

A GUIDE TO PERTHES DISEASE

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What Is It?

In 1910, Drs. Legg, Calve, and Perthes independently described a disorder of the hip in young children. Many theories have been proposed as to the cause including inflammation, infection, trauma, and even “hip problems” noted at birth. The most popular theory at the present time is that for some unknown reason, there is a temporary loss of the blood supply to the femoral head (ball portion of the hip joint).

In the growing child, the two major components of the hip joint are the ball (femoral head) and the socket (acetabulum). These two structures are pictured in figure (1).

The femoral head in the growing child has several distinct zones, figure (2). The metaphysis is the spongy bone area in the femoral neck or that portion of bone next to the femoral head. The physis is the growth center of the femur (thigh bone). It is composed of bone forming cells that enable the bone to grow in length. The epiphysis is located between the physis and the acetabulum (socket), and it actively contributes to the growth of the femoral head (ball portion). As stated earlier, for some unknown reason, there is a temporary interruption in the blood supply to the epiphyseal, physeal, and sometimes metaphyseal portion of the femur of the hip joint. This is shown by the stippled area in figure (3). When this happens, a part or all of the area involved temporarily dies. It is important to understand that the acetabulum (socket) is not affected or involved in this loss of blood supply.

Who Is Affected?

Perthes disease most commonly is seen in the 4-8 year age group, but children age 2 years up to teenagers, can be involved. Boys are involved more than girls by a ratio of 4 or 5 to 1. Perthes can involve both hips in 10-12% of the youngsters but does not occur simultaneously at the exact same stage. Reports exist showing positive family histories (up to 20%), but it has never been proven to be inherited. Generally speaking, children with Perthes disease tend to be shorter in stature than average.

How Is It Detected?

Children with Perthes disease most commonly have a limp that has been slow and intermittent in its appearance. Pain is not usually a major factor, and often seems more activity related. Because of slow onset and minimal pain, if any, it may be months before the parents seek help in determining the cause of the problem. Often, children will complain of knee pain, when the problem is located in the hip. Initially, the doctor, during his examination, will find a child that limps, has loss of motion about the hip, and in cases of longer duration, evidence of muscle wasting or atrophy about the involved hip and thigh.

The Radiograph

The radiograph (or X-ray) of the involved hip is key in diagnosing Perthes disease. With today's modern equipment and appropriate shielding, risk from radiation is at a minimum and unfortunately, the radiograph is the only way to detect and monitor stages and progress of Perthes disease.

There are basically four stages of this disease, each having its unique characteristics on the radiograph.

The first or **initial phase** is characterized by a smaller femoral head and apparent widening of the hip joint. Usually, the femoral head appears whiter (or more dense) than the normal hip.

The second or **fragmentation phase** actually results from early return of the blood supply to the involved area. The dense femoral head appears to be separating into pieces or look “moth eaten” as new blood vessels form.

The third or **reossification phase** is marked by new bone formation as the healing commences. The femoral head starts to take on a new appearance and give some insight as to the final shape it will have.

The fourth or **healed phase** is just what it says. The repair process has been completed and the radiograph will show you any residual deformity in the femoral head and neck.

It is important to understand that every affected child goes through each phase, from beginning to end. This whole process can extend over many months to years.

FOUR STAGES OF THE DISEASE

1. **initial**
2. **fragmentation**
3. **reossification**
4. **healed**

****note- the above 5 lines need to be displayed in a small box with a border around the description of the four stages****

Important Factors – Outcome

Probably the most important factor influencing the end result is the age of the child at the onset of the disease. Current studies use seven years of age as the cut-off. The younger they are, the better they do because more growth remains to help reshape and remodel the involved areas. Females with Perthes disease do not do as well as males once the disease has healed – reason unknown.

Another very important factor influencing the outcome relates to how much of the physis (growth center) loses its blood supply. Studies have shown that when the major portion of the physis is involved, the disease process is prolonged and the end result poor. At other times, the physis can be permanently damaged (i.e., stops growing) due to lack of blood supply; this directly and permanently alters the shape of the femoral head once healing has completed.

Quite often, due to delay in diagnosis, the child will already be in the healing or reossification phase. Once this point in time has been reached, treatment can no longer affect the eventual end result.

What Can Be Done – Treatment?

Drs. Stulber and Salter did a long term study to determine what factors are important relative to the final outcome or end result. They found that the ***four major factors of significance*** are:

1. The child’s age at onset.
2. The extent of involvement of the femoral head (i.e., percent losing its blood supply)
3. The ability to keep the femoral head in the acetabulum (ball in the socket) during the disease process – also called containment.
4. Loss of motion of the hip joint.

It’s quite obvious that the physician has no control over the age of onset or the extent of blood supply. This leaves the latter two problems to concentrate on in the management or treatment of the disease.

Initial treatment focuses on trying to decrease the inflammation, pain, and muscle spasm about the hip. Therapies such as Ibuprofen and rest are started, the latter being difficult to control in the young child. Re-evaluation at scheduled intervals allows the physician to insure this is being accomplished. It is important to know that good hip motion and containment

(keeping the ball in the socket) go hand in hand. The major reason for this emphasis is that the acetabulum (not involved in Perthes disease) acts as a template or mold for the deformed femoral head to help influence its shape as healing progresses. Physical therapy may be used in those youngsters who do not regain satisfactory hip motion.

If these goals are not reached and maintained, further measures may be necessary. Occasionally, the child may have to be admitted to the hospital, taken to the Operating Room and have an arthrogram (dye injected into the hip joint) to determine the shape of the femoral head and decide whether or not the femoral head is containable. Once this is decided, a brace is used to maintain containment. Plaster casts are often used temporarily while brace is being manufactured. Surgery may eventually be necessary in the more difficult cases but this decision is made much later in the treatment process.

Eventual Outcome – End Result

In addition to understanding the basics of Perthes disease, parents are anxious to know what effect Perthes will have on their child in future years. Many long term studies have now been completed and it is the overwhelming consensus that thirty to forty years after the onset, as high as 90% of the children are active and pain free. This means they will enjoy the ability to keep up with their friends as they progress through immediate and future years. All of this, in spite of the fact that most hips do not return to normal shape, once completely healed.

****Note- logo for Children's Orthopaedics of Atlanta, P.C. is displayed on bottom right side of the page****