usually affect the ventricular end of the catheter rather than the peritoneal end. Many children develop abdominal distension after shunting. This is due to the unusual load of CSF. In the absence of vomiting and constipation, it should be treated conservatively.

- Shunt infections can present acutely with features of cerebral irritation, fever and seizures if there is major ventriculitis occurring within a few days of insertion; however, this is relatively rare. The only method that is guaranteed to eliminate shunt infection is removal of all components, including any loose or retained fragments from earlier procedures, interval external drainage, appropriate antibiotics and shunt reinsertion through fresh incisions. As with all serious infections, success is dependent upon accurate microbiological diagnosis. The most frequently encountered organisms are Staphylococcus epidermidis and Staphylococcus aureus. The most useful antibiotic is vancomycin by IV injection children 1 month to 18 years 15 mg/kg every 8 hours to a maximum daily dose of 2 grams. The duration of treatment depends on how rapidly the CSF becomes sterile, but a minimum of 7 days is recommended.

### Myelomeningocele

Myelomeningocele is the commonest major congenital malformation compatible with survival. Its incidence has been progressively falling for 20 years. Although there are regional variations, the overall frequency is 0.7–0.8 per 1000 live births. The objective of management in the immediate postnatal period is the prevention of infection of the central nervous system. This is achieved by early closure of the lesion.

- The level of the open lesion is noted and an assessment made of the sensorimotor level, the state of the sphincters, any orthopaedic deformity, and the presence of major hydrocephalus, as evidenced by signs of raised intracranial pressure (see above).
- The ideal is to achieve closure within 24 hours of birth. The majority of lesions have adequate skin in the wall of the sac, as long as this is not unnecessarily sacrificed by a wide incision. The technique employed involves mobilisation of the neural placode, watertight dural repair and closure of the skin. While awaiting closure, the lesion should be protected with a dressing of moist sterile 0.9% saline, which must be replaced every few hours to prevent desiccation.
- Most babies will require surgical treatment for hydrocephalus in the first few weeks of postnatal life.

In children who are paralysed and without urinary or bowel control, the commitment is a lifelong one, and this is a challenge to families and healthcare systems. Before offering treatment to these children, it is important that their future prognosis and quality of life is discussed with the parents. The aim should be to prevent as many as possible of these anomalies by adequate maternal nutrition prior to conception and during pregnancy.

**Folic acid taken prior to conception and for the first trimester of pregnancy abolishes 75% of cases of myelomeningocele and anencephaly.**

See Section 5.10.A.

### 5.17 Orthopaedic problems

**80X 5.17.1 Minimum standards**

- Antibiotics.
- X-rays.
- Erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP).
- Antituberculous drugs.
- Orthopaedic procedures.

**Introduction**

Injuries are by no means the only paediatric orthopaedic problems in resource-limited countries. There is a great burden of orthopaedic infective conditions which, if treated suboptimally, can lead to considerable handicap. Furthermore, there is the same spectrum of non-infective conditions as is seen in well-resourced countries which, due to the limited resources available in under-resourced healthcare systems, represents a considerable diagnostic and therapeutic challenge.

**Infections**

Paediatric musculoskeletal infections are a common presentation in resource-limited countries. Morbidity and mortality can be prevented by prompt diagnosis, antibiotics and surgery where indicated. Infection should be suspected in any child presenting with pain or swelling in the limbs, spine or pelvis.

**Pyomyositis**

- Pus is present within skeletal muscle, most commonly in the thigh and gluteal regions.
- It is caused by bacterial infection of muscle, in nearly 70% of cases due to Staphylococcus aureus.
- It is common in the tropics, but exceedingly rare in the developed world.
- There may be a history of previous injection or trauma to the site.
- Signs include general malaise, swinging fever, decreased range of motion, fluctuant swelling in the later stages, and tenderness.
Investigations

- In resource-limited countries, clinical examination is the mainstay of diagnosis.
- White blood cell count is unreliable.
- Erythrocyte sedimentation rate is raised in 90% of cases.
- Blood culture is positive in 40–50% of cases.
- Plain X-rays: bony changes take 7–14 days.
- Aspiration and Gram stain; look for acid-fast bacilli.
- Bone scan (if available).

**Acute haematogenous osteomyelitis**

- The causes are unknown.
- Infection starts in metaphyseal venous sinusoids.
- There is thrombosis of the vessels.
- Pus develops in the medullary cavity, leading to a build-up of pressure.
- If untreated, pus bursts through the cortex and spreads under the periosteum, rendering bone ischaemic (see chronic osteomyelitis below).
- In infants and children the pathogen is almost always *Staphylococcus aureus* (for neonates, see below). The exception is in sickle-cell disease, where *Salmonella paratyphi* is common. In this situation, use cefotaxime or ciprofloxacin.

**Diagnosis**

- Any child with fever and unexplained bone pain.
  - High index of suspicion.
  - Around 50% of cases will have a history of recent infection.
  - The child refuses to move the affected limb.

**Operations**

- Undertake incision, drilling and drainage of the osteomyelitic abscess.
- Mark the area of maximal swelling and tenderness prior to anaesthesia.
- Make a longitudinal incision.
- Dissect on to and incise the periosteum.
- Drill the cortex of the bone. If there is no pus at one site, drill further holes proximally and/or distally until pus is obtained.
- Copious irrigation is needed.
- Leave the wound open, and apply a dry or antiseptic dressing.
- Monitor post-operatively for recurrence and other foci of infection, and leave the wound to granulate.

**Neonatal osteomyelitis**

- There are several features unique to neonatal osteomyelitis.
  - In the neonate, metaphyseal vessels communicate with epiphyseal ones, thus permitting the spread of infection into the epiphysis and ultimately into the joint. Therefore acute haematogenous osteomyelitis and septic arthritis may occur together. This can lead to complete lysis of areas such as the femoral head and neck and the proximal humerus, or premature physeal arrest.
  - As the immune system of the neonate is immature, there may be a less marked inflammatory response to infection, with an absence of fever, raised white blood cell count or erythrocyte sedimentation rate.
  - Multiple foci of infection are more common.
  - A wider spectrum of infecting organisms is found (not only *Staphylococcus aureus* but also group B streptococci and Gram-negative coliforms).
  - Antibiotic treatment consists of gentamicin plus flucloxacillin.

**Subacute haematogenous osteomyelitis**

- This differs in presentation from acute haematogenous osteomyelitis in the following ways:
  - It often has an insidious onset.
  - The clinical signs are less marked.
  - Investigations may be inconclusive or equivocal.
  - The location is usually metaphyseal, with plain X-rays showing a solitary lytic lesion (abscess) with a sclerotic margin.
  - The differential diagnosis includes a neoplasm.

The usual causative organism is, as for acute haematogenous osteomyelitis, *Staphylococcus aureus*.

**Treatment**

- Prior to the formation of pus in the medullary cavity, antibiotics alone may suffice.
- Due to the predominance of *Staphylococcus aureus* as the causative organism, the initial antibiotic should be flucloxacillin while culture results are awaited (50 mg/kg IV or orally (maximum individual dose 2 grams) 6-hourly for 3 weeks).
- Once an abscess has formed this should be drained surgically.

**Operative treatment**

- If diagnosed early (which is unusual), pyomyositis may respond to antibiotic therapy (flucloxacillin), but most cases will require incision and drainage of the abscess under general anaesthesia.

**Pathogenesis**

- The usual causative organism is *Staphylococcus aureus*.
- This differs in presentation from acute haematogenous osteomyelitis, in that the infection may spread more rapidly and involve multiple bones and joints.

**Treatment**

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**Operative treatment**

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Chronic osteomyelitis

If acute osteomyelitis goes untreated, the pressure due to the intramedullary pus eventually increases until it bursts through the cortical bone into the subperiosteal space. If still undercompressed, the pus spreads proximally and distally, stripping the periosteum and thus rendering this cortical bone ischaemic (having been deprived of both intramedullary and periosteal blood supply).

The avascular cortical bone therefore dies and becomes a focus of chronic infection called a ‘sequestrum’. Simultaneously, a periosteal reaction occurs under the stripped periosteum, resulting in the laying down of new bone or ‘involucrum’.

The appearance on plain X-ray is characteristic, with sclerotic sequestrum separated (by the abscess cavity) from an irregular and enveloping involucrum. Chronic osteomyelitis is difficult to treat even with optimal resources. Some guidelines on its management are as follows:

- If an osteomyelitic abscess is beginning to point, or there are signs of an underlying abscess, this should be incised and drained.
- In weight-bearing bones there should be no attempt at removal of sequestrum until the overlying involucrum is mature. This maintains the potential for weight bearing and ambulation.
- Periods of immobilisation should be minimised in order to retain ranges of motion and function of nearby joints.
- Sequestrum that begins to point through the skin can be removed or excised.
- In many cases the clinical picture that results is one of intermittent flare-ups of infection which can be treated by incision and drainage of abscesses, excision of sequestrate, and antibiotic (flucloxacillin) suppression of infection as required.
- Curative treatment is often elusive even in specialised centres, and a degree of morbidity is unfortunately inevitable.

Septic arthritis

Septic arthritis is infection of a synovial joint.

Features

- It is more common in males than in females.
- The peak incidence is at around 2 years of age.
- The first symptom may be a reluctance to use the limb.
- There is a swollen tender warm joint with a restricted range of motion.
- There is commonly fever (38–40°C).
- The patient is usually systemically unwell.

Diagnosis

- The mainstay of diagnosis is clinical examination.
- The white blood cell count is raised in 30–60% of cases.
- Elevation of the erythrocyte sedimentation rate is more sensitive (except in the neonate).

Plain X-rays are often normal until there is evidence of bone destruction at 7–14 days. Common pathogens include Staphylococcus aureus, Haemophilus influenzae, group A and B streptococci, pneumococci and Gram-negative coliforms (in neonates).

Aspiration of the joint is the definitive test.

Treatment

- Antibiotic therapy should not begin until after joint aspiration and blood cultures have been taken.
- Start with flucloxacillin (infants and children) or flucloxacillin and gentamicin (neonates).
- Some studies have shown that a combination of aspiration and antibiotic therapy is sufficient treatment, but this must be followed by close monitoring to ensure improvement.
- If the child fails to improve, surgical washout and drainage is required either via open arthrotomy or by arthroscopic means (but only if a skilled operator and equipment are available).

Post-operative care

- Continue antibiotic therapy, and monitor for recurrence.
- Early mobilisation of the affected joint is needed to prevent stiffness.
- If treated early, the prognosis for functional recovery is good. However, if presenting late there may already have been destruction of the articular surface.
- Be alert for coexisting osteomyelitis, which is present in around 15% of cases of septic arthritis.

Tuberculosis

Tuberculosis as an entity is covered in detail in Section 6.1.N, but it is important to remember the potential orthopaedic manifestations.

- It can cause both osteomyelitis and septic arthritis.
- In both cases the signs are less marked than in their non-mycobacterial forms, and the history is usually more chronic.
- It may be associated with systemic manifestations of tuberculous disease (respiratory and renal).
- Spinal tuberculosis (Pott’s disease) can be the cause of both paraplegia and scoliotic deformity.
- Treatment consists of surgical drainage and curettage of abscess cavities combined with antituberculous chemotherapy. For chronic disease and joint destruction, spinal stabilisation and joint arthrodesis may be indicated.

Non-infective conditions

The non-infective paediatric orthopaedic conditions described below can be extremely difficult to treat in resource-limited settings. First, without any form of population screening procedure in place or comprehensive primary healthcare provision, many cases will present late. Secondly, the advanced diagnostic modalities (ultrasound and arthrography) that are needed to direct treatment may not be available. Finally, where surgery is indicated, the operative techniques often need highly specialist training and/or specialised resources such as internal fixation and perioperative fluoroscopy, which are unlikely to be available in most resource-limited countries.

Fortunately, the conditions described are rare, typically occurring at a rate of less than 0.1%. They thus present far less commonly than the orthopaedic paediatric infections, and cause a lesser burden of handicap overall.

Developmental dysplasia of the hip

Formerly known as ‘congenital dislocation of the hip’, this complex condition has now been renamed ‘developmental dysplasia’ in recognition of its variable characteristics, such
as the fact that it is not always present at birth, nor does it always feature hip dislocation.
- Reported initial (neonatal) rates range from 3 to 17 per 1000 live births, but the rate of established dislocation is much lower, at around 1 per 1000.
- The aetiology is multifactorial; increased rates are seen in female children, firstborns, breech position and oligohydramnios, and there is undoubtedly a genetic influence (increased rates are associated with a positive family history and affected siblings).
- Early detection depends upon neonatal screening, which is often not available in resource-limited countries.
- If screening is to be carried out, it should involve Barlow and Ortolani tests for newborns followed by subsequent re-examination and ultrasonography of suspected cases at 1 month of age.
- Plain X-rays are of limited use before 6 months of age.

**Treatment**

As mentioned above, treatment of this condition in resource-limited settings is extremely difficult.
- Up to 6 months of age, gentle closed reduction can be undertaken, and then maintained in a Pavlik harness.
- If a Pavlik harness is unavailable, a plaster spica, maintaining the hips in flexion and abduction, will achieve the same objective.
- In children over 6 months of age, closed reduction can still be attempted, but is increasingly less likely to be successful due to the interposed joint capsule preventing stable concentric reduction.
- If closed reduction fails, open reduction can be attempted if surgical skills allow and infection is avoided.
- Later presentation with proximal femoral and acetabular abnormalities may require complex secondary reconstructive procedures, but these can only be undertaken in specialist hospitals by a specialist surgeon.

The reality of developmental dysplasia of the hip in resource-limited countries is that cases will often not present until after the age of 18 months, when the child has failed to walk or has an obviously abnormal gait. By this time bony abnormalities may require complex secondary reconstructive procedures, but these can only be undertaken by an orthopaedic specialist surgeon.

**Congenital talipes equinovarus**

More than two-thirds of cases of talipes equinovarus occur in developing countries. Most children receive either no treatment or substandard care. This results in physical disability that is entirely preventable.

There are three classes of talipes equinovarus (clubfoot):
- **Postural:** this arises from intrauterine positioning, and resolves fully with passive stretching within a few weeks of birth. Parents can be trained to do this.
- **Congenital:** this arises in an otherwise normal child, and has varying degrees of severity. It occurs in 1 in 1000 live births, and is bilateral in 30–40% of cases.
- **Syndromic:** this is associated with other syndromes, such as arthrogryposis, is often severe and is refractive to treatment.

**Treatment**

- The goal of talipes treatment is to obtain a functional plantigrade stable foot by the time the child begins to walk (i.e. before 1 year of age).
- If it is recognised in the neonatal period, gentle daily parental manipulation may be successful, or alternatively manipulation and taping by qualified healthcare professionals (e.g. a physiotherapist). Ponseti management has gained popularity as it does not require surgery and is easily learned (www.global-help.org/publications/books/help_cponseili.pdf).
- For cases that fail to resolve in the first 6–12 weeks, serial manipulation and plaster casting is indicated, with cast changes every 2–4 weeks.
- If the deformity still fails to resolve, there may be a place for limited percutaneous soft tissue releases (Achilles tendon or plantar fascia) at the age of 3–9 months. These techniques are relatively easily learned, have low morbidity, and are user-friendly in resource-limited settings. They should be combined with manipulation and casting.
- For the case that still fails to resolve, more extensive surgery, such as a postero-medial release, is required. The timing of this surgery is usually between 6 months and 1 year of age. Although specialist training is required to learn this operation, it can be relatively easily assimilated by the non-orthopaedic surgeon and, being only a soft tissue release, does not require any ‘high-tech’ surgical resources.
- Unfortunately, as with developmental dysplasia of the hip, children with this condition in resource-limited countries commonly present late (over 18 months of age), when the deformity is fixed and secondary bony changes have occurred. Correction at this stage requires a combination of bony and soft tissue surgery which can really only be undertaken by an orthopaedic specialist surgeon.
- In the adolescent child with fixed chronic deformity, the procedure of choice may be an arthrodesis (fusion) combined with correction of deformity performed at skeletal maturity.

**Perthes disease (Legg–Calve–Perthes disease)**

Perthes disease is a disease of uncertain aetiology involving a process of fragmentation and repair of the femoral head, possibly due to underlying idiopathic osteonecrosis.
- It usually occurs in susceptible children between 4 and 8 years of age, but can occur in children as young as 2 years or as old as 12 years.
- It is five times more common in boys, 10% of cases are bilateral, and it is associated with hyperactivity.
- It presents with a limping or waddling gait with groin, thigh or knee pain.
- X-rays show varying degrees and stages of fragmentation and repair of the femoral head.
- The prognosis depends on the degree of fragmentation and the potential for repair and remodelling prior to epiphyseal closure. A good prognosis is therefore associated with early onset and male gender (as the epiphyses close later).

**Treatment**

- In the majority of cases no specific treatment is indicated. The femoral head will heal and remodel satisfactorily, and the eventual outcome will be good. Bed rest, activity
Restriction and abduction braces have no proven impact on the natural history of the disease.

- In the small proportion of cases that may benefit from surgery, the issue is containment. A very deformed femoral head may not sit or move properly in the acetabulum, and thus leads to secondary arthrosis. A proportion of these cases may benefit from varus osteotomies of the proximal femur or pelvic osteotomies. Assessment for these procedures requires arthrography at the very least, and the procedures themselves are very much the preserve of the orthopaedic specialist surgeon in a specialist hospital.

**Slipped upper femoral epiphysis**

Slipped upper femoral epiphysis (SUFE) (also known as slipped capital femoral epiphysis, SCFE) is a disease in which the epiphysis becomes posteriorly displaced on the femoral neck.

- The prevalence is 1–10 per 100,000, and is higher in black populations.
- It is twice as common in boys as in girls, the at-risk age group being 10–17 years for boys, and 8–15 years for girls. Most affected children are obese, and in 40% of cases there is bilateral hip involvement.
- The aetiology is unknown, but is possibly endocrine related.
- The onset may be abrupt or gradual. Sudden slips present with severe pain and inability to walk; chronic slips present with pain often referred to the knees, a slight limp, and limited internal rotation of the hip.
- Plain antero-posterior and lateral X-rays are the most important diagnostic investigations. Severity can be classified according to the degree of epiphyseal displacement. Greater than 30% displacement can result in premature osteoarthrosis.

**Treatment**

- The goal of treatment for SUFE is to stabilise the slippage and to promote premature fusion of the epiphysis if possible.
- Ideal treatment is fixation in situ with a single cannulated screw. Given the posterior position of slippage, the point of entry of the screw needs to be anterior on the femoral neck. This procedure needs to be done under fluoroscopic or X-ray control in a specialist hospital.
- Where internal fixation or peri-operative imaging is not available, an alternative would be spica cast immobilisation. However, this is often logistically difficult, and the physis may still be open even after cast removal.
- For the most severe degrees of slippage, in the hands of a specialist surgeon, reduction and fixation of the slip on the femoral neck realignment osteotomies may be indicated.
- The commonest complications of operative treatment for SUFE are chondrolysis and osteonecrosis of the femoral head due to vascular compromise.

**Genu varum and genu valgum**

Varying degrees of bowed knees and knock-knee are common in the paediatric population. Most of these are merely variants of the normal physiological knee-angle development appropriate to the child’s age. Very few will require any form of intervention.

- Normal development: Babies are born with a varus knee angle that reduces with growth to become neutral at 18 months to 2 years of age. Thereafter the knee becomes increasingly valgus, reaching a peak at 5–7 years, after which the angle gradually declines to the 5–9 degrees of valgus seen in most adults.
- Blount’s disease is a developmental condition that affects the proximal tibial physis and results in progressive varus deformity.
- Treatment of degrees of genu valgum and genu varum depends upon the age of the child and the severity of the condition. Bracing is of no proven benefit. Various corrective osteotomies are possible, but these should be restricted to those cases with functional handicap, and are certainly not indicated merely on cosmetic grounds.

**Scoliosis**

Scoliosis is deformity of the spine characterised by lateral curvature and rotation.

- The commonest cause of paediatric scoliosis in resource-limited countries is probably tuberculosis (Pott’s disease). X-ray appearances can be strongly suggestive of this diagnosis, and then antituberculous chemotherapy is commenced.
- The scoliotic deformity is described as idiopathic where there is no known aetiology. Contrary to popular belief, most idiopathic scoliosis is only of cosmetic significance; only the most severe cases will have any degree of cardiorespiratory compromise.
- Scoliotic bracing is expensive, has compliance problems and is unlikely to be available in resource-limited countries. If available it may have a role in slowing the progression of curves which are between 20 and 40 degrees.
- Curves that are under 40 degrees at the time of skeletal maturity are unlikely to progress further.
- Surgical correction requires a specialised hospital and a skilled fully trained surgeon and support staff.