



POSNA

The Core Curriculum

Legg-Calvé-Perthes syndrome

Objectives

1. Describe the blood supply to the immature hip
2. Describe symptoms, signs, and age of onset of Legg-Calvé-Perthes syndrome
3. Describe the 4 stages of Legg-Calvé-Perthes syndrome
4. Discuss classification systems for Legg-Calvé-Perthes syndrome, including reproducibility and ease of clinical usage
5. Describe "head at risk"
6. Discuss the present treatment choices for Legg-Calvé-Perthes syndrome
7. Discuss the effect, if any, of treatment on the natural history of Legg-Calvé-Perthes syndrome

Discussion points

1. What are current theories of etiology of Legg-Calvé-Perthes syndrome?
2. Why call it a syndrome and not a disease?
3. What is Gage's sign?

Discussion

In 1910, Legg, Calvé, Perthes, and Waldenstrom independently published a description of what we now call Legg-Calvé-Perthes syndrome (LCPS). They all had different ideas about what they saw, and that basically set the tone for the rest of the twentieth century. We still have a very limited concept of etiology, uncertainties about the pathophysiology, a better concept of the natural history, and a modest understanding of the effectiveness of treatment. Chung and Trueta both noted the more tenuous blood supply to the anterior half of the proximal femoral epiphysis, and the anterolateral quadrant is most vulnerable to vascular embarrassment. Present thought is centered around mechanisms of disruption of this blood supply, more likely repeated insults than one major event. Various treatment programs including prolonged bedrest, a variety of braces, and a variety of surgical procedures have been promulgated for treatment, but especially over the last 20 years, there have been more systematic investigation of treatment results. Much effort has been expended trying to devise a method to quantify results. It is important to remember that there is a limited relationship of radiographic stages to the timing of the onset of symptoms. Classification systems to better evaluate treatment suffer from the lack of uniform positioning for radiographs, and the subjectivity of the classifications. About 30 years ago, Catterall proposed dividing cases of LCPS into 4 groups, primarily based on the amount of the femoral head undergoing necrosis. Since this could often be hard to ascertain at the outset, the classification could change during treatment. Salter and Thompson reduced the classification to two possibilities, either more than half or less

than half of the bony epiphysis undergoing necrosis. Herring focused on the lateral portion of the femoral head, called the lateral pillar. There are three choices - the lateral pillar is intact, it is sustained some collapse which is less than 50% of the original height, or greater than 50% collapse. Collapse of the lateral pillar has a more clear relationship to outcome than the percentage of head involvement, and there is more interobserver agreement than with the Catterall. Many investigators judge outcome by the Stulberg method, but the subjectivity of this method also has been documented by a recent thoughtful study from Iowa.

Just about every possible idea has been floated as a possible etiology of LCPS. Two possible causative factors are presently being intensely studied, passive smoking, and thrombophilia, although this idea has not been corroborated by any group except the originators of the concept. The increased incidence of LCPS in lower socioeconomic groups has been a recurrent theme, but this is not an etiology, just an observation. The list of proposed etiologies is very long.

Presenting symptoms include anterior hip, thigh, and/or knee pain, or a painless limp. Careful examination will always reveal loss of motion, particularly in rotation and abduction, and thigh atrophy.

The clinical course of LCPS has traditionally been divided into 4 stages - initial, fragmentation, reossification, and healed. There is generally less disagreement about the stage of the process than the pattern and quantity of necrosis of the femoral epiphysis. Catterall also described "head at risk" radiographic signs which appear to have some connection with a poorer prognosis. Head at risk signs are 1) Gage's sign, a radiolucent defect between the lateral epiphysis and metaphysis 2) calcification lateral to the epiphysis 3) metaphyseal cysts 4) lateral subluxation and 5) horizontal growth plate. Taken collectively, these signs indicate a more severe injury to the physis.

With our present concepts, we try to assess the severity of the vascular insult, noting whether the critical lateral portion of the femoral head has maintained height. If less than half the head is necrotic, and the lateral pillar is intact, no treatment is necessary, as it would not improve on the natural course of the condition. If more than half the head is necrotic, and the lateral pillar is collapse, some attempt to alter the natural history appears warranted. Simply maintaining a range of motion aids tremendously, imagine the shape of clay rolled back and forth between your two hands and the different shape of clay rolled in a circular motion. If we believe the femoral head is biologically plastic, range of motion is a mainstay of effective management. Bracing is less popular now than at any time during the last half century, end results of bracing appear to offer no improvement over the natural history. Surgery on the femoral or pelvic sides both have their advocates. The rationale is to reduce the laterally subluxated portion of the femoral head to better distribute joint forces. This is often called "containment". If the femoral head is in the acetabulum, there is a good range of motion, and there is sufficient growth remaining to allow some remodeling, the outlook should be good. Girls are said to have a worse prognosis, this could be related to their earlier skeletal maturity with less time remaining for remodeling. Caxa magna is inevitable following any more than minimal LCPS. Long-term studies indicate that hips affected with LCPS perform reasonably well until the fifth decade of life.

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