SportsMedRef – Pediatrics

Legg-Calvé-Perthes Disease (LCPD)

1. Pathogenesis

Legg-Calvé-Perthes Disease (LCPD) is an idiopathic avascular necrosis of the proximal capital femoral epiphysis in the pediatric population.[1,2] LCPD develops after a double insult of at least two episodes of proximal femoral epiphysis ischemia.[3,4] The epiphysis undergoes collapse, resorption, reossification, and eventually, healing.[2] The course of this disease is denoted by the Waldenström stages: synovitic, avascular, fragmentation (collapse), reossification (healing), and healed (residual).[2]

2. Risk Factors

The cause of LCPD remains unknown, though several factors have been hypothesized to play a role[1-5]:

- Micro-trauma
- ADHD
- Passive smoke exposure
- Low birth weight
- Obesity
- Hypercoagulability
- Positive family history

However, no causal link between these factors and LCPD has been definitively demonstrated, suggesting a multifactorial pathogenic process.[1-5] The predominant theory is that LCPD is caused by a combination of genetic and environmental factors.[4,6] Genetic factors may make the blood supply to the capital femoral epiphysis susceptible to disruption, while environmental factors, such as repeated subclinical trauma, may trigger the disease.[6]

3. Epidemiology

Although most cases of LCPD are unilateral, approximately 15% of cases are bilateral.[7] The peak incidence of LCPD occurs in children between the ages of 4 and 8 years.[1,6,8] The reported incidence of LCPD varies from 4 to 32 per 100,000, with the condition being 5 times more common in males than females.[6,8] Racial differences in incidence of LCPD have been observed, with LCPD being more common among White children compared to East Asian and Black children.[6,8] Interestingly, the incidence of LCPD is lower in equatorial regions and with increasing with latitude.[6,8] A higher incidence of LCPD has also been found in less densely populated areas and among individuals from lower socioeconomic backgrounds.[1,2,6]

4. Initial Presentation

Patients with LCPD usually present with a painless limp for a few weeks to months.[9] Pain may or may not be present, and the location of the pain can vary.[9] Due to involvement of multiple nerves, pain may be referred to the knee (femoral nerve), medial thigh (obturator nerve), or buttock (sciatic nerve).[9] Range of hip motion may be affected, first with limited abduction and internal rotation, followed by a

Trendelenburg gait.[5] In unilateral cases, patients may appear to have shortening of the affected extremity.[9] On physical examination, the most consistent signs of LCPD are limited hip abduction and internal rotation (best tested with the hip extended).[9] Patients often have weak quadriceps and hip abductors with muscle atrophy.[9] In severe cases, adduction contracture may form, but hip flexion and extension are rarely affected.[5]

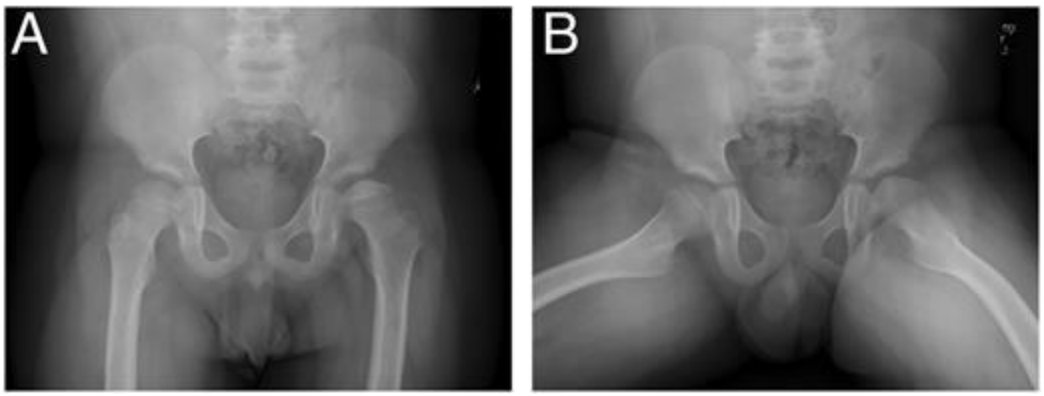
5. **Clinical Differential Diagnosis[10,11]**

    a. Slipped Capital Femoral Epiphyses (SCFE)
    b. Developmental dysplasia of the hip (DDH)
    c. Limb-length discrepancy
    d. Inflammatory etiologies:
        - Transient synovitis
        - Juvenile idiopathic arthritis (JIA)
        - Reactive arthritis
        - Chondrolysis
    e. Infectious etiologies:
        - Septic arthritis
        - Osteomyelitis
        - Pyomyositis
    f. Overuse injuries:
        - Muscle strains and tendinopathies
        - Apophysitis and apophyseal avulsion injuries
        - Stress fracture
        - Snapping hip syndrome
    g. Tumors or Malignancies:
        - Ewing sarcoma
        - Osteoid osteoma
        - Acute leukemia
        - Neuroblastoma
    h. Referred pain (e.g. low back, intra-abdominal, or pelvic pathology):
        - Discoid lateral meniscus
        - Osteochondritis dissecans

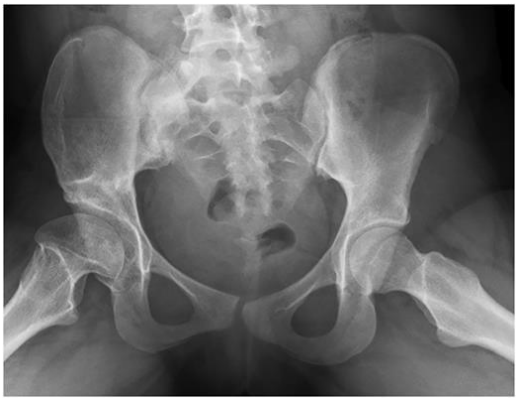
6. **Imaging**

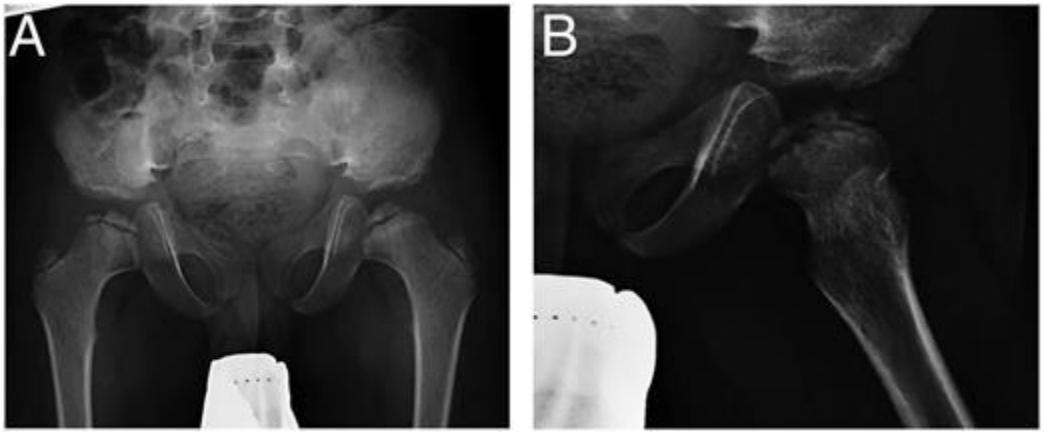
<u>Radiographs</u>

Radiographs serve as the primary imaging modality for diagnosis and surveillance of LCPD.[6] Initial imaging should include an AP pelvic radiograph with bilateral frog-leg lateral views (hips flexed and abducted).[9] A single hip radiograph is not adequate because comparison with the contralateral hip cannot be made, and bilateral disease may thus be missed.[9] Early radiographic signs include flattening of the femoral head and subchondral sclerosis, while later signs include extrusion of the femoral head laterally such that it is not contained by the acetabulum.[9] Of note, prognostic radiographic signs rarely appear until LCPD is established, and usually take over six months after the onset of the disease.[5]



AP pelvic radiograph with bilateral frog-leg lateral views showing early radiographic signs of LCPD on the left, with femoral head flattening and subchondral sclerosis. Image from the American Academy of Pediatrics.[9]


Bilateral frog-leg lateral view pelvic radiograph shows flattening and extrusion of the right femoral head. Image from the American Academy of Pediatrics.[9]

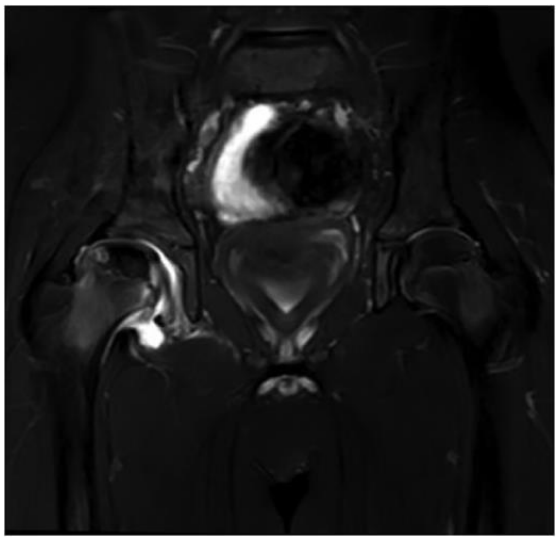


A. AP pelvic radiograph showing bilateral LCPD with fragmentation of bilateral femoral heads. B. Fragmentation on lateral view of the left hip. Image from the American Academy of Pediatrics.[9]

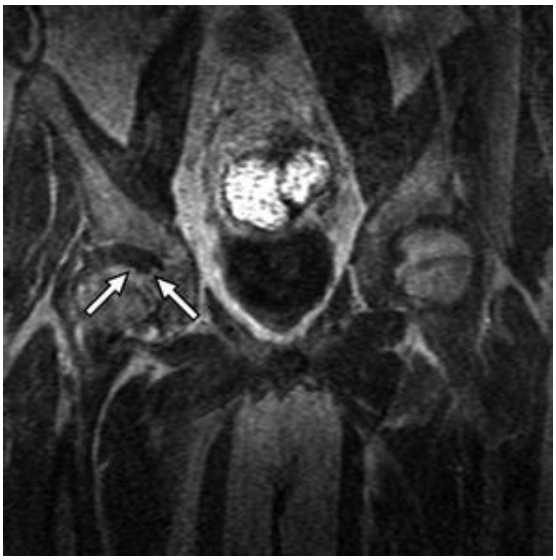
MRI

MRI can be used to diagnose LCPD at a very early stage, when radiographs are still normal.[3] Contrast-enhanced MRI has been shown to be more sensitive than radiography and provide more

specific information about the blood flow to the femoral head for detection of early ischemia.[6,7] Importantly, MRI does not expose the pediatric patient to the harmful effects of ionizing radiation.[7]



Perfusion-weighted MRI of bilateral hips showing LCPD of the right hip, with femoral head flattening and lateral extrusion relative to the acetabulum, decreased contrast uptake (dark signal), and a hip joint effusion (bright signal). Image from the American Academy of Pediatrics.[9]



3D T1-weighted spoiled gradient-recalled echo fat-saturated image acquired after IV administration of contrast showing central nonenhancement of subchondral right proximal femoral ossific nucleus (arrows). Of note, right hip radiographs were normal. Image from the American Journal of Roentgenology.[7]

Radiographic Outcome Classification Systems

Stulberg Classification

The Stulberg classification system is the most commonly used radiographic outcome classification system for evaluating the long-term prognosis of LCPD after skeletal maturity.[3,6] It divides patients into 5 classes, which are further collapsed into 3 groups, based on the shape and size of the femoral head, the shape and dimension of the acetabulum, and the congruency between the femoral

head and the acetabulum at skeletal maturity on AP and lateral radiographs.[3,6] Class I/II patients are predicted to have no osteoarthritis, Class III/IV patients, mild-to-moderate osteoarthritis, and class V patients, severe osteoarthritis before 50 years of age.[3]

Lateral Pillar Classification

Another commonly used radiographic outcome classification system, the lateral pillar classification, described by Herring in 1992, distinguishes four groups based on a simple pelvic radiograph. This classification relies on the appearance of the lateral pillar of the proximal femoral epiphysis on AP radiograph obtained at the onset of fragmentation.[3] The lateral pillar classification system was initially divided into three groups: A, normal lateral pillar; B, lateral pillar >50% of its original height; and C, lateral pillar ≤ 50% of its original height.[3] In 2004, the classification was modified to include a fourth group: B/C border group.[3] Patients in group A have favorable outcomes, while outcomes for patients in group B depend on age at onset: most patients ≤ 9 years at onset have good outcomes, whereas most patients > 9 years at onset have unfavorable outcomes.[3] Outcomes are usually unfavorable in group C patients.[3] Of note, the lateral pillar classification system is applied during the fragmentation stage, when significant femoral head deformity has already occurred.[6] Thus, the clinical applicability is hampered by the need to wait until the fragmentation phase.[6]

7. Radiographic Differential Diagnosis[12,13]

a. Infectious etiology (e.g. septic arthritis, osteomyelitis, pericapsular pyomyositis)
b. Transient synovitis
c. Multiple epiphyseal dysplasia (MED)
d. Spondyloepiphyseal dysplasia (SED)
e. Sickle cell disease
f. Gaucher disease
g. Hypothyroidism
h. Meyers dysplasia

8. Treatment

Goals of treatment

The goal of treatment for LCPD is to minimize femoral head deformity and thus reduce the risk of secondary degenerative osteoarthritis later in life.[6] The choice between operative and non-operative treatment is based on the concept of containment, which involves maintaining the femoral head within the acetabulum throughout the entire evolution of the disease, thereby protecting the vulnerable segment of the epiphysis from being subjected to deforming forces.[6]

Non-operative treatment

Non-operative treatment of LCPD focuses on elimination of weightbearing such as through use of crutches or a wheelchair.[6] Clinical studies of non-weight-bearing for treatment of LCPD have shown conflicting results: some studies suggesting a benefit, others do not.[6] Although orthoses have been designed to contain the femoral head in the acetabulum, current evidence in the literature does not support its use for the treatment of LCPD.[5,6] Recently, bisphosphonates, which decrease bone resorption by limiting osteoclastogenesis, have been shown to delay the resorption of necrotic bone and to decrease femoral head deformity in animal models of LCPD.[4,6] A recent experimental study has suggested that exogenous bone morphogenetic protein (BMP)-2 administration may hasten the

restorative process by stimulating bone healing.[4] However, the use of bisphosphonates and bone anabolic therapy to treat LCPD is experimental, and additional studies are required prior to clinical use.[6]

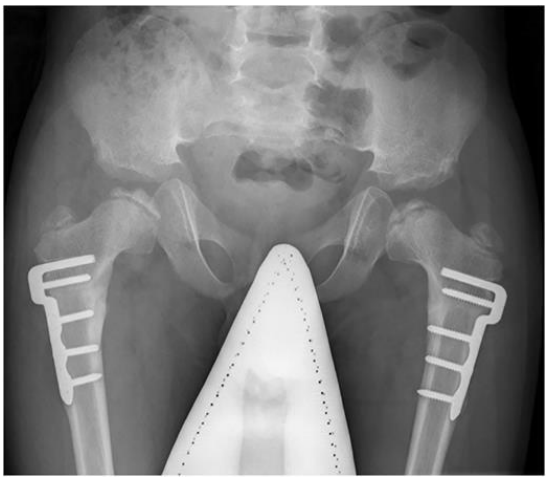
Making the choice between conservative vs. surgical treatment

Children with a skeletal age of 6 years or less at the onset of LCPD often do well without surgical treatment.[1] Surgical treatment should be considered in children 6 years or older who have over 50% femoral head necrosis at the time of diagnosis.[1]

Patients who fall under lateral pillar classification group A (see "Radiographic Outcome Classification Systems") consistently have favorable outcomes, so surgery is not recommended.[3] Surgery improves the outcomes of patients with extensive epiphyseal involvement, such as those in lateral pillar group B and B/C.[3] However, surgery is not recommended for patients with lateral pillar group C disease, as surgical management has not been effective in preventing the nearly consistently unfavorable outcome of Group C disease.[3]

Surgical treatment

The goal of surgical management of LCPD is to prevent loss of joint congruence by restoring the epiphysis to its central position within the acetabulum.[3] Containment can be achieved by means of an osteotomy of the femur and/or pelvis.[6] Several surgical techniques have been described, including femoral varus osteotomy (FVO), Salter's innominate osteotomy, and triple pelvic osteotomy, among others.[3] The best approach is controversial, with conflicting opinions on optimum treatment for LCPD.[1,6]



AP pelvic radiograph status post bilateral proximal femoral varus osteotomies. Image from the American Academy of Pediatrics.[9]

9. Prognosis

Factors associated with prognosis

The prognosis of the hip joint affected by LCPD depends on the age of the patient at the time of onset, the stage of the disease, the extent of epiphyseal involvement, and the lateral extrusion of the femoral head.[6]

Age at onset and the lateral pillar classification (see "Radiographic Outcome Classification Systems") are the two main prognostic predictors that can help guide surgical decision-making.[3] Prognosis is consistently favorable when onset occurs before six years of age, whereas severe residual abnormalities can be expected in patients older than eight years at onset, regardless of treatment pursued.[3,5,6]

Girls have been found to have a worse prognosis compared with boys.[6] Other clinical features, such as heavier weight, stiffness with progressive loss of hip range of motion, adduction contracture, and longer duration from onset to completion of the healing phase, have been associated with a poor prognosis.[6]

Outcomes in Adulthood

LCPD can cause pain, femoroacetabular impingement, and hip osteoarthritis in adulthood.[3] However, hip osteoarthritis rarely develops before 50 years of age.[3] The risk of osteoarthrosis depends chiefly on the degree of joint incongruence.[3]

Direct Links

1. https://www.ncbi.nlm.nih.gov/pmc/articles/PMC6429000/
2. https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4063164/
3. https://www.sciencedirect.com/science/article/pii/S1877056817303249
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5. https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4151449/
6. https://journals.lww.com/jbjsreviews/FullText/2016/07000/The_Pathogenesis_and_Treatment_of.4.aspx?casa_token=MbyWMXlkUvMAAAAA:KtPfxGt7w0dDM4shucoCFS5AZgaLJ_VMobYh4MiFUBDE5UGHvXC5cK_f1f8lwQK3RXTP4-Hskvn8dM4-1QKq61Dd
7. https://www.ajronline.org/doi/10.2214/AJR.09.2444
8. https://academic.oup.com/aje/article/175/3/159/105365
9. https://publications.aap.org/pediatricsinreview/article/39/9/454/35194/Pediatric-Hip-Disorders-Slipped-Capital-Femoral?autologincheck=redirected
10. https://www.ncbi.nlm.nih.gov/pmc/articles/PMC2686695/
11. https://publications.aap.org/aapbooks/book/659/chapter/7948991/The-Limping-Child-General-Approach-and
12. https://www.orthobullets.com/pediatrics/4119/legg-calve-perthes-disease
13. https://europepmc.org/article/nbk/nbk513230

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