General Surgery in Childhood

7

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INTRODUCTION

It is common for children to present with surgical conditions in childhood. Roughly, one in ten children undergo surgery, a much larger number present either as emergencies or to outpatients for surgical problems; until recently, the majority of these children were not operated upon by specialist paediatric surgeons. Whilst not seeking to elaborate on areas covered in other chapters, it is important to possess a working knowledge of the surgical problems that present commonly in childhood. This is particularly relevant to practice in remote and rural settings where an understanding of these problems is particularly useful to allow appropriate recognition and timely referral of problems requiring specialist intervention. In such settings, these conditions are often managed by general surgeons and remote and regional medical specialists rather than specialist paediatric surgeons; it is useful to remember that one of the founding principles of the British Association of Paediatric Surgeons was 'to set a standard, not create a monopoly', and this sentiment is as relevant to practice today as it was in 1953; this is particularly true in our practice where patients and resources are spread over a large geographical area.

PRESENTATION

For the purposes of this chapter it is convenient to divide patients into different groups, as an understanding of the common presenting features of each condition is often helpful in establishing the correct diagnosis. Surgical problems often present as emergencies and in an unplanned fashion. The symptoms are often acute in onset, distressing for children and lead parents to seek medical attention promptly.

THE CHILD WITH ABDOMINAL PAIN

Patients presenting with abdominal pain make up a significant number of attendances to see both family doctors and emergency departments. Abdominal pain accounts for 5–10% of all presentations to paediatric emergency departments.

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Assessment of the Child with Acute Abdominal Pain

The first and most important component in the successful management of a child with abdominal pain is to take a careful, and where necessary, detailed history from the child and the parents. Acute abdominal pain is defined as a pain of non-traumatic origin with a maximum duration of 5 days. While most emergency visits presenting with acute abdominal pain are self-limiting a surgical cause may be found in up to 20%, it is important to consider that in younger children, the incidence of surgical pathologies decreases, and paediatric medical diagnoses are much more common. In infants (children under the age of 1 year), the two most common surgical diagnoses are incarcerated inguinal hernia and ileocolic intussusception. These two conditions account for 90% of surgical cases in children under 1 year of age presenting as emergencies with acute abdominal pain.

In children under over 1 year of age, the most common surgical diagnosis is acute appendicitis (which accounts for roughly 2/3 of cases ultimately requiring surgery). The other common causes requiring surgery are incarcerated hernias, trauma and ileocolic intussusception. In contrast to what is seen in adults, the majority of children (2/3) present with suppurative or gangrenous appendicitis. It is often possible after taking a history and after a careful examination to establish a non-surgical cause of abdominal pain.

Acute Abdominal Pain in Children

It can be seen from Table 7.1 that the majority of conditions requiring surgery would not only be expected to manifest as abdominal pain, but also that, in general, the nature of that pain would be severe, not self-limiting, often becoming progressively worse and would be associated with abdominal tenderness and in particular guarding. These cardinal features of surgical causes of abdominal pain should lead to prompt assessment and referral to surgeon. Of particular significance to paediatric surgeons is the sign of bile-stained vomiting. Dark-green vomit is of particular concern to surgeons as it can be the only sign of intestinal obstruction – and of particular concern, a small bowel volvulus secondary to malrotation which requires urgent emergency surgery. Patients often present with more nebulous signs and symptoms, and in such cases, it is important to consider medical causes of abdominal pain, as failure to adequately treat and identify these problems (particularly sepsis) can lead to poor outcomes for children.

TABLE 7.1 Common causes of abdominal pain in children

Surgical	• Acute appendicitis • Ileocolic intussusception • Incarcerated inguinal hernia • Torsion of the testis • Meckel's diverticulum • Pancreatitis • Trauma • Psoas abscess
Medical	• Non-specific abdominal pain • Mesenteric adenitis • Gastroenteritis • Urosepsis • Hepatitis • Psychological • Non-accidental injury/child abuse/neglect • Lower lobe pneumonia • Henoch–Schonlein purpura • Diabetic ketoacidosis
Other	• Gynaecological • Musculoskeletal e.g. osteomyelitis, referred pain from hip

The Infant with Abdominal Pain

It is uncommon for children under 12 months of age present with surgical problems. The history is presumptive and often difficult, and the diagnosis of abdominal pain is often made on the basis of distress and crying. The pain is often described as colicky and may be associated with intermittent drawing up of the legs and settling between episodes of pain. It is important to recognise surgical causes as if they go unrecognised and untreated, there can be rapid clinical deterioration. The specific features that point to a surgical pathology are:

- Bile-stained vomiting
- The passage of blood per rectum
- Abdominal tenderness
- The presence of tender swellings in the abdomen or groin

Children THAT are critically unwell upon presentation require empiric resuscitation with fluids and antibiotics concomitant to the establishment of a definitive diagnosis.

ACUTE APPENDICITIS

Acute appendicitis is the most common surgical cause of patients presenting with acute abdominal pain. Around 3–5% of children undergo appendectomy during childhood, and the most common age of presentation is between the ages of 10 and 17 years. Reported mortality in higher income countries is low, but morbidity is higher in children than adults, with perforated or complicated appendicitis occurring in around 1:3 children. The risk of complicated appendicitis is higher in younger children, and perforated appendicitis is very common in patients under 5 years of age.

Clinical Features of Appendicitis

The common presenting features are of malaise and decrease appetite with associated abdominal pain and often vomiting. Classically, the pain is characterised as intermittent and periumbilical in nature initially with a subsequent localisation to the right lower quadrant. Unfortunately, younger children are often unreliable historians; however, the careful elucidation of a history of the nature of the pain is the cornerstone of making the correct diagnosis. When asked about symptoms, families will often volunteer that the child has had a fever with nausea and a loss of appetite (although anorexia is not universal, particularly in teenage boys!). By the time of presentation to hospital, there is often pain on movement, with an abnormal gait, which is exacerbated by sudden movements or bumps in the road during transfer. The vital signs that support acute appendicitis are a low-grade fever and a mild tachycardia.

Although the common features of appendicitis are widely known, only around 50% of children present with these classical features. If the diagnosis is delayed beyond 48 h (which is often the case in remote and rural communities), the perforation rate is around 2/3. After perforation, the localising signs are often less obvious, and the child may present with sepsis or obstructive symptoms.

Investigations

In general, clinical diagnosis is more accurate than the most widely performed investigations. In cases in which there is diagnostic uncertainty then a number of tests can be helpful, but often, repeated clinical examination is a better option.

A leucocytosis may occur as appendicitis worsens, but is often normal in early appendicitis. Urinalysis is commonly performed in children as urinary tract infections are a common cause of abdominal pain. Often in appendicitis the urinalysis is normal, but around 25% of patients have white cells or red cells present in their urine at the time of presentation, and so it is important to also look for bacteria in a clean catch urine specimen with formal urine microscopy and culture.

Around 50% of patients can be diagnosed clinically without the need for further investigations.

Radiological investigations

Ultrasound scanning (USS) is widely used in the assessment of children with acute abdominal pain. It has the advantage of being widely available, even in remote and rural settings, it is relatively cheap and has been reported as high levels of sensitivity (80–85%) and specificity (90–95%) when diagnosing appendicitis. It is important to note that the majority of these studies have been performed by skilled operators in children's hospitals, and it is unlikely that such high levels of sensitivity and specificity can be replicated in the hands of those less accustomed to performing USS examinations in children on a regular basis. The ultrasonographic features suggestive of appendicitis include probe tenderness over the appendix, a transverse diameter of the appendix greater than 6 mm and a non-compressible appendix. Ultrasound is most useful in looking for ovarian pathology in post-pubertal females, as it is important to rule this out prior to planning for surgery. It is important to remember to only use supporting investigations when clinical diagnosis alone is not able to come to a satisfactory conclusion.

CT scanning is more accurate than USS with reported sensitivity and specificity of around 95%. It can be of particular value in obese children but is not commonly used due to the long-term risks of cancer development as a consequence of radiation exposure. It very rarely is indicated. Paediatric surgeons have been repeatedly demonstrated to be about 95% accurate in diagnosing appendicitis using selective observation, repeated clinical examination and selected blood tests. The use of poorly timed and indiscriminate investigations increases costs, causes diagnostic confusion and delay and should be avoided.

Treatment of Appendicitis

The treatment of non-complicated appendicitis consists of:

- Preoperative resuscitation as required
- The administration of appropriate analgesia (including opiates)
- Early institution of antibiotics after coming to a definitive clinical diagnosis
- Appendectomy (discussed later in this book)

There is currently insufficient data to support conservative treatment of appendicitis in children. Although this is becoming more common in the adult population, early data suggests that recidivism with further episodes of appendicitis is more common in the paediatric population.

In the case of complicated appendicitis (generalised peritonitis supporting a diagnosis of perforation), surgical exploration and aggressive irrigation should be performed, once the patient has been adequately resuscitated as in the treatment of simple appendicitis. For those patients with signs of shock or intestinal

obstruction, the placement of a urinary catheter to monitor urine output, routine bloods and the placement of a nasogastric tube to decompress the stomach should all be considered. These measures are particularly important in remote settings where there may be considerable delay in transferring patients to a tertiary level facility and the use of aeromedical retrieval means intestinal decompression is necessary.

In rare cases of perforated appendicitis with subsequent appendix mass formation treatment should initially be supportive and conservative rather than surgical. Physiological support (including adequate pain relief and nutrition) and intravenous antibiotics guided by local antimicrobial policies are often successful, although the data suggests that subsequent interval appendectomy is probably indicated. This is particularly true in patients coming from remote areas.

THE ACUTE SCROTUM

Presentation to the emergency department because of concerns about acute pain in the testis is common in children. The acute scrotum is defined as the constellation of sudden onset of pain and or tenderness or swelling and redness in the scrotum or its contents. It is important to expeditiously assess children to determine the cause of the pain, as testicular torsion must be considered in any patient who complains of acute scrotal pain and swelling.

Testicular Torsion

Torsion of the testis is a surgical emergency as the likelihood of testicular salvage decreases as the duration of torsion increases. The initiation of investigations should never delay the prompt referral to the relevant surgical team to prevent unnecessary testicular loss. Whilst it is important not to delay treatment, certain features both in the clinical history and signs found on physical examination can help differentiate between the different causes of acute testicular pain. A number of conditions can mimic testicular torsion, most commonly a torsion of a testicular appendage (particularly in prepubertal boys) and less commonly in children epididymo-orchitis, trauma, hernia, hydrocoele, Henoch–Schonlein purpura (HSP) and idiopathic scrotal oedema (ISE).

The following signs and symptoms are the cardinal features of testicular torsion:

- Acute onset of severe pain
- Vomiting/anorexia/nausea
- Testicular tenderness
- A horizontal testicular lie
- Abolition of the cremasteric reflex

It is quite possible for none of these features to be present and to still find a torsion of the testis at surgical exploration. In regional and rural practice, and, in particular, in remote tropical areas, practical considerations often mean that patients cannot undergo immediate scrotal exploration. In such cases, urinalysis and Doppler USS examination of the scrotum may have a limited role in allowing the relative likelihood of a genuine testicular torsion to be present when immediate surgical exploration is not available. There is unfortunately considerable overlap in the clinical features of acute testicular torsion and torsion of a testicular or epididymal appendage. It is the author's practice to urgently explore almost all boys with testicular tenderness. The clinical features of the common causes of the scrotum are listed below (Table 7.2).

TABLE 7.2 Common causes of the acute scrotum

FEATURES OF COMMON CAUSES OF THE ACUTE SCROTUM				
CONDITION	*ONSET*	*AGE*	*TENDERNESS*	*URINALYSIS*
Testicular torsion	Acute	Post-pubertal	Diffuse and cord	–
Epidiymo-orchitis	Insidious	Post-pubertal	Epididymal	+/– leuc
Appendix torsion	Subacute	Prepubertal	Discrete area	–
HSP	Subacute	Prepubertal	Diffuse	+/– rbc
Idiopathic SE	Subacute	Prepubertal	None	–

Torsion of Testicular Appendages

The appendix testis is a Mullerian duct remnant located at the superior pole of the testis. It is the most common appendage to undergo torsion; the epididymal appendage (a Wolffian duct remnant) may also become twisted. Torsion of either appendage produces pain and tenderness similar to that seen in a true testicular torsion; patients are usually prepubertal in age, and the onset and severity of the pain is often less than that seen in acute testicular torsion. Unfortunately, USS is not sufficiently specific as a test to completely exclude testicular torsion. On occasion, a 'blue dot' may be visible through the scrotal skin, particularly if patients present early prior to the development of scrotal oedema of the overlying skin, this allows a clinical diagnosis of a torsion of a testicular appendage to be made with reasonable certainty, and it is possible to offer the family a conservative approach to management. This is particularly useful in remote practice as it may avoid the need for urgent exploration and transfer of the patient. Boys with this condition can sometimes experience ongoing debilitating pain and worsening of symptoms even with simple analgesia, and a conservative strategy may be unsuccessful in such cases. In prepubertal males, the incidence of torsion of the testicular or epididymal appendage is significantly more common than a genuine torsion of the testis.

Epididymo-Orchitis

Epididymo-orchitis is uncommon in prepubertal males. It has long been recommended that urinalysis is performed in males with an acute scrotum to try and identify patients with epididymo-orchitis and thus avoid unnecessary urgent scrotal exploration. However, urinalysis may be negative even in the presence of bacterial epididymo-orchitis. Urine microscopy, culture and sensitivities can be helpful in guiding subsequent antimicrobial therapy if epididymo-orchitis is suspected. Any episode of epididymo-orchitis in childhood should be investigated with renal tract ultrasonography to look for structural renal tract abnormalities. Treatment is with antibiotics; it is not uncommon for patients to require an extended course of antibiotics to successfully treat bacterial epididymo-orchitis, and usually, we recommend a course of 2 weeks ciprofloxacin in the first instance. The majority of cases in younger patients are viral in nature, but it is difficult to exclude bacterial infections as urinalysis is often normal. Patients with recurrent epididymo-orchitis may be due to abnormalities in the proximal urethra such as an enlarged prostatic utricle with abnormal vasal reflux, such patients may require further investigations such as cystoscopy to investigate for possible anatomical abnormalities.

Scrotal Trauma

Severe testicular injury is fortunately uncommon in childhood, but a direct blow to the scrotum or a straddle injury can result in an intratesticular haematoma or laceration of the tunica albuginea. Such cases require immediate drainage and repair. Traumatic epididymitis is one potential sequela of scrotal trauma,

and a conservative strategy may be adopted if there are no concerns about damage to the tunica albuginea. Ultrasonography in skilled hands can be useful in such cases.

Henoch–Schonlein Purpura (HSP)

HSP is a systemic vasculitic syndrome. It is characterised by a non-thrombocytopenic purpura. This can cause a variety of symptoms, including arthralgia, renal disease (manifesting as a microscopic haematuria on urinalysis and microscopy), abdominal pain and gastrointestinal bleeding. Classically, a purpuric rash is often noticed on the shins. HSP occasionally presents as acute testicular pain, and if there are other supporting features or the diagnosis is suspected, then they should be assessed by a paediatrician as well.

Idiopathic Scrotal (O)Edema (ISE)

Acute idiopathic scrotal oedema (ISE) is another possible cause of an acute scrotum. The aetiology of this condition is unclear, but the patient presents with a red or swollen scrotum. Classically, the oedema and redness are described as bilateral and symmetrical; however, it is the author's experience that often it can start on one side and then spread across the median raphe over time. Although the appearance of the scrotum may be quite florid and certainly alarming to the parents and clinicians, the cardinal feature of this condition is a distinct lack of tenderness of either testis or the cord. The cremasteric reflex is usually maintained. Treatment is with bed rest/elevation/analgesia (although pain is often not a clinical feature). There is some evidence that antihistamines may reduce the duration of clinical features. (An example of a male infant with ISE can be seen in Figure 7.1.)

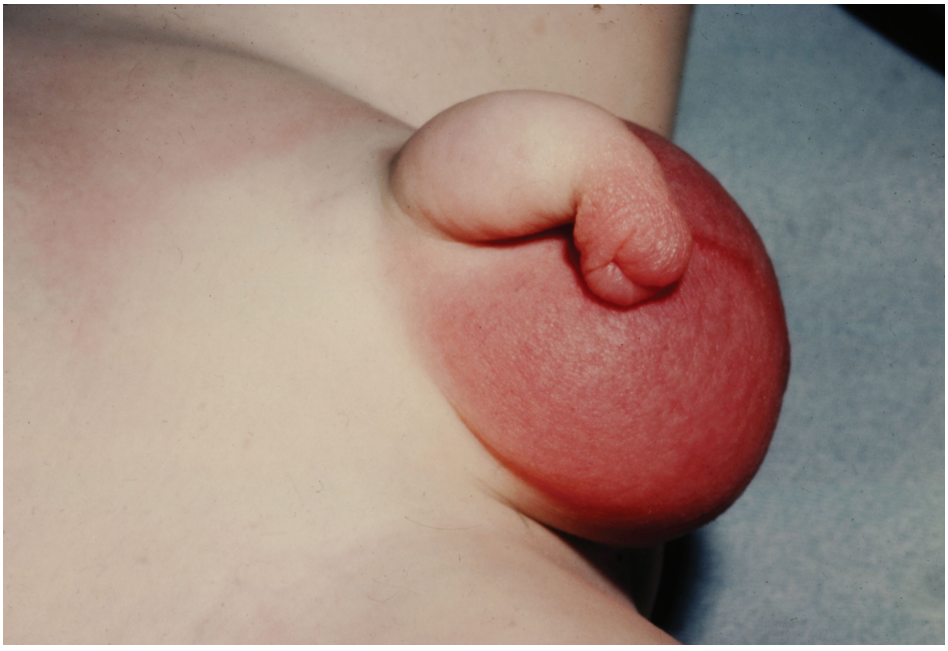

FIGURE 7.1 Idiopathic scrotal oedema.

Inguinal hernia/Hydrocoeles

Occasionally, obstructed or incarcerated inguinal hernia can manifest as an acute scrotum. A fuller description of the management of these conditions can be found in Chapter 6.

UMBILICAL HERNIA

It is a very common occurrence to find an umbilical abnormality at birth, particularly in premature infants. The estimated incidence of umbilical hernia in neonates is between 20 and 30% at birth, and the incidence is higher in premature infants with a reported incidence of 85% in children with a birth weight between 1000 and 1500 g. The incidence is higher in some ethnic groups; it is also more commonly seen in patients with a variety of other medical conditions, including trisomy 21, mucopolysaccharidoses and other congenital abnormalities.

The commonest umbilical abnormality at birth is an umbilical hernia; the vast majority of these will close spontaneously by the age of 4–5 years (over 90%). It is important to understand the aetiology and embryology, as well as to be able to recognise other umbilical pathologies to allow for appropriate advice and information to be given to families and to ensure that patients are referred appropriately to paediatric surgical services.

Embryology and Anatomy

During early fetal development, the primitive umbilical ring forms on the ventral surface of the fetus between the 4th and 5th weeks of fetal life. The primitive umbilical ring contains a number of structures: the umbilical vessels (two arteries and one vein), the vitelline duct and vessels and the allantois as well as a loop of midgut. The herniated midgut returns to the abdomen and, in doing so, leads to the development of the definitive umbilical cord which contains the umbilical vessels encased in Wharton's jelly. The umbilical vessels obliterate soon after birth and are replaced with a ligamentous structure. Failure of these normal embryological processes can result in a number of congenital disorders of the umbilicus, the commonest being umbilical hernia. The commonest abnormalities are:

- Umbilical hernia
- Patent urachus
- Omphalomesenteric fistula
- Umbilical polyp
- Umbilical granuloma
- Hernia of the cord

Assessment of Umbilical Abnormalities

Although umbilical abnormalities are commonplace, it is important to be able to recognise other umbilical problems as they may require more specialised assessment and intervention. Any abnormal finding in a neonate should prompt a more detailed evaluation of the infant, including the taking of an

antenatal and family history. Umbilical hernia is more common in a number of important conditions, including:

- Trisomy 21 and 18 and other chromosomal abnormalities
- Beckwith–Wiedemann syndrome
- Marfan's syndrome
- Hypothyroidism
- Mucopolysaccharidoses

It is important to look for other stigmata of these conditions and to examine the umbilical region carefully. It is important to look at the size of the defect; typically, very large defects (2–3 cm) are uncommon but are less likely to close spontaneously; it is important to distinguish between the size of the defect in the fascia rather than the size of the skin defect, which can often be large even with relatively small defect. It is important to examine the umbilicus carefully to look for persistent discharges or ectopic mucosae which are signs of a patent urachus or persistent vitellointestinal duct remnant. These abnormalities require further assessment by a paediatric surgeon.

Management of Umbilical Abnormalities

Surgical repair of umbilical defects is usually deferred until children are between 2 and 5 years of age. The risk of complications due to incarceration of intestinal contents is low in childhood. It is possible for intestinal contents or omentum to become incarcerated and require emergent surgical treatment, but this is uncommon with an incidence of around 1:10,000 cases per year. Many parents are concerned about the size of the swelling, particularly if the hernia is large and protuberant, although these herniae are less likely to become incarcerated or obstructed. A number of attempts have been made to strap or tape umbilical herniae to encourage them to close, but unfortunately, these techniques have been of little demonstrable value and are associated with an increase in the incidence of potential complications. In our practice in remote and regional areas, it is often practical to repair uncomplicated umbilical herniae in children towards the younger end of the age range to prevent unnecessary parental anxiety as patients often live significant distances from a paediatric surgeon. (An example of an infant with a large umbilical hernia can be seen in Figure 7.2.)

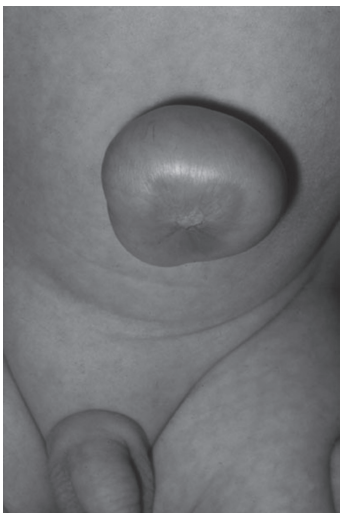

FIGURE 7.2 Umbilical hernia.

Emergency surgery is indicated for complications of the hernia, which are fortunately uncommon. Occasionally, an umbilical hernia may become incarcerated (often with omentum present in a smaller defect), and there are reports of intestinal strangulation and obstruction and even of rupture secondary to trauma. In children with larger fascial defects (>1.5 cm) that persist beyond 2 years of age, these rarely close spontaneously, and so repair is recommended. There is little value in deferring surgery in these patients as the risk of surgery is little changed between the age of 2 and 5 years.

Umbilical hernia repair is more correctly termed an umbilical herniotomy. An infra-umbilical incision is made, and then the hernial sac and defect are circumscribed. The hernia sac is then opened, and the fascial defect is closed with an absorbable suture. This can typically be performed as a day case surgery. It is uncommon to have to use mesh to facilitate a repair in children, but it may be necessary in very large defects (>5 cm); these are probably more correctly thought of as covered anterior abdominal wall defects. Umbilicoplasty may be performed for those patients particularly with large skin defects that are protuberant in nature.

Complications and Long-Term Management

Unfortunately, it is difficult to ascertain the true rate of complications in patients with untreated umbilical hernias as there is significant selection bias in the published data. From our experience, in those patients who have umbilical hernia but only have limited access to surgery (e.g. in developing countries) we know that complications of untreated umbilical hernias are rare in childhood. The reported incidence of complications in untreated umbilical hernias is between 1:300 and 1000. As children progress into adult life, the risk of complications becomes higher, particularly in pregnancy; the rationale behind fixing these hernias in childhood is to prevent these complications later, and the surgery is relatively safe and straightforward in younger children. Umbilical hernia repairs have a low postoperative complication rate. The commonest early complication is a superficial wound infection, prompting the use of routine perioperative antibiotics. After routine repair, recurrence is uncommon (1–2%) except in those patients with underlying predisposition to hernia formation (trisomy 21, mucopolysaccharidoses, connective tissue disorders). Longer-term follow-up is recommended for this selected group of patients.

ABNORMALITIES OF TESTICULAR DESCENT

Abnormalities of testicular descent are a common problem in everyday practice. A failure of the normal processes of testicular descent is termed *testicular maldescent*. This can be further classified as:

- Undescended testis
- Ectopic testis
- Ascending testis
- Retractile testis
- Absent testis

Perhaps most usefully testes can be divided up practically into two main groups, *palpable* (but abnormally positioned) and *impalpable*.

Undescended Testes

The second most common condition requiring paediatric surgery in childhood is testicular maldescent. The reported incidence varies according to the population being studied (e.g. age and other factors such

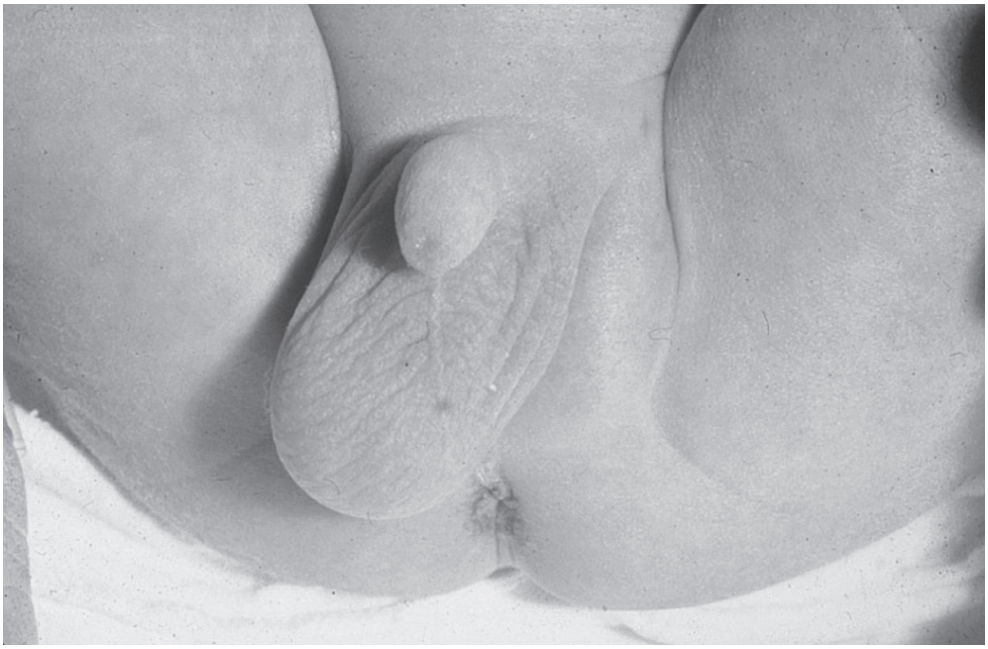

FIGURE 7.3 Empty left hemiscrotum (left undescended testis).

as prematurity), but a widely accepted figure is that around 1:60 boys undergo surgery for undescended testis in the first 2 years of life. An example of a child with an undescended testis with a clearly empty left hemiscrotum can be seen in Figure 7.3.

At birth, up to 10% of term infants are identified as having some abnormality of testicular descent. By the age of 3–6 months, this falls to around 2–3% of boys. It is important that these patients are followed up carefully, and that boys who are determined clinically to have testicular maldescent at 3 months of age or older are referred to a paediatric surgeon for assessment and ongoing management.

In the majority of cases, testicular descent is unilateral; however, it is important to acknowledge the increased clinical significance of boys with bilateral testicular maldescent; in such cases, examination of the external genitalia should be performed by a specialist to exclude disorders of sexual differentiation. In cases of bilateral impalpable testes, or in cases with other abnormalities of the external genitalia (e.g. hypospadias, bifid scrotum), urgent paediatric surgical/urological referral is warranted.

To fully understand the different forms of testicular maldescent, a basic understanding of the embryology and pathophysiology of normal testicular descent is required.

Normal Mechanisms of Testicular Descent

There are two independent stages of testicular descent; the first stage termed the *abdominal phase* occurs between 8 and 15 weeks of gestation. The gubernaculum tethers the developing gonad to the region of the internal ring resulting in its migration to the location in the developing male fetus during the second trimester. This phase is dependent upon anti-Mullerian hormone (AMH) and functioning AMH receptors but is independent of androgens and functional androgen receptors. If this first stage of testicular descent fails them, the testis remains intra-abdominal.

The second phase of testicular descent is more complex, and as such, more commonly goes awry. The second phase of testicular descent is termed the *inguinal phase*. After the testis has reached the internal ring, the gubernaculum continues to expand and extends into the scrotum. Under the influence of androgens, virilisation of the genitofemoral nerve results in the local production of calcitonin gene

related peptide (CGRP) which is through to cause the gubernaculum to rhythmically contract, drawing the testis through the inguinal canal to its ultimate destination within the scrotum. It is clear that testicular maldescent can occur for a variety of different reasons as it is dependent upon complex series of sequential events. For successful descent of the testis into the scrotum to occur, we must have:

- A correctly sited gubernaculum
- Adequate fetal virilisation
- Normal hormonal balance during the third trimester

Emerging evidence suggests that disruptions in the normal fetal hormonal balance caused by disturbances in intrauterine hormonal function are important contributors to testicular maldescent. Commonly cited risks for testicular maldescent include:

- Family history of undescended testis
- Maternal history
- IUGR
- Prematurity
- Intrauterine exposure to high levels of oestrogen

However, the causal mechanisms behind these observations are yet to be established.

There is evidence that testes that fail to descend normally are intrinsically abnormal (both macroscopically and microscopically). Histological studies have demonstrated that dysplastic elements are commonly seen. These abnormalities in combination with the non-physiological temperature environment of the undescended testis are thought to contribute to the increased risk of malignant transformation of undescended testes. The temperature within the scrotum is around 3°C lower than body temperature. The complex nature of the mechanisms underpinning normal testicular descent into the scrotum results in the relatively common incidence of testicular maldescent.

Diagnosis

It is important to take every opportunity to examine the external genitalia of male infants when they come to see a doctor or healthcare professional. This is of particular importance in areas where healthcare resources are unevenly distributed such as remote practice. The first opportunity to do so is at the routine baby check. For the vast majority of male infants, it is relatively straightforward at this stage to palpate both testes in the scrotum, and it is usual for it to remain straightforward for the first few months of life. Once children are older, and the cremasteric reflex becomes more established in male infants over 6 months of age, the assessment of testicular position is more challenging. This is particularly true if the child is frightened, upset, cold or crying, and in such instances, parents will often be able to recall if they have seen the testes in the scrotum. Ideally, the physical examination should take place in an unhurried manner, in a warm room with a calm child. A well-developed hemiscrotum is usually a sign that a testis has at least at some point been within the scrotum. Conversely, an underdeveloped hemiscrotum suggests that the testis has not come to occupy a satisfactory scrotal position, as normal scrotal development and virilisation are dependent upon local production and tissue response to androgens by a testicle situated within the hemiscrotum. If the testis is palpable, then it may be mobile, but it is important to determine testicular volume and position and to recognise whether it can be drawn to the base of the scrotum and whether it reascends upon release.

Classification of Testicular Maldescent

If a testis is maldescended, it can be divided into two major categories, *palpable* (80%) or *impalpable* (20%). For palpable testes, these can then be classified according to their position in relation to the inguinal canal. Impalpable testes are either absent (50%), intra-abdominal (40%) or canalicular (10%). Most commonly undescended testes sit in the superficial inguinal pouch, whereas retractile testes occupy a high scrotal position but can be drawn into the hemiscrotum and remain there without tension upon release. When a testis has been documented as palpable and descended but comes to occupy a less satisfactory position as the child grows, this is termed the *ascending testis*, although the author prefers the term 'stationary testis' as it more precisely describes the fixed position of the testis in relation to the external ring.

Management of Testicular Maldescent

Current guidelines recommend referral between the ages of 3 and 6 months for boys with unilateral, uncomplicated testicular maldescent. Those with complicating features, such as bilateral impalpable testes or additional abnormalities of the external genitalia (such as hypospadias or bifid scrotum), should be referred urgently to paediatric surgical or paediatric urological services dependent upon local availability.

Patients with testicular maldescent should undergo corrective surgery as soon as is safe and practical. Some surgeons now advocate earlier surgery than previously recommended (between the ages of 3 and 6 months). Theoretically, this will improve long-term prospects for fertility and reduce the longer-term risk of carcinogenesis; however, most surgeons outside of large specialist centres aim to perform surgery around 9–12 months of age when the risk of complications and general anaesthesia are considered to be somewhat lower. In practice, it is important to refer patients as soon as is practical if testicular maldescent persists beyond 3 months of age to allow for timely surgery.

Investigations

USS is widely performed prior to referral; however, it is not usually useful. It has a low sensitivity (78%) and even lower specificity (45%) in determining testicular position and in locating an impalpable testis. In general, clinical examination is superior in determining the need for surgery and in deciding what surgery will be required.

Surgical Management

Testes that are palpable are usually relatively straightforward to deal with operatively. These patients should ideally undergo an open orchidopexy between 9 and 12 months of age. Through an inguinal incision, the testis is identified, and any hernia sac or fibrotic remains of the patent processus vaginalis are divided. This manoeuvre usually allows for the testis to be easily delivered to the base of the scrotum via a scrotal incision without tension. The testis is then placed in a subdartos pouch and secured to the median raphe with absorbable sutures. Closure of the scrotum is best performed with an absorbable suture such as chromic as it is my experience that rapidly absorbing sutures such as vicryl rapide absorb too quickly in the tropical environment. The success rate for an open orchidopexy is reported to be around 90–95%, although complications such as testicular atrophy and reascent do occur and should be openly discussed with the family as part of the consent process, particularly in patients with bilateral testicular maldescent.

Patients with impalpable gonads require a slightly more involved approach. Most surgeons choose to examine the child under anaesthesia and if the testis remains impalpable under these conditions proceed to laparoscopy to determine whether the testis is absent, intra-abdominal or canalicular. In the case of a solitary testis, it is my usual practice to fix the remaining testis because of the subsequent risk of testicular torsion. This is particularly pertinent in remote practice, as the chances of being able to get to hospital for scrotal exploration in a timely manner are significantly lower than for those patients living in metropolitan areas. If the testis is intra-abdominal, it is possible to a single-stage or more commonly a two-stage laparoscopic Fowler–Stephens procedure. In this operation, pioneered in Melbourne, the testicular vessels are divided and the testis freed and mobilised on the vas deferens (and it's attendant vessels), prior to subsequent laparoscopic or open placement into the scrotum. Very high success rates have been reported for the two-stage procedures, but I suspect the true rates of long-term testicular survival without reascent are in the range of 70–80%.

Prognosis

Parents are usually quite reasonably concerned about the outlook for fertility and the longer-term risk of testicular cancer in boys with testicular maldescent. As previously mentioned, histological studies of the undescended testis have demonstrated histological abnormalities in the undescended testis from a very early age. It is not uncommon during operation to demonstrate testicular and epididymal abnormalities (e.g. testiculo-epidiymal dislocation), and although the clinical significance of these findings has not been examined in any great detail, then it would seem likely that these will have some impact on sperm production or delivery into the ejaculate. A complete lack of germ cells has been reported in patients as young as 15 months, and there is some understandable concern that testes should be brought into the scrotum as soon as possible.

Despite these concerns, the outlook for fertility is quite good for boys with unilateral testicular maldescent. In larger studies, only small differences in sperm count have been demonstrated in men who have undergone unilateral orchidopexy and even smaller reported differences in self-reported paternity (89% compared to 94% in the normal population).

Unsurprisingly, for boys with bilateral testicular maldescent, the long-term results of surgery have been less encouraging. The infertility rate has been reported to be up to 50–60%, with associated significantly lower sperm density when compared to controls. Theoretically, early surgery may be of some benefit to this group, although currently, there is a paucity of long-term follow-up studies to support this.

The subsequent long-term risk of testicular cancer has been approximated as between 1 and 2% in patients with undescended testes. It is for this reason that it is essential to ensure a satisfactory scrotal position of the testis as soon as possible to allow young men to adequately perform routine testicular self-examination. Estimates as to the relative risks of malignancy lie between 5 and 10 compared to the background population, but this is dependent upon a number of other factors most notably testicular position at the time of operation with the highest risk being seen in those boys with late recognition and treatment of intra-abdominal testes.

Summary

- Testicular maldescent is common (1:30–60).
- Surgery is the only effective treatment currently for testicular maldescent.
- Subfertility is largely a concern for boys with bilateral testicular maldescent.
- The increased relative risk of malignancy requires long-term follow-up, and routine testicular self-examination should be encouraged.

NECK SWELLINGS

It is common for children to present with parental concerns regarding a lump in the neck. Whilst they are usually managed in primary care, by paediatricians or by ENT surgeons, it is not uncommon for children to be referred to paediatric surgeons for further investigation and management. In general, it is a helpful approach to divide masses up in relation to their position in the neck. Swellings may be midline or lateral and may increase rapidly in size or be present over a longer period of time. Careful history taking and physical examination often allow a differential diagnosis to be established prior to deciding the appropriate sequence of investigations. Some of the commoner lesions are listed in the following table:

CONGENITAL	*INFECTIVE*	*NEOPLASTIC*
Midline	Acute	Benign
Thyroglossal cyst	Reactive lymphadenopathy	Pilomatrixoma
Ectopic thyroid	Lymphadenitis	Lipomas
Dermoid cyst	Abscess	Neural tumours
Lateral	Chronic	Malignant
Branchial cleft/arch anomalies	Atypical/non-TB mycobacterial	Lymphoma
Torticollis	TB	Thyroid malignancies
SCM tumour	Toxoplasmosis/HIV	
	Goitre	
Other		
Haemangiomas		
Vascular: AVM/venous		
Lymphangioma		

Conditions of the Thyroid

Persistent thyroglossal duct and thyroglossal cysts

A persistent thyroglossal duct is one of the most common lesions in the midline of the neck and occurs in approximately 7% of the population. They generally present with symptoms typically seen in the pre-school age: 3–5 years old. An example of a child with an infected thyroglossal cyst can be seen in Figure 7.4.

Basic embryology

The thyroglossal duct is a congenital remnant which connects the posterior aspect of the tongue to the thyroid gland during embryological development. It originates from the foramen caecum which is where the thyroid diverticulum develops. At weeks 4–7, the thyroid migrates inferiorly from the foramen caecum past the base of the tongue and hyoid bone. The thyroid remains connected to the foramen caecum by the thyroglossal duct which is lined by lymphoid and epithelial cells. By the 3rd month, the duct typically disintegrates as the ductal walls adhere to itself and obliterate the canal.

Pathophysiology of thyroglossal duct remnants/cysts

In some children, the thyroglossal duct fails to involute completely. When mucus is secreted from the epithelial cells lining the duct, this subsequently prevents the adherence of the ductal walls. Collections of mucus in pockets within the duct allow cysts to form.

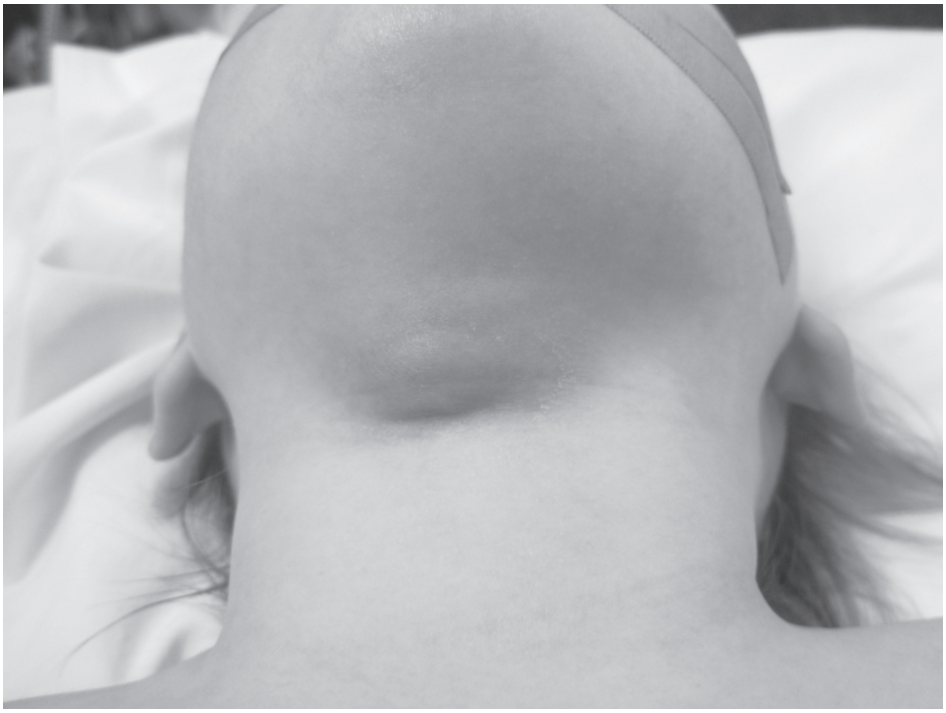

FIGURE 7.4 Infected thyoglossal cyst.

Clinical presentation

Overall, 75% of these occur near or attached to the hyoid bone, whilst others occur in the suprahyoid or subhyoid position. Children will typically present with a painless swelling in the midline of the anterior neck. This mass moves with swallowing or protrusion of the tongue due to the embryological attachment discussed earlier.

Due to its proximity and the presence of lymphoid cells, local inflammation stimulates mucus secretion and thus increases the size of the cyst. Other symptoms or complications include erythema and pain (33%), a discharging sinus (25%) or a foul taste depending on the patency of the duct. Erythema and pain are symptoms associated with an infected cyst. Thyroglossal cysts are usually diagnosed clinically but can be further evaluated with a US or CT scan if unclear.

Management of thyroglossal cyst

Primarily, the goal of treatment is to excise the entire tract – the Sistrunk procedure. But if it is infected, a course of antibiotics may be required prior to excision.

Ectopic thyroid tissue

Occasionally, cells of the thyroid gland can be found in the thyroglossal duct. This is concerning as it may indicate a hypoplastic thyroid and increases the risk of a thyroid tumour developing in the cyst. Early suspicion of an ectopic thyroid arises from abnormal thyroid function levels which are completed as part of the newborn screen. On palpation, they may be softer than a thyroglossal cyst but may only be discovered on excision. Radioisotopes can be used to determine if the ectopic thyroid is the only thyroid tissue present at which excision should not occur.

Dermoid and Epidermoid Cysts

Dermoid cysts can be difficult to delineate from thyroglossal cysts. Like thyroglossal cysts, they also present as midline masses which move with swallowing. However, dermoid cysts are typically more superficial and mobile. Most dermoid cysts are in the suprahyoid or submental area and filled with sebaceous material. During surgical excision, examination of the contents of the cyst may help differentiate the two pathologies.

Epidermoid cysts are lined with epidermis and do not contain sebaceous or mucous material. These can arise congenitally or are acquired due to traumatic entrapment of epidermis into the dermis/subcutaneous tissue. They are associated with sinus tracts and can be mistaken as branchial cleft remnants. The presence of multiple epidermoid cysts should raise the suspicion of Gardner's syndrome. Both dermoid and epidermoid cysts are excised due to their risk of rupture and infection which follows.

Thyroid Malignancies

Thyroid malignancies are rare in childhood. They usually come to the attention of medical professionals due to parental concerns about a new swelling appearing in the neck. Thyroid malignancies are more likely to present with cervical lymphadenopathy than a swelling due to a goitre. Risk factors include radiation exposure including CT scans and radiation treatment for other conditions. Overall, 90% of paediatric thyroid cancer comprises papillary thyroid cancer. Follicular thyroid malignancies are rare, whilst medullary carcinomas are frequently diagnosed in association with MEN syndrome. Hodgkin's lymphoma (HL) is the most common malignancy associated with secondary thyroid malignancies. The main causes of goitres include Hashimoto thyroiditis, Graves' disease, iodine deficiency or excessive maternal iodine intake during pregnancy.

BRANCHIAL CLEFT/ARCH ANOMALIES

Basic Embryology of Brachial Cleft Abnormalities

During week 3 of embryological development, gastrulation occurs, and the three pluripotent germ cell layers develop – the endoderm, mesoderm and ectoderm. Within the pharyngeal apparatus which forms the primitive pharynx, these three layers correlate to the pouch, cleft and arch. Initially, there are six branchial arches from the pharyngeal apparatus; however, the fifth typically regresses; thus, only five remain. Branchial cleft or arch anomalies usually develop from remnants of the first, second and third branchial systems.

The second branchial system is the most common pathology and starts in the tonsillar fossa. It then passes next to the glossopharyngeal nerve, between the internal and external carotids to the anterior border of the lower third of the sternocleidomastoid muscle (SCM).

The first branchial system anomalies manifest uncommonly, and its path begins at the external auditory canal and passes to the cutaneous surface below the inferior border of mandible.

Rarely, the third branchial system persists with the opening lying internally at the piriform sinus and traces to the cutaneous surface of the anterior border of the sternocleidomastoid.

Branchial arch anomalies present as atypical mesodermal remnants along the arch's development. The most common is the second branchial arch remnant which presents as a skin tag at the cutaneous opening. These are excised generally for cosmetic reasons.

Persisting branchial clefts (between arches) can form sinuses, fistulas or cysts. Branchial sinuses are the most common and are blind-ending canals with a cutaneous orifice whilst fistulas occur when there

is a communication between two epithelial lined surfaces. They typically present with mucous leaking from the opening, or a recurrent wet spot noted on clothing. Tracts can occasionally be palpable and may excrete mucus when massaged.

Sinus and fistulas are usually lined by respiratory epithelium, whilst cysts are typically lined by squamous epithelium. Rarely, branchial cysts persist and are remnants of sinuses without a cutaneous orifice. All three pathologies are at risk of infection and require surgical complete excision to prevent recurrence. Depending on the clinical suspicion, further investigations including injection of radio-opaque liquid into the orifice, or a US/CT/MRI can be used to define the tract.

Sternocleidomastoid pathology

Congenital torticollis is a postural deformity commonly caused by the fibrosis of the SCM, causing shortening and the deviation of the neck *towards the side of the lesion*. It has been associated with a muscular injury sustained during a difficult birth or even prior to delivery. A suspicion of acquired torticollis should involve an investigation into ophthalmological (strabismus), otolaryngological (infection) or gastrointestinal pathology (gastro-oesophageal reflux disease [GORD]).

It is typically seen unilaterally but may occur bilaterally. Unilaterally, the infant has a classical appearance – ***the head is rotated away from the side of the lesion and with contralateral lateral flexion***. Most cases resolve with conservative management involving physiotherapy. Surgical intervention is considered when conservative management fails, for patients with a severe rotational deficit, or for those with prolonged symptoms.

Physical examination of a unilateral torticollis with a sternocleidomastoid mass is clinically suspicious of a sternocleidomastoid tumour. Key features include a 2–3 cm long hard, painless and spindle-shaped swelling in neonates 2–3 weeks old. It is commonly associated with plagiocephaly due to the impressionable skull and prolonged lying with the positional preference.

Management of torticollis

Similar to congenital torticollis, conservative treatment until 6 months allows 90% of cases to self-resolve. Indications for surgical intervention include persistent torticollis >12 months or signs of facial hemi-hypoplasia. Despite surgical intervention, facial asymmetry will improve over years but may not completely resolve.

Cervical Lymphadenopathy

Lymphadenopathy of the neck manifests in the acute period as a reactive response usually to an upper respiratory tract or ear infection. This may be due to a bacterial, viral or mycobacterial cause. Palpable lymph nodes may be tender and are expected to resolve as the infection improves. Occasionally, reactive hyperplasia occurs as a non-tender mass with a non-specific cause.

Acute lymphadenopathy can progress to lymphadenitis when the lymph nodes become infected. Lymphadenitis requires rest and simple analgesia, and if precipitated by a bacterial infection, IV antibiotics are recommended. With IV antibiotics, lymphadenitis may progress into a lymph node abscess. These are typically seen in the 6-month to 3-year-old age group whereby the infected lymph node continues to enlarge despite antibiotics. They may be fluctuant on palpation, but deeper nodes may not be palpable. It is typically associated with superficial skin changes such as overlying erythema, and if left treated, an abscess may form. In cases of clinical uncertainty, an USS of the area may be used to assess an enlarged lymph node to look for evidence of a collection forming. Lymph node abscesses require an incision and drainage.

Mycobacterial Infections of the Head and Neck

Mycobacterial infections can be divided into tuberculosis and atypical (non-tuberculosis) mycobacteria. Atypical mycobacteria include *Mycobacterium avium-intracellulare*, *Mycobacterium scrofulaceum, Mycobacterium fortuitum* and *Mycobacterium chelonae*.

These infections are rarely seen in Western countries and are transmitted through oral ingestion of infected soil. Atypical mycobacterial lymphadenopathy is non-tender and affects lymph nodes of the cervical and submandibular chains higher up in the neck. Up to 50% of these infections with atypical mycobacterium subsequently form *cold abscesses*. As these lymph nodes tend to be deep, they do not display the expected superficial erythema or palpable fluctuance. Instead, atypical mycobacterial lymph node abscesses rupture in a collar-stud distribution within the subcutaneous space causing a superficial blue-purple discolouration. Overall, 10% of these then ulcerate to form multiple discharging sinuses at the skin.

Atypical mycobacterial lymph node abscesses respond poorly to antibiotics and require surgical excision of the lymph node. Their surgical management can be difficult, and different approaches have been advocated by some surgeons, particularly for MAIS lesions on the face where complete excision risks damage to the facial nerve.

In lymphadenopathy caused by tuberculosis, the condition is primarily a pulmonary infection which causes supraclavicular lymphadenopathy. Because of this, the typical management for non-TB mycobacterial lymphadenopathy differs from TB lymphadenopathy. TB responds well with chemotherapy over a span of 2 years whilst atypical mycobacterial lymphadenopathy requires surgical excision and could be considered for antibiotic treatment if an excision would be anatomically challenging – e.g. in a deep location or near the facial nerve.

Tuberculosis is the world's second leading infectious killer second to COVID-19. Multi-drug-resistant (MDR) TB is a public health crisis and poses a challenge for TB eradication. In Australia, there is a low incidence of TB; however, there is an over-representation of immigrants and First Nation's Australians. Epidemiological studies have found that the highest rates are found in children born overseas, and the annual notification rate was three times higher in First Nation's Australians compared to Australians not of First Nation background. Papua New Guinea (PNG) has been speculated to be epidemic with MDR TB and HIV. The cross-border spread into Australia from PNG is highlighted where 77% of paediatric TB cases in 2003–2012 were from PNG. Treatment outcomes for children of PNG heritage with TB in the Torres Strait were less favourable compared to Australian children. In a study evaluating transmission pathways within a community outbreak, cultural and family relationships were identified as key factors contributing to TB transmission. With a strong emphasis in cultural and family relationships in both First Nation Australian and PNG cultures, the elimination of TB becomes more difficult. Queensland faces a unique challenge in TB control compared to the rest of Australia given its proximity to PNG and First Nation Australian population, particularly in North Queensland. It is not unexpected that there is an over-representation of both cross-border and First Nation Australian children with TB.

Rarer Causes of Cervical Lymphadenopathy

Other conditions for consideration include cat scratch disease (*Bartonella henselae*), toxoplasmosis, Kawasaki disease and AIDS. Lymphadenopathy which persists in the subacute period (2–6 weeks) is likely to be viral. Viral lymphadenopathy like viral illnesses is treated with supportive measurements, such as rest, hydration and analgesia. Biopsies are indicated in cases of chronic lymphadenopathy (>6 weeks), an unclear diagnosis or when lymph nodes are >3 cm. Lymphadenopathy in the supraclavicular and posterior triangle are more concerning for malignancy or tuberculosis, whilst those located in the submandibular and anterior cervical are commonly infective.

MALIGNANCIES OF THE HEAD AND NECK IN CHILDREN

Lymphoma

Lymphoma occurs when there is uncontrolled proliferation of lymphoid cells in lymphoid tissue. Primary causes of lymphoma are categorised as *Hodgkin's* and *non-Hodgkin's lymphoma* (*NHL*) which combined the third most common cause of cancer in adolescents.

NHL typically causes lymphadenopathy in the mediastinum (thymus) or abdomen. At the time of diagnosis, the child is usually symptomatic from nodal enlargement, and the NHL has likely spread to regional nodes and to other locations such as the neck or axilla. NHL responds well to chemotherapy and steroids. The surgical role of NHL is to aid in biopsy and cytological diagnosis rather than curative treatment.

HL originates in one group of lymph nodes and progresses following lymphatic pathways. Similar to NHL, surgery has limited involvement often only providing the tissue diagnosis in biopsies. Histologically, Reed–Sternberg cells are seen in HL. Surgical excision may be considered in certain cases whereby lymphoma is solely located in one area.

Cervical Rhabdomyosarcoma

Rhabdomyosarcoma is the most common form of sarcoma found in the paediatric population. It is usually diagnosed by the age of 14 and may be associated with familial syndromes – neurofibromatosis 1 and Li-Fraumeni. They arise from immature or satellite cells expected to become striated skeletal muscle cells but may also be found in atypical locations.

Within the neck, they may present as a painless mass with overlying skin changes. Occasionally, rhabdomyosarcomas may be suspected when a child presents with symptoms due to a mass effect on important structures. Excision of such lesions can be challenging due to its proximity to these structures and the overall cosmetic effect. Complete excision is typically recommended, but this should be conferred with a paediatric oncological specialist to discuss the role of chemotherapy or radiotherapy. Local lymph node biopsies may also be suggested in conjunction with imaging to determine the staging of a suspected rhabdomyosarcoma.

Secondary Malignancies

Secondary malignancies which may cause cervical lymphadenopathy include nasopharyngeal, thyroid neoplasms and neuroblastoma. These are present with hard and hyperplastic nodes which may be difficult to differentiate from reactive hyperplasia. Lymphadenopathies in these scenarios are biopsied for histopathological evaluation.

Skin Lesions of the Head and Neck

Pilomatrixoma or pilomatricoma, also known as calcifying epithelioma of Malherbe, is a benign tumour of hair follicles histologically defined by calcifications and ghost cells. It is the most common acquired soft tissue mass in children with 60–70% located in the head and neck. Pilomatrixomas are firm and non-tender mass that slowly grow and may be well circumscribed or irregular. They are located in the

intradermal or subcutaneous space producing a white, yellow or blue discolouration superficially with no overlying punctum. These are typically seen as solitary lesions; however, in 2–3% of cases, multiple lesions are associated with Gardner's syndrome. Pilomatrixomas are managed with an excision which can occur under local anaesthetic for superficial lesions. As they do not have a capsule, rupture of the lesion during excision is not unexpected and does not predispose recurrence.

Summary

- Neck lumps are common in childhood.
- Anatomical location and history often point to the examination.
- Targeted radiological investigations are often helpful in confirming a diagnosis.
- It is important to formulate a differential diagnosis to guide investigations.

FURTHER READING

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