

Legg Calvé Perthes Disease: An Overview for Parents

Introduction:

Legg Calvé Perthes disease (LCPD)—also known as ischemic (avascular) necrosis of the hip—is a condition characterized by a temporary loss of blood supply to the femoral head (top of the femur). The typical course of LCPD is an initial ischemic necrosis of the femoral head (loss of blood supply), followed by collapse and subsequent remodeling (re-shaping to normal sphere shape) **[Figure 1A &B]**. Perthes disease occurs most commonly in boys between the ages of 4-8 years and the overall incidence is approximately 1 in 1200 children. It is bilateral (in both hips) in approximately 10-15% of cases, but typically the hips will show different stages of involvement.



Figure 1A: Left LCPD with early collapse of femoral head

Figure 1B: Bilateral LCPD R>L [note different states of involvement]

What are the Symptoms?

Common symptoms include a painless limp, mild or intermittent pain in the hip, knee, groin, or anterior thigh, a painful limp that is worsened by activity, mild restriction of motion (on abduction and internal rotation), and possible mild quadriceps/thigh weakness or atrophy. Based on the child's history and age, x-rays of the pelvis will usually be obtained which will often pick up the changes in the femoral head that are associated with Perthes disease. Sometimes the changes are too subtle to see on x-ray and an MRI (magnetic resonance imaging) might be used to confirm early avascular necrosis or Perthes disease.

What Causes Perthes Disease?

Although we do not know a direct cause for Perthes disease, there are several known risk factors. Perthes is much more common in boys (approximately 5:1 male-female ratio). There is also an association of Perthes with ADHD (attention deficit hyperactivity disorder) and in children who are, in general, more active than average (running, jumping, sports, etc). There is also an increased incidence in children who are small for their age (delayed bone age). Finally, there have been some recent studies that connect Perthes disease to prolonged exposure to second hand smoke. The prolonged inhalation of cigarette smoke can increase the risk of ischemia

leading to the avascular necrosis/Perthes disease. In addition, the condition is found more often in the Asian, Eskimo, and Caucasian population.

What are the Four Stages of LCPD/Perthes Disease?

- 1.) **Necrosis:** Initial period of ischemia/loss of blood supply to femoral head
- 2.) **Fragmentation:** Re-absorption of bone with femoral head collapse
- 3.) **Re-ossification:** New bone re-grows to reshape the femoral head
- 4.) **Remodeling:** Femoral head reshapes itself into normal spherical shape

What is the Treatment for Perthes Disease?

The overall goal of treatment is to:

- 1.) to reduce hip irritability and pain
- 2.) restore and maintain hip mobility
- 3.) to prevent the femoral head from extruding or collapsing
- 4.) To regain a spherical shape of the femoral head



Once the diagnosis is confirmed (usually via x-rays), a decision is made as to whether any active treatment is needed. The natural history of Perthes is such that many children do well without any treatment and can be treated with observation every 6-12 months. In the past, children with Perthes disease were treated with prolonged casting and immobilization requiring extensive hospitalization. The goal of treatment today is to preserve the roundness of the femoral head and to prevent deformity while the condition runs its course. Prolonged casting and immobilization are usually avoided as it will generally not change the course of the disease.

Instead, the contemporary concept in treating Perthes disease is "containment" of the femoral head in the acetabulum (socket). Containment is a very simple concept. The femoral head can be molded as it heals. This is very similar to molding clay or plastic. The acetabulum (socket) is not affected when the femoral head loses its blood supply. It can be used as a mold to shape the femoral head as it heals. The femoral head must be held in the joint socket as much as possible during the re-ossification and re-modeling phase. It is better if the hip is allowed to move and is not immobilized in one position. Joint motion is necessary for nutrition of the cartilage and for healthy growth of the joint. All treatment options (non-surgical and surgical) for Perthes disease try to position and hold the femoral head in the acetabulum as much as possible.

Overview of Treatment:

- 1.) Observation
- 2.) Physical Therapy/Traction to improve range of motion
- 3.) Surgery: Femoral/Acetabular osteotomy with internal fixation

Most children with Perthes disease are treated with observation. Young children (4-6 years) who are pain-free and have full range of motion are generally observed with follow up visits/intermittent x-rays every 6-12 months. However, if there is stiffness or restriction of motion, other treatment options will likely be discussed or recommended. These include physical therapy, traction, and possible use of anti-inflammatory medication to quiet symptoms. In some cases, surgery might be the best option to obtain adequate containment. Surgical treatment for



containment in Perthes usually consists of procedures that re-align either the femur or the acetabulum or both. Re-alignment of the femur is called a femoral osteotomy. This procedure changes the angle for the femoral neck so that the femoral head points more towards the socket. The bone of the femur is cut and re-aligned and internal fixation (plate/screws) is then inserted to hold the position until the bone has healed (**Figure 2**). The plate and screws may need to be removed once the bone has healed (generally 6-12 months post-op)



Figure 2: 10 year old boy s/p bilateral femoral osteotomies for LCPD

Long-Term Prognosis in LCPD:

The long-term prognosis for Perthes disease is generally quite good—especially for children who develop the condition at a very young age (4-5 years). The younger the child, the more time there is to reshape the affected hip bone. If the femoral head maintains its spherical shape and is contained in the acetabulum, the prognosis for a normal hip is excellent. The two most critical factors that determine the outcome are the child's age at diagnosis and how much of the femoral head is affected. Older children (diagnosis at age 7-10 years) with greater involvement are generally more likely to have limited hip motion, difference in leg lengths, and arthritis in early adulthood. The goals of treatment are to keep the hip pain free, allow full participation in all sports and activities, and prevent or delay the development of early hip degenerative arthritis.

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