

legg-calve-perthes disease

for the clinician



the global incidence of Perthes is **5 in 100,000** children



males are affected 4 times as often as females



average age at diagnosis is **4-8 years**, though this statistic is variable



Treatment is variable from **weight bearing restriction** to **surgery**



Perthes progresses in **stages: initial, fragmentation, reossification, healed**



Prognosis depends on shape of femoral head and articulation with acetabulum

Presentation

Due to young age at onset and activity level of children, presentation can be variable.

- unilateral hip, knee, leg pain
- limp or gait abnormalities
- decreased range of motion

History may reveal a very active child, small for age, with no history of specific trauma.

Differential

- osteomyelitis
- pyogenic arthritis
- **transient synovitis**
- juvenile rheumatoid arthritis
- hemophilia
- **slipped capital femoral epiphysis**
- **infection**
- **neoplasm**
- Gaucher's Disease

Work-up

- CBC, ESR
- AP&Lat hip xray with pelvis



Treatment focuses on containing the femoral head in the acetabulum and preserving range of motion. Methods to achieve this vary based on the age of the patient at onset and stage of disease. Younger age at onset is associated with positive outcomes and may warrant conservative treatment such as activity restriction and watchful waiting.

An older child lacks time prior to closure of the epiphyseal plates, thus more aggressive approach may be recommended. Surgical options include osteotomy, tenotomy, and ORIF. All ages and stages require regular imaging surveillance.

Supporting the patient

a medical-surgical team

- PT
- Psychological Counseling
- OT
- Pediatrician
- Nutrition
- Pediatric Orthopedic Surgeon



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Stages

- 1 Initial / necrosis:** blood supply is compromised and osteonecrosis begins. The hip becomes inflamed and the patient may appear symptomatic with pain and a limp. This may last for months.
- 2 Fragmentation:** Osteoclasts are activated and resorption occurs over a period of 1 to 2 years. Proper alignment within the acetabulum further molds the new, woven bone of the femoral head and protects it from collapse.
- 3 Reossification:** Remodeling continues and bone strengthens. This is often the longest stage.
- 4 Healed:** Bone regrowth is complete and the femoral head has reached its final shape. Prognosis can be made based on the shape of the femoral head and articulation with the acetabulum.

What parents say...

“ No one talks about the grieving associated with Perthes. In one moment my son lost his "normal" childhood. He could no longer run onto the field with his team; we were discussing a 504 plan with the school. And the guilt I felt as a parent...did I wait too long; should I have been more persistent; what is the right treatment? It impacted our whole family.”

-LK Texas

“ My daughter was diagnosed at 3. She sometimes complained of her "leg" hurting but she remained active. When her dance teacher mentioned a limp, we went back to the doctor. Eventually she was diagnosed with Perthes in her Rt. hip. That was three years ago. I wasn't prepared for the amount of missed work due to specialist visits or the cost of treatment. Our house is one story but we had to have a ramp built just to get her wheelchair in. Her cast required special pants with snaps. Insurance covered less than expected. My daughter couldn't fit into our sedan in her Petrie casts, so we traded with a friend. Treatment takes creativity and perseverance.”

-CJ GA



Prognosis for children diagnosed with Perthes disease is generally good. Males and those diagnosed prior to age 6-8 are reported to have the most favorable outcomes.

The Stulberg system classifies the shape of the femoral head as round to ovoid to flat based on radiographic appearance and is used to estimate likelihood of potential sequelae such as early onset, severe osteoarthritis and hip replacement.

Q&A

Is Perthes disease genetic?

There has been no inheritance pattern established for Perthes however familial clustering is noted. In these cases Perthes can be linked to a defect in the COL2A1 gene.

Are there pharmaceutical treatments available?

Trials have shown promise in IL-6 inhibitors. The role of bisphosphonates are also being studied.

Encourage your patients to participate in the Perthes Contact Registry at perthes.org/perthes-contact-registry.



PERTHES.ORG