small children, hyperstimulability is a sign of pain.

■ MASSES

- Abdominal – This mass is usually already large when a diagnosis is made:
  - Abdominal masses are malignant until proven otherwise.
- Masses at unusual sites, e.g. on the cheeks or head, are suspicious.
- Masses around the knee or shoulder joint are cause for concern.
- Persistent lymphadenopathy – indications for biopsy:
  - Gland enlarges within 2 months
  - Gland does not shrink within 4–6 weeks
  - Gland not normal size within 8–12 weeks
  - No response to a course of oral antibiotics
  - Matted glands
  - Associated hepatosplenomegaly
  - Generalised lymphadenopathy
  - Glands in the posterior triangle of the neck and supraclavicular are very suspicious.

■ ABNORMAL BLEEDING

The abnormal bleeding can include epistaxis (nose bleeds), petechiae (point bleeding in the skin) and purpura (larger bleeding in the skin). A primary disease, e.g. leukaemia, a secondary infiltration by lymphoma or any other small round cell tumour, will destroy the bone marrow with a consequent cytopenia of red blood cells or platelets.

A platelet deficiency has the following effects:

- bruising out of proportion to the level of injury
- multiple petichiae
- epistaxis (nosebleed) – repetitive or constant

- bilateral peri-orbital ecchymosis and proptosis

■ PALLOR

A primary disease, e.g. leukaemia, a secondary infiltration by lymphoma or any other small round cell tumour, will destroy the bone marrow with a consequent cytopenia of red blood cells or platelets.

Cytopenia of red blood cells has the following effects:

- normochrome normocytic anaemia
- reticulocytopenia

■ CHANGE IN BALANCE, GAIT OR PERSONALITY

The following early signs must draw your attention to the possible development of intracranial tumours:

- Posterior fossa tumours – abnormal gait and balance
- Supratentorial tumours – convulsions
- Midline tumours – personality changes which take place suddenly in a previously normal child

Some parents just find 'something wrong' with the child.

Raised intracranial pressure is a late sign of brain tumours.

■ EYE CHANGES

- Cat's eye reflex – white pupil in case of a retinoblastoma
- Strabismus (squint) – developed suddenly in a previously normal child
- Decrease in vision, usually of sudden onset

■ FACIAL NERVE PARALYSIS

Sudden increased pressure at any site along the path of the nerve must be regarded with great suspicion, e.g.:

- Middle ear – rhabdomyosarcoma
- A basal nucleus which is compressed between the brain, the meninges and the base of the skull, by
  - brain tumours
  - leukaemia
- In the course through the parotid, the nerve path may be infiltrated by a parotid carcinoma.

Special investigations that should be conducted in any paediatric patient with a suspected cancer:

- When leukaemia/lymphoma is suspected:
  - full blood count
  - urea, uric acid, calcium, electrolytes (**NB:** urate)
  - liver function tests and enzymes
  - X-rays of the chest
  - X-rays of areas of severe bone pains

- When solid tumours are suspected:
  - the site is clinically documented
  - ultrasound determines: size, anatomical position and whether solid, cystic or necrotic
  - overview X-ray of the abdomen, chest or limb

Special investigations – how, where and when (DeVita *et al.* 2005; Pizzo and Poplack 2002)

Refer the child as soon as possible to a paediatric oncology unit at the nearest academic hospital where the paediatric oncologist, paediatric surgeon and other members of a highly specialised multi-disciplinary team will take over the management of the patient.

Requests by the paediatric oncologists that primary care doctors refer children with suspected cancers speedily to a central academic hospital can easily be motivated.

The chance of survival is much higher when the primary management is conducted in a central facility rather than on the periphery.

Unnecessary surgery is avoided in this manner because it is often necessary for the child at an academic hospital to undergo repeated procedures:

- For complete staging of the disease
- To obtain fresh diagnostic material for electron microscopic studies, tissue culture, biochemical analysis, surface marker studies and cytogenetics.

It is difficult to justify surgery in an institution which is not equipped to offer pre-operative diagnostic studies or definitive post-operative therapy, especially if a doctor who seldom sees a tumour in children performs the surgery.

Even the experienced paediatric pathologist sometimes finds it difficult to distinguish between a mesoblastic nephroma or an anaplastic Wilm's tumour (the histologically unfavourable type).

Electron microscopy is essential, e.g. in the identification of ultrastructures of small round cell tumours. This is an investigation that is not extensively carried out.

Specific chromosomal abnormalities have now been identified in a whole range of paediatric tumours, but chromosomal studies cannot be performed on samples that have been contaminated or fixated.

Precise anatomical and clinical staging of subgroup classification is an essential part of the planning of therapy and can only be done during primary surgery.

For most paediatric cancers, the primary therapy is a very important and decisive factor which will determine the survival of the child.

If there has already been a relapse of the disease, there is a notably smaller survival rate. The involvement of the primary care

doctor can actually be supportive to the child and the family, especially if much of the treatment is conducted outside the hospital. The costs of follow-up treatment can be cut by up to 40% if the primary doctor is involved in the treatment of the patient. Good communication between the central facility and the primary care doctor is therefore of the utmost importance.

4.3 Malignancies in children

In South Africa, mortality due to malignancy is the third highest cause next to trauma and infectious disease.

In comparison with malignancies in adults, there are many differences in children in terms of:

- cell type – often an embryonal cell type
- biological behaviour – active tumour metabolites with clinical symptoms (see Table 4.1)
- more favourable response to therapy
- organs affected:
  - organs affected in the child are the kidneys, adrenals, brain, liver, muscle, bone, etc.
  - organs usually involved in the adult are those of procreation or those subject to abuse, e.g. the uterus, prostate, mammary glands, or organs that undergo metaplasia due to long-standing exposure to carcinogens, e.g. the oesophagus, stomach, colon, lung and liver (cirrhosis).

Prognosis

The basis for therapy is the combination of different modalities, e.g. surgery, radiotherapy and chemotherapy. It is very important and advantageous for the child that he or she be immediately referred to a paediatric oncology unit where all therapy can be conducted under one roof.

The danger signs, as indicated by the Cancer Association, are important as they draw attention to possible malignancy. Early diagnosis remains an important factor in the prognosis of a patient.

There are three main groups of malignancy:

- Reticulo-endothelial origin:
  - Leukaemia 27%
  - Lymphoma 9,5 – 36,5%
- Central nervous system 13%
- The rest
  - neuroblastoma 8%
  - nephroblastoma 12%
  - soft tissue sarcoma 7%
  - bone tumours 3,5%

- teratoma 2%
- retinoblastoma 3%
- hepatoblastoma 1%
- diverse 14% – 50,5%

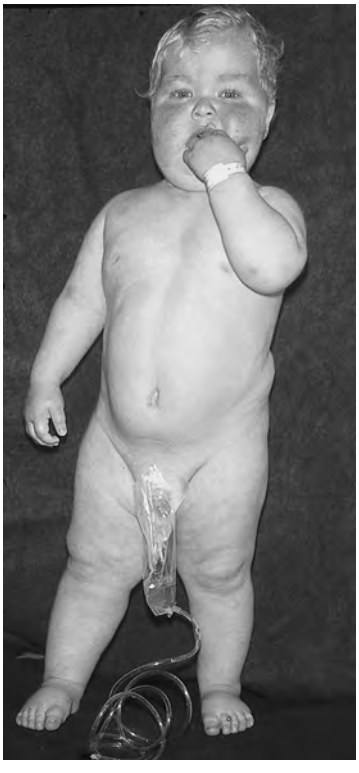


Fig. 4.5 Cushing's syndrome.

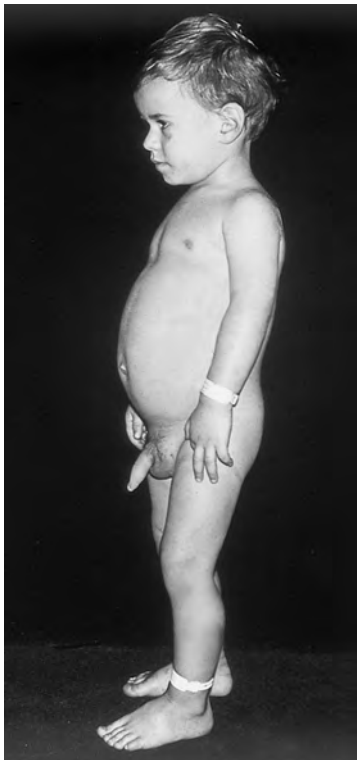


Fig. 4.6 Virilising adrenal tumour in a 2-year-old boy.

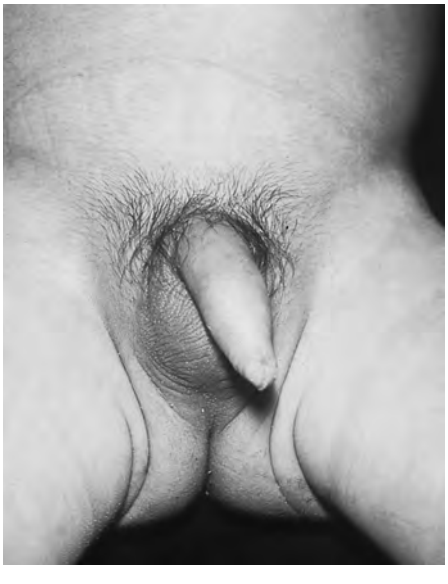


Fig. 4.7
Virilising adrenal tumour in a 2-year-old boy (close-up).

TABLE 4.1 Tumour metabolites with clinical symptoms

Tumour	Metabolite	Symptoms
Neuroblastoma	Catecholamines VIP	Hypertension Diarrhoea
Nephroblastoma	Renin	Hypertension
Pheochromocytoma	Catecholamines	Hypertension
Beta cell tumour	Insulin	Hypoglycaemia
Carcinoid	5HIAA	Carcinoid syndrome
Granulosa cell tumour	Oestrogen	Precocious puberty
Adrenal carcinoma	Cortisone Androgens	Cushing's disease Precocious puberty

The above-mentioned metabolites can also serve as a biological or biochemical marker, in other words the effective control of the tumour will allow the concentration of the metabolite to decrease or disappear. It is a highly effective way of monitoring the course of the disease.

There are also other biochemical markers, but they are not biologically active.

Alpha-fetoprotein (AFP) increases dramatically with:

- hepatoblastoma
- yolk sac tumours:
  - embryonal carcinoma
  - endodermal sinus tumour

Degeneration of a tumour

It is often in this way that the tumour comes to the attention of the parent or the doctor because the tumour suddenly enlarges due to bleeding or necrosis and pain is experienced. A tumour that previously grew uninhibited now becomes palpable and can lead to a diagnosis.

Special investigations that will be conducted by the referring hospital

X-RAYS

- Abnormal calcifications will be visible.
- Displacement of other viscera in the abdomen will be visible.

CONTRAST STUDIES

- An excretory urogram can be diagnostic of a nephroblastoma.
- A barium meal or enema can demonstrate the displacement and infiltration of hollow viscera.
- An arteriogram can be invaluable in determining the blood supply prior to surgery.
- Ordinary chest plates and skeletal plates can demonstrate metastases.

■ FULL BLOOD COUNT

This is cardinal in the diagnosis of leukaemia, and anaemia is commonly present with other tumours.

■ BONE MARROW ASPIRATION

This is diagnostic in leukaemia, metastatic neuroblastoma, rhabdomyosarcoma, non-Hodgkin's lymphoma and Ewing's sarcoma.

■ BIOCHEMICAL INVESTIGATIONS

The metabolites described in Table 4.1 must be evaluated, e.g.:

- urea, uric acid, creatinine, electrolytes – particularly in diagnosing and monitoring uric acid nephropathy and renal failure
- liver function tests, particularly LDH, which is a very important non-specific marker
- urine microscopy – haematuria may indicate bladder rhabdomyosarcoma or nephroblastoma.

■ ULTRASOUND

This is a non-invasive investigation which can be applied to advantage in the diagnosis of a tumour. It does in fact have one disadvantage, namely that it is operator-dependent and not all operators are capable of interpreting the image correctly.

4.4 Staging of tumours

The general principles of staging are the following, with small individual variations in different tumours:

Stage I

Tumour limited to the organ.

Stage II

Local infiltration of the tumour around the organ of origin, with or without local lymph nodes being affected. At this stage, it is completely resectable.

Stage III

Infiltration of the tumour into the surrounding organs and direct spread into the lymph nodes. It cannot be completely removed surgically. Massive intra-abdominal tumours are also viewed as Stage III tumours, not due to infiltration of glands, but due to the irresectability of the tumour.

Stage IV

Systemic dissemination of the tumour via haematogenous or lymphogenic spread.

Management in view of the staging

If the tumour can be surgically completely removed during Stage I or II, then the prognosis is very good. The chemotherapist must be involved at all times in the treatment of the tumour as some tumours will require chemotherapy during treatment, before and/or after surgery, depending on the completeness or incompleteness of the removal of the tumour. Tumours already in Stage III or IV have spread so far that chemotherapy and radiotherapy are sometimes all that can be offered to the patient. The successful treatment of a malignancy with chemotherapy and radiotherapy has long-term side effects.

4.5 Nephroblastoma (Wilm's tumour)

This is a tumour of embryonal renal tissue. It commonly appears before the age of 5 years. The tumour is mostly discovered by chance when the mother dries the child after a bath. Signs such as anorexia, haematuria, fever, hypertension and weight loss are common when sought.

Clinically, it is a flank tumour (differential diagnosis is neuroblastoma, hydronephrosis, polycystic kidney). These tumours may already be massive during the first examination and have aptly been described as a "watermelon under the skin".

Staging

The same as mentioned above, except that an additional Stage V is added when the tumour is present in both kidneys either synchronously or metachronously.

Special investigations

- Abdominal overview X-ray:
  - A small percentage show calcifications in the tumour
- Lung X-rays:
  - Possible metastases
- Ultrasound of the abdominal organs:
  - Confirms whether the tumour is in the kidney or not
  - Look for lymph node metastases
  - Liver metastases
  - Examine the contralateral kidney for a second primary tumour
  - Tumour in IVC
- A CT scan can replace an excretory urogram to demonstrate excretion and to guide the surgeon in planning the operation
- Urea, electrolytes and creatinine
- Liver function tests and LDH – non-specific tumour markers and indicators of the severity of the disease present
- Histology

Management

Combination therapy of surgery and chemotherapy for all stages. Radiotherapy is added for Stages III and V and, where indicated, for Stage IV.

Prognosis

The availability of chemotherapy has dramatically improved the prognosis of these children. In the past when only surgery was offered, the overall survival figure was only 10%. The combination of chemotherapy and radiotherapy, as well as surgery, has dramatically improved the prognosis. In Stage I there is, e.g., an 85–90% chance of 2-year disease-free survival.

4.6 Neuroblastoma

This originates from embrional nerve cells in the neural crest, in other words, the adrenal medulla and sympathetic ganglia along the vertebral column. The course is as follows:

- There is a flank tumour with a differential diagnosis of nephroblastoma, hydronephrosis and polycystic kidney.
- These children are usually obviously ill.
- The tumour spreads early to the bone marrow, the bone cortex of the long bones, the skull, the lymph nodes and the liver. If the tumour is located along the vertebral bodies, it can infiltrate via the intervertebral foramina into the spinal canal.

This is a highly malignant tumour. The most common site of appearance is the adrenals, but it may appear anywhere along the sympathetic chain.

Due to early metastases, it must be suspected in the following cases:

- Lymph node swellings in the posterior triangle of the neck
- Sudden paraplegia
- Proptosis, nodule on the skull
- Pain and swelling in the long bones
- Failure to thrive
- Malignant malaise, namely fever, weight loss, fatigue and fleeting pains in the limbs

In 65% of all cases there are already metastases during the first diagnosis of the disease.

Staging

Much like the previously discussed tumour, except:

- Stage IV–S – Like a Stage I or II, both with distant disease limited to one or more of the following systems: liver, skin and bone

marrow, in a baby less than 1 year of age without radiological proof of bone metastases.

Special investigations

- Abdominal X-ray – shows typical calcifications in the tumour
- Excretory urogram – shows a tumour on the outside of the kidney that displaces the kidney
- Ultrasound – determines the size of the tumour and the presence or absence of metastases in the liver and lymph nodes
- Bone marrow aspiration for malignant cells
- Skull and long bone X-rays for metastases
- 24-hour urine sample to determine the presence of active metabolites (VMA, metanephrines and normetanephrines)
- CT scan and MRI according to discretion
- Histology

Management

Combination of surgery, chemotherapy and radiotherapy.

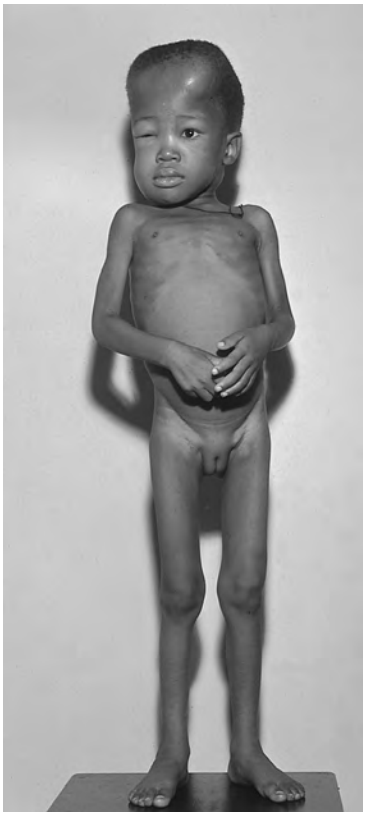


Fig. 4.8 Neuroblastoma metastasis.

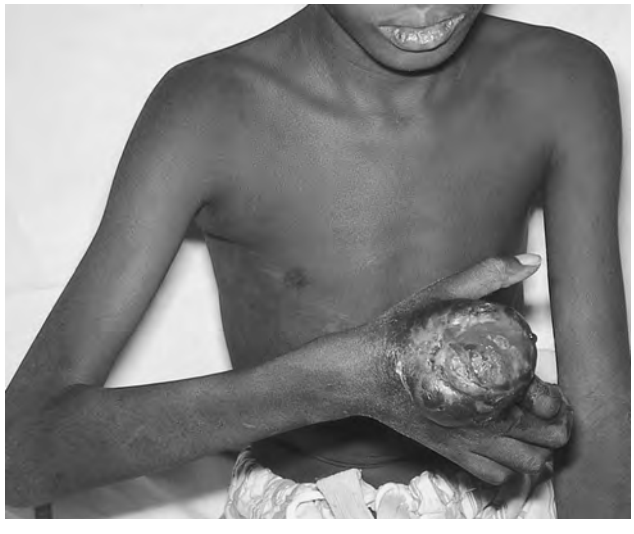


Fig. 4.9 Sarcoma, right hand.

Prognosis

The prognosis is generally poor (in comparison with nephroblastoma, chemotherapy has not improved the prognosis in neuroblastoma).

■ TWO-YEAR SURVIVAL

- Stage I = 100%
- Stage II = 87%
- Stage III = 67%
- Stage IV = 20%

Stage IV–S tumours have the best prognosis, sometimes with complete recovery.

Extra-abdominal tumours, e.g. the neck, thorax and pelvis, have a better prognosis because these cases are diagnosed earlier due to local pressure symptoms.

4.7 Hepatoblastoma

This develops from embryonal liver cells. It is the most common primitive liver tumour in children. It always appears before the child's fifth birthday, with a boy-to-girl ratio of 2:1.

Clinically, there is generalised malaise, weight loss, vomiting, abdominal pain and swelling.

Special investigations

- Ultrasound confirms a mass in the liver.
- The serum alpha-fetoprotein concentration is high, usually very high.
- A CT scan will determine tumour size.
- Ultrasound will determine tumour size.
- Histological confirmation is needed.

Management

The only chance of survival with these children is complete surgical resection of the tumour. At present, chemotherapy is given preoperatively for 2–4 months in order to simplify the surgery.

Prognosis

The prognosis depends on the extent of the tumour in the liver. If it can be surgically completely resected, then the chance of survival is 100%. If it is incompletely removed or is inoperable, then all children die within a year.

Dictum

A liver mass in a child under the age of 5 years with a raised serum alpha-fetoprotein level must be considered to be a hepatoblastoma until this diagnosis is proved incorrect.

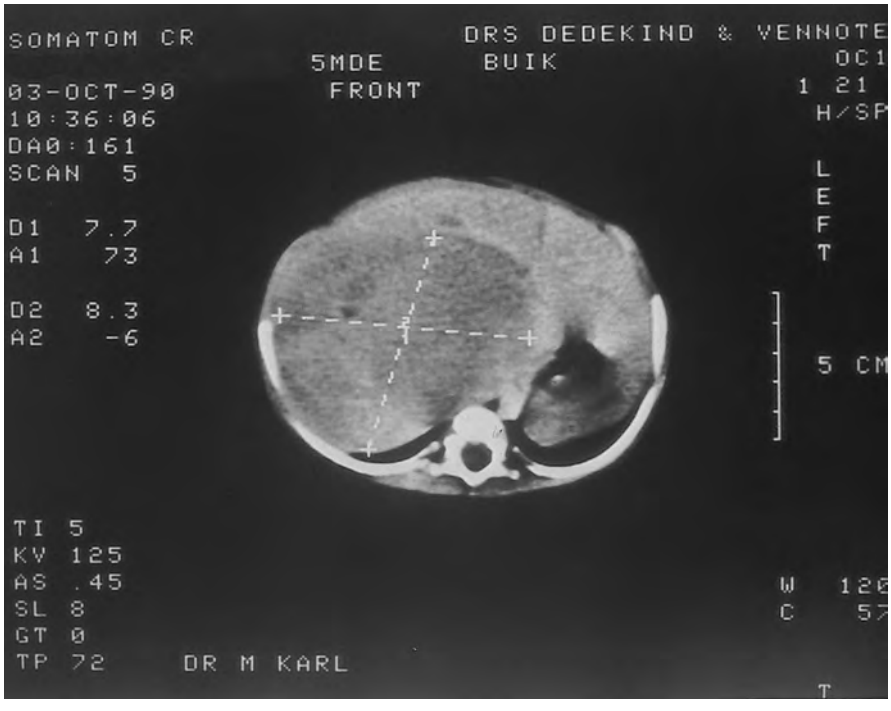


Fig. 4.10 Hepatoblastoma – right lobe of the liver.

4.8 Rhabdomyosarcoma

This is a malignant tumour of muscle and appears as commonly as nephro- and neuroblastomas.

The most common site of appearance is the head and neck region, which includes the orbit. The rest appear in the genito-urinary tract or on the trunk or limbs.

There are four important histological types:

- embryonal
- alveolar
- pleomorphic
- anaplastic

Embryonal rhabdomyosarcoma

- This is common in the genito-urinary system and to a lesser extent in the head and neck region.
- The prognosis is much better than that of the alveolar type.

Genito-urinary embryonal rhabdomyosarcoma

- In the genito-urinary tract it presents as:
  - urinary obstruction
  - suprapubic mass
  - vaginal bleeding
- It often looks polypoid or botryoid.
- The management is a combination of:
  - surgery
  - chemotherapy
  - radiotherapy
- Many of the tumours are particularly sensitive to chemotherapy, which in some series would be the primary form of treatment.
- Radiotherapy and radical surgery are relevant if chemotherapy fails.
- The recovery figures for genito-urinary rhabdomyosarcoma are in the region of 75%.

Head and neck rhabdomyosarcoma

Embryonal rhabdomyosarcoma of the scalp, face and parotid, where radical surgery is a possibility, has a good prognosis. By contrast, lesions of the nasopharynx, pterygoid fossa and inner ear have a poor prognosis due to the inaccessibility of the tumours.

Orbital rhabdomyosarcoma is usually diagnosed early and lends itself to radical surgery, e.g. a total evisceration of the eye which improves the prognosis to a survival of 90%.

Alveolar rhabdomyosarcoma

- Mostly located on the trunk and limbs.
- The natural course is less favourable than with the embryonal type.
- In comparison with the embryonal type, which reacts well to chemotherapy, the first choice of management is total removal by means of surgery, followed by radiotherapy and chemotherapy.
- The overall survival is only 50%.

4.9 Teratoma

Teratoma consists of three germ cell layers and can therefore contain any type of tissue (e.g. brain, thyroid, ovaria, etc.).

The sites where it commonly appears are:

- sacrococcygeal
- abdominal
- mediastinal

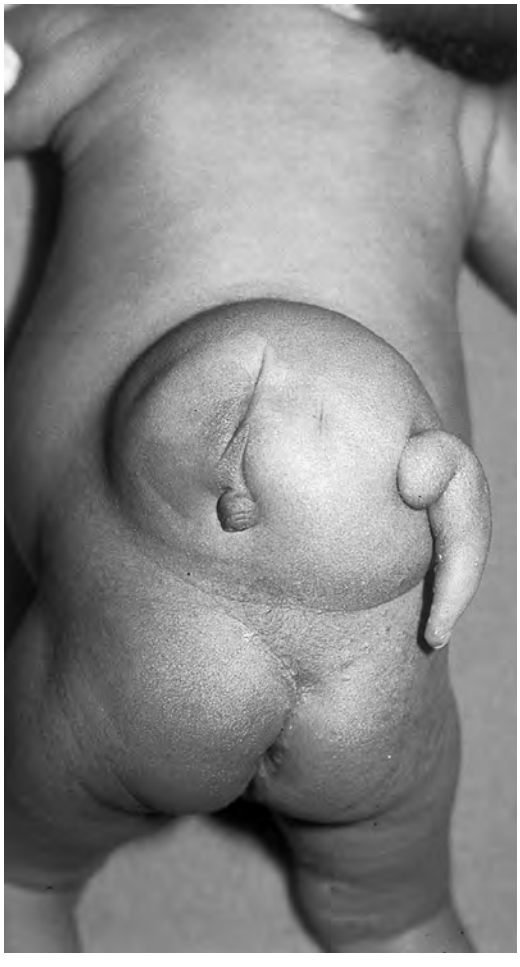


Fig. 4.11
Posterior midline teratoma.

Sacrococcygeal teratoma

The tumour can be located precoccygeally, with an extension into the abdomen – palpable as an abdominal tumour or an extension posteriorly (outside). The latter type is visible as a massive appendage between the anus and the coccyx. The importance of this tumour is its malignant potential, which is given as high as 35% in some reports. Early surgery is very important to limit progress to malignancy. The longer the tumour is left, the greater the malignant potential.

The endodermal sinus or yolk sac tumour is the most common malignant tumour of the teratomas. This is the reason for the associated rise in serum alpha-fetoprotein.

■ EARLY MANAGEMENT

Surgery is recommended in all cases. Removal of the tumour in its entirety is the absolute ideal and therefore an attempt to biopsy and make a histological diagnosis is not recommended.

NB: Surgery must be conducted before the age of 1 month.

■ PROGNOSIS

This depends on the local malignant potential of the residual tumour. If the tumour is more solid than cystic, then there is a greater possibility for malignant local recurrence.

NB: Surgery of this nature must not be attempted by the coincidental paediatric surgeon.

Abdominal teratoma

It appears retroperitoneally in boys and girls and must be distinguished from other flank tumours (nephroblastoma, neuroblastoma, hydronephrosis). In girls it also commonly occurs in the ovaries.

On X-rays, the abdominal teratoma can demonstrate typical calcifications of a tooth or bone in the stroma.

Like the sacrococcygeal teratoma, the abdominal teratoma also has malignant potential and surgery should be performed as soon as possible.

Mediastinal teratoma

The same principles as for the other teratomas also apply here.

The differential diagnosis of anterior mediastinal tumours is the Three Ts, namely:

- Thyroid
- Thymus
- Teratoma

4.10 Brain tumours in children

HP SHAPIRO

Brain tumours are the second most common tumour in children, leukaemia being the commonest form of cancer. Central nervous system tumours are the commonest solid tumour in children and account for 20–30% of tumours in children younger than 18 years of age. Approximately 60% of brain tumours in children are infratentorial.

Classification

Primary brain tumours have been classified by the WHO according to their cell of origin.

Tumours occurring in children include astrocytomas, optic nerve gliomas, brainstem gliomas, ependymomas, choroid plexus tumours, medulloblastomas, germinomas and craniopharyngiomas. (Note: The term 'glioma' includes astrocytomas, ependymomas and oligodendrogliomas).

Primary intracranial childhood tumours according to predominant site:

- Supratentorial
  * Cerebral hemispheres:
    – Astrocytoma
    – Ependymoma
    – Primitive neuroectodermal tumours
  * Midline tumours:
    – Craniopharyngiomas
    – Optic nerve and hypothalamic gliomas
    – Pineal tumours, e.g. germinomas
- Infratentorial
  * Cerebellum and IV ventricle:
    – Cerebellar astrocytoma
    – Medulloblastoma
    – Ependymoma
    – Brainstem glioma

Clinical manifestations

Brain tumours present in many ways depending on age, location, type of tumour and rate of growth.

Patients can present with signs and symptoms of raised intracranial pressure or focal neurological signs.

Signs and symptoms of raised intracranial pressure (ICP) include:

Headaches: Often worse in the morning, improving through the course of the day. Headaches should be viewed with suspicion in a child.

Vomiting: Particularly associated with tumours in the region of the IV ventricle. Vomiting is often unexpected, projectile and may not be preceded by nausea. The patient's appetite often remains good despite the vomiting. Gastro-intestinal causes of vomiting must be excluded.

- Papilloedema - not always present
- A full fontanelle
  * Macrocephaly or rapid growth in head circumference in infants with an open fontanelle and sutures.
- Abducens nerve paresis
  * Decreasing visual acuity either as a result of chronic raised intracranial pressure or direct pressure on the optic apparatus
- Upward gaze palsy (Parinaud's syndrome)

Supratentorial tumours

Clinical signs depend on the location of the tumour. Signs and symptoms of raised ICP may be present. Focal neurological signs and epilepsy are signs of hemispheral tumours. Altered level of

mentation, deterioration in school performance and personality changes are also seen. Compression or involvement of the visual apparatus, e.g. craniopharyngiomas and optic gliomas may result in deteriorating visual acuity and visual field defects. Endocrine involvement may result in obesity, polyphagia, or delayed or precocious puberty. Pineal region tumours may present with a Parinaud's syndrome.

Infratentorial tumours

Obstruction of CSF pathways occurs more commonly with cerebellar tumours and results in hydrocephalus. Truncal ataxia occurs with vermis (midline) tumours and limb ataxia with hemispheral involvement. Vertical nystagmus occurs with brainstem involvement, and horizontal nystagmus with cerebellar tumours. Head tilt and neck stiffness may indicate tonsillar herniation through foramen magnum. The older child should be examined for the presence of an intention tremor, dysdiadokokinesia and dysmetria.

Brainstem involvement may present with a mixed picture of long tract signs and cranial nerve involvement.

■ DIAGNOSIS

When the clinical history and physical examination suggest the presence of an intracranial lesion, a CT scan or MRI with contrast is indicated. A lumbar puncture in a patient with a suspected intracranial lesion is contra-indicated.

■ MANAGEMENT

Surgery is the mainstay in the treatment of intracranial tumours. The extent of the removal has an impact on long-term outcome. Complete resection is often curative as in the case of a cerebellar astrocytoma. Radiation therapy is often used as an adjunct in the management of incompletely resected tumours.

The increasing role of chemotherapeutic agents in the management of brain tumours is still under investigation.

Dexamethasone is prescribed peri-operatively for cerebral oedema. Anti-epileptic agents are prescribed as necessary, as is hormone replacement therapy.

■ PROGNOSIS

Prognosis and long-term survival depend on the histological type of tumour, clinical condition of the child and the extent of tumour resection.

Low-grade tumours have a better prognosis than high-grade tumours.

A multi-disciplinary team is required in the management of these patients and includes neurosurgeons, radiotherapists, chemotherapists, nursing personnel, rehabilitation personnel, social workers and neuropsychologists.

BGP LINDEQUE

Musculoskeletal tumours refer to soft tissue tumours and benign tumours which occur in the musculoskeletal system. A wide variety of tumours are seen, namely benign bone tumours, malignant bone tumours, and benign tumours of fatty tissue, connective tissue and muscle, as well as malignant tumours of fat, connective tissue and muscle.

Relative to malignancies of the bronchus, breast, prostate and colon, tumours of the musculoskeletal system are rare. It is therefore absolutely necessary that these tumours be managed in a specialised unit where the necessary skill is available to give proper attention to these patients.

Musculoskeletal tumours grow along the path of least resistance, which is determined by anatomical borders. The medullary canal of long bones offers very little resistance to marrow cell tumours and it is for this reason that Ewing's sarcoma, lymphoma and leukaemia easily spread intramedullarily. On the other hand, the periosteum is an effective barrier to an aggressive tumour and can contain a tumour for a while within the bone. A tumour also tends to spread longitudinally in a muscle before it penetrates the thick fascia layer to enter a neighbouring muscle group. Blood vessels and nerves are usually pushed aside by the tumour and it is seldom that a large blood vessel or nerve is penetrated by the tumour.

Of great importance is the fact that a tumour can be much more accurately assessed if the clinical course, the radiological image and the histological level can be taken into consideration (as opposed to histology only).

Definitions

■ LATENT STATIC TUMOURS

These lesions are usually asymptomatic and are discovered by chance; they do not enlarge at all within the bone.

■ LOCALISED ACTIVE BENIGN LESIONS

These tumours are mildly symptomatic, grow very slowly and may present with a pathological fracture or mechanical dysfunction.

■ AGGRESSIVE BENIGN TUMOURS

These tumours are often discovered due to pain, a pathological fracture or a large growing mass. These tumours penetrate through the surrounding capsule and enter the muscle.

■ LOW-GRADE SARCOMAS

These lesions are slow-growing masses and may present with pain

or are sometimes discovered due to a mass. The surrounding tissue has time to form a reactive zone and is displaced by the slow-growing malignant tumour.

■ HIGH-GRADE SARCOMAS

These are painful, rapidly growing inflammatory masses, often associated with pathological fractures or severe limb dysfunction. They grow so rapidly that surrounding tissue does not always have time to form a capsule and natural barriers in a limb are ineffective in barricading them.

Staging system

It is absolutely necessary that tumours of the musculoskeletal system, just like those of other regions, e.g. the thyroid or breast, are properly staged before they can be treated. Results with regard to management cannot be reported accurately if the tumours have not been properly staged. Staging of musculoskeletal tumours consists not only of the histological grading, but also takes into consideration the clinical course, radiological image and blood profile, as well as the histological image.

It is also absolutely necessary that the staging of any tumour should precede histological confirmation of diagnosis due to the fact that biopsies cause bleeding and disturb the tissue planes, and in doing so jeopardise staging.

Staging of musculoskeletal tumours depends on:

- grading
- site
- metastases

■ GRADING

By this term we understand the biological aggressiveness, including the histologically aggressive nature, radiological investigations and clinical course in terms of growth time, size, temperature and biological markers.

■ SITE

Here we see whether the tumour has spread intracapsularly, intracompartmentally or extracompartmentally.

■ METASTASES

Lung metastases, distant metastases and regional lymph node metastases are considered to be part of metastatic disease.

■ RADIOGRAPHIC STAGING MODALITIES

The modalities that are used to stage musculoskeletal tumours include an ordinary X-ray film, bone isotope scans, arteriography, magnetic resonance imaging (MRI) and CT scans.

■ X-RAYS

Ordinary X-rays are still the most important modality in the initial diagnosis of musculoskeletal tumours. The differential diagnosis of bone tumours is made on the basis of the radiological image taken with ordinary X-ray apparatus. The other special investigations indicate the precise spread and anatomical extent of the tumour.

■ ISOTOPE SCANS

Technetium scintigraphy is used to indicate the multiplicity or the spread of metastases in bone or soft tissue sarcomas to other bones in the skeletal system.

■ ARTERIOGRAPHY

Nowadays arteriography is only used to determine the effect of chemotherapy and to evaluate and localise large blood vessels in relation to the tumour.

■ CT SCANS

CT or MRI plays a very important role in the staging of musculoskeletal tumours. Anatomically, these tumours can be perfectly localised by means of these scans.

■ BIOPSIES

Taking a biopsy of musculoskeletal tumours is of cardinal importance. The placement of a biopsy can jeopardise a limb-sparing procedure and cause a patient to lose that limb, instead of simply undergoing a local resection. The following rules are of cardinal importance in taking a biopsy:

- A biopsy should never interfere with the final excision of a tumour.
- The incision should always be removed *en bloc* with the tumour during final excision.
- The biopsy should be done by the same surgeon who will perform the final excision of the tumour.
- An incision biopsy is used for suspected malignant lesions.
- Excision biopsies (marginal excision) are indicated for suspicious benign lesions.
- A biopsy incision should always be along the long axis of a limb and never parallel to it or transverse. It should run parallel to the neurovascular bundles. It should be placed far enough from a neurovascular bundle so that the neurovascular bundle does not have to be included in the final excision of a tumour operation.
- The biopsy should be properly marked in terms of where proximal, distal, medial and lateral is to simplify the examination by the pathologist.
- A pathologist should preferably be present in the operating theatre when the biopsy is done so that he can determine whether

sufficient representative tissue has been taken for a final diagnosis.

- If a Portovac is used, then the exit of the Portovac on the skin should be in line with the longitudinal biopsy incision and not to either side of it. This Portovac exit should be included during the final excision.
- It is preferable not to do biopsies under local anaesthesia, but rather to use regional blocks or general anaesthesia. Local anaesthesia injected into the area can make the diagnosis more difficult.
- It is important to give the pathologist a piece of tissue of sufficient size, and also to inform him or her properly about the clinical history, special investigations and X-rays that have gone before. This information will aid the pathologist in making a diagnosis.
- A biopsy must not be delayed for weeks after a patient has been seen with the possible diagnosis of malignancy. All staging tests should be completed within a week and the biopsy should be done the following week.
- If there is any suspicion that the diagnosis might be an infection, then tissue should be sent away for culture and sensitivity tests.
- Frozen sections are of importance in theatre if the tissue does not have much bone. A frozen section can be a very good indication of what the tumour contains. If it is a malignancy, then the incision biopsy is sufficient for the time being. If the diagnosis is a benign tumour, one can go ahead directly with the final surgery.

Principles in the management of musculoskeletal tumours

Benign tumours can be removed with local excision in the soft tissue or by curettage in the bone. The cavities are filled with bone transplantation.

If low-grade malignant tumours are diagnosed, then these tumours are resected with a border of normal muscle tissue, connective tissue or bone between the tumour and the line of resection.

Highly malignant aggressive tumours are resected with radical operations, e.g. an amputation or radical excision of the tumour. Chemotherapy is initially administered in these cases to shrink the tumour's inflammatory reaction and to establish better margins. Surgery is then performed and the sample analysed for the percentage of tumour necrosis, which is of prognostic importance.

The management of malignant tumours is done by a team consisting of the surgeon, medical oncologist, radiotherapist, pathologist, nursing staff, physiotherapist, occupational therapist and trained cancer counsellors. These patients are discussed at multidisciplinary team meetings before management is begun as the

treatment must be individualised. As a rule, patients with highly malignant tumours receive chemotherapy first before an operation, followed by chemotherapy.

CAVEAT

It is of cardinal importance that if the patient presents with a swelling of a limb and a tumour is suspected, then the patient should be referred to the nearest referral centre where he or she can be diagnosed for further management.

4.12 *Leukaemia* (Greaves 2002; Kersey 1997)

MARIANA KRUGER & RUELLYN L COCKCROFT

Definition

- Leukaemia is the rapid uncontrolled proliferation of white blood cells, which can be increased in the circulating blood. The disease is fatal if untreated.
- Leukaemia is a primary malignancy of blood-forming organs in which the normal blood-forming cells are replaced with blast cells.

Incidence

Leukaemia is the most common malignant condition occurring in childhood. It occurs mostly before the age of 15 years, with a peak incidence at 4 years of age.

Types of leukaemia in children and occurrence

Acute lymphocytic leukaemia	75%
Acute myeloid leukaemia	20%
Chronic myeloid leukaemia	3%
Other rare types	2%

Aetiology

Leukaemia is a clonal disease, derived from a single cell, and characterised by an uncontrolled proliferation of this immature haematopoietic cell with arrested or aberrant differentiation. The causes of leukaemia are mostly unknown, but recent studies on neonatal blood spots and monozygotic twins suggest a prenatal origin for acute lymphocytic leukaemia (ALL). Certain syndromes are associated with an increased risk of developing acute leukaemia, of which Down syndrome (trisomy 21) is the most common. Viruses are associated with T-cell leukaemia (HTLV = human T-cell lymphoma leukaemia virus) and B-cell leukaemia (Burkitt-type leukaemia and lymphoma). The chances that a brother or a sister

will develop the disease are approximately 1:700 and 1:5 for the other member of an identical twin.

■ CLINICAL PROFILE

The general symptoms are:

- malaise
- fatigue
- persistent or unexplained fever which does not respond to antibiotics

Symptoms due to bone marrow disease:

- pallor (normochromic normocytic anaemia) – decreased red blood cells
- petechia, purpura, ecchymoses, epistaxis, bleeding gums – decreased platelets
- infections – upper respiratory tract and lungs due to decreased neutrophils
- severe diffuse bone pain
- arthralgia or joint pain

Organs that are involved and that enlarge:

- lymph nodes – generalised lymphadenopathy
- liver
- spleen
- mediastinum – thymus in T-cell leukaemia

Organs that are infiltrated:

- brain – present as a meningitis or with a cranial nerve paralysis, especially N. VI
- testes – in boys
- ovaria – in girls (very rare)
- kidneys

■ SPECIAL INVESTIGATIONS

If you suspect that a child has leukaemia or a lymphoma, then you should refer the child as soon as possible to an established paediatric oncology centre, and do the minimum of special investigations before referral as these may be time-consuming and fatal. Such special investigations are:

- Full blood count
  - haemoglobin (Hb) decreased or normal
  - white blood cells raised, normal or decreased
  - platelets decreased or normal
  - blasts on peripheral smear
- Urea, electrolytes, calcium, magnesium and phosphate – urea can be raised with direct infiltration of the kidney

- Uric acid pool is overloaded due to the excessive breakdown of malignant cells; this causes a uric acid nephropathy which can lead to renal failure
- Liver function tests showing a raised LDH indicate liver infiltration. LDH is a non-specific marker for disseminated malignancy in children, particularly leukaemia, lymphoma and neuroblastoma.
- A chest X-ray may demonstrate an enlarged thymus and hilar lymph nodes – these can cause a superior vena cava syndrome which requires emergency chemotherapy.

Refer the child as soon as possible to the nearest academic hospital, with the following treatment:

- Analgesia (also helps for fever) – paracetamol, codeine, morphine. **NB**: No salicylates are administered because they may encourage a bleeding tendency.
- Good oral/intravenous fluid intake
- Allopurinol 8 hourly *per os* to help prevent uric acid nephropathy
- Stop active bleeding by platelet transfusion or nasal plugs
- Red blood cell transfusion in severe anaemia (< 5 g/dl) or if in cardiac failure

■ SPECIAL INVESTIGATIONS AT THE PAEDIATRIC ONCOLOGY CENTRE

- Bone marrow aspirate: An increase in immature white blood cells (blasts) of more than 25% in a bone marrow aspirate is diagnostic of leukaemia (ALL or AML). The French-American-British (FAB) morphologic classification system is used to diagnose the three morphologic subtypes in ALL.
  - morphology and staining (PAS) (esterases)
  - immunocytochemistry (to distinguish between the subtypes of leukaemia)
  - cytogenetics
- Bone marrow biopsy if the above-mentioned has failed
- Blood for molecular genetic and cytogenetic studies is a valuable diagnostic and prognostic tool in the diagnosis of ALL.
- Flowcytometry: Immunophenotyping of the blasts with lineage-associated monoclonal antibodies to differentiate between T- or B-cell ALL. The antibodies are used to further subdivide into subtypes according to stages of maturation.
- Blood culture
- Urine MCS
- Throat swabs
- X-rays of long bones
- Central lines are preferably placed before treatment commences, e.g. a Broviac catheter

■ SPECIAL PROTECTION DURING HOSPITALISATION

- Measles immunisation must be completed because it would be too late to immunise at this stage – measles is a deadly disease in these patients.
- Varicella vaccine can be given soon after diagnosis. Alternatively, the child should receive varicella immunoglobulins after exposure.
- Nurse the child in a paediatric oncology unit, which should be removed from general paediatric wards with infections.
- Limit visitors.

■ MANAGEMENT

The treatment of ALL is intensive, with chemotherapy according to the subtype of leukaemia. This is dependent on the prognostic group into which the child falls, namely:

- standard
- intermediate
- high-risk group

This is determined as soon as all the special investigations have been completed. The higher the white cell count, the more urgent it is for the intensive treatment to begin – in a good unit usually within 24 hours.

■ PROGNOSIS OF ACUTE LYMPHOBLASTIC LEUKAEMIA

ALL has a good prognosis with a cure rate of more than 75% overall. The cure rate in ALL is determined by the following:

- Age: Children between 3 and 10 years of age have a better prognosis.
- Gender: Girls have a better prognosis.
- Initial white blood cell count: < 20 000 has a better prognosis
- Ploidy (number of chromosomes per cell): hyperdiploidy is a good prognostic feature.

The prognosis is also dependent on the quality of treatment, which should be administered only under the authority of a specialised paediatric oncology unit.

Acute Myeloid Leukaemia (AML) and Acute Non-Lymphocytic Leukaemia (ANLL)

■ CLINICAL PROFILE

Children present with the following symptoms and clinical signs:

- Abnormal masses: AML is not commonly associated with either enlarged lymph nodes or hepatosplenomegaly, but can present with chloroma (granulocytic sarcoma), a soft tissue infiltration by abnormal blasts, especially periorbital or in the gums.

- Abnormal bleeding:
  - epistaxis
  - petechiae
  - ecchymosis
- Pallor
- Unexplained fever
- Weight loss
- Disseminated intravascular coagulopathy: especially in subtype M3

■ DIAGNOSIS

- Full blood count and differential count: An increased white blood cell count with predominantly immature white blood cells (blasts), but can also be normal or low in the early phase of the disease.
- Bone marrow aspirate: An increase in immature white blood cells (blasts) of more than 25% in a bone marrow aspirate is diagnostic of AML. The French-American-British (FAB) morphological classification system is used to diagnose the subtypes. There are eight morphological subtypes in AML (see below).
- Flowcytometry: Immunophenotyping of the blasts with lineage-associated monoclonal antibodies differentiates between the subtypes of AML according to the predominant blast in the myeloid lineage.
- Molecular and cytogenetic studies are valuable for diagnosis and prognosis.

■ DIFFERENT GROUPS

- M0 – Undifferentiated myeloblastic leukaemia
- M1 – Acute myeloblastic leukaemia without maturation
- M2 – Acute myeloblastic leukaemia with maturation
- M3 – Acute promyelocytic leukaemia
- M4 – Acute myelomonocytic leukaemia
- M5 – Acute monocytic leukaemia (AMOL)
- M6 – Erythroleukaemia
- M7 – Acute megakaryocytic leukaemia

M1 and M2 ANLL

These types usually occur in older children. Up to 40% of children will be cured with intensive chemotherapy with or without bone marrow transplantation. Intensive supportive care is necessary at all times during these phases of treatment.

M3

This type is the most curable of the AML subtypes. The disease typically presents with a widespread intravascular consumption coagulopathy, namely:

- prolonged prothrombin and thrombin time
- hypofibrinogenaemia
- deficiencies of factor V and VIII
- raised concentrations of fibrinogen-breakdown products. Bleeding does not react simply to platelet transfusions, but requires the complete treatment of a consumption coagulopathy.

M4 and M5

These types are prevalent in younger children and present with hypertrophic bleeding gums. In comparison with M1 to M3, the patient can also have, amongst others, a lymphadenopathy and a hepatosplenomegaly.

Occasionally, subcutaneous nodules are noted. The response to combination therapy is an aplastic phase of the bone marrow during which the child must be well supported. The median survival is approximately 1 year.

M6 and M7

These do not occur or rarely occur in children.

■ PROGNOSIS

The prognosis depends on the type of acute non-lymphatic leukaemia which the patient has, e.g. M1 does better than M6, while M3 is the most curable.

Chronic myelogenic leukaemia (CML)

Chronic myelogenic leukaemia in children consists of two definitive separate entities, namely:

- typical adult-type CML in older children
- juvenile CML in toddlers and young children

■ ADULT-TYPE CML

Clinical manifestations

The clinical profile takes months to develop. The early symptoms are caused by:

- anaemia (fatigue, irritability)
- a packed bone marrow (bone pain and tenderness)
- splenomegaly, upper abdominal discomfort and fullness, as well as hypersplenism with sequestration of red cells and platelets

Laboratory investigations

- Moderate normochromic, normocytic anaemia
- Leukocytosis, usually more than 100 000/ml^3
- Normal to raised platelet count
- Left shift on the differential count with cells in all stages of maturation

The decreased leukocyte alkaline phosphatase is an important test for distinguishing the condition from infectious conditions, which also cause a left shift on a differential full blood count.

The characteristic karyotype abnormality, namely the Philadelphia chromosome, is present in the haemopoietic cells. It is an acquired abnormality. This chromosome is present at diagnosis in 90–100% of the cells and persists even if the condition is in remission. Only in individual cases, where intensive chemotherapy has caused severe marrow hypoplasia, does the chromosome occasionally disappear for varying time periods.

Management

Various cytostatics are used, as well as Interferon. Radiotherapy is of value in patients who have a poor follow-up risk.

The terminal phase of this disease is a blastic transformation of the lymphoid or of the myeloid cell series, which may respond to treatment.

The suggested treatment is a bone marrow transplant (should a suitable donor be available) in the chronic phase before blastic transformation takes place.

■ JUVENILE-TYPE CML

Clinical manifestations

Juvenile CML occurs in young children and toddlers and is characterised by leukocytosis, granulocyte hyperplasia and a decreased leukocyte alkaline phosphatase. This condition differs remarkably from classic CML in the following points:

- absence of Philadelphia chromosome
- more prominent lymphadenopathy and skin lesions
- raised incidence of thrombocytopenia
- monocytosis in blood and bone marrow

Chemotherapy is of limited value in these patients and juvenile CML has a more acute and fatal natural course than the classic CML, with a median survival of 9 months.

4.13 Lymphoma

Non-Hodgkin's lymphoma (NHL)

Ten per cent of all childhood cancers are lymphomas, of which two thirds are NHL. This is a diverse group of malignancies of lymphoreticular cell origin, with a multi-focal origin. Boys are more often affected. Children incur almost exclusively high-grade NHL, which has a poorer prognosis if it is not rapidly treated with intensive chemotherapy since it is already an extensive systemic disease when diagnosed.

TABLE 4.2 St. Jude Children's Research Hospital Staging System for Paediatric Non-Hodgkin's Lymphoma

Stage	Description
I	A single tumour (extranodal) or single anatomic area (nodal), with the exclusion of mediastinum or abdomen
II	A single tumour (extranodal) with regional node involvement Two or more nodal areas on the same side of the diaphragm Two single (extranodal) tumours with or without regional node involvement on the same side of the diaphragm A primary gastro-intestinal tract tumour, usually in the ileocaecal area, with or without involvement of associated mesenteric nodes only
III	Two single tumours (extranodal) on opposite sides of the diaphragm Two or more nodal areas above and below the diaphragm All the primary intra-thoracic tumors (mediastinal, pleural, thymic) All extensive primary intra-abdominal disease All paraspinal or epidural tumours, regardless of other tumour sites
IV	Any of the above with initial central nervous system or bone marrow involvement

Stage II abdominal disease is typically limited to a segment of the gut (usually distal ileum) plus or minus the associated mesenteric nodes only, and the primary tumour can be completely removed grossly by segmental excision. Stage III abdominal disease typically exhibits spread to the para-aortic and retroperitoneal areas by implants and plaques in the mesentery or peritoneum, or by direct infiltration of structures adjacent to the primary tumour. Ascites may be present, and complete resection of all gross tumour is not possible.

If bone marrow involvement is present at diagnosis, the percent blasts or abnormal cells must be 25% or less to be classified as a Stage IV non-Hodgkin's lymphoma. If there are more than 25% blasts, the patient is classified as having acute leukaemia (either precursor B or T acute lymphoblastic leukaemia or L3 acute lymphoblastic leukaemia).

■ CLINICAL PROFILE, TREATMENT AND PROGNOSIS

Patient presents with:

- lymphadenopathy which affects one or more groups of glands
- hepatosplenomegaly:
  - in 30–40% of patients the primary disease is in the abdomen
- fever

There is further systemic spread to the:

- central nervous system
- bone marrow, where it is diffusely infiltrated by 'leukaemic transformation'

CAVEAT

Superior vena cava syndrome is an oncological emergency. This is caused by glands in the lung hilum and mediastinal regions being affected. As a result of the cells duplicating every 48 hours, it can cause acute pressure symptoms, with upper airway obstruction. The children present with the following:

- cough
- wheezing or shortness of breath
- facial swelling
- cyanosis of the head and neck, but no distal cyanosis

These children must receive emergency treatment with high-dose steroids with or without radiotherapy.

■ MANAGEMENT

This is determined by:

- staging of the tumour
- histology and immunocytochemistry of the tumour cells (T- and B-cells)

■ CHARACTERISTICS AND OUTCOME OF THE DIFFERENT TYPES OF NHL

Lymphoblastic NHL

Rapidly enlarging neck or mediastinal lymph nodes, rarely below the diaphragm. Superior vena cava syndrome is an emergency and needs high-dose steroids with or without radiotherapy (see above caveat). Treatment is with intensive chemotherapy and 90% cure rates are obtained in Stage I and II disease. More extensive disease has an improved prognosis with more intensive chemotherapy regimens.

Burkitt NHL

There are two typical clinical presentations:

1. *Endemic (African):* This is more common with an incidence of 100 per 1 million children and is associated with Epstein-Barr virus. The typical presentation is in the jaw or other facial bones.
2. *Sporadic:* Extensive intra-abdominal disease with or without bone marrow involvement. The tumour is often in the lower right quadrant of the abdomen.

With intensive chemotherapy the cure rate is more than 80% for all stages, excluding CNS disease, and therefore the least invasive method should be used for diagnosis.

Diffuse large B-cell and anaplastic large cell lymphoma

Clinical presentation is varied. Diffuse large B-cell lymphoma can be present in the abdomen, bone, peripheral lymph nodes and the mediastinum, while anaplastic large cell lymphoma involves nodal and extranodal sites such as skin, bone and soft tissue. Treatment is intensive chemotherapy and even Stage III and IV disease can be cured in 70% of patients.

Hodgkin's disease (Hodgkin's lymphoma)

There is progressive, pain-free enlargement of the lymph nodes, most probably unifocal in origin and with a predictable pattern of spread.

■ INCIDENCE

Sixty per cent occurs in boys and it is seldom seen before 5 years of age; there is a progressive increase until 11 years of age. There is a markedly higher incidence in adolescents and young adults up to the age of 30 years.

■ PROGNOSIS

This is dependent on:

- staging (Ann Arbor)
- histological type

TABLE 4.3 Ann Arbor staging: Hodgkin's disease

Stage	Affliction
I	A single lymph gland area (I) of extra-lymphatic organ or an area (IE) through direct spread.
II	Two or more affected lymph gland areas on the same side of the diaphragm (II) or a localised extra-lymphatic area affected on the same side of the diaphragm (IIE).
III	Lymph gland groups affected on both sides of the diaphragm. A localised extra-lymphatic area may be affected (IIIE) or the spleen may be affected (IIIS) or both (IIISE).
IV	One or more extra-lymphatic organs or tissues, diffuse or disseminated, with or without lymph nodes affected.
A	Absence of systemic symptoms (fever, night sweats, pruritis, weight loss); presence of systemic symptoms.

Management is planned for each individual according to age, stage of the disease and the histopathology of the lesion, and consists of cytostatics with or without radiotherapy.

Prognosis is dependent on:

- stage (I is better than IV)
- systemic symptoms (poorer if present)

- histopathology (lymphocyte predominant is best)

■ HISTOLOGICAL VARIATIONS

- lymphocyte-predominant HD
- nodular sclerosing HD
- mixed-cell type HD
- lymphocyte-depleted HD

Gastro-intestinal tract

5

JHR BECKER & ML VAN NIEKERK

5.1 Congenital digestive tract obstructions – approach and management

Definitions

Atresia: This is an absent opening in a portion of a hollow viscus. Proximally and distally of the atresia is a patent lumen, for example atresia of the ileum, atresia of the aorta or tricuspid valve atresia.

Agenesis: The organ involved is absent, for example sacral agenesis or anorectal agenesis. (The primordium has never developed in the embryo.)

Background principles

The digestive tract (gastro-intestinal or GI tract) is a tube-shaped hollow viscus that stretches from the mouth opening to the anus.

The foetus begins at an early stage *in utero* to use the gastro-intestinal tract by swallowing amniotic fluid. The effective use of the gastro-intestinal tract leads to a decrease in the amniotic fluid and positive growth in the baby.

The opposite is also true, in other words if the baby has an obstruction and the amniotic fluid cannot be satisfactorily absorbed, this leads to the development of maternal hydramnios and a small-for-dates baby. The obstruction consequently produces a "malnourished", dysmature, small baby. To what extent the foetus is affected by the obstruction depends on the surface area of gastro-intestinal tract that is available for the absorption and digestion of amniotic fluid. Proximal obstructions will result in more malnourished (smaller) babies in comparison with more distal obstructions – in which case the baby may be completely normal.

The most common cause of non-canalisation of the gastro-intestinal tract is atresia. According to Louw and Barnard (1955), the main cause of atresia is a vascular incident (unknown aetiology) that the foetus experiences during pregnancy. Due to this incident there is anoxia, ischaemia and gangrene of a segment of the bowel, with consequent luminal closure and the image of atresia.

5.2 Cardinal signs of digestive tract obstruction

■ HYDRAMNIOS

The foetal gastro-intestinal cause of hydramnios is as explained above, but one must never lose sight of the fact that there may also be maternal causes for hydramnios. Not all cases of hydramnios necessarily have a gastro-intestinal tract obstruction.

■ VOMITING

Prograde movement of the content is not possible and it is by means of retrograde peristalsis that the content is transported proximally to be expelled with the vomit reflex.

- First, the type of vomiting must be observed. Is it
  - ordinary vomiting associated with other disease states, e.g. fever?
  - overflow vomiting from too much feeding?
  - primary vomiting – with the child having no transit of gastro-intestinal content?
  - projectile vomiting caused by hypertrophic pyloric stenosis?
  - related to meals if the baby is consuming solids?
- Secondly, the content of the vomitus must be noted, for example bile, digested food or bowel content. (The presence or absence of bile determines whether the obstruction is proximal or distal to the ampulla of Vater.)

NB: Vomiting does not always mean obstruction: infective, neurological, metabolic, physiological and feeding causes must also be sought (Shawis *et al*. 1984), for example

- too much food intake
- food intolerances
- food allergies and other causes

■ NO MECONIUM PER RECTUM

The foetus urinates actively *in utero*, but no meconium is normally passed *in utero*. The first stool is passed within the initial 24 hours post-partum. If no meconium is passed, this indicates a mechanical obstruction (atresia, agenesis, etc.), a functional obstruction or Hirschprung's disease.

Functional obstruction (Sieber and Girdany 1963) implies that the effectiveness of the bowel wall has been lost due to the effect on the bowel of a systemic illness, for example:

- electrolyte disturbances
- prematurity
- jaundice
- diabetes mellitus
- medication

With a functional obstruction and Hirschprung's disease there is an open lumen, but no passage, and the pathology is located in the bowel wall. Therefore, to call it an 'obstruction' is a terminological paradox.

The fact that a baby passes meconium does not rule out a total atresia. If the vascular incident occurred after a lumen had already been created, then it is possible for the content to be passed despite the presence of a complete obstruction. This happens very rarely but it is still a possibility. *Nil per rectum* remains an important sign of atresia, or any obstruction, regardless of the reason.

■ ABDOMINAL DISTENSION

The gastro-intestinal tract proximal to an obstruction actively dilates and distally to the obstruction it collapses, or in the neonate it remains hypoplastic and small. The more distally the obstruction is located in the gastro-intestinal tract, the greater the possibility of abdominal distension. If it is located more proximally, the opposite will be true and, for example, only the epigastrium will be distended.

The abdominal distension sounds resonant when percussed, in comparison with abdominal distension due to, for example, ascites, tumours, cysts, etc., which would give a dull tone.

A number of conditions will now be discussed in detail.

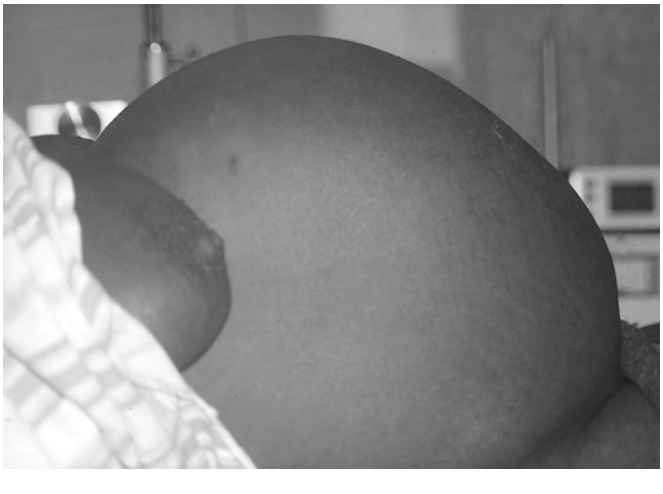


Fig. 5.1 Maternal hydramnios.

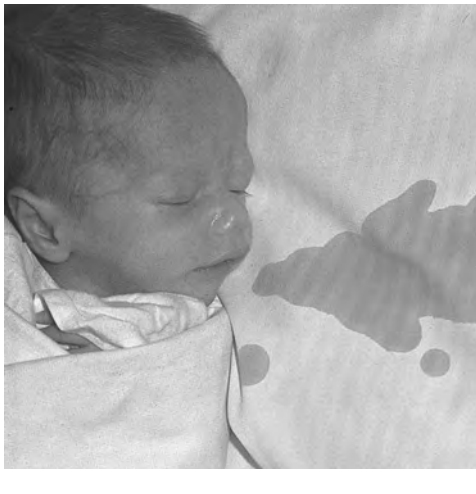


Fig. 5.2 Vomiting of bile.

5.3 Proximal oesophageal atresia with distal tracheo-oesophageal fistula (TOF)

The commonest form (90%) is as indicated in the heading, namely a proximal oesophageal atresia with a distal tracheo-oesophageal fistula. The other types account for, in total, less than 10% of cases, but the principles of management remain the same as for the common type.

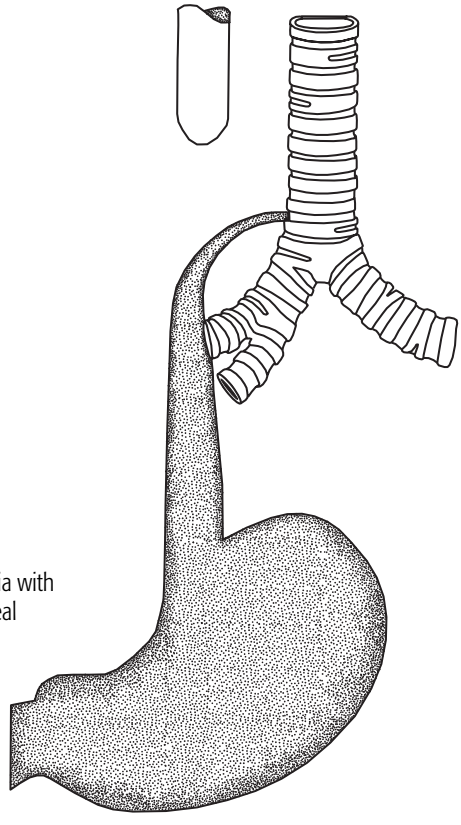


Fig. 5.3 Oesophageal atresia with tracheo-oesophageal fistula.

The other types of TOF (10%) are:

- Distal oesophageal atresia with a proximal tracheo-oesophageal fistula
- Proximal and distal oesophageal atresia with no fistula
- No atresia with a tracheo-oesophageal fistula (so-called 'h-fistula')
- Oesophageal atresia with proximal and distal tracheo-oesophageal fistulae

Problems that can present are:

- Aspiration of saliva from the proximal blind sac
- Aspiration of stomach juices via the tracheo-oesophageal communication
- Associated defects that make up the VACTERL syndrome.

The final result of the above-mentioned problems is that these children develop lung problems which are largely preventable if correct management is applied from the beginning.

Diagnosis

These conditions must be suspected when a baby cannot swallow his or her spittle and froth appears at the mouth.

The mother may present with polyhydramnios due to the inability of the foetus to swallow amniotic fluid.

Classic Triple C

C = Coughing
C = Choking
C = Cyanosis

These symptoms are the result of the aspiration of saliva and possibly water or milk that the baby cannot swallow.

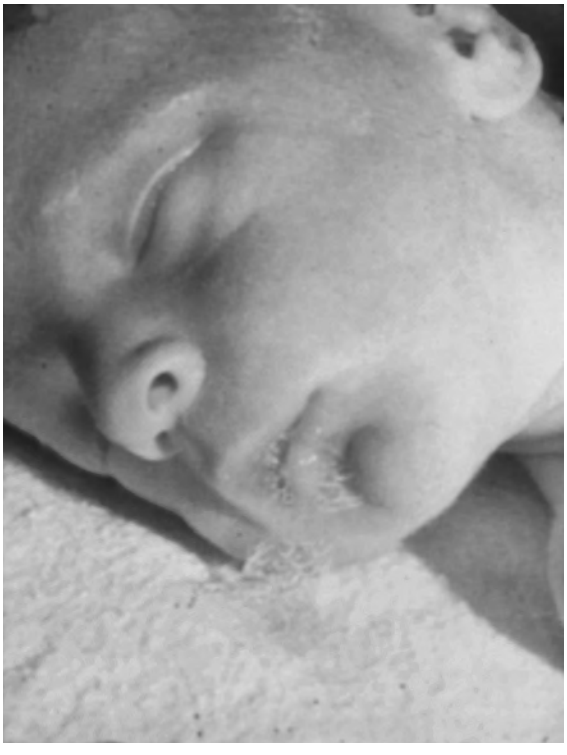


Fig. 5.4
Patient with oesophageal atresia 'spitting bubbles'.

Confirming the diagnosis

Pass a firm nasogastric tube (Fr 12–16) via the mouth into the proximal oesophagus. If an atresia is present, the tube will come to a halt. If too thin a tube is used, it will curl up and give a false impression of length. A thin tube may even end up in the stomach via the trachea and the fistula – confirmed radiologically and also clinically by the aspiration of acidic stomach juice. The consequences of an incorrect diagnosis can be catastrophic: inappropriate feeding by mouth, leading to aspiration, pneumonia and death. With the thick tube *in situ*, a single hanging (erect) abdomino-thoracic X-ray plate

is taken (so-called 'babygram') which will provide all the necessary information, *inter alia:*

- The stomach tube has halted very proximally in the oesophagus.
- Gas in the gastro-intestinal tract means that there is in fact a fistula present.

Besides the above-mentioned possible findings, the procedure will also determine whether the lungs, heart, skeleton and the rest of the GI tract are normal. It must always be remembered that one defect is usually associated with other defects, the so-called VACTERL syndrome (Muraji and Mahour 1984). This association is seen in 5-10% of patients.

V – vertebral anomalies – hemivertebrae
– 13 thoracic vertebrae
A – anal anomalies
– other gastro-intestinal atresias
C – cardiac
T – trachea
E – oesophagus
R – renal and radius
L – limbs (the radius may be absent)

Emergency management follows after confirmation of the diagnosis.

Emergency management

1. Use a naso-oesophageal Replogle tube (Argyle) No. 8 or 9 – all centres where babies are delivered should have a Replogle tube (Argyle Division of Sherwood Medical Supplies) close at hand. (It should be ordered in advance.) Constant suctioning on this tube, even during transport of the baby, prevents aspiration of saliva.
2. The baby is nursed in a 45° prone, head-up position. (This position is necessary to prevent reflux of stomach juice to the lungs – the so-called 'anti-reflux position'.)
3. Intravenous infusion of 5% dextrose or 0,25% NaCl is administered. This must be done carefully, taking the correct precautions, to prevent over-hydration of the baby.
4. The baby must be transferred to a paediatric surgical unit.

Surgical management

Primary repair can usually be accomplished at birth, even in very small infants.

Procedure:

- Thoracotomy through the third or fourth intercostal space on the right side of the baby, except for patients with an H-type fistula, in which case the thoracotomy is done through a neck incision

- Transsection of the fistula
- Closing of the opening in the trachea
- Anastomosis of the atretic segment to the distal oesophagus

Prognosis

The overall survival rate of patients born with a TOF is 95%. The prognosis is good if the opening at the anastomosis is wide and supple. A wide, supple anastomosis is not always immediately present and it tends rather to become a hard, fibrotic stenosis requiring repeated dilatations under general anaesthesia. The complication of a stenosis is overflow and the aspiration of saliva and food in the lungs.

In addition to a stenotic anastomosis, these children have a tendency to suffer from gastro-oesophageal reflux, which could lead to lung complications. As a consequence, a stenosis in the presence of reflux will never heal.

Tracheomalacia is sometimes an associated developmental fault of the tracheal cartilage rings that causes a soft, weak trachea. The trachea tends to collapse, with resultant airway obstruction. These children have a typical cough that may sound like a neglected croup cough. Apart from the airway obstruction, children with tracheomalacia also experience recurrent lung infections. It can therefore be very difficult to distinguish whether the airway infection arose due to gastro-oesophageal reflux, tracheomalacia or stenosis with aspiration.

Reasons for lung complications after TOF repair:

- Stenosis of the anastomosis with overflow of food into the trachea
- Gastro-oesophageal reflux
- Tracheomalacia
- Persistent fistula

5.4 Upper gastro-intestinal obstruction

Obstructions proximal to the ligament of Treitz

■ ABOVE THE PAPILLA OF VATER

- Stomach outlet obstruction
- Proximal duodenal atresia (first part of the duodenum).

Clinically

- These babies usually vomit a clear vomitus that contains no bile.
- The overwhelming majority are usually small-for-dates babies.

Confirmation of the diagnosis

A hanging (erect) babygram (abdomino-thoracic) X-ray shows a single stomach air bell. The so-called 'single bubble' indicates a single fluid level in the stomach, together with a gasless abdomen.

Emergency management

- Intravenous infusion of 5% dextrose water or 0,25% NaCl
- Nasogastric suctioning (Fr 8)
- *Nil per os* (NPO)
- Transfer the baby to a paediatric surgeon

■ BELOW THE PAPILLA OF VATER

This is usually a duodenal atresia.
Clinically

- Bile is vomited.
- It is usually a small-for-dates baby.
- There is mild jaundice.
- The baby has a distended epigastrium.

Confirmation of the diagnosis

A hanging abdomino-thoracic plate shows the classical *double bubble:* one bubble in the distended duodenum and one in the distended stomach. It is a double fluid level in the upper abdomen, with no gas in the rest of the abdomen.

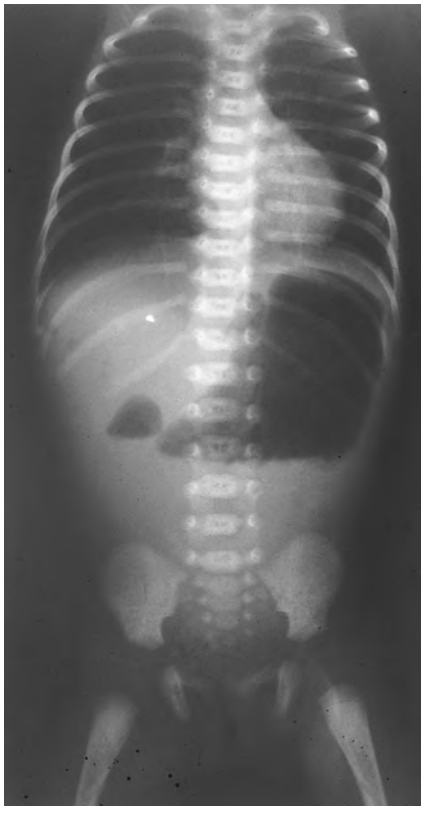


Fig. 5.5 Double bubble in a patient with duodenal atresia.

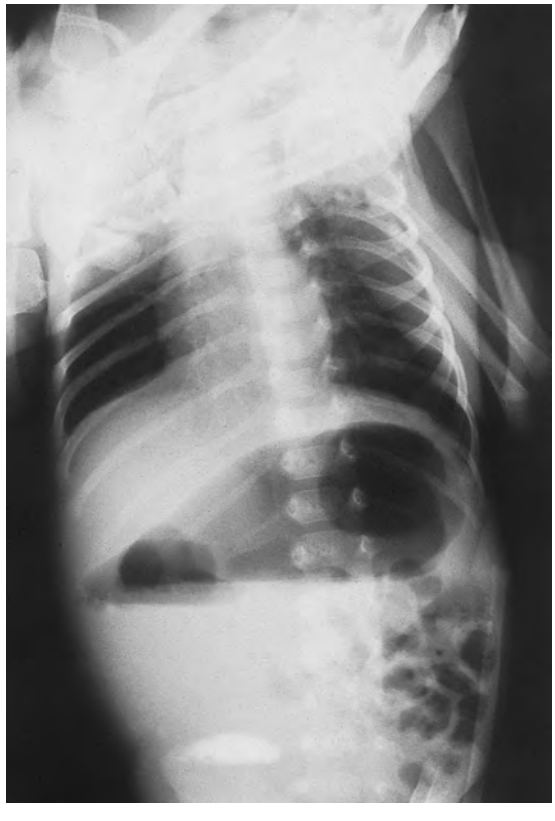


Fig. 5.6 Double bubble in a patient with a perforated duodenal membrane; note air in the distal bowel and the contrast in the dilated duodenum.

- Intravenous infusion of 5% dextrose water or 0,25% NaCl
- Nasogastric tube (Fr 8)
- *Nil per os* (NPO)
- Transfer the baby to a paediatric surgeon

Obstructions in the rest of the small bowel, up to and including the colon

Clinically

- Vomiting – bile containing vomitus
- No meconium per rectum
- Obvious abdominal distension – the lower the obstruction is located in the digestive tract, the greater the distension

Possibilities:

- Atresia (at any level in the digestive tract)
- Meconium ileus
- Volvulus
- Hirschprung's disease

There are other possibilities, but the above-mentioned are the most common.

■ CONFIRMATION OF THE DIAGNOSIS

- A hanging abdomino-thoracic plate (babygram) shows gas and multiple fluid levels. The fluid levels are located in the distended loops proximally to the obstruction. The fluid levels increase progressively according to how low down the obstruction is located in the digestive tract.
- If there is still doubt, then a gastrograffin enema can be requested, which will show a microcolon if there is indeed a proximal obstruction. The enema is diagnostic in the case of an obstruction (microcolon) and therapeutic in the case of meconium plug syndrome (neonatal constipation).

CAVEAT

The baby must have an intravenous infusion before the gastrograffin enema is requested because, due to the fact that the gastrograffin is hygroscopic, it could lead to a state of shock.

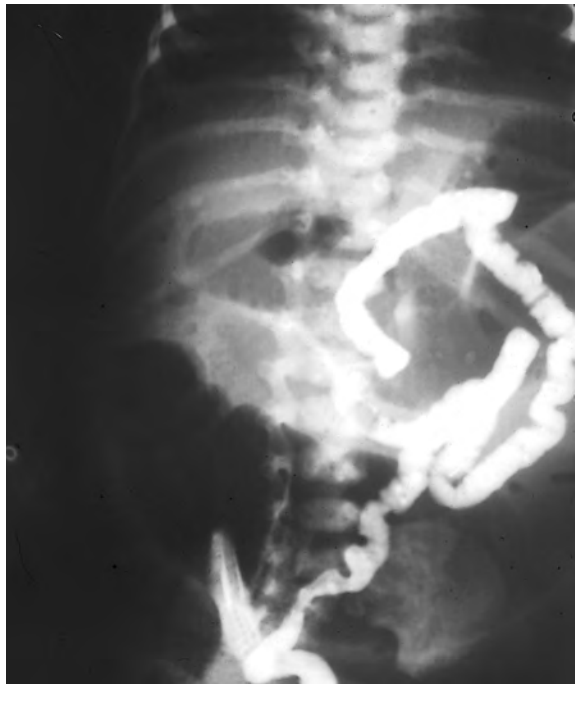


Fig. 5.7 Microcolon with malrotation; note the small calibre of the colon and the caecum in the epigastrium.
Source: Shawis *et al.* 1984

■ DIFFERENTIAL DIAGNOSIS OF A MICROCOLON

- Small bowel atresia
- Proximal colon atresia
- Meconium ileus
- Small left colon syndrome, such as that found with diabetic mothers
- Long-segment Hirschprung's disease

■ EMERGENCY MANAGEMENT (SMALL BOWEL OR COLON OBSTRUCTION)

- Nasogastric suctioning (Fr 8)
- Intravenous infusion with 5% dextrose water or 0,25% NaCl
- *Nil per os* (NPO)
- Transfer the baby to a paediatric surgeon

■ SURGICAL MANAGEMENT OF THE ABOVE OBSTRUCTIONS

Usually the baby will undergo a laparotomy with an anastomosis of the dilated proximal segment to the distal hypoplastic segment. Due to the differences in the calibre of the lumens, these babies are parenterally fed for a period until the proximal and distal lumens are equal in size. The proximal wider segment will, according to the law of Laplace, simply expand with no effective transit to the narrow segment, despite an open anastomosis, if the patient is fed too soon before adaptation of the distal segment has taken place.

5.5 Anal anomalies

Anal abnormalities (Muraji and Mahour 1984, Smith 1987) usually appear in full-term infants with normal mass, but with abdominal distension and vomiting, and inability to pass meconium per rectum. Local examination will often reveal the anal anomaly because the anus will be either absent or tiny or just covered. A fistula in the perineum, vestibulum, vagina, urethra or bladder may also be present or absent. The presence of a fistula can usually be demonstrated with great accuracy – the meconium is black and is found in the perineum, vestibulum or vagina, or it is found in the urine.

Most babies with an imperforate anus have one or more abnormalities that affect other systems. The incidence varies from 50–60%. These abnormalities include genito-urinary, gastro-intestinal, cardiovascular and vertebral abnormalities.

After the diagnosis of an anal anomaly has been made, no further investigations are necessary; the baby is immediately referred.

■ EMERGENCY MANAGEMENT

- Intravenous infusion of 5% dextrose water or 0,25% NaCl
- Nasogastric suctioning (Fr 8)

- *Nil per os* (NPO)
- Transfer the baby to a paediatric surgeon as soon as possible within the first 24 hours

■ TREATMENT

In assessing a newborn infant, two factors need to be considered: the need for a colostomy and the presence of other malformations. The need for a colostomy can be determined from the following factors:

- A good clinical examination.
- An invertogram. This is a lateral pelvic radiograph obtained with the baby prone and the hips slightly raised. A waiting period of 24 hours after birth is necessary to allow gas to reach the most distal part of the bowel. A gap between the skin and the gas of more than 1 cm indicates a high abnormality. A colostomy is necessary for all high abnormalities. Local perineal surgery without colostomy is done for low abnormalities.
- Sonar, RT and MRI scans – these can also be used to determine the position of the rectum and the pelvic anatomy.
- The gender of the patient. In female infants, a colostomy is done for all patients with a vestibular or vaginal fistula. For a perineal fistula, a local perineal repair is done. In male infants with a perineal fistula, a bucket handle midline fistula, an anal stenosis or an anal membrane, local surgery is done. If a male infant has a flat bottom, meconium in the urine or air in the bladder, this means a high abnormality which needs initial colostomy.

The anal sphincter mechanism is very specialised and the best initial management is absolutely essential. A general surgeon should only do a colostomy for a high abnormality in an emergency situation, and should not attempt repair.

■ COURSE AND PROGNOSIS

Usually only an anoplasty is done for a low anomaly. These patients require anal dilatations with Hegar dilators until the anus is supple and allows at least a Hegar 16 at the age of 6 months. These children have good anal sphincter function and ought to be socially acceptable.

A primary colostomy is done on patients with high anomalies. As soon as the baby's weight is 5 kg, a pull-through procedure is done. During this operation an anus and anal sphincters are created. Anal dilatations are also necessary post-operatively. The colostomy is closed as soon as the anus accommodates a Hegar 12 with ease. The anus must eventually also allow a Hegar 16.

Children with a high anomaly do not have an internal anal sphincter and can therefore never be completely continent. They must be encouraged all their lives and rapidly placed on a bowel routine:

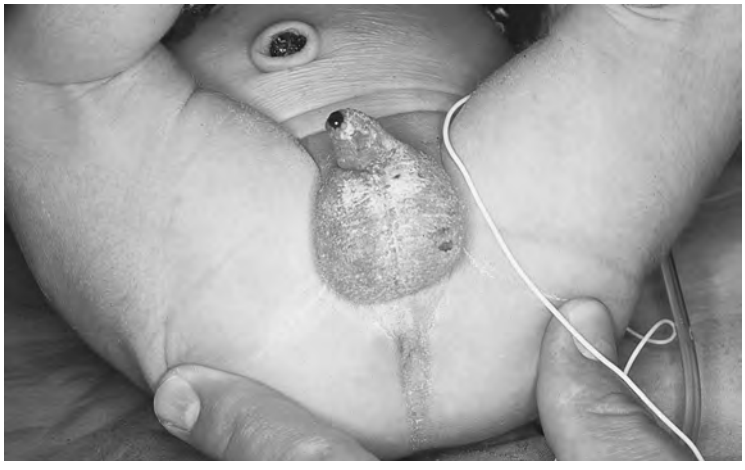


Fig. 5.8 Anal anomaly with meconium at the external urethral meatus due to a probable rectovesical fistula.

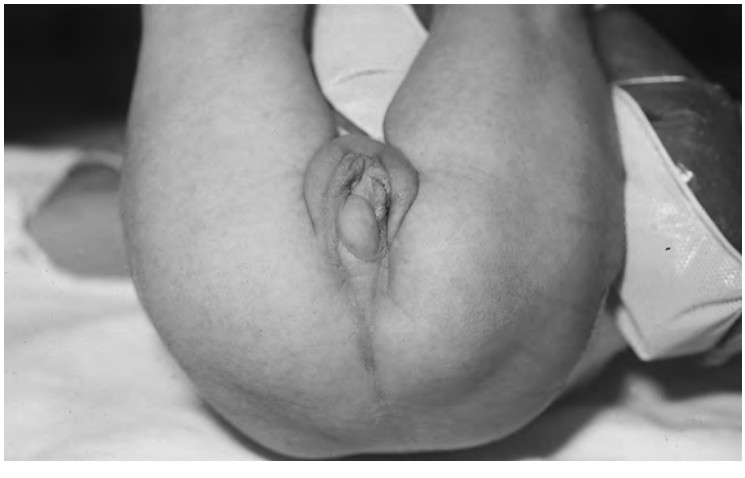


Fig. 5.9 Genital abnormalities associated with an anal anomaly.

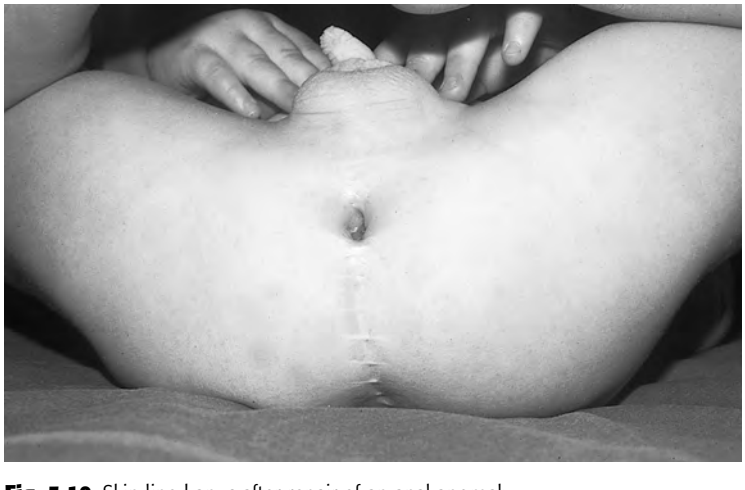


Fig. 5.10 Skin-lined anus after repair of an anal anomaly.

- Bran (nature's laxative) with breakfast
- Bowel emptying shortly after breakfast (the reflex and the routine are learned by means of enemas)
- A small disposable nappy in the underpants (a small square cut out of a linen saver is usually sufficient)

Other digestive tract obstructions that are not mentioned or discussed here can be diagnosed and managed using the above principles. Transfer to a paediatric surgeon must occur as soon as possible within the first 24 hours of life.

5.6 Hirschprung's disease

The following facts should be borne in mind:

- Incidence 1:5 000 births
- Short segment 4:1 male to female ratio
- Long segment 1:1 male to female ratio

Hirschprung's disease is a congenital disease that is caused by insufficient cephalo caudal migration of ganglion cells. The affected segment is therefore always located distally in the bowel, with a varying length being affected. The bowel wall contains no ganglion cells and the plexuses of Meissner and Auerbach are absent. In addition, there is an overgrowth of nerve bundles which makes this disease a neuromatosis of the bowel.

Diagnostically

- Ganglion cells will never be histologically shown at the anorectum.
- Hypertrophic nerve bundles are seen histologically.
- Histochemically, an excess of acetylcholine esterase can be shown in the mucosa.
- Anal manometry indicates poor relaxation of the internal sphincter – absence of the inhibitory reflex.

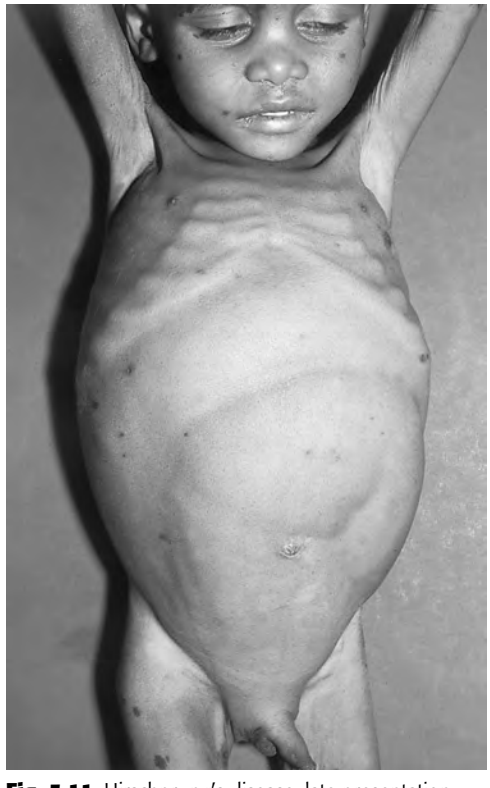


Fig. 5.11 Hirschprung's disease, late presentation; note the cachexia and dilated visible colon.

Clinically

- Unable to pass meconium stool in the first 24 hours of life
- Resonant abdominal distension
- Vomiting
- Rectal examination reveals a spastic anus – the rectum is empty and when the examination ends, the colon discharges explosively

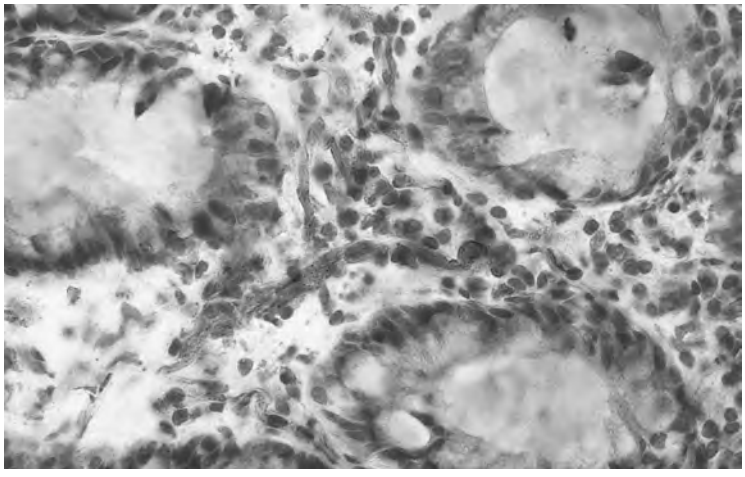


Fig. 5.12 Special staining microscopy of a rectal mucosal biopsy, demonstrating the hypertrophied nerve bundles (brown bundles) confirming Hirschsprung's disease.

- If the condition presents late, stasis occurs in the proximal dilated bowel with resultant stasis enteritis, which gives rise to:
  * toxic dilatation of the bowel
  * systemic toxic manifestations, such as:
    – fever, tachycardia, tachypnoea
    – hypotension
    – death

■ MANAGEMENT OF STASIS ENTERITIS

If circulatory collapse is present, then it must be rapidly attended to in the following way:

- Administer intravenous infusions
  – fresh frozen plasma – begin with 20 ml/kg
  – crystalloid, for example Ringer's lactate, until circulation has returned to normal
  – maintenance fluid.
- Monitor vital functions.
- Empty the colon of toxic debris using saline enemas at body temperature until the fluid returns clear.
  – Also empty it often by means of a rectal examination to aid relaxation of the internal sphincter.
  – It is important that all fluid that is inserted comes out again and that all is clear with the last washout.
- Refer the baby as soon as possible to a paediatric surgeon for biopsies and a colostomy.

■ CONFIRMATION OF THE DIAGNOSIS

Barium enema: This may be falsely negative for Hirschprung's disease in the first 6 weeks of life, except for the microcolon of a long-

segment Hirschprung's which may already be suspected on giving the first enema. The narrow segment of the affected bowel is visible distally in the colon, with an obviously dilated proximal ganglion-containing colon. It is important that the barium enema is done on an unprepared bowel. After a period of 24 hours and sometimes even 48 hours, a post-evacuation abdominal overview plate is taken which will show residual barium in the colon.

■ HISTOLOGY

This is the final arbitrator in determining the diagnosis of Hirschprung's disease. The procedure is:

- A full-thickness rectal muscle biopsy is done which confirms the absence of ganglia, as well as the presence of hypertrophied nerve bundles.
- If the facilities are available, then an increase in acetylcholine esterase can be shown histochemically, simply on a rectal mucosal biopsy, which is then diagnostic.

■ EMERGENCY MANAGEMENT

Emergency management by the general practitioner involves thorough daily saline enemas which decrease the risk of stasis enteritis in the proximal dilated viscus. Evacuation must sometimes be initiated by placing a finger in the rectum due to the fact that these children do not have a normal evacuation mechanism. The child must be transferred as soon as possible to a paediatric surgical unit where definitive therapy must follow.

■ MANAGEMENT

- The standard treatment today is a primary pull-through operation in the neonatal period. This can be done with or without laparoscopic assistance.
- In many institutions an initial colostomy is still done, followed by removal of the aganglionic segment a few weeks later. At operation a proximal ganglion-containing viscus is attached to the anus.
- One of three conventional operations is done, namely:
  - Duhamel (Livaditis 1981)
  - Soave
  - Swenson (Sherman *et al.* 1989).
- An initial colostomy is also indicated for enterocholitis, or when there is an inability to obtain adequate decompression of the bowel with irrigations.

The results are very good and 80% of these children can lead a normal life post-operatively. The other 20% require lifelong support of some nature.

One of the problems of concern with the above-mentioned 20% of patients is the persistence of the stasis enteritis, which, as discussed, must be treated. This group of patients must undergo a repeat rectal biopsy to ensure that the operation was complete and was conducted in a ganglion-containing segment. If ganglia are present, then no further surgery is required and the patient can be treated with a pro-kinetic.

5.7 Hypertrophic pyloric stenosis

This condition is a fibro-muscular thickening of the pyloric muscle that manifests clinically from the first week post-partum. It is speculated that it is already present *in utero* and can be diagnosed with the use of ultrasound examinations which show a hugely dilated stomach.

■ CLINICAL PROFILE

- There is classic projectile vomiting.
- The baby is hungry after vomiting.
- No signs of raised intracranial pressure, sepsis or reason for vomiting are found.
- There is some level of dehydration.
- The stools are dry (so-called 'hungry stools').
- There is obvious peristalsis visible from left to right in the left hypochondrium and epigastrium.
- A walnut-sized tumour can be palpated in the right hypochondrium. This examination must be done while the baby is lying peacefully on the mother's lap, drinking sugar water, and the examiner is kneeling in front of the mother with two fingers palpating the right hypochondrium.

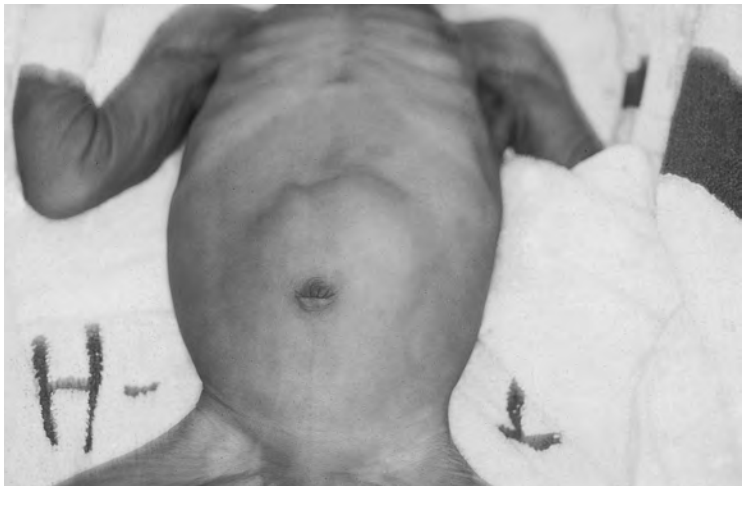


Fig. 5.13 Hypertrophic pyloric stenosis with visible peristalsis in the epigastrium.

■ DIAGNOSIS

The diagnosis can usually, in 80% of cases, be made from a medical history and clinical examination. If there is doubt (average one in five cases), a diluted barium contrast study is advised.

The classic signs are those of a distended stomach, relatively little gas in the rest of the abdomen, an elongated pyloric channel (longer than 1,5 cm), the Chinese umbrella in the duodenal cap and on the side of the stomach, the shoulder, beak and teat signs. Biochemically, these children usually have a hypokalemic, hyponatremic alkalosis and dehydration that must first be corrected before an operation can be performed. It is therefore not an urgent operation. If the facilities and expertise are available, an abdominal sonar scan will further confirm the diagnosis.

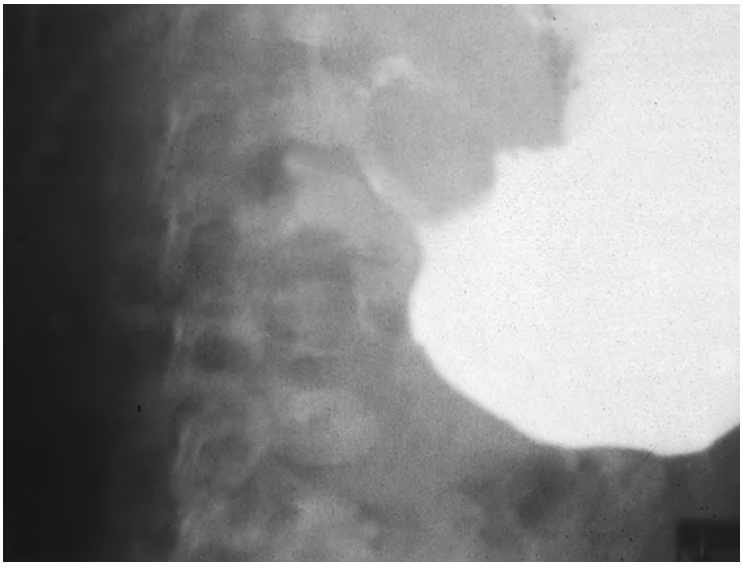


Fig. 5.14 Barium meal confirming the presence of hypertrophic pyloric stenosis; note the elongated pyloric canal as well as the shoulder formation on the gastric surface.

■ TREATMENT

The operation is usually a Ramsted pyloromyotomy. By implication this means that only the pyloric muscle is split and an intact mucosa is left. No sutures are placed in the pylorus. The operation is performed with minimal trauma through a small transverse incision in the right upper abdominal cavity. In many centres around the world, the laparoscopic pyolorotomy is the method of choice. This procedure is done through three 2–3 mm abdominal incisions. This method has a cosmetic advantage over the open procedure. After a pyloromyotomy, babies can commence steadily with feeds as soon as 4–6 hours post-operatively. Feeds are introduced 3 hours after completion of the procedure and gradually increased.

5.8 Meconium ileus (mucoviscidosis) ('Sticky slime disease')

This is a common form of small bowel obstruction which appears in 15% of patients with mucoviscidosis or rather *cystic fibrosis*, which is a congenital disease of the exocrine glands with an abnormal loss of electrolytes in sweat, tears and saliva, as well as a particularly thick and sticky mucus in the digestive tract and lungs. This condition is an autosomal recessive disorder. It appears in people of European descent, but not in Africans.

■ SYMPTOMS

- The bronchi are blocked with thick slime plugs which leads to recurrent infections.
- The digestive tract becomes blocked with meconium in the neonate. This leads to abdominal distention, vomiting and failure to pass meconium in the first 24 hours after birth.
- It usually arises in the distal ileum.
- Severe constipation is experienced by the older child and adult.
- The pancreas is progressively replaced by fibrosis and later a total loss of exocrine functions develops.

Clinically, the profile of a small bowel obstruction is apparent in the baby:

X-rays indicate:

- upper abdominal fluid levels and a gasless lower abdomen
- a grainy image of the thickened meconium in the RIF
- sometimes eggshell calcifications in the abdomen
- sometimes dotted calcifications in the abdomen, indicating meconium peritonitis secondary to intra-uterine intestinal perforation

A gastrograffin enema indicates the typical microcolon, with occasional small hard plugs of meconium visible.

■ EMERGENCY MANAGEMENT

- Infusion of 5% dextrose water or 0,25% NaCl
- Stomach suctioning (Fr 8)
- Refer the patient immediately to a paediatric surgeon

■ DEFINITIVE MANAGEMENT

- Therapeutic gastrograffin enema after rehydration and in the absence of clinical or radiological indications of complications. This is sometimes successful in relieving the obstruction.
- Laparotomy
- Resection and primary anastomosis if necessary. Indications are: volvulus, bowel necrosis and perforation.

- A T-tube is left in the bowel, through which irrigation is done post-operatively with gastrograffin to relieve the remaining distal obstruction.
- Hyperalimentation until the anastomosis heals and the baby can take special feeds.

■ PROGNOSIS

The long-term prognosis is not good as a result of the other organs being affected; in particular, there are recurrent lung infections. Some patients do reach adulthood. These patients' parents must all receive genetic counselling.

5.9 Meconium plug syndrome (neonatal constipation)

The baby has the following symptoms:

- a distended abdomen
- signs of a low digestive tract or colon obstruction
- little or no meconium per rectum
- X-rays show gas up to the colon
- a gastrograffin enema shows a dilated colon with many meconium plugs that pass spontaneously after the enema

The baby usually requires no further treatment thereafter.

NB: Be observant of the following possibilities:

- Hirschprung's disease – if there is doubt, then do a biopsy.
- Mucoviscidosis – if there is doubt, then do a sweat test.

5.10 Short frenulum of the tongue

The frenulum is located in the midline anterior, below the point of the tongue between the external openings of the submandibular salivary gland ducts.

The frenulum is normally very mobile and allows free movement of the tongue. In some cases, however, it is short and fibrotic, and acts as a band that limits the mobility of the tongue.

This limitation of the tongue has the consequences that the child's tongue:

- cannot be stuck out
- cannot normally lick something, for example ice-cream
- sometimes cannot formulate some sounds
- causes pain where the band moves over the lower teeth with normal use of the tongue

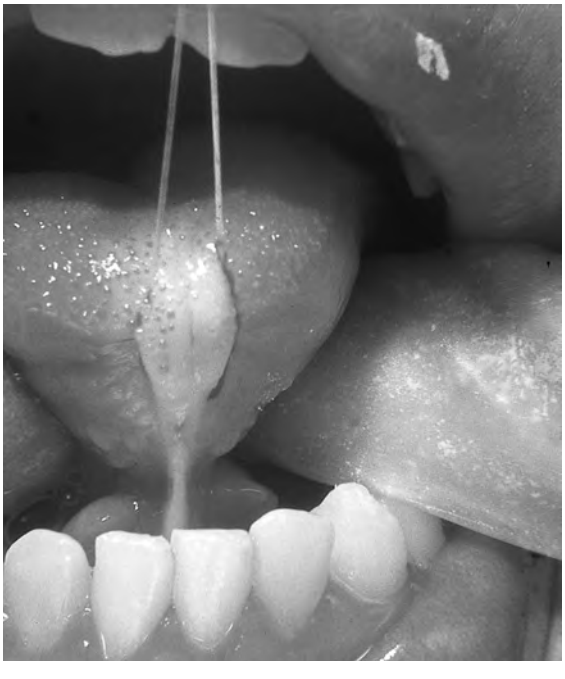
When the band is very short, it can even pull the lower teeth skew, with the roots inward and the teeth outward.

A short frenulum is therefore not life-threatening and must be left alone if the child is not functionally impaired in any way.

If, however, it is symptomatic and some of the above-mentioned points are present and cause concern, then the band can be cleaved during a small operation under general anaesthesia.

CAVEAT

The precautions with small operations must at all times be irreproachable because to experience anaesthetic complications from a small non-life-threatening operation is unacceptable.



Figs. 5.15 and **5.16** Short frenulum ('tongue tie') in a young boy.

Burn wounds

6

JHR BECKER, JAN J VAN WINGERDEN & H RODE

Definition of burn wounds: Coagulation of proteins caused by thermal, chemical or electrical injury (Du Plessis and Becker, 1983a; Du Plessis and Becker 1983b; Becker 1987).

6.1 Thermal burn wounds

Pathophysiology

- Loss of the skin barrier between the unsterile outside world and the sterile interior milieu
- Increased capillary permeability with subsequent fluid losses leading to hypovolaemia and shock
- Cardiac suppression due to myocardial suppression factor
- Red blood cell loss – anaemia (due to heat injury, bleeding, bone marrow suppression)
- Susceptibility to infection
- Catabolism with increased energy expenditure
- Respiratory distress syndrome due to:
  - carbon monoxide or cyanide poisoning
  - sepsis
  - multi organ failure

Emergency management

- Cool the burn with running cold tap water at 16 °C for 20–30 minutes or use Burnshield (*Melaleuca alternifolia* Hydrogel); do not use ice water – the vasoconstriction that results may deepen the burn wound.
- Remove smouldering or hot clothing immediately and ensure an open airway.

- Irrigate copiously with water in the case of chemical burns.
- Administer 100% oxygen if an inhalation injury is suspected.
- Place a sterile or clean dressing on the wound and dress with crêpe bandages.
- Administer an intravenous infusion; begin with Ringer's lactate. Start IV immediately if burn wound covers more than 10% of the total body surface area (TBSA).
- Transfer the patient to a burn wound centre (a patient will tolerate transfer better shortly after the burn wound has been sustained than later).

6.2 Clinical determination of the depth of a burn wound

First degree

- Red, painful, swollen
- Analogous to severe sunburn
- Heals spontaneously
- *Keep the skin supple with skin creams or oils and cover with paraffin gauze (Jelonet S+N) and crêpe bandaging.*

Second degree, superficial

- Red, painful, swollen, blistered, loosening epidermis, serum oozing from wounds
- Will heal by the 7th to 14th day
- *Cover with paraffin gauze (Jelonet (S+N) or Granuflex® (Squibb)) or a polyurethane dressing and remove the bandaging when required.*

Second degree, deep

- Painful, thin layer of parchment (dead tissue) on the surface; serum oozes from the wounds
- If it is not contaminated, then it will heal between the 14th and 21st day
- Heals with scarring
- *Cover with Granuflex® (Squibb) or topical antiseptics*
- *A skin graft must be done on any wound that does not show signs of epithelialisation by 21 days.*

Third degree

- Deep burn wounds
- All skin rests and appendages are destroyed
- Burns are less painful
- Dry surface, appears parched, brown or charred, depending on the agent that caused the burn wound:

  - boiling water: parchment-like
  - open flame: brown or carbonised
- *These wounds will not epithelialise spontaneously, and a skin graft will be needed.*

Fourth degree

- Loss of tissue from the skin down to deeper structures, e.g. muscle, tendon or bone
- *These patients require radical debridement of all burnt tissue.*

DICTUM

A burn wound from an open flame is always deep – third or fourth degree. If it is not like this, it should be considered a bonus.

6.3 Definitive management

Resuscitation

- Ensure good venous access (central or peripheral lines).
- Administer sufficient and correct fluid therapy.
- Monitor effective resuscitation.
- Administer topical treatment.

Venous access

- Follow a large-bore central or peripheral line
- Preferably not through a burnt area
- Preferably not in the lower limb

Total body surface area burnt (TBSA)

■ PERCENTAGE IN THE ADULT – RULE OF 9s

For determination of the surface area of smaller burn wounds, an open hand can be used; this is equivalent to 1% of the body surface area.

■ PERCENTAGE IN THE CHILD – ADAPTED TO AGE-RELATED DIFFERENCES

The chart of Lundt and Browder (next page) is used and not the Rule of 9s because a child's body surface area ratios differ from those of an adult.

Fluid therapy

The volume of fluid lost can be calculated on the basis of the percentage of the body surface that has been burnt.

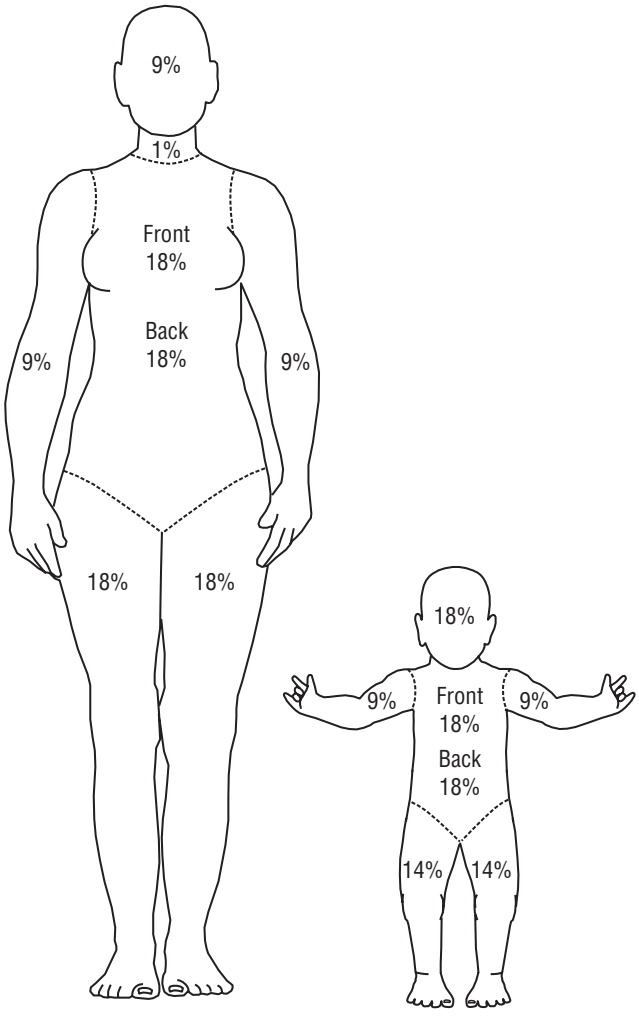


Fig. 6.1 Diagram to calculate percentage burn wound in adults and children.

■ ADULTS AND CHILDREN: PARKLAND FORMULA

- 4 ml/kg/%burn = Ringer's lactate per 24 hours required.
- Give half over the first 8 hours and the remainder over the following 16 hours.
- The total percentage of the burn wound is used in the formula, up to 50% TBSA.

The best, easiest and most reliable methods of monitoring fluid resuscitation are:

1. Urine output adult 0,5 ml/kg/hour = 30–50 ml/h
   children (<30 kg) 1,0 ml/kg/h
   (range 0,5–2 ml/kg/h)
2. Blood pressure + pulse rate
   Adapted for age and prior hypertension
   Children = age (in years) × 2 + 80 = systolic pressure

3. Central venous pressure 5–10 cm H_2O – seldom necessary
4. Blood gas analysis – acidaemia (pH<7,39) commonly indicates inadequate tissue perfusion
5. Restlessness, mental obtundation and anxiety are often indicators of hypovolaemia

Topical treatment

The general principles with the placing of the first dressing are:

- Remove all debris, e.g. dead skin, blisters and contaminants.
- Rinse with chlorhexidine in water.
- Cover with a topical cream – bandaging.

Five topical antiseptics are available: Topical therapy is used to reduce bacterial and fungal proliferation but cannot sterilise a burn wound. Intra- and sub-eschar organisms can be reduced to $<10^3$ organisms/g of tissue.

- Betadine® cream (povidone iodine – 5%). This is a good prophylactic cream with a broad spectrum. The maximum effect is achieved within 24 hours, with good tissue penetration.
- Sulfamylon® (magenide acetate). This is good for gram negative wound infections. The maximum effect is achieved within 12 hours, with good tissue penetration. However, there may be undesirable side-effects.
- Silver sulphadiazine (AgSD) (flamazine – 1%). This is effective for 24 hours but does not give good penetration. It is good for gram positive and negative organisms. This is the most commonly used antiseptic. Nystatin 1:1 can be added to AgSD to reduce the incidence of invasive candidal growth.
- Silver nitrate solution – 0,5%. Application is messy but effective and is complicated by electrolyte abnormalities.
- Mupirocin/Chlorhexidine in combination. This is effective for 24 hours and offers broad spectrum protection with no resistance, good tissue penetration and minimal side-effects. It is an excellent combination for resistant wound organisms or as a topical antiseptic for grafted areas.

Alternatively, especially in superficial wounds, tullegras, a polyurethane dressing, or another occlusive material can be used, such as Granuflex® or Coloplast®.

Wound management with dressing changes

- All patients must shower rather than bath because the running water removes the dressings and cleanses the wound without contamination from the perineum.
- An ordinary bath, and even the special whirlpool burns bath in which the patient sits, is not recommended because perineal organisms are splashed from the bath water onto the wounds.

Analgesia

Analgesia must be given with caution because it influences the sensorium and respiration and, in so doing, neutralises one of the important monitors.

There are three components to be considered, namely acute, procedural and chronic pain. Pain management is an integral part of the programme and must be initiated from the beginning. Commonly used analgesic agents are:

Oral: Tilidine HCI (Valoron) – 1 mg/kg/6 hourly (each drop = 2,5 mg)

Paracetamol – Loading dose is 20 mg/kg. Maintenance dose is 15 mg/kg

Morphine – 0,3 mg/kg/dose 8–12 hourly

Parenteral: Morphine – 0,2 mg/kg in 20 ml of 5% D + W. Infusion rate must be 1–4 ml/hour.

Ketamine – 2–3 mg/kg/dose. For use during procedures

Prophylactic antibiotics

This is not recommended. Treat systemic infections as necessary when they appear (specifically according to culture and sensitivity).

Definitive management

■ MINOR BURNS

Initial therapy for minor burns should include administering analgesics, cleaning the wound with bland soap and water or detergent, removing topically applied agents and shaving of hair where necessary. Dead tissue should be debrided and any tar removed with soft paraffin in a water base. Topical antibacterial agents and occlusive dressings (gauze and Elastoform) or an adhesive polyurethane sheet (Omiderm) should be used to dress the wound and tetanus toxoid should be administered. The dressings can be left for 7–10 days until the underlying wound has healed. With regard to follow-up therapy, the patient should be encouraged to move the affected area. Local topical therapy should be applied every 2–3 days until the wound has healed or Omiderm can be left in place until the wound has healed.

■ MAJOR BURNS

Early excision and grafting should be considered as the best treatment option for all burn wounds that do not heal by primary intention within 3 weeks. Once the burn wound has been excised, rapid burn wound closure is essential. The depth, extent and post-burn day of the burn wound will determine the surgical approach.

■ SUPERFICIAL WOUNDS

These usually epithelialise within 3 weeks. Excision is contra-indicated. Satisfactory functional and cosmetic results are achieved.

■ DEEP PARTIAL THICKNESS WOUNDS

These wounds are difficult to assess within the first few days. The cosmetic and functional results are unsatisfactory. Early tangential excision and grafting is advised or there can be a delay for 14 days, especially with scald burns in children.

■ UNEQUIVOCALLY FULL-THICKNESS WOUNDS

Smaller burns are a surgical priority, with complete excision and immediate split skin grafting. Larger wounds should be serially excised within the first 10 days. The usual procedure entails the excision of 15–20% of the burn surface area, with immediate skin grafting.

■ DEEP BURN WOUNDS

The primary-care physician should be absolutely clear about the difference between an escharotomy and an escharectomy.

An **escharotomy** can be life-saving or limb saving and is a procedure that *every* clinician should be able to perform. The tight, full-thickness dead tissue, i.e. *eschar*, is split with a surgical knife, at least once longitudinally, e.g. down the sternum or length of a limb, and then vertically. Properly performed, it will allow the constricted underlying structures, e.g. the chest or abdomen, to expand.

A *specialist* should preferably perform an **escharectomy,** i.e. complete removal of all dead tissue. Great care will be taken not to remove potentially viable tissue, e.g. the female breast bud that would prevent normal breast development later (Neale *et al.* 1982). The resulting raw areas are normally covered with split-thickness skin grafts as soon as possible after the escharectomy.

Tangential or sequential excision

This method entails the sequential excision of thin layers of burn eschar until a viable bed is encountered. Once excision to the appropriate level has been done and haemostasis has been secured, an immediate split-thickness skin graft is performed. Sheet grafts are placed on important cosmetic and functional areas. To prevent the desiccation of exposed and viable tissue, mesh grafts should not be expanded more than – 2 :1. If greater expansion is needed, temporary skin substitutes (cadaver or synthetic skin substitutes) should overlie the autograft.

Fascial excision

This method is generally reserved for very large, life-threatening or deep full-thickness burns. The excision is performed using a combination of sharp dissection, traction and haemorrhage control.

The excision is preferably limited to approximately 10–20% TBSA at any one time. Fascial excision assures a viable bed for skin grafting with moderate blood loss, especially if it is done under tourniquet control; the graft can be expected to take very well if fascial excision is done within the first few days after injury.

Delayed escharectomy

This is done after spontaneous separation of the eschar has allowed a bed of granulation tissue to form. Daily debridement by means of hydrotherapy (showering or bathing), or coarse mesh gauze dressings, will hasten separation of the eschar. The burn wound is ready for split skin grafting when there is a shiny, slightly granular, pinkish-red, uniform bed of granulation tissue, with no debris or evidence of infection. This method is most often used for old, neglected burn wounds.

■ CONTRA-INDICATIONS FOR SKIN GRAFTS

- Unsatisfactory, infected, recipient area
- The presence of beta-haemolytic *Streptococci* and *Pseudomonas aerugenosa*
- Anaemia – Hb less than 8 (full blood count twice a week)
- Severely catabolic patient – negative nitrogen balance
- Medical contra-indications for general anaesthesia

6.4 Chemical burn wounds

JAN J VAN WINGERDEN

Chemicals may harm the child in three possible ways:

- Local effect:
  - by directly denaturing protein
  - by producing heat (*exothermic reaction*), especially when the burn is exposed to other, indiscriminately chosen chemicals
- Systemic (*toxic*) effect: hepatotoxicity or nephrotoxicity

The *extent* of the injury is dependent on:

- the pH of the chemical
- the concentration of the chemical
- the volume of the chemical
- the physical form of the chemical
- the duration of contact
- the surface area exposed

Alkalis (also known as *bases*) are proton acceptors (OH-) and cause *liquefaction necrosis.*

On exposure, proteins denaturate and fat saponificates; this allows the alkaline proteinates to penetrate deeper into tissue, causing extensive local destruction. Dangerous alkalis usually have a pH above 10.

Acids are protein donors (H+) and cause *coagulation necrosis*.

On exposure, proteins denaturate and a coagulum forms, which limits penetration. Dangerous acids usually have a pH below 2.

One *exception to the rule* is hydrofluoric acid, which causes liquefaction necrosis.

Emergency management

- Remember the ATLS principles.
- Immediately remove all clothing.
- Scrape off the granules or powder of the offending chemical.
- Irrigate with copious amounts of water.
- The treatment dictum is:
  - 'Irrigate until water comes out of your (the treating physician's) eyes. Continue until water comes out of the eyes of everyone in theatre and when everybody is crying, continue some more!'
- Only apply very specific surface antidotes for very specific chemicals (e.g. calcium gluconate for hydrofluoric acids).
- Once you are sure that the local damage has been contained, continue management as for thermal burns.

6.5 Electrical burn wounds

The six factors

1. Type of current – alternating current allows the person to 'freeze' to the contact point due to muscle spasm
2. Volts (power)
3. Amperes (current)
4. Resistance of the body
5. Pathway of current through the body
6. Duration of exposure

Pathology

Tissue damage results from the generation of heat, which is a function of the resistance of the tissue, the duration of the contact and the square of the current.

Emergency management

- Remove the patient from the current in one of the following manners:
  - Switch off the current
  - Pull the patient away from the point of contact. Use a non-conductor of electricity, e.g. a stick.
- Administer cardiopulmonary resuscitation (e.g. fibrillation or asystole). Dysrhythmias are common.

- Start fluid therapy early – the urine outflow must be more than 1 ml/kg/h. The crush syndrome caused by severe muscle and tissue damage will lead to myoglobinuria and haemoglobinuria, with consequent kidney failure.
- Perform fasciotomy if circulation to distal parts is threatened or with compartment syndrome.
- All dead tissue, skin, muscle, etc. must be removed within 24–30 hours.
- A second evaluation of all wounds within 24–48 hours must be done, with re-evaluation every 48 hours until there is certainty that all dead tissue has been removed.

6.6 Burn wounds and feeding

Resuscitation of the patient and treatment of the wounds is often conscientiously seen to, whereas the feeding of the patient is neglected. The basal metabolic rate (BMR) doubles with a 30% third-degree burn wound. If the calorie intake is not supplemented, these patients will soon have no more energy and will die, i.e. "The machine will run out of fuel".

As soon as resuscitation is completed, introduce parenteral feeding and enteral feeding as soon as the gastro-intestinal tract is available.

Proteins are of particular importance and 2 g per kg of body mass is given. Raw eggs in a base of cow's milk as enteral feeding and Vamin as parenteral protein feeding source are recommended (1 egg is equivalent to 7 g of protein). Patients who are regularly taken to theatre for procedures are usually fasted for a period before and after the anaesthesia. With these patients, switch over to parenteral feeding during the fasting periods until enteral feeding can be resumed.

Remember

- No patient's wounds will heal,
- no skin graft will take,
- no sepsis will clear up,
- no anaemia will resolve itself,
- if the patient is insufficiently fed during the acute and chronic phases of burn wound healing.

6.7 Burn wounds of special areas

Eyes

- Rinse out thoroughly with 0,9% NaCl solution.
- Consult an ophthalmologist to evaluate whether the cornea has been burnt.
- Bathe the eye in a broad spectrum antibiotic ointment.

Ears

- Place the ear in a ring of orthopaedic wool (a 'doughnut') so that when the child lies on the ear, pressure sores will not develop on the thin burnt skin overlying the cartilage.
- Consult a plastic surgeon.

Face

- Consult a plastic surgeon immediately.

Hands

- Place the hands in surgical gloves filled with antiseptic cream.
- Elevate the hands above the level of the right atrium so that swelling can drain with the aid of gravity.
- Encourage the patient to move the hand so that the hand acts as its own pump to maintain suppleness and to pump out oedema fluid.
- Consult an orthopaedic surgeon immediately or a plastic surgeon with a special interest in the treatment of hands. Early debridement and skin graft is a priority.

Penis

- A balloon catheter is not inserted if the patient is able to urinate spontaneously despite the level of swelling.
- If a catheter is essential for the monitoring of effective resuscitation, it must be inserted only for this purpose.
- Never cut off the prepuce because this may be folded back at a later stage to cover the penile shaft if a skin graft is necessary.
- The perineum is also treated with an antiseptic cream.

Peri-anal region

- The patient is nursed on his or her stomach; the burn wound in the region of the anus is left open and only covered with an antiseptic cream.
- The patient is encouraged to empty the colon once a day, after which the region is cleansed (showered) and covered as described.
- Emptying of the colon can be facilitated with a small enema or suppository at a controlled time of day.
- A temporary colostomy should be considered when there is faecal contamination of deep perineal, gluteal, anal or inner thigh burns, or when there is invasive sepsis in these regions.

Fig. 6.2 Superficial second degree burn; note the healing in the retained hair follicles.

Fig. 6.3 Singing of nasal hair in a patient with facial flash burn.

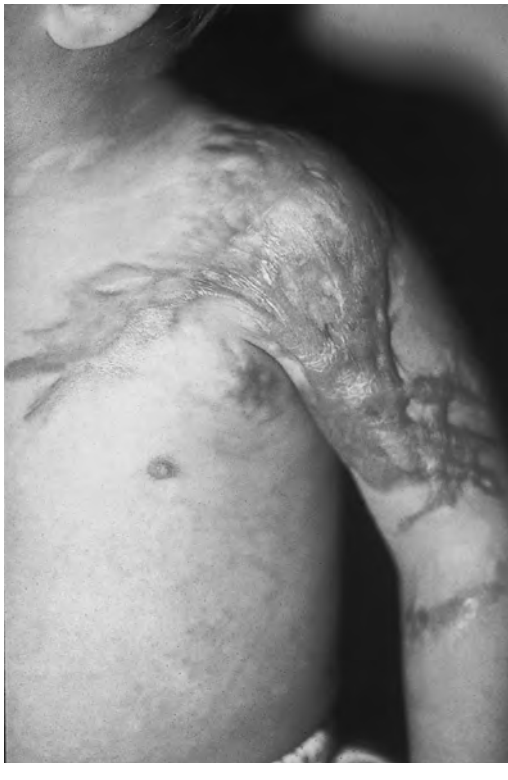


Fig. 6.4 Hypertrophied scar tissue in a deep superficial burn wound that healed with primary intent without skin grafting.

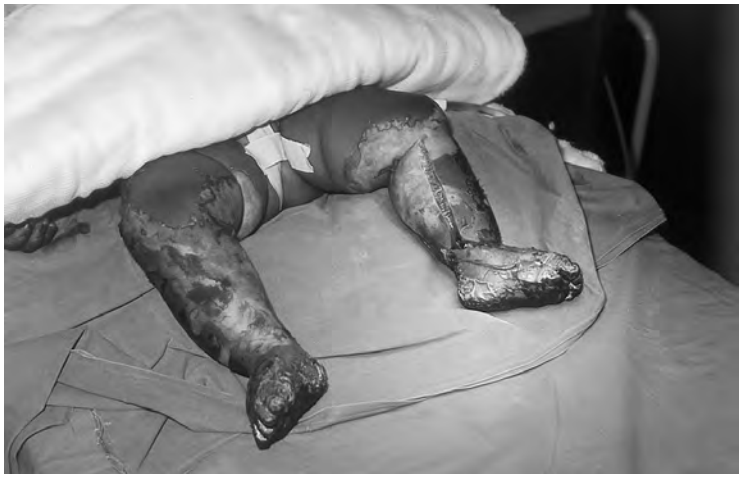


Fig. 6.5 Escharotomy in full-thickness circumferential burn wounds.

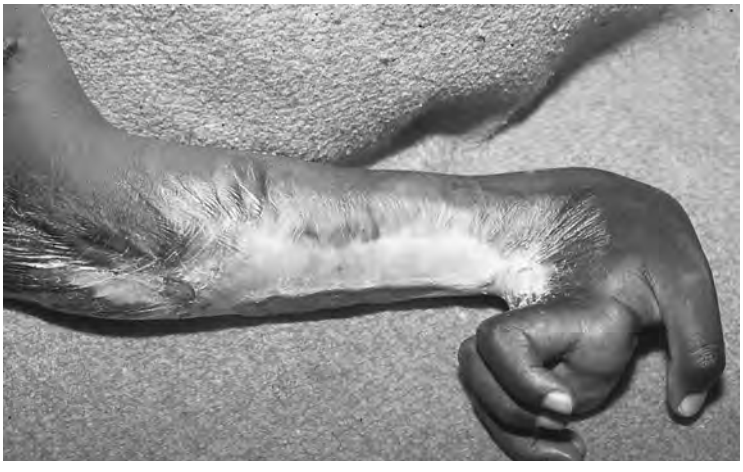


Fig. 6.6 Contractures due to scar tissue in a healed third degree wound.

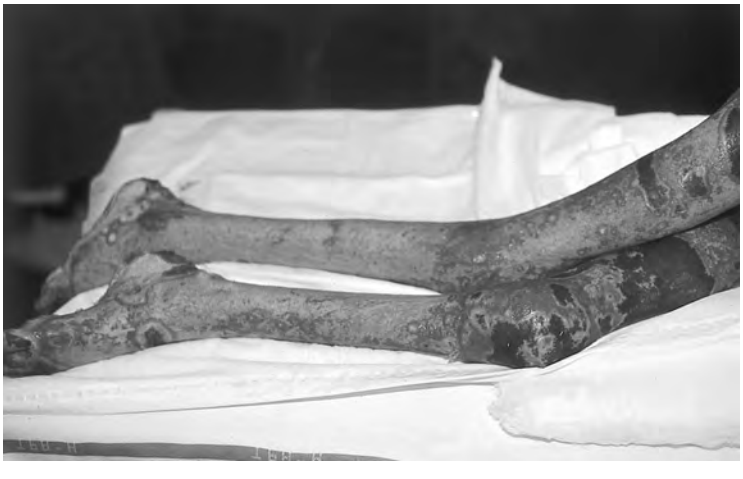


Fig. 6.7 Extensive third degree burns; note the total muscle atrophy due to catabolism caused by the injury.

Fig. 6.8 Limb loss in an electrical burn.

Hernias

7

JHR BECKER, GP HADLEY & PG BEALE

Definition: An inguinal hernia may be an indirect inguinal hernia or a femoral hernia (Berliner 1984).

7.1 Indirect inguinal hernia

Pathology

The pathology is a patent processus vaginalis between the internal opening of the inguinal canal and the scrotum. The width of the neck at the internal ring will determine what the content of the hernia sac will be. If the neck is narrow and no viscus is allowed, then only peritoneal fluid will pass through and it becomes a communicating hydrocele or fluid hernia. If the neck is wide enough to allow viscus through, then the content will usually be bowel or, in the case of a girl, the ovary and fallopian tube.

Symptoms

- A swelling due to peritoneal fluid or viscus in the hernia sac
- Pain due to
  - distension of the hernial sac
  - incarceration (see below)
  - strangulation of the content (see below)

Examination

- Undress the boy
- Palpate the testes in both hemi-scrotums
  - Determine if there are any other swellings present in the region of the testis
  - Determine the size, mobility and position of the testis

- Examine the cord in the following manner: the child lies on his back on the examination bed. With the flat hand on the abdomen, roll the cord with all its structures over the symphysis pubis with the index finger of the examining hand. The following points should be noted:
  - There may be thickening of the cord on the affected side.
  - When the opposing surfaces of the hernia sac are rubbed against each other, there is a feeling of silk – this is the so-called 'silk sign of Gross'.
  - If there is bowel present in the sac, it can often be deduced from a bubbling or gurgling sound or sensation.
- The contralateral testicle and hernial orifice must be thoroughly examined using the above method. The incidence of bilaterality is ±20% when the presenting hernia is right sided and double this when left sided. In premature baby boys the incidence of bilaterality exceeds 50%.

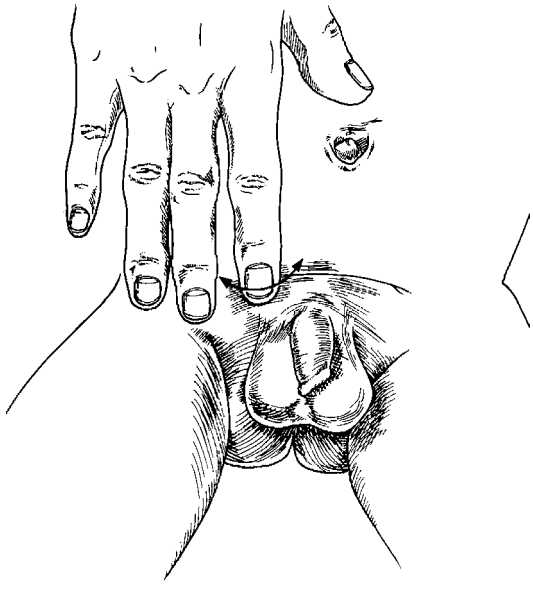


Fig. 7.1 Eliciting the 'silk sign'.

Management

Only the hernia sac is dissected free and tied off at the internal opening in the child, i.e. herniotomy. No herniorraphy or repair of the posterior wall of the canal is necessary because the pathology is not a weakened posterior wall, but rather a patent processus vaginalis.

■ WHEN MUST THIS OPERATION BE DONE?

On the first convenient elective schedule with the proviso that the following conditions have been met:

- All the facilities for paediatric anaesthesia are available.
- There are no medical contra-indications for surgery.
- A paediatric surgeon and anaesthetist are available.

■ HOW OLD SHOULD THE CHILD BE?

If the baby (premature or neonate) is medically fit to leave the hospital, he or she is also fit for an inguinal herniotomy.

7.2 Incarcerated indirect inguinal hernia

Incarcerated – irreducible
Strangulated – non-viable hernial content due to ischaemic compromise of blood supply

An indirect hernia in a child is a narrow canal that incarcerates easily. Even a premature baby's hernia can become incarcerated and must be observed. Due to the significant risk of an episode of incarceration, herniotomy is indicated as soon as there are no medical contra-indications to anaesthesia. Incarceration can progress to strangulation if not resolved without prolonged delay.

Clinical symptoms

- There is a painful, tender, firm swelling in the groin.
- The content is bowel, omentum or ovary.
- It becomes red and can look like an abscess if it has been present for a long time.
- The abdominal symptoms are similar to those of a bowel obstruction:
  - abdominal distension
  - vomiting
  - no flatus per rectum
  - abdominal tenderness (late sign)
  - increased bowel sounds

Differential diagnosis

- Inguinal adenitis or abscess
- Torsion of the testis
- Epididimo-orchitis
- Encysted hydrocele of the cord

Special investigations

- Blood gas
- Electrolytes
- X-rays of the abdomen – may show inguino-scrotal intra-luminal bowel gas
- Sonar

Emergency management

- *Nil per os* (NPO)
- Intravenous infusion
  - maintenance fluid
  - replace fluid deficit and abnormal losses
- Sedation and then elevation of the legs in a Gallows traction frame. The baby must sleep undisturbed in this position so that the muscles can relax and the hernia can slide back. The reason for this ultra-conservative approach is that there are not enough experienced medical practitioners who can safely distinguish between incarceration and strangulation with the possibility of dead bowel.

If the content does not slip back, it can be assumed that there is sufficient oedema at the neck of the hernia to compromise the blood supply, and an operation is therefore indicated.

If the baby has to travel some distance, the same principle can be adopted by allowing the baby to sleep on the mother's lap with the legs elevated against her chest. If the baby falls asleep en route and the hernia does not reduce by itself, an operation is indicated.

Most paediatric surgeons throughout the world practice reduction of paediatric incarcerated inguinal hernias by manipulation or taxis. Approximately 90% are reducible in experienced hands. The rationale is that infant incarcerated hernias are very infrequently strangulated, and if they are, they are most unlikely to reduce. This avoids an emergency anaesthetic and procedure in less-than-ideal emergency circumstances.

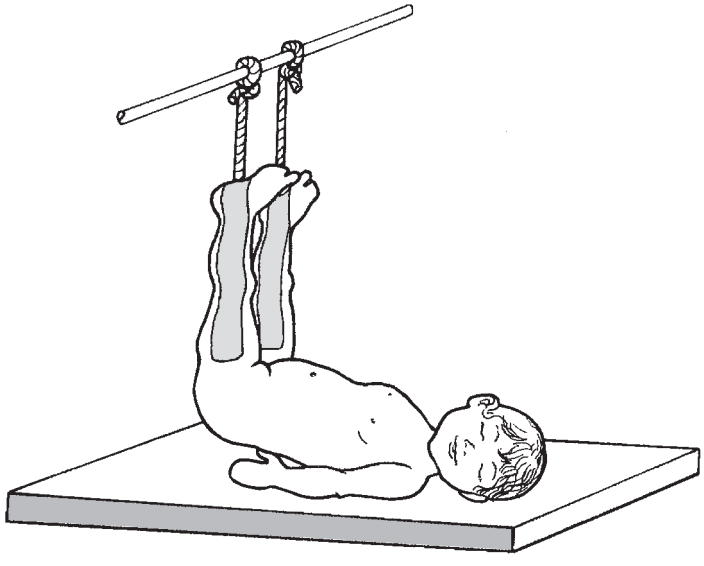


Fig. 7.2 Gallows traction in a sedated baby of less than 15 kg with an incarcerated (NB not strangulated) left inguinal hernia.

Fig. 7.3 Transport (by the parents) of a baby with an incarcerated (NB not strangulated) left inguinal hernia.

Indications for an emergency operation

- Reduction by manipulation or sedation in gallows has failed.
- Incarceration that has been present for a long time.
- Red, swollen inguinal area. If present, no attempt at hernia reduction should be made.
- Acute abdominal symptoms with incarceration.
- If the hernia, despite the above management, remains irreducible, then the baby is submitted for emergency operative repair.

7.3 Femoral hernia and direct hernia (Marshall 1983; Zaman and Taylor 1985)

These types are exceptional in paediatric practice.

7.4 Umbilical and para-umbilical hernias (James 1982)

Both of these hernias appear in the region of the umbilicus.

- The umbilical hernia appears through the umbilical ring and is obviously centrally placed in the umbilicus.
- The para-umbilical hernia is in the linea alba adjacent to and above the umbilicus; the hernia ring usually also includes the

umbilical opening. Clinically, such a hernia hangs like a little elephant's trunk because there is more skin at the top than at the bottom of the umbilical skin scar.

Natural course

Most umbilical hernias present in early infancy and tend to close spontaneously. Those that have not closed by the age of 5 years will not do so and require surgical closure. Umbilical hernia closure may be offered earlier to those that predictably have no chance of spontaneous closure. A useful guideline is a diameter of 1 cm at 2 years of age. The ultimate cosmetic result of large protuberant umbilical hernias with no prospect of spontaneous closure is improved by an early procedure.

The para-umbilical hernia does not close spontaneously and incarcerates more easily in comparison with the umbilical hernia. It is operated on when diagnosed on the next available theatre list.

Indications for surgery

- Incarceration
- Strangulation
- The para-umbilical hernia, because it does not heal spontaneously and tends to complicate, is operated on electively once the diagnosis has been made.

Surgery

The surgery entails:

- An intra-umbilical transverse skin incision is made.
- The hernia sac is dissected out.
- If it is not too large, a purse string suture is placed in the ring and the defect is pulled closed.
- If the opening is too large, interrupted non-absorbable sutures are placed to close the defect.
- Although not a cosmetic operation, the surgeon should achieve a cosmetic result with little or no evidence of surgical intervention. It is important to preserve the umbilicus for psychological reasons.

7.5 Omphalocele (Kim 1976)

Definition

An omphalocele or exomphalos is a membrane-covered central abdominal wall defect. Depending on the size, varying amounts of the abdominal organs, including bowel and liver, can be present in the hernia sac. Other congenital defects, especially associated heart defects, may co-exist. Beckwith Wiedeman syndrome or EMG

(exomphalos macroglossia) is associated in about 12% of patients and it is important to identify it due to associated hyperinsulinaemic hypoglycaemia and its consequences. Exomphalos major measures > 4 cm at the base and has solid and hollow visceral content. Exomphalos minor is < 4 cm with hollow visceral content only. It is important to recognise the lesser form – a so-called hernia into the base of the cord – and to avoid damage to a loop of bowel in the base of the cord when the cord is tied after delivery.

Diagnosis

An obvious large congenital membrane-covered umbilical hernia that contains abdominal organs with:

- intact membrane or
- ruptured membrane

Emergency management

■ RUPTURED MEMBRANE

- Give an intravenous infusion of 5% dextrose water or 0,25% NaCl.
- Monitor blood glucose (vene-puncture or heel prick) + dextrostix.
- Protect the intestines lying alongside the baby outside the ruptured membrane sac against:
  - drying out
  - hypothermia
  - sepsis
- Use the following methods:
  - Rinse the bowel with warm saline or diluted lukewarm Betadine® solution.
  - Place the baby in a sterile transparent plastic bag with a wide neck to cover exposed bowel and insulate against heat and fluid loss.
- Place the baby in an incubator with oxygen.
- Transfer the baby to a paediatric surgical unit, informing the unit that the baby is on the way.
- The baby must be accompanied by a medical doctor or skilled nursing staff.

■ INTACT MEMBRANE

- Give an intravenous infusion of 5% dextrose water or 0,25% NaCl.
- Monitor blood glucose.
- Paint the membrane with Betadine® solution.
- Place the baby in an incubator with oxygen.
- Transfer the baby to a paediatric surgical unit.

Fig. 7.4 Omphalocele; note the intact membranes and umbilicus.

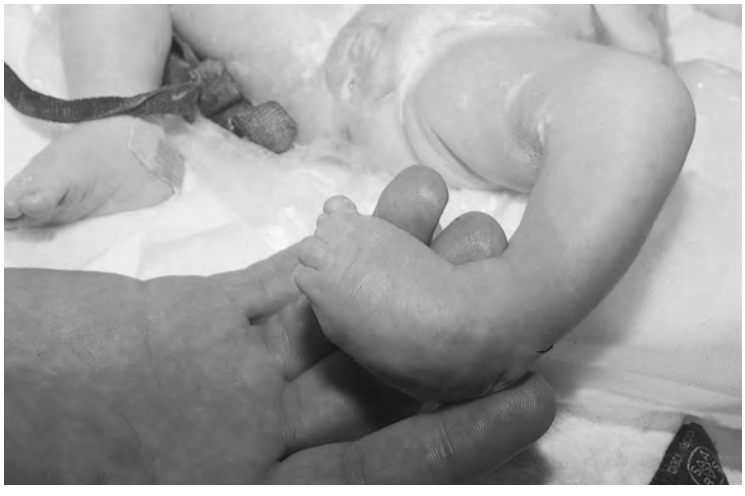


Fig. 7.5 Omphalocele with associated limb anomalies.

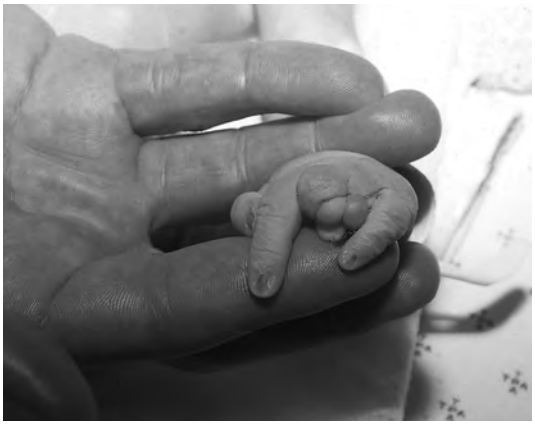


Fig. 7.6 Omphalocele with associated limb anomalies.

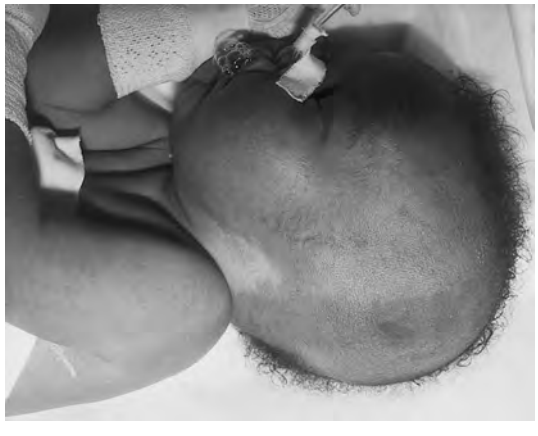


Fig. 7.7 Omphalocele, absent ears.

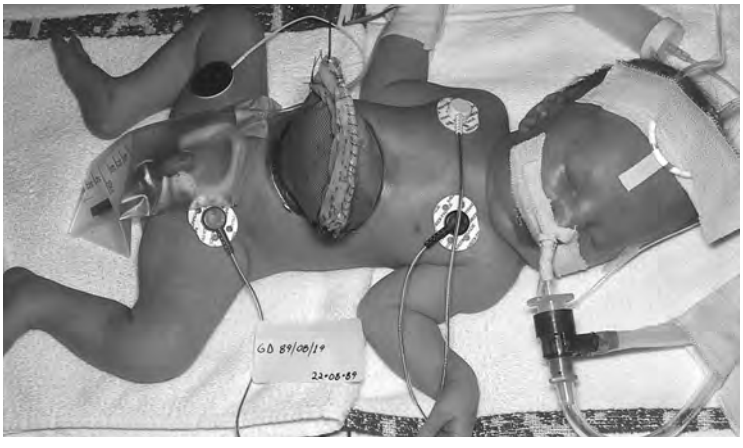


Fig. 7.8 Omphalocele/gastroschisis. 'Silo' to facilitate the reduction of 'excess bowel' into the abdominal cavity.

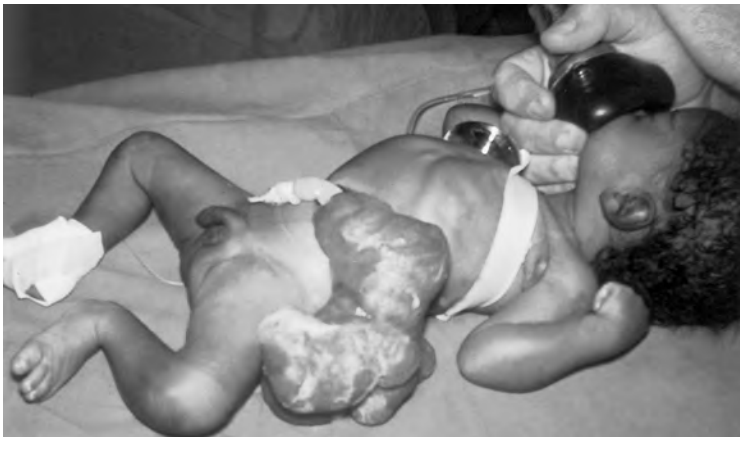


Fig. 7.9 Gastroschisis; note the abdominal defect, no membranes, normal umbilical cord, extra-abdominal bowel and adhesive fibrin on the bowel due to intra-uterine exposure to amniotic fluid.

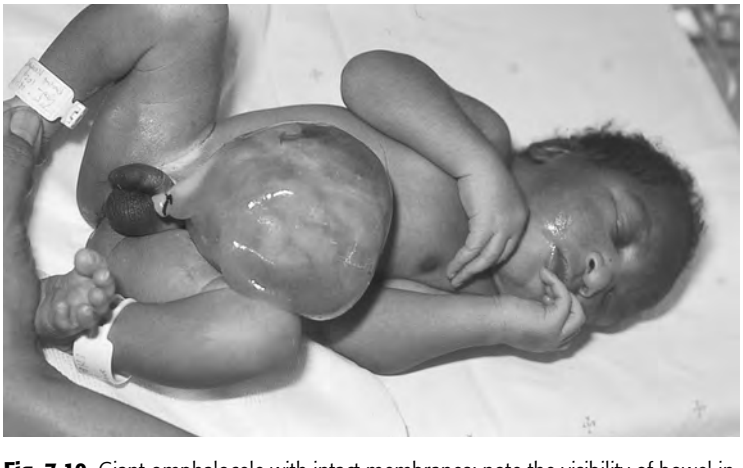


Fig. 7.10 Giant omphalocele with intact membranes; note the visibility of bowel in the sack.

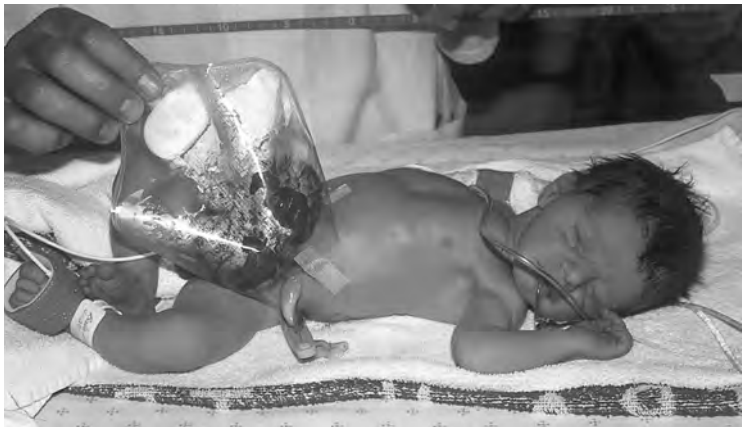


Fig. 7.11 Emergency treatment of exposed extra-abdominal bowel with a plastic bag; note the fog on the inside of the bag due to evaporation of fluid from the bowel surface.

7.6 Gastroschisis

World-wide the incidence of gastroschisis has become two to three times more common than exomphalos. In contra-distinction to exomphalos it is not a genetically related congenital abnormality and does not tend to have genetically associated abnormalities.

Definition

This is a congenital abdominal wall defect, lateral to the right of the umbilicus in 98% of cases. The bowel is always exposed and there is never a covering membrane present. Due to the open abdominal cavity and exposed bowel, these neonates are particularly prone to fluid and heat loss. The interval between delivery and closure of the abdominal wall should be limited and the baby protected with adequate infusion plus prevention of hypothermia as described for ruptured exomphalos. Many abdominal wall defects are identified prenatally by ultrasound.

Where gastroschisis is diagnosed, the baby should be transferred prenatally and delivered in a specialised centre where a neonatal ICU and neonatal surgical care are available. A select group can be reduced in the neonatal ICU without anaesthesia. The majority, namely 60–70%, are closed by primary closure. The rest cannot be closed without causing respiratory failure or abdominal compartment syndrome, and require staged closure with the use of a prosthetic bag or 'silo' on the anterior abdominal wall. An American company, BENTEC, produces a custom-made bag with a spring ring in the base which can be applied without anaesthesia or operative intervention. Gastroschisis patients have prolonged recovery of intestinal transit and require extended maintenance of intravenous alimentation. The current expected survival rate is approximately 90%.

7.7 **Bochdalek hernia** (Reynolds et al. 1984)

JHR BECKER & GP HADLEY

Definition

This is a congenital diaphragmatic hernia with abdominal content herniating into the chest cavity.

Depending on the size of the defect and the amount of pressure from the abdominal content on the heart and lungs, the baby may become cyanotic and die unless there is effective intervention. Cyanosis may already be present at birth or may develop later (even weeks to months later).

The classic picture is that of a newborn baby with:

- cyanosis
- scaphoid abdomen
- pectus carinatum

Confirm the diagnosis with a chest X-ray, which will clearly show bowel in the relevant hemi-thorax:

82% on the left
12% on the right
6% other combinations

Emergency management

The baby has cyanosis, respiratory distress and develops a state of shock. Therefore:

- Intubate and administer oxygen quickly and continuously.
- Give an intravenous infusion of 5% dextrose water or 0,25% NaCl.
- Confirm the diagnosis with a chest X-ray.
- Pass a No. 8 or 10 nasogastric tube in order to allow the stomach content and air to escape.
- Note whether a pneumothorax is present on the contralateral side and drain if necessary (Hansen *et al.* 1984). (Underwater drainage apparatus must travel with the patient during transport.)
- Place the baby in an incubator and ventilate continuously.
- Transfer the patient rapidly to a paediatric surgical unit, informing them that the patient is already en route. A medical doctor should accompany the patient.

General

The prognosis for a Bochdalek hernia (Becker 1987) depends on how soon after birth the baby became cyanotic. The sooner this happens, the worse the prognosis. There is practically 100% mortality (Reynolds *et al.* 1984) if the baby is already cyanotic at birth

and almost 100% survival if the baby only develops symptoms after the first 24 hours. Associated congenital abnormalities must also be considered when determining the prognosis.

The decision about the baby's prognosis must be left to the paediatric surgeon. The baby must be transferred as soon as possible to the paediatric surgical unit after emergency management has begun.

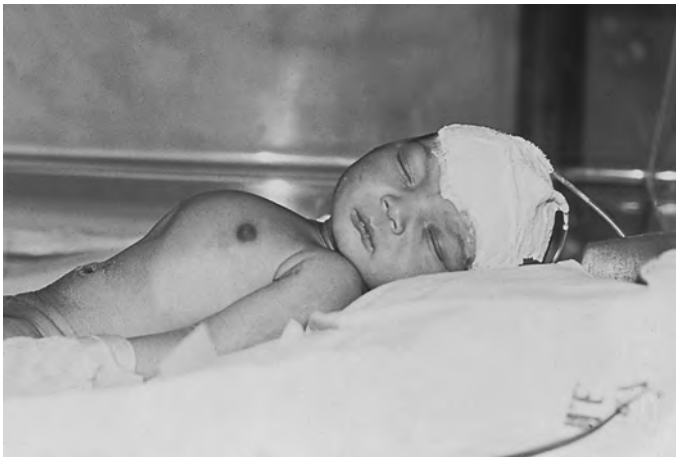


Fig 7.12 Bochdalek hernia; note the scaphoid abdomen and prominent chest due to the displaced bowel.

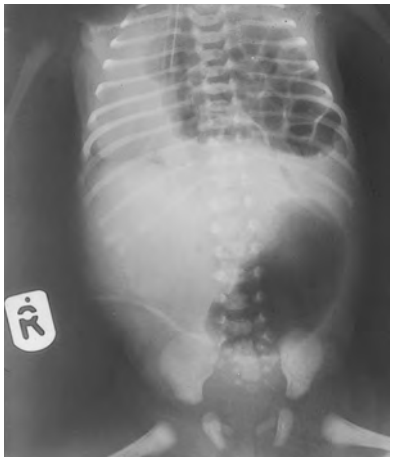


Fig 7.13 X-ray of a baby with a Bochdalek hernia; note the bowel in the left hemi-thorax.

Jaundice

8

JHR BECKER & MH SHEIK-GAFOOR

Jaundice in the newborn is a rather general condition, and the art for the clinician lies in establishing whether it is normal physiological jaundice or the complex problem of neonatal obstructive cholangiopathy (neonatal hepatitis and biliary atresia).

8.1 Causes of jaundice

Medical

Medical jaundice is a condition managed without invasive surgery, in other words the aetiology is not surgically managed. Examples are given below.

■ HAEMATOLOGICAL (RAISED BREAKDOWN OF RBC LEADING TO JAUNDICE)

- Problems outside of the red blood cell:
  - One of many problems is the presence of antibodies, e.g. ABO incompatibility or hypersplenism, etc.
- Problems in the cell wall of the red blood cell:
  - This means that the morphology of the red blood cell is abnormal, e.g. spherocytosis, elliptosis, etc.
- Problems in the red blood cell:
  - Enzyme deficiencies, e.g. Pyruvate-kinase deficiency

■ INFECTIVE (NEONATAL HEPATITIS)

- Viral hepatitis, e.g. cytomegalovirus, herpes, etc.
- Toxoplasmosis and congenital syphilis

■ DIVERSE

- Rare diseases, e.g. Gilbert's syndrome, Gaucher's disease, Niemann-Pick diesease or galactosaemia

Surgical

This usually presents as an obstructive jaundice, for example:

- Biliary atresia
- Biliary hypoplasia
- Choledochus cyst
- Inspissated bile syndrome (cholestasis) (alpha 1 antitrypsin deficiency) (Balistreri 1985)

8.2 Management and treatment of jaundice

If the cause is medical in nature, e.g. with a raised breakdown in red blood cells in congenital spherocytosis, septicaemia or ABO incompatibility, then the blood analysis will give an indication of where the pathology lies, e.g. morphology of the red blood cells, abnormal enzymes or positive viral studies.

A thorough examination, i.e. good medical history-taking and judicious biochemical investigations, is the method to follow to make a diagnosis. The most important test for differentiating between medical or surgical jaundice is the presence or absence of bile pigments, stercobilinogen in the stool or urobilinogen in the urine. If the latter pigments are present, then they could only have got into the digestive tract via the bile ducts and in no other manner – but it is unlikely that there would be a surgical cause. The exceptions are a choledochal cyst which would be easy to detect with a sonographic investigation.

The absence of bile pigment in the stool or of urobilinogen in the urine indicates the absence of communication between haepatocyte and bowel, which in turn indicates a surgical cause, e.g. atresia or the medical condition of neonatal hepatitis. In some cases it is almost impossible to differentiate pre-operatively between neonatal hepatitis and neonatal obstructive cholangiopathies (biliary atresia, biliary hypoplasia).

The liver biopsy may image identical patterns in neonatal hepatitis and in neonatal obstructive cholangiopathies, i.e.

- With a sonographic image there is no dilatation of the intra- and extra-hepatic bile ducts.
- The stools are white and occasionally have a streak of yellow discoloration coming from pigment derived from the bowel canal.

If the above-mentioned stool picture is found and the neonatal jaundice does not improve within 3 weeks after birth, then the child requires rapid surgical intervention. Surgical exploration must be done at a paediatric surgery unit with an open liver biopsy and an intra-operative cholangiogram done through the gallbladder.

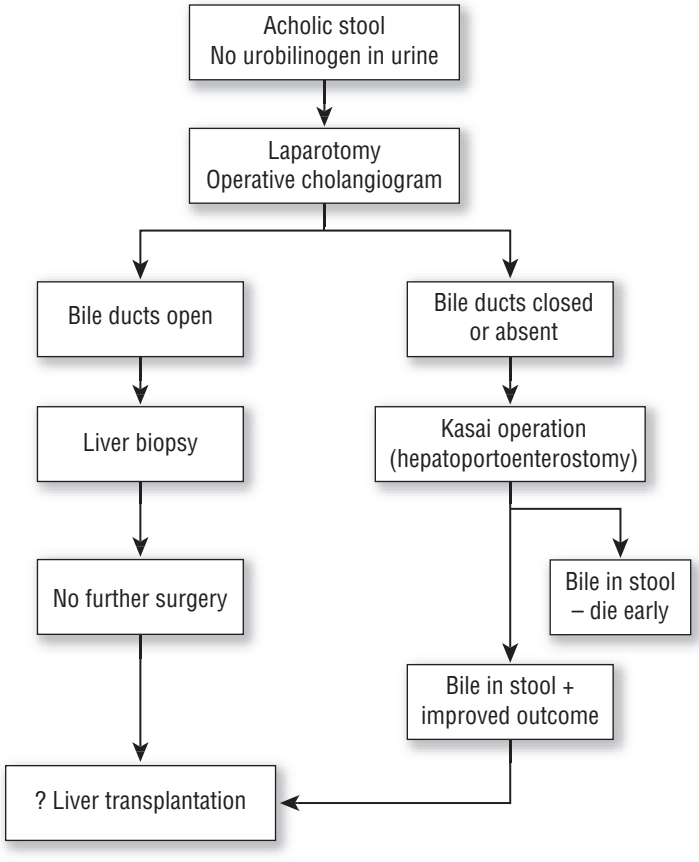


Fig. 8.1 Flow diagram for diagnosis and treatment of jaundice.

If atresia of the bile ducts is diagnosed with laparotomy, then there must be immediate progression to a porta-enterostomy (the Kasai operation (Kasai 1974)). If bile ducts are present, then proceedings are halted to perform an open-liver biopsy as this would more than likely be a case of neonatal hepatitis. Ultrasound is of value in the pre-operative work-up of these patients because the ultrasound investigation can image if there is a choledochus cyst present, which is managed like biliary atresia with a porta-enterostomy. The choledochus cyst which is usually present in 30% of cases has the classic triad of abdominal pain, right upper abdominal mass and obstructive jaundice.

8.3 Prognosis of the Kasai operation

The Kasai operation (porta-enterostomy) will, with expertise, allow bile flow in more than half of the patients, provided that they are operated on before the age of 2 months. Patients with bile flow to the bowel survive for a longer period of time (as long as 10 years) and in this way become good candidates for liver transplantation. Those who had unsuccessful surgery, those who had surgery too

late (after the age of 2 months) and those not operated on at all, die in their second year of life due to liver failure. Adjuvant measures that have been shown to improve results include the use of steroids, antibiotics, urodeoxycholic acid and special diets containing medium-chain triglycerides.

8.4 Gallbladder disease in the child

Gallbladder disease is rare in a child, except in the case of haemolytic disease where there is a raised incidence of gallstones (Bunyapen *et al.* 1986; Pollak *et al.* 1982). An ultrasound investigation of the gallbladder is always recommended when planning a splenectomy for haemolytic disease. Acute cholecystitis and gallstones are actually not unknown in children (Pollak *et al.* 1982; Bunyapen *et al.* 1986). They are often incorrectly diagnosed as acute appendicitis. The management is usually initially medical with the use of antibiotics. Confirm the diagnosis with an ultrasound investigation and if more information is required, then do an oral cholecystogram. A cholecystectomy can be done after clearing of the initial acute phase. Acalculous cholecystitis should also be considered in the ill, septic, intensive care patient. The early signs of the disease are frequently missed as the patient is often intubated and the commonest presentation is deterioration in the condition of a previously stable patient. The diagnosis should be considered and confirmed by ultrasound examination, which demonstrates a distended thickened gallbladder with intraluminal echogenic debris. Management is usually a cholecystectomy if the patient's condition allows or percutaneous cholecystostomy in the unstable patient.

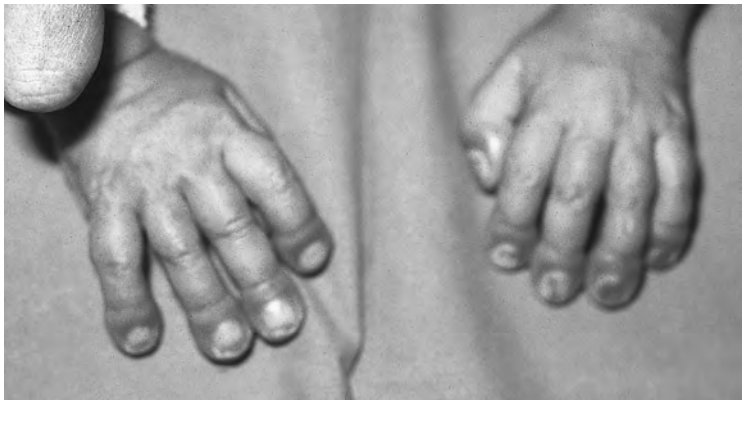


Fig. 8.2 Finger clubbing in a patient with terminal liver disease due to biliary atresia.

Gastro-oesophageal reflux (GOR)

9

JHR BECKER & ML VAN NIEKERK

9.1 Pathophysiology of gastro-oesophageal reflux

The anatomical and physiological factors of importance in overcoming GOR are as discussed below.

The angle of His

This is the angle between the fundus of the stomach and the oesophagus. The natural response to a pressure increase in the fundus of the stomach is closure of the oesophagus, which prevents reflux. Conversely, if the angle is not present, such as with a hiatus hernia, then raised intragastric pressure will allow stomach contents to reflux into the oesophagus.

Intra-abdominal segment of the oesophagus

This segment is exposed to positive intra-abdominal pressure in comparison with the negative pressure of the intra-thoracic oesophagus.

Zone of high pressure

At the gastro-oesophageal transition zone, there is an area with higher tone that performs a sphincter action to prevent reflux. This area can be regarded as being the lower oesophageal sphincter (LES).

Mucosal rosettes

These are mucosal folds in the lumen that seal the region tightly during raised pressure in the high-pressure zone.

Closure pressure

The above mechanisms all work together to create a critical closure pressure.

Intrinsic function of the oesophagus

This is the ability of the oesophagus to empty content via peristalsis.

Opening pressure

This is the ability of the oesophagus to generate sufficient pressure to overcome all the mechanisms mentioned above (analogous to the pressure exerted to open a door).

Disturbance in any of the above mechanisms will lead to pathological reflux.

Clinical reflux

All newborn babies reflux. The amount and extent of reflux decreases progressively during the first year of life.

Reflux sometimes occurs with such ease that it is difficult to differentiate it from projectile vomiting as found in hypertrophic pyloric stenosis.

Reflux may even cause the baby to become irritated and uncomfortable, with arching of the back and sleep disturbances due to the pain it causes.

The majority of babies should be symptom-free at the age of 2 years, by which time the above-mentioned mechanisms will have matured. In the interim, medical management is of great value.

9.2 Main aspects of pathological gastro-oesophageal reflux

The main problems are lung complications, systemic effects and local complications.

Lung complications

Aspiration of gastric contents can cause the following:

- recurrent pneumonias
- chronic fibrotic lung changes
- pulmonary abscesses
- bronchiectasis
- apnoea spells secondary to laryngospasm
- Sudden Infant Death Syndrome (SIDS)
- asthma and bronchospasm

Systemic effects

- Insufficient weight gain or failure to thrive. The mechanism is a significant calorie deficit due to vomiting, or refusal to eat as a consequence of oesophagitis.
- Irritability
- Sleep disturbance
- Neurological symptoms, e.g. seizures or hypotonia

Local complications

What is meant here is that complications arise in the oesophagus as a result of the acidity, or sometimes even the alkalis, to which the mucosa is exposed. These may manifest as:

- ulceration with bleeding and anaemia
- stricture as a result of oedema and later as a result of fibrosis
- stomach mucosa metaplasia (Barrett's oesophagus)
- adenocarcinoma

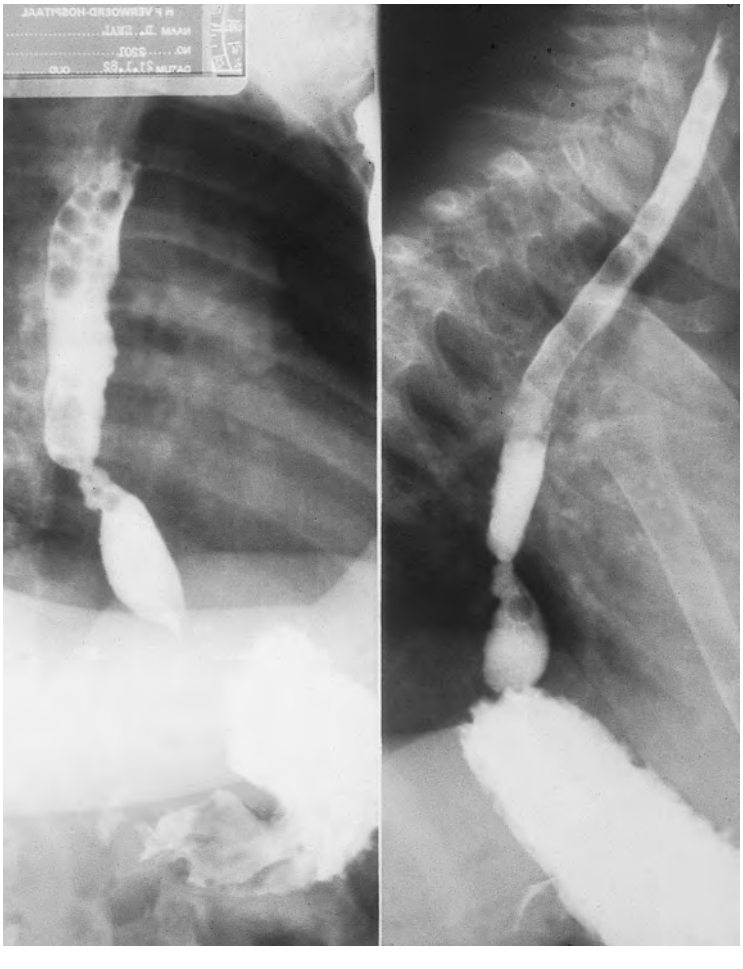


Fig. 9.1 Oesophageal strictures due to gastro-oesophageal reflux disease in a baby.

9.3 Basis for treating the baby or child with gastro-oesophageal reflux

Reflux in a child whose lungs have developed normally and without complications, both systemically and locally, can be examined using the necessary special investigations to set a base line. Manage these children with medical support, reassurance and regular follow-up examinations.

9.4 Medical management

Anti-reflux position

This is a 30–40° prone head-up position. This position must be maintained in the bed, as well as when the baby is carried. When the baby is carried, he or she will lie across the forearm with the stomach and anterior chest facing downward, the head alongside the cubital fossa, and the face turned downwards and outwards. The person carrying the baby will hold him or her between the legs and the baby's body will lie antero-laterally against the carrier's abdomen.

In the anti-reflux position, the gastro-oesophageal junction is the highest point and the pylorus the lowest. This causes winds to be passed more easily ('burping') and the solids content (e.g. milk) to move to the lowest point, in the direction of the pylorus.

Milk thickeners and anti-regurgitation milk formula

Nestargel® (Nestlé), adding of infant breakfast cereal to the feed and antireflux milk formula help to prevent the stomach contents from pushing up into the oesophagus, and are valuable first-line measures to prevent reflux.

Gaviscon

This reacts with saliva and gastric acid to produce a viscous gel which floats on the gastric content and suppresses gastric reflux.

Antacid drugs

H_2-receptor antagonists and proton pump inhibitors are the next line of treatment when the above measures still fail to control reflux.

Prokinetics

Methoclopramide, Domperidone and Erythromycin are prokinetic drugs. They have limited value in the treatment of gastro-oesophageal reflux.

9.5 Special investigations of importance

Barium swallow and barium meal

This is an important functional anatomical examination. The following should be noted:

■ PHARYNGEAL SWALLOW PHASE

The effectiveness of this function is important because it regulates the passage of food away from the airway and to the oesophagus. You must check that there is no aspiration here due to a malfunction of the pharynx (bulbar or pseudo-bulbar paralysis).

■ OESOPHAGEAL SWALLOW PHASE

Check:

- peristalsis
- prograde movement of the content
- emptying of the oesophagus
- retention in the oesophagus
- local effects of reflux, e.g. ulceration, stenosis or stricture

■ GASTRO-OESOPHAGEAL JUNCTION

Check for:

- presence of a hiatus hernia
- spasticity (non-relaxation) at the junction
- gastro-oesophageal reflux

■ STOMACH AND DUODENUM

Check:

- stomach emptying to the duodenum
- co-ordination of peristalsis in the stomach with receptive relaxation of the pyloric muscle

Ambulant oesophageal pH measurement

An electrode is placed in the oesophagus to measure the pH for 24 hours.

Measurements of importance

- Percentage of time which pH is less than 4
- Mean duration of reflux episodes
- Number of reflux episodes per 24 hours
- Number of reflux episodes lasting more than 5 minutes

This investigation will help you to determine the quality and quantity of the reflux, as well as the ability of the oesophagus to rid itself of the content.

Lung X-rays

This is an important way of determining radiologically the extent of lung damage.

Lung suctioning

After suctioning the patient's airway, the physiotherapist must send a specimen of the secretion obtained to the laboratory for analysis. Macrophages containing milk should be sought because their presence is pathognomonic for milk aspiration.

Endoscopy (oesophagogastroscopy)

The damage done in the oesophagus is visualised and documented, together with the presence or absence of a hiatus hernia.

Other investigations that may be used

- Manometry – when motility disorders are inspected
- A radio-isotope study

9.6 Surgical treatment

The goal of surgery is to re-establish the antireflux barrier.

Indications for surgery

Gastro-oesophageal reflux and its complications are unmistakeably confirmed with special investigations by one of the following:

- failed medical management
- serious lung complications
- serious local complications
- serious systemic complications
- hiatus hernia with GOR

Operation of choice – Nissan fundoplication

The laparoscopic Nissan fundoplication is replacing the open procedure worldwide. The Nissan fundoplication is a complete 360° wrap. This is done by mobilising the fundus and intra-abdominal oesophagus. The mobilised portion of the fundus is brought behind the oesophagus, and the anterior wall of the fundus is brought anterior to the oesophagus to form the antireflux barrier. The left and right crurae are also sutured together, using non-absorbable sutures.

The child and trauma

10

JHR BECKER, AB VAN AS, BGP LINDEQUE, HP SHAPIRO & JAN J VAN WINGERDEN

Trauma in children is the greatest cause of child deaths worldwide. Every year, motor vehicle accidents alone claim lives equivalent to the number of children in four primary schools, and maim and disable children equivalent to the number of children in 48 primary schools. Blunt trauma is present in 80% of all accidents and head injuries are the major cause of death.

According to the Medical Research Council (5th annual NIMSS report, December 2004), approximately 3 000 children under the age of 15 years die each year in South Africa from unnatural causes. The most common cause again is motor vehicle accidents (see Table 10.1).

TABLE 10.1 Causes of non-natural childhood death: the big killers in South Africa

Motor vehicle pedestrian fatalities	28,2%
Motor vehicle passenger fatalities	7,6%
Drowning	13,7%
Burns	13,3%

Polytrauma is more common than in adults (due to the proximity of all body parts) but fortunately is not as dangerous in the child as in the adult. The presence of extensive head injuries is a strong determining factor in the chance of survival.

A child's organs are still undamaged by bad habits, e.g. smoking and drinking, and the ability of the tissue to regenerate gives the child an exceptional chance of recovery.

The protective structures of the vital organs are, however, not yet completely developed, e.g. the ribs are soft and supple, and

underlying organs can be injured even without a fracture. The liver and spleen are not yet protected by the rib cage in the young child as they are in the adult.

Similarly, the kidneys are protected in the adult in the paravertebral gutter, but in the child this gutter is shallow and the kidneys are relatively large, which then exposes the kidneys to blunt trauma.

In addition, the bladder is an abdominal organ in children which is easily burst by a blow to the lower abdomen, whereas in an adult the bladder is a pelvic organ and therefore less vulnerable. The baby's skull suture lines are not yet closed, which means the brain is particularly exposed to trauma.

When managing trauma in a child, the physiological differences between a child and an adult have to be borne in mind and the factors involved in effective fluid therapy (Chapter 3) are also important here.

The monitors for successful resuscitation as described in Chapter 6 on *Burn wounds* should also be used here.

10.1 Emergency management

The principles of resuscitation as described by the APLS apply here:

A Airway

Ensure an open airway, free of foreign bodies, e.g. a sweet lying on top of the vocal cords or a peanut lodged in the trachea. Other foreign bodies are blood, saliva and vomitus. Use *chin lift* (larger child) or *jaw thrust* (smaller child) to pull the tongue out of the airway.

B Breathing

After the airway has been opened, it must be determined whether the patient is breathing properly. If not, then artificial ventilation is commenced immediately.

B Bleeding

Stop bleeding at any visible point of blood loss and be aware of occult blood loss.

C Circulation

Note whether the heartbeat is still present and react appropriately if it is not, e.g. do external cardiac massage.
Get an IV going.
Do compatibility and other blood tests.

D Disability

Consciousness, GCS
Neurotrauma precautions should be taken.

E Exposure, evacuation

When exposing the child, keep in mind the dangers of hypothermia. Should the patient be transported while lying down, sitting or walking, or by road or air? Should the doctor accompany the patient? Should the child be covered, etc.?

F Finish

Everything has been done and the patient has passed away – this is an active decision which must be taken.

10.2 Traumatic brain injuries

HP SHAPIRO

Traumatic brain injuries (TBI) are common in the paediatric age group. The majority of patients with brain injuries, whether the result of accidental or non-accidental trauma, are not managed primarily by a neurosurgeon.

Classification of traumatic brain injuries (TBI)

There are numerous classifications of TBI:

- Mechanism of injury: blunt vs. penetrating
- Open vs. closed
- Severity: as determined by the Glasgow Coma Scale
- Primary vs. secondary injuries

Primary injuries occur at the time of trauma and include diffuse axonal injury, cerebral contusion and brain laceration.

Secondary injuries occur as a result of secondary intracranial or extracranial insults. It is for these secondary injuries that intervention can be instituted either to prevent or treat them.

Secondary intracranial injuries of importance include extradural and subdural haematomas and cerebral oedema. These insults result in an increase in intracranial pressure, brain shift and herniation. In a child hyperaemia as a result of vasoparalysis is an important treatable cause of neurological deterioration and can occur up to 72 hours after the initial trauma.

Important preventable extracranial causes of neurological deterioration include hypoxia and hypotension.

Acute management

Acute management of the TBI patient begins with the airway and with in-line spine immobilisation. The prevention of hypoxia via supplemental oxygen or, in the case of comatose patients, by intubation and ventilation is of paramount importance in determining outcome. Respiratory causes of hypoxia must be assessed and treated.

A cause of circulatory compromise, i.e. hypotension, must be sought and treated appropriately. The blood pressure should be appropriate for age in order to maintain adequate cerebral perfusion. TBI in older children does not result in shock and an alternate cause of the shock must be sought.

The *pitfall* is in the neonate and young baby where an intracranial bleed may result in shock and anaemia.

Once the patient is stable from a cardio-respiratory point of view, he or she is assessed from a neurological point of view. In trauma the Glasgow Coma Scale (motor, verbal and eye response) or in paediatrics a modified Glasgow Coma Scale has become the accepted method of evaluation. Pupils are assessed for size and reaction to light.

Once the TBI patient has been assessed and resuscitated and is stable, he or she is referred for a computerised tomography (CT) scan of the brain.

Adjuncts to treatment

Where raised intracranial pressure (ICP) is suspected and there is deterioration in the neurological condition, measures must be taken to prevent a further increase in ICP. These measures include:

- Elevation of the head of the bed 30° (NOTE: Beware of spinal injuries!) Hyperventilation, to a $PaCO_2$ 28–32 mmHg, is excellent for hyperaemia as it results in vasoconstriction and a decrease in intracranial blood volume.
- Mannitol 20% (0,25–1,0 g/kg IVI) may be given but must be used with caution in children as a paradoxical increase in ICP may occur as a result of increased cerebral blood flow (Professor Van Rensburg, personal communication).
- There is no role for corticosteroids in the management of acute trauma at present.
- An intracranial haematoma must, of course, be evacuated.
- Analgesia and short-acting sedation may be used where appropriate.

Specific injuries

■ SCALP LACERATIONS

Because of the rich blood supply to the scalp, bleeding can be most profuse and can result in shock. Wounds should be cleaned, debrided and sutured and are best treated by primary closure.

■ SKULL FRACTURES

Skull fractures can be open or closed, linear or stellate and depressed or non-depressed. They may also involve the vault or skull base. Simple depressed closed skull fractures are of importance if associated with an intracranial bleed or are of cosmetic importance.

Open skull fractures require debridement and elevation of the bone fragments with repair of the dural defect.

Base of skull fractures can be classified into anterior fossa fractures and middle fossa fractures and can be diagnosed from their clinical presentation.

Anterior fossa fractures can present with:

- periorbital haematomas ('raccoon eye')
- rhinorrhea
- subconjunctival haemorrhage
- loss of smell

Middle fossa fractures may present with:

- Battle's sign
- otorrhea
- haemotympanum
- hearing loss
- a lower motor neuron seventh cranial nerve palsy

■ EXTRADURAL/EPIDURAL HAEMATOMA

Extradural haematomas are often associated with a skull fracture. They are usually bleeds of arterial origin. On history, a lucid interval may have been present. As there is usually no underlying primary brain injury, the prognosis is good if the haematoma is evacuated as soon as possible. It is for this reason that a general practitioner must be able to perform exploratory burr holes.

■ SUBDURAL HAEMATOMA

This is often associated with a primary brain injury and therefore the child is unconscious from the beginning. Subdural haematomas are often of venous origin. Evacuation of the haematoma is important. The prognosis is often not as favourable as that of an extradural haematoma due to the underlying primary brain injury.

■ CONCUSSION

Transient loss of consciousness followed by recovery is present in a concussion. Children may complain of headache, vomit, or be apathetic or irritable after the trauma. These symptoms are often transient and children often recover, fully or partially, by the time they are seen by a doctor. It is prudent to observe the child for neurological deterioration. Admission to hospital is therefore often indicated.

10.3 Chest injuries

AB VAN AS

The fact that the child's chest is very supple and that underlying organ injuries can occur without fractures must always be borne in

mind. This is in contrast to the adult, in whom rib fractures usually draw attention to an underlying injury.

Three main groups are considered in the examination of the chest:

- lungs
- heart
- mediastinum

Rib fractures

Rib fractures can provide a good indication of underlying injuries.

■ 1ST AND 2ND RIB FRACTURES

These are indicative of high-velocity injuries, and are frequently associated with injuries of structures in the thoracic outlet, such as the great vessels and/or brachial plexus.

■ 3RD TO 9TH RIB FRACTURES

These are usually the result of direct trauma to the chest wall and often associated with injuries of the internal organs of the chest.

■ 10TH TO 12TH FRACTURES

These usually also occur as a result of direct trauma. However, these fractures are commonly associated with injuries to the contents of the upper abdomen, such as the liver, spleen and, retroperitonially, the kidneys.

Haemothorax, pneumothorax, haemopneumothorax

If the patient is dyspnoeic, confirm the clinical examination with a standing or sitting chest X-ray and act appropriately, with underwater drainage if necessary.

An AP radiograph of the chest only can easily miss a pneumothorax (40% according to some studies!) Therefore, in a polytraumatised patient with respiratory distress (and no other obvious causes for hypoxia) insertion of bilateral chest drains is advocated.

- Tension pneumothorax
  - severe respiratory distress and air hunger
  - trachea displaced contralaterally
  - clinically, no breathing sounds auscultated ipsilaterally
  - signs of raised intrathoracic pressure, with neck vein congestion – the emergency treatment is placement of an underwater drain or needle aspiration before the necessary X-ray is taken

Heart

Heart contusion can have the same effect on the function of the myocardium as that of a myocardial infarction. If during resuscitation it is found that the patient is not responding to the resuscita-

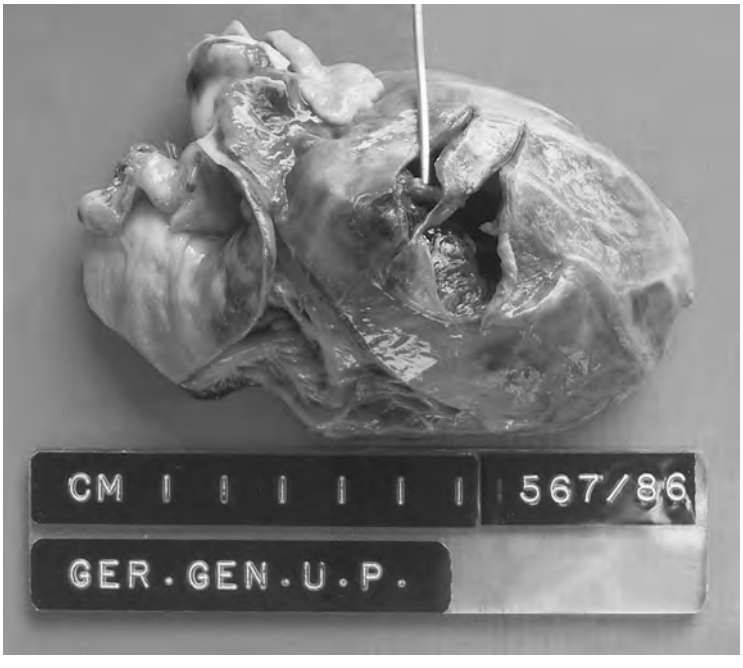


Fig. 10.1 Blunt trauma of the heart; note the cardiac contusion.

tion as expected (perfusion remains weak), then the patient must be examined for heart contusion. The clinical signs are a persistently low blood pressure in spite of satisfactory resuscitation. The assistance of a cardiologist must be sought without delay and the possibility of using an aortic balloon pump and placing the patient in intensive care should be considered immediately.

Mediastinum

Many major structures are located here, any of which can be easily injured. Be aware of pathology in the following cases:

- Broad mediastinum – more than one-third of the cross-section of the thorax at the height of the fourth vertebra, as seen on the AP standing or sitting chest X-ray. In children under 8 years, however, this is most commonly caused by the projection of the thymus, while a rupture of the aorta or major vessels is very rare, due to the elasticity of the organs.
- Air in the mediastinum – indicates a trachea, bronchus, oesophagus or lung injury and must be investigated further.

If any of the above factors are present, then the mediastinum must receive attention from a thoracic surgeon.

Endotracheal intubation

All casualty units ought to have available tables showing the sizes of endotracheal tubes to be used according to age. As a rule of thumb

the following formula can be used for children under the age of 12 years:

$$\text{ETT size} = \frac{\text{Age} + 4}{4}$$

Additionally, the cross-section of the last digit of the right hand can be used as a standard guide. The diameter of the finger represents the outer diameter of the endotracheal tube required.

Intubation is advised in all cases in which the patient ventilates poorly. The intervention must be carried out gently, because injured oedematous vocal cords may delay extubatation. Control the position of the tube in the trachea at all times after intubation with an X-ray of the chest. In children, the endotracheal tube is likely to be inserted too deep and down the right main bronchus. The ideal position is approximately 2 cm above the carina in small children, and 4 cm above the carina in bigger children.

10.4 Blunt abdominal trauma

JHR BECKER & AB VAN AS

Blunt abdominal trauma is one of the most common injuries in today's society. It most frequently occurs during transport (motor vehicle, bicycle, pedestrian accidents), sports and falls.

In this discussion of blunt abdominal trauma, it is accepted that an overall evaluation of the patient, both physically and psychologically, has already been done. The general principles of resuscitation (APLS) apply. An open airway and sufficient intravenous fluid are exceptionally important. The pulse rate, blood pressure and pulse pressure must be determined repeatedly.

The first step is to determine the mechanism of injury, namely:

- What is the nature of the injury? Are there external indicators of underlying pathology, e.g. abrasions, lacerations, contusions or a haematoma?
- In a vehicle accident, was the patient wearing a safety belt?
- In a road accident, was the patient a pedestrian or passenger?

The nature of the injury (as set out above) will lead the practitioner to the underlying pathology. Abrasions over the lower ribs of the left hypochondrium, together with a pale, cold, clammy patient, are extremely suspicious, as they could be indicative of a splenic rupture. A plan of action can then immediately be put in place without the emergency worker having to have great diagnostic skills.

Abdominal organs are divided into five main groups:

1. Parenchymal organs, such as the liver and spleen, that bleed excessively with injury

2. Organs involved in the production, transport and storage of urine
3. Hollow viscera (bowel), excluding the extra-peritoneal duodenum and the extra-peritoneal part of the ascending and descending colons
4. Retroperitoneum: pancreas, major blood vessels, duodenum
5. Diaphragm

The abdomen is approached with reference to the above five systems; in this way you can approach the injury systematically and there is no danger that a system will be overlooked. However, it must be stressed that the above groups are only a scheme for the management of abdominal injuries. The emergency worker must have a similar scheme to divide the whole body into systems, e.g.

- head
- neck
- chest
- abdomen
- vertebral column
- pelvis
- upper limbs
- lower limbs

Parenchymal organs that bleed (liver and spleen)

In the case of an injury with rupture of the normal parenchyma, bleeding must be expected. The level of bleeding is related directly to the severity of the injury. If the injury is mild, the body's haemostatic mechanisms will be able to seal off the injury and it will heal without surgical intervention. The following scheme is handy for placing the injured patient into one of three groups:

- Group for vigilant observation
- Group for vigilant observation with an operation at a later stage if necessary
- Group for immediate emergency surgery

The following scheme (Figure 10.2) is particularly important in patients under the age of 18 years:

Patients are divided into three groups:

■ 1. THE STABLE PATIENT WITH ONLY A HISTORY OF TRAUMA

Liver and/or splenic injury may be present. Vital signs are regularly checked – ultrasound or a CT scan investigation will confirm the presence or absence of a rupture. If a rupture is confirmed, then the patient must be placed in a high-care nursing unit. If the patient remains persistently stable, then surgery is not recommended.

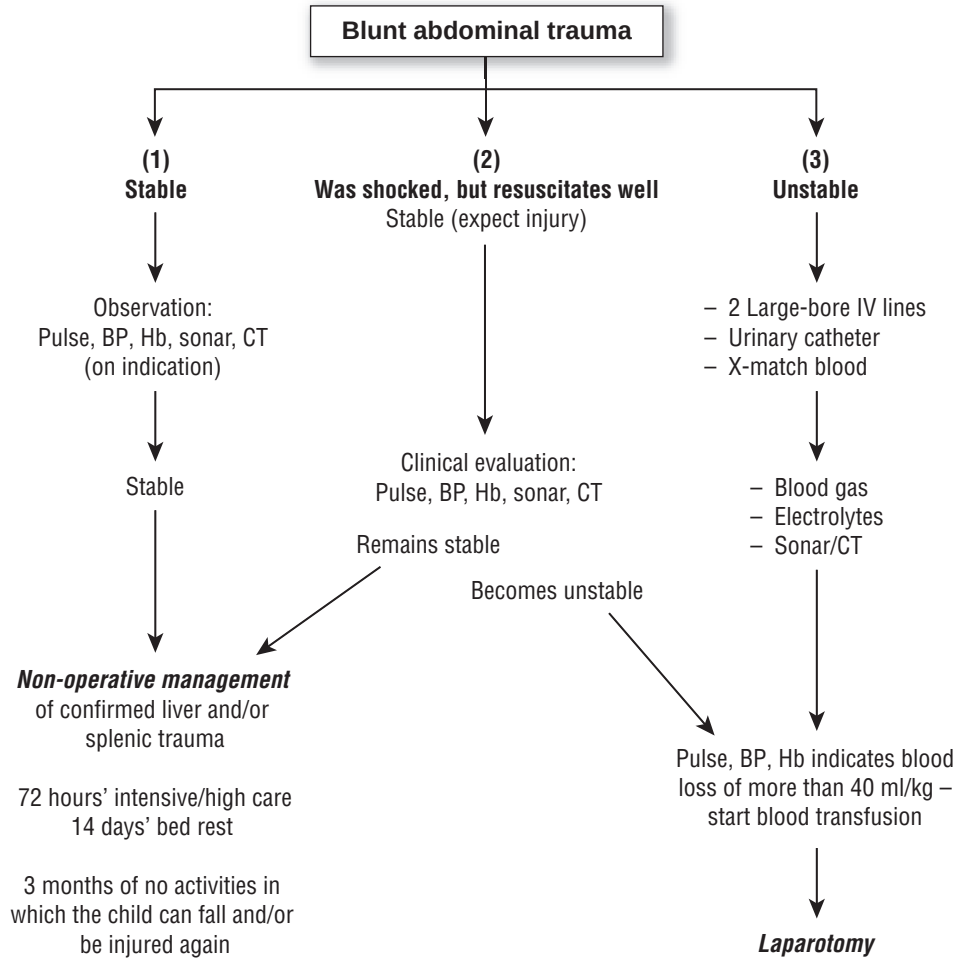


Fig. 10.2 Management of blunt abdominal trauma in children.

■ 2. THE INITIALLY UNSTABLE PATIENT WITH A HISTORY OF BLUNT ABDOMINAL TRAUMA, NOW STABILISED

The patient was shocked but, after administration of intravenous fluid (colloid and/or crystalloid), recovered and the vital signs have returned to normal. While the patient is under continuous observation, ultrasound or a CT scan investigation of the abdomen is done. This will confirm liver and/or splenic rupture. If the patient is stable, a liver and/or splenic rupture is indeed present and there are no other organ injuries that require surgery, then the patient is transferred to high care for constant observation and serial haemoglobin determinations.

The patient may now remain stable and recover without any further surgery or he or she may bleed progressively until as much as 40 ml/kg is needed for resuscitation. Such a patient must undergo a laparotomy to repair the rupture. A patient in this category usually

has an excellent prognosis with a low morbidity and mortality because the injury is repairable.

The patient who qualifies for continuous observation must be managed according to the following regime:

- Intensive/high care observation for 72 hours
- Bed rest in hospital and repeat the ultrasound and/or CT scan in order to monitor the progression of the injury for 2 weeks
- No contact sport or activity that may lead to further injury for 3 months

■ 3. THE UNSTABLE PATIENT WITH A HISTORY OF BLUNT ABDOMINAL INJURY

A patient with a raised pulse rate, low blood pressure, low haematocrit and an unsatisfactory response to sufficient resuscitation must be taken directly to theatre for an emergency laparotomy.

Patients in this group usually have severe injuries which will test the skill of the surgeon. The morbidity and mortality are much higher.

If the necessary infrastructure for high care of the patient is not available, then the surgeon must perform a laparotomy on the patient with a liver and/or splenic injury immediately because these patients may bleed suddenly and massively, and die.

An adult above the age of 18 years should preferably be operated on immediately because even through this group qualifies for vigilant observation (if the facilities are available), 70% of these patients will have to undergo surgery and splenectomy anyway. The older patient with a labile cardiovascular system must be operated on immediately because a laparotomy is better tolerated than a second state of shock plus a laparotomy. Surgery to conserve the spleen is not so important in the adult because overwhelming post-splenectomy sepsis is less common and also splenectomy does not have a high mortality. Splenography, if possible, can be done.

Organs involved in the production, transport and storage of urine

Here the starting point is testing for the presence of microscopic (dipstick) or macroscopic haematuria, irrespective of whether it was present before the traumatic incident or not.

In penetrating trauma it is mandatory to investigate microscopic haematuria, since a penetrating injury to the ureter and/or urethra might not always result in macroscopic haematuria. However, blunt abdominal injury often results in *microscopic* haematuria, without major anatomical damage. In general therefore, only *macroscopic* haematuria should be investigated thoroughly after blunt trauma.

If haematuria is present, then the urinary tract must be radiologically and/or sonographically examined as follows, in this order:

- urethrogram
- cystogram (urethro-cystogram)

- excretory urogram (IVP)
- ultrasound and/or CT scan
- arteriogram

If an injury is identified, appropriate management by the best qualified person, preferably a urologist, is imperative. If anuria or blood is present at the external meatus of the urethra, then the patient must not be catheterised before an urethrogram has been done because of the possibility of a rupture of the urethra.

Hollow viscera (bowel) (excluding extra-peritoneal duodenum)

An injury to the hollow abdominal organs with leakage of contents will cause chemical or bacterial peritonitis. Peritonitis leads to peritonism, a tender abdomen with positive rebound, an increase in the pulse rate with a later rise in temperature and, finally, a very tender, hard abdomen that is painful with movement.

The most common areas of perforation are the proximal jejunum (first 30 cm from the ligament of Treitz) and the last 30 cm of the ileum, since these areas are fixed and likely to be compressed between the anterior abdominal wall and the vertebral column. The injuries are usually small and result from extensive localised pressure.

A ruptured stomach usually results from a 'blow-out' rupture of a full stomach; generally the rupture is large in size and occurs in the vicinity of the greater curvature.

Peritoneal irritation is a very important clinical sign and it is of the utmost importance to identify it at an early stage. The patient must be examined at least 4-hourly by the same doctor to exclude a perforation. Worsening of signs is an indication for a laparotomy. If there are clinical indications for a laparotomy and nothing is found amiss during surgery, it should be regarded as a positive finding that there is nothing wrong and the surgeon should not feel guilty. An erect or supine abdominal X-ray will be helpful only in approximately one-third of children. CT scanning can be of more assistance, especially in experienced hands. The basis for evaluation is regular examination by the same doctor.

Retroperitoneum

■ PANCREAS

The pancreas can be injured in blunt abdominal trauma, particularly when the abdomen is compressed in antero-posterior fashion. The neck of the pancreas is then compressed between the anterior abdominal wall and the vertebral column posterior, and the pancreas can fracture at that level. The key question with pancreatic injuries is whether the major duct is intact or not.

Due to the position of the pancreas in the retroperitoneal space behind the stomach, with the body of the pancreas over the vertebral column, this organ is sometimes very difficult to evaluate.

Therefore special aids must be used, namely:

- ultrasound
- CT scan
- serum and urine amylase which rise with injury

Both ultrasound and a CT scan will indicate a fracture of the pancreas. If this type of injury is demonstrated, conservative management is likely to fail. ERCP is recommended by some experts, but if an injury is shown on the ultrasound or CT scan, this in itself is an indication for surgery. There are various options according to the severity of the injury. Procedures range from simple drainage if only parenchymal injury is present to more complicated resections such as distal pancreatectomy (with or without splenectomy) and even Whipple's procedure (pacreaticoduodenectomy) if the main duct is severed.

■ LARGE BLOOD VESSELS

Retroperitoneal haematoma and large blood vessel injury are never isolated entities, but are usually accompanied by other multiple organ injuries each of which may need surgery in their own right. Retroperitoneal haematoma is classified into:

Pelvic A haematoma limited to the pelvic cavity with the apex of the bladder anterior and the promontorium posterior. Bleeding is usually from the pelvic blood vessels that are torn by pelvic fractures.

Flank This is lateral to the psoas muscles. Bleeding is usually from the kidney or lumbar arteries and veins.

Central This can either be an inferior haematoma over the aorta and inferior vena cava or a superior haematoma over the pancreas, duodenum and superior mesenteric artery.

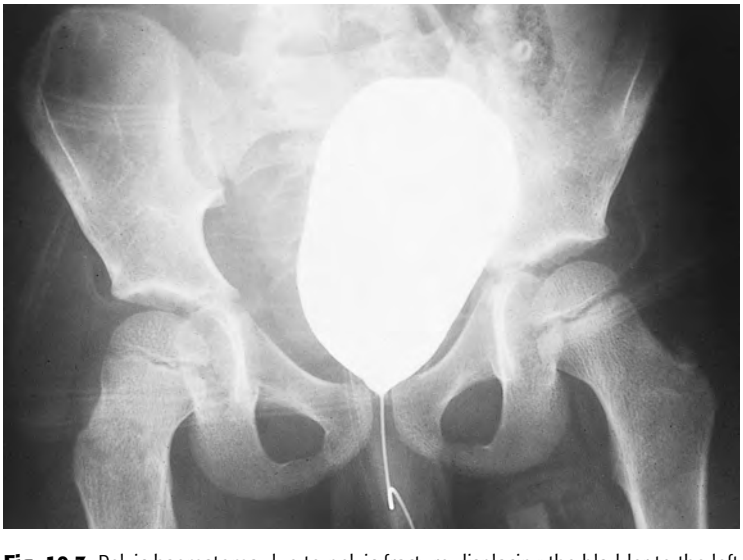


Fig. 10.3 Pelvic haematoma due to pelvic fracture displacing the bladder to the left.

A rupture of the aorta will cause a pulsating retroperitoneal *expanding* haematoma. The patient is shocked and has poor peripheral circulation. These patients need immediate vascular surgery.

The inferior vena cava is seldom injured during blunt abdominal trauma. It is in fact a low-pressure bleed that speedily tamponades unless vena cava thromboses and stasis symptoms are present. A non-expanding haematoma in an otherwise stable patient is best left unopened, unless other organs involved need to be repaired.

Bleeding from the pelvic vessels usually goes with fractures of the pelvis and the bleeding is often very difficult to control. If this is found with an open abdomen, then tying off the internal iliac arteries is recommended.

Duodenum

Duodenal injuries occur in trauma of the right abdomen with superficial abrasions or with injury to the right kidney, pancreas or liver.

Fractures of the 10th and 12th right ribs or of the transverse processes of the 1st and 2nd right lumbar vertebrae increase the chances of duodenal injuries.

Two types of injuries are found:

- duodenal haematoma
- duodenal rupture

Both can be diagnosed with gastrografin-duodenography. A haematoma shows the typical spiral spring appearance and a rupture causes extravasation. There may also be gas around the right kidney – the Freehman Dahl sign. A duodenal haematoma can be treated non-operatively and managed by means of:

- parenteral feeding
- *nil per os* (NPO)
- nasogastric suctioning as required

It usually resolves over 3 weeks. It is sometimes also possible to drain the haematoma endoscopically into the bowel lumen.

Duodenal rupture always requires immediate surgical repair (usually with drainage). Duodenoscopy is not recommended with a perforation.

Diaphragm

The left diaphragm is more commonly involved. This diagnosis should be suspected if bowel sounds are auscultated in the chest.

In these cases it is advised to pass a nasogastric tube immediately since:

- it empties the stomach
- it is also visible in the chest in X-rays of the abdomen and chest

The diagnosis can be further confirmed with diaphragmatic screening or sonar. It is very important that the diagnosis be made, other-

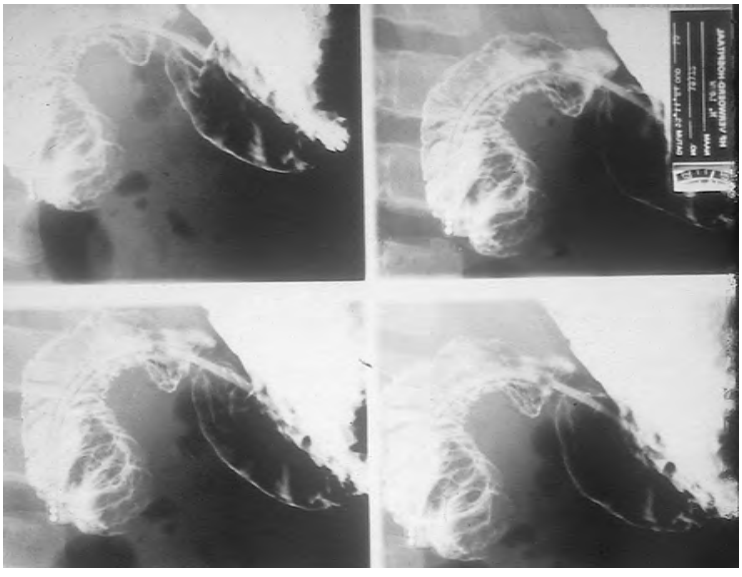


Fig. 10.4 Typical 'coiled spring' appearance of the duodenal mucosa due to a mural haematoma.

wise an underwater drainage tube may be placed in a hollow viscus or the spleen.

A ruptured diaphragm in children occurs according to a different mechanism than a ruptured diaphragm in adults. In adults the diaphragm usually ruptures as a 'blow-out' rupture, typically a large tear in the central (tendinous) part of the diaphragm. In children, however, rupture typically occurs along the chest wall, probably as a result of the elasticity of the chest, and therefore can easily be missed during a laparotomy.

A positive diagnosis means that the diaphragm must be repaired immediately by means of a laparotomy. If the patient with blunt abdominal trauma is approached systematically and sensibly, then few injuries that require active therapy will be overlooked.

10.5 Soft tissue injuries

Skin abrasions

The depth of the injury can be determined in the same way that burn wounds are categorised into degrees. The chances of healing are closely associated with the amount of viable epithelium that is present in the wound.

Emergency management is the same as for burn wounds. Cover the wound with paraffin gauze or Bactrigras® and circular crêpe bandages.

The definitive treatment consists of thorough cleansing of the wound, preferably in theatre under general anaesthesia in order to

remove all debris. In particular, pigment from soil, tar, grass, etc. must be scrubbed off with a soft nail-brush before the wound is again well covered and bound up. If the pigment is not completely removed, permanent 'tattooing' can develop. Deep abrasions will require skin grafts, but superficial wounds will epithelialise. Once the wound has been thoroughly rinsed, it must be covered with paraffin gauze and crêpe bandage similar to the cleaned burn wound in the emergency situation. If the wound is to regenerate and heal on its own, then it must be kept covered and the bandages must only be removed after healing has taken place. If the wound is deep without any epithelial remains, then it will require a skin graft, which should be done as soon as possible.

Lacerations caused by sharp or blunt objects, with or without serious bleeding or tissue loss, must be managed in theatre, with or without general anaesthesia, depending on the circumstances and the extent of the injury. It is often possible to conduct many procedures under local anaesthesia.

Theatre management of wounds

The management of all surface wounds should be according to the following principles:

- Haemostasis
- Wound cleaning of all debris (stones, soil and grass) and dead tissue with running tap water or 0,9% NaCl
- Surgical removal of all non-viable tissue
- Anatomical suturing if the wound is clean
- Delayed suturing after follow-up inspections if the wound is not satisfactory after the first treatment

Nerve, tendon and blood vessel injuries

Nerve, tendon and blood vessel injuries must be referred immediately to a person who is qualified to handle the case. The wound is rinsed with running tap water or 0,9% NaCl and packed with paraffin gauze before the patient is transferred, which must be as soon as possible.

Especially all cases in which there is compromise of the circulation need emergency care since delay can result in unnecessary amputation.

10.6 Orthopaedic injuries

BGP LINDEQUE

Orthopaedic injuries are stabilised and referred for further management to an orthopaedic surgeon after emergency management and wound care have been applied.

10.7 Human and animal bites

JAN J VAN WINGERDEN

Human bite injuries

These occur occasionally in the older child during a fist fight. The flexed metacarpophalangeal joint may be punctured by the opponent's tooth. With full extension of the finger, the wound that extends from skin and fascia through the extensor tendon into the joint is sealed off and the damage is thus underestimated. Primary or delayed tendon rupture may occur.

These wounds are always contaminated. Management is as follows:

- Always clean the wound (irrigate with povidon-iodine and debride).
- Never close the wound primarily.
- Always prescribe an antibiotic.

The infection-causing bacteria are *streptococci*, *staphylococci* and the gram-negative anaerobe *Eikenella corrodens* (Goldstein *et al.* 1983). The antibiotic of choice is penicillin.

Dog bites

Humans bitten by dogs frequently underestimate the potentially unfavourable sequelae.

General guidelines

- All wounds, regardless of age, should preferably be cleansed and debrided in the operating theatre:
  - Irrigate with normal saline solution (0,9%) or Ringer's lactate.
  - Debride wound edges and remove devitalised tissue.
- Do not attempt primary closure in:
  - Wounds older than 8 hours
  - Puncture wounds
  - Wounds anywhere on the body apart from face and scalp.
- Fractures are uncommon but may be found in the child bitten near the orbit, nose or cheek (Tu *et al.* 2002).
- Tetanus prophylaxis should be given if the last vaccination was less than 5 years previously.
- Prophylactic antibiotics should be considered but always prescribed in cases where:
  - Injury was sustained more than 8 hours previously (Cummings 1994)
  - The head, neck or hand is involved
  - Contra-indications to immediate debridement exist
  - Immunodeficiency (HIV, immunosuppresion, etc.), diabetes, splenectomy)

- Potentially harmful bacteria include: *staphylococci*, *streptococci*, *Pasteurella multocida* and *Capnocytophaga canimorsus* (Talan *et al.* 1999).
- Pasteurellosis is the most common dog bite-associated infection; it develops within a few hours and presents as a rapidly spreading red swelling around the wound.
- Capnocytophaga infection is especially dangerous in the immunocompromised patient and presents with headache and muscle ache, followed by the development of a red skin rash and septicaemia.
- The antibiotic of choice (in all cases) is amoxicillin/clavulonic acid (Chait and Spitz 1975; Goldstein *et al.* 1987).
- Exclude the possibility that the dog is rabid. Signs suggestive of rabies are:
  - Excessive salivation
  - Difficulty in swallowing
  - Indiscriminate biting
  - Progressive paralysis
- These dogs should be captured, isolated and if in doubt, euthanised and decapitated for examination of the brain tissue.

CAVEAT

Every second child bitten by a dog will suffer from significant post-traumatic stress disorder – do not underestimate the seriousness of the injury (Peters *et al.* 2004; Morris and Bernstein 2004).

10.8 Hand injuries in children

JAN J VAN WINGERDEN

The hand is frequently injured in the child. The diagnosis and treatment can be difficult – the main responsibility of the primary care physician is to make an accurate diagnosis.

Special considerations

- All penetrating hand injuries should be explored.
- All badly bruised or swollen hands should be radiologically examined.
- If you are going to refer the child, do not cover the wound with antiseptic cream. This applies to all open wounds, including burn wounds. You would be clouding the objective evaluation for your colleague. The optimal referral bandage is moist gauze, soaked in a physiological solution (preferably Ringer's lactate or 0,9% NaCl solution) and covered loosely with a single layer of Gladwrap® to prevent it from drying out, cotton swabs and a loosely wrapped comfortable bandage. The end-result is a thick,

soft covering without excess pressure on the soft tissue and blood supply. The hand should be kept elevated for the full duration of the transport.

- Be aware of the correct transport medium for any amputated parts (see below).

Fingertip injuries

The fingertip, by definition being the portion of the finger distal to the implantation of the flexor and extensor tendons, is the most common hand injury in the younger child (Fetter-Zarzeka and Joseph 2002). Usually the finger is jammed in a door at home. It is extremely painful and, as the 'eye' of the hand, it always warrants special consideration.

Injuries distal to the nail bed usually heal uncomplicated by secondary intention provided the fingertip, as a whole, was not crushed. It should be carefully cleansed and then covered by a thin dressing, such as Opsite® or hydrocolloid, such as Granuflex®. The dressing is left undisturbed for as long as possible and is only replaced if it has been accidentally removed or has become smelly (Mennen and Wiese 1993).

If the nail bed is involved, an underlying fracture should be excluded. When present, both the nail bed and fracture are dealt with operatively. The nail bed is repaired, under magnification, after removal of the remnants of the nail. A spatulated needle and rapidly absorbable thread, 6–0 or thinner, is used. Remember, the fingernail serves as a 'radar' to the finger – good adherence to the nail bed is therefore absolutely necessary.

If the whole nail bed needs replacement, a thick split-thickness nail bed graft is harvested from the big toe.

A closed fracture of the distal phalanx is less common than an open fracture or partial amputation. In both instances, the aim would be to maintain maximum length and to prevent formation of a hook nail (so called '*parrot beaking*'). Soft tissue coverage by means of either a full-thickness skin graft or one or more local flaps follow on to conservative bone debridement whereby only loose fragments are removed to prevent late sequestrae and sharp tips are trimmed.

Traumatic amputations

All traumatic amputations in children must be viewed as potentially revascularisable. Leave the decision to the specialist as to whether it is possible or not.

Absolute contra-indications to replantation are:

- a badly crushed or destroyed finger
- a complete ring avulsion
- a concomitant life-threatening injury

There are two factors that influence a successful end result, namely:

- management of the amputation wound
- management of the amputated finger

■ MANAGEMENT OF THE AMPUTATION WOUND

- Elevate the bleeding limb.
- Dress with sterile, thick, soft compression bandaging.
- Do not tie or clamp off vessels.

■ MANAGEMENT OF THE AMPUTATED FINGER OR PART THEREOF

- Find it, clean it and keep it clean.
- Rinse thoroughly with sterile, physiological solution (e.g. Ringer's lactate).
- Do not perfuse vessels.
- Do not tie off vessels.
- Transport the finger in saline or saline-soaked gauze in a plastic bag on ice (coin bags as used by the bank are ideal). Never place the finger directly on ice.
- Where more than one finger has been amputated, all are transported, each in a separate bag, on ice.

Although the survival rate for transplanted parts is lower in children than in adults in general, two-point discrimination and function of the successfully replanted finger is often better with continuance of longitudinal growth (provided the epiphyseal growth plate was not damaged initially) (Cheng *et al.* 1998).

Flexor tendon injuries

Flexor tendon injuries should be repaired as soon as possible after diagnosis by a surgeon specifically trained in hand surgery (e.g. a plastic or orthopaedic surgeon).

The aim is always to repair whatever is transected. If both flexor digitorum profundus (FDP) and superficialis (FDS) are found transected, both should be repaired. In general, this gives a better result than repairing the FDP alone (Grobbelaar and Hudson 1994). Flexor tendon injuries with concomitant neurovascular injuries tend to give poorer results, regardless of adequate repair of all the structures involved (O'Connell *et al.* 1994).

Tenolysis, i.e. careful, surgical release of all adhesions, is seldom necessary in children. When required in the child less than 10 years of age, the results are not uncommonly disappointing (Birnie and Idler 1995).

Extensor tendon injuries

These occur frequently due to the thin overlying skin cover in the child. Accurate surgical reconstruction is always necessary.

Nerve injuries

The possibility of, and difficulty in diagnosing, a nerve injury in the

child is another important reason for recommending a routine exploration of all penetrated injuries of the hand. Sensation can roughly be tested by allowing the older child to distinguish between a fingernail scratch and a simple touch (children are not tested with pins!). Immersion of the hand of a younger child in lukewarm water for 5 minutes will result in a wrinkled, healthy region – the denervated area, if present, will remain smooth. Peripheral nerve deficits are associated with sympathetic nerve abnormalities, resulting in a dry region in the denervated area. Passing a plastic pen across the suspected and surrounding areas can clinically elicit this sign.

Fractures of the hand

Suspect a closed fracture of the hand or digit in the older child who presents late after a sport-related injury.

The *younger* the child, the more *distal* the fracture (e.g. distal phalanx/fingertip) – the *older* the child, the more *proximal* and ulnar the fracture (e.g. the fifth metacarpal) (Rajesh *et al.* 2001).

Observe whether the hand is swollen, bruised or whether any of the digits are deformed in any way.

Check the *active* range of movement and observe whether each finger touches the same spot at the palmar base of the thumb when flexed; exclude rotation of any of the fingernails of the fully extended hand.

Examine for tenderness over the growth plate or at the joints (ligament attachment) and *passive* range of movement.

Diagnose the fracture accurately. For this purpose, *at least* two clear radiographs of the injured hand and digit are necessary: an antero-posterior and a 'true' lateral view. Radiographs of the other hand are taken for comparison when in doubt, or in the very young child in whom the epiphysial growth plate may not yet be visible.

Exclude involvement of the epiphysial growth plate (incidence 2:5). (Mahabir *et al.* 2001). Apart from the usual potential complications related to all fractures (e.g. angulation, rotation and seldom malunion), growth retardation of a specific digit or post-traumatic arthritis of its joint may be a late result of inadequate treatment of an injury perpendicular through, or compression of, the epiphysis (Torre 1988).

The epiphysial plate is found proximally in each phalanx, as well as in the metacarpal of the thumb and distally in the other four metacarpals. The epiphyseal plate closes at an average age of 16,5 years in boys and 13,5 years in girls.

Refer to a specialist centre:

- when in doubt
- in all open fractures
- in all closed fractures with:
  - **A**ngulation
  - **R**otation
  - (possible) **E**piphysial plate involvement

(These **ARE** the complicated fractures that may require open reduction and Kirschner wire fixation. There is evidence that small, smooth Kirschner wires perpendicular to the epiphyseal plate cause hardly, if any, growth disturbances.)

Treat the other fractures where specialist facilities are not available by strictly adhering to the principles of all fracture management: adequate (closed) reduction and immobilisation. The principles are simple, but carrying out these procedures can be exceedingly difficult because the structures are small, and healing, regardless of the position, is usually rapid. Fingers must never be 'individually' immobilised – the nearest finger helps to prevent angulation and rotation. A functional position, with the metacarpophalangeal joints in 60—70° flexion and 0–15° flexion at the proximal interphalangeal phalangeal joints, is required and should be maintained with a plaster of paris.

Rehabilitate: Start within three to four weeks *irrespective* of what the radiographs show (all K wires are removed by this time).

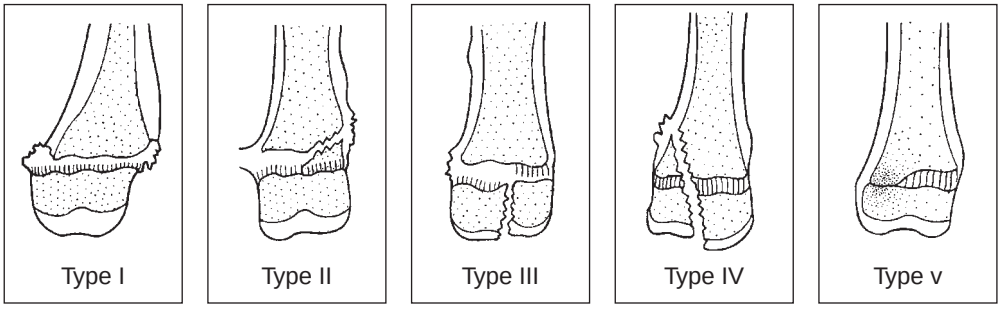


Fig. 10.5 Salter–Harris classification.
Source: Salter and Harris (1963)

Paediatric infections

11

JHR BECKER & E MULLER

It is not the aim of this chapter to educate the reader regarding gram positive, gram negative, aerobic and anaerobic organisms. For this you should consult the various literature sources.

Infection (sepsis) is the successful overriding of the body's defence mechanisms, locally or systemically, by a micro-organism, with consequent local or systemic manifestations that are indicative of an infection.

11.1 Local signs and symptoms of sepsis

- Redness – shiny surface and a change in skin tone
- Swelling
- Heat
- Pain and tenderness
- Loss of function
- Suppuration

11.2 Systemic signs and symptoms of sepsis

- Generalised malaise and apathy
- Fever and shivering
- Loss of appetite, nausea and vomiting
- Warm, clammy skin, dry mouth with halitosis
- Thirst
- Tachycardia
- Later in the disease process: hypotension, delirium, stupor, coma and anuria

The baby's temperature may be rather unstable and it can fluctuate and change by as much as 3°C from normal within minutes.

A dictum never to be forgotten is that cellulitis of a child's limb in an unusual place must be considered to be osteomyelitis until otherwise proven. Radiological signs will be absent in the early stages and the only diagnostic test (provided the facility is available) is a radioisotope study. The bone scintigram will undoubtedly indicate whether the infection is located in the bone or the soft tissue.

If osteomyelitis is present, then an orthopaedic surgeon should immediately relieve the tension in the bone by means of boreholes or else permanent damage to the bone or epiphyseal growth plates could follow.

If there is doubt about the diagnosis, then the patient should be referred to an orthopaedic surgeon to make the diagnosis.

11.3 Septicaemia

Small, local infections in the newborn, e.g. of the umbilicus, skin, nose, ear or bladder, can lead to septicaemia which causes a rapid deterioration in the patient's status.

All babies and children should therefore be thoroughly examined from head to toe – examine the ears, nose, throat, skin, urine, central nervous system and any other imaginable place in the case of a possible septicaemia or bouts of fever.

DICTUM

Septicaemia must be considered in all cases in which babies suddenly develop circulatory collapse, a fluctuating temperature, jaundice, abdominal distension or coma.

Emergency treatment for septicaemia

- Administer an intravenous infusion:
  - Administer Ringer's lactate or 0,9% NaCl at 20 ml/kg as a bolus.
  - Re-evaluate the perfusion and give more fluid as necessary.
  - Requirements up to 60 ml/kg in the first hour are not unusual.
  - Successful resuscitation is indicated by a urine output of >1 ml/kg/h, normal mental status, normal blood pressure and pulse, and capillary refill of <2 seconds.
  - During rapid fluid resuscitation, constant monitoring of the patient for rales, hepatomegaly and increased work of breathing is required. Avoid fluid overloading.
- Ventilation – ensure an open airway and give oxygen.
- Do blood cultures (prior to administering any antibiotics) for culture and sensitivity.
- Take a urine sample from a catheter and test for culture and sensitivity.

- Do a lumbar puncture and CSF for culture and sensitivity.
- Begin immediately with intravenous administration of a broad spectrum antibiotic.
- If the source of sepsis is localised, e.g. an abscess or osteomyelitis, it must be treated immediately.

Phases of the infection

- Cellulitis – redness, throbbing pain, swelling
- Suppuration – liquefaction of the content, clinical fluctuation, surface may appear yellowish
- Draining

11.4 Abscesses

An abscess is a collection of pus with a surrounding inflammatory reaction. The body has therefore succeeded in satisfactorily localising the septic focus.

If the abscess is allowed to follow its natural course, the pus will dissect to the surface and drain itself, but usually with damage to the surrounding organs.

DICTUM

As soon as pus has formed, in other words when the mass fluctuates or suppuration can be confirmed clinically or by special investigations (ultrasound or CT scan), then it must be surgically drained.

If the infection is in the cellulitis phase – red, swollen and painful – there is no point in surgical intervention because this will merely cause bleeding without drainage of pus. Antibiotics can be given or topical agents that will penetrate the cellulites, e.g. Bactroban® topical or Fucidin® ointment.

Surgical drainage of an abscess

- The area is sterilised under general or local anaesthesia.
- Facilities to take pus swabs for aerobic and anaerobic cultures and sensitivity testing must be available.
- The surgical incision is made in the lines of Langer – no saucerisation of the abscess.
- The abscess cavity is cleared of all pus (samples taken) and non-communicating septae in the cavity are broken down.
- The cavity is rinsed with 0,9% NaCl.
- A light, thin foreign body (latex drainage tube or a piece of surgical gauze) is placed between the skin edges of the wound, because:
  - it ensures that the cavity continues to drain and collapses
  - it prevents the skin edges from adhering before the abscess cavity has completely disappeared.

- The cavity must never be plugged because this is painful and the aim of drainage is defeated.
- The gauze drainage must be replaced daily and the wound must be rinsed with an antiseptic solution or by gentle irrigation with running tap water.
- The administration of a systemic antibiotic in the management of abscesses is a controversial issue.

It is recommended that antibiotics be administered in the following cases:

- as prophylaxis in patients with heart valve lesions
- in immune-compromised patients
- if systemic signs of bacteraemia or septicaemia are present
- for patients after a splenectomy
- if severe surrounding cellulitis is present

In practice, the patient usually feels better immediately after the pus has been drained and then antibiotics are unnecessary.

In layman's terms, an abscess can be compared with a pimple of which most people have had experience. All the above-mentioned principles apply to a pimple which is in fact also an abscess – the size differs, but the characteristics are:

- It is a painful red swelling.
- It will not drain before it shows yellow.
- If it is yellow, it drains easily.
- Healing follows shortly after drainage.
- Scarring occurs if drainage is incorrectly carried out.

Types of antibiotics to be administered with abscesses

The big question that must always be asked is: Which organism is most likely to be present?

The commonest organism present with abscesses is *Staphylococcus aureus*. According to modern rules, penicillin is no longer used in the management of these organisms.

The choice of antibiotic is:

- cloxacillin
- erythromycin
- trimethoprim
- a first-generation cephalosporin

If the abscess is located in the lower half of the body and a mixed infection is suspected – gram positive, gram negative, aerobic or anaerobic – then switch to the triple regimen of:

Aminoglycoside
Metronidazole
A penicillin, e.g. ampicillin, or
A first-generation cephalosporin
} Administered simultaneously

Mixed infections with gas formation and gangrene must be managed aggressively and invasively. The patient is immediately referred to a surgeon who will operate invasively and provide supportive therapy in a high-care unit. These infections spread very quickly and the patient may rapidly become toxic and die.

11.5 Carriers of Staphylococcus aureus

There is probably a source from where repeated re-infection takes place in children who develop carbuncles, pimples and abscesses.

The commonest site is the nose and nasopharynx. Children scratch in their noses and spread the organisms with their hands to the rest of the skin. Nasal secretions may also be swallowed, which may cause peri-anal and gluteal pimples.

It is important that the nose itself also be 'sterilised' in the management of the abscess. The products presently available are:

- Mupirosin® nasal ointment (not the topical ointment Bactroban®, Beecham, S4)
- Fucidin® cream (Leo, S4) (not the topical ointment)

A small amount of either of the above products is applied twice a day in each nostril.

INCIDENTALLY

In veterinary practice, anti-flea or anti-tick poison ('pour on' or 'spot on') is only applied to the back or neck of an animal; it then spreads over the entire body to keep the animal free of ticks and fleas. In the same manner, the entire body can be infested with organisms as a result of a single abscess. Elective surgery cannot be conducted if there is a source of pus elsewhere on the body.

11.6 Erysipelas

This is usually caused by the beta-haemolytic *Streptococci*. The organism has enzymes (hyaluronidase and streptokinase) which assist the spread of the infection. The skin is red and oedematous. There is a demarcated edge between the infected and non-infected skin, and pain and fever are present. The patient is actually less ill and the area is less swollen and tender than in the case of a soft tissue cellulitis of the same extent.

Management

- Penicillin per mouth or intravenously
- Bed rest

There must be a dramatic change within 24 hours or else the diagnosis must be reconsidered or the antibiotics must be changed.

11.7 Mastitis

The neonatal breast tissue is exposed to high levels of maternal oestrogen and progesterone, with consequent breast development and even post-partum milk secretion (so-called 'witch's milk'). The breasts may become infected post-partum with resulting mastitis and abscess formation. Mastitis is treated successfully in the vast majority of patients with a broad spectrum antibiotic, e.g. erythromycin or cloxacillin. If an abscess develops, it should be drained immediately using a submammary incision so that the young breast tissue does not suffer too much damage. If the diagnosis is doubtful, prior sonar investigation or needle aspiration helps to avoid unnecessary (and potentially harmful) surgical exploration.

11.8 Gynaecomastia

This condition is discussed here because it must be distinguished from a breast tumour or breast abscess. In the prepubertal period it is normal for breast tissue to become palpable and tender in boys and girls. The breasts may even enlarge unilaterally. This is not an abscess, neither is it a benign or malignant breast tumour. While special investigations are not necessary, a thorough general examination should be done and a good medical history taken because it is important to bear in mind rare hormone-producing conditions such as:

- hypophysis tumours
- testis tumours
- liver diseases
- medication, drugs or alcohol

The patient and the parents should be reassured and the patient must undergo regular follow-up examinations to ensure that an important factor has not been overlooked. The breast must not be drained, neither should a biopsy be taken, as this will lead to permanent damage.

Needle aspiration cytology is recommended if the doctor is concerned and the 'tumour' feels suspicious.

11.9 Gluteal abscesses and pimples

The most common form arises as a result of complications with nappy rash. The abscess is dealt with in a routine manner as discussed above. The rash must also be treated in its own right.

Such abscesses usually originate from *Staphylococci* rather than gram negative organisms – remember the nasal carrier.

11.10 Peri-anal abscesses

Peri-anal abscesses are not as common in children as in adults, but they do occur. The abscess may merely be a pimple or a boil located next to the anus. This is then also treated as such.

NB: Remember the nose as a source of infection. The nose is the carrier – organisms are swallowed (*S. aureus*) and this causes little abscesses in the gluteal region. Sterilise the nose with Mupirosin® nasal ointment (Beecham, S4) or Fucidin® cream (Leo, S4).

It is not unusual to find that the abscess, as in the adult case, has given rise to fistula formation – this must be completely excised.

There is also a possibility that the fistula may be a congenital developmental abnormality with a secondary infection. These fistulae must be managed by an expert, preferably a paediatric surgeon.

11.11 Nappy rash

This is a rash in the region that is covered by a nappy and develops as a result of long exposure of the skin to a combination of stool and urine. The soaps, soap powders and fabric softeners in which nappies are washed are contributing factors. Often the cause is that the nappies have not been sufficiently rinsed to get rid of all the soap.

The very irritated, tender skin is now exposed to secondary infections by organisms such as S*taphylococci* (if there are pustules) and *Candida* (if there is simply a red, painful, infected, moist skin).

Treatment

- Remove the irritating contact of excreta and urine from the skin:
  - Wash the skin by hand or with cotton wool after each nappy change.
  - The water must be at body temperature.
  - The soap that should be used is Elizabeth Anne baby shampoo and it should be applied gently.
  - Rinse the skin and wash away all remnants of urine, excreta and soap.
  - Pat (do not rub) the skin dry with a soft hand towel.
- Now create a barrier between the irritant (excreta and urine) and the skin. Smooth a barrier cream, e.g. Fissan®, Penaten® or zinc oxide, onto the clean, prepared skin.

It is absolutely useless to apply the cream on top of excreta which is simply wiped away with the nappy.

If a secondary infection is present, then Mupirosin® topical ointment (Beecham, S4) or Fucidin® ointment (Leo, S4) must be used for abscesses and Mycostatin® ointment (Squibb, S2) for *Candida* infection. The latter two ointments are applied first before the barrier cream.

Abdominal pain

12

JHR BECKER & SM LE GRANGE

Abdominal pain is a reality for a child – a child will seldom pretend to have pain, especially if the pain limits his or her normal activities. Caretakers often fear that abdominal pain may be a symptom of acute appendicitis. If you experience problems in determining the cause of the pain, ask the following questions:

- How urgently do you need to take action?
- Which special investigations should be conducted?
- Is the child often absent from school due to recurrent abdominal pain?

12.1 Differential diagnosis

- Acute appendicitis
- Primary peritonitis
- Mesenteric adenitis
- Gynaecological conditions
- Intussusception
- Malrotation of the bowel
- Constipation and faecal impaction
- Extra-abdominal pathology

The pain is of importance in the following cases:

- Severe, recurrent pain which causes the child to cry
- Pain that wakes the child up at night
- Pain that limits favourite activities
- Pain associated with vomiting
- Colicky pain, with or without associated:
  - bowel habit changes

  - blood per rectum
  - mucus per rectum
- Pain with
  - fever
  - pallor
  - confinement to the bed
- Pain associated with the following physical signs
  - abdominal tenderness
  - abdominal muscle rigidity
  - palpable mass
  - positive rebound sign

Babies and neonates are particularly difficult to examine, therefore the medical history is of special importance. Note, e.g., whether there are:

- crying attacks
- drawing up of legs
- abdominal examination has to be deferred

The medical practitioner will require all his or her skills, and at times even a second opinion, to make a diagnosis.

12.2 Acute appendicitis

Quotations on appendicitis

- "The appendix is like a lion: It does not do as it is told, it does as it wishes."
- "At the apex of all intra-abdominal calamities stands the appendix."

These quotations arise from many years' experience of this diagnosis being rather problematic.

Definition

Inflammatory disease of the appendix that may result in a variety of clinical pictures, depending on the severity, as well as the underlying condition of the patient. Variables such as the position of the appendix may also play a role.

Pathophysiology

The lumen of the appendix has a small volume of 1–2 ml. Any obstruction of the lumen, e.g.

- lymphoid swelling
- faecolith
- foreign body
- worms

will allow the normal secretion in the lumen to accumulate. The mucus-secreting glands of the appendix are able to secrete against a pressure gradient. This secretion, along with the multiplication of enteral organisms, increases the luminal pressure, which causes venous obstruction and further congestion. The pressure rises above capillary and even arterial pressure, leading to arterial insufficiency and consequent necrosis with perforation.

Appendicitis follows a seasonal pattern – the incidence rises during changes in season, when more cases of upper respiratory tract infections are reported. The presence of a systemic infection causes generalised lymphoid hyperplasia, also in the appendix, which may result in appendicitis.

Symptoms and signs

Appendicitis has symptoms and signs pointing to the following:

- gastro-intestinal upset
- abdominal pain
- an inflammatory process

■ SYMPTOMS

- Gastrointestinal symptoms:
  - *Anorexia* is the most consistent complaint in appendicitis. *Nausea* is usually present and the patient may *vomit*. Some of these patients complain of *constipation* and some even of *diarrhoea*.
- Abdominal pain:
  - If they are old enough, these patients may give the classic description of a pain that started somewhere around the *umbilicus* and later located as a sharp pain in the *right fossa iliaca (RFI)*. (If only the appendix is inflamed, somatic pain becomes a peritonitic pain when the overlying peritoneum gets involved.)
- Inflammatory process:
  - A caretaker or older child may complain of *fever*. If *rigors* are present, this usually indicates that perforation and/or abscess formation have already taken place.

■ SIGNS

- General examination (inflammatory process):
  - *Tachycardia* is the most consistent sign of appendicitis. A low-grade *fever* (38,5 °C) is often present. If a patient has a temperature of 39 °C or more, the diagnosis must be re-considered. A viraemia is far more likely, but complicated appendicitis (abscess or peritonitis) may present with a high fever.
- Abdominal examination:
  - Asymmetry due to a mass may in some instances be visible. On light touch, *tenderness* in the RFI is elicited. The presence of a

palpable *mass* indicates either an appendiceal mass or an abscess. Tenderness in the RFI on deep touch in the left fossa iliaca (LFI), is called the *Rovsing sign*. *Diffuse tenderness*, *muscle rigidity* and a positive *rebound sign* are associated with general peritonitis following perforation and free pus in the abdomen. A rebound sign may be difficult to elicit in children. Try to distract their attention by talking about things at school. It is sometimes helpful to ask them to jump or cough.

A rectal examination is not indicated in children because they tolerate it so poorly and the yield as regards finding more signs than with a well-performed abdominal examination is rather small.

Differential diagnosis

The differential diagnosis may be found in:

- Other gastro-intestinal organs:
  - distal ileitis (Crohn's disease)
  - Meckel diverticulum
  - cholecystitis (rare in children)
- Extra gastro-intestinal, intra-abdominal organs:
  - urinary tract infection
  - torsion of ovarian cyst
  - primary peritonitis
- Extra-abdominal organs:
  - right lower lobe pneumonia

These conditions should be ruled out by way of thorough physical examination, side-room investigation of the urine, as well as selected special investigations. Remember that children with a febrile disease of any origin may present with proteinuria. Likewise, they may present with haematuria due to an inflamed appendix in close proximity to the ureter.

Special investigations

Classic appendicitis is a clinical diagnosis. In these instances no special investigations are probably necessary. This is unfortunately not always the case and in an uncomfortable percentage of cases the clinician is not immediately certain of the diagnosis. A few special investigations may be helpful:

- Full blood count:
  - A low-grade *white cell response* (15×10^9) may be helpful; like temperature, a very high white cell count usually points to a viraemia (a differential count may help) or a pneumonia, etc., or, if it is very high, even to leukaemia
- C-reactive protein (CRP) or procalcitonin:
  - Both are indicators of an inflammatory process. CRP is rather

non-specific for any form of inflammation, but procalcitonin is more specific for an inflammatory process associated with an infection

- Abdomen X-ray:
  - If a faecolith is visible on an abdominal X-ray, it may be regarded as pathognomonic for appendicitis. Other signs to look for include: a mass in the RFI, signs of small bowel obstruction, wiped-out right psoas shadow, and scoliosis to the right
- Ultrasound:
  - Very helpful in skilled hands
- Computerised tomography or magnetic resonance:
  - May be helpful in really challenging cases

Natural history

Central peri-umbilical pain is soon followed by nausea and vomiting. As the pain increases, it shifts to the RFI where it remains, eliciting local symptoms and signs. If left untreated, the local pathology will gradually increase, leading to:

- appendix mass
- appendix abscess
- perforation
- general peritonitis
- death

Management

The management of acute appendicitis rests on four 'legs':

- fluid resuscitation
- intravenous antibiotics
- proper analgesics
- surgery

These patients are dehydrated to different extents. Due to their gastro-intestinal symptoms, as well as in preparation for surgery, they should be kept *nil per os*. A proper intravenous catheter should be placed and maintenance fluid should be calculated and administered. Resuscitation fluid should be given until an acceptable urinary output is obtained.

Due to the mixed enteral organism aetiology, intravenous antibiotics covering gram positive organisms (*Streptococcus faecalis*), gram negative organisms (*E. coli*, *Klebsiella pneumonia*, etc.) and anaerobic organisms (*Bacteroides* spp.) should be commenced and continued until the diagnosis has been proved wrong. If the diagnosis is unlikely, but the clinician prefers to admit the patient for observation, antibiotics may be omitted until the diagnosis becomes certain.

The view that analgesics will mask peritonitis is not only wrong, but also cruel towards the patient. Throughout the period set aside for either resuscitation or observation, proper analgesia (IV morphine infusion) should be given. (Please note that intramuscular injections are unacceptable in children.)

An appendectomy should be performed by a skilled person when the diagnosis is certain and the child is properly resuscitated.

Subsets of appendicitis

■ APPENDIX MASS

If an inflammatory mass has formed, but no pus is present (usually after about 3 days' history of disease), the fever is still not very high and no rigors are present, the patient may be treated conservatively with IV antibiotics (Osner-Scherin regimen). The pre-requisites are as follows:

- The diagnosis must be confirmed (pus excluded by ultrasound).
- The responsible clinician must re-evaluate the patient at least 6-hourly.
- The tachycardia and fever should respond within 12 hours.
- The mass must become smaller in size.
- If the above do not occur, the patient should be taken to the operating theatre.

The regimen consists of the following:

- *Nil per os* (NPO)
- IV infusion (maintenance, as well as resuscitation)
- IV antibiotics (as pointed out above)
- Pain management (as outlined above)

■ ABNORMAL LOCATION OF THE APPENDIX

The appendix can be in different anatomical positions. For this reason the pain is not necessarily restricted to the RFI but will occur in the area where the appendix resides. Examples are given below:

- Pelvic appendix:
  - This can be confused with gynaecological, urological and rectal conditions.
- Retrocaecal appendix:
  - The symptoms can be very similar to those of kidney or vertebral conditions.
- Upper abdominal appendix:
  - The appendix is located in the upper abdomen as a result of malrotation of the mid-gut and, due to this position, may imitate inflammatory conditions of the gall bladder or pancreas.
- Left-sided appendix:
  - With *situs inversus*, in which the abdominal organs lie on the reverse side, the appendix will be found in the LFI.

Appendicitis in the neonate, baby and nursery school child

- Neonates:
  - When there appear to be signs of appendicitis in the neonate, always bear in mind the possibility of enterocolitis or associated Hirschprung's disease or necrotising enterocolitis.
- Babies and toddlers:
  - In babies and toddlers, the diagnosis can be difficult due to communication problems, as well as atypical presentation. As a result, in 50% of cases less than 5 years of age, the appendix has already perforated when management starts. Therefore, exercise extreme caution with patients in this age group.

The 'white' appendix ('lily white')

This term refers to an appendectomy of an uninflamed appendix. In other words, the cause of all the symptoms and signs indicating an appendectomy was not appendicitis, but something else. At laparotomy, a limited inspection of the abdominal cavity should be carried out to determine other possibilities, e.g.

- Meckel's diverticulum
- gynaecological conditions
- mesenteric adenitis
- Crohn's disease
- *Yersinia ileitis*
- free pus from elsewhere

If no other cause can be found, the appendix should probably still be removed due to the possibility of a carcinoid appendix causing the symptoms. Sometimes one is also surprised by the histologist with reports of parasitic ova. It is therefore imperative that all appendices should be sent for histology. If, however, other pathology can be found, that should be addressed and the appendix should be left in situ.

The old assumption that a surgeon should have a 10% lily white rate is incorrect. Appendectomies should be performed only if the clinician is certain beyond reasonable doubt of the diagnosis. There are enough aids available to the clinician to avoid removing white appendices.

Classification

An intra-operative classification to aid in the management of the patient after appendectomy has been proposed:

Grade 1: erithematous
Grade 2: exudative
Grade 3: purulent
Grade 4: necrotic

The antibiotics of a patient with Grade 1 or 2 disease may probably be stopped post-operatively and he or she may be discharged on day 1. Patients with Grade 3 or 4 disease, however, will most likely need a longer course of antibiotics, will suffer from a period of ileus and have a higher incidence of wound sepsis.

Prognosis

The prognosis of acute appendicitis should be very good and the morbidity due to the operation should, especially in the event of laparoscopic surgery, be very low. Unfortunately, we still see many patients with Grade 3 and 4 disease in whom not only does the morbidity rise, but even mortality may occur.

12.3 Primary peritonitis

Definition

This is a condition characterised by pus in the abdomen with all the symptoms and signs of peritonitis, without a clear cause. It is more prevalent among girls than boys. The organism present is often *Streptococcus pneumoniae*, but other organisms have also been isolated. The diagnosis is usually made at laparotomy when it is found that a mistaken diagnosis has been made, e.g. a suspected appendicitis. Only one organism is cultured with primary peritonitis. Multiple organisms are usually involved in cases of secondary peritonitis (polymicrobial).

Pathophysiology

There is a list of suggested postulates as to how the organisms reach the abdominal cavity:

- haematological spread
- urinary tract
- fallopian tubes
- lymphatic spread
- micro-perforation

However, none of these mechanisms has been proved. Peritonitis does not develop from a bacteraemia – the urine is normally clear and there is usually no gynaecological pathology.

Symptoms and signs

These children present with peritonitis, which is often difficult to differentiate from acute appendicitis. However, they are normally girls, they usually do not have any gastro-intestinal complaints, and the level of tenderness often does not fit the rest of the general disease (they are often not very sick).

Management

- Resuscitate with fluid and electrolytes.
- Remember the diagnosis in atypical cases, especially girls.
- Try to rule out appendicitis with ultrasound or even a CT scan.
- If a laparotomy is not done, then:
  - take a pus swab
  - look for any pathology
  - do not remove the appendix unnecessarily
  - do an abdominal lavage (rinse copiously with 0,9% NaCl)
  - close the abdomen
  - give a broad spectrum antibiotic, covering gram positive organisms – adjust according to culture

12.4 Mesenteric adenitis

Definition

The mesenteric lymph nodes are enlarged due to viraemia, often associated with an upper respiratory tract infection.

Symptoms and signs

These children present with either a current or recent upper airway infection and vague abdominal complaints. The tenderness is usually diffuse, but not serious enough to suspect a perforated appendix. Only on very rare occasions will they present with a positive rebound sign. They do not have any gastro-intestinal complaints.

Management

Every effort should be made not to take these patients unnecessarily to theatre, because they suffer from airway infection and may develop bronchospasm with an anaesthetic.

12.5 Gynaecological conditions

In a sexually inactive patient group, inflammatory gynaecological problems seldom present a problem. One should, however, keep in mind possibilities such as a foreign body inserted into the vagina with consequent infection.

Remember that girls may start menstruating by the age of 11–12 years, but they seldom ovulate before the age of 15–16 years.

Other gynaecological factors that may cause abdominal symptoms and signs are:

- rupture of an ovarian cyst or follicle
- ovarian or adnexal torsion

The diagnosis of a gynaecological cause for abdominal pain is usually made with ultrasound or, sometimes, retrospectively after a laparotomy.

12.6 Intussusception

Definition

This is a condition where the proximal bowel moves into the distal bowel, like an inverted sock. The proximal bowel is called the intussusceptum and the distal or receiver bowel the intussuscipiens.

This phenomenon follows conditions in the bowel wall causing a so-called 'lead point', which is pulled into the lumen by peristalsis. Intussusception may be ileo-ileal, ileo-colonic (the usual presentation) or colo-colonic. In rare instances the intussusceptum may protrude through the anus, mimicking a rectal prolapse.

Incidence

This is a common condition in healthy, well-fed and non-immune-compromised children. There are two incidence peaks:

- Age 5–9 months
- Age 3–4 years

Pathophysiology

In the first age group, enlarged bowel lymphoid tissue (Peyer's plaques), following a viral infection (usually Rota virus), is believed to pull the bowel into an intussusception.

In the older group, a definitive lead point can usually be found:

- Meckel's diverticulum (80% of cases)
- polyps
- lymphoma
- duplication cysts
- worms or other foreign bodies

If left untreated, complete small bowel obstruction may develop and the risk of necrosis and perforation arises.

Symptoms and signs

■ SYMPTOMS

The patient is usually a *well-fed* baby with a recent upper respiratory tract infection or gastro-enteritis and abdominal *cramps*. These patients classically pass a *bloody*, *slimy stool* ('red current jelly' stool). The patient may *vomit* bile.

■ SIGNS

In between attacks of abdominal cramps, the patient may be calm

and easy to examine. Subtle signs of *dehydration* may be visible early on. The abdomen is *soft* and *non-tender* in the uncomplicated phase and a *sausage-shaped mass* can often be palpated in the right flank. After perforation has set in, the patient will present with peritonitis.

Special investigations

- Full blood count:
  - white cell response will set in after perforation
- Biochemistry:
  - high urea and creatinine point at dehydration
  - electrolyte disturbances, in line with vomiting
- Abdominal X-ray:
  - small bowel obstruction
  - mass effect in right flank
  - no air in colon
  - 'tram line' sign
  - free air if perforation is present
- Ultrasound:
  - imaging study of choice
  - 'pseudo-kidney' sign
  - 'target' sign

Management

- *Nil per os* (NPO)
- Nasogastric tube
- IV infusion with maintenance, as well as rehydration fluid
- Analgesics (antispasmodic is a good idea)
- Antibiotics (treat all children with bowel obstruction with broad spectrum antibiotics to counteract the possibility of translocation)

Pneumatic reduction

■ PRE-REQUISITES:

- The patient must be fully resuscitated.
- The abdomen must be soft (no peritonitis).
- There may not be any free air on the abdominal X-ray.
- The operating theatre must be ready to start a laparotomy immediately if it becomes necessary.

■ TECHNIQUE:

- This procedure should be performed by a skilled paediatric radiologist.
- The surgeon must be in the radiology suite during the procedure.

- If a perforation occurs during the trial of pneumatic reduction, large-bore needles should be inserted into the abdominal cavity immediately and the child taken to theatre without delay.

■ POST-PNEUMATIC REDUCTION:

- Start feeds when the patient is awake – if tolerated well, discharge the patient the next morning.
- Toddlers should be examined on an elective basis for a lead point.

Laparotomy

- Indications:
  - peritonitis or free air on X-ray (perforation has already occurred)
  - failed pneumatic reduction (usually means the bowel is necrotic)
- Operation:
  - manual reduction may be tried, if the bowel is not necrotic
  - *en masse* resection of necrotic bowel with primary anastomosis

12.7 Malrotation of the bowel

The bowel of the embryo was originally partially outside the abdominal cavity. It is a long tube that is proximal at the foregut and distal at the hindgut in the abdomen, with the rest outside in a membrane-covered cavity. The digestive tract undergoes gradual rotation and return of the content to the abdominal cavity; this process can be artificially divided into three phases.

First phase of rotation

The liver, due to its weight, comes to rest in the right hypochondrium and a 90° rotation takes place.

Second phase of rotation

- The mid-gut returns to the abdominal cavity.
- The duodenum moves in behind the superior mesenterial artery.
- The small bowel comes to rest in the right middle abdomen and from here, as a result of the volume, the hindgut is displaced to the left.
- The caecum and right colon return to the abdominal cavity last and come to rest anterior to all the loops of small bowel, but are at this stage located in the middle upper abdomen.

Third phase of rotation

- The abdominal caecum moves from the epigastrium (where it stopped at the end of the second phase of rotation) down to the right fossa iliaca (RFI).
- Fixation of the duodenum, with the right and left colon against the posterior abdominal wall.

The above is a brief explanation, but it is essential to know how the actions proceed so that the associated problems can be understood, e.g.

- The viscera remain outside and the umbilicus does not close as in exomphalos.
- If the third phase is not completed, the caecum remains in the middle abdomen and the bands that are intended to fix the caecum in the RFI now stretch over the duodenum up to the abnormally positioned caecum. These bands (so-called 'Ladd bands') may cause duodenal obstruction.
- Insufficient fixation in the third phase of rotation causes a loose and mobile bowel – especially the midgut which may then undergo a volvulus (twisting itself around the stalk of the superior mesenteric artery) with consequent gangrene.

Ladd bands

These are congenital bands that run over the duodenum to the caecum and are located in the middle abdomen in malrotation. These bands can press the lumen of the duodenum closed, completely or incompletely.

Babies with this condition show signs of an incomplete to complete obstruction, with subsequent:

- feeding problems
- vomiting of bile
- failure to thrive

■ INVESTIGATIONS

- If the obstruction is incomplete, then a diluted barium meal is recommended – this will indicate the site of the incomplete obstruction in the duodenum.
- If the obstruction is more complete, then a barium enema will indicate the caecum in an abnormal position in the upper abdomen.

■ MANAGEMENT

An operation is always recommended when the diagnosis of a malrotation with symptoms is made. The so-called 'Ladd operation' is done.

Poor fixation of hollow viscera to the posterior abdominal wall

In this case, the third stage of rotation is incomplete. The caecum is located in the mid-abdomen and therefore not fixed in the RFI. The result is a very mobile mid-gut and a very small so-called 'duodenocolonic isthmus'. This is the reason why volvulus around the superior mesenteric artery occurs easily. The volvulus may be intermittent with associated attacks of pain that warrant further investigations.

All children who have attacks of acute abdominal pain associated with sweating, fainting and pallor, and children who are doubled up with pain should have a barium enema in order to exclude a malrotation and poor fixation.

The reason for this drastic investigation is that malrotation and poor fixation can cause midgut volvulus, possible gangrene and a non-viable short bowel syndrome.

All cases of malrotation in which the above-mentioned attacks of pain are experienced must be operated on. Many children have recurrent attacks of severe abdominal pain, but only once all organic pathology has been excluded, e.g. appendicitis, intussusception and malrotation, may the child be managed conservatively.

Constipation

13

JHR BECKER & M KIRSTEN

13.1 Constipation

Patients' and physicians' concepts of constipation differ in most cases. There are great variations in normal bowel patterns and a thorough history is essential to make the diagnosis. The history must include details concerning frequency and consistency of faeces.

Definition

- Infrequent passage of hard stools (less than 3 times a week)
- Difficulty in passing stools/painful bowel movement
- Rectal impaction/abdominal mass

Remember that constipation is a symptom and not an illness.

Aetiology

Constipation is multifactoral in the majority of cases and often referred to as chronic functional or idiopathic constipation.

There is no connection between constipation and/or encopresis and IQ or social standing, but there can be a genetic predisposition. Many children's history goes back to the first 6 months of life.

■ SEVERE SLOWING OF COLONIC TRANSIT

- Metabolic disturbances (hypothyroidism, renal tubular acidosis)
- Electrolyte disturbance
- Side-effects of medication
- Mental retardation

- Neurological disturbances (spina bifida, hypotonia)
- Congenital aganglionosis (Hirschsprung's disease)

■ DISORDERS OF ANORECTAL FUNCTION

- Anal stenosis
- Anorectal malformation
- Failure of internal sphincter relaxation (Hirschsprung's disease)
- Voluntary inhibition of defecation
- Painful disorders: fissures, dermatitis
- Idiopathic mega-rectum

■ FUNCTIONAL DISORDERS

Incorrect eating habits:

- Diet low in fibre:
  - 'Milkoholics', juice-addicts, tea-lovers
- Refined foodstuffs:
  - Low liquid intake
- Lack of colon routine and discipline
- Lack of exercise (due to chronic illness, watching too much TV)
- Psychological (e.g. incorrect potty training, depression, sexual molestation)
- Unavailability of toilets

Initial evaluation

- Careful history
- Physical examination
- Plain abdominal X-rays

■ HISTORY

Determine the duration of the problem:

- Did it start at birth? Was meconium delayed?
- Did it start after a specific event?
- When was the child weaned from the breast?
- When were solids introduced?
- When did the child start potty-training?
- Has the child recently started nursery school?

Does the child pass stools spontaneously or only after medicine or an enema?

- Take a complete dietary history.
- Does the child have faecal soiling?
- How did the child react to previous treatment?
- Does the child have any other diseases or use any medication?

Medical history is a method of excluding Hirschsprung's disease. Ask specifically whether the child was able as a baby to pass normal

stools. Hirschsprung's disease is a congenital disorder that is present from birth. These children have an abnormal evacuation mechanism and will never be able to pass stools normally.

If there is still doubt, then give a saline enema as a therapeutic test. Any normal rectum empties easily with water and the contents squirts out. This does not happen with Hirschsprung's disease and these patients are able to walk about for up to an hour without any evacuation. If there is still some remaining doubt, then anorectal biopsies must be done to test for the presence of ganglion cells.

■ PHYSICAL EXAMINATION

General appearance

- Nutritional status?

Abdomen

- Distended abdomen?
- Faecal mass?
- Intra-abdominal tumour?

Anal inspection

- Normal position of anus?
- Evidence of faecal soiling?
- Peri-anal infection or anal fissures?

Rectal examination

- Anal tone and sensation?
- Faecal mass present?
- Presence and consistency of stools?
- Other masses present?
- Back and spinal examination

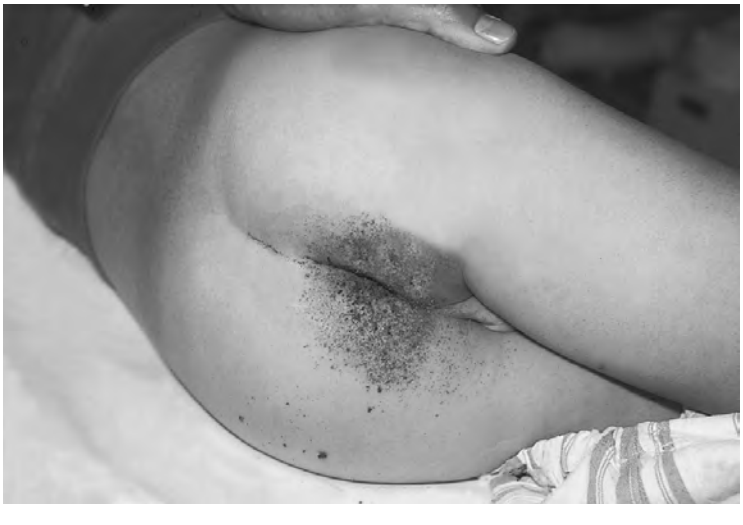


Fig. 13.1 Peri-anal soiling due to encopresis.

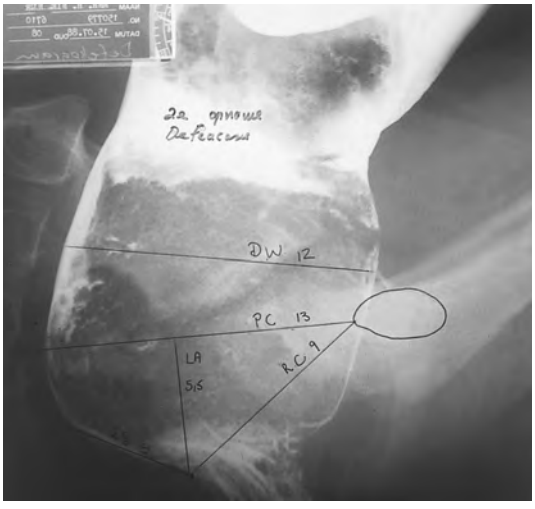


Fig. 13.2 Primary encopresis due to faecal impaction with overflow incontinence.

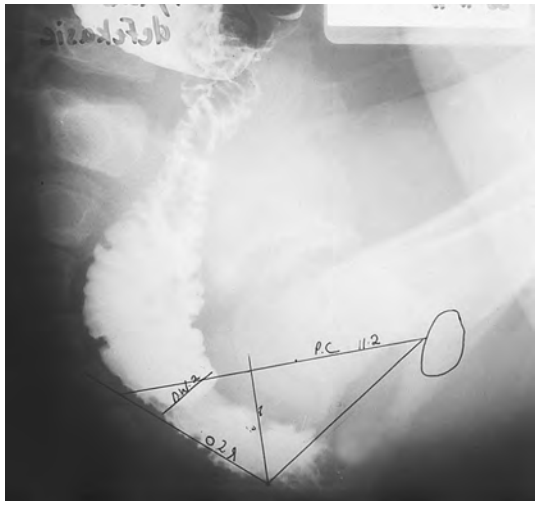


Fig. 13.3 Secondary encopresis due to psychological problems with a normal-calibre rectum.

Special investigations

In most cases the diagnosis of functional constipation is made without any special investigations. If there are any abnormal findings from the history or clinical examination, or if the patient does not improve on treatment, further investigations should be done. These are:

- plain abdominal X-rays
- contrast studies
- anorectal manometry
- rectal biopsy
- test for transit time of the colon

Complications of untreated constipation

Children:	Chronic repeated abdominal pain
	Anal prolapse and fissure
	Rectal bleeding
	Encopresis
	Enuresis
	Failure to thrive
Adults:	Haemorrhoids
	Spastic colon syndrome
	Diverticular disease
	Colon carcinoma

The diagnosis of abdominal pain as a result of an overloaded colon is an exclusion diagnosis. The other dangerous causes, e.g. appendicitis, intussusception or malrotation, must first be excluded before a constipated colon is suggested as a diagnosis.

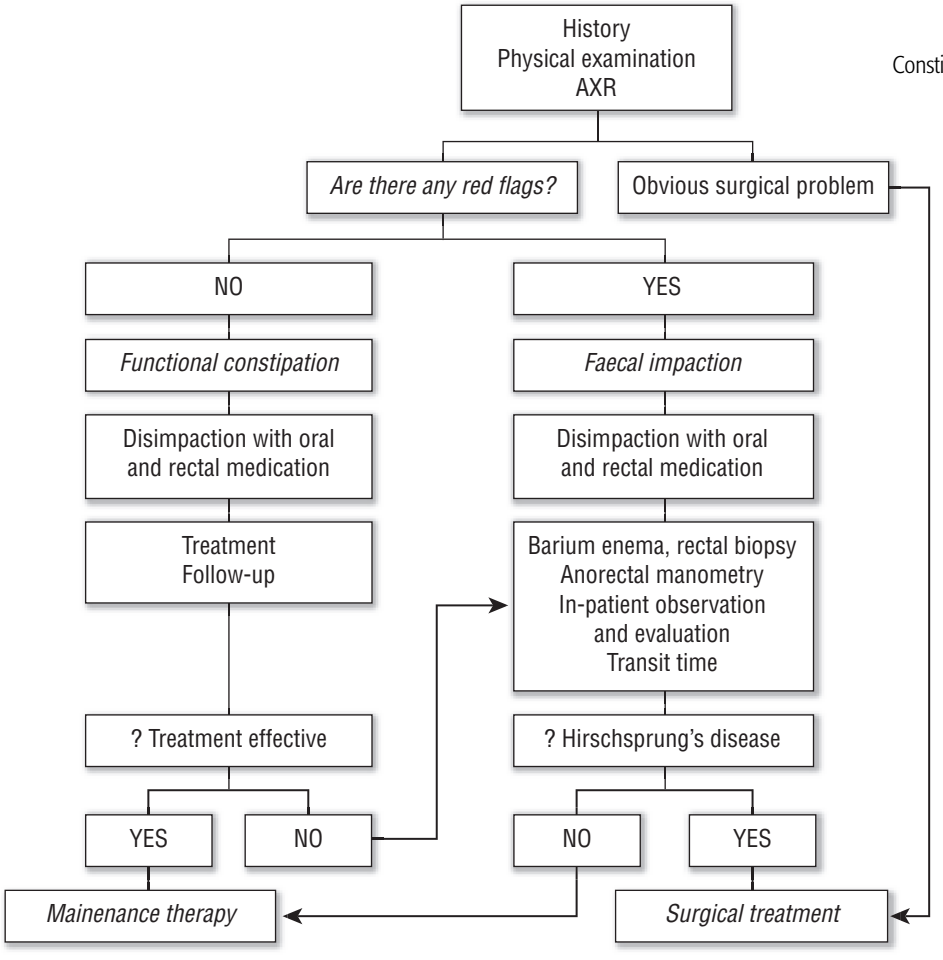


Fig. 13.4 Approach to constipation.
Source: Baker *et al.* 1999

Constipation can be confirmed with a survey abdominal X-ray on which an overloaded colon will be shown if present.

Treatment of constipation

- Disimpact the colon.
- Adjust the diet.
- Use faecal softeners when necessary.
- Encourage good bowel habits.
- Educate parents or family.
- Ensure regular follow-up and support.

Management of an overloaded colon

A high saline (0,9% NaCl) enema is indicated. (This is preferred to a Fleet® (Lennon) enema because the latter empties only the rectum.) A saline enema empties the entire colon.

Why a soap-and-water enema should not be used:

- There have been accidents in which the soap has caused gangrene of the colon.
- If the colon is greatly distended, then not all the soapy water will be emptied, with consequent absorption of the remaining water and the risk of water intoxication.

A single purgative by mouth, e.g. X-Prep® (Rio Self Med) or Epsom salts ($MgSO_4$) is recommended to ensure that the proximal colon is also emptied of accumulated content. As soon as the colon has been emptied, colon discipline (see below) should commence and it must be continued throughout the patient's life.

Diet adjustment

The typical modern Western diet no longer contains nature's laxative, namely bran. Bran is hygroscopic, attracts water to the colon and therefore keeps stool softened.

In addition, bran

- lowers cholesterol by 15%
- helps with the regulation of a diabetic diet
- prevents haemorrhoids
- reduces the incidence of colon carcinoma
- reduces diverticular disease
- reduces the symptoms of the so-called 'spastic colon syndrome'

The recommended dose is one tablespoon of raw bran ('digestive bran') per day. Begin with small amounts as soon as the baby starts solids and adjust the dose gradually up to one tablespoon per day. In small infants, bran can be replaced with oat bran or oatmeal porridge.

Most people claim to have sufficient fibre intake, but a detailed dietary history often reveals that this is not true. A tablespoon of bran must be taken daily for the rest of one's life, including weekends and holidays, especially when travelling.

The raw bran is taken every morning at breakfast with one's cereal or porridge. It can also be mixed with any sandwich spread. Bran must not be seen as a medication, but rather as a part of the family's diet. Excuses such as "I don't eat breakfast" or "I don't eat grass" should not be accepted. A responsible lifestyle includes a balanced diet with sufficient fibre.

■ DIETARY SOURCES OF NATURAL BRAN

- Fruit – do not peel (if appropriate) – eat fresh or dried
- Vegetables, especially beans, peas, cabbage, spinach, carrots, etc.
- High-fibre breakfast cereal, e.g. Weet Bix, All Bran, oats
- Corn or popcorn
- Brown or whole-wheat bread
- Potatoes with their skins

- Brown (unpolished) rice
- Lentils (add to soup, rice, pasta, salads, mincemeat)

How do you convince people to eat a high-fibre diet?

- Remember: 'Hunger has no preferences'.
- Excessive intake of cane sugar should be avoided.
- Eliminate refined foods (white bread, Rice Crispies, etc.) from the diet.
- A low-calorie diet stimulates the appetite which will encourage people to eat healthy food.

Good bowel habits

Most human beings have a normal gastrocolonic reflex that is fairly regular (one could almost set the clock by it). This reflex can be acquired so that it occurs every morning after breakfast.

The colon is usually emptied once a day and most people do not find it necessary to empty the colon again before the next day at the same time.

If you get up too late and have only a quick bite to eat before leaving home, the consequences are:

- The reflex occurs when it is inconvenient or when a toilet is not readily available.
- The person then holds back.
- The reflex vanishes.
- The reflex then occurs when a toilet is not available or only the next day.

The results of this are:

- gradual accumulation of stool
- constipation
- colon distension
- repeated abdominal pains
- passage of hard stools which can give rise to
  - fissures *in ano*
  - haemorrhoids
  - pain and bleeding per rectum

Colon discipline therefore entails a rescheduling of the routine of the family, namely:

- Get up early enough.
- Eat breakfast with bran before commencing the morning routine of washing and dressing.
- There will now be enough time for the gastrocolonic reflex to occur so that the colon can be emptied before the daily tasks begin.

This programme is recommended for both adults and children.

13.2 Encopresis

Definition

Encopresis is the antisocial passage of stools in children older than 4 years, in the absence of organic pathology, unlike incontinence due to organic pathology.

There are two types of encopresis: primary and secondary.

Primary encopresis is encopresis due to faecal impaction with overflow incontinence. The faecal soiling causes secondary psychological problems as a result of teasing at home and at school.

With *secondary encopresis* children have psychological problems and as a result a normal stool is passed antisocially. This usually happens in the clothing, or in the bath, on a seat, in the garden or in any other place deemed to be antisocial.

Clinically, identical complaints occur in both these groups of children, namely:

- soiling
- psychological problems

In primary encopresis, the soiling occurs first and the psychological problems follow, but with secondary encopresis it is precisely the reverse.

In the management of the two types, you should bear in mind that the child's psychological problems resulting from primary encopresis will usually fade away of their own accord when the soiling has been effectively managed and is under control. Conversely, the child with secondary encopresis requires comprehensive psychological support at the same time, as this is the primary problem.

Primary encopresis

■ SYMPTOMS

- Soiling is the first symptom.
- There are psychological problems.
- The child becomes constipated as a result of:
  - faecal impaction
  - an overloaded colon
  - overflow incontinence.
- The child passes continuous runny, watery, foul-smelling stools – day and night.
- The child cannot sense the need to pass stool because the rectum is constantly overstretched.

■ DIAGNOSIS

- A clinical examination and history will exclude other causes of diarrhoea.
- An abdominal examination confirms an overloaded colon.

- A rectal examination confirms an overloaded rectum.
- X-rays confirm an overloaded colon.
- A barium enema on an unprepared colon confirms the overloaded rectum and colon.

■ MANAGEMENT

- Administer a saline enema until the colon is clean.
- Give an oral purgative to empty the caecum.
- The colon must now be emptied daily before school.
- Adjust the diet to keep the stool soft.
- Regular follow-up and emotional support is of the utmost importance to avoid setbacks.

A child who is managed in this way should not soil again.

DICTUM

An empty colon cannot soil. In other words, when the bowel is kept empty, the child will not soil and the psychological problems will become manageable.

■ LEARNING THE BOWEL ROUTINE

These children cannot be allowed to defecate without assistance because the rectum and colon are now overstretched and have low tone, as well as the child having poor sensation. A large overstretched colon will simply fill up progressively and the vicious cycle of impaction and overflow incontinence will repeat itself.

These patients require a daily enema to empty the rectum. The enemas are continued daily until sensation returns and the child begins to defecate on his or her own in the morning just after breakfast. The process of learning a bowel routine can take from one week up to several months.

The routine is as follows:

- Get up early enough.
- Eat breakfast with bran.
- Apply a Lenolax® enema (paediatric or adult, depending on the age).
- Spend time on the toilet (check that the colon is completely empty – sometimes children get up too soon).
- Since an empty bowel cannot soil, the child is then ready for the day.

Secondary encopresis

The primary problem is psychological.

■ DIAGNOSIS

- The abdomen is usually soft.
- There are soft stools in the rectum or even an empty rectum.

- A barium enema shows a colon of normal calibre, not overloaded.
- The psychological cause of the problem must be found.

■ MANAGEMENT

The management of the soiling aspect is precisely the same as with primary encopresis because an empty bowel cannot soil – even if the child wishes to. This child must be given a bowel routine to follow, as described above. Referral for psychological evaluation and therapy must be done simultaneously.

13.3 Repeated abdominal pain in children

This is a very difficult problem that leaves many doctors racking their brains. The children lose many hours of school time, the problem spoils many holidays and many practitioners have had their sleep interrupted by this complaint.

Approach

A thorough medical history must be documented and a complete examination conducted.

- Never conclude that this is a matter of 'crying wolf', even if it has happened a hundred times before this particular episode.
- Examine the abdomen thoroughly.
- Check for urological complaints and do investigations.
- Take abdominal X-rays.
- Do an abdominal sonar.
- Place the child on a high-fibre diet and a bowel routine.
- Do a barium enema and exclude malrotation. Ask the radiologist to determine whether the appendix fills with barium; if this is the case, then the cause is unlikely to be appendicitis.

If malrotation is radiologically diagnosed, then the child should be booked for an elective Ladd operation and appendectomy.

Necrotising enterocolitis

14

JHR BECKER & SM LE GRANGE

14.1 Pathophysiology

Necrotising enterocolitis (NEC) is the result of ischemic damage to the hollow viscera of the abdominal cavity due to an adverse incident at birth (peri-partum). Irrespective of the nature of this 'attack', the baby goes into a state of shock, during which time a compensatory vasoconstriction takes place, with a redistribution of blood from the so-called 'non-vital organs' to the vital organs. This is known as the *diving reflex*.

The non-vital organs become ischemic, which can lead to cell damage. The extent of the damage is directly related to the level and duration of the initial ischemia. The bowel is regarded by the body as non-vital and is exceptionally sensitive to ischemia. The extent of the damage varies from mucosal damage only, to full-thickness transmural gangrene and loss of the bowel wall.

As a result of the loss of integrity of the bowel wall, colonisation of bacteria from the ulceration takes place. A vicious cycle develops: bleeding from the ulcer, followed by the multiplication of bacteria in the ulcer, which exacerbates the damage and causes gas in the bowel wall, some of which, together with bacteria, ends up in the portal system with consequent portal septicaemia. The outcome of this spread of the disease is perforation, peritonitis, septic shock and death.

There are various theories about the cause of such attacks on a baby at birth, namely:

- hypothermia
- prematurity
- low birth weight
- respiratory distress syndrome of the neonate
- foetal distress during birth
- asphyxia neonatorum

- umbilical vessel catheterisation
- exchange transfusion
- hypoglycaemia
- symptomatic congenital heart disease
- early formula feeds

14.2 Symptoms and signs

The high-risk baby should be closely observed. The first sign is usually that the baby becomes *intolerant of feeds*. *Blood per rectum* may be the only manifestation. General *signs of septicaemia* are difficult in a neonate and include things such as unstable vital signs, erratic breathing, etc. The abdomen becomes *distended* and in severe cases may *splint* the diaphragm, causing *ventilatory problems*. When the abdominal wall becomes *red* (umbilical flaring) and/or *oedematous*, or if a *mass* is palpable, these usually point towards necrotic bowel.

Special investigations

- Full blood count:
  - White cells can be either high or low
  - There is a tendency to thrombocytopenia with severe disease (can be used to monitor treatment)
- Biochemistry
- A persistent hyperkalaemia usually indicates necrotic bowel
- C-reactive protein/procalcitonin
  - CRP is a non-specific marker of inflammatory disease – may be used to monitor treatment

Procalcitonin is a more specific marker for infections

- Abdominal X-ray
  - Gas in the bowel wall – this is called *pneumatosis intestinalis* and is pathognomic
  - Paralytic ileus
  - Gas in the portal venous system
  - Sentinel loop
  - Ascites
  - Free air

Classification

This simple classification not only gives an idea of what the prognosis of the baby with NEC is, but also serves as a guideline for surgical intervention:

Grade Ia: non-specific signs
Grade Ib: bloody stools
Grade IIa: pneumatosis intestinalis

Grade IIb: air in portal vein
Grade IIIa: ascites
Grade IIIb: free air/perforation

The surgeon is most probably of use only in Grade III.

14.3 Management

Supportive

This is the mainstay of the treatment.

- These babies usually are already in a neonatal intensive care unit – if not, transfer immediately
- *Nil per os* (NPO)
- Nasogastric tube on suction/hourly aspirates
- Intravenous infusions:
  1. maintenance fluid
  2. parenteral feeding
  3. packed cells or platelets as necessary
- Broad spectrum intravenous antibiotics, covering all enteric organisms, are mandatory
- Ventilation, if necessary

14.4 Surgical intervention

Necrotising enterocolitis is being seen more and more often as a medical condition. There are only a few indications for surgical intervention:

- Clinical:
  - Distended abdomen, splinting the diaphragm
  - Umbilical flaring
  - Oedematous abdominal wall
  - Mass palpable
- Blood tests:
  - Plummeting platelet count
  - CRP/procalcitonin remains high
  - Persistent hyperkalaemia
- X-rays:
  - Sentinel loop (bowel loop stays stagnant over a series of X-rays)
  - Free air

Type of surgery

More and more surgeons will only place peritoneal drains in these babies. This technique usually leads to dramatic improvement and

no definitive surgery is necessary. If, however, the baby does not improve after peritoneal drainage, a laparotomy becomes mandatory. The theory behind this is that the bowel is not necessarily necrotic and very often the surgeon finds nothing to do. If drainage does not help, it indicates that large pieces of bowel are necrotic, and should be resected.

Babies treated with drains only should have a contrast enema before discharge to rule out colonic stricture-formation as part of the healing process of NEC.

14.5 Prognosis

The mortality in this group of patients remains high. Not only may NEC be a serious disease, but it also occurs in a very fragile subgroup of patients.

Important factors:

- Early recognition of the disease
- Appropriate management
- Prompt referral

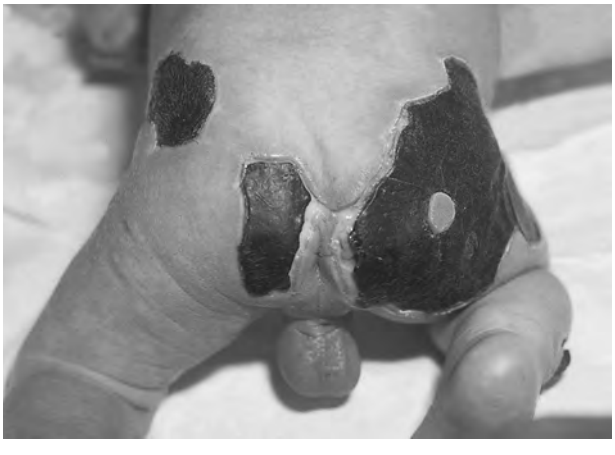


Fig. 14.1 Skin necrosis caused by thrombosis of small cutaneous blood vessels due to disseminated intravascular coagulation (DIC) in a patient with necrotising enterocolitis.

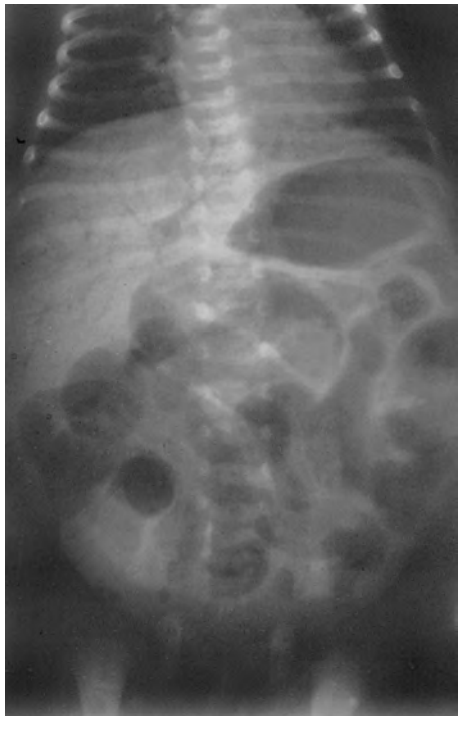


Fig. 14.2 Air in the bile ducts translocated from the bowel in a patient with necrotising enterocolitis.

Glandular swellings in children

15

JHR BECKER & LZ MARCISZ

Swellings in the parotid gland appear from time to time and can occasionally be a diagnostic dilemma.

15.1 Acute parotitis

Mumps

This is a common childhood disease that is treated symptomatically.

Acute suppurative parotitis

This is a mixed infection of the parotid with drainage of pus from Stenson's tube, usually in children under the age of 3 years, where the opening is injured by the suckling process.

■ MANAGEMENT

- Prescribe a broad spectrum antibiotic.
- Massage the gland in the direction of the mouth.
- The patient should drink lemon juice – it stimulates the salivary secretion which effectively rinses the gland.
- Check that there is no stenosis of the papilla in the mouth, as this would have to be dilated or incised.

Recurrent parotitis in children

This is most likely an auto-immune disease that may present unilaterally or bilaterally, usually in children between the ages of 4 years and puberty. The swelling is cyclical in nature – it comes and goes.

Surgery is not recommended because the condition usually burns itself out by the time puberty is reached.

■ MANAGEMENT

- Massage the parotid gland from the back to the front.
- The patient should drink lemon juice to stimulate secretion (salivation).
- Prescribe a broad spectrum antibiotic if signs of a secondary infection are present.
- Keep the patient under close observation.

15.2 Congenital abnormalities

Haemangioma

This is one of the most common benign swellings of the parotid. The management is conservative (non-operative) because there may be spontaneous remission. If there is any doubt, then refer the patient to a paediatric surgeon for a second opinion. The latest treatment is bleomycin intralesional injections.

Lymphangioma (macrocystic lymphatic malformations)

This is part of a cystic hygroma. It must be operated on by a paediatric surgeon with experience in this field. Bleomycin intralesional injections are also recommended.

Brachial cyst

This is a remnant of the first brachial arch and should also be operated on by a skilled paediatric surgeon.

15.3 Tumours of the parotid gland

Suspect any swelling in the parotid to be a tumour if it shows the following signs:

- firm consistency
- discrete lump
- unilocular
- rapid growth
- infiltration of N VII
- on a sonar or CT scan the swelling appears to be different from the rest of the parotid

All swellings showing such signs should be further investigated and not just dismissed as benign swellings.

Actions

- Obtain a second opinion.
- Do aspiration cytology.
- Do ultrasound or a CT scan to chart the extent of the tumour.

NB: Children also get malignant tumours of the parotid; they are rare, but be aware of this possibility.

15.4 Submandibular salivary gland problems

The common problem that occurs here is a stone. If the stone is in the oral part of the duct, a suture is placed behind the stone (to prevent it from sliding backwards into the main gland) before it is removed by making a small incision over the stone. If the stone is located within the gland, then the entire salivary gland must be removed.

15.5 Conditions of the thyroid

Neonatal goitre

In this case, the baby is born with a greatly enlarged thyroid that could compromise respiration. The neonate's neck is rather short and the thyroid partially encases the trachea – with the risk of an airway obstruction.

■ CAUSES

- Lowered maternal intake of iodine
- Raised maternal intake of iodine – use of medication
- Congenital enzyme deficiency of the thyroid
- Maternal intake of a goitrogen, e.g. para-aminosalicylic acid
- Intake of thyroid-suppressing drugs, e.g. thiouracil
- Exopthalmic goitre

In the latter case, the baby shows signs of exophthalmos, such as are found in Basedow/Graves' disease. There are few cardinal signs, but clear signs of a raised metabolic rate which manifests mainly as weight loss or poor weight gain, tremors and irritation. The diagnosis is confirmed if the serum thyroid hormone levels are raised and if raised thyroid uptake of I^{131} is present. It is managed by administering anti-thyroid drugs until the patient is euthyroid, and then with surgical intervention.

The same management is appropriate for the older patient with primary hyperthyroidism.

■ MANAGEMENT OF NEONATAL GOITRE

This excludes primary hyperthyrotoxicosis.

The respiratory distress is managed by:

- extending the neck
- giving oxygen
- giving exogenous hormone orally

The thyroid should shrink and the symptoms should improve. Tracheostomy is not recommended. Rather intubate and operate on the thyroid if respiratory distress does not respond to conservative measures.

■ INDICATIONS FOR SURGERY

- Respiratory distress that does not respond to conservative treatment
- A goitre that improves after treatment with exogenous hormone, but does not shrink further and is still large and cosmetically unacceptable

NB: These patients must all be followed up to prevent them from becoming hypothyroid.

Physiological goitre

This normally occurs during puberty and clears up spontaneously. It may be necessary in certain cases to provide exogenous hormone temporarily.

■ THYROID NODULES

These fall into three categories:

- cysts
- adenomas
- carcinomas

A malignancy in the thyroid must be considered in all cases of a single, solid, 'cold' nodule, particularly in the euthyroid girl of 8–12 years old.

■ RADIOACTIVE I^{131} UPTAKE IS IMPORTANT

A functioning nodule is most likely not to be malignant (it is probably an adenoma), but with a solid cold nodule there is a 27% chance of malignancy. If the nodule is cystic, then it could be aspirated and left alone if:

- the cytology is negative
- the fluid is clear and not blood-stained
- the cyst does not fill up again
- no residual mass is palpable after aspiration

■ MANAGEMENT OF A SINGLE THYROID NODULE

- Do an excision biopsy, leaving a border of normal tissue.

- If it is malignant, then the same principles apply as in adult surgery, namely:
  - less than 2 cm requires an ipsilateral lobectomy
  - larger than 2 cm requires an ipsilateral lobectomy, as well as removal of the isthmus and contralateral subtotal resection.
- Multi-centricity implies the need for a bilateral total thyroidectomy.
- All affected lymph nodes are removed.

NB: Do not forget chest X-rays to show metastases.

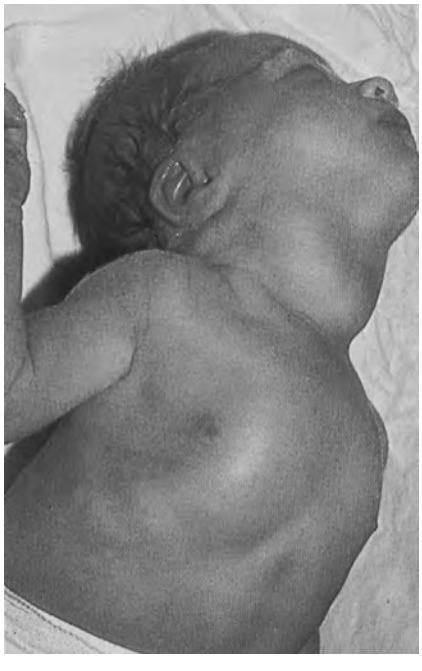


Fig. 15.1 Neonatal goitre causing respiratory distress.

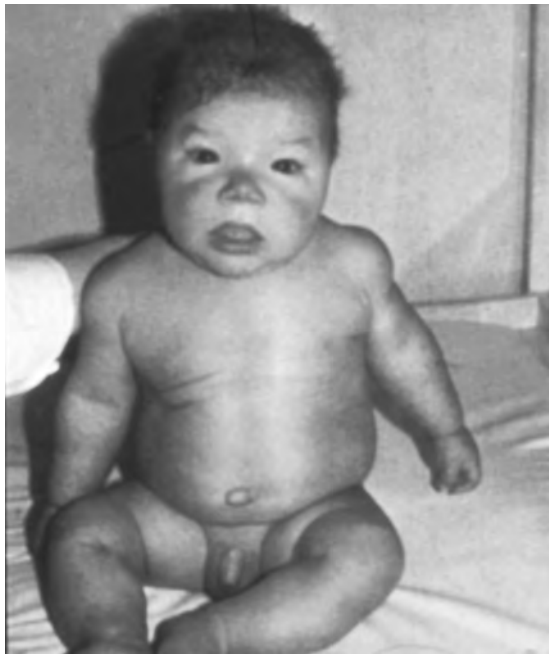


Fig. 15.2 Baby with typical signs of hypothyroidism (cretinism).

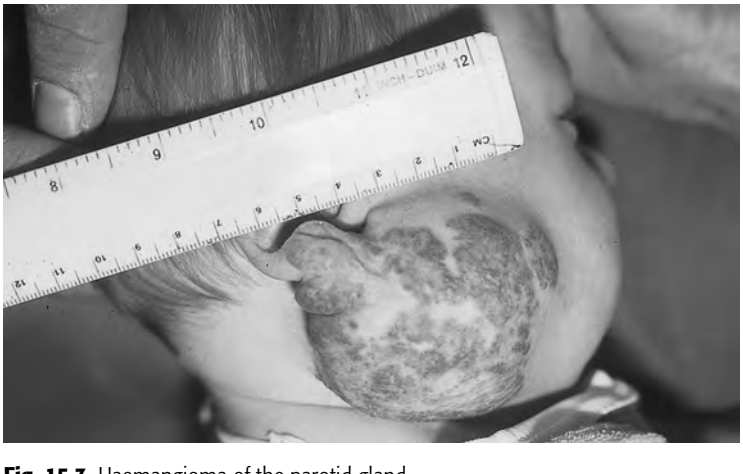


Fig. 15.3 Haemangioma of the parotid gland.

Portal hypertension

16

JHR BECKER & MH SHEIK-GAFOOR

The symptomatology of portal hypertension is twofold. Symptoms can be caused by either:

- the presence or absence of liver pathology, or
- portal hypertension

16.1 Liver pathology

The pathology in the liver (the cause of portal hypertension) and the level of residual liver function each have their own plethora of symptoms, e.g. oedema, ascites, malnutrition, jaundice, etc.

If the pathology is in the liver, with hepatocyte damage, the prognosis depends on the amount of residual liver that is left. The effects of the portal hypertension are an added problem ('double trouble').

If the pathology is prehepatic (which is often the case), then there is no hepatocyte damage and the liver function is normal. This means that these children have a normal life expectancy, provided the effects of the portal hypertension are effectively managed.

16.2 Effects of portal hypertension

Raised pressure in the portal system has the following effects:

- oesophageal varices with consequential bleeding
- hypertensive stomach mucosa with consequential bleeding from erosions
- possible poor absorption of nutrients from the bowel

- hypersplenism, which could cause a cytopenia of one or more of the three main blood cell components: red blood corpuscles (RBC), white blood corpuscles (WBC) or platelets
- ascites

16.3 Aetiology of portal hypertension

The major cause is pathology in the flow of blood through the portal venous system – the obstruction can be pre-, intra- or post-hepatic.

- *Pre-hepatic* – e.g. portal vein thrombosis
- *Intra-hepatic* – liver parenchyma pathology, e.g. cirrhosis due to hepatitis or the eventual outcome of biliary atresia
- *Post-hepatic* – hepatic vein obstruction, intravenous cholangiography (IVC) obstruction or cardiac pathology

16.4 Haematemesis due to oesophageal varices

The child vomits bright red blood in large quantities, until he or she enters a state of shock and becomes anaemic. Thereafter, the varices often cease bleeding spontaneously, after which the child requires active resuscitation and blood transfusion.

The diagnosis must be confirmed by means of an endoscopy as it must be determined whether the bleeding arose from varices, gastric erosions or an ulcer.

The endoscopy must be done under general anaesthesia and by a doctor skilled in doing paediatric endoscopies and capable of simultaneously injecting a sclerosing agent into the varices.

There is no sense in delaying sclerotherapy because follow-up endoscopy and sclerotherapy will be necessary until the varices have been completely eradicated.

If the bleeding cannot be stopped, further therapy will be necessary. This may include the following:

- intravenous pitressin
- intravenous somatostatin
- Sengstaken-Blakemore tube (paediatric)
- endotracheal intubation and ventilation
- active monitoring and support of liver functions
- repetition of endoscopy and sclerotherapy

Undoubtedly most patients will achieve permanent control of the bleeding from oesophageal varices through the use of the above intensive management techniques, endoscopy and sclerotherapy. Patients who continue to bleed or who bleed repeatedly must undergo a portal systemic shunt operation to lower the pressure in the portal system.

The major problem with lowering pressure in the portal system is the lowered perfusion pressure of the liver. This in turn decreases the metabolism of protein breakdown products, which leads to an increase in ammonia in the circulation, with symptomatic encephalopathy.

The ideal portal systemic shunt creates two surgical compartments in the portal drainage system in order to lower the risk of encephalopathy – one as a high-pressure system that maintains the perfusion to the liver, and the other as a low-pressure system that selectively drains the oesophageal varices and the spleen to prevent bleeding and hypersplenism.

The shunt is called the Warren shunt and it is technically a distal Lieno renal shunt. This entails:

- Attaching a distal splenic vein end-to-side to the left renal vein – the spleen and oesophageal varices drain selectively to the low-pressure IVC (low-pressure system).
- Tying off *the proximal splenic vein*, *the coronary vein and the right gastro-epiploic vein* prevents the high-pressure system that perfuses the liver from communicating with the low-pressure system that decompresses the oesophageal varices and the spleen.

This operation must only be done in a unit that has the necessary skills.

Bleeding from the anus

17

JHR BECKER & RA BROWN

Unlike in adults, the cause is often "transient, almost invariably benign, and usually simply treated" (Raine 1991).

The classic questions are:

- Colour?
- Volume?
- Blood mixed with stool?
- Blood at the end of defecation?
- Is the patient shocked?
- What is the medical history – personal or familial – of polyps, ulcerative colitis or Crohn's disease? Drug ingestion?
- Is there associated pain?

The examination should consist of:

- a full general examination
- inspection of the perineum
- a rectal examination

17.1 Anal fissures

These appear with the painful passage of a very hard stool. The blood is bright red with only a little streak on the outside of the stools. It is diagnosed by peri-anal inspection.

Management

- Analgesia of the anus is achieved with Remicaine® jelly – this covers the anus and anaesthetises it. The jelly is applied regularly during the day until the fissure has healed. Remicaine® jelly must be applied during all procedures (therapeutic or diagnostic) done on these patients.

- The colon must be completely emptied of all hard stool by means of a saline enema at body temperature.
- Colon discipline and the daily routine are important.
- Early-morning rectal emptying is encouraged, using a glycerine suppository covered with Remicaine® jelly, shortly after breakfast. If this is ineffective, a phosphate enema covered with Remicaine® jelly may be effective.
- Bran (nature's laxative) is strongly recommended.

17.2 Polyps

These are usually juvenile polyps located within 20 cm of the dendate line. Polyps usually occur singly, but they may be multiple (20%).

A polyp can protrude from the anal opening and look like mucosal prolapse or a haemorrhoid. Usually it bleeds very little but regularly, and there is fresh red blood on the sides of the stool.

The treatment involves general anaesthesia, tying off the stalk and amputating it surgically. During the anaesthesia, a sigmoidoscopy should be conducted to exclude pathology higher up in the rectum, particularly further polyps.

17.3 Intussusception

This is the typical mixed bright red blood and slime from the anus that looks like jelly, the so-called 'red currant jelly'. This condition has already been discussed in Chapter 12.

17.4 Malaena

This is black, tarry, foul-smelling stool that is indicative of bleeding proximal to the ligament of Treitz. Investigations of the upper gastro-intestinal tract are therefore required to determine the cause, e.g. an endoscopy should be conducted under general anaesthesia.

Differential diagnosis

- Oesophageal varices
- Peptic ulcer
- Erosive gastritis
- Ingested blood, e.g. as a result of epistaxis

Management

This will depend on the cause, e.g.

- *Oesophageal varices:* Manage the portal hypertension as discussed.

- *Peptic ulcer:* Medical management with an H_2 receptor antagonist, Tagamet® (SK+F), proton pump inhibitor, Losec® (Astra Zeneca) or Sucralphate® (Continental Ethicals).
- *Erosive gastritis:* Exclude an aetiological agent, e.g. salicylates used for fever. Treat by suppressing the acid as in a peptic ulcer.

17.5 Meckel's diverticulum

This is the remnant of the vitelline intestinal duct, located in the distal small bowel at the apex of the midgut. The diverticulum may contain 30% of ectopic gastro-intestinal tissue, e.g.

- stomach mucosa
- pancreatic tissue
- colonic mucosa

The stomach mucosa may contain acid-secreting cells, which can produce hydrochloric acid. The non-neutralised hydrochloric acid will erode the small bowel mucosa and cause a peptic ulcer, which may bleed profusely.

Fig. 17.1 Peptic ulceration at the neck of a Meckel's diverticulum. The diverticulum had ectopic gastric mucosa in it.

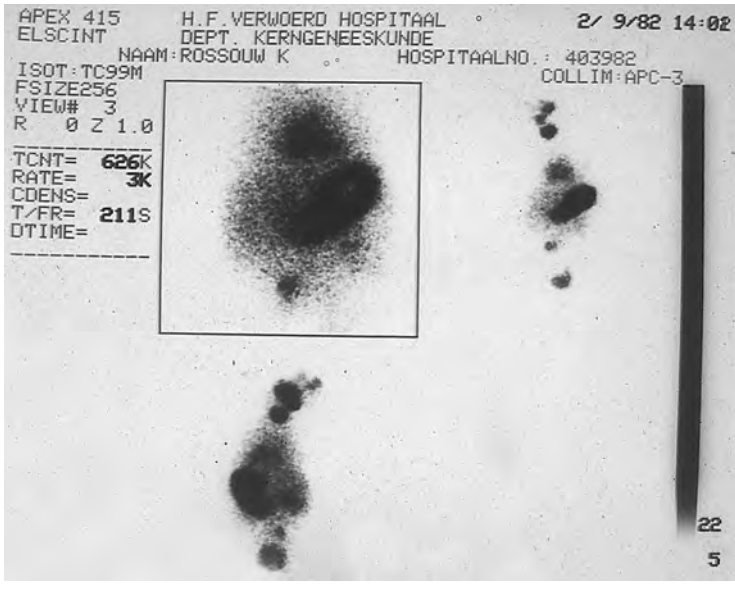


Fig. 17.2 Isotope scan demonstrating a 'hot spot' in the ectopic gastric mucosa of a Meckel's diverticulum.

Typical history

Typical age of presentation is 2–5 years. These children present with significant passage of dark red blood per rectum. This is painless and results in a drop in haemoglobin.

The patient may not bleed sufficiently to cause a state of shock

because the volume can vary from occult to a great deal, as mentioned above.

The bleeding is usually painless, without abdominal pain or any other symptoms.

NB: A Meckel's diverticulum can also present as:

- intussusception, acting as a lead point
- diverticulitis
- volvulus and gangrene of the bowel because of a band to the umbilicus

It may also be the location for tumours, e.g. carcinoid.

Diagnosis of a Meckel's diverticulum

- Typical medical history of bleeding, as described above.
- In a radio-isotope scan, the radio-isotope, such as technetium pertechnitate, concentrates in the ectopic gastric mucosa located in the diverticula and will show up as a 'hot spot' on the scintigram.

Management

The condition is managed by laparotomy and excision of the Meckel's diverticulum with the part of the small bowel that contains the ulcer.

What to do with a diverticulum incidentally discovered at laparotomy: This must be removed, except if there is a surgical contra-indication, e.g. a vascular prosthesis that has just been inserted or any other foreign body (e.g. mesh) that has been inserted during the present laparotomy or if the surgeon is inexperienced with compromised patients.

CAVEAT

The mid-bowel volvulus with severe abdominal pain, a history of abdominal discomfort and bleeding per rectum must not be forgotten.

17.6 Peptic ulcers

These may occur in babies and children, with the usual upper abdominal pain and haematemesis (bright or coffee grounds). The diagnosis must be confirmed by endoscopy. Peptic ulcers are managed with medication (H_2 receptor antagonists, proton pump inhibitors or Sucralphate®). If the ulcer does not heal, then gastrin levels must be determined to exclude the possibility of a gastrin-producing tumour.

Peptic ulcer surgery (if ever necessary) in children should consist of no more than a highly selective vagotomy. If more invasive procedures are conducted, there should be strong motivation for them.

17.7 Prolapse of the anus

Prolapse of the anus means that the normal configuration of the anus has been disturbed by an abnormal object.

The possibilities are as follows:

- mucosal prolapse
- intussusception of the anorectum
- intussusception of the rectum or colon
- prolapsing or protruding polyp

Mucosal prolapse

This is a common occurrence in babies and toddlers who are beginning to pass formed stools, become constipated or are learning to use a potty.

The baby's anorectal mucosa is quite mobile and can easily extend to prolapse from the anus. Apart from the mobility of the anorectal mucosa, the following factors may contribute to a mucosal prolapse:

- The anorectum is straighter in contrast to the sigmoid form of the adult anorectum.
- The sacrum is straighter in the child than the adult.
- The child sometimes sits for prolonged periods on the potty bearing down to pass a stool when there is nothing to be passed. The posture predisposes to prolapse.
- There may be a worm infestation.
- There may be diarrhoea with frequent passage of stool and hyperperistalsis.
- There may be severe constipation.

Clinically,

- There is red-pink mucosa hanging out of the external anal opening.
- There is bloody slime on the nappy and/or undergarments.
- It is often painless.
- The child actually continues to bear down because there is a sensation of fullness in the anus, resulting in a sensation of incomplete evacuation.

■ EXAMINATION

- The findings will be as above.
- The examiner must ensure that it is in fact a mucosal prolapse and not a full-thickness prolapse.
- A space between the mucosa and the anal opening cannot be found on digital palpation.

■ MANAGEMENT

- The mucosa is pushed back with a gloved finger.
- The mucosa must not be left outside for a long time as it then congests and reduction may be extremely difficult.
- Treat the cause, e.g. incorrect potty training, worms, constipation or diarrhoea.
- The buttocks are taped together to prevent the prolapse recurring while the cause is being addressed.
- Ninety per cent of cases will rectify themselves after conservative measures, as set out above, have been taken.
- Ten per cent of cases require an injection of ethanolamine-oleate to fixate the mucosa so that it does not prolapse again.

Intussusception of the anorectum

Unlike the common mucosal prolapse, this is a case of prolapse of all the layers of the anorectum, in other words of the muscle layers as well.

Clinically, this can look similar to a mucosal prolapse, but on examination it can be clearly seen that the prolapse is coming from inside the anal opening and that there is a space between the intussusseptum and the anal opening. A finger can even be placed in this space.

■ MANAGEMENT

The management is the same as for mucosal prolapse, including the injection, but it is less successful.

These cases often require surgery to fix the rectum to reduce its mobility. The operation is usually successful. Sigmoid resection with mobilisation of the rectum is the operation of choice.

Underlying predisposing causes (*Tricburis* infestation, cystic fibrosis, meningomyelocoele) must be excluded.

Intussusception of the rectum or colon

- There is obvious prolapse of bowel at the anus.
- There is a clear space between the intussusseptum and the anus into which the examining finger can be placed.
- The protruding intussusseptum is curled backwards because the mesentery, which is also part of the intussusseptum, pulls it cephelad.

■ MANAGEMENT

- Manual or hydrostatic reduction with barium.
- If the intussusseptum does not reduce, then it requires operative reduction.
- If no lead point is present, e.g. a polyp, then reduction will be sufficient. If there is indeed a lead point, then the intussusception

will recur, and all cases with a lead point must be operated on and the cause removed.

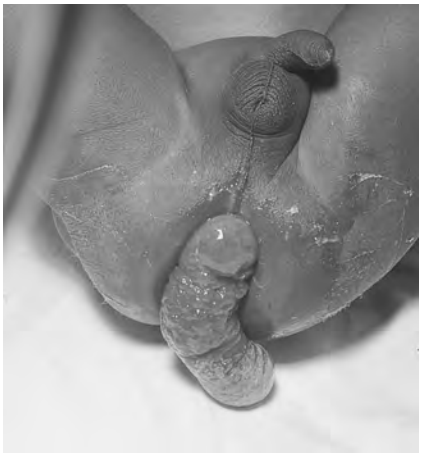


Fig. 17.3 Intussusception of the ano-rectum; note the posterior curvature of the intussusceptum due to traction by the mesentery.

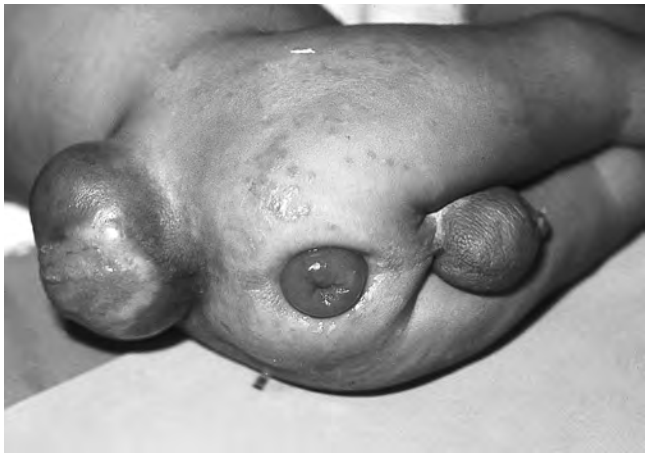


Fig. 17.4 Anal mucosal prolapse due to a patulous anus caused by a spina bifida and myelomeningocoele.

17.8 Haemorrhoids

The condition is rare in babies and even in children, but must be distinguished from a haemangioma or submucosal haematoma of the anus ('external pile').

Haemorrhoids cause bleeding at the end of the passing of stool, normally visible when wiping the anus or on the sides of the toilet bowl – the blood is usually bright red.

The treatment is similar to that for constipation, i.e.

- colon discipline
- a high-fibre diet

However, surgery is not usually indicated.

Naevi

18

JAN J VAN WINGERDEN

A *hamartoma* is a tumour-like overgrowth of *mature* cells and tissue normally found in the affected area. A *naevus* is a hamartoma of the skin, composed of a proliferation of vascular tissue (vascular naevus), sebaceous tissue (sebaceous naevus), or melanocytes (pigmented naevus).

18.1 Vascular naevi

Portwine stain

This is a diffuse (capillary) *vascular malformation* (see Chapter 4, Section 4.1) usually found on the face and, to a lesser extent, the extremities.

When discovered in the face, in particular in the trigeminal nerve distribution, it may be associated with leptomeningeal angiomatosis (Sturge-Weber syndrome). These children may suffer from severe epilepsy and early referral is advisable.

Klippel-Trenaunay syndrome consists of a triad of portwine stains, anomalous veins and overgrowth of a limb. It is progressive and may require stapling epiphysiodesis of the knee cartilages to contain the growth of the affected leg. It should be distinguished from a non-progressive variant, which consists of portwine stains and congenital hypertrophy of the limb (no vein anomalies associated) (Enjolras *et al.*, 2004).

Two-thirds of facial portwine stains improve with pulsed dye laser therapy and in approximately 10% of patients, complete ablation can be expected. The rest would benefit from cosmetic camouflage (Stone 2001).

Strawberry naevus

This is a capillary *haemangioma* and therefore usually discovered at birth or within the first month of life. It has a recognisable growth pattern (see Chapter 4, Section 4.1). It presents as an elevated red, vascular tumour, anywhere on the body but more commonly in the region of the head and neck. Referral to a plastic surgeon, who will consider the various management options in consultation with the parents, is recommended.

Early or immediate referral is required in cases of:

- haemangiomas in close proximity to the airway
- peri-orbital haemangiomas
- cases of children with a suspected haemangioma who present with severe bleeding (Kasabach-Merritt syndrome – see Chapter 4, Section 4.1).

18.2 Sebaceous naevi

This uncommon hamartoma, also known as the *naevus sebaceous of Jadassohn* (NSJ), presents as a yellow, wart-like lesion, which changes from an epithelial hyperplasia initially, with age to a lesion consisting mainly of hyperplasia of the sebaceous glands. Sebaceous naevi are most commonly found in the region of the head and neck.

Their clinical importance is twofold:

- Malignant transformation, although uncommon in childhood, has been described. This is usually a basal cell carcinoma, but a squamous cell carcinoma has been described. This is the main reason why total excision of these naevi is recommended (Hagan 1987; Hidvegi *et al.* 2003).
- When an NSJ presents as a linear lesion in the face, a difficult tracheal intubation due to facial asymmetry can be expected. These cases are not uncommonly associated with intractable seizures, which will challenge peri-operative management (Diaz 2000).

18.3 Pigmented naevi

Five types of pigmented (*melanocytic*) naevi will be described:

1. Junctional naevus
2. Compound naevus
3. Spitz naevus
4. Dysplastic naevus
5. Large congenital melanocytic naevus

Junctional naevus

This is a flat (macular) lesion, with colour variations of shades of brown. It is sometimes slightly elevated, with a small light or dark surrounding halo. Such naevi are common in children and adolescents.

Compound naevus

This is a maculopapular-pigmented naevus, which is more common in adolescents.

Spitz naevus

This is a darkly pigmented naevus and thus often clinically indistinguishable from a melanoma. It is usually found in the region of the head and neck and may clinically resemble a small haemangioma. Complete excision is advocated.

Dysplastic naevus

Dysplastic naevi are larger than 5 mm in diameter, have indistinct borders and are various shades of brown. They may be discovered on the scalp first. Children with *dysplastic naevus syndrome* present with numerous naevi by school-going age and have an increased risk of melanoma. Regular dermoscopy (epilumenescence) by an experienced dermatologist may be of value in these children.

Large congenital melanocytic naevus

This is a giant, pigmented, hairy naevus that involves a large body-surface area and is also known as a 'garment' naevus.

The correct, internationally accepted scientific name – a 'large congenital melanocytic naevus' – is preferable. This is, by defini-

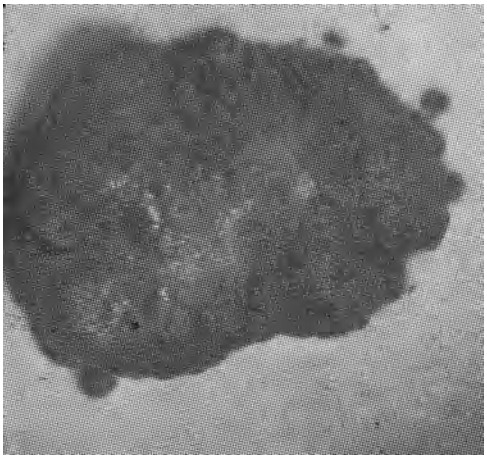


Fig. 18.1 Strawberry naevus.

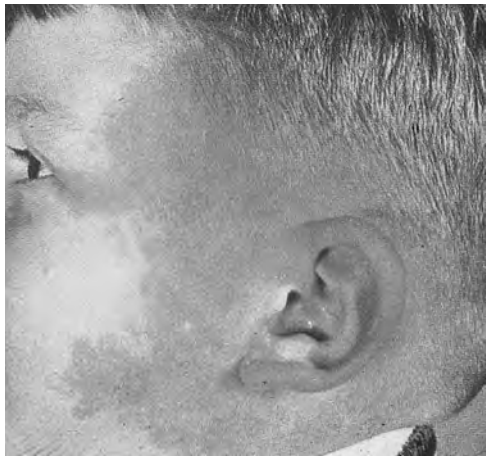


Fig. 18.2 Portwine naevus.

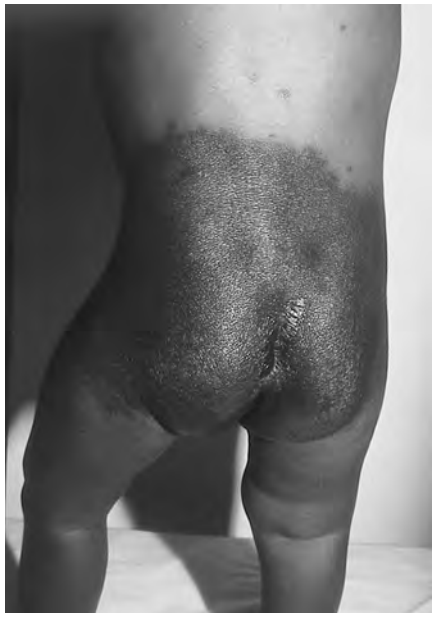


Fig. 18.3 'Bathing trunk' naevus ('garment' naevus).

tion, a lesion whose largest diameter in adults is at least 20 cm or a lesion that is predicted, by growth chart analysis, to attain a largest diameter of at least 20 cm in adulthood (Bittencourt *et al.* 2000). Although numerous studies have suggested a significantly increased risk of melanoma in these patients, the exact incidence remains unknown (Zaal *et al.* 2004; Watt *et al.* 2004).

Treatment, if the patient is referred within the first 6 months of life, would be by curettage or dermabrasion. These two methods will improve the cosmetic appearance but there is no evidence that they will decrease the risk of malignant transformation (Bauer *et al.* 2001).

If the patient is referred only at a later stage, the plastic surgeon will consider prophylactic excision, either serial or complete. The excision will extend at least to muscle fascia as most of the melanomas found in large congenital naevi have non-epidermal origins. The defect will be closed with tissue (either a skin graft or a flap) obtained with the aid of tissue expansion.

18.4 Melanoma

Malignant melanoma in childhood is fortunately rare. It may arise:

- congenitally, through transplacental spread
- in specific conditions or under specific circumstances:
  - large congenital naevi
  - dysplastic naevus syndrome
  - xeroderma pigmentosa
  - immunodeficiency
- *de novo*

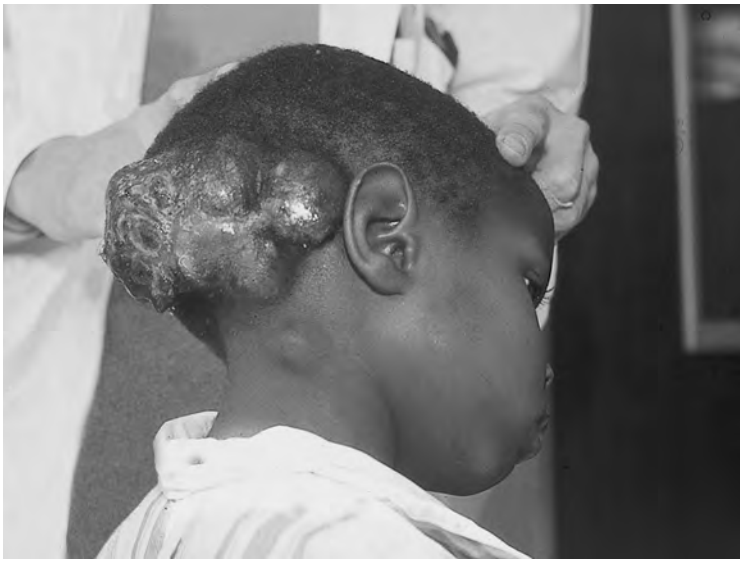


Fig. 18.4 Melanoma of the occiput; note the metastasis in the apex of the posterior triangle of the neck.

Lentigo maligna melanomas are unknown in childhood and acral lentiginous melanomas are highly unusual. Superficial, spreading melanomas are most frequently found and nodular melanomas occasionally.

It would be prudent for the general practitioner to hone his or her skill in detecting the potentially malignant pigmented lesion. After careful clinical assessment, the practitioner should be able to triage patients and lesions into one of three groups (mnemonic – 'ART'): **A**ct (biopsy), **R**eassure (advise and ignore) and **T**rack (re-evaluate at 2 months, and again at 6 months) (Weinstock *et al.* 1996).

Act

Two *checklists* have been proposed to aid the practitioner in the diagnosis of melanoma:

The ***ABCD rule*** stands for:

A = *Asymmetry:* One half of the naevus does not match the other.

B = *Border:* The edges are irregular, notched or blurred.

C = *Colour:* The colour is uneven, with various shades of brown, black and even blue or red present.

D = *Diameter:* The diameter is larger than 5 mm (the size of a pencil eraser)

The presence of any one of the above would be an indication for biopsy or at least the patient should be referred to a plastic surgeon or dermatologist.

The 7-*point checklist* differentiates between major and minor features:

Major features:

1. change in size
2. change in shape
3. change in colour

Minor features:

4. diameter more than 6 mm
5. inflammation
6. bleeding or oozing
7. mild itch or altered sensation

The presence of at least one major feature in a pigmented lesion should be an indication for removal (excision biopsy) and the presence of an additional minor feature should arouse suspicion.

Reassure

Reassure the patient and parent about

- typical benign lesions, such as freckles, café-au-lait spots, etc.
- lesions less than 3 mm in diameter, flat and unchanged
- lesions that have been present for less than 3 weeks

Track

Track all other lesions.

Caustic burn wounds of the oesophagus

19

JHR BECKER & ALP NUMANOGLU

As many as 80% of caustic burn wounds of the oesophagus occur in children under the age of 5 years. The agents are fluids or granules stored in cold drink bottles or in brightly coloured containers that appear attractive to a toddler.

19.1 Agents

- Strong alkalis, e.g. sodium and potassium hydroxide in liquid, granular or solid form, such as drain cleaners, washing powders and soaps
- Strong acids, e.g. hydrochloric acid and sulphuric acid which are used in batteries and swimming pools

19.2 Pathogenesis

The level of damage will depend on:

- type of agent
- concentration (pH)
- amount ingested
- duration of exposure:
  - acids cause immediate pain and are quickly spat out
  - alkalis are odourless and tasteless and cause damage before the protective reflexes come into play
  - powders and granules adhere to the mucosa and therefore burn more proximally and locally in the mouth and throat. The presence of food or fluid in the stomach will work against the caustic agent to some extent, unlike in an empty stomach

Alkalis cause liquefaction necrosis and saponification of membranes at a tremendous rate, e.g. 1 ml of a 30,5% solution gives full-thickness damage in the cat's oesophagus within 1 second. Acids cause coagulation necrosis, which is to some extent protective. Granules cling to the mucosa and cause deep local burns. The mouth, pharynx, oesophagus and stomach are burnt to a similar extent by acids and alkalis, unlike granules and powders which burn mainly the mouth, pharynx and proximal oesophagus.

19.3 Degrees of burn wounds

These are analogous to burn wounds of the skin.

First degree

Superficial, red, oedematous, heals spontaneously and completely.

Second degree

Damage throughout the mucosa into the muscle layers. Healing takes place with fibrosis and stenosis – particularly in the case of circumferential burn wounds.

Third degree

Transmural damage with perforation, mediastinitis and peritonitis.

19.4 General clinical profile

Early signs

- Often asymptomatic without signs in the mouth or oropharynx – this is no guarantee that no damage has occurred further down
- Persistent hypersalivation
- Stridor and hoarseness
- Dysphagia and odynophagia
- Epigastric pain
- In the case of third degree burns, perforation

■ MEDIASTINITIS

- tachycardia
- tachypnoea
- dyspnoea
- stridor
- shock

- abdominal muscle rigidity
- positive rebound
- pain, etc.

Later signs

- Stenosis of the oesophagus and/or stomach
- Weight loss
- Dysphagia
- Lung complications

19.5 Mortality

With good management in a good medical unit, only ± 3% of cases die.

19.6 Morbidity

Fifteen to 25% of cases develop oesophageal strictures.

19.7 Diagnostic studies

There is poor correlation between symptomatic signs and true pathology, and it is for this reason that special investigations are recommended.

Oesophagogastroduodenoscopy

This must be done under general anaesthesia because the oesophagus and stomach are friable and oedematous, and can easily perforate in a struggling child.

The aim of the endoscopy is to determine the extent of the burn wound rather than the depth. Only first degree burn wounds can be accurately determined with endoscopy as it is not endoscopically possible to distinguish between second and third degree burn wounds.

Radiology

- Contrast studies are not of much value in the acute phase, but they are of value in the follow-up management of the patient to indicate the presence of strictures.
- Survey chest and abdomen X-rays are important in the acute phase to indicate the presence of perforation, e.g. free air in the mediastinum or in the abdomen.

19.8 Emergency management

Neutralising agents are of no value as:

- the heat produced can cause further damage
- the damage takes place so rapidly that there is no time to neutralise the agent before full-thickness damage has occurred

All that may help is for the patient to drink large volumes of water or milk immediately to rinse away and dilute the agent. No emetic must be given because this may once again expose the oesophagus and orophaynx to the agent.

19.9 Recommended management

Early

- Emergency management as above
- Intravenous infusion and resuscitation
- Nothing by mouth
- Intubate and ventilate if the airway is compromised
- General anaesthesia and endoscopic evaluation of the extent and degree of damage: evaluate the mouth, throat, oesophagus, stomach and duodenum
- Broad spectrum antibiotic for 3 weeks
- Emergency surgery if there are signs of perforation
- Follow-up contrast studies to determine the presence of strictures (video oesophagogram) at 2–3 weeks
- Parenteral feeding from the outset until the enteral route can be used again
- Dilate a single, short stricture
- When long and multiple strictures do not respond to dilatations, the oesophagus is removed and replaced with stomach or colon
- A stomach stricture is excised, bypassed or a stricture-plasty (a V-Y plasty) can be performed

Long-term follow-up

It is assumed that the patient's problems are now under control and he or she is able to eat normally. Bear the following points in mind:

- These patients have a 1 000–3 000 times higher risk of developing oesophageal cancer.
- There is a better prognosis than for the traditional oesophageal carcinoma and it develops approximately 40 years later.
- Regular life-long surveillance is needed.
- Prophylactic oesophagectomy and bypass may be required.

Foreign bodies that are swallowed

20

JHR BECKER & LZ MARCISZ

This is a very common occurrence amongst children because they place foreign bodies in their mouths to suck on them.

The objects are very varied: money, pins, bottle tops, button batteries, nails, bones, fish bones, even sticks have been swallowed.

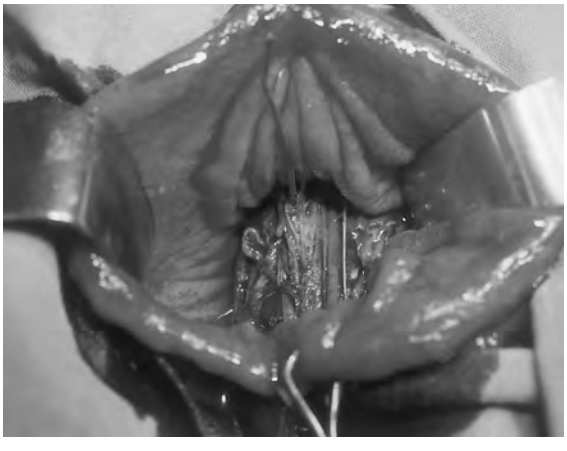


Fig. 20.1 Psychological patient eating grass, sticks, plastic, etc. – open stomach containing the foreign bodies.

Fig. 20.2 Foreign bodies that were taken out of the previous patient's stomach (the cigarette box simply demonstrates the size).

20.1 General

A foreign body that has spontaneously passed through the oesophagus will also pass through the digestive tract, except if it is too long to manage the curvatures of the duodenum, ligament of Treitz and ileocaecal valve.

A foreign body that progresses through the body without symptoms must be left alone as it will be passed spontaneously.

However, any object that remains stationary or causes symptoms must be removed – surgically or endoscopically. In the latter case, the object must be within the reach of an endoscope and an experienced endoscopist must conduct the removal.

20.2 Oesophagus

Common places where foreign bodies lodge:

- crycopharyngeus
- aortic arch
- epiphrenic area

Management of the foreign body

- The patient (or the parent) tells the doctor how the foreign body came to be swallowed.
- The nature of the foreign body will determine whether it is visible on X-rays of the chest and abdomen.
- If it is visible on X-rays and is stuck, then it must be removed.
- If it is not visible on X-rays and the history and symptoms are convincing, then the patient must receive an endoscopy.
- An endoscopy is conducted under general anaesthesia because this reduces the risk of perforation due to the child struggling.
- The foreign body is removed.
- After removal, the oesophagus is again inspected for possible perforation.
- Foreign bodies that are particularly hazardous with regard to perforation are toothpicks and other bones, especially fish and chicken bones.
- If perforation is suspected, then contrast radiology must be performed and a thoracic or paediatric surgeon is consulted.
- If the foreign body is lodged in the pharynx or proximal oesophagus, an attempt can be made to remove it under radiological control with the inflated balloon of a Foley urological catheter.

20.3 Stomach

Objects in the stomach that have passed spontaneously through the oesophagus will pass through the digestive tract, except if they are too long for the curves lower in the tract.

Indications for endoscopic removal:

- The expertise for endoscopy is available.
- Sharp objects, e.g. pins, blades, etc., are involved.
- Button batteries have been swallowed.
- Long objects, e.g. wire, nails, spoons, sticks, etc., are involved.

- There are foreign bodies that have remained stationary for a long time: the danger is that if left alone, they may become embedded inside the stomach and perforate it later.

20.4 Duodenum and small bowel

Common places where foreign bodies lodge:

- duodenal loop
- ligament of Treitz
- ileocaecal valve

Objects that are too long, e.g. wires, nails and sticks, become stuck and may perforate the organs. They must be removed by means of a laparotomy.

20.5 Colon

Foreign bodies seldom lodge here due to the calibre of the colon and the fact that the objects have already passed through the rest of the digestive tract.

Colonoscopy is of value if problems occur.

20.6 Button batteries

- They must be removed if they become stuck in one place and remain stationary.
- If a button battery lodges in the oesophagus, then it must be removed very quickly.
- If the battery is in the stomach, then it is only necessary to remove it if it does not progress and the expertise is available.
- In the rest of the digestive tract, the battery will progress on its own, especially if the passage is assisted with liquid paraffin.
- If the battery appears to be breaking up on radiological images, then urine and blood specimens must be taken to determine whether the patient has developed mercury poisoning or heavy metal poisoning.

Paediatric plastic surgery

21

JAN J VAN WINGERDEN

21.1 Introduction

Paediatric plastic surgery deals with the repair of the ravages of trauma (see Chapter 10) and the correction of numerous congenital anomalies. The many challenges faced range from finding creative and technically feasible reconstructive solutions to determining the ideal moment for implementation, which will not be too early and harm the child but will be early enough to encourage further, normal growth. The rewards are immense: a normally functioning child, who is accepted by, and feels accepted by, society.

Some of the more familiar or life-threatening congenital anomalies are highlighted.

21.2 Craniofacial anomalies

Skull anomalies, either in size, shape or both, usually present during the first year of life. They may be harmless but could indicate a congenital abnormality such as a craniostenosis, especially if accompanied by a number of symptoms. These include changes in behaviour (e.g. increased drowsiness or irritability), feeding-related problems (poor intake or projectile vomiting) and seizures.

Change in size

The infant's head may appear *uniformly enlarged* (for example, in hydrocephalus) or *unilaterally enlarged* (for example, due to a cephalohaematoma).

In other instances, the head may appear uniformly smaller (for example, in microcephaly) or unilaterally smaller (for example, in craniostenosis).

Although the exact measurement should always be taken, the following may serve as an *aide-mémoire* for average skull circumferences at the 50th percentile:

Birth	34,5 cm
6 months	43,5 cm (i.e. 34 in reverse)
(×2) 12 months	46,5 cm (i.e. 43,5 + 3)
(×2) 24 months	49,5 cm (i.e. 46,5 + 3)

Change in shape

Immediately after delivery, mild asymmetry may be the result of skull bone overlapping during birth.

In the developed world, the commonest cause of a misshapen head during the first year of life is *positional plagiocephaly* (Greek *plagio* = oblique and *cephale* = head). The occipitoparietal flattening develops secondary to the infant always being placed to sleep in the same supine position. This, in turn, has arisen from a misunderstanding of a finding by some researchers who linked SIDS (sudden infant death syndrome) with infants placed with, or preferring to sleep with, their faces down.

Other causes of a flat head, such as found in the *hypotonic infant* (Freeman and Carson 2003) and the highly uncommon premature fusion of the lambdoid suture (*'true' lambdoid synostosis*) (Huang *et al.* 1996) should be excluded (see below). In the latter, the hair growth tends to be normal over the whole scalp in comparison with the scanty growth on the side that the infant with positional plagiocephaly lies on, the ear is displaced posteriorly (instead of anteriorly such as in positional plagiocephaly) and, often, a thick ridge is palpable over the fused suture compared with the infant with positional plagiocephaly in whom the sutures are open. *Torticollis* (Latin *torti* = twisted and *collum* = neck) is occasionally found, either as a contributing factor (e.g. the congenital muscular torticollis) or as an associated factor (e.g. the non-muscular, craniovertebral types).

A radiograph only may suffice to confirm the diagnosis of positional plagiocephaly but, should there be any doubt, the infant should be referred to a specialist centre for further evaluation and computed tomography scans.

Once the diagnosis of positional plagiocephaly has been established, reassurance and prevention of further progression of the condition are the most important steps. These include changing and maintaining the changed position of the baby's head with the aid of a rolled towel on the ipsilateral side, or having a special, small foam rubber pad carved by a therapist (Freeman and Carson 2003). Physiotherapy may be useful in the treatment of congenital muscular torticollis. Severe cases may benefit from cranial orthosis, i.e. with the aid of a remoulding helmet.

Other, unusual craniofacial anomalies

These will be discussed under the following headings:

Craniostenosis
Craniosynostosis syndrome
Orbital displacement
Craniofacial clefts
Encephalocoele
Mandibulofacial dysostosis
Hemifacial microsomia

■ CRANIOSTENOSIS

This refers to an inability of the cranium to accommodate the growing brain. The term is limited to congenital (primary) premature closure of one or more of the skull sutures, a condition known as craniosynostosis. The diagnosis is made after a complete history has been taken, followed by a careful and thorough clinical evaluation. It must be distinguished from secondary causes of premature closure of the sutures, such as various metabolic, haematological or endocrine abnormalities, as well as cranial trauma, which may also require early intracranial decompression.

The origin of craniostenosis is currently attributed to a dysmorphology of the skull base (*Theory of Moss*). The physical consequence, known as *Virchow's Law*, implies that:

- Normal growth ceases perpendicular to the suture.
- Compensatory growth takes place parallel to the suture involved.

In this way the following abnormalities, for example, may develop:

Scaphocephaly (long axis from front to back)

- Sagittal suture synostosis – the most common form of craniostenosis

Plagiocephaly (flat head)

- Frontal: synostosis of one half of the coronal suture
- Occipital: synostosis of the lambdoid suture

Trigonocephaly (long axis is vertical)

- Synostosis of the suture between the frontal bones, the metopic suture

■ CRANIOSYNOSTOSIS SYNDROME

Although suture synostosis forms an important part of this condition, different sutures may be involved in different patients with the same syndrome. A specific pattern of genetic inheritance may be present and genetic assessment and counselling are necessary.

Crouzon's syndrome

Autosomal dominant. Consists of craniosynostosis with:

- mid-face hypoplasia
- exophthalmos

The hand is usually normal.

Apert's syndrome

Autosomally dominant, but more often sporadic; arises from mutations in the fibroblast growth factor receptor 2 (FGFR2) (Ibrahimi *et al.* 2005). FGFRs have been implicated in several craniosynostosis syndromes and skeletal disorders.

Usually presents with:

- high skull dome which flattens posteriorly (due to bicoronal synostosis)
- mid-face retrusion with orbital proptosis
- syndactaly of all four limbs (especially of the hand)

In general, it could be stated that the more sutures involved (synostosed), the higher the risk of raised intracranial pressure, visual disturbances and mental retardation.

A high index of suspicion is required to diagnose raised intracranial pressure timeously. Late signs of raised intracranial pressure are:

- Frequent, projectile vomiting
- Papilloedema (on fundoscopy)
- Skull radiography may show a 'beaten copper' appearance (also known as thumb-printing)
- Early referral of these cases will allow for adequate planning of the reconstruction by a multidisciplinary team

■ ORBITAL DISPLACEMENT (TELORBITISM)

Telorbitism refers to an abnormally wide distance between the orbits and is etymologically preferred above the old term 'hypertelorism', which accentuates the distance between the eyes and not the true pathology, which is really an increased interorbital distance.

The average interorbital distance (IOD) at birth is 16 mm (Hansman 1966). Seventy per cent of the adult IOD is achieved by the age of 2 years (Laestadius *et al.* 1969). Normal IOD in women is on average 25 mm and in men 28 mm. As telorbitism can occur unilaterally, the distance from the centre of the bridge of the nose to each posterior lacrimal crest is measured.

Seventy per cent of the adult inter-orbital distance is achieved by the age of 2 years. Normal inter-orbital distance in women is on average 25 mm and in men 28 mm. As telorbitism can occur unilaterally, the distance from the centre of the bridge of the nose to each posterior lacrimal crest is measured.

Telorbitism must be distinguished from *telecanthus*, which is lateral displacement of the medial canthi and may be post-traumatic or congenital, e.g. as part of Waardenburg's syndrome, blepharophymosis or canthus inversus, etc., in origin.

The cells of the anterior ethmoidal sinus are overexpanded but not those of the posterior ethmoidal cells or sphenoid sinus. Furthermore, the smaller wing of the sphenoid bone, in which the optic foramen lies, is also not affected. The combination of these factors, amongst others, allows orbital replacement surgery to be done without the danger of optic nerve injury.

■ CRANIOFACIAL CLEFTS

These are clefts other than those of the lip or palate only. They occur in approximately three per 100 000 births. There is no fixed pattern of inheritance. In some cases, amniotic bands are blamed.

Tessier (1976) summed up his observations and incorporated them into a classification, the main points of which are as follows:

- The clefts can involve the face or the cranium (skull) or both.
- A fixed pattern is observed in the natural course of the cleft.
- The origin of the cleft is located at the skull base.
- Not all tissues are affected to the same degree.
- Soft tissue defects are most commonly located between the midline and the infra-orbital foramen.
- Bony impairment is more commonly located from the infra-orbital foramen to the temporal bone.
- Sometimes an underlying cleft is only betrayed by an abnormal hairline or sclerodermic skin marking.
- Clefts do not run through or include neurovascular bundles.

■ ENCEPHALOCOELE

This is readily recognisable as a soft tissue mass and occurs particularly nasofrontally and naso-ethmoidally. A small encephalocoele must be distinguished from a dermoid cyst (see p 33). The patient with an encephalocoele must preferably be referred as soon as discovered due to the danger of ulceration and rupture, with a consequent risk of intracranial infection.

■ MANDIBULOFACIAL DYSOSTOSIS

This is also known as *Treacher-Collins-Franceschetti syndrome*. It is an autosomal-dominant inheritance with a genetic basis on chromosome 5. The face is bilaterally and *symmetrically* affected, with hypoplasia of the malar and maxillary bones and zygomatic arches. The condition is not life-threatening, except in some cases where the airway is threatened by a combination of maxillary atresia and mandibular hypoplasia. Repositioning and/or reconstruction of the mandible, maxilla and zygomas, as well as of the lower eyelids, is required.

■ HEMIFACIAL MICROSOMIA

In contrast to mandibulofacial dysostosis, this condition has no specific inheritance pattern and usually affects the face unilaterally (if bilateral, the deformity is *asymmetrical*).

The acronym OMENS (suggested by Vento *et al*. 1991) indicates the five major manifestations of hemifacial microsomia and is also used for classification purposes: ***O***rbital distortion (all the bones involved); ***M***andibular hypoplasia; ***E***ar defects, e.g. microtia (defective development of the external ear); ***N***erve involvement (facial nerve deficits); ***S***oft tissue deficiencies. Reconstruction is planned around the severity of the structures involved.

Surgical management of craniofacial conditions

■ AIMS

- Repositioning of the structures involved to stimulate normal growth and development
- Immediate relief of the threatened airway and raised intracranial pressure (where present)

■ PRINCIPLES

- Repair bony elements first and then soft tissue.
- Obtain satisfactory, complete exposure with minimal facial scarring by using special approaches, such as bicoronal, transconjunctival and oral incisions.
- Bone segments can be mobilised in any direction.
- Complete rigid stabilisation after mobilisation is essential.
- Additional bone (also from the cranium) is usually required for:
  - stability
  - filling of defects
  - smooth contours.
- Regular long-term follow-up is essential.

21.3 Cleft lip and palate

This is a mid-face abnormality with attendant functional, growth and cosmetic problems, each of which condition requires attention in its own right or in combination.

Classification

■ CLEFT LIP

- Incomplete (partial, vertical lip shortness with an intact nasal floor)
- Complete (cleft through the entire lip, including the nasal floor; with/without alveolar cleft)

The defect may present unilaterally or bilaterally, with or without a cleft palate.

■ CLEFT PALATE

Various classifications are available. It is important is to ascertain whether the cleft extends through either the soft or hard palate only, or through both.

The *prepalate*, i.e. all the structures anterior to the incisive foramen, including the alveolus, upper lip and columella, and the tip of the nose, is either unilaterally or bilaterally involved.

A ***submucous cleft*** should be suspected in the child who presents with a bifid uvula. A notch is palpable in the posterior edge of the hard palate. Despite an otherwise normal appearance of the palate, these children may develop hearing problems and, in approximately 15%, velopharyngeal insufficiency (see below).

Surgical timetable

0–3 months	Lip and nose repair
5–12 months	Palate repair (one procedure)
Up to 5 years	Small lip adjustments (where necessary)
6–9 years	Pharyngoplasty for nasal escape
9–11 years	Alveolar bone transplantation Maxillary osteotomies
15–18 years	Small nasal adjustments (where necessary)

Pierre Robin sequence

This is an uncommon condition that requires *emergency management* of the airway. The abnormalities include:

- micrognatia (mandibular hypoplasia)
- glossoptosis (falling back of the tongue)
- cleft palate

These patients die due to respiratory distress.

Emergency management includes ('N2T2'):

- Nurse sitting up in a forward position
- Nasotracheal intubation
- Tongue-to-lip adhesion (see below)
- Tracheostomy

Cleft lip

- If only the lip is involved, neither breathing nor feeding should pose a problem.
- Where a cleft lip occurs in combination with a cleft palate, a better result is obtained with
  - pre-operative orthodontics
  - an initial surgical lip adhesion procedure.
- Both provide a better bony platform upon which a later lip reconstruction can be performed.

■ TIMING OF REPAIR (RULE OF 10s)

- The infant is at least 10 weeks old
- with a mass of, at least 4,5 kg (10 pounds),
- a haemoglobin of more than 10 g/dl, and
- a white cell count of less than 10 000

Definitive reconstruction includes careful repositioning of the displaced and hypoplastic structures, namely:

- skin and vermilion
- orbicularis oris muscle
- hypoplastic nostrils
- displaced nasal septum

■ AIM OF REPAIR

The aim is *improved appearance*, recognised by:

- (an almost) normal appearance of the *lip* and *nose*
- minimal growth inhibition of the mid-face

Cleft palate

It is essential to realise that most of the controversies surrounding the surgical management of the cleft palate revolve around how best to achieve the aim of an ideal repair.

■ AIM OF REPAIR

The aim is *improved function*, in terms of:

- normal speech, which is achieved by complete closure of the cleft and optimal velopharyngeal closure
- minimal growth inhibition of the mid-face

Techniques have been described, ranging from straight-line closures to Z-plasties, all taking special care to realign the muscles of the soft palate. It is essential to realise that this is not only a tissue deformity, but that existing structures, i.e. the velar muscles, are also displaced with consequent abnormal implantation at the back of the hard palate. Reconstruction includes the dissection of the velar muscles and suturing in the midline of the reconstructed soft palate. Aggressive stripping of the periosteum and oral mucosa from the hard palate can result in scarring and can harm the growth potential of the maxilla. Extremely careful surgery is therefore of the utmost importance.

Of interest to the primary care physician are the following problems and complications, which can be expected in these patients:

■ MAINTENANCE OF AIRWAY

This is critical where mandibular development is unsatisfactory, for example with retrognatia or micrognatia, such as in the Pierre Robin sequence. These newborns must be nursed on their sides in a slanting chair and referred to a plastic surgeon as soon as possible.

During transfer, the availability of an oropharyngeal airway and competent nursing staff are essential.

Often the surgeon will perform a tongue-lip adhesion procedure (modified *Douglas-Routledge procedure*) whereby the tip of the tongue is fixed to the inner surface of the lower lip, so helping to prevent glossoptosis and avoid the need for a tracheostomy.

■ FEEDING

Feeding of the baby is the most important aspect that the practitioner should attend to. In the presence of a cleft palate, the newborn will not be able to suck efficiently. However, the swallowing mechanism does function. It is recommended that the mother is actively involved in developing a method by which milk is effectively administered. In this way, she will become the expert in feeding her child and a healthy mother-child relationship will develop which will quickly override the initial shock of a 'malformed' baby. Counselling by the general practitioner and an experienced nurse is advisable without the mother feeling that they are dominating her – at this stage, the baby is very dependent on the mother.

Feeding must be done in a semi-sitting position – this applies to breast-feeding as well. With bottle feeding it is advisable to enlarge the teat opening, allowing for an easier flow of milk into the child's throat. The angle at which the bottle is held will determine the flow rate. A careful balance between too fast and too little flow will be developed between mother and child. Naturally, more air will be swallowed, feeding will take longer and the child will need to be held upright longer to get rid of winds.

A feeding plate (also known as an 'obturator') made by an orthodontist could be of value to separate the nose and oral cavity and to guide both dental arches to a more favourable position before surgical closure of the palate. However, the obturator will have to be regularly altered as the child grows. A growth percentile chart is indispensable in monitoring whether the child is receiving adequate nutrition.

■ OTITIS MEDIA

The abnormal course and implantation of the tensor palati and levator palati muscles from the Eustachian tube onto the palate of cleft patients prevent normal drainage of the middle ear. The consequence is fluid accumulation, increased incidence of middle ear infections and, later, conduction deafness.

Active, immediate management of all possible ear infections is necessary. Decongestants, antibiotics, myringotomy and grommets may be indicated. Regular inspection of the eardrums during each consultation is essential.

■ SPEECH

Normal speech depends on a competent speech mechanism (achieved through successful palatal closure and a good muscle

sphincter), normal hearing, intelligence, and motivation of the patient in the home and school environments that encourages correct speech.

The normal mobile and muscled soft palate (*velum*) moves upward and backward during speech. Together with the lateral and posterior walls of the pharynx, it forms the velopharyngeal sphincter. A normally functioning velopharyngeal sphincter will prevent air from escaping through the nose during speech.

Even with a good palate repair, a certain amount of air may still escape from the nose, which the child attempts to overcome by movements of the tongue.

If the velopharyngeal closure is ineffective, allowing for excessive nasal escape or hypernasality (*rhinolalia*) during speech, it is known as *velopharyngeal insufficiency* (VPI).

The extent of the VPI will determine the need for and the type of surgical procedure required to correct the problem. The following are necessary:

- Intensive speech and hearing assessment
- Nasendoscopy
- Videofluoroscopy

VPI is repaired, when indicated, ideally between the ages of 5 and 12 years, either by means of a *pharyngeal flap*, a *sphincter pharyngoplasty* or a re-repair of the palate, depending on where the weakness in the sphincter mechanism lies (Jackson 2005).

CAVEAT

It is critical that the tonsils or adenoids are not removed in cleft palate patients without a complete evaluation by the 'cleft team'. Apart from the plastic surgeon, this team includes, at least, a speech therapist and an ear, nose and throat specialist. The danger is that such an operation will enlarge the oronasal cavity, with worsening of the speech abnormality.

■ ORTHODONTICS

An orthodontist assesses and quantifies the growth and development of the upper and lower jaws in these patients. He or she also ensures that the teeth and dental arches are optimally aligned for growth. Where the growth deformity is so great that orthodontic treatment alone is not sufficient, then the patient will have to undergo some osteotomies when the maxillary or mandibular growth is complete.

■ THE NOSE

This is the most neglected anatomical part in cleft patients because surgery is so often performed by surgeons who are not trained in all aspects of facial surgery. The patient ends up with a closed lip but also a flat nose and deviated septum, which are difficult to improve at a later stage.

The best time to address the rotated, hypoplastic alar cartilage and nasal floor is during the primary repair of the cleft lip. If done properly, it could simplify further nasal reconstruction at a later stage significantly.

21.4 Congenital deformities of the ear

A normal external ear is characterised by the following (from the 'outside in'):

- helix (a)
- antihelix (d), which divides into a superior (b) and inferior crus (c)
- concha (e)
- tragus (f), which serves to protect the external auditory canal
- lobule (g)

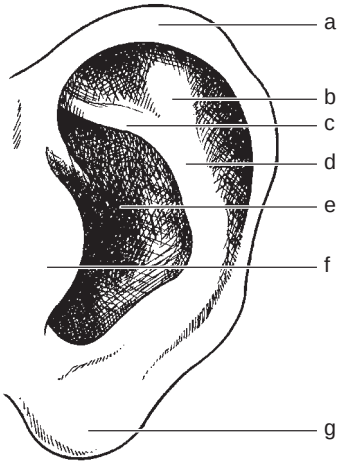


Fig. 21.1 Diagram of the normal ear.

Development of the ear

One of the commonest ear-related anomalies the general practitioner will encounter is pre-auricular tags (PATs) and pits. The ear develops from six embryonic hillocks, which fuse to form the external ear. PATs represent accessory hillocks and therefore often contain cartilage.

The presence of one or more PATs may indicate:

- an associated urinary tract anomaly – a renal ultrasound should be performed, especially if a positive family history of deafness, renal malformations or maternal gestational diabetes is present (Kohelet and Arbel 2000; Wang *et al.* 2001)
- a hearing impairment – audiometry (hearing assessment) is recommended (Kankkunen and Thiringer 1987; Kugelman *et al.* 1997)

CAVEAT

The enthusiasm to attempt to simply tie off a PAT, immediately after birth, should be suppressed as this commonly leaves an unsightly scar that will require revision. A small titanium clip, placed by a plastic surgeon, would leave a neater scar (in suitable patients). This can be performed under local anaesthetic. PATs with a broader attachment require excision.

The most challenging deformity of the ear is partial or complete absence of the external ear or *microtia*. The lobule, embryologically

the last structure to form during normal ear development, is usually present.

Microtia can occur as a solitary anomaly or as a part of hemifacial microsomia (see above) (Bennun *et al*. 1985). A complete assessment is required to exclude associated mandibular, condylar and middle ear abnormalities. Ordinary radiographs may suggest the presence of a middle ear. The absence of a middle ear on the one side makes confirmation of a properly functioning middle ear on the other side essential. An early hearing assessment is advisable.

Reconstruction of the external ear is not embarked upon before the age of 5 years when the other, normal ear has attained approximately 85% of its adult size. Regarded by some as the most challenging of all reconstructive procedures, it entails the sculpting of an ear framework from rib cartilage, covering it with a local, fascial flap and, if necessary, a full-thickness skin graft.

Position of the ear

With regard to the position of the external ear, one must distinguish between an underdeveloped ear (microtia) that appears optically to be set too low, and a relatively normal ear that is set too low and can be indicative of a rare congenital anomaly like bilateral renal agenesis, known as Potter's syndrome.

Shape of the ear

The abnormally shaped ear lends itself to conservative measures immediately after birth because the ear cartilage is still supple as a result of the influence of maternal oestrogen and the ear muscles are still poorly developed. It is thus advisable that a newborn with a malformed external ear is referred to the plastic surgeon as soon as the abnormality is recognised. The surgeon will attempt to influence the still soft malformed ear favourably using external splints. The earlier this is begun, the greater the chance of success.

Prominent ears

Prominent ears are caused by:

- absence of the superior crus of the antihelix
- absence of the antihelix and both crura
- abnormally enlarged concha
- a combination of the above

This is a congenital condition that, for the reasons set out, should be correctable when the child is older. In the meanwhile, it can cause definite emotional problems if the parents and the general practitioner do not provide careful guidance and support.

The specialist's first duty is also guidance. The very young child should be ensured that this condition could be corrected properly whenever they are ready. The parents, however, should be assured

that a successful result requires the patient's compliance – this is why it is advisable to wait until the child is old enough to co-operate but still young enough to avoid permanent psychological problems.

A second important consideration is that the ear reaches almost full adult size at the age of 6 years and then a more permanent operation can be performed.

The parents are requested to make the ear abnormality a point of as little debate as possible and, where necessary, the child must learn to defend himself or herself psychologically prior to reconstruction.

Usually, the ears are not equally prominent which, together with the identification of specific anatomical abnormalities, requires specialised surgical correction for each ear. The operator must be not only technically skilled, but also possess a good sense of aesthetics and balance.

21.5 Congenital hand deformities

A congenital hand deformity is usually diagnosed on the first complete examination of the newborn.

The next step following the recognition of the hand deformity should be to exclude any other congenital anomalies. The assistance of a paediatrician, especially a neonatologist, is indispensable. Furthermore, most parents would request genetic counselling which should be provided at the earliest opportunity.

Referral to a plastic (hand) surgeon with a special interest in this type of surgery must take place as soon as possible. A thorough, unhurried examination is conducted by the surgeon (usually assisted by a hand therapist). Maximum information is required to form a complete image of the anatomical defect and to be able to present the parents with a realistic treatment plan.

CAVEAT

It should be emphasised to the parents that good function, rather than appearance, is the standard according to which success will be measured. The newborn with a congenital hand deformity will have far fewer problems with the deformity than the parents will.

The surgeon has three main objectives:

- Optimise spatial orientation and placement of the hand.
- Provide good skin cover with satisfactory sensation.
- Obtain a good power grip and precision handling.

There are also three factors that determine the surgeon's timetable:

- If growth is impaired – repair within 12 months.
- If the developmental pattern (of hand use) requires it – repair before 30 months.
- If patient compliance (post-operatively) is necessary – repair before 60 months.

Classification (Swanson 1976)

The international classification of congenital hand deformities is given here in a simplified form.

I Defective development of parts (transverse or longitudinal)
II Defective differentiation of parts (for example, syndactaly)
III Duplication (for example, polydactaly)
IV Overgrowth
V Undergrowth
VI Constriction ring syndrome
VII General skeletal defects

The most common abnormalities are syndactaly and polydactaly. One of the more frightening, that of constriction ring syndrome, will be discussed in more detail.

Polydactaly

This is the commonest congenital hand abnormality, which in Africans occurs in as many as 1 in 300 live births. In Caucasians and Asians, the incidence is 1 in 3 000 births (Flatt 1994).

The ulnar form of polydactyly is much more common in Africans in comparison with radial (thumb) duplications, which have a higher incidence in Caucasians and Asians.

(Note: When discussing the hand, radial, central and ulnar aspects are preferred to the terms pre- and postaxial).

■ DESCRIPTIVE CLASSIFICATION (STELLING AND TUREK),

(as quoted in Flatt 1994)

Type I Duplication consists of soft tissue only

Type II Duplication is incomplete with normal anatomical structures; partially articulates with either the phalanx or metacarpal

Type III Complete duplication with separate metacarpals

Thumb duplications (as an example of a radial duplication) are usually rather complicated to reconstruct and referral should preferably be done before the first year of life so that surgery can be performed and completed before the child reaches school-going age.

Central duplication involves the ring finger in at least 50% of cases. Especially in the case of a type II deformity, it is associated with syndactaly to form a so-called 'hidden' syndactylous polydactaly, the management of which also requires special skills.

CAVEAT

Ulnar duplication must always serve as a warning of associated anomalies.

■ MANAGEMENT

This always entails the surgical removal of either a part of or the

whole duplication. Where partial removal is performed, the retained tissue is used to augment or replace part of the remaining digits. A type I duplication should rather be surgically amputated as the old-fashioned method of tying off often leaves a wart-like lesion which may later require surgical excision.

Syndactaly

This is recognised by a web between at least two fingers which, in practical terms, is described as being:

- *Complete:* Fingers are joined along their entire length.
- *Incomplete:* Fingers are joined from the distal half of the proximal phalanx up to the proximal half of the distal phalanx.
- *Complex:* Fusion of the bones (synostosis) is present.
- *Simple:* Only soft tissue is involved.
- *Acrosyndactaly:* Distal fusion with proximal fenestration.

Apart from the above, in a thorough examination attention will also be paid to the nails (as part of the syndactaly), the length of the fingers (they may be shorter as in brachysyndactaly) and the suppleness of the joints (or lack thereof, e.g. in symphalangism).

Where syndactaly occurs alone, it is inherited as a dominant gene with a decreased penetration and varying manifestation so that 10–40% of these patients have a positive family history and 50% of the cases are bilaterally symmetrical.

The usual involvement of the four interdigital spaces from thumb to little finger is 5% (between thumb and index finger, etc.), 15%, 50% and 30%.

Early surgery is essential:

- in acrosyndactaly
- where fingers are of different lengths (for example thumb – index finger) in order to prevent rotation and flexion abnormalities of the longer finger

■ SURGICAL PRINCIPLES

- Interdigital webs are reconstructed with local *flaps*.
- Full-thickness skin transplants are always needed distally.
- *Z-shaped incisions* prevent contractures during further growth.
- The shorter the finger, the more proximal the new web should be made (but not proximal to the metacarpophalangeal joint).
- Release only one side of a finger at a time.
- Bilateral surgery is desirable in order to limit the number of procedures necessary.

Constriction ring syndrome

This is a rare, sporadic phenomenon, which in some cases requires urgent action and can present as one of the following:

CLASSIFICATION (PATTERSON 1961)

- Simple constriction ring(s)
- Rings with associated distal deformity, with or without lymphoedema
- Rings with distal fusion (acrosyndactaly)
- Amputations

There is much speculation about the origin of these deep grooves, which encircle the limb partially or completely: both exogenous and endogenous factors have been blamed.

Kino (1975) was able to elicit a similar condition in laboratory animals with amniocentesis and early uterine irritation, hence it was believed that the condition was due to so-called 'amniotic bands'.

TREATMENT

A careful examination will determine how urgent the need is for surgical intervention by noting particularly the levels of lymphoedema, circulatory and neurological function.

Possible surgery includes (alone or in combination) (Emmett and Morris 1998):

- excision of the groove with blood vessel and nerve decompression by means of a Z-plasty
- release of the syndactaly
- amputation of functionless 'mushroom' fingers
- resection of dorsal, overly thickened soft tissue to improve the so-called 'lollipop' appearance
- transposition of stumps on neurovascular pedicles
- lengthening of the phalanx or metacarpal by means of bone transplants

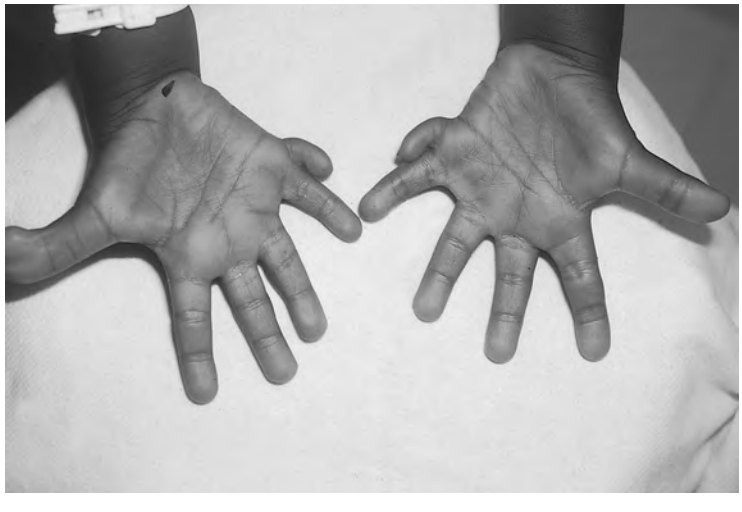


Fig. 21.2 Polydactyly.

Paediatric urology

22

D J DU PLESSIS & R WIERSMA

Symptoms and signs are an important part of evaluating a child's urogenital system. The following symptoms and signs may indicate disease affecting the kidneys, renal pelvis, ureters and bladder.

22.1 General symptoms

Fever, rigors, vomiting, weight loss, failure to thrive, as well as abdominal pain and fatigue may be symptoms of a variety of kidney and bladder conditions.

22.2 Specific symptoms of urinary tract disease

Urinary frequency

Frequent urination is normal in early childhood during which the bladder acts reflexically. Older children pass urine approximately four times a day. If urinary frequency persists, this may be an indication of bladder irritability due to an underlying urinary system or spinal cord pathology.

Nocturia

This is when urine is passed when the child is asleep at night and is not considered normal.

Urge urination

This is a strong urge to urinate, which precedes the physical passage of urine. This may indicate an infection of the urinary system, but may be seen with neurogenic conditions, such as myelomeningocoele.

Burning urine

This is the sensation of burning pain during the passage of urine. It usually indicates an infection of the bladder or urethra.

Bedwetting

This is when the child wets the bed during sleep. Until the age of about 3–4 years, this can be viewed as normal. If the bedwetting persists after 4 years, the child must be medically examined to exclude psychological problems or underlying abnormalities of the brain, spinal cord and urinary system.

Hesitancy

This occurs when the child, while attempting to urinate, has to wait a few seconds before spontaneous urination takes place.

Post-micturation dribble

This is when a few drops escape from the urethra after urination, wetting the underwear.

Urinary incontinence

This is spontaneous involuntary loss of urine which leads to wet underwear.

Oliguria

This is when the child passes less than 0,5 ml/kg/hour of urine per 24 hours. This may be the result of pre-renal hypovolaemia or may be due to renal impairment.

Pneumaturia

This is where the child passes air while urinating. This is the consequence of a fistula, congenital or acquired, between the colon and the bladder or is due to an infection of the urinary tract with a gas-producing organism.

Haematuria

This is the result of pathological conditions of the kidneys, ureters, bladder and urethra, which lead to blood in the urine.

Urine output

Normally, the child produces a minimum of 1 ml/kg of body weight/hour. If the child produces less, this would be a sign of pre-renal hypovolaemia.

Specific gravity

The renal function is dependent on the renal tubular concentrating ability, a function that improves with aging of the kidney. This can be measured by assessing the specific gravity of the urine; normal urine concentration is 1,005–1,020 mOsm. The total number of active particles is measured with an osmometer. Damage to the renal tissue leads to the loss of concentrating ability.

Serum urea

Urea is a by-product of amino-acid breakdown in the liver and is excreted by the kidneys. The capacity of the kidney to excrete urea is great and therefore a rise in serum urea levels is indicative of impairment of renal function.

Serum creatinine

This is a product of tissue creatine metabolism. It is not related to diet and is actively excreted by the kidneys with no reabsorption, and is therefore a good measure of renal function.

Ultrasonography

This is a safe investigation for assessing the urinary system. It can accurately measure the size and shape of the kidney. It should clearly demonstrate calyceal dilatation and hydronephrosis, the size of the renal pelvic, the size of the hydro-ureters, the thickness of the bladder wall and the prostatic urethral dilatation.

Intravenous pyelogram

This is an X-ray-based investigation, which utilises the concentrating ability of the kidney to excrete an intravenously injected, iodine-based compound, called 'contrast'. The morphology, and the concentrating and excreting ability of the contrast are investigated. Iodine compounds may cause allergic problems. Gonadal protections should be used wherever possible when using X-rays on children.

Intravenous pyelograms have largely been superseded by the use of CT scanning and isotopic renograms.

Radio-isotopic renogram

This is a safe method of acquiring quantitative functional information on the renal system. These studies, using different isotopes, can provide information on renal structure, perfusion, relative function and clearing from the kidney. A renogram can also measure the glomerular filtration rate (GFR).

Micturating cysto-urethrogram

This is an X-ray-based study in which the bladder is filled with an iodine-based fluid per urethral catheter, and the size, shape and contractility of the bladder are studied. This test is useful in the investigation of bladder outlet obstruction and vesico-ureteral reflux.

22.4 Congenital abnormalities of the urinary tract

A number of congenital abnormalities can occur in the child's urinary tract. Common among them are the renal pathologies and the obstructive uropathies. With the advent of ultrasonography during pregnancy, it is now possible to diagnose such congenital defects ante-natally.

22.5 Renal pathologies

Renal agenesis

Unilateral renal agenesis is found in approximately 1 per 1000 of the population. It is associated with a hemi-trigone of the bladder, which can be cystoscopically viewed. Due to the increased workload on the remaining kidney, compensatory hypertrophy may occur. In boys, agenesis of the kidney may be accompanied by an absent testis and seminal vesicle on the same side. In girls, this may be seen as a missing ovary and fallopian tube. Abnormalities of the uterus and vagina are occasionally seen.

The diagnosis is made coincidentally or during the investigation of a urinary tract infection. The missing kidney will be noted on the initial ultrasound and confirmed on isotopic renogram, and the hemi-trigone is seen cystoscopically.

Abnormalities in the remaining kidney must be managed. Contact sport must be avoided to prevent injury to the remaining kidney.

Malrotation of the kidney

During foetal development, the kidney moves from a pelvic position to the normal retroperitoneal position. With the upward movement there is a medial rotation of the renal pelvis. If the rota-

tion process is incomplete, a malrotation of the kidney follows. The most common abnormality is an anteriorly positioned renal pelvis. The abnormality is usually visible on ultrasound and an excretory urogram. Hydronephrosis is sometimes associated with this abnormality.

Treatment is only indicated for complicated cases with hydronephrosis or stones.

Pelvic kidney

During the ascent, the kidney passes through the arterial fork formed by the umbilical vessels. Failure to pass through this arterial fork leads to a pelvic or low-lying abdominal kidney. Patients with this abnormality present with an abdominal mass, vesico-ureteral reflux, urinary tract infections and sometimes obstruction at the pelvi-ureteral junction. This may be co-incidentally found during investigation for urinary tract infections.

The diagnosis depends on clinical suspicion which is confirmed with an ultrasonogram and isotope renogram. A voiding cystourethrogram can define the presence of vesico-ureteral reflux.

Treatment is only necessary if abnormalities are associated with the pelvic kidney.

Crossed renal ectopy

This is where one kidney moves over to the contra-lateral side during embryological development, and fusion takes place at the upper or lower pole of the contra-lateral kidney. Diagnosis is usually made co-incidentally during examination of a child with a urinary tract infection or renal trauma. Treatment is seldom necessary for these children.

Horseshoe kidney

The ascending kidneys may be pushed so close together by the umbilical vessels that the lower poles fuse. Due to the union, the inferior mesenteric artery prevents the normal upward migration of both kidneys and leads to the lower position of the 'horseshoe' kidney.

Horseshoe kidneys are uncommon. Patients may present with urinary tract infections due to the vesico-ureteral reflux, kidney stone formation or hydronephrosis. The diagnosis may be confirmed on ultrasonography, isotopic renography and CT scan. No treatment is needed in uncomplicated cases. Treatment of the complications, namely vesico-ureteral reflux, kidney stone formation and pelvi-ureteral obstruction, is managed in the routine manner.

Renal hypoplasia

This is a small kidney with normal renal parenchyma.

22.6 Cystic disease of the kidney

Renal dysplasia

This condition is usually associated with obstruction of the urinary tract during foetal life. Macroscopical cysts are present on the surface of the kidney. Histologically, the condition confirms the presence of primitive glomeruli, tubuli, tubes and cartilage. Renal dysplasia may be diffuse or be seen focally throughout the kidney parenchyma.

Multi-cystic kidney

This is a severe level of renal dysplasia, which can present unilaterally or bilaterally. Bilateral multi-cystic kidneys are incompatible with life. A unilateral multi-cystic kidney may give rise to a palpable loin mass in children. Ultrasonography shows a typical cystic kidney. A photopenic area is demonstrated on isotope renography. Atresia of the ureter is usually associated with a multi-cystic kidney. These kidneys involute with time – failure to involute or even growth in size are indications for nephrectomy. Prophylactic nephrectomy is seldom indicated.

Autosomal recessive (infantile) polycystic kidney

Infantile polycystic kidney disease is an autosomal recessive inherited condition that affects newborns. They usually present with bilaterally enlarged kidneys and an associated jaundice due to hepatic liver fibrosis. Owing to progressive disease of the renal parenchyma, these children die in renal failure. Boys and girls are equally affected. Ultrasonography of the kidneys confirms the bilateral diffuse kidney enlargement and prominent renal pyramids are also seen on excretory urography.

Due to the severe level of kidney and lung impairment, renal and respiratory failure follow in the early post-natal period. A few children survive for years, but later die of hypertension. Management involves treatment of the renal failure and hypertension.

22.7 Upper tracts

Duplication of kidneys, renal pelvis and ureters

Duplication of the upper urinary tract is one of the most common congenital abnormalities in the child. It is seen most commonly in girls, and in 20% of cases it is found bilaterally. The upper ureter, if it inserts into the bladder separately, will insert in a lower position than the ureter from the lower position.

Duplication of the ureters is classified into complete and incomplete duplication:

- *Complete duplication* exists when both ureters from the same kidney open on the ipsilateral side in the bladder.

- *Incomplete duplication* is when the two ureters from the same kidney join before entering the bladder. In these cases only one ureteral opening is found on the ipsilateral side; this joined ureter is referred to as a 'bifid ureter'.

The following conditions are frequently found in association with duplication of the ureters.

Vesico-ureteral reflux

Vesico-ureteral reflux is most commonly associated with duplication of the ureters. Due to the lateral ectopy and the short submucosal path, reflux usually takes place in the lower system. Vesico-ureteral reflux rarely occurs in the upper system.

Ectopic ureters

If the ureter does not open at the trigone, this is referred to as an 'ectopic ureter'. Ectopic ureters are usually associated with complete duplication of the ureters in girls, who seldom have a single ectopic ureter. An ectopic ureter in a duplicated system usually affects the upper pole ureter. Boys usually have single ectopic ureters. Both sexes frequently present with urinary tract infections.

If the ectopic ureteral opening appears in the proximal portion of the girl's urethra, it is associated with vesico-ureteral reflux, urinary obstruction or both. Sometimes the ectopic ureter may open in the vulva, uterus or vagina. Ectopic ureters situated distal to the external sphincter may be the cause of urinary incontinence.

In boys, ectopic ureters are seldom seen with a duplicated ureteral system, but, if present, the ureter opens in the prostatic urethra, *ductus ejaculatorius*, seminal vesicles or *vas deferens*. This condition is not the cause of urinary incontinence in boys. Surgical repair of proven ectopic ureters is indicated.

Ureterocoele

This is a condition in which there is distension of the terminal or intermediate portion of the ureter due to a weakness in the superficial portion of the trigone. This usually involves the upper pole ureter in a complete duplex system. This may also be seen in a single ureter system in boys. In a small percentage of patients, ureterocoeles may appear bilaterally.

If the ureterocoele occurs in the bladder, it is referred to as an 'orthotopic ureterocoele'. If it occurs at the bladder neck or in the urethra, it is referred to as an 'ectopic ureterocoele'. In these cases it may lead to the obstruction of urine flow and to infection.

The diagnosis is confirmed with ultrasound or a cysto-urethrogram.

The management of ureteral duplication depends on the presence of vesico-ureteral reflux or ureterocoele, as well as on the level of renal functional impairment. If irreversible damage of the upper

or lower segment has taken place secondary to an ureterocoele or vesico-ureteral reflux, then hemi-nephrectomy is indicated. The presence of reflux in the lower segment justifies surgical re-implantation of both ureters, since both make up a single ureter sheath. A ureterocoele that causes obstruction of the upper segment justifies excision and re-implantation of both ureters in the common ureteral system. Total nephrectomy is seldom indicated.

Mega-ureter

In this condition there is dilation of part, or the whole, of the ureter due to primary or secondary causes. A primary mega-ureter is indicative of vesico-ureteral junctional pathology.

A primary obstructive mega-ureter is caused by a functional obstruction at the vesico-ureteral junction. Although a urinary catheter can be passed with ease, it does lead to hydro-ureter and hydronephrosis. It is more commonly found on the left side and affects more boys than girls. It occurs bilaterally in about 20% of cases. The child presents with a proven urinary tract infection, with or without haematuria, sometimes accompanied by abdominal pain.

A secondary obstructive mega-ureter is caused by an obstructive uropathy, such as posterior urethral valves, a neurogenic bladder (e.g. from a myelomeningocoele) or abnormalities of a functioning bladder. In these patients the bladder hypertrophy leads to vesico-ureteral obstruction.

Other secondary causes of unilateral or bilateral mega-ureter are: vesico-ureteral reflux, diabetes insipidis, polyuria or bacterial infections of the urinary tract.

The condition is confirmed by means of excretory urography, with or without ultrasonography. The secondary renal impairment is determined by isotopic renography, which also determines the treatment. A cysto-urethrogram must be done to eliminate the possibility of vesico-ureteral reflux, as well as to confirm the presence or absence of an obstruction of the bladder outlet.

In most cases, surgical repair is indicated. However, if remodelling causes severe dilation of the lower part of the ureter, ureteral re-implantation is indicated. If there is an outlet obstruction, this must be repaired before surgical re-implantation is carried out. Lower grades (I–III) of vesico-ureteral reflux do resolve spontaneously with the growth of the bladder in a high percentage of cases.

22.8 Obstructive uropathies

Pelvi-ureteric junction obstruction (PUJO)

The ureteropelvic junction is the most common cause of urinary tract obstruction in children. These children present with an abdominal mass as a result of hydronephrosis. As a result of the routine use of ultrasonography during pregnancy, this condition is commonly diagnosed ante-natally.

The condition is more common among boys and more frequently seen on the left side. A contralateral ureteropelvic obstruction may be seen in 10–15% of cases.

The causes of ureteropelvic obstruction are:

- Intrinsic conduction defects, which lead to inadequate peristalsis
- Accessory blood vessels that usually run behind the ureteropelvic transition zone
- Secondary ureteropelvic reflux, seen with severe levels of vesico-ureteral reflux; in these cases a badly twisted ureter is usually present
- Stenosis, stricture, angulation, fibrosis or external pressure – these are rare causes of ureteropelvic obstruction

■ SYMPTOMS AND SIGNS

Where this condition has not been diagnosed ante-natally, it usually presents with nausea, vomiting and failure to thrive, as well as a palpable flank mass. In older children, there is intermittent abdominal pain, especially during a high fluid intake. Microscopic or macroscopic haematuria may be present. In 25% of children, there is a documented urinary tract infection. With severe bilateral hydronephrosis due to ureteropelvic obstruction, the first symptoms may be those of end-stage renal failure.

Diagnosis is made on ultrasonography and a diuretic isotope renogram confirms the presence of a unilateral or bilateral ureteropelvic obstruction. Badly affected kidneys will not concentrate sufficient contrast or isotope to show up the kidney on either an excretory or isotopic renogram. These would be seen as photopenic areas. If the ureters appear distended, a voiding cysto-urethrogram should be done to exclude vesico-ureteral reflux.

A flow pressure study (Whitaker test) can be performed to distinguish between a functional and a mechanical obstruction in the ureteropelvic transition zone. However, due to the invasive nature of this investigation (it requires percutaneous nephrostomy), it is not routinely done.

■ MANAGEMENT

- Pelvi-ureteral obstruction with small renal pelvis (<2 cm) and >40% function on the isotopic renogram may be managed conservatively.
- With a near-total obstruction at the ureteropelvic transition, or with deteriorating function shown on the isotopic renogram, the child will need an open pyeloplasty or endopyeloplasty.
- If the kidney has been totally destroyed, a nephrectomy is indicated.

Posterior urethral valves

This condition is the most common cause of bladder outlet obstruction in boys in early childhood. The inferior mucosal folds

emanating from the verumontanum obstruct the flow of urine from the time the foetus commences to produce urine.

■ SYMPTOMS AND SIGNS

Severe levels of obstruction may lead to renal dysplasia and uraemia. This, in turn, may lead to an associated oligohydramnios and hypoplasia of the lungs. The level of obstruction determines the symptoms of posterior urethral valves. Newborns may present with a dribbling urinary stream, urinary ascites, patent urachus and failure to thrive. They may be shown to have uraemia, high blood pressure and anaemia. Older boys present with documented urinary tract infections, as well as haematuria. Sometimes overflow urinary incontinence is present, with vague abdominal pain. In these cases a midline lower abdominal swelling is present due to a distended bladder.

■ INVESTIGATIONS

- Ultrasonography during pregnancy confirms the presence of posterior urethral valves if the following is found: thickening of the bladder wall with bilateral hydro-ureters and hydronephrosis. No active intervention during pregnancy is indicated to manage the condition.
- Ultrasonography in the newborn with neonatal uraemia may confirm enlargement of the prostatic urethra, thickening of the bladder with bilateral hydro-ureters and hydronephrosis.
- Excretory urography is seldom indicated in the newborn due to poor tubular function.
- A typical voiding cysto-urethrogram shows a trabeculated bladder with a prominent bladder neck, a dilated posterior urethra and hold-up of urine in the prostatic urethra.
- Urethral cystoscopy confirms the presence of the valves in the posterior urethra.

■ MANAGEMENT

The primary management is the correction of the obstructive uropathy and biochemical disturbance. Catheterisation of the bladder and intravenous fluid management, with or without antibiotics, must precede any investigation or therapeutic intervention.

The surgical treatment is endoscopic disruption of the posterior urethral valves. If this is not technically possible, then a cutaneous vesicostomy may be indicated.

If the uraemia does not clear on simple bladder drainage, a supravesicle decompression by means of a percutaneous nephrostomy or cutaneous ureterostomy is indicated. In a small number of patients, there is a serious renal dysplasia with an associated high mortality. Any vesico-ureteral reflux that does not clear up after the management of posterior urethral valves requires correction.

Hydrocalycosis

This term describes urinary dilatation of the renal calyces, associated with obstruction in the urinary tract. This may be diffuse or focal.

Diffuse hydrocalycosis occurs with urethropelvic obstruction or obstructed mega-ureters. Local or segmental hydrocalycosis occurs with infundibular stenosis, or intrinsic or extrinsic pressure on calyx necks.

The management is determined by the cause of the condition.

Megacalycosis

This is a unilateral, congenital malformation of the kidney with dilated kidney calyces showing on excretory urography. No clear mechanical or functional obstruction can be demonstrated. There is no indication for surgical intervention with this condition. Treatment may be required with complications such as infection or stone formation.

Calyceal diverticle

This is a cavity in the renal parenchyma; it communicates with an upper calyx via a narrow connection. Most children with this condition are asymptomatic. If infection or stone formation should follow, then a partial nephrectomy may be indicated.

22.9 Bladder pathologies

Vesico-ureteral reflux

During the normal flow of urine into the bladder, the intravesicle pressure compresses the intramural ureter and prevents reflux of urine into the ureters.

Vesico-ureteral reflux is not observed in normal healthy people. Fifty per cent of children with a documented urinary tract infection have an underlying vesico-ureteral reflux.

The causes of vesico-ureteral reflux are:

- Primary congenital reflux, which is usually found in girls due to a poorly developed trigonal muscle and lateral displacement of the ureter opening
- A neurogenic bladder with high intravesicle pressure
- Outlet obstruction, such as with posterior urethral valves and detrussor hyper-reflexia
- Trauma to the intramural passage of the ureter
- Specific and non-specific urinary tract infections
- Congenital causes, namely complete duplication of the ureter, ectopy of the ureters and 'prune belly' syndrome
- Iatrogenic reflux due to surgical procedures that damage the trigonal muscle

The effect of vesico-ureteral reflux is the retention of urine and that may be the cause of urinary tract infections. It is thought that infection consequent on vesico-ureteral reflux in childhood causes renal scar formation, with functional impairment of the kidney and the development of hypertension.

Vesico-ureteral reflux should be part of the differential diagnosis of any child with a documented urinary tract infection. In such cases a voiding cysto-urethrogram (with contrast medium or isotope) is the gold standard for diagnosing vesico-ureteral reflux. The use of pressure cystograms to demonstrate vesico-ureteral reflux is not recommended. A DMSA isotope renogram will show any renal scars. Endoscopic evaluation of the bladder outlet, bladder wall and appearance of the ureteral opening is occasionally indicated.

The level of reflux and the age of the child will determine the treatment method indicated. The levels of reflux are:

Level I – ureter only
Level II – ureter and pelvis without dilatation
Level III – slight dilatation, no blunting of renal calyces
Level IV – mild dilatation, blunting of renal calyces
Level V – severe dilatation with tortuosity of the ureter

Medical management is recommended for vesico-ureteral reflux Levels I to III in children under 5 years of age. The child is placed on *nocte* doses of Trimethoprim® or Nitrofuration®. As the bladder grows, more of the ureter becomes incorporated into the bladder wall and this leads to spontaneous recovery of the vesico-ureteral reflux in the majority of patients. Three-monthly urine cultures are recommended.

An initial period of medical management is also recommended for Levels IV and V reflux.

Surgical management is indicated in the following groups of children:

- Levels III to V reflux, which do not improve with time
- Low-pressure reflux with severe hydro-ureter
- Unsuccessful medical treatment – defined as breakthrough infections on low doses of a urinary antiseptic or the development of renal scarring on treatment, deteriorating renal function and irresponsible parents

Surgical management may consist of cystoscopic injection of an inert material (collagen/Uropaque) into the bladder wall overlying the ureter ('Sting' procedure) or operative lengthening of the intramural passage of the affected ureter. If wide ureters are present, then remodelling of the wide ureter precedes the anti-reflux procedure.

The complications of re-implantation are stricture formation with hydro-ureter or hydronephrosis and even progression to a non-functioning kidney. Persistent vesico-ureteral reflux may occur in a small percentage of patients after anti-reflux procedures.

Re-operation or repeat cystoscopic injections are indicated with either complication in children.

Exstrophy of the bladder

This is part of a syndrome consisting of anterior abdominal wall anomaly with a hypogastric exomphalos, a small exposed (exstrophic) bladder, a short urethra, abnormal development of the external genitalia and diastasis of the pubic rami. Epispadias with a small penis in boys and bifid clitoris in girls is usually associated with this condition. Occasionally, inguinal hernia and undescended testes are also noted.

Ante-natal diagnosis with high-resolution ultrasound will note:

- lack of bladder filling
- low-set umbilicus
- small genitalia
- diastasis of the pubic rami

Transfer the baby ante-natally, or when initially the diagnosis was missed post-delivery, to a unit where closure can be conducted within 24 hours. Where the bladder is too small for initial closure, waiting for a 4–6 month period is recommended. Once the bladder is closed, epispadia repair in boys is done after a 6-month period. Thereafter an anti-incontinence procedure is done to bring about urinary continence.

22.10 Hypospadias

Hypospadias is a congenital condition of the penis consisting of the following three components:

- an abnormal ventral opening of the urethral meatus
- a ventral curvature or chordee of the penis
- a dorsal displacement of the prepuce

The level of the hypospadias is named after the site of the abnormal opening of the urethral meatus, i.e. glandular, coronal, penile, penis-scrotal, scrotal or perineal.

Undescended testis is relatively common in boys with hypospadias. A severe level of hypospadias, i.e. penis-scrotal or perineal, is observed in approximately one-third of cases. In this group of children, an underlying intersex should be investigated.

Management consists of correcting the chordee and reconstructing the urethra. For the reconstruction of the urethra, non-hair-bearing skin may be needed. Consequently, it is important not to do circumcision prior to hypospadias repair. The optimal age changes with newer repair methods and ideas. At present, most are attempted during the first year of life. Urethrocutaneous fistula formation is observed in approximately 15% of cases.

22.11 The empty scrotum

As the name indicates, the scrotum is empty due to the absence of one or both testes. The common causes of a unilateral or bilateral empty scrotum are:

- rectractile testes
- undescended testes
- ectopic testes
- loss of the testis due to torsion, destruction or orchidectomy
- agenesis of the testis

Retractile testes

Here the testes have descended normally into the scrotum, but due to an overactive cremaster muscle, they are pulled up into the inguinal canal. In such cases the testes can be 'milked down' into the scrotum on examination or may come down spontaneously if the boy is placed in a warm bath. The management is conservative, setting the parent's minds at rest. Surgery is not indicated.

Undescended testes

An undescended testis is characterised by a failure of testicular descent anywhere along the normal path between the kidney and the intrascrotal position.

The cause of the condition is intrinsic testicular abnormalities. The majority of undescended testes are located at the external inguinal ring. Bilateral undescended testes are seen in approximately 15% of cases.

Normal descent of the testes into the scrotum usually takes place at approximately 7–8 months' gestation. Consequently, undescended testes are seen in 30% of premature boys. At the age of 1 year, only 1% of boys are found to have undescended testes.

Unilateral undescended testis usually occurs as an isolated case. Bilateral undescended testes usually form part of intersex, exstrophy of the bladder and pituitary defects.

As the diagnosis is obvious in most cases, special investigations are not routinely done. A chorionic gonadotropin (HCG) testicular stimulation test can be used to distinguish between bilateral undescended testes and bilateral agenesis of the testes. Two thousand units of HCG are given intramuscularly, and a rise in serum testosterone over a 4-day period confirms the presence of bilateral undescended testes.

Undescended testes are managed surgically by orchidopexy once the child is older than 1 year. Failure to find the testes on surgical exploration should mandate a laparoscopic inspection of the posterior abdomen and pelvis to locate unilateral or bilateral gonads.

The complications of undescended testes are:

- malignancy of the testis, which is found in approximately 7% of patients with undescended testes

- infertility
- torsion of the testis
- trauma to the testis
- psychological problems resulting from an empty scrotum

Ectopic testis

Ectopic testis is an uncommon condition in which the testis lodges outside the normal path of descent. The most common place that it occurs is at the superficial inguinal sac of McGregor. Other sites at which the ectopic testis may be found are: the base of the penis, the perineum and the femoral triangle. The management of choice is an orchidopexy.

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