5.19 Surgical disorders

BOX 5.19.1 Minimum standards
- Surgeon experienced in working with children’s disorders.
- Anaesthetist experienced in children’s anaesthesia.
- Equipped theatre with appropriately sized instruments where relevant.
- Pain relief.
- Fluid management.
- Ultrasound.
- Radiography.
- Blood transfusion
- Cytology.
- Chest drain insertion.

Introduction
Children’s surgery is a specialist subject. There are some emergency operations that may have to be performed by a competent general surgeon, such as appendectomy and surgery for a strangulated inguinal hernia, but most of the operations that are needed on very young children and infants require specialist knowledge and experience. Children’s surgery is therefore likely to be a tertiary-level referral service.

Indirect inguinal hernia
This is the protrusion of the abdominal viscus into a peritoneal sac (the processus vaginalis) in the inguinal canal. The contents of the sac are usually intestines, but may be omentum, Meckel’s diverticulum, or ovary and Fallopian tube in females.
- Around 50% of cases are seen in the first year of life, mostly before 6 months of age.
- Patent processus vaginalis (not a hernia) is present in 80% of boys at birth, in 40% at 2 years, and in 20% of adult men.
- A bulge in the groin, which sometimes extends to the scrotum, and which appears when the child cries or strains but disappears when he relaxes, is certainly a hernia. Hernias are seldom symptomatic except when they are very large or are incarcerated or strangulated.
- On physical examination, cough or crying impulse is the most important sign. A soft bulge that is reducible on digital pressure is also a diagnostic feature. Hernia in neonates may be transilluminant, so it is not a very reliable test to differentiate with hydrocoele.

Needle aspiration is contraindicated in any inguinal swelling because of the risk of perforating the intestines.

Differential diagnosis
This should include the following:
- Lymphadenopathy: firm, immobile, non-reducible, and no cough impulse.

Hydrocoele: can reach the upper pole of the swelling, transillumination, no cough impulse is present.

Hydrocoele of the cord: separate from testes, non-reducible, no cough impulse, upper limit is reachable, moves on pulling on the same-sided testis.

Undescended testis: scrotum empty and hypoplastic, cough impulse, may be reducible.

Femoral and direct inguinal hernias: rare, but should be kept in mind.

Treatment
All inguinal hernias should be promptly repaired unless there is another medical condition that makes the anaesthetic risks prohibitive. Premature infants with hernia should not be discharged without a repair of the hernia, as the risk of incarceration is high. An anaesthetist with paediatric anaesthesia experience is required, as anaesthesia-related risks are higher in children. Post-operative apnoea may occur in premature babies and at times may require ventilatory support. If these facilities are not available, the baby should be referred to higher centres or the surgery deferred until the risks associated with anaesthesia are low.

There are reasons for avoiding delay, especially in infants.
- Spontaneous disappearance of inguinal hernia does not occur.
- The risk of incarceration is greater in infants.
- Operation is technically more difficult and the risk of injury to the vas and testicular vessels is greater in longstanding and incarcerated hernia.
- Increasing age does not affect the risk of anaesthesia so long as an experienced anaesthetist is available.

A herniotomy is performed through an incision in the lowermost transverse inguinal skin crease. The sac is identified and transfixed. Herniorrhaphy is not required, as the cause is a patent processus vaginalis. Bilateral exploration and repair are indicated in patients with bilateral hernias, but routine contralateral prophylactic exploration is no longer recommended.

Incarcerated hernia
This occurs when the intestine becomes stuck at the internal inguinal ring. If it is prolonged, the blood supply may also become compromised, causing strangulation. There is a sudden increase in the size of the hernia with severe pain and symptoms of bowel obstruction (vomiting and abdominal distension). On examination a hard tender fixed mass in the groin is palpable, with increased bowel sounds on auscultation. It may be confused with the torsion of testis, acute inguinal lymphadenitis and tense infected hydrocoele.

Treatment
This includes the following:
Hydrocoele
This is accumulation of fluid in the scrotum; there is communication via a patent processus vaginalis (PPV) with the peritoneal cavity. Rarely hydrocoele is secondary to epididymo-orchitis, tumour and torsion of the testis.
- It is usually asymptomatic.
- The testis is not palpable separately, and the upper pole of the swelling is reachable, reduces on lying down and is transilluminant (hernia in a neonate may also be transilluminant).
- No cough impulse is present.

Differential diagnosis
Hydrocoele should be differentiated from inguinal hernia, and underlying pathology such as tumours and torsion of testis should not be missed. In older children, spermatocele and varicocele are non-transilluminant, have a worm-like feeling on palpation, and are separate from the testes.

Surgical treatment
This is rarely indicated. More than 90% of hydrocoele will spontaneously disappear. Surgery is indicated if it has not disappeared by the age of 2 years, and for hydrocoele that are larger and symptomatic. Herniotomy or PPV ligation, as performed for inguinal hernia, is the procedure of choice.

Undescended testis (cryptorchidism)
An undescended testis is one that cannot be made to reach the bottom of the scrotum. It is the second most common problem in paediatric surgery after indirect inguinal hernia, and should be distinguished from the more common retractile testis.

The incidence of undescended testis is 2.7–3% at birth in full-term infants, decreasing to 1.5% after 1 year of age, and thereafter the incidence remains the same. It is more common in premature infants, approaching 100% at a gestational age of 32 weeks or less.

- An ectopic testis is one that has strayed from the inguinal canal, usually to the thigh, perineum, base of the penis, or femoral or abdominal region.
- An ascending testis is one that is in the scrotum at birth, but the spermatic cord fails to elongate at the same rate as body growth, so the testis becomes progressively higher in the inguinal canal during childhood.

An impalpable testis is quite uncommon (less than 10%), and agenesis is rare (20% of all impalpable testes). A fully descended but grossly hypoplastic testis may be impalpable and only identified by exploration. Normal descent of testes occurs around the seventh month of fetal life when the gubernaculum swells and shortens, drawing the testis through the inguinal canal into the scrotum. Failure of descent may occur because of hormonal failure (inadequate gonadotrophins and testosterone), a dysgenetic testis, or an anatomical abnormality such as an abnormal or misplaced gubernaculum, obstruction of the inguinal canal or scrotum, or a short vas or vasa.

Sequelae of non-descent
- The higher temperature of the extrascrotal testis causes testicular dysplasia with interstitial fibrosis and poor development of seminiferous tubules, thus hampering spermatogenesis. Testosterone production is unaffected by position. Thus a male with bilateral undescended testes will develop secondary sexual characteristics, but will be sterile.
- Due to dysplasia, there is an increased risk of malignancy (10- to 20-fold higher). The risk of malignant degeneration is not altered greatly by orchidopexy, but a position where it is palpable helps early diagnosis and gives a better prognosis. Malignancy usually develops in the second or third decade of life.
- A testis in the inguinal region is more prone to direct trauma and torsion.

Examination
An unhurried examination with warm hands and environment greatly helps in picking up a testis in an abnormal position. A hypoplastic scrotum may suggest that it has never housed a testis. In older children, squatting may coax the testis into the normal position, thus differentiating a retractile testis. Always look for an associated hernia. The position and size of the testis should be noted. If it is palpable, ectopic locations of the testis should be examined. For a testis that cannot be felt at all, an ultrasound examination may be helpful. Bilateral non-palpable testes may require laparoscopic examination and hormonal profiles in a higher centre.

Treatment
The histological changes in the testes occur as early as 6 months of postnatal life, and therefore a child who has an undescended testis should be operated at the earliest time possible to prevent such changes.
- The best time for orchidopexy is about 1 year of age, and preferably before the child’s second birthday.
- The hernial sac should be dissected from the cord structures and a high ligation done.
- The testis is placed in an extra-dartos pouch in the scrotum after an adequate dissection and mobilisation of the vas and vessels. Retroperitoneal dissection and careful snipping off of lateral peritoneal bands will give an adequate length to the cord.
- In about 50% of cases of impalpable testis a useful testis can be brought down, and in the remaining 50% there is either no testis present (testicular agenesis or intrauterine torsion-vanishing testis), or there is a useless and potentially neoplastic testis, which is removed.
- For an abdominal testis, laparoscopy is useful for identifying and confirming the position of the testis and
simultaneously permitting the ligation of spermatic vessels (Fowler–Stephen’s stage I operation). Later the testis can be brought into the scrotum after dividing the artery, the testicular blood supply being supported by the artery to the vas.

- For psychological reasons, if orchidectomy has been undertaken, prosthetic replacement should be performed later on.
- In bilateral undescended testes, especially with hypospadias, an intersex disorder should be suspected and the child should be further investigated.

**Prognosis**

There is a 2% recurrence rate, 2–5% incidence of atrophy, 70–80% fertility after unilateral orchidopexy, and 40% fertility after bilateral orchidopexy.

**Hypospadias**

This is a condition where the urethra opens on the ventral aspect of the penis at a point proximal to the normal site. When it opens on the dorsal aspect (termed ‘epispadias’) there is usually associated exstrophy of the bladder. If there is no infection, two-layered closure of the bladder is sufficient, requiring no catheter or suprapubic drainage. If there is no infection, two-layered closure of the bladder is sufficient, requiring no catheter or suprapubic drainage. Open stone surgery is the modality of choice. Endoscopic removal can be performed in some children if the necessary equipment and expertise are available.

- Hypospadias is one of the commonest congenital anomalies of the male genitalia, occurring in 1 in 300 male births. There are various degrees of severity depending on how far back the urethral meatus lies. It may be associated with undescended testes, and in severe cases there is a possibility of an intersex problem.
- Ventral curvature of the shaft of the penis is called a ‘chordee’. It is due to fibrosis of the urethral plate, shortened skin, or fibrosis of the corpora cavernosa. The prepuce is deficient ventrally, and an unsightly dorsal hood of redundant skin is present.
- Congenital short urethra is a deformity where there is ventral curvature of the shaft of the penis without hypospadias.

The disabilities of hypospadias are cosmesis of the penis, a stream that is deflected downwards or splashes, and in severe hypospadias, boys have to void in a sitting position (like females). Uncorrected chordee interferes with intercourse, and there is infertility in severe hypospadias (penoscrotal and perineal), as semen is not directed into the vagina.

**Treatment**

Hypospadias should be corrected before school age so that the child does not feel ostracised in society. In severe cases of hypospadias, intersex disorders and associated urological abnormalities such as pelvic-ureteric junction obstruction or renal agenesis should be ruled out.

**Principles of surgery**

These are as follows:
- correction of chordee to straighten the penis (orthoplasty)
- movement of the urinary meatus to its normal position on the tip of the penis (urethroplasty)
- correction of the deformity of the glans to give it a conical shape (glansplasty).

No infant with hypospadias should be circumcised, as the prepuce is essential for the repair. Repair can be undertaken as a one-time or staged procedure. It depends on the degree of chordee and the severity of the hypospadias.

**Bladder stones**

In resource-limited countries, bladder stones are quite common due to the prevalence of malnutrition. The stones are composed of ammonium acid urate and oxalate, and are seen in lower socio-economic groups. Such stones are usually related to a high dietary intake of rice or wheat and low intake of milk and animal protein (see Section 5.10.B).

Children present with increased frequency of urine and strangury or haematuria (the child usually holds the penis and rubs it with the finger and cries during micturition). Children may present with an episode of retention of urine if the bladder stone becomes impacted at the bladder neck or in the urethra.

- During rectal examination, a stone may be palpable on bimanual palpation. A plain abdominal X-ray may reveal calcified stones. Abdominal ultrasonography will detect non-calcified stones.

**Cervical swellings**

The neck is one of the commonest sites of cystic and solid swellings during childhood. Lesions are either developmental anomalies arising from the remnants of branchial arches, thyroglossal tract, jugular lymphatics or the skin, or acquired as in diseases of the salivary gland, lymph nodes or thyroid gland.
- Lymphangiomas (cystic hygroma).
- Branchial cysts/fistulae.
- Thyroglossal cyst.
Lymphadenopathy

Enlargement of the lymph nodes may result from acute or chronic infection and from primary or secondary neoplasia.

- Infection is the commonest cause of lymph node enlargement in childhood, secondary to scalp and skin infections, including lice.
- Tuberculosis is the most important pathogen in resource-limited countries.
- In many cases the lymph nodes are reacting to an upper respiratory tract infection or an ear infection. This is known as non-specific reactive hyperplasia, and is much more common. Thus not every enlarged lymph node needs a surgical biopsy.
- Primary tumours of the lymph nodes include lymphoma and leukaemia.

Enlargement of a lymph node by more than 1 cm is significant, and a persistent node more than 3 cm in diameter requires fine-needle aspiration cytology or surgical biopsy.

A careful history with regard to repeated upper respiratory tract infections, boils on the scalp or drainage area, and ear discharge, should be taken. A positive family history of tuberculosis is an important feature of tubercular lymphadenitis. A history of the pattern of fever, loss of weight and appetite, and the presence of night sweats are important features when making a differential diagnosis.

On careful physical examination, all sites of lymph nodes (cervical, axillary, inguinal and abdominal) should be examined. The size, number, consistency, tenderness, and presence or absence of fluctuations should be noted. On abdominal examination, liver, spleen and mesenteric lymph nodes should be palpated. The drainage area of the lymph nodes should be examined for boils, furuncles, injury or neoplastic swelling. The tonsils should be inspected for enlargement and suppuration.

In acute lymphadenitis, the affected nodes are enlarged, painful and tender, restricting movement of local areas of the body. Fever and leukocytosis are common. Untreated infections may resolve spontaneously, progress to suppuration and abscess formation, or become chronic.

In tubercular lymphadenitis, lymph nodes are enlarged and painless, and become matted together and fixed to adjacent structures. Caseation leads to the formation of ‘cold’ abscesses, which lack the local and systemic signs of acute inflammation (fever, tenderness and erythema). When a cold abscess ruptures through the deep fascia (a ‘collar-stud abscess’) the skin becomes red and thin, takes on a blue tinge and then gives way to establish an indolent tubercular sinus. On aspiration, straw-coloured fluid is present, in contrast to the thick pus that is usually present in an acute abscess. Confirmation depends on culture of the organisms or visualisation of acid-fast bacilli on microscopy.

In primary neoplasia (e.g., leukaemia) the nodes are painless, rubbery in consistency and discrete. Liver and spleen enlargement may or may not be present.

Systemic features of low-grade fever, night sweats, or loss of weight and appetite point towards the diagnosis.

Secondary enlargement of the lymph nodes due to neoplasia is rare in childhood.

Primary cancers are soft-tissue sarcomas and very rare. The nodes are large, firm to hard in consistency, and fixed to underlying structures.

Investigations

- Full blood count.
- The erythrocyte sedimentation rate is usually raised in chronic infection and neoplasms. Leukocytosis is seen in acute lymphadenitis and abscess formation. Leukaemia will usually be diagnosed by the appearance of leukemic cells in peripheral blood.
- Mantoux test. To diagnose tuberculosis, start with 1 in 10,000 and then 1 in 1000. A strongly positive test is a pointer towards the diagnosis; if the test is negative, it does not rule out the disease (especially in the presence of HIV infection).
- X-ray of the chest.
- To identify there is the pulmonary lesion of primary complex or the hilar lymphadenopathy seen in cases of tuberculosis. Mediastinal widening is seen in patients with lymphomas.
- Fine-needle aspiration cytology (FNAC) is helpful if there are persistent lymph nodes that do not decrease in size after a 1-week course of antibiotics and another week of observation. Lymphomas cannot be definitely diagnosed on FNAC, and a surgical biopsy is mandatory.

Treatment

Acute lymphadenitis

- Antibiotics are prescribed. Penicillin is usually appropriate, as most infections occur outside the hospital setting. Oral or IV preparations may be used. If improvement has not occurred within 48 hours, a broad-spectrum antibiotic such as an oral or IV cephalosporin may be started.
- Anti-inflammatory medication (to relieve the pain and reduce the swelling).
- Hot fomentation (to relieve the pain and reduce the swelling).

Fluctuation, or other local signs of abscess formation, indicate the need for incision and drainage of pus, which is best performed under general anaesthesia. All of the loculi are broken and necrotic material is curetted out. Always visualise and remember the important structures nearby. A sample should be sent for microscopy (including Ziehl–Neelsen staining), culture and sensitivity, and appropriate antibiotics prescribed. The precipitating cause of acute lymphadenitis should also be treated.

Tubercular lymphadenitis

Antitubercular treatment leads to resolution (a full course of 9 months should be undertaken, with four drugs for 2 months and two drugs for the next 7 months; see Section 6.1.N).

Cold abscesses require drainage, and repeated aspirations may be preferable to avoid sinus formation, pending diagnosis and initiation of treatment.
**Lymphomas and leukaemias**

After diagnosis, further investigations will be required to stage the disease and its treatment (see Section 5.14).

**Cystic hygroma**

This is a hamartoma of the jugular lymph sac which presents in infancy and is more common in boys than in girls. It produces a major neck swelling and is diagnosed by inspection. The swelling is usually found as a unilateral, fluctuant, transluminant swelling centred on the carotid triangle. The cysts are of varying sizes and contain clear fluid. A haemangiomatous element may be present in the swelling, giving it a reddish tinge instead of a light blue colour. Cysts may enlarge suddenly due to viral or bacterial infection or haemorrhage. If the cyst compresses airways and vessels, it may cause stridor, respiratory distress and superior vena caval syndrome. Initially these lesions can be treated by aspiration and intralusal injection of bleomycin (a less expensive anti-cancer drug), at a dose of 300–600 micrograms/kg; these procedures can be repeated every 2–6 weeks, producing excellent results in the majority of cases. Surgical excision is difficult, and removal should be attempted without sacrificing important structures, in some cases in conjunction with sclerotherapy.

**Branchial cysts, sinuses and fistulae**

Sinuses and fistulae most commonly arise from the second branchial cleft, and occasionally from the first or third one. They present as a small discharging sinus on the skin overlying the lower third of the sternomastoid muscle. Parents often notice a drop of clear fluid coming from a very small opening. Sinuses and fistulae usually present in early childhood, and may sometimes be complicated by infection and abscess formation. Treatment consists of surgical excision of the whole tract up to the pyriform fossa to prevent recurrence. Methylene blue is injected or a nylon thread guided in the fistula to delineate it during surgical dissection for appropriate excision.

**Thyroglossal cyst**

The descent of the thyroid gland from the floor of the fetal mouth leaves a tract from the foramen caecum of the tongue to the thyroid isthmus. A cyst lined by respiratory epithelium may arise anywhere along the tract, but is usually subhyoid (75%). The swelling is in the midline and moves with swallowing and also with protrusion of the tongue. An infected cyst may be mistaken for acute bacterial lymphadenitis, or an ectopic thyroid may cause a similar swelling. The thyroglossal cyst and the entire tract along with the central portion of hyoid bone should be excised to minimise the risk of recurrence (Sistrunk’s operation).

**Epidermoid cyst**

Inclusion dermoid cysts arise from ectodermal cells that become detached during fetal growth. They are often in the midline or along lines of embryonic fusion. They contain sebaceous cheesy material surrounded by squamous epithelium. They enlarge slowly and should be removed completely; the capsule should not be breached to prevent recurrence.

**Haemangiomas**

These are the most common tumours of infancy and the most common congenital anomalies. They are present in around 1–3% of all newborn infants. This figure increases to 10% by 1 year of age. Haemangiomas can be capillary or cavernous, although both types may be present.

The natural history of capillary haemangiomas is as follows:

- They initially present shortly after birth as a pale pink or bright red spot or patch on the skin.
- There is subsequently rapid growth in infancy for 3–6 months, followed by a static phase.
- At 18–24 months the lesion starts to involute. Around 50% will involute by 5 years and 90% by 7 years. Rarely the lesion persists and requires excision.

A cavernous haemangioma has a deeper component in subcutaneous tissues or muscles, and is less likely to regress completely.

**Management**

Management of these lesions consists of an accurate diagnosis and careful observation. Parents need reassurance when the lesion is growing rapidly. Problems of ulceration, bleeding (and rarely) infection occur secondary to minor trauma. These are best treated non-operatively.

- Surgical excision is indicated when there is functional or gross cosmetic disability (e.g. a haemangioma on the eyelid), or a vital organ is threatened.
- Steroids may be used to induce involution in large haemangiomas (prednisolone 1–2 mg/kg/day for 2–4 weeks; the dose is tapered off before stopping the therapy). These can be repeated in cycles, with a gap of 4–6 weeks. Intra-lesional steroids can be used to induce regression in the size of haemangiomas in and around the eye.

**Obstructive jaundice in infancy**

This is most commonly caused by extrahepatic biliary atresia, choledochal cyst or inspissated bile syndrome.

- The most difficult differential diagnosis is neonatal hepatitis.
- If jaundice in the newborn persists, the stools are never yellow or green, and the urine is brown, a conjugated bilirubin level should be measured and urobilinogen and bilirubin looked for in the urine.
- Ultrasound may help in diagnosis. Strongly suspected cases need referral for radioactive scan and further management.

**Empyema thoracis (see Section 5.3.B)**

This is defined as an accumulation of pus in the pleural space. In most children this results from an infected pleural effusion associated with ongoing uncontrolled pulmonary sepsis or pneumonia. An infection of the pleural space is unlikely when there is a healthy underlying lung that is completely expanded. Empyemas and effusions may be diffuse and involve the entire pleural space, or they may be intralobar, diaphragmatic or paramediastinal.

Before the advent of antibiotic therapy, *Pneumococcus* and *Streptococcus* species were the organisms most frequently associated with empyema. Currently
**Umbilical hernia**

- This is a defect in the umbilical ring, which generally closes at birth, leading to protrusion of a loop of bowel or omentum through it. Some degree of herniation is seen in 20% of newborn babies, and still more in premature babies or when there is any increase in intra-abdominal pressure (e.g., due to ascites or VP shunt).
- Swelling appears on crying and straining, and decreases when the child is calm.
- It can be reduced with an audible gurgle.

Most umbilical hernias close spontaneously in the first 12 months of life, but they may take up to 3 years. Strangulation and incarceration are virtually unknown; therefore it is safe to wait. **Strapping with coin application is contraindicated**, as it leads to maceration of skin and infection, without any real advantage of inducing closure.

Surgical indications are a large hernia that has not closed by 3 years of age or an incarceration.

**Umbilical discharge**

- **Purulent discharge** is seen in umbilical sepsis. **Neonatal tetanus** is a serious condition in which mortality is very high (see Section 3.4) (cow dung application, as practised in rural India, is one cause). Portal thrombosis may occur secondary to it and manifest later as portal hypertension. Appropriate antibiotics (benzylpenicillin) should be instituted at the earliest possible stage, and local hygiene maintained.
- **Mucus/serous discharge** is seen in umbilical polyps and granulomas. Silver nitrate application will enable these to epithelialise. If these persist, excision will be required. Umbilical fistula may be present and require exploration and excision.
- **Urinary discharge** is seen with a patent urachus in association with a lower urinary tract obstruction. It is quite rare. Surgical treatment involves excision of the urachal remnant after investigation and relief of any underlying outlet obstruction.
- **Faecal discharge** is seen with a patent vitello-intestinal duct. This is a persistence of the connection between the yolk sac and the midgut, which normally disappears at about the sixth week of gestation. All remnants need to be excised, which may necessitate a laparotomy to search for any discontinuous segments of the tract.

**Appendicitis**

Appendicitis is the most common abdominal surgical emergency. Although diagnosis and treatment have improved,
appendicitis continues to cause significant morbidity, and is still (although rarely) a cause of death. However, abdominal pain unrelated to appendicitis is also common, and in many cases a few hours of active observation are recommended before proceeding to surgery.

Appendicitis results from luminal obstruction following infection or impaction by a faecolith. Inflammation of the appendix does not inevitably lead to perforation, as spontaneous resolution may occur.

Clinical presentation

- Presentation is very variable.
- Pain is invariably present and nearly always the first symptom. Early visceral pain is non-specific in the epigastric or umbilical region, and only later does pain become localised over the appendix, most typically at McBurney’s point. Pain with a pelvic appendix is often delayed in onset because the inflamed appendix does not contact the peritoneum until rupture occurs and infection spreads. Pain of a retrocaecal appendix may be in the flank or back.
- Anorexia, nausea and vomiting typically follow the onset of pain within a few hours.
- Diarrhoea occurs more frequently in children than in adults, and can result in misdiagnosis. It may indicate a pelvic abscess.
- The child with acute appendicitis lies in bed with minimal movement. There may be fever and tachycardia.
- The patient may be asymptomatic before perforation occurs, and symptoms may be present for longer than 48 hours without perforation. In general, however, the longer the duration of symptoms, the greater the risk of perforation.

Examination

Examination of the chest to rule out a lower respiratory tract infection is essential.

The single most important aspect of evaluation is serial examination undertaken by the same person. This decreases the number of unnecessary operations. Analgesia should not be withheld as was previously advised.

Investigation

There may be an increase in the white blood cell count, but this is unreliable.

Ultrasonography is an effective diagnostic aid, with a sensitivity of about 85% and a specificity of about 90%. Demonstration of a non-compressible appendix that is 7 mm or larger in antero-posterior diameter is the primary criterion.

Management

- The initial management involves IV fluids and adequate analgesia.
- In a patient who presents with peritonitis, adequate fluid resuscitation (see Section 5.5.B) must be performed before surgery is undertaken.
- For early non-raptured appendicitis, peri-operative antibiotics (cefuroxime and metronidazole) should be given.
- For perforated appendicitis after appendicectomy, saline irrigation of the peritoneal cavity with the patient in the head-high position is advisable in an attempt to remove as much infected material as possible. Intravenous antibiotics should be given for at least 5 days:
  - Cefuroxime (50 mg/kg 8- to 12-hourly) plus metronidazole (7.5 mg/kg 8-hourly IV over 20 minutes)
  - Ampicillin IV (25–50 mg/kg 8-hourly; maximum 4 grams/day) plus gentamicin (7 mg/kg once daily) plus metronidazole (7.5 mg/kg 8-hourly).
- If the initial presentation is with an appendicular mass, conservative treatment with IV antibiotics is given until the symptoms subside, with a plan for an interval appendicectomy.

Complications

Complications following appendicectomy include wound infection, abscess formation (local, subphrenic or pelvic) and paralytic ileus. A late complication may be an adhesive bowel obstruction.

Pyloric stenosis

This is a classical cause of gastric outlet obstruction in infants. It has a prevalence rate of about 1.5 to 4 in 1000 live births among white populations, but is less common in Africans and Asians. It is more common in males than in females, with a ratio of between 2:1 and 5:1. There appears to be an increased risk to firstborn infants with a positive family history.

Cause

No definite cause has been established. Pathologically there is marked muscle hypertrophy, primarily involving the circular layer, which produces partial or complete luminal obstruction.

Presentation

Pyloric stenosis typically presents at 2–8 weeks of age, with a peak occurrence at 3–5 weeks. The vomiting is projectile and non-bilious. Occasionally there is coffee-ground vomiting due to gastritis or oesophagitis. The child remains hungry after vomiting, and is otherwise not ill looking or febrile. Around 2–5% of infants have jaundice associated with indirect hyperbilirubinaemia. Non-bilious projectile vomiting, visible gastric peristalsis in the left upper abdomen, and in those presenting late a hypochloroaemic hypokalaemic metabolic alkalosis are the cardinal features of pyloric stenosis.

Diagnosis

A definite diagnosis can be made in 75% of infants with pyloric stenosis by careful physical examination of the upper abdomen. An absolute prerequisite for this is a calm and cooperative child, a warm environment, good light and patience. With the patient in the supine position, in the mother’s left arm and sucking on the left breast, and the surgeon sitting on the left side of the patient, the left hand is used to feel the classically described ‘olive’ to the right of the rectus muscle, often palpated against the spinal column. Visible gastric peristalsis is often noticed.

Investigations

- Ultrasonography is the most commonly used imaging technique for diagnosis. A positive finding is a pyloric canal length of 16 mm or more and a pyloric muscle thickness of 4 mm or more. A diameter of more than 14 mm is also considered abnormal.
Clinical presentation

- The infant is suddenly disturbed by what appears to be violent abdominal pain. The pain is colicky, intermittent and severe. With spasms the infant draws up the knees to the abdomen, screams, becomes pale and may sweat, and vomiting occurs soon afterwards. The infant may pass a normal stool, appears to recover immediately, and may resume normal eating habits, until stricken by another bout of colicky abdominal pain. The vomiting is initially reflex, but with a delayed diagnosis becomes secondary to intestinal obstruction and is often bile-stained.
- Classically, the infant passes stool that resembles redcurrant jelly. Many parents describe this as the presenting symptom, and consequently it is often treated as bacillary dysentery initially.
- The triad of pain, vomiting and blood per rectum is present in only one-third of patients. One in 10 infants with intussusception will have diarrhoea before signs and symptoms attributable to intussusception become obvious. This is often a cause for delay in diagnosis.
- Pallor, persistent apathy and dehydration are common signs.
- Abdominal examination reveals emptiness in the right lower quadrant and a sausage-shaped mass in the right hypochondrium, extending along the line of the transverse colon. The mass is not always easy to palpate, and its absence does not rule out an intussusception.
- Blood investigations in an advanced situation may show the typical hypochloraemic hypokalaemic metabolic alkalosis.

Management

- It is most important to prepare the patient appropriately and adequately for anaesthesia and surgery.
- Intravenous fluid resuscitation with 5% glucose in 0.9% saline with 20–40 mEq/litre of potassium chloride is the optimal fluid.
- Urine output and serum electrolytes should be monitored.
- The stomach should be aspirated before the operation.
- Ramstedt’s pyloromyotomy performed through a right upper quadrant or supraumbilical incision is curative, and is associated with a low morbidity.
- The majority of these patients can be started on feeds about 6 hours after surgery.
- Those who present with haematemesis from gastritis may benefit from delay of feeding for an additional 6–12 hours after surgery.
- Vomiting in the early post-operative period is thought to be secondary to discordant gastric peristalsis or atony.

Intussusception

This is the telescoping of a portion of the intestine into the lumen of an immediately adjoining part. Typically it occurs in a well-nourished child aged 4–12 months. The male:female ratio is 3:2, and it is more common in Caucasians.

The pathogenesis of intussusception is unclear. It usually originates in the ileum close to the ileoceleal junction and proceeds into the ascending colon. In 2–8% of cases there is a specific lead point such as a Meckel’s diverticulum, polyp or duplication cyst. Adenoviral infection resulting in lymphoid hyperplasia may act as a lead point.

Clinical presentation

- The infant is suddenly disturbed by what appears to be violent abdominal pain. The pain is colicky, intermittent and severe. With spasms the infant draws up the knees to the abdomen, screams, becomes pale and may sweat, and vomiting occurs soon afterwards. The infant may pass a normal stool, appears to recover immediately, and may resume normal eating habits, until stricken by another bout of colicky abdominal pain. The vomiting is initially reflex, but with a delayed diagnosis becomes secondary to intestinal obstruction and is often bile-stained.
- Classically, the infant passes stool that resembles redcurrant jelly. Many parents describe this as the presenting symptom, and consequently it is often treated as bacillary dysentery initially.
- The triad of pain, vomiting and blood per rectum is present in only one-third of patients. One in 10 infants with intussusception will have diarrhoea before signs and symptoms attributable to intussusception become obvious. This is often a cause for delay in diagnosis.
- Pallor, persistent apathy and dehydration are common signs.
- Abdominal examination reveals emptiness in the right lower quadrant and a sausage-shaped mass in the right hypochondrium, extending along the line of the transverse colon. The mass is not always easy to palpate, and its absence does not rule out an intussusception.
- Fever and leukocytosis are common, and tachycardia results from episodes of colic and hypovolaemia from dehydration.

Investigations

- Abdominal X-ray may show a soft tissue mass across the central abdomen with dilated loops of bowel.
- Ultrasonography has become the standard non-invasive diagnostic test, and is very reliable. Doughnut (target or concentric ring) and pseudo-kidney sign suggest a diagnosis of intussusception.

Management

The most important aspect of treatment is adequate resuscitation prior to intervention. This involves establishing reliable IV access, collecting blood for baseline investigations and for cross-matching, passing a nasogastric tube for decompression, and giving IV fluids and analgesia. Some patients may require one or more boluses of 10–20 mL/kg of albumin or Ringer-lactate solution when first seen.

Broad-spectrum IV antibiotics such as a combination of cefuroxime (25–50 mg/kg 8-hourly, depending on the degree of infection) and metronidazole (7.5 mg/kg 8-hourly IV over 20 minutes) are started, and the urine output is monitored. Management is initially non-surgical (i.e. with the use of air or barium enema). Sedation should be used for the procedure.

- A surgeon and theatre should be ready when the radiologist attempts reduction. If perforation occurs, surgery should be performed immediately.
- An absolute contraindication to rectal reduction is evidence of peritonitis, indicating the presence of a gangrenous intestine.

If hydrostatic reduction fails and if the patient is stable, a repeat reduction may be attempted. Once the intussusception reduces, the child should be observed overnight with careful monitoring of fluid and electrolytes.

If reduction fails, the child is taken for surgery, where by gentle manipulation (pushing and not pulling) the intussusception can be reduced. The appendix may be removed, recorded and the parents informed. If a pathological lead point is found, a resection anastomosis is performed. If the bowel is not viable, it is resected and a primary anastomosis is performed. Feeds are started the day after the operation and increased gradually.

Intravenous antibiotics should be given for at least 48 hours, and longer (for 7 days) if peritonitis is present.

The interval between the onset of symptoms and institution of treatment is of paramount importance, and mortality can be reduced if the condition is recognised and treated early.

Intestinal obstruction

This is the most common condition requiring emergency surgery in infants and children. Most causes result from complications of congenital anomalies or from inflammatory conditions that affect the bowel.
Causes
- **Extrinsic causes:** incarcerated hernia and vascular bands, intussusception, anomalies of rotation (volvulus and Ladd's bands, paraduodenal and paracaeal hernias), post-operative adhesions.
- **Intrinsic causes:** inspissation of bowel contents (meconium ileus, distal intestinal obstruction syndrome in patients with cystic fibrosis), roundworm obstruction.
- **Peristaltic dysfunction:** Hirschsprung's disease.
- **Inflammatory lesions:** tuberculosis, Crohn's disease.

Symptoms and signs
Patients present with cramping abdominal pain with anorexia, nausea and vomiting, which progresses to become bile-stained. Abdominal distension occurs, with the degree being directly related to the site of obstruction in the gastrointestinal tract, such that the distension is greater the more distal the obstruction.

On examination, the patient may have tachycardia and signs of dehydration. Tenderness and hyperactive bowel sounds are present on abdominal examination.

Chest and abdominal films are taken to confirm the diagnosis of obstruction and rule out the presence of free air.

Treatment
- The goal of treatment is to relieve obstruction before ischaemic bowel injury occurs.
- Intravenous access is established and blood collected for baseline investigations, including a full blood count, urea, creatinine and electrolytes, and cross-matching.
  - Intravenous fluids (Ringer-lactate or Hartmann's solution with 10% glucose) are started according to the guidelines of 4 mL/kg/hour for the first 10 kg, 2 mL/kg/hour for the next 10 kg, and 1 mL/kg/hour for the next 10 kg.
  - For example, a child weighing 22 kg would need 40 + 20 + 2 = 62 mL/hour.
- Some patients may need one or more IV boluses (10–20 mL/kg) with Ringer-lactate or Hartmann's solution or albumin at the start of resuscitation.
- A nasogastric tube is passed for decompression.
- Give broad-spectrum IV antibiotics such as:
  - cefuroxime 50 mg/kg 8-hourly or 12-hourly in the neonate, and metronidazole 7.5 mg/kg 8-hourly IV or
  - benzylpenicillin 50 mg/kg 6-hourly plus gentamicin 7 mg/kg once daily plus metronidazole 7.5 mg/kg 8-hourly.
- Once the patient is adequately resuscitated and fluid and electrolyte imbalances have been corrected, laparotomy is performed and the cause treated. Transfer to a facility where paediatric surgical and anaesthetic skills are available should be undertaken if the patient's condition will tolerate this. Otherwise, or in the absence of such a facility in the country, surgery should be performed.
- At all times adequate analgesia should be given (see Section 1.15).

Hirschsprung's disease
This is characterised by an absence of ganglion cells in the affected intestine. The incidence is about 1 in 4400–7000 live births; the male:female ratio is about 4:1, and in long segment disease it approaches 1:1. The longer the segment of aganglionosis, the higher is the familial incidence.

Associated conditions
These include Down's syndrome (4–16%), Waardenburg syndrome, multiple endocrine neoplasia 2A and Von Recklinghausen's disease. A higher incidence of enterocolitis has been noted in patients with Hirschsprung's disease and Down's syndrome.

Presentation
The usual presentation is with delay of passage of meconium beyond 48 hours after birth. (Around 95% of full-term infants pass meconium within 24 hours after birth, and the remainder pass it within 48 hours.) The child then has episodes of constipation, abdominal distension, vomiting and poor feeding, and fails to thrive. They may also present with a history of constipation with explosive diarrhoea, the latter indicating the development of enterocolitis.

Differential diagnosis
Hirschsprung's disease should be considered in the differential diagnosis of any child who has constipation dating back to the newborn period. However, childhood constipation related to dietary and habitual problems needs to be carefully ruled out in order to avoid unnecessary X-rays and biopsies.

Examination
On examination the child has a distended abdomen, and after a rectal examination there is often explosive passage of flatus and faeces.

- A plain X-ray of the abdomen may show dilated bowel loops with paucity of air in the location of the rectum. Barium enema may show the characteristic coning, although a simple colonic dilatation can occur in any chronic constipation.
- Rectal biopsy remains the gold standard for diagnosis. It should be performed at least 2 cm above the anal valves, as the normal anus has a paucity or absence of ganglion cells at the level of the internal sphincter. Although suction rectal biopsy with acetylcholinesterase staining has become the accepted standard for diagnosis in most centres, a full-thickness rectal biopsy under general anaesthesia is equally useful if such facilities are not available.

Treatment
Enteroctolitis remains the major cause of morbidity, and has a mortality rate of around 6–30%. It manifests clinically as explosive diarrhoea, abdominal distension and fever. The pathophysiology is not fully understood. The diagnosis is made on clinical grounds, and treatment is conservative, consisting of IV fluids and rectal washouts to decompress the colon.

Surgery
The surgical treatment of Hirschsprung's disease has evolved from a three-stage procedure (initial colostomy with multiple seromuscular biopsies, pull-through of the ganglionic colon as the second stage, and closure of colostomy as the third stage) through a two-stage procedure (colostomy at the transition zone initially, and pull-through as a second stage) to a one-stage procedure without a colostomy. The essential prerequisite for a primary pull-through is adequate preparation with colonic washouts.
Perforative peritonitis

The causes of perforation include amoebiasis, typhoid, tuberculosis, roundworm perforation and Hirschsprung's disease (see Section 6).

Management starts with an adequate history and clinical examination, followed by chest and abdominal X-rays. Adequate resuscitation should be carried out as outlined in the section on intestinal obstruction. After this a laparotomy is performed and the cause treated. Treatment includes fluid resuscitation if necessary, and antibiotics (either a third-generation cephalosporin or an aminoglycoside plus metronidazole).

Further reading