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Legg-Calve'-Perthes Disease

The National Osteonecrosis Foundation

Legg-Calvé-Perthes disease is a rare disease of the hip that afflicts approximately 1 in 1200 children. Of those children, only about one in four are girls. About 5% of all diagnosed develop the disease in both hips (bilaterally). Most of these children are very active and often very athletic. The age of diagnosis is usually between 2 and 12 years old, with the average age of 6. Legg-Calve'-Perthes children tend to be of shorter stature due to delayed bone age. The purpose of this pamphlet is to provide you with more information to help you understand this condition and some of the treatments.

What is Legg-Perthes Disease?

Legg-Calvé-Perthes disease (LCPD) is a form of osteonecrosis of the hip that is found only in children. It is known by a few other names such as ischemic necrosis of the hip, coxa plana, osteochondritis and avascular necrosis of the femoral head. Most commonly it is called Legg-Perthes disease, LCPD, or Perthes.

LCPD is of unknown origin. It is known that bone death occurs in the ball of the hip due to an interruption in blood flow. As bone death occurs, the ball develops a fracture of the supporting bone. This fracture signals the beginning of bone reabsorption by the body. As bone is slowly absorbed, it is replaced by new tissue and bone.



Initial Phase



Reabsorption Phase



Reossification Phase/Healed

Four Stages of LCPD

1. Femoral head becomes more dense with possible fracture of supporting bone;
2. Fragmentation and reabsorption of bone;
3. Reossification when new bone has regrown; and
4. Healing, when new bone reshapes.

Phase I takes about 6-2 months, Phase 2 takes one year or more, and Phase 3 and 4 may go on for many years.

Who is at Risk?

There is no specific cause known for LCPD, however, there are some risk factors. Some of the factors identified as possible links include children who are small for their age and are extremely active. The disease is found more often in Asians, Eskimos, and

Whites, with a much lower incidence found in Australian aboriginals, Native American, Polynesians and Blacks. Exposure to secondhand smoke is correlated with LCPD.

First Symptoms

The first symptoms characterized in LCPD are usually a limp and perhaps pain in the hip, groin, or knee (known as a referred pain). Often you will first notice limping during your child's active play. They usually cannot tell you an instance when they hurt themselves. They may not be able to tell you exactly where they hurt, especially if the pain is referred toward the knee area. They may not even experience much pain. Other cases may not be diagnosed until some precipitating event (fall, twisting injury) leads to an x-ray that uncovers the previously undiagnosed Legg-Calve'-Perthes disease.

Diagnosis

Initial diagnosis will require an x-ray, magnetic resonance imaging (MRI) or bone scan. Other diagnostic measures may include tests for limitation of abduction, a measurement of the thigh to determine muscle atrophy, and tests to determine the child's range of motion.

Extent of Disease

It is rare for a patient to have whole head involvement. However, age can play an important role in the prognosis of the disease. New bone growth typically reshapes better in younger children and it may improve with growth.

There are several different classifications used to determine severity of disease and prognosis.

The **Catteral Classification** specifies four different groups to define radiographic appearance during the period of greatest bone loss.

The **Salter-Thomson Classification** simplifies the Catteral Classifications by reducing them down to two groups: Group A (Catteral I, II) which shows that less than 50% of the ball is involved, and Group B (Catteral III, IV) where more than 50% of the ball is involved. Both classifications share the view that if less than 50% of the ball is involved, the prognosis is good,

while more than 50% involvement indicates a potentially poor prognosis.

The **Herring Classification** studies the integrity of the lateral pillar of the ball. In the Lateral Pillar Group A, there is no loss of height in the lateral 1/3 of the head and little density change. In Lateral Pillar Group B, there is lucency and loss of height of less than 50% of the lateral height. Sometimes the ball is beginning to extrude the socket. In Lateral Pillar Group C, there is more than 50% loss of lateral height.

Many doctors utilize these classifications as they provide an accurate method of determining prognosis and help in determining the appropriate form of treatment.

Prevention

There is no known effective preventative measure.

TREATMENT The Goal

The goal of treatment is four-fold:

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

Types of Treatment

Often at the initial diagnosis, the physician may take a "wait and see" approach to get a clearer picture of the progression of the disease. As long as the patient's symptoms are mild, the physician may only prescribe physical therapy exercises to help maintain good range of motion. If the patient's mobility changes, then the physician may prescribe either non-surgical or surgical treatment.

Non-Surgical Treatment

Non-surgical treatments come in varying forms. Crutches are used for non-weight bearing treatment for pain. Casts, traction, and braces help return range of motion and mobility. Range of motion exercises may be given to you by your physical therapist to do with your child in the home.

Surgical Treatment

Tenotomy

A "Tenotomy" is a surgery that is performed to release an atrophied muscle that has shortened due to limping. Once released, a cast is applied allowing the muscle to regrow to a more natural length. Cast time is about six to eight weeks.

Osteotomy

There are different types of "osteotomies" (cutting the bone to reposition it) and, depending on the need they are performed at different stages of the disease. At times with the softening of the ball, there is the possibility of the ball slipping out of the socket. To protect it, a femoral varus osteotomy, with or without rotation partially redirects the ball into the socket.

Another approach to surgically treating Legg-Calvé-Perthes is to do an osteotomy above the hip socket. This allows the surgeon to reposition the hip socket in such a way that the femoral head will have less tendency to become deformed. The shelf arthroplasty gives added coverage of the ball from the top lip of the socket. Both the innominate and the shelf arthroplasty help in reshaping.

LOOKING TO THE FUTURE

Studies on long-term results of LCPD indicate that the incidence of late degenerative osteoarthritis is dependent on two factors. If the ball reshapes well and fits well in the socket, arthritis is usually not a concern. If the ball does not reshape well, but the socket's shape still conforms to the ball, the patient will tend to develop mild arthritis in later adulthood. Patients who~ femoral head does not shape well and does not fit well in the socket usually develop degenerative arthritis before the age of 50.

Although Legg-Calvé-Perthes disease cannot be prevented, much has been accomplished toward minimizing its effects. Research and clinical studies continue to provide patients with better long-term results.

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