Abstract

Legg-Calve Perthes is a hip disorder in children due to an interruption of blood supply to the head of the femur causing it to deteriorate. Due to the complexity of the disease, there are several imaging modalities used to aid physicians in assessing the severity of the disease to provide guidance to therapy. Those affected by LCP could have a good or poor prognosis in future years depending on how old the child was when diagnosed, how they were treated, and the progression of the disease.
Introduction

Legg-Calve Perthes (LCP) is a disease process that causes avascular necrosis of the femoral head affecting children from 3 to 11 years of age. LCP occurs most commonly in boys than girls by a ratio of 4 to 1. Children diagnosed with LCP need both clinical and radiological attention to administer proper treatment. Depending on the age of onset and severity of disease progression, the prognosis in patients with LCP could be good or bad. Those with a more severe form of LCP may develop early osteoarthritis leading to hip replacement surgery.

Literature Review

Clinical Presentation

Children with suspected LCP complain of pain in the affected hip. The pain is generally experienced in the groin following the obturator nerve’s innervations area. The pain, in the affected hip, increases upon physical exertion. Symptoms experienced by those with LCP include walking with a limp, and limited range of motion of the affected hip particularly abduction and medial rotation.

Imaging

Radiological examination plays an important role in establishing the diagnosis, evolution, and prognosis of the patient. Imaging LCP to assess disease progression is imperative to help physicians administer proper therapy. The imaging modalities used to diagnose LCP are: plain radiographs, Magnetic Resonance Imaging (MRI), Arthrography, Computed Tomography (CT), and Ultrasonography (US).

According to Dimeglio and Canavese (2011) “It is generally accepted that it is best to initiate imaging of LCP disease with anteroposterior (AP) and frog lateral plain radiographs, and to image both hips to detect any contralateral disease” (p.297). Diagnostic x-rays are a less
expensive form of imaging therefore plain x-rays are considered to be a first choice test that assists physicians to initially determine if LCP is the cause of the patient’s hip pain.

Magnetic Resonance (MR) imaging is extremely useful in the initial diagnosis of LCP along with assessing disease progression and planning therapy. According to Dwek (2009) “MR imaging has a unique advantage as an imaging tool because it is able to image both the ossified and nonossified structures, such as the labrum, in a single examination and without exposure to ionizing radiation” (p.997).

MR scans provide good anatomic images of the cartilaginous femoral head including its shape. Also, MR has been shown to be more accurate in evaluating the extent of epiphyseal necrosis. MR can be used to stage the hip and identify when the revascularization period begins. MR imaging with intravenous (IV) contrast enhancement is capable of identifying ischemia to the femoral head. If interruption of the blood supply has occurred, the portion of the head that is ischemic does not enhance and it remains dark on T1 fat-saturated sequences. MR provides images in three different planes; coronal, sagittal, and axial. The coronal plane is best for comparing one hip with the other. The sagittal plane is most accurate at evaluating the volume of necrotic bone. The axial plane is useful in examination of the surrounding structures of the femoral head.

Arthrography is unique because it allows the physician to evaluate mobility of the hip under direct vision using fluoroscopy. Arthrography helps to identify the best position for femoral head containment and to demonstrate absence of hinge abduction prior to containment surgery if necessary. During the late stages of remodeling, arthrography may be used to determine if non-united fragments of bone are free or covered by cartilage.
CT scans allow early diagnosis of bone collapse and subtle changes in the bone trabecular pattern. CT can also identify intraosseous cysts in later stages of LCP disease. CT gives precise information concerning the femoral head and acetabulum relationship, along with showing a three dimensional nature of the femoral head deformity. However, the use of CT is limited because of high radiation dose to the patient.

US is primarily used to provide clues to diagnose early LCP disease by showing continuous effusion of the hip. US also can demonstrate flattening of the femoral head and deformation of the bony epiphysis without exposing the patient to ionizing radiation.

**Phases of LCP**

The three phases of LCP disease progression are avascular necrosis, fragmentation, and the healing phase. The avascular necrosis phase is characterized by the femoral head not receiving adequate blood supply causing deterioration of bone. Fragmentation phase is the aftermath of the avascular necrosis phase where pieces of bone break away from the femoral head epiphysis. The healing phase consists of reabsorbed fragmented bone being replaced with new bone growth.

**Prognosis**

The prognosis in LCP disease varies considerably according to risk factors such as age, the amount of femoral head involvement, and femoral head cover. According to Albers et al. (2012) “Patients after Legg-Calve Perthes disease often develop pain, impaired range of motion, abductor weakness, and progression of osteoarthritis in early adulthood” (p.2450).

To distinguish the severity of LCP, the disease is classified in four groups named after Catteral. In Catteral groups I-II less than half of the femoral head is diseased. In Catteral groups I-II it is possible to have full anatomical and functional recovery with the femoral head regaining
spherical form. In Catteral groups III-IV more than half of the femoral head is diseased. Catteral groups III-IV are severe forms of LCP which lead to severe deformations including femoral head flattening and slightly external subluxation with poor prognosis which may lead to early osteoarthritis and eventual hip replacement. According to Orban and Razvan (2007) “The starting age has an important role in prognosis. Therefore, the younger the patient, the more favorable the prognosis” (p.138).

Treatment

A child who is diagnosed with LCP regardless of the extent of epiphyseal involvement should receive treatment. Treatment must be started early within the necrosis or fragmentation phases and maintained during the whole evolution period.

The basic principle is that of containment. Containment can prevent deformities of the diseased epiphysis. The main goal is relief of weight bearing on the affected femoral head and preventing its extrusion. To equalize the pressure on the head and subject it to the molding action of the acetabulum, the femoral head must be contained within the acetabulum. The final goal is to have a spherical femoral head at the time of healing.

Radiographs should be taken every 3 to 4 months to follow the radiographic stages and the degree of disease progression. If the disease has already progressed to the reossification stage, it may be too late for treatment to be beneficial. A long term outcome for patients with a more severe form of LCP is a standard total hip replacement. As stated by Traina et al. (2011):

The results of the present study allow us to recommend standard total hip replacement for the treatment of osteoarthritis of the hip secondary to Legg-Calve Perthes disease if careful preoperative planning is performed to overcome the technical pitfalls related to the abnormal proximal femoral and acetabular anatomy of these patients (p.6).
Studies on LCP Patients

A study on LCP was conducted in Norway by Terjesen, Wiig, and Svenningsen (2010) over a period of 5 years. The study involved 212 children (mean age 5.1 years, 77% boys) who were affected unilaterally and who had been treated with physiotherapy, which consisted of range of motion exercises and muscle strengthening exercises. They were followed by taking radiographs at the time of diagnosis and after 5 years.

The radiographic outcome at the 5-year follow-up was good (i.e. spherical femoral head) in 114 patients (54%), fair (ovoid femoral head) in 60 children (28%), and poor (flat femoral head) in 38 children (18%). There was a strong relationship between radiographic outcome and Catteral group as no hips with <50% femoral head necrosis had a poor outcome whereas 37% of those with total necrosis had a poor result.

According to Terjesen, Wiig, and Svenningsen (2010) “There was a clear association between the radiographic outcome and the age of the patients at the time of diagnosis. Younger children had better results than older children, no matter whether the age was 5, 6, or 7 years” (p.711). The average age of patients with a poor result after the 5-year radiographic outcome was 6.1 years and those with a good result had a mean age of 4.8 years.

In the most severe forms of Perthes disease with Catteral groups III-IV risk factors, containment surgery seems advisable. This means that the great majority of the children in the present study had an adequate non-containment treatment, whereas those with the most severe form of the disease and relatively older age would probably have had a better outcome if containment surgery had been performed.

In another study by Dimeglio and Canavese (2008) findings of LCP in children under 6 years of age were reported. The study consisted of 166 patients who were affected unilaterally.
The mean age at onset of the disease was between 3-4 years of age. Mild forms (Catteral grades I-II) were treated conservatively, and more severe forms (Catteral grades III-IV) were treated conservatively or operatively.

All the patients were followed to skeletal maturity with a mean follow-up of 11 years (8 to 15). A total of 50 hips were Catteral grades I-II, 65 Catteral grade III and 51 Catteral grade IV. All hips with mild disease had a good result at skeletal maturity. In Catteral grade III hips, 38 were treated conservatively of which 31 (81.6%) had a good result, 6 (15.8%) a fair result, and 1 (2.6%) had a poor result.

Operative treatment was carried out on 27 Catteral grade III hips resulting in 21 (77.8%) good, 4 (14.8%) fair, and 2 (7.4%) poor. By comparison conservative treatment of 19 Catteral grade IV hips led to 10 (52.7%) good, 7 (36.8%) fair, and 2 (10.5%) poor results. Operative treatment was carried out on 32 Catteral grade IV hips resulting in 16 (50%) good, 9 (28.1%) fair, and 7 (21.9%) poor.

It was confirmed that the prognosis in Perthes disease is generally good when the age at onset is less than 6 years. It was also concluded that in severe disease there is no significant difference in outcome after conservative or operative treatment. According to Canavese and Dimeglio (2008) “Children presenting with Perthes disease before their sixth birthday are considered to have a good prognosis” (p.940)

A study focusing on the long term prognosis for patients with LCP who have had little or no treatment was conducted by physicians at the Texas Scottish Right Hospital for Children. Fifty six patients were followed over the course of 20 years to determine the functional and radiographic outcome of the affected hip.
Classic teaching in regard to the clinical outcome of patients with LCP has been that they do well until the fifth or sixth decade of life before experiencing a decline in their hip function. According to Larson et al. (2012) “Good long-term functional outcomes have been reported despite persistent radiographic abnormalities. However, clinical experience commonly reveals a subset of patients in their second and third decades of life with substantial dysfunction and pain” (p.585). Of the 56 patients studied, there was a high prevalence of arthritis and low clinical outcome scores after 20 years of follow-up.

Case Report

The patient is a 25 year old Caucasian male who was diagnosed with LCP at three years of age. The patient’s parents became concerned because he couldn’t walk without pain. Between the ages of 3 and 7, he periodically had x-rays taken to assess the progression of the diseased femoral head. During this time of disease progression the patient underwent physiotherapy. He was encouraged to keep the blood flow moving in both hips by lying down on his back and pretending to pedal a bicycle for 10 minutes every morning and night. Also, the patient was told to avoid climbing stairs, jumping, and playing contact sports.

The LCP disease initially affected the right femoral head at the age of 3, and by the age of 5 to 6 the disease expressed itself in the left femoral head. Symptoms in his childhood include limping, aching, pain, and limited range of motion. The patient specifically remembered having to have an excuse from his physician because he was not able to sit with his legs crossed on the floor for reading time.

Now, at the age of 25, the patient still suffers chronic aching, limited range of motion, and pain of the left hip when running, hiking, walking, or standing for long periods of time. Interestingly, the right hip, which had the disease first at the onset of three years of age, does not
give him pain at the present time. Along with these symptoms, the patient also experiences chronic lower back pain due to favoring his weak hip joints. To help prevent daily aching of the left hip, the patient has a special pillow shaped to fit between his legs that he sleeps with. The patient has been able to lead an active and normal lifestyle with the exception of not being able to play contact sports in high school or riding a horse on the farm.

To detect LCP, AP pelvis and frog leg plain radiographic views were taken. The radiographs documented the patient’s disease progression from ages 3, 4, 7, and 25 years of age. A radiologist after reviewing the images said that “from the ages 3 to 7 there is developing fragmentation of the right femoral head with loss of epiphyseal height” (personal communication, October 26, 2012). (See Figure 1-6) He then continued “the current x-rays, 20 years later, at the age of 25 demonstrate flattening and remodeling of the femoral heads with secondary acetabular remodeling”. (See Figure 7-8).

When the patient reached middle school age, his physician highly recommended that the patient not play football because there was too much risk of fracturing or dislocating a hip. The physician told the patient that if he were to fracture his hip, the consequences could be stunted growth and extensive surgery because of the abnormality of the ball and socket joint. The physician also told the patient that he should try to get all the growth and height he could because he may need hip replacements at the age of 18. Now at the age of 25, the radiologist suspects early osteoarthritis with multiple hip replacements in the future. The recommendation is that the patient should hold off getting hip replacements as long as can be tolerated, because the life span of an initial hip replacement is 10 years with each subsequent replacement having a decreased life span. From the current hip x-rays spurring on the inferior vertebral bodies of L4-L5 is due to the patient favoring his hips causing more stress to his back.
Discussion

In conclusion, LCP disease is a hip disorder in children due to a lack of blood flow to the affected hip causing osteonecrosis of the femoral head epiphysis. The typical presentation of a child with LCP is a limping child who refers pain to the hip and limitation of range of motion. Physicians utilize medical imaging to aid in the diagnosis and treatment of LCP. Plain radiographs, MRI, Arthrography, CT, and US are used to assess the severity of the disease, and to provide guidance to therapy. The Physician determines, with the help of imaging, which of the three phases of the disease the patient is experiencing. Treatment options vary depending on whether the patient is in the avascular necrosis, fragmentation, or healing phase. Depending on the severity of the disease, the patient may require containment surgery for an optimal outcome. The earlier the onset of the disease, the better the outcome for the patient. Patients with LCP who are in their second or third decade of life who are treated nonoperatively usually experience femoroacetabular impingement, instability, labral disease, and early osteoarthritis. A total hip replacement has proven to be a feasible option for patients suffering osteoarthritis of the hip as a result of LCP disease.
Figures

**Figure 1.** AP pelvis view of a three year old male showing developing fragmentation of the right femoral head with loss of epiphyseal height.

**Figure 2.** Frog lateral image of bilateral hips on the same patient.
**Figure 3.** AP right hip view of the same patient at the age of 4. This image also demonstrates developing fragmentation of the right femoral head with loss of epiphyseal height. Note the fragmented pieces of bone above where the epiphyseal plate should be.

**Figure 4.** Frog lateral of the right hip on the same patient.
Figure 5. AP pelvis view of the same patient at 7 years of age. This view also shows developing fragmentation of the right femoral head with loss of epiphyseal height.

Figure 6. Frog lateral bilateral hips view on the same patient.
Figure 7. AP pelvis view on the same patient at 25 years of age. This view shows flattening and remodeling of the femoral heads with secondary acetabular remodeling.

Figure 8. Frog lateral views of bilateral hips on the same patient.
References


