

Bilateral hip pain with right proximal femoral lesion







Legg-Calve-Perthes

- Idiopathic osteonecrosis of the femoral head epiphysis during childhood
- First described by Arthur Thorton Legg in 1909 and published in 1910 the same year that Frenchman Jacques Calve and German Georg Perthes produced similar descriptions
- Exact pathology is not clear
 - Larger cartilage: LCP patients tend to have a delayed bone age, on average 2 yrs in girls and 1 yr in boys → femoral head ossific center is smaller than those in children of similar chronologic age → cartilaginous component of epiphysis relatively larger and traversing blood vessels are more vulnerable to mechanical compression
 - Vascular insufficiency: Lateral epiphyseal artery (LEA) extends into the epiphysis (ultimately arises from the medial femoral circumflex artery). Angiography showed interuption of the origin of the lateral epiphyseal artery in 68% of LCP patients. Perfusion is position dependent. The combination of abduction with internal rotation interrupts flow through the LEA as it traverses the capsule.
 - **Repetitive loading:** associated with increased flattening of the femoral head
 - Association with hypercoagulable disorders debated

Classic findings of LCP



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THE "SAGGING ROPE SIGN" IN AVASCULAR NECROSIS IN CHILDREN'S HIP DISEASES - CONFIRMATION BY 3D CT STUDIES

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ABSTRACT

Growth disturbance of the proximal femoral epiphysis and physis secondary to avascular necrosis (AVN) in a variety of children's hip disorders produces changes in the femoral head and neck that make radiographic interpretation difficult. The enlarged overhanging femoral head produces radiographic markings on the femoral neck which are sometimes confusing. These have sometimes been misinterpreted as growth arrest lines. Apley and Wientroub reintroduced Perkins' description of the "sagging rope" sign in AVN of the femoral head, and Clarke clarified that this puzzling radiographic transverse metaphyseal line overlying the femoral neck in fact represents the margin of the femoral head rather than a growth arrest line. Their report was made after studying plain and stereoscopic radiographs alone. Our review of 23 cases of femoral head AVN in children, documented by 3 dimensional computerized tomographic (3D CT) radiographs of the femoral head and pelvis, confirms Clarke's view of the nature of the "sagging rope" sign. These sophisticated radiographic studies provide new detail and understanding of head-neck relationship in AVN which allows better planning for surgical correction of hip disorders in children.



Figure 1 Depiction of "sagging rope" sign: a concept proposed by Perkins to define the lower margin of the femoral head in AVN in children. The radiopaque line, which resembles a loosely held jump rope, represents the margin of the true femoral head.

of the hip in children with AVN. The line is seen overlying the femoral neck and extends laterally from the inferior border of the neck for a variable distance, frequently reaching the superolateral border of the neck. Usually the line is curved and sags at its center but is sometimes almost straight.

Waldenstrom Stages

- Initial Stage: small sclerotic epiphysis with medial joint widening, clinically occult
- Fragmentation: period of resorption lasts 6 months, collapse occurs in the softened femoral head from the osteoclastic removal of dead bone without reformation, inhomogenous dense physis, metaphyseal cysts, most symptomatic
- Reossification: new bone formation proceeding from lateral to medial and posterior to anterior, last 18 months
- Healing or remodeling: trabecular patterns reform, eventually take on the final shape of the femoral head and acetabulum

Epidemiology

- Clinical onset 3-12 yrs of age (median peak incidence 6 yo)
- 15-20% bilateral (usually asynchronous)
- Incidence:
 - Highest in whites and lowest in African Americans
 - Boys 5x more than girls
- Low twin concordance pointing more towards environmental causes as opposed to genetic predisposition
- Role of nurture over nature: incidence 4 to 32 per 100,000 in highest and lowest social classes, respectively
- Associated with congenital malformations such as inguinal hernia, GU abnormalities, undescended testes, and Down's syndrome

Clinical Presentation

- Variable from a painless limp to irritable hip
- Knee pain referred to hip
- Limited ROM especially with abduction and internal rotation
- Late in disease- early OA, femoroacetabular impingement

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- Clue to diagnosis:
 - Bilateral involvement almost always demonstrates hips to be in different stages of disease
 - Hand radiograph for bone age often shows delayed bone age in LCP
- Symmetric involvement suggests other conditions such as epiphyseal dysplasia
- Atypical presentations consider osteonecrotic conditions, such as hypothyroidism, Gaucher's Disease, glycogen storage defects, sickle cell anemia, Meyer's dysplasia
- Septic arthritis

Prognosis

- 60-70% heal spontanouesly without functional impairement at maturity
- RF with worse outcome
 - Older skeletal age at time of presentation
 - XR:
 - More horizontal physis seen with almost total head involvement
 - Lateral pillar= lateral 30% of the epiphysis
 - >75% of the lateral pillar demonstrates significantly poorer prognosis
 - Lateral subluxation
 - Calcification lateral to epiphysis
 - Gage sign- radiolucent "v" in lateral epiphysis
 - Metaphyseal "cyst" formation
 - >20% epiphyseal extrusion
 - >50% femoral involvement
 - Physeal bar





Treatment

- Conservative for those with decrease risk for progression
 - Bed rest +abduction stretching and bracing
- Surgical with goal of preservation of ROM

 proximal femoral and pelvic osteotomy to cover
 femoral head

References

Bull Hosp Jt Dis (2013). 2014;72(1):18-27. Legg-Calvé-Perthes disease: an overview with recent literature. Chaudhry S, Phillips D, Feldman D.

Iowa Ortho J. 1995; 15:101-111. The "sagging rope sign" in avascular necrosis in children's hip diseases— confirmation by 3D CT studies. Kim et al.

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Learning Radiology

Prognosis

- Perthes study group, multicenter prospective study with 438 patients
- Non-operative group managed with one of three different protocols and operative group with two surgical protocols
- LCP children between 6-12 yo
- Children with symptom onset prior to 8 yo are likely to have good result regardless of treatment
- Onset after age 8 with at least 50% lateral pillar height maintenance (Herring Groups B and B/C), operatively managed hips had better outcomes than non-operatively managed hips
- Group C hips with greater than 50% collapse of lateral pillar height had poor outcome regardless of treatment