

Legg Calve Perthe's Disease

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The femoral head in a child undergoes many changes as it grows and develops. Along with the many changes are alterations in the vascular supply to the femoral head. During certain periods of rapid growth, the femoral head is more vulnerable to ischemia. When the blood flow is interrupted, the bone "dies". This event leads to avascular necrosis (AVN) of the femoral head. This is commonly called Legg Calve Perthes Disease (named after the three doctors who described the condition nearly simultaneously and independently in 1910). Avascular necrosis of the femoral head differs from the AVN in adults by the fact that in the young there is possibility through growth for the complete restoration of the femoral head. In adults, once the femoral head becomes avascular it would surely lead to bony collapse and deformity.

The etiology for this ischemic event is probably multifactorial and still unclear. It is also unclear why it is most prevalent between the age of 4 to 8 years. It has been noted that the incidence is higher among boys than girls (4;10). Around 10% can involve both sides. These children are more likely to be of short stature. Their bone age is delayed compared to their chronological age. The area of involvement can cover the entire femoral head or just part of it. The avascular bone would over time become fragmented. By creeping substitution, the avascular bone is replaced by new bone. This process of repair and remodeling occurs over a long period of time with huge variation. Typically at least 2 years is needed. The final shape of the reconstituted or remodeled femoral head would be dictated by many factors, including: extent of bone involvement; +/- extrusion of femoral head from acetabulum or "containment" of femoral head in acetabulum; hip movement retained during process of healing; age of disease onset; and +/- premature closure of physal plate.

The typical initial presentation is a painful limp or the so-called antalgic gait. The child would be seen favoring the painful limb. One would note that there is avoidance of putting weight on the said limb. During walking, the time spent on standing on one leg is called the stance phase while the time spent with the leg off the floor is called the swing phase. In short, this antalgic gait can be best described as shortening the stance phase of the painful limb. In many cases, they would just complain of inguinal pain upon waking up and exhibit antalgic gait. There are no precipitating incident or factor. Commonly, no other systemic symptom is experienced.

At the initial onset, the disease entities that are needed to be ruled out are Septic Hip Arthritis and the benign Transient Synovitis. Septic arthritis is frequently accompanied by other systemic symptoms like fever. The pain is described to be more severe, hip movement is extremely limited with hip in a flexed position and the child would completely refuse to stand or walk. Laboratory exams for inflammatory markers like CBC, ESR and CRP are elevated. Transient synovitis on the other hand would present just like Legg Calve Perthes (LCP). The course though differs. After a few days of rest, a child with transient synovitis will recover fully. In LCP, there would be improvement with rest, but the symptoms of pain and limp will recur and persist. The initial similarity is due to the fact that the first stage in LCP is also synovitis. Upon examination, the hip joint will be found to have limited range of motion, particularly hip abduction and internal rotation.

X-rays are most helpful in diagnosing LCP. The x-ray views requested are AP and frog leg lateral. During the initial stage, nothing can be seen on x-ray except maybe for a thin lucent line

("crescent sign") underneath the subchondral bone of the femoral head. This signifies the area of ischemic involvement. Over time, the involved bone segment becomes sclerotic and fragmented. During the next stage of LCP, these fragments become more lucent as new bone replaces the dead bone. Later the epiphyses reforms and remodels. The x-rays do not just show the stage of the disease but also the extent of bone involvement. The femoral neck would exhibit varying amount of cyst formation and the orientation of the physes can also be shown. The x-rays can also show whether the femoral head is being extruded out or subluxating. During the initial stage when there are still no significant x-ray findings, an MRI is helpful. It can show the area of ischemia.

For prognostication and management purposes, LCP is classified using the x-rays. The amount and area of involvement (eg. Catterall Classification) are important indicators of outcome. A global involvement of the femoral head points to very poor outcome. LCP can also be classified as to the amount of lateral pillar affected (Herring's Lateral Pillar Classification). The femoral head can be divided into central, medial and lateral pillars. The most critical in LCP is the amount of involvement or collapse of the most lateral pillar. An intact lateral pillar acts as a weight-bearing support to the central avascular segment. The lesser the Involvement, the better. The outcome is expected to be poor if 50% or more of the lateral pillar are involved.

Going back to the list of factors governing the outcome, we can see that there are only two factors which the surgeon can have any influence. They are the range of hip motion and 'containment' of the hip. Thus, treatment is geared towards these factors. A child who is limping and complaining of hip pain is put to rest. Physical therapy is instituted to regain lost hip motion. The femoral head is maintained inside the acetabulum or 'contained' by all means. The weak and fragmented femoral can remodel to the shape of the acetabulum if the femoral head remains contained all throughout the healing period. Containment strategies may take the form of physical therapy, traction, bilateral long leg cast bridged in hip abduction (Petrie cast) or by osteotomies of either the pelvis or the femur.

The eventual outcome is varied. The younger the onset, the better results are expected. The femoral head may regain good sphericity though generally become bigger or coxa magna. The femoral head may become flat or coxa plana or the shape may become irregular or coxa irregularis. It goes without saying that the result is much better if the head is spherical than if it is not. Another factor to consider also is congruency of the femoral head to the acetabulum. The best outcome is a spherical head congruent to the acetabulum. The worst outcome would be the incongruent fit of an irregular head. Early arthritis is the result.

References:

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