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The Diagnostic Calendar
Birth  congenital dislocation
0-5   Perthes’ disease
10-20  slipped epiphysis
Adult  osteoarthritis, AVN, rheumatoid arthritis

Developmental Dysplasia of the Hip
- 5 to 20 per live 1000 births exhibit instability (up to 2%), but 80-90% resolve spontaneously by 3 weeks.
- girls are affected 7 times more than boys; 1 in 5 bilateral (20%)
- left > right perhaps due to normal vaginal delivery being left occiput anterior – that is the left hip against the maternal spine?
- Genetic factors:
  - Northern & eastern Mediterranean
  - Generalized joint laxity (dominant)
  - Shallow acetabula (polygenic trait)
- Risk factors:
  - High levels of relaxin present in last few weeks of pregnancy
  - Rarer in Chinese & Africans who carry children astride their shoulders
  - Breech presentation
  - Oligohydramnios
  - Other congenital anomalies
- Otolani’s test: the hip is dislocated and there is a loss of smooth abduction of the flexed hip, though pressure beneath the trochanters may allow the head to relocate with a “clunk” and fully abduct.
- Barlow’s test: the hip is dislocatable – combined these are 60% sensitive and 100% specific (Jones 1998)
- Galeazzi sign for age 3 to 6 months, demonstrates LLD with the hips and knees flexed.
- X-ray signs:
  - Epiphysis should lie medial to a vertical line which defines the outer edge of the acetabulum (Perkins line), and below a horizontal line which passes through the triradiate cartilage (Hilgenreiner’s line)
  - The acetabular roof angle should not be > 30°
  - The femoral shafts should point to the acetabulum with the hips abducted 45° (Von Rosen’s lines)
- 0-6 months:
  - screen high risk babies or those with signs of instability
  - if dislocated start abduction splintage immediately, but if simply unstable then nurse in double nappies or abduction pillow
  - re-examine at 6 weeks and observe if stable, or else place in abduction splint if unstable, until x-ray evidence of satisfactory position (6 months)
  - risk factors for failure of Pavlik Harness include bilateral pathology, higher age at treatment onset, failure to reduce manually, and <20% coverage of femoral head on ultrasound.
  - If already dislocated and closed reduction works – apply Pavlik’s in reduced position and re-scan at weekly intervals.
- 6-18 months:
if delayed presentation, then closed reduction is preferable over three weeks using a vertical frame with gradual abduction ± adductor tenotomy

- splint with plaster spica at 60° flexion, 40° abduction and 20° internal rotation; replace at 6 weeks with a Pavlik harness for 3-6 months.
- X-ray follow-up to check position, and gradually abandon splint.
- Or closed reduction under GA carefully, with measurement of safe zone of Ramsay (maximum and minimum abduction in which hip is reduced). If Safe Zone < 25°, then release of adductor longus tendon, followed by plaster spica for 6-12 weeks with immediate CT scan to check reduction.

- Open reduction may be needed in which case the psoas is divided and redundant capsule ± thickened ligamentum teres (limbus) is excised. A derotation subtrochanteric osteotomy may be needed if stability can only be achieved by marked internal rotation of the hip.

18 months to 4 years:
- open reduction ± derotation osteotomy ± excision of segment of femur to reduce pressure on the hip and AVN.

![Figure 1 - Medial Approach](example.png)

- For a acetabular dysplasia which persists after 12-18 months (failed acetabular remodelling)
  - Pemberton’s capsular reconstruction of acetabular roof
  - Salter osteotomy where an osteotomy is performed through the ischium above the acetabulum. The acetabulum is then tilted inferiorly and anteriorly, with the defect filled with bone graft from the ilium and secured with a K-wire.

- Splint in plaster spica for 3 months and then a harness for further 1-3 months initially full-time and then at night, until acetabulum has remodelled and hip stable.

Over 4 years:
- If open reduction performed, often combined with pelvic osteotomy, as less potential for acetabular remodelling.
- Increasingly difficult to reduce and stabilize, with AVN and stiffness rates as high as 25%
- Non-intervention in child over 8 is justified as the child often has a mobile hip which is pain free, and only suffers from a waddling gait. In bilateral dislocations the waddling gait is less noticeable, and failure rates greater.
- AVN is the feared complication for which there is no treatment once established
  - Estimates up to 73%, just 1% with Pavlik Harness alone.
  - Mild cases are confined to the ossific nucleus, and supported by NWB while avoiding manipulation until the epiphyses heal. In severe cases the growth plates suffer and there is shortening and deformity of the femoral neck ± coxa vara.
- Late complications may require THR. Lengthening beyond 3 cm may lead to stretching of sciatic nerve, or damaging the profunda femoris artery.

- National guidelines 2008
  - Otolani and Barlow's test within 24 hours
  - If concern should have USS within 2 weeks and see Orthopaedic Surgeon by 4 weeks.
  - If breach or family history, then should have USS within 6 weeks and if abnormal should see an orthopaedic surgeon within 8 weeks.
Acetabular Dysplasia ± Subluxation

- May be congenital or follow incomplete reduction of DDH, or maldevelopment of femoral head (e.g. Perthes disease).
  - Also neuromuscular disorders, primary acetabular dysplasia, skeletal dysplasias.
- The acetabulum is usually shallow and open, with deficient coverage of the femoral head supero-laterally.
- In children it is usually asymptomatic and picked up radiographically. There may be a limp, groin pain or +ve Trendelenberg following strenuous activity.
- Older adolescents may complain of lateral hip pain from muscle fatigue, while adults are at risk of early secondary osteoarthritis.
- On x-ray Wilberg’s Centre Edge Angle should not be $< 30^\circ$ (normal $40^\circ$)
  - As the CRE falls, there is increased pressure and point loading on the lateral part of the acetabulum leading to degenerative changes.
  - OA associated with CRE$<15^\circ$, uncoverage $> 30\%$ or AI$<15^\circ$
- Crow Classification is assesses amount of proximal migration of the femoral head, and is based on the ratio distance migrated to the femoral head height
  - 1 = $< 50 \%$ subluxation
  - 2 = 50-75$\%$ subluxed
  - 3 = 75-100$\%$ subluxed
  - 4 = $> 100\%$ or superiorly dislocated.
- Treat infants as for DDH with abduction splintage, while older children may simply need muscle strengthening exercises.
- Surgical options for persistent pain, failure of AI to improve over 12 months or reach normal by 4 years. In older children, surgical indication is inadequate acetabular cover (CRE$<15^\circ$ or $>30\%$ uncoverage)
  - Goal is to change the shape ± direction, if joint is enlocated and congruent with a normal ROM.
Change in shape:

- *Pemberton* – lever acetabulum inferiorly with a wedge from crest to stabilise, but allows to reduce the acetabular volume and useful for a small femoral head.
- *Dega* – similar but osteotomy made closer to acetabulum and more laterally, giving more anterior or posterior adjustment

Change in direction:

- *Salter’s innominate osteotomy* – single
  - Maximum improvement in CRE is 25°, so femoral head needs to be containable with maximum of 25° abduction pre-operatively.
  - Unlike the Pemberton, the acetabular volume is not reduced.
- *Sutherland* – double (not often used)
- *Steele / Tonnis* – triple
  - Femoral head needs to be reducable as a pre-requisite
- *Ganz*
  - Peri-acetabular osteotomy and 3D-reorientation of entire acetabulum
  - Keeps posterior column intact

Salvage osteotomies – reserved for instances where concentric reduction not possible.

- *Chiari’s pelvic osteotomy*
  - step created to increase coverage
  - but results in fibrocartilage instead of articular cartilage.
- *Shelf procedure (Staheli)*
  - Can be used when complete reduction is not obtainable
- Total hip arthroplasty

**Femoral Anteversion (In-toe Gait)**

- Children usually walk awkwardly and trip over their own feet, and sit in the television position with their knees pointing inwards. There may also be a “squinting” patella on standing.
- < 3 years – due to forefoot adduction or tibial torsion
- >3 years – due to anteversion of the femoral neck (internal rotation of leg)
- Femoral anteversion seen with hips and knees extended, but not when both are flexed 90° (tibial torsion or forefoot adduction)
- The condition usually improves with growth with no bar on athletic prowess. Treatment is seldom indicated, but a femoral osteotomy may be considered in children over 8 years with a clumsy gait.

**Protrusio Acetabuli**

- Deep socket which may bulge into the cavity of the pelvis. X-rays show inner wall beyond the iliopectineal line.
- Primary form shows familial tendency, more common in girls developing after puberty.
  - Usually asymptomatic with the exception of reduced ROM
  - May lead to early secondary osteoarthritis
Secondary forms associated with softening disorder such as osteomalacia, Paget’s disease or chronic rheumatoid arthritis.

**Coxa Vara**
- Congenital form due to a defect in endochondral ossification of medial part of femoral neck. Continues to bend or fracture on crawling/standing
  - Leads to medial collapse with varus ± retroversion
  - Bilateral in one third of cases
  - Leg is often shortened and thigh may bow.
  - Look for a Hilgenreiner’s epiphyseal angle > 30°
  - Observe if angle 40 to 60°, but consider subtrochanteric valgus osteotomy if greater.
- Acquired causes:
  - Rickets
  - Perthes’ disease
  - Epiphysiolysis in adolescents
  - Osteomalacia
  - Fibrous dysplasia
  - Pathological fracture of malunited fractures
  - Paget’s disease
  - PFFD – Aitken A

**Proximal Femoral Focal Deficiency**
- Rare non-genetic disease, possibly teratogenic origin
- Aitken classification:
  - A – segment of unossified cartilage give appearance of a gap in the femoral neck or sub-trochanteric region. It will eventually ossify but only after proximal femur has developed a varus and shortening.
  - B – gap persists leading to acetabular and femoral dysplasia
  - C – femoral head is missing and acetabulum underdeveloped
  - D – agenesis of entire proximal femur and acetabulum
- Treatment:
  - A & B – joint is mechanically functional and can be treated with sub-trochanteric osteotomy and bone grafting to promote ossification. Limb-lengthening also possible.
  - C & D – Van Ness rotationplasty which is a tibial rotational osteotomy to turn the foot “back to front” to allow fitting of a prosthesis.
The Irritable Hip

- A transient synovitis of the hip which is more common in boys, usually presenting with hip/groin pain and a limp. Resolves within 2 weeks
- A diagnosis of exclusion with a systemically well child with normal blood parameters and x-rays; USS may show a small effusion. Exclude:
  - Perthes
  - SUFE
  - Tuberculous or septic arthritis (>50 x 10⁹ WCC/litre on aspirate)
  - Initial presentation of juvenile chronic arthritis and ankylosing spondylitis (uncommon)
- Usually treat with bed rest at home, but for severe effusions traction may be required in slight flexion and external rotation to reduce intra-articular pressure on the femoral head which can otherwise lead to AVN.
  - Sequential USS useful, and continue traction until both effusion and symptoms have resolved.
  - Repeat x-rays if symptoms persist beyond 2 weeks

Kocher’s Criteria for Septic Arthritis
Taken from JBJS (1999)

1. History of fever (oral temperature > 38.5°C)
2. Non weight bearing
3. ESR > 40 mm/hour
4. WCC > 12 x 10⁹ litre⁻¹

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<th>Incidence Septic Arthritis</th>
<th>Probability Septic Arthritis</th>
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<tbody>
<tr>
<td>0</td>
<td>22%</td>
<td>0%</td>
<td>&lt;0.2%</td>
</tr>
<tr>
<td>1</td>
<td>55%</td>
<td>1%</td>
<td>3%</td>
</tr>
<tr>
<td>2</td>
<td>19%</td>
<td>15%</td>
<td>40%</td>
</tr>
<tr>
<td>3</td>
<td>5%</td>
<td>54%</td>
<td>93%</td>
</tr>
<tr>
<td>4</td>
<td>0%</td>
<td>31%</td>
<td>99.6%</td>
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Another good predictor to distinguish between true septic arthritis (+ve joint fluid microbiology) compared with presumed septic arthritis (aspirate WCC>5010⁹ litre⁻¹ and +ve blood culture only) is history of rigors.

Infection in Paediatric Orthopaedics

- Purpura fulminans – meningococcal septicaemia complication with osteomyelitis, growth arrests and amputations
- Metaphysis susceptible to infection
  - High turbulent flow, low pressure and low oxygen tension
- Staph aureus
Most common pathogen
- 75% carriage
- gram positive, and coagulase positive
- 80% will produce penicillinase
- PVL (panton-valentine leukocidin) secreting results in severe septicaemia
  - Destroys host neutrophils and results in intra-vascular coagulopathy
  - Very difficult to drain as adherent and widespread bone involvement
  - Cavitating lung lesions as well

Types of osteomyelitis
- Acute pyogenic
- Subacute (Brodie’s)
- Chronic
- Periostitis (e.g. Typhoid)
- Tuberculous
- Multifocal

Primary haematogenous (blunt trauma and haematoma)
- Secondary haematogenous
- Direct inoculation
- Compound fracture
- Foreign body / implant

Natural progression:
- Bacteria divide in metaphysis (inflammation)
- Suppuration where there is metaphyseal abscess and intra-osseous pressure with pain
- Exits via nutrient foramen and spread sub-periosteally.
- Pressure and disruption of periosteal and endosteal blood supply → bone necrosis and sequestrum.
- Surgical Clearance requires a 5 mm margin – but in a child this would obliterate the growth plate. Thus we cannot achieve a good clearance and long-term antibiotics required.

Hip is different as the metaphysis is intra-capsular. Therefore when assessing a septic arthritis it is important to ask for a history of pro-dromal pain. This could represent an osteomyelitis and an MRI is needed.
- If a hyperacute history, then probably a pure septic arthritis.

Primary epiphyseal osteomyelitis can spread from metaphysis – as blood vessels may traverse the growth plate for first 18 months of life.

Management:
- History and examination
- Admit and observe
- Anti-pyretics and anti-inflammatory drugs – beware the effects
- Splintage and rest
- Bloods, blood cultures, throat swab and urine culture
- Antibiotics only if clinically unwell and unstable; otherwise a tissue diagnosis.
Perthes’ Disease

- Painful childhood disease characterized by AVN of the femoral head
  - Age group is 4-8
  - 4:1 ratio in boys
  - Incidence is 1 in 10,000 (very rare in black Africans)
  - Common in lower socio-economic class, or failure to thrive
  - Genetic link amongst those with an acquired thrombophilia (e.g. hypofibrinolysis, factor V Leiden & protein C&S deficiencies), low birth weight or increased maternal age.
  - 4% associated with a urogenital anomaly, and most have a reduced bone age.

- Blood supply to the femoral head:
  - Metaphyseal vessels penetrating growth plate
  - Lateral epiphyseal vessels within the retinacula (from medial circumflex)
  - Vessels of the ligamentum teres
  - The metaphyseal vessels gradually decline until virtually absent by age 4, and the vasculature of the ligamentum teres only matures by age 7. Therefore in between the femoral head relies on the retinacula vessels which are susceptible to stretching and venous occlusion by capsular tamponade (e.g. effusions).

- Stages:
  - 1: ischaemia and bone death; cartilaginous part being nourished by synovial fluid thickens (increased joint space on x-ray)
  - 2: revascularisation and repair with alternating zones of new lamellae and fibrous tissue (increased density, but fragmented appearance on x-ray)
  - 3: distortion and remodelling; if repair process is complete before the femoral head loses shape, the bony architecture can be saved. If tardy, the epiphysis collapses (crescentic subarticular fracture on x-ray) and the femoral head becomes mushroom-shaped and displaces laterally in the acetabulum.
  - Waldenstrom also described stages as sclerosis → fragmentation → deformation → re-ossification.

- Catterall Classification (of the femoral head)
  - 1: < 50% ossific nucleus sclerotic (anterior), epiphysis retains height
  - 2: < 50% sclerotic, with some central collapse
  - 3: most of the nucleus is involved, with fragmentation and collapse
  - 4: whole head involved, ossific nucleus is flattened & dense, with marked metaphyseal resorption
  - 1 and 2 has good prognosis, but 3 & 4 have poor prognosis

- Catterall Head at Risk criteria (if >2 indicate surgery)
  - Radiographic:
    - Cysts and Gage sign
      - V-shaped radiolucency in the lateral portion of the epiphysis and/or adjacent metaphysis
- Calcification lateral epiphysis
  
  *
  
  *loss of weight-bearing pillar allows collapse*

- Lateral subluxation
- Diffuse metaphyseal reaction
- Horizontal growth plate / physis

- Clinical:
  - Older or heavier child
  - Adduction ± flexion contracture
  - Progressive reduced ROM

- An arthrogram may demonstrate whether the hip is containable (i.e. the area of disease can be located satisfactorily within the acetabulum) or in non-containable hips may demonstrate the position of best fit prior to valgus or valgus-extension osteotomy.

  - A – all do well without treatment
  - B + bone age < 6 – uniform outcome irrespective of type of treatment
  - B + bone age > 6
    - containment does better than no treatment
    - surgery better than brace
  - C – poor outcome irrespective of treatment

- Age under 6 – 75% do well
- Age 6-8 take x-rays of the wrist to estimate bone age:
  - Bone age < 6:
    - A: normal height → symptomatic treatment with traction
    - B: <50% collapse → symptomatic treatment with traction
    - C: >50% collapse → containment with abduction brace
  - Bone age > 6:
    - A & B → containment with brace or surgery
    - C → probably unaffected regardless of treatment, but some would advocate surgery.

- Age over 9 years → except in very mild cases, surgery is recommended.

- Stulberg (JBJS 1981)
  - Spherical congruity → arthritis does not develop
  - Aspherical congruity → mild to moderate arthritis in mid to late adulthood
  - Aspherical incongruity → severe arthritis before age 50

- Management:
  - Initially while the hip is still “irritable” it should be kept in traction for up to 3 weeks, in a position of slight flexion and external rotation.
  - Thereafter either symptomatic treatment can be given (analgesia ± further traction followed by gentle physiotherapy) or
  - *Early Disease* – head is still plastic, so try to influence femoral head deformation
- Non-weight bearing does not abolish joint reaction force from muscle and gravity → does not work
- **Containment** of the head within the acetabulum to prevent dysplasia or loss of shape of the head during the healing & remodelling phase.
  - Held in wide abduction by plaster or ambulation brace (up to 1 year)
  - Surgical option: varus osteotomy of femur, or pelvic osteotomy.
    - for patients who have restricted abduction when awake due to pain
    - must perform arthrogram and EUA to ensure hip can physically abduct without mechanical block and containable → in which case can perform a varus osteotomy of the femur if child < 8.
    - Lateral shelf osteotomy if child > 8 years.
- **Late Disease** – deformed femoral epiphysis and head is no longer plastic
  - usually develop a short neck due to damage and growth arrest, and have limited abduction while awake, with a weak abductor lever arm
  - perform an arthrogram to reveal hinge abduction, and then check whether adduction offers better congruence – as central and medial columns usually better preserved
  - thus a valgus osteotomy to improve congruence and lengthen leg (if oblique osteotomy).

- The natural history has been described by Waldenstrom’s stages of sclerosis, fragmentation, healing and healed.
  - Ideally containment should begin before or during fragmentation to maintain the sphericity of the femoral head by keeping it within the acetabulum and allow movement to mould the softened head back into a sphere.
  - If caught late in the healing phase, then salvage procedures can be considered such as abduction osteotomies to correct hinge-abduction pain, where the extruded segment of the distorted femoral head impinges on the acetabulum during abduction, or extension osteotomies to correct fixed-flexion deformity.
  - There is a 10-fold increased risk of secondary osteoarthritis, though often only 50% will require arthroplasty after 40 years have passed. Between 20 and 40 years, up to 90% function with minimal pain.

- Herring (JBJS Am, 2004) basically found that most patients in lateral pillar group A do well, and in group C do poorly based on the Stullberg classification of the shape of the healed femoral head, and this is regardless of chronological or bone age at presentation.
  - However, in group B and group B/C patients older than 8 years at presentation, surgery via either pelvic or femoral osteotomy to contain the head, improved Stullberg radiographic outcome. The surgery was followed by 6 weeks in a hip spica or allowed to mobilise NWB (pelvic) and PWB (femoral).
**Slipped Upper (Capital) Femoral Epiphysis**

- Epidemiology – 1 to 2 in 100,000. With 1.5:1 male to female ratio, and bilaterality in 40%. Usually at pubertal growth spurt when physis is widest.

- “Epiphysiolyis” is usually confined to the pubertal growth spurt, again favouring boys and the left side
  - Unilateral slip through the hypertrophic zone of the cartilaginous growth plate increases the risk of a contralateral slip.

- Aetiology – widened physis and weakened perichondrial ring:
  - Hormonal due to an imbalance between pituitary stimulated physical growth – adolescence
  - gonadal hormone mediated physeal maturation and epiphyseal fusion.
    - (Seen in cases of juvenile hypothyroidism and craniopharyngioma)
  - Growth hormone treatment
  - Trauma: 30% acute slip, 70% acute-on-chronic slip.
  - Mechanical factors:
    - Trauma
    - Obesity (weight >80th centile) – more shear force
    - Femoral retroversion

- In a SUFE, the epiphysis slides posteriorly on the femoral neck, which tends to cause the shaft to roll into external rotation. The anterior retinacular vessels are disrupted. Therefore the only blood supply to the femoral head is derived from the posterior vessels which tend to be lifted off the bone with the retinaculum, and hence more prone to damage by surgery or manipulation.

- Physeal disruption usually leads to premature fusion of the epiphysis within 2 years.

- Clinical signs:
  - Groin or knee pain
  - Pubertal, overweight, or particularly tall & thin child
  - Prolonged limp
  - Leg is externally rotated, shortened by 1-2 cm (look at patellae)
  - Limitation in abduction, internal rotation (and flexion)
  - An acute slip is accompanied by pain and limitation of all movements (irritable)

- X-rays:
  - Very early sign may be a reduction in epiphyseal height, as slips posterior
  - Trethowen’s sign on the AP view shows a line along the superior femoral neck remaining above the head, instead of passing through it.
  - In the lateral view, the angle between the centre of the femoral shaft and the femoral epiphysis is 90°. An angle <90° indicates a posterior slip.

- Classification:
  - Pre-slip = pain and no radiology
  - Acute = < 3 weeks
Chronic = >3 weeks  
Stable – weight bearing is possible with or without crutches → AVN 0%  
Unstable – weight bearing not possible → AVN 47% (Loder, JBJS 1993)

- Treatment goals:
  - Preserve the epiphyseal (posterior retinaculum) vessels
  - Stabilise or fix the physis
  - Correct any residual deformity for improved function and prevention of early 2° osteoarthritis.
  - Consensus is to pin in-situ and re-align if required 1 to 2 years later (90% good or excellent results – Bellman 1996)

- Mild = <1/3 width of epiphysis on AP view, and <20° on lateral x-ray.
  - Minimal deformity is accepted
  - Stabilise with threaded pins through femoral neck into the epiphysis

- Moderate = one to two thirds lateral slip, and posterior tilt 20-40°
  - Accept deformity initially and stabilise with short-threaded pins to protect the posterior retinacular vessels, or a less risky method is fusion using a bone graft epiphyseodesis (Heyman & Herndon)
  - If deformity still noticeable at 1 year despite remodelling, then proceed with a corrective sub-trochanteric osteotomy.

- Severe = >2/3 lateral slip (AP) and >40° posterior tilt (lateral film)
  - Marked deformity and risk of early 2° osteoarthritis
  - Open reduction even in specialist centres carries 5-10% risk of AVN
  - Staged fixation and corrective osteotomy is a safe option, with the cuts of the osteotomy aiming to restore valgus, internal rotation and flexion, while realigning the femoral head into the acetabulum.

- Single pin vs. multiple pins – complication rate reduced from 40% to 7%

- Management of unstable slip:
  - Reduction and pinning should either be early <24 hours (7% AVN rate), or later at 1 week. Attempts at between 24-72 hours results in AVN rate of 20%.
  - If decision for delayed treatment: open reduction to relieve haematoma and avoid stress on posterior retinacula vessels. Often anterior approach, and also perform femoral neck shortening osteotomy.

- Complications:
  - 20% risk of contralateral slip (therefore should be radiographically observed) – usually within 12-18 months of index slip.
  - Avascular necrosis – actually rarely occurs in the absence of treatment
  - Articular chondrolysis results in narrowing of the joint space and stiffness; again usually iatrogenic.
  - Coxa vara – apparent as opposed to real, as the actual deformity is femoral retroversion with external rotation & shortening
  - Early secondary osteoarthritis – which needs early THR. During a THR the femoral head may be difficult to dislocate, requiring an in-situ femoral neck cut ± reaming of the head if it cannot be levered out.
AVN Causes

- Idiopathic is most common.
- Congenital – Perthes, Multiple Epiphyseal Dysplasia
- Acquired
  - **Traumatic** (e.g. NOF#, scaphoid #, Neer 3 or above proximal humeral #)
  - **Infection** (septic arthritis or TB)
  - **Neoplastic / paraneoplastic** (during a hypercoaguable state)
  - **Circulatory** – haemoglobinopathies, myeloma, thrombophilia, Caisson’s
  - **Autoimmune** – vasculitis
  - **Pulmonary** – COPD, chronic lung disease
  - **Metabolic** – Gaucher’s disease
  - **Endocrine** – Diabetes
  - **Drugs** – steroids or alcohol
  - **Degenerative**
  - **Iatrogenic** – during manipulation of reduction of SUFE or CDH.
  - **Psychogenic** – nil
Normal Foot Variants

- Infant foot appears flat with mild calcaneovalgus, but retains passive movement at the ankle.
  - Infant does not tend to actively move ankle, as “kicking” achieved by moving hip and knees.
  - Abundant fat on the plantar aspect of foot exaggerates flat appearance.
- Medial longitudinal arch appears gradually between age 1 and 2, as toddler starts walking.
  - To help with balance gait is stamping and broad, with feet externally rotated and pronated – giving an appearance of hindfoot valgus.
  - Only as the gait becomes narrower and smoother between ages 2 and 3, does the medial arch properly develop.
- Gait becomes more adult-like by age 6: propulsive (toe-off) and normal base.
- 97% have flat feet at 18 months, but only 4% persist by 10 years.
- Toe walking can occur in normal individuals and is usually habitual related to tight Achilles tendon. But need to rule out other pathologies:
  - Creatine phosphokinase - Duchenne muscular dystrophy
  - Hip X-ray - DDH (unilateral toe walkers)
  - EMG’s - differentiates normal children from those with cerebral palsy
  - Charcot Marie Tooth
  - Cerebral Palsy
- Treatment of normal paediatric variants is aimed at parents: ERO = explanation, reassurance and offer to follow-up.

Fracture Management

- Paediatric bone has lower Young’s modulus and bending strength – but absorbs more energy
  - Fracture ends tend to be more spiculated → better stability on reduction
Greenstick fractures are a combination of tension and compression – periosteum intact on compressions side.

Spiral fractures tend to be lower energy, because bone fails earlier to shear.
- If managing conservatively, control both rotation and length by flexing joint above and below and incorporating into plaster.

Bone remodelling:
- Wolff's law – bone responds to the force placed on it
- Heuter Volkman – growth plate responds to forces on it
- Angular remodelling factors:
  - Skeletal age 8-10 years
  - Closer to growth plate
  - In the plane of motion
- Fracture overgrowth – related to younger age, larger surface area, and more deformity. Less in upper limb.
  - Average of 1-2 cm in femoral fractures

Growth plate – area of cartilage between metaphysis and epiphysis that allows for longitudinal growth:
- Note that the epiphysis technically represents the secondary ossification centre.
- Blood supply from distal to proximal (i.e. epiphyseal entry)
- Look for Harris Growth arrest lines – if these are parallel to the physis then this is reassuring that growth is occurring and is occurring equally.
- Uneven surfaces (Mamillary processes) and complex shape (W-distal femur, L-proximal tibia) – increase stability on load bearing areas
  - Upper limb growth plates have more simple shapes
- But complex growth plates that are injured (e.g. distal femur) require more force. Thus higher risk of growth plate arrest/injury regardless of timing or type of treatment.
- Treatment:
  - If bony bar or arrest touches the periphery, up to 20% can be resected and normal growth expected
  - If bony bar central, then up to 50% can be resected.
  - Alternatives are growth plate ablation – but leaves any existing deformity and results in shortening → needs subsequent treatment.

Growth plate:
- Resting zone – storage disorders (e.g. Gaucher’s)
- Proliferative zone – longitudinal growth disorders (e.g. achondroplasia)
- Hypertrophic zone – reduced collagen in maturation layer (e.g. SUFE, fracture), and problems with calification (e.g. rickets)

Normal Variants in Children

Growth velocity fastest at birth and decreases by 5 years.
- Between 5-10, growth is slower and steady
- Rapid in adolescence (10-12 in girls, 12-14 in boys)
Close to complete by 16 (girls) and 18 (boys)

- Sitting height
  - Birth 70%
  - 1 year 63%
  - 2 years 60%
  - maturity 52%

- Flat feet can be normal up to the age of 10
  - Is it flexible? Rigid is pathological from congenital vertical talus, tarsal coalition or juvenile chronic arthritis.
  - Insoles or footwear modification do not change natural history
  - But a tight Achilles tendon is worth stretching.
  - Symptomatic (pain) flexible flat feet can be helped with a medial heel counter. A pure arch support will load the arch and be uncomfortable.
  - A flexible flat foot in a child older than 10 – think of generalised laxity (e.g. Marfan’s, Ehlers Danlos).
    - There are surgical options such as a sinus tarsi wedge.

- Bow legs and knock knees:

  ![Graph showing changes in body weight over time]

  - Signs of pathology: asymmetry, short stature, more than 2 standard deviations from normal, or an inter-malleolar/inter-condylar distance over 8cm.
  - Blount’s Disease – early and late onset
    - Early onset difficult to distinguish
    - Metaphyseal / diaphyseal angle > 11° (Drennan)
  - Knock knees common at age 4, but usually resolved by 8.
    - Weight can result in persistence due to abnormal pressure on growth plate.

- In-toeing and Out-towing gait:
  - Most children under 4 will have a –ve (internal) foot progression angle
  - Most adults have a +10° (external) foot progression angle
  - In-toe common and resolves
    - 30% age 2-3, 8% in adolescents
  - Out-toe is less common except in first year of life due to foetal external rotation contracture.
  - Femoral anteverision 50° at birth, and about 15° adults.
- Surgical indicators: significant cosmetic and functional deformity, age > 8 years – also internal rotation > 85°, external rotation < 15°

- Tibial torsion:
  - 4° at birth, 10° at age 1, 12° adults (external)

- miserable mal-alignment:
  - persistant femoral anteversion
  - compensatory external tibial torsion
  - oblique knee axis and squinting patella

- Metatarsus adductus
  - Heel bisector should hit 2nd toe
  - Increased severity as the heel bisector line hit more lateral lesser toes

- Tip toe walking:
  - Exclude neuromuscular problems, CP, muscular dystrophy or spinal cord pathology.
  - Idiopathic – ensure no contractures of TA (which needs serial casting)
    - Serial casting usually result in recurrence,
    - Later if CHILD complains – consider TA lengthening.

- Growing Pain:
  - Red-flags: same location, constant or progressive and at night.
    - Exclude malignancy, inflammatory arthritis, Muscular Dystrophy
  - Usually – zonal and vague, intermittent, long history
    - Usually pre-tibial
    - Scream at night and responds to massage, but the next day all gone.
  - Worth checking CPK to exclude Muscular Dystrophy.

**Paediatric Upper Limb:**

- Radial Longitudinal Deficiency (PRE-axial) – spectrum from complete loss to floating thumb
  - Associations: VACTERL, TAR (thrombocytopenia – absent radius), Holt Oram (heart), Fanconi
  - Defect in Sonic Hedgehog gene.

  - Treatment principles:
    - Correct radial deviation
    - Balance wrist
    - Maintain wrist and finger motion
    - Promote forearm growth
    - Reconstruct thumb deficiency
    - Improve function

  - Algorithm:
    - Passive stretching and splinting as infant
    - Centralisation with tendon transfers
    - Thereafter think about pollicisation
    - Growing rod (ideally by age 2 for developmental reasons)
    - High risk of recurrence – but long-term stabilisation sacrifices wrist mobility and risks stunting growth
- If absent thumb – linked with absent radial nerve

- Ulna Longitudinal Deficiency (POST-axial) – affects elbow more than wrist
  - Associations: lower limb longitudinal deficiencies → examine the feet
  - Priority is to improve elbow function and then position wrist (abnormally) but to allow function.
  - Ulnar sided syndactyly common
    - 1 = hypoplasia
    - 2 = partial aplasia (stable elbow)
    - 3 = complete aplasia (unstable elbow)
    - 4 = synostosis (radius to humerus, stable elbow)

- Madlung’s
  - Growth arrest of volar ulnar side of distal radius – with a normal ulna
    - Abnormal Vicker’s Ligament from ulnar side of radius to lunate results in compression of the physis on this side.
  - MRI to quantify size of bony bar and see if resectable
  - Lengthen radius or shorten ulna / epiphysiodesis

- Congenital Ulna Synostosis:
  - Even if surgically released, the muscle of biceps is weak – usually have compensatory movements at shoulder to allow function.
  - Bilaterality is a surgical indication
  - 33% associated with Apert’s¹ and Klinefelter’s syndromes

- Cleft hand:
  - Typical (V-shape)
    - bilateral, autosomal-linked dominant
    - associated with cleft lip/palate
    - central longitudinal deficiency
    - functional triumph and social disaster
    - lack of microvascular bed prevent ability to perform toe transfer
  - Atypical (U-shape)
    - Form of transverse arrest and symbrachydactyly
    - Spectrum from small fingers (brachydactyly) and connected fingers (syndactyly) → complete absence of central three fingers
    - Unilateral, sporadic and no foot involvement
    - Can be associated with Poland’s syndrome (congenital absence of sternal head of pectoralis major – with unequal nipples)

- Polydactyly:
  - Pre-axial (radial), central or post-axial (ulna)
  - Wassell’s classification
    - Based on split or duplication of phalanges
    - Type 7 associated with heart and lung complications

¹ Autosomal dominant mutation of FGFR-2 resulting in abnormal fusion / synostosis of cranial sutures. Main issues are with facial / dental abnormalities and may impact on intellect. Also results in fused digits.
Thumb Hypoplasia – a type of radial longitudinal deficiency (see above)
- 1 in 100,000, right more common (60% bilateral)
- 80% association with Holt Oram, Fanconi, VACTERL, thrombocytopenia absent radius (TAR)
- Blauth classification
  - 1 = minor hypoplasia → leave
  - 2 = bones present, MCP-UCL instability, thenar hypoplasia → reconstruct
  - 3 = musculotendinous and osseous deficiencies → 3A (CMCJ intact) reconstruct; 3B (absent CMCJ) amputate & pollicisation
  - 4 = floating thumb → amputate & pollicisation
  - 5 = complete absence → pollicisation

Camptodactyly – little finger sagittal plane deformity (<1% population)
- Painless flexion contracture at PIP
- Two-thirds bilateral
- Pathology with lumbrical not inserting onto extensor hood
- Treatment only if >30° and impaired function
- Volar release and augment extension with tendon transfer.

Clinodactyly – autosomal dominant, often bilateral.
- Coronal plane deformity of finger (often little)

Congenital Trigger Thumb
- Hypertrophic flexor tendon (Notta Node)

Trigger Digit
- Could be abnormality of A1 pulley, or an abnormality of FDS where it traps FDP that passes through the area it splits.

Syndactyly
- Simple – soft tissue abnormality
- Complex – bone abnormality
- Complicated – Poland’s (absent pec major sternal head; difference in nipple level) and Apert’s
- Border digits of unequal lengths and therefore should have early release within few months age – allows thumb function and prevents tethering / flexion contracture of longer digit
Central digits have similar lengths and can be released later (12-18 months)
- If all fused – staged release start with 1st and 5th ray

- **Congenital Clavicle Pseudoarthrosis**
  - Infants – excise
  - Teens – fix
  - Adults – fix and graft

- **Sprengel Deformity**
  - High scapula C5-T1 instead of T2-T7
  - Persistence of omovertebral bar
  - Associated with:
    - Klippel Fiel - short neck, low hairline, and stiff neck.
    - Fused ribs
    - Cervical rib with omovertebral connection blocking normal caudal migration
    - Congential scoliosis

- **FSHD – facial-scapula-humeral dystrophy**
  - AD
  - Selective muscular defects
  - Winging

- **CP in hand**
  - Weakness and spasticity → imbalance around joint → correctible contracture → fixed contracture → secondary skeletal deformity.

- **OBPI**
  - Traction that affects proximal roots first
  - Narrakas 1 – C5, C6 → shoulder problem
  - Narrakas 2 – C5, C6, C7 → wrist drop
  - If no biceps function by 3 months = indication for surgery.

- **Arthrogryposis:**

- **Congenital pseudoarthrosis of clavicle:**

- **Congenital dislocation of radial head:**

- **Delta phalanx:**

- **Congenital clasp thumb:**
**Paediatric Knee**

- Knee responsible for 2/3 of growth in leg – 15mm per year
  - Distal femur 9 mm/yr, Proximal tibia 6mm / year

- Congenital dislocation of knee
  - Spectrum from hyper-extension to anterior dislocation
  - 1 in 100,000
  - aetiology: oligohydramnios, extended breach, quads contracture
  - associations: DDH, club foot, myelodysplasia, arthrogryposis
  - pathology
    - fixed hyper-extension
    - absent supra-patellar pouch
    - hamstrings elongated and change from flexors to extensors

- Treatments:
  - Conservative closed reduction and serial casting followed by Pavlik’s harness
  - V-Y plasty of quadriceps – before walking age (6-12 months)

- Congenital Patella dislocation (flexed knee)
  - Failure of myotome containing the quadriceps from internally rotating during 1st trimester ventrally.
  - Extensor mechanism inserted anterolaterally, resulting in contracture of ITB, lateral capsule and vastus lateralis.
  - Loose medial capsule and VMO
  - Patella pulled laterally.
  - Treatment – before age 1 (before walking age)
    - Extensive lateral release of whole Vastus Lateralis and knee capsule
    - Reef medial structures onto lateral structures once pulled medially
    - ± Roux Goldthwaith (patella tendon re-alignment, not bony correction)

- Discoid Meniscus – almost always lateral meniscus, with 20% bilaterality
  - Watanabe:
    - 1 = complete
    - 2 = incomplete
    - 3 = Wrisberg (loss of ligamentous stabilisers) ➔ painless snapping knee
  - X-ray shows widening of lateral joint space, and flattening of surface of tibial condyle
  - Treatment –
    - Observe if asymptomatic from pain
    - But warn more at risk of tears
    - Preserve as much meniscus with saucerisation ± peripheral rim stabilisation procedure

- Popliteal Cysts:
Medial based between medial gastrocnemius and semimembranosus
Asymptomatic – often quite hard, but will still transilluminate
Does NOT involve joint
50% recurrence with surgery
resolve by 2.5 years.
Ultrasound to confirm no joint involvement.

Infantile Blount’s Disease:
Continuation of physiological bowing – usually with obesity, family history and early walking hx. Often bilateral
Excessive pressure on posteromedial physis will disrupt normal growth leading to varus
<9° metaphyseal-diaphyseal angle and 95% will resolve
>16°, then 95% will not resolve
Initially starts with bowing, but as progresses there is bar formation and then depression.
Treatments:
- Brace treatment if younger than 2.5 years with early stage (Langenskiold 1-2)
- Guided Growth
- Surgery once over 3 – Langenskiold 5-6 or failed bracing
  - Osteotomies
  - Bar Resections
  - Medial plateau elevation if significantly depressed
  - Aim to over correct by 10-15° to avoid recurrence

Adolescent Blount’s Disease:
Morbid obesity / fat thigh syndrome. Often unilateral.
Femur is also involved
There is tri-planar deformity with internal rotation and pro-curvatum as well as varus.
Options: guided growth less effective → osteotomies, circular frames

Knee Apophysitis: (an apophysis is a growth plate in tension)
Tibial tuberosity – Osgood Schlatters
- Usually self limiting with waxing-waning symptoms for up to 2 years
- Male = female
- Activity or over use.
- Look for abnormal forces through the patella (e.g. pes planus)
- Always check the hips
- Rest and activity modification; NSAIDs; strapping has little evidence
- If failed conservative rx – can consider drilling to fuse the apophysis

Inferior Patella – Sinding-Larsen-Johansson
- 10-13 year old males
- palpable gap at inferior patella + extensor lag → sleeve fracture?
- Self-limiting but shorter duration than OSG (3-18 months)
- NSAIDs, RICE → physio (strengthen quads & keep knee supple)

Osteochondritis Dissecans
Localised lesion of subchondral bone of an articular surface, without breach of articular cartilage – underlying AVN
Best seen on tunnel view AP of knee. MRI to assess stability
Pain ± mechanical symptoms
Classical lateral surface of medial femoral condyle (normal variant has similar appearance on medial condyle)
Stable injuries – conservative (if physis is open, most will heal with activity modification)
Unstable –
- drill base and fix (K-wire)
- reduce and fix with headless screw
- complex options (e.g. mosaicplasty) for complete loose bodies that are unsalvageable

ACL injury:
- If > 3 years of growth left, try conservative, and then combined intra-and extra articular reconstruction sparing physis
- If > 2 years of growth – partial sparing fixation (tibia has less growth than femur)
- If > 1 year – trans-physeal fixation (adult)

Idiopathic anterior knee pain – “chondromalaciae patellae”
- Adolescent girls – sedentary overweight, or very sporty
- Poorly localised anterior pain
- No significant examination findings
- Normal x-ray, but tend not to MRI
- Diagnosis of exclusion
- Natural history – self-limiting, with physiotherapy and NSAIDs

Non-accidental Orthopaedic Injury

- NAI involves acts of commission or omission which directly or indirectly cause harm
  - 7% children experience serious physical abuse by parents/carers, and 6% experience neglect.
  - Majority in children under 2 years
    - 80% of NAI victims may be under 18 months
    - whereas 85% of accidental injury may be in children over 5 years
  - Infants younger than 4 months with fractures are more likely due to abuse
  - NAI more common in children who have not yet walked
  - 55% of fatally abused children have been seen by a healthcare professional within previous 1 month.

- Suspicious clinical history:
  - Vague, inconsistent, inexplicable
Un-witnessed mechanism
Delay in seeking medical advice
Evasive, aggressive or unusual parental responses

Risk factors:
Lower socioeconomic groups
Unplanned pregnancy

Metabolic disorders that need to be excluded:
Copper deficiency (serum caeruloplasmin, magnesium levels)
Rickets (↓calcium & phosphate levels, ↑parathormone, ↓fasting Vit-D)
Osteopaenia of prematurity – higher risks if co-existent:
- cholestatic jaundice
- chronic lung disease...
- >2 weeks diuretic treatment
- >3 weeks parenteral nutrition
Chronic illness / failure to thrive
Osteogenesis imperfecta – genetic testing
Caffey’s disease – infantile cortical hyperostosis (first 3 months, multifocal swelling, bone lesions and irritability – self-limiting)
Osteomyelitis
Malignancy and neurofibromatosis 1
Physiological periosteal reaction – symmetrical, infant and confined to diaphysis only.

Bruising:
Away from bony prominences, larger/clustered bruises
Shapes of implements
Lack of bruising is characteristic of pathological fracture
but acute accidental fractures may have minimal bruising

Most common accidental fractures in children are the distal radius and ulna.

Multiple rib fractures without a clear explanation have the highest specificity for NAI
Note post-resuscitation rib fracture incidence is < 2%
Rib fractures not due to NAI:
- Prematurity – but on review of films at Women’s Hospital (Liverpool), actually rare.
- Osteogenesis imperfecta
- Birth injury
- Physiotherapy
- CPR
No clear relationship between fracture configuration in the femur and NAI; in fact mid-shaft femur fractures are common in both accidental and non-accidental injury.

Although diaphyseal fractures 4 times more common than metaphyseal, it is the metaphyseal fractures that are highly specific for NAI in under 2’s.

Other suspicious pattern: disproportionate callus formation and fracture through the callus.

<table>
<thead>
<tr>
<th>HIGH Specificity Fractures</th>
<th>FREQUENT fractures but LOW Specificity</th>
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<tbody>
<tr>
<td>Metaphyseal</td>
<td>Mid-shaft clavicle</td>
</tr>
<tr>
<td>Multiple Ribs</td>
<td>Simple linear skull fractures</td>
</tr>
<tr>
<td>any Scapula fracture</td>
<td>Single long bone fracture</td>
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<tr>
<td><em>Lateral</em> Clavicle (non ambulant)</td>
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<td>Fractures of different ages</td>
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<td>Vertebral</td>
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<td>Digital injuries in non-mobile children</td>
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<tr>
<td>Bilateral fractures</td>
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<tr>
<td>Complex skull fractures</td>
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</tbody>
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Full skeletal surveys recommended in children under 2 years to diagnose occult injury, and to be reported by a paediatric musculoskeletal radiologist, with repeat survey at 2 weeks.

(up to 20 exposures)

- **Skull**
  - AP, lateral ± Towne’s view
  - even if CT already performed
- **Chest**
  - AP including clavicles
  - Oblique views (left and right) to assess for rib fractures
- **Spine**
  - AP & lateral chest and abdomen
  - Lateral only of C-spine
- **Limbs**
  - AP humerus
  - AP radius & ulna
  - AP femurs
  - AP tibia & fibula
  - PA hands
  - Dorso-plantar views of feet

Increasing evidence for use of CT-scanning if x-rays normal with high clinical suspicion, and for CT brain to investigate NAI in pre-ambulant children (because of high incidence of subdural haemorrhages)
Mean Times for long bones:

- Soft tissue swelling: 7 days
- Periosteal reaction: 10 days
- Early callus (density < cortex): 14 days
- Marginal sclerosis: 8 weeks
- Hard callus (density > cortex): 10 weeks
- Remodelling: 12 months
- Altered pattern of healing for repetitive injury, lack of immobilisation, concurrent head injuries and nutrition.

Sub-periosteal new bone is a normal finding in children up to the age of 6 months. Usually single layer and not extending below the metaphysis-diaphysis junction. Consider NAI if thickened or multi-layered.

**Limb Length Discrepancy**

**Signs to compensate:**
- Gait asymmetry – toe walking i/l, and knee flexion c/l
- Pelvis not level
- C/L OA hip due to point loading from longer leg being point loaded due to uncoverage as placed in adduction
- acquired scoliosis

**Causes:**
- Congenital – longitudinal deficiency, hemi-hypertrophy, PFFD
  - Femur – coxa vara, PFFD, lateral condyle hypoplasia
  - Tibia/Fibula – shorter, external tibial torsion, **fibular hemimelia**, cruciate deficiency
  - Feet – missing rays, ball and socket joint, tarsal coalition
- Acquired – Trauma, Infection
  - meningococcal infection & physeal infarction
- Neuromuscular – CP and Polio
  - Gait lab analysis to assess if equalisation will compromise compensatory mechanisms to inherent weakness
- Apparent – DDH, joint contractures

**Distinguish hemihypertrophy from hemi-atrophy**
- Look for cutaneous manifestations
- Compare to normal anthropometric data –need to match race
- Look for syndromic conditions:
  - Beckwith Weidemann – macroglossia, hyperglycaemia, 5% Wilm’s tumour, port wine stain (11p15 mutation over-expresses IGF-2)
  - Klippel Trenaunay Weber – vascular
  - Neurofibromatosis
- Non-syndromic:
  - Look for skeletal dysplasias: HME, OI, Olliers, Rickets
  - Could have Wilm’s tumour – so need regular USS abdomen until age 7
Assessment:
- History
- Examination
  - General – gait, abdominal exam, cutaneous signs
  - Specific – site, size, joint status
  - Contractures usually result in apparent LLD
- Difference between bone age and chronological age tends to be constant over time
- Boys stop main growth at 16, and girls at 14.
- Estimate discrepancy at maturity
  - Menelaus rule of thumb (15 mm/yr – 9mm femur, 6 mm tibia) only useful for complete growth arrests
  - Eastwood & Cole – clinical assessment of discrepancy and fit to best fit Shapiro curve
  - Paley’s Multiplier – also used for Shapiro Type 1 inhibition (linear)
- Useful to know predicted height of child – may influence whether you choose to lengthen or perform a contra-lateral epiphysiodesis

Management
- LLD < 2 cm → Observe, Shoe raise, but beware compliance issues
- LLD 2-5 cm
  - With deformity – correction and lengthen
  - Average height – shorten long leg
    - Problems with reducing muscle power, non-union, need implant removal?
    - Maximum 5 cm in femur and 3 cm in tibia
  - Below average height – lengthen short leg, unless short leg has been affected by infection, vascular/nerve issues, radiation scarring
- LLD > 5 cm → lengthening procedure
  - Ilizarov method – 1mm/day in aliquots, assess regenerate
  - Hybrid frames
  - Mono-lateral external fixators
  - Intramedullary Skeletal Kinetic Device (ISKD)
    - approx 35-40 days /cm in normal children
    - 25-30 days / cm in achondroplastics (membranous growth)
    - 40-60 days / cm in adults

Obstacles / complications:
- Pin site infections nearly 100%
- Wound problems
- Lysis and loosening of pins (usually due to pin-site infections)
- Early regenerate consolidation
- Soft tissues not yielding
- Non-compliance with physiotherapy and joint contractures
- Fracture
- Rare pseudo-aneurysm or compartment syndrome
- Psychosocial issues for children AND parents.
Cerebral Palsy

- Group of non-progressive disorders in which disease on the brain causes an impairment of motor function
  - Static cerebral pathology with progressive musculoskeletal sequelae
  - 0.25% of all live births

- Causes:
  - Pre-natal – intrauterine infection, congenital malformations
  - Peri-natal – birth trauma / asphyxia, kernicterus
  - Post-natal (up to age 2) – infections (meningitis), NAI, haemorrhage

- Classification:
  - Topographical – unilateral, bilateral or total body involvement (includes trunk and head control)
  - Neurological – spastic, athetoid, dystonic, rigid, mixed
    - Spasticity = pyramidal system injury with predictable increased tone and contracture
    - Dystonia = extra-pyramidal system injury with alternating tone and rare contracture
  - Functional – ambulant, non-ambulant
    - Ability to sit independently by age 2 → ambulant
    - Bleck’s test (5 neuro, 2 postural reflexes) if 2+ by age 1 then walking unlikely
    - Type of CP
      - Walkers – community, domestic, therapeutic (with physio sessions)
      - Standers – independent, transfers
      - Sitters – with trunk / head control or not
Problems:

- **Primary – injury to CNS**
  - Loss of normal motor development
  - Maintenance of primitive reflexes
  - Loss of selective motor control
  - Weakness $\leftrightarrow$ treatable with stamina training
  - Problems with balance
  - Abnormal tone – spasticity (pyramidal injury), athetosis (extra pyramidal injury)
    - Spasticity treatable with Botulinum toxin
    - but beware permanent effect on motor end plates, but with age and reduced end-plate re-cycling there is permanent muscle weakness.
    - Selective Dorsal Rhizotomy – breaks stretch reflex and involves neurosurgery. Ideal for spastic diplegia with good antigravity strength and adequate cognition/motivation to comply with rehab
    - Intrathecal Baclofen – GABA inhibition but does not cross blood-brain barrier and so needs to be placed intra-thecal.

- **Secondary – from growth**
  - Muscle contractures
  - Over-long muscles
  - Bony deformity
  - Lever arm dysfunction – short, flexible, mal-rotated, abnormal fulcrum, positional.
    - if potential to walk consider single-event multi-level surgery
The Hip in Cerebral Palsy

- Hip dislocations and subluxations untreated lead to pain, scoliosis and problems with hygiene/cleaning
  - Very problematic as 80% of modern CP patients are living till age 40
  - Higher rates in non-walkers (higher GMFCS)
  - Why – failure to remodel birth anatomy as not walking, combined with weakness/spasticity
  - Children who walk independently by 30 months, rarely develop hip instability
  - If total body involvement – >50% affected by age 5
- Monitor with x-rays every 2 years and clinical review annually
- Older children with an established and painless hip dislocation can be left alone
- Not affected by botulinum or SDR. Bracing will temporarily halt progression if applied for >16 hours per day.

Surgical options:
- Soft tissue – adductor tenotomise (50% useful for GMFCS 3 only), if Reimer uncoverage < 40%
- Femoral osteotomy ± hip reconstruction
- Salvage – Schanz abduction osteotomy (creates a straight neck-shaft)

Spinal adjunct:
- Take patient out of chair and sit on couch.
- Assess if both buttocks on couch (Group 1)– otherwise functional pelvic obliquity (Group 2)
- Group 1 – treat hip first and then observe spine and treat as needed
- Group 2 – treat spine first and then hip (if can be positioned for spinal surgery, otherwise hip first)

Talipes Equinovarus

- Heel in equinus and hindfoot in varus, with forefoot adducted and supinated
- 1-2 per 1000 births; male twice as common, and 33% bilateral
- Idiopathic - 10% sibling risk (Wynne-Davies 1973)
  - Multifactorial, polygenic inheritance
- Cause unknown:
  - Postural deformity from overcrowding in uterus
  - Neural tube defects (meningomyelocele)
  - Genetic halted development
  - Maternal smoking, drug abuse, ?alcohol
- Syndromic:
  - Spina bifida
  - Arthrogryposis (congenital contractures in 2+ areas)
- Sacral agenesis
- Foetal alcohol syndrome
- Congenital myopathy
- Down’s syndrome (may include vertical talus)

- Clinical features:
  - Obvious at birth with foot twisted inwards and soles facing each other
  - Calf may be abnormally thin
  - Normal babies can dorsi-flex ankle until toe touches shin – reduced ROM in affected neonates; or can be fixed deformity
  - Always examine to exclude CDH or spina bifida

- Pathoanatomy is a short calcaneo-fibular ligament which rotates calcaneus.

- Pirani Score (max 6)
  - Midfoot – lateral border, medial crease, TNJ reduction
  - Hindfoot – posterior crease, fixed equinus, empty fat pad sign.

- X-ray:
  - 30 degree AP film
  - Kite’s angle is angle between medial border of talus and lateral border of calcaneus (normal is 20-40, reduced in talipes)
  - On lateral film, the talo-calcaneal angle (between long axis of talus and lower border of calcaneus) should be 40 degrees in any position. <20 degrees shows the calcaneus cannot be tilted up into true dorsi-flexion

- Conservative rx:
  - Within 48 hours of birth, manipulate and strap into correct position
  - Teach parents to repeat stretching while strapping in place
  - Monitor (with physio) as strapping can be altered as correction obtained, or use weekly POP casts to hold.
  - Manipulation should over-correct (CAVE)
    - Increase supination deformity to bring forefoot into alignment with more supinated hindfoot
    - Reduce both hindfoot and forefoot out of varus and supination
    - Dorsi-flex the foot
    - Start by correcting cavus by lifting metatarsals and increasing supination. Then correct adduction by ab-ducting the foot. Continue abduction until extreme hyper-abduction which unlocks the calcaneus and corrects hindfoot varus. Finally address equinus by Achilles tendon tenotomy (1 to 1.5 cm above insertion percutaneously).
  - Percutaneous Achilles tendon lengthening may be required – heals by gap healing in the young with pleuripotent cells forming new tendon as opposed to scar tissue.

- Casting protocol:
  - 3-5 weeks in cast
  - 3 months in boots and bars full time
  - 3 -4 years night time bracing.
Surgery is usually deferred for 6 months or until child is walking, as this allows for bigger bones to aid surgery, and walking itself helps maintain correction:

- Posterior incisions (Turco, Cincinatti-Crawford, or Caroll)
- Achilles tendon and Tibialis posterior lengthened with z-divisions
- Division of posterior capsules of ankle and subtalar joints
- Lengthening of FDL and FHL sometimes needed
- Release of calcaneo-fibular ligament with complete subtalar release
- Superficial part of deltoïd ligament released, but deep part preserved to maintain ankle stability
- If a K-wire is used to maintain reduction, it is removed at 6-8 weeks

Recurrent cases that fail to respond to soft tissue release, can be treated with a protracted regime using the Ilizarov frame.

- Alternative include: repeat casting for 6-8 weeks to obtain abduction, repeat tenotomy, or tibialis anterior transfer to lateral cuneiform.

**Ponseti Methods for CTEV**

**Under 2:**
- Gentle manipulation
- Serial casting
- Achilles tendon tenotomy
- Regime of splinting in boots & bars

**Over 2 years:**
- Serial casting
- Anterior tibial tendon transfer

**Anatomy of club foot and order of treatment:**
- C – cavus
- A – adductus midfoot
- V – varus of hindfoot
- E – equinus of hindfoot

**The acetabulum pedis – navicular, subtalar surfaces + inter-osseous ligament**

**Manipulation:**
- Foot moves around head of talus
- There use head of talus as fulcrum for all manipulation
- Correct cavus by supinating the pronated-forefoot to bring in line with supinated-hindfoot
- Correct adductus by moving forefoot and midfoot as one into Abduction, which also moves the hindfoot from varus to valgus
- Then can correct equinus once calcaneus has swung into valgus, otherwise the attempt will result in movement at the midfoot. 90% need Achilles tenotomy – achieves 20º dorsiflexion.
- Plaster used above knee to avoid falling off, and then release dorsum of toes. Usually 5 weekly plasters.
- Final plaster usually has 70° abduction and 15° dorsiflexion

- Boots and Bars (e.g. Piedro boot)
  - Full time for 3 months
  - Nights and naps for at least 4 years, with normal footwear during the day without splints

- Recurrences defined as supination of foot during swing phase:
  - treated by repeat casting and return to boots up to age 2
  - after age 2
    - treated with anterior tibial transfer
    - above knee cast for 5 weeks
    - no need for boots & bars

- [www.ponseti.org.uk](http://www.ponseti.org.uk)

**Flat foot in Children**

- Tip-toe test confirms a mobile subtalar joint and tibialis posterior function
  - Tibialis posterior will plantar flex the ankle and invert at the subtalar joint. Isolate by placing foot in plantar-flexed, everted position. Ask patient to invert against resistance in this position (to stop tibialis anterior masking a weakness)

- 90% of under-2s will have flexible pes planus, which gradually corrects between ages 3-5. This is due to natural joint hyper-mobility, and the broad-based gait with the mechanical axis falling slightly medial to the 1st or 2nd ray. By age 10, only 4% will have persistent flat foot.

- Assessment should include:
  - Is there a tight TA – seen if the heel is not flat during stance phase
  - Gastrocnemius tightness – assess TA in flexion and extension; tiptoe gait?
  - Family history
  - Signs of hyperlaxity

- Painful flatfoot in child:
  - Congenital vertical talus
  - Tarsal coalition
  - Tumour
  - Foreign body
  - Accessory navicular

- Normal lateral x-ray of foot will show a straight line through the long axis of the talus, navicular and 1st MT. Sagging at the talo-navicular joint indicates a flat foot:
  - <15° = mild
  - 15 to 40° = moderate (associated with lateral subluxation of the navicular
  - >40° = severe off the head of the talus)
Calcaneovalgus foot:
- 30-50% neonates
- Usually result of intra-uterine positioning
- Normal anatomy AND flexible

Flexible flat foot
- Normal in toddlers and usually resolves within a few years when medial arch development is complete
- Arch restored by dorsi-flexing the hallux (Jack test) perhaps by putting the FHL tendon onto a stretch which supports the medial longitudinal arch?
- Usually re-assure and no treatment needed – insoles reduce shoe-wear by changing pattern of weight bearing and do not help the feet
- If painful: plantar medial, sinus tarsi or from callosities
- Treatment:
  - TA stretching – not effective
  - Pure soft tissue releases – not effective
  - Orthotics make no difference to shape of foot (Wenger)
  - Sinus Tarsi prosthesis (arthroreisis) – blocks hindfoot valgus
  - Lateral column lengthening (using wedge shape graft to also rotate foot internally) ± medial plication and Tibialis posterior advancement (equivalent to Windlass mechanism)
  - Subtalar arthrodesis or triple arthrodesis for salvage.

Stiff flat foot – consider congenital vertical talus, an inflammatory joint disorder, tarsal coalition or a neurological disorder

Congenital Vertical Talus
- Fixed dorsal dislocation of talar head and neck
- Causes:
  - Neurological – Arthrogryposis, SMA, Neurofibromatosis, Sacral Agenesis, Myelomeningocele
  - Idiopathic
  - Genetic
- Usually bilateral with medial arch not simply flat, but most prominent part of sole resulting in a rocker-bottom appearance
- Hindfoot in valgus with talus pointing vertically – contracted TA and peronei
- Forefoot then abducts, pronates and dorsiflexes – contracted extensors
- Usually a fixed deformity
- On x-ray the talus is vertical with the navicular dislocated dorsally.
  - Always repeat lateral film with foot plantarflexed
  - In flexible flat foot, the navicular returns to normal position (often not ossified so look at 1st MT)
- Untreated – increasingly rigid deformity with painful pushoff and callosities under talar head.
- Treatment = surgery < 2 year old
  - Traditional
• Achilles tendon lengthened + capsulotomies of ankle and subtalar joints
• Reduction of talonavicular joint with transfer of tibialis anterior tendon to neck of talus
• Lateral structures may need to be lengthened or released
• K-wire transfixes talonavicular joint (removed at 6 weeks)
• 8-12 weeks POP
  ▪ Dobbs Technique = reverse Ponseti
    • Weekly casts (approx. 5)
    • Mini-open reduction of talonavicular joint and fix
    • Now correct equinus.

❖ Skew Foot = hindfoot valgus with midfoot varus & metatarsus adductus
  ➢ X-ray
    ▪ 1st ray translated in respect to axis of talus
    ▪ Increased talonavicular coverage angle
  ➢ Treatment:
    ▪ Closing wedge of cuboid and opening wedge of medial cuneiform = treatment of metatarsus adductus
    ▪ Mosca = calcaneal lengthening + opening wedge medial cuneiform + TA lengthening

❖ Accessory Navicular:
  ➢ Tenderness over medial prominence – with x-ray showing an extra ossicle
  ➢ Symptoms relate to a bursitis and pressure over the area
  ➢ Bone scanning is used to exclude infection or osteoid osteoma, but can also identify a “hot” accessory navicular.
  ➢ Excision by shelling out ossicle from Tibialis Post tendon, and then hitching it to the navicular, by looping suture around it and then passing it through 2 drill holes in the navicular, tying it on the dorsal surface of the navicular → this hitches up the medial longitudinal arch.

❖ Kohler’s Disease = AVN of navicular.

❖ Tarsal Coalition:
  ➢ Fibrous, cartilaginous or bony pathological connection between 2+ tarsal bones
  ➢ Can result in a variety of partial or complete unions that represent a failure of segmentation of the tarsal bones: talo-calcaneal, calcaneo-navicular and talo-navicular.
  ➢ Incidence 1%; bilateral in 50% of those affected.
  ➢ Association:
    ▪ Fibular hemimelia
    ▪ Club foot
    ▪ Apert syndrome (early closure of cranial sutures, reduced IQ, syndactyly, autosomal dominance)
Painful stiff flat foot accompanied by spasm of extensor tendons and peroneal tendons, as patients tries to compensate and correct overall hindfoot alignment (*peroneal spastic flatfoot*)

Autosomal dominant condition, with an initial fibrous syndesmosis maturing into cartilage and later into a bony bar.
- Usually happen in puberty or early adolescence, as the cartilaginous interface matures into a rigid bony structure
- Symptoms may only start with trauma that fractures bar, or from abnormal tarsal stress

Treatment:
- Initial 6 weeks conservative mx with plantigrade walking POP cast, as that the condition may be an incidental finding and not the root cause of symptoms – 30% persistent relief.
- Calcaneo-navicular bars can be excised with a piece of muscle (EDB) interposed to prevent recurrence.
- When not to surgically resect:
  - >50% involvement of posterior facet by a sub-talar bar
  - >21° heel valgus
  - pre-existing OA
- Talo-calcaneal bars are more difficult, and it may be wiser to wait until after puberty to perform a triple arthrodesis. If resecting, a fat graft is preferred for interposition.

### Pes Cavus

**Aetiology:**
- Muscle: muscle dystrophies
- Peripheral nerve: hereditary neuropathy
- Spinal cord: viral disease, structural
- Brain: congenital
- Post-traumatic: compartment syndrome

**Clinical features**
- Age 8-10
- Usually bilateral with high arches and toes drawn into clawed position
- Metatarsal heads forced down ± callosities on sole
- Giving way of ankle due to varus of hindfoot.

**Tripod analogy:**
- Normal = relation between calcaneus, 1st and 5th metatarsal
- Plantaris = then both rays are drawn towards heel
- Cavovarus = 1st ray alone pulled towards heel + varus
- Calcaneus = heel is pushed plantarwards
- Calcaneo-varus = heel pushed plantarwards, and 1st ray pulled in to heel

Initially flexible deformity with hyperextension of MTPJ, subluxation of MT-heads and clawing of toes → become fixed
- Pain on lateral aspect of foot, over 5th MT head, and even ankle instability with severe hindfoot varus.

- **Aetiology = muscle imbalance**
  - E.g. in HSMN there is overactivity of Peroneus Longus compared with Tibialis Anterior resulting in cavus, and overactivity of Tibialis posterior compared with Peroneus Brevis resulting in varus
  - Association of bilateral pes cavus with Charcot Marie Tooth is 78% in the absence of family history, and 91% if family history present.

- **Symptoms & signs:**
  - Pain
  - Callosities / Corns
  - Walking tolerance
  - Giving way
  - Footwear problems
  - Coleman block test – will reverse hindfoot varus that is driven by the forefoot 1st ray
  - Always check spine

- **X-rays**
  - Meary’s angle normal = 0 degrees (angle between axis of talus and 1st MT)
  - Calcaneal pitch normally 10 to 30 degrees from horizontal
  - Both increased in pes cavus

- **Other Ix:**
  - Blood CPK to exclude dystrophy
  - MRI and EMG to look for neurological cause
  - Genetic testing or muscle biopsy

- **Treatment:**
  - Look for correctible higher causes – once treated first there is more confidence that any residual deformity will be static and unlikely to recur.
  - Non-operative options such as custom made shoes, and moulded inserts treat sx, but do not alter disease progression. This will also act as a shock absorber as the rigid foot can no longer do this alone.
  - Flexible deformity can be treated with a lateral heel counter, or a medial bar and lateral T-strap to pull the heel out of varus.
  - Surgical aim is to provide a pain-free, plantigrade, supple but stable foot:
    - Timing of surgery is when orthotics have failed, but foot remains flexible.
    - Soft tissue release
      - Lengthening of Achilles tendon, and posterior capsulotomy
      - Release of plantar fascia for cavo-carus foot (in children only)
    - Osteotomies (for fixed deformity and to avoid fusion)
      - Lateral closing-wedge calcaneal osteotomy to correct varus (Dwyer)
      - Correct cavus with 1st MT dorsiflexion osteotomy
    - Tendon transfers if joints are mobile
      - Tibialis anterior transfer from base of 1st MT to calcaneum
- EHL from anterior surface of fibula to hallux distal phalanx, used as a sling through the neck of 1st MT, to elevate it
- Peroneus longus runs from superior 1/3 of fibula to 1st MT-base and medial cuneiform – splitting it and attaching it to peroneus brevis will reduce pull on 1st MT and improves eversion
- *Principles of tendon transfer*
  - Loss of power by one grade
  - *Straight line of pull*
  - Tendon to bone preferable
  - *Not in presence of fixed deformity*
  - Expendable muscle
  - *Ensure balance achieved to avoid recurrence*

- Correction of claw toes by Robert Jones tendon transfer
  - Fusion of IP joint with transfer of EHL from distal phalanx to neck of 1st MT- acts as a sling to pull up head of 1st MT. Must be combined with release of Peroneus Longus onto Peroneus Brevis (removes deforming plantar flexion force on 1st ray, and augments eversion to offset any varus.

- Late options for fixed deformity = triple fusion (never for asymptomatic)

**Neuromuscular Disorders**

- Intrinsic Muscle disease:
  - Myopathy – structural abnormality
  - Dystrophy - Protein deficiency

- Any toe walker (especially boys) – should have a CPK test.

- Muscle weakness drives deformity. Interacting factors:
  - Changing pattern of relative weakness
  - Speed of progression
  - Growth relative to weakness

  - Thus changing sites and evolution in pathology, and prone to recurrences.

- Pathologies:
  - Scoliosis
  - Contractures
  - Hip dysplasia
  - Knee – contractures and lengthening of extensor mechanism
  - Foot Deformity – cavus foot, ankle instability
  - Sensory defects – pressure sores
  - Pain – muscle, deformity, osteoporosis, or spurious post-operative pains

- Principles:
  - Diagnosis
  - Define functional objective
  - Management plan
Assessment:
- Gait pre-requisites – foot pre-position, foot clearance, stability in stance and adequate step length
- Spine
- LLD and pelvic obliquity
- Sensation

Supracondylar Fracture:
- Gartland with Wilkins modification
  - 1A undisplaced
  - 1B collapse / comminution of medial column
  - 2A hinged → can be left alone as will remodel since in plane of joint movement
  - 2B hinged + rotation → leads to varus/valgus
  - 3A complete + posteromedial – reduce in pronation
  - 3B complete + posterolateral – reduce in supination

Timing
- Ideally within 24 hours if pulse maintained
- If pulseless → notify vascular surgeon and emergency closed reduction
- If fails and cool → open reduction (lazy S approach)
- If remains pink & pulseless
  - “rubbery” reduction or median nerve symptoms → more likely to have entrapment → anterior open reduction
  - if perfect reduction and no nerve symptoms → careful exploration (Mangat JBJS)
- BSCOS
  - Gartland 3 → MUA, K-wire, POP
  - Crossed K-wire
  - Medial mini-open
  - Ulnar nerve palsy – explore if <24 hours
  - AIN or absent radial pulse → immediate reduction
  - After surgery a pink-pulseless hand can be observed

Wires should not cross at fracture site, otherwise simply acting as one wire and reduced stability.

Skeletal Dysplasias
Please read separate document on common Skeletal Dysplasias (MACHO MEN OF GOD)
- Disordered osteochondral development – 1 in 10,000 approx
- Where is the abnormality?
  - Rhizomelic
  - Mesomelic – middle
  - Acromelic
Site:
- Epiphyseal → joint problem
- Spine – instability and kyphoscoliosis
- Metaphysis → short, deformity (+ 2 joint issues from malalignment)

Achondroplasia:
- >85% are new mutations
- AD complete penetrance
- Homozygous is lethal
- Fibroblast Growth Factor Receptor – 3
- Point mutation on Chr 4p16.3
- Pathology:
  - Abnormal cartilage
  - Failure of proliferative zone in physis (do not stack up)
  - Inhibition of endochondral bone formation (not membranous bone)
  - Increased bone suppression (so called a gain in function)
- Signs:
- X-ray:
  - Horionatal acetabular
  - Coxa valga
  - V-shaped distal femur epiphysis
  - Long fibula
  - Arnold chiari malformation
  - Throracolumbar kyphosis
  - Short pedicles → spinal stenosis

Pseudoachondroplasia
- Normal at birth – but become short
- Soft tissue laxity
- Joint problems
- Spinal abnormalities

Multiple Epiphyseal Dysplasia:
- Chr 19 abnormality resulting in abnormal COMP protein and Collagen IX
- Linked to: Perthes, Premature OA

Spondylo-epiphyseal Dysplasia
- Collagen 2 or 11 problems
- Display a dominant negative effect

Osteogenesis Imperfecta
- Type 1 Collagen defect
- Type 1a – the mutant allele is non-functional so you simply have 50% of your collagen and mild symptoms
- Other lethal forms have a mutant allele with bad collagen → dominant negative effect.
- Though they fracture easily, they do heal well.

Diaphyseal Achalasia
- Chromosomes 8, 11 and 19
- Affects perichondral ring resulting in osteochondromas
- Grow as you grow
- Deformity and leg length discrepancies
- Potential for malignant transformation

- Treatment principles
  - Correct deformity
  - Support brittle or weak bones – medically or surgically
  - Equalise leg lengths and possible increase height

**Paediatric Spine**

**Idiopathic Paediatric Scoliosis**

- Often familial
- Incidence of curves > 30° and needing treatment is 0.3%
- 2-3% overall prevalence for smaller curves < 10° with less female dominance
- More trivial curves more common
- Can be divided into adolescent, juvenile and infantile

- Adolescent (age 10+):
  - 90% in girls
  - usually primary right thoracic curves, with lumbar curves to the left
  - curves under 20° usually remain static or resolve spontaneously
  - once progression begins → usually continues until growth period over
  - predictors of progression:
    - very young age
    - marked curvature
    - incomplete Risser sign (0 to 5)
      - Female Risser 0-2 and COBB 20-30° → 65% progression risk
      - Female Risser 3+ and COBB 20-30° → 20% progression risk
      - But Males have high chance of progression up to Risser 4

- treatment aims to prevent a mild deformity from becoming severe, and to correct cosmetic appearance which can have psychosocial implications to the patient as well as future cardiorespiratory issues.
  - Most referrals are initially monitored to gauge progression with 4-monthly x-rays of the standing whole spine
  - Well balanced curves under 30° can be monitored
  - If progression seen for curves between 20° and 30°, then consider bracing to prevent further deterioration, but be aware bracing does not achieve any reduction. Milwaukee or Boston brace worn for 23 hours of the day, and does not preclude mild exercise.
    - Issues with compliance and psychohology
  - Surgical correction reserved for curves > 30° that are likely to progress or are that are cosmetically unacceptable. Double major balances curves corrected if > 60°
    - ABC principle: arthrodesis, balancing, correction of deformity.
- **Harrington rods** – placed posteriorly on concave side of curve, attaching to upper and lower ribs to distract the curve if flexible. Fusion then performed along length of curve to hold in place. Does not correct rotation and consequent rib hump.

- **Luque rod and sublaminar wires** – sublaminar wires added in this modification of Harrington system to provide extra support along curve and try to pull backwards on each vertebra to reduce rotation. However, risk of neurological injury increased.

- **Cotrel-Dubousset system** – double rods to distract concavity and compress convexity. Rigid enough to not need post-operative bracing.

- Current practice is to consider growing rods (Isola or VEPTR) to halt curve progression in growing children, lengthening every 6 months to reduce shortened stature, and performing a final fusion after skeletally mature. Alternatively, for untreated significant curves in a skeletally mature patient use posterior double rod instrumentation (e.g. DePuy Expedium or K2M), and consider anterior release and discectomies over apex of curve for better correction and to shorten anterior segment to reduce any distraction injury of cord during straightening. Neurophysiological cord monitoring mandatory.

- **Juvenile (age 4-9):**
  - Uncommon but with worse prognosis
  - Surgical correction may be necessary before puberty
  - However a brace may hold the curve until age 10, for a final fusion

- **Infantile (age < 3):**
  - Rare and reducing prevalence
  - More common in boys – with left thoracic curves
  - 90% can resolve spontaneously, but those with **rib-vertebra angles which differ by >20° between sides** will progress and deteriorate severely with cardiopulmonary sequelae.
  - Treatment:
    - Serial elongation-derotation-flexion plaster casting under GA.
    - Once age 4+ should be able to fit and comply with a brace.
    - If bracing fails to control curve progression, then surgical intervention needed to control curve using growing rods.
    - Avoid posterior fusion to allow as much height as possible.

- **ICEOS 2012 Conference:**
  - New classification system or early onset (EOS) and late onset scoliosis.
  - Cut off age is 7 years
  - Reflects importance of alveolar hyperplasia during the first 5 years following birth (rise from 20 million to 300 million)
  - 35° Cobb angle is a limit for moderate scoliosis under which there is a role for bracing – this can control or improve Cobb angle, or at the very least delay the need for surgery and allow pedicles to grow enough to accommodate screws.
  - Treatment stratification:
    - Bracing
- Growing rods - manual lengthening, controlled self lengthening (SHILLA) or external lengthening (MAGEC)
- Vertical expanding prosthetic rib (VEPTR) better for primarily chest wall problems.
- Growing rods susceptible to law of diminishing returns where most only grow for 5 years.
- Risk of auto-fusion over the years, but always need assessment for definitive surgical fusion (with or without instrumentation) on completion of majority of growth

Scheuermann’s Kyphosis
- Most common cause of thoracic (>45°) or thoracolumbar (>30°) hyperkyphosis in adolescence
  - Normal thoracic kyphosis is 25-45°
  - 4-8% incidence and no gender bias
  - Minimum of 3 vertebra wedged >5°, with irregular endplate changes
- Present in late childhood and adolescence with increasing kyphosis
  - often with compensatory lumbar or cervical “goose-neck” hyper-lordosis
  - This hyper-lordosis can also lead to hamstring tightness
- Symptoms may be minimal and simply a cosmetic issue, or...
  - Dull non-radiating back pain ± local tenderness
- Investigate with supine and hyper-extended x-rays over a bolster to assess mobility. MRI prior to any surgery
- Natural history untreated of curves < 72°:
  - No difference in types of employment
  - No difference in pain medications / incidence of back pain
  - No difference in sick leave
- Curves >85° had lower inspiratory capacity, without any major life interference.
  - Disc degeneration tends to occur at the apex of the curve
  - Neurological deficits can correlate with shorter-segmented curves
- Symptoms:
  - Deformity (fixed)
  - Pain
  - Neurology is rare
  - Association with spondylolysis
- Treatment options:
  - Physiotherapy – 16-32% reduction in pain
  - Brace therapy:
    - useful if flexible deformity <65°
    - early instigation of brace, with an initial correction of >15°
    - worn for 16-23 hours a day for 18 months, followed by 18 months of gradual weaning.
Return of deformity in as much as 30%

- Indications for surgery:
  - Severe deformities >72°
  - Failed conservative management for 6 months

- Harrington rods:
  - In addition to pre-operative traction and 6-9 months post-operative casting.
  - High complication rate: rod breakage, loss of correction, wound infection, junctional kyphosis at adjacent vertebral joints.
  - Fusion should extend from T2 to 1st lordotic segment.
  - Concurrent anterior release did not improve symptoms or function, but more risks.

Neuromuscular Curves

- Deformity tends to be progressive even after skeletal maturity, with inadequate compensatory curves producing a loss of balance
  - Typical paralytic curve is convex towards the side of the weaker spinal / thoracic / abdominal musculature
  - Can result in imbalance and instability to the spine, which may make sitting or hygiene problematic (costo-iliac impingement).

- Treatment poses ethical dilemmas in terms of consent and balancing the prognosis of the underlying pathology with the risks of surgery.
  - Mild curves may require no treatment.
  - Moderate curves can be monitored initially and then braced either externally or internally.
  - Progressive or severe curves may simply be managed with sitting orthoses, but may require staged surgical release and stabilisation.
  - May not help with respiratory or GI problems, which are contributed to from reflux, and the curve tends to be below the diaphragm.
  - Aim for improved sitting and independency, along with pain relief from impingement and better hygiene.
  - More risks and complications:
    - Poorer bone quality for screw fixation
    - Soft tissues oedematosus and non-compliant
    - Respiratory problems and cardiomyopathies common
    - Nutritional status poor – check for albumin or lymphocyte deficiency
    - Uncooperative, epilepsy or other involuntary movements may increase risks of non-union
    - Higher infection rates (up to 10%)

- Classification:
  - Neuropathic
    - Upper motor neurone
      - Cerebral palsy
      - Spino-cerebellar degeneration – CP, Friedreich’s, CMT
      - Syringomyelia
• Spinal cord tumour / trauma
  ▪ Lower motor neurone
  ▪ Poliomyelitis
  ▪ Other viral myelitis
  ▪ Traumatic
  ▪ Spinal muscular atrophy

➢ Myopathic
  ▪ Arthrogryposis
  ▪ Muscular dystrophies – Duchenne’s
  ▪ Congential hypotonia
  ▪ Myotonia dystrophica

❖ Treatment principles are to restore function, taking into account any likely neurological progression from the primary disease, and the patient’s mobility and prognosis
  ➢ Bracing is used to hold a curve static to allow younger patients some growth before definitive surgery.
  ➢ Recently the VEPTR (vertical expanding titanium rib) can allow indirect internal control.
  ➢ Wheelchair adaptations can achieve a similar effect to a brace without restricting chest movements
  ➢ Physiotherapy with regular stretching important but will not alter curve progression.

❖ Surgery tends to involve long posterior corrections to prevent junctional recurrence above or below the fused segments. Anterior releases at the apex of very severe curves, and anterior surgery reserved for children under 10, where further anterior growth in the presence of posterior fusion would otherwise result in crankshaft failure.

❖ Cerebral Palsy – 25% have spinal deformity, frequency increases with level of disability.
  ➢ Group 1 have balanced curves – treated surgically when curve has reached 60°, and usually does not need to be extended to pelvis
  ➢ Group 2 have unbalanced curves with pelvic obliquity
    ▪ Usually more severely disabled with stiff curves needing anterior release and posterior correction from T2-T3 all the way to pelvis.

❖ Neurofibromatosis:
  ➢ Scoliosis can affect up to 33% of patients with neurofibromatosis
  ➢ Usually a short, sharp curve
  ➢ **Diagnostic Criteria 2 or more:**
    ▪ 6+ café-au-lait spots (>5mm pre-pubertal, >15mm post-pubertal
    ▪ 2+ any neurofibromas or 1+ plexiform neurofibroma
    ▪ Crowe’s sign = freckling in axillary or inguinal regions
    ▪ Optic glioma
    ▪ 2+ Lisch nodules (iris hamartomas)
    ▪ distinctive bony lesion
      (e.g. thinning of long bone cortex or sphenoid dysplasia)
- an affected 1st degree relative
- Managed as described above, but there is a higher rate of graft dissolution and pseudoarthrosis to be considered when planning final surgery.
- BEWARE paralysis from dislocated rib head in the foramen, and intradural neurofibromas which tether cord

- Dystrophic curves
  - Short sharp curve
  - Scalloping of vertebra from dural ectasia
  - Pencil ribs due to expansion of intercostal nerves

- Ehlers-Danlos – avoid anterior surgery, due to friable anterior vessels.

- Duchenne’s Muscular Dystrophy = X-linked affecting 1 in 3500 males.
  - Progressive scoliosis in 95%, with initial hyperlordosis in 5%
  - Usually have problems in teenage years, when deteriorate from standing and go into a chair.
    - Muscle weakness more progressive at this stage, and lose compensatory hyperlordosis when walking – which tensions the ALL to stabilise the spine
  - Beware: dilated cardiomyopathy and reduced lung function → plan surgery when curve is around 20° (i.e. quite early)
  - But normally eat by equal movements or arm to mouth, and flexing spine down to arm → spinal fusion can mean they lose this “last independent action” of self feeding
  - Usually only posterior correction to avoid further chest function impairment

- Marfan’s
  - Aortic defects mean anaesthetic risk
  - Dural ectasia results in dilated dura and scalloped pedicles – difficult to place screws.

**Congenital Curves**

- Associations:
  - Klippel-Feil 25%
  - Renal 30%
  - Cardiac 10%
  - Neural axis 15%
  - 25% static, 50% moderate progression, 25% severe progression

- Failure of formation
  - Wedge Vertebra – usually in lower thoracic and lumbar regions, again progressing slowly at 1-2° per year
  - Hemivertebra (unsegmented) – little growth potential and slow growth, usually <30° at maturity
  - Multiple or segmented hemivertebra – as the site of the anomaly goes down the side, the progression rate increases.
  - Sacral agenesis – in diabetic mothers
- Failure of separation / segmentation
- Block vertebra – often multiple and stunt trunk growth, but progress at a slow rate as themselves have little growth potential
- Unilateral unsegmented bar – progression rate increases further down the spine, and tend to result in trunk imbalance, pelvic obliquity and large secondary curves.
  - Resection not effective because of re-fusion, but will respond to distraction (e.g. VEPTR – vertical expanding titanium rib)

- Mixed defects:
  - Potential for rapid deterioration in excess of 14° per year.
- Complex / unidentifiable pattern
- Other causes:
  - Absent or fused ribs
  - Fractures from rickets or osteogenesis imperfect

- Rate of progression based on type and site:
  - Failure of formation – 1 to 2° per year upper thoracic, 2 to 3° in lower thoracic, >3° per year in thoracolumbar region.
  - Failure of segmentation – 3 to 6° per year in thoracic region, 6 to 9° in thoracolumbar region, and about 6° per year in the lumbar region.

- Assess for clues such as tufts of excessive hair, skin dimples, naevi.
- Spina bifida may be associated along with visceral anomalies
- MRI to assess for Arnold-Chiari malformations, diastematomyelia, cord tethering, syrinx or conditions that put the cord at risk during correction.
- Often require staged anterior release and posterior correction to reduce the risk of cord traction injuries.
- No increased risk of next child having a congenital scoliosis – reassure parents.

**Spondylolysis**

- Defect in Pars Interarticularis
- Listhesis is the subsequent forward slip.

- In children – most happen at age 1-2 when start walking
  - Infant gait is hyperlordotic with abdomen pushed out
  - Pressure and fracture of Pars Interarticularaes
  - Pre-requisite is ambulant child
  - 2nd group of children are the elite athletes age 16.

- Most common area is L5 (90%)

- Symptoms:
  - Back pain from pseudoarthrosis
  - Reduced exercise tolerance – hamstring tightness resulting in Pelvic waddle
  - Radicular symptoms
  - Deformity

- Treatment options:
- Restrict activity, brace, physiotherapy
- Repair
- Fuse above and below stabilisation

**Spinal Trauma**

- Factors:
  - Large head
  - Flexible spine
  - Ligamentous laxity
  - Facet orientation
- Lap belt injuries – look for Naked Facet Sign
  - Unstable and require stabilisation
  - Beware intra-abdominal or vascular pathology
- SCIWORA
  - Can result from traction and end plate separation
  - Vascular infarct may occur
  - Variable prognosis – but usually poor
- Back pain in children
  - Mechanical – postural or rare disc herniation
  - Developmental – Sheuermann’s or spondyloysis
  - Neoplasms
  - Inflammatory – Rheumatoid, discitis, infection
  - Psychosomatic
- Red Flags ➔ MRI or bone scan
  - Night pain
  - Continuous
  - Off school
  - Off sports

**Musculoskeletal Tumours in Children**

- Needle biopsy:
  - Less traumatic with less pain and infection
  - Can achieve multiple sites from single entry point
  - Can be done in the OPD
  - Less contamination
- Incisional Biopsy:
  - Incision in longitudinal line of planned resection
  - Dissection directly down to the growing edge – NO FLAPS
  - Meticulous haemostasis
  - Water-tight closure in layers
  - Pre-determined transport arrangements and correct medium
Staging – local and systemic.

Management depends on
- Tumour type
- Natural history
- Malignant potential
- Distant metastasis

Management options:
- Observation
- Intrallesional
- Marginal – reactive zone
- Wide excision – 2cm periphery
- Radical – compartment
- Adjuvant therapies

Desmoid tumour
- Local malignant
- Surgical, radiotherapy and chemotherapy options

Digital Fibromatosis
- Unique lesion of fingers and toes
- High recurrence
- Unknown malignant potential

Synoviosarcoma
- 2nd commonest paediatric sarcoma
- Surgery + chemotherapy
- Enblock compartment resection offers 5 year survival of 75%

Rhabdomyosarcoma
- Lower limbs > upper lombs
- Surgical, Chemo
- If no mets → 70% 3-year survival, If mets → 20%-30%

Malignant Bone tumours:
- 0-5: metastatic (renal or neuroblastoma common), leukaemia, Ewing
- 5-10: osteosarcoma, rhabomyosarcoma
- 10-20: chondrosarcoma, Ewing, osteosarcoma, rhabomyosarcoma
MTS Character type of lesions:
- 1 – superficial, static
- 2 – local and contained
- 3 – active and local destruction

Enneking thinking: where is it, what is it doing the bone, and how is the bone responding?

Metaphyseal-diaphyseal lesions tend to be benign cartilage or bony lesions that are still locally aggressive.

Risks of osteochondroma:
- Single – 1%
- Multiple – 10%
- Traditional excision should include base of tumour and be an extra-periosteal excision – beware damage to perichondrial ring and growth arrest

Unicameral cysts treated by aspiration and injection of bone marrow or methylprednisolone. But if already fractures, then wait 6 months.

ABC
- 50% recurrence with curettage alone
- 20% recurrence with curettage and grafting

Enneking:
- Grade 1 low
- Grade 2 high

Adamantinoma – tibia and mandible (90%)
- Age 10-50

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<table>
<thead>
<tr>
<th>Age (years)</th>
<th>Benign</th>
<th>Malignant</th>
</tr>
</thead>
<tbody>
<tr>
<td>0-5</td>
<td>Osteomyelitis</td>
<td>Osteosarcoma</td>
</tr>
<tr>
<td>5-10</td>
<td>Unicameral bone cyst, Aneurysmal bone cyst, Nonossifying fibroma, Osteoid osteoma</td>
<td>Osteosarcoma, Rhabdomyosarcoma</td>
</tr>
<tr>
<td>10-20</td>
<td>Fibrous dysplasia, Osteoid osteoma, Aneurysmal bone cyst, Chondroblastoma, Osteofibrous dysplasia</td>
<td>Osteosarcoma, Ewing sarcoma, Chondrosarcoma, Synovial Sarcoma, Rhabdomyosarcoma</td>
</tr>
</tbody>
</table>
- Large and expansile
- Wide excision

- Osteosarcoma – long bone diaphysis, bimodal distribution

- Ewing's:
  - < 30 years
  - any long bone, pelvis or spine
  - metaphyseal / diaphyseal
  - lytic
  - differential is infection

- Chondrosarcoma
  - Rare in children, usually in >50
  - Painful and slow growing
  - Any grade, and may be de-differentiated
  - Surgical without adjuvant therapy
  - Pink and blue on slides