

Legg-Calvé-Perthes disease

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Incidence and aetiology This rare condition, characterised by the development of AVN of the proximal femoral epiphysis, predominantly affects boys aged 4–7 years; 10% develop bilateral disease. Although the aetiology is unclear, factors such as socioeconomic deprivation and passive smoking have been implicated. Other causes of femoral head AVN must be considered, particularly in bilateral cases (Table 44.6).

Pathology Once established the process follows a well-described course. The avascular change may affect all or part of the femoral epiphysis. If the avascular bone collapses, this is followed by revascularisation, resorption and fragmentation of the dead ossific nucleus within the cartilaginous femoral head, and finally by reossification and regeneration ('healing') of the bony epiphysis. In this respect, Perthes' disease is a self-limiting condition but, during the collapse and fragmentation phases, femoral head deformity occurs as the cartilage 'follows' the Legg, Calvé and Perthes all described osteochondritis of the head of the femur independently in 1910. Arthur Thornton Legg , 1874–1939, orthopaedic surgeon, The Children's Hospital, Boston, MA, USA. Jacques Calvé , 1875–1927, orthopaedic surgeon, La Fondation Franco-Américaine, Berck Plage, Pas-de-Calais, France. Georg Clemens Perthes , 1869–1927, Professor of Surgery , Tübingen, Germany .

shape of the reossifying epiphysis. This change in shape is irreversible and has a permanent effect on hip function. **Diagnosis** The history , clinical examination and anteroposterior and 'frog' lateral pelvic radiographs make the diagnosis. An intermittently painful hip (or knee) with a limp and irritability or restriction of hip movements requires investigation. The radiographic features vary with the disease stage and may not correlate with the clinical condition (Figure 44.21).

TABLE 44.6 Causes of avascular necrosis of the femoral head. Steroids Infection/surgery/previous injury Perthes' disease Sick cell disease Hypothyroidism Multiple epiphyseal dysplasia will show AVN-like appearances in both femoral heads

AI CE (b) Figure 44.19 Anteroposterior pelvic radiographs demonstrating the acetabular index (AI) and the centre-edge (CE) angle: (a) normal hips; (b) the left hip shows residual dysplasia. The AI is increased when compared with the normal right hip. The left CE angle would be smaller too, but it has not been measured on this radiograph.

Management The prognosis, and hence the management, is influenced by the extent of AVN and the degree of collapse. The Herring classification is popular but can only be applied when the head is in the fragmentation phase. If the anterolateral portion of the head is preserved, the prognosis is good. Treatment aims to minimise femoral head deformity and the risk of secondary acetabular dysplasia by maintaining a good range of joint movement with analgesia and physiotherapy. The use of crutches and/or wheelchairs is discouraged because they promote a flexion/adduction posture. Brace management does not alter the natural history. The role of operative treatment is controversial. Surgery ordered early to prevent deformity secondary to can be performed to 'salvage' a poor mechanical femoral head collapse or in a situation when deformity is limiting movement (Table 44.7 Summary box 44.9 Legg-Calvé-Perthes disease

/uni25CF John A Herring , contemporary , pediatric orthopedic surgeon, chief of staff emeritus, Texas Scottish Rite Hospital for Children, and professor, UT Southwestern Medical Center, Dallas, TX, USA. - -). Not all hips with deformity require 'salvage' surgery: young children, with more time to remodel, have a better prognosis as the acetabular changes (in response to the altered femoral head shape) result in an aspherical but congruent joint. Degenerative change may occur in adult life.

Figure 44.20 Anteroposterior pelvic radiograph of a 7-year-old girl who developed avascular necrosis secondary to a postoperative wound infection following a closed reduction of her dislocated left hip. Note the destruction of the femoral head and the proximal femoral physis, so that the femoral neck is short and the greater trochanter relatively high (arrow). Most common in boys aged 4–7 years AVN leads to femoral head collapse; the return of the blood supply heralds the resorption and reossification phases that allow the femoral head to 'heal' The prognosis is better in younger children (and in boys), who have more remodelling potential before skeletal maturity Management aims to maintain femoral head sphericity Treatment may be non-surgical (to maximise range of movement) or surgical (early for containment or late for 'salvage') (b) Figure 44.21 Anteroposterior pelvic radiographs of Perthes' disease demonstrating whole head involvement: (a) right-sided disease; the process is in an early phase and the area of dense necrotic bone is visible; (b) there has been collapse and fragmentation. The Herring classification relates to the height of the lateral pillar (lateral portion of the epiphysis) in the fragmentation phase of the disease.

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Revision #1

Created 2025-12-31 15:16:48 UTC by Omar Ayman

Updated 2025-12-31 15:16:48 UTC by Omar Ayman