

# **Legg-Calvé-Perthes Disease** **(Avascular Necrosis of Femoral Head)**

## **Background**

### 1. Definition

- Idiopathic osteonecrosis or avascular necrosis of capital femoral epiphysis of femoral head

### 2. General info

- Described independently in 1910 by Legg, Calvé, and Perthes

## **Pathophysiology**

### 1. Pathology of disease

- 4 phases:<sup>1,2</sup>
  - Necrosis
  - Fragmentation
  - Re-ossification
  - Remodeling
- Necrosis
  - Disruption of blood supply leads to infarction of femoral capital physis
    - Affects subchondral cortical bone
  - Ossific nucleus ceases growth
  - Infarcted bone softens/dies
- Fragmentation
  - Dead bone reabsorbed
- Reossification
  - New bone deposited to reestablish femoral head
- Remodeling
  - New femoral head may be enlarged or flattened
  - Reshapes during growth
  - Complete healing usually occurs in 2-4 years

### 2. Incidence, prevalence

- Age: 3-12 years
  - Peak incidence 5-7 years
- Affects 1 in 1200 children under age 15
- Bilateral in 10-20%<sup>3</sup>
- Male: Female ratio of 4:1
- Caucasians, Asians most affected

### 3. Risk factors

- 10% Familial<sup>3</sup>
  - Delayed bone age by ~2 years<sup>4</sup>
- HIV
  - Up to 5% of HIV patients have avascular necrosis of hip<sup>5</sup>
- Factor V Leiden / other inherited coagulopathies<sup>6</sup>
- Thrombophilias
  - Increased clotting<sup>7</sup>

- Hypofibrinolysis
    - Decreased ability to dissolve clots<sup>7</sup>
  - Secondhand smoke exposure<sup>8</sup>
  - Low socio-economic status<sup>9</sup>
  - Birth weight less than 2.5 kg in boys<sup>10</sup>
  - Short stature
4. Morbidity
- Outcome depends on:
    - Age at onset
    - Degree of femoral head involvement
    - Younger age at diagnosis
    - Better outcome
    - Newly formed bone may have residual deformity
    - "Coxa Plana" - flattening of epiphysis
    - Femoral head "mushrooming" around femoral neck
    - Femoral head changes can lead to disability, collapse, early OA of hip

## Diagnosics

### 1. History

- Limp of acute/insidious onset 1-3 months
  - Often painless
- If pain present:
  - Can be localized to hip
  - Referred to knee, thigh, abdomen
- With progression, pain worsens with activity
- No systemic symptoms

### 2. Physical exam

- Decreased internal rotation/abduction of hip
- Pain on rotation referred to anteromedial thigh and/or knee<sup>3</sup>
- Atrophy of thighs/buttocks
  - Due to disuse from pain
- Afebrile<sup>11</sup>
- Leg length discrepancy
- Gait Evaluation-The Limping Child:<sup>12</sup>
  - <http://www.medscape.com/viewarticle/490135?src=mp>
- Acute: antalgic gait: short-stance phase secondary to pain in weight-bearing leg
- Chronic: Trendelenburg gait - downward pelvic tilt away from affected hip during swing phase

### 3. Diagnostic testing

- Demands high index of suspicion
- Labs: used to exclude other diagnoses (CBC, ESR normal in LCP)<sup>11</sup>
- Diagnostic imaging - early radiographs can be normal
- Plain films-initial imaging choice (SOR:C)<sup>13,14</sup>
  - Standard AP Pelvis, Frog-Leg lateral (Lauenstein View)
  - Early findings:
    - Widening of joint space - epiphyseal cartilage hypertrophy

- Changes in epiphysis - smaller, more dense-appearing<sup>11</sup>
- "Crescent sign": subchondral radiolucent zone of anterolateral epiphysis - subchondral fracture
  - Late findings:
    - Flattening of femoral head, fragmentation, healing – sclerosis<sup>11</sup>
- Bone scan:
  - Decreased perfusion to femoral head
- MRI:
  - Marrow changes suggestive of Legg-Calve-Perthes<sup>15</sup>
  - MRI also seems to be superior to bone scan for depicting the extent of involvement in the early/evolutionary stage

### **Differential Diagnosis<sup>16</sup>**

1. Transient Synovitis / Toxic Synovitis
  - Similarities: same age, sex, symptoms
  - Differences: effusion; self-limited
2. Infectious
  - Septic Arthritis
    - Elevated ESR/WBC
    - Fever
    - Erythema
  - Osteomyelitis
    - Systemic symptoms
    - Elevated ESR/WBC
  - Lyme disease
3. Inflammatory
  - Juvenile Rheumatoid Arthritis
    - Elevated ESR/WBC
    - Fever
    - Joint effusion
  - Lupus
4. Slipped Capital Femoral Epiphysis
  - X-ray findings
  - Obese adolescent males
5. Malignancy
  - Bone tumors
  - Blood neoplasias
    - Leukemia
      - Low-grade fever, malaise, weight loss
6. Metabolic
  - Renal disease
  - Hyperparathyroidism
7. Hematologic
  - Sickle-Cell Anemia
    - Symmetric changes
    - Anemia

- African American
  - Family history
8. Developmental Dysplasia of Hip
- Female > Male
  - <5 years old
    - Referred pain
    - Trauma

### Therapeutics

#### 1. Acute treatment

- Non-weight bearing
  - Containment of femoral head within acetabulum<sup>17</sup>
- Strict "bed rest" is not recommended
  - Ambulate with crutches
- NSAIDs
- Referral to pediatric orthopedist
- Physical therapy to maintain range of motion
- Splints and braces
  - Thomas-type splint
  - Atlanta Scottish Rite Brace

### Further Management (days)

1. Containment
2. Brace treatment
3. Spica cast immobilization<sup>18</sup>
  - Reduces force through hip by actual or relative varus positioning<sup>19</sup>
  - Allows for remodeling of epiphysis in more normal shape
4. Without treatment
  - 24% will have spherical femoral head
  - 52% will have irregularly-shaped femoral head<sup>20</sup>
5. Range-of-motion therapy<sup>11,18</sup>
6. Surgery - various procedures recommended

### Long-term Care

1. Follow position of femoral head in relation to acetabulum on x-ray<sup>11</sup>
2. No large controlled trials available
3. Long-term consequences only available after decades

### Prognosis

1. Age at onset<sup>21</sup>
  - Younger age at diagnosis
    - Better outcome
  - <6 years old at diagnosis
    - Normal hip joint
  - >6 years old at diagnosis
    - Pain may continue
    - Arthritis may develop

## 2. Lateral Pillar Classification

- Degree of femoral head involvement:
    - A-least to C-most<sup>18</sup>
  - >8 years old and lateral pillar group B & B/C
    - Do better with surgery than with non-operative treatment
  - <8 years old and group B
    - Do well regardless of treatment choice
  - Group C hips
    - Poor outcomes, regardless of treatment
3. 50% almost fully recover
4. 50% develop pain/disability in their 40s and 50s
- Degenerative joint disease in 60s and 70s
  - Often require hip replacement
5. Females with onset >8 years of age have worse prognosis than males<sup>18</sup>

## Prevention

1. Recommend cessation of smoking for parents
2. Not clearly proven for prevention
3. May be associated with thrombophilia
4. Anti-coagulant therapy for prevention of recurrences not indicated<sup>9</sup>

## Patient Education

1. With appropriate intervention and follow-up, usually good outcomes
2. Websites for Education:
  - Legg-Calve-Perthes Disease
    - [http://www.seattlechildrens.org/child\\_health\\_safety/pdf/flyers/PE247.pdf](http://www.seattlechildrens.org/child_health_safety/pdf/flyers/PE247.pdf)
  - Legg-Calve-Perthes Disease
    - <http://www.chw.org/display/PPF/DocID/22573/router.asp>
  - Perthes Disease
    - <http://orthoinfo.aaos.org/topic.cfm?topic=A00070>

## References

1. Herring JA, Williams JJ, Neustadt JN, et al. Evolution of femoral head deformity during the healing phase of Legg-Calve-Perthes disease. *J Pediatr Orthop.* 1993; 13:41-45.
2. Legg-Calve-Perthes Disease: Patient and Family Education. (2006). Retrieved February 1, 2009 from Seattle Children's Hospital. Website: [http://www.seattlechildrens.org/child\\_health\\_safety/pdf/flyers/PE247.pdf](http://www.seattlechildrens.org/child_health_safety/pdf/flyers/PE247.pdf).
3. Wenger DR, Ward WT, Herring JA. Legg-Calve-Perthes disease. *J Bone Joint Surg Am* 1991; 73:778.
4. Wang NH, Lee FT, Chin LS, Lo WH. Legg-Calve-Perthes disease: clinical analysis of 57 cases. *J Formos Med Assoc.* 1990; 89:764.
5. Gaughan DM, Mofenson LM, Hughes MD, et al. Osteonecrosis of the hip (Legg-Calve-Perthes disease) in human immunodeficiency virus-infected children. *Pediatrics* 2002; 109:E74.
6. Glueck CJ, Crawford A, Roy D, et al. Association of antithrombotic factor deficiencies and hypofibrinolysis with Legg-Perthes disease. *J Bone Joint Surg Am.* 1996; 78:3.

7. Eldridge J, Dilley A, Austin H, et al. The role of protein C, protein S, and resistance to activated protein C in Legg-Perthes disease. *Pediatrics*. 2001; 107:1329-1334.
8. Mata SG, Aicua EA, Ovejero AH, Grande MM. Legg-Calve-Perthes disease and passive smoking. *J Pediatr Orthop*. 2000; 20: 326.
9. Margetts BM, Perry CA, Taylor JF, Dangerfield PH. The incidence and distribution of Legg-Calve-Perthes disease in Liverpool, 1982-95. *Arch Dis Child*. 2001; 84:351.
10. Molloy MK, MacMahon B. Birth weight and Legg-Perthes' disease. *J Bone Joint Surg Am*. 1967; 49:498-506.
11. Adkins SB, Figler RA. Hip Pain in Athletes. *Am Fam Phys*. 1 Apr 2000.
12. Leung AKC, & Lemay JF. The limping child. *J Pediatr Health Care*. 2004; 208:339-343.
13. Fordham L, Gunderman R, et al. Limping Child – Ages 0-5 years. ACR Appropriateness Criteria 2008. American College of Radiology Website. Available [www.acr.org/ac](http://www.acr.org/ac). Accessed March 15, 2009.
14. De Smet AA, Dalinka MK, et al. Avascular Necrosis of the Hip. ACR Appropriateness Criteria 2008. American College of Radiology Website. Available [www.acr.org/ac](http://www.acr.org/ac). Accessed March 15, 2009.
15. Uno A, Hattori T, Noritake K, Suda H. Legg-Calve-Perthes disease in the evolutionary period: comparison of magnetic resonance imaging with bone scintigraphy. *J Pediatr Orthop* 1995; 15:362.
16. Kermond S, Fink M, Graham K, et al. A randomized clinical trial: should the child with transient synovitis of the hip be treated with nonsteroidal anti-inflammatory drugs? *Ann Emerg Med*. 2002; 40:294-299.
17. Weinstein SL. Bristol-Myers Squibb/Simmer award for distinguished achievement in orthopaedic research. Long-term follow-up of pediatric orthopaedic conditions. Natural history and outcomes of treatment. *J Bone Joint Surg Am*. 2000; 82-A: 980.
18. Herring JA, et al. Legg-Calvé-Perthes Disease. Part II: Prospective Multicenter Study on the Effect of Treatment on Outcome. *Journal of Bone and Joint Surgery, Inc*. 2004; 86-A(10):2121-2134.
19. Kamegaya M, Saisu T, Ochiai N, et al. A paired study of Perthes' disease comparing conservative and surgical treatment. *J Bone Joint Surg Br*. 2004; 86:1176-1181.
20. Joseph B, Varghese G, Mulpuri K, et al. Natural evolution of Perthes disease: a study of 610 children under 12 years of age at disease onset. *J Pediatr Orthop*. 2003; 23:590-600.
21. Yrjonen, T. Long-term prognosis of Legg-Calve-Perthes disease: a meta-analysis. *J Pediatr Orthop B*. 1999; 8:169.

**Authors: Heather C. Kompanik, MD, & Kevin E. Burroughs, MD, Cabarrus FMRP, NC**

**Editor: Carol Scott, MD, University of Nevada Reno FPRP**