Evolution in diagnosis and treatment of Legg-Calve-Perthes disease

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Abstract

Legg-Calvé-Perthes disease (LCPD) is an idiopathic osteonecrosis of the femoral head with variable complications and resultant deformity of the femoral head and osteoarthritis. Suggested risk factors are acetabular retroversion, obesity, latitude, hyperactivity, and coagulopathy. The most commonly applied classification is based on radiolucency in the lateral pillar of the femoral head, which is strongly correlated with the outcome. In the fragmentation stage of the disease involvement can be classified into three groups. After skeletal maturity, the outcome can be classified using the Stulberg classification based on the sphericity and congruence of the femoral head in relation to the acetabulum. The early goal of treatment is to prevent head deformation by weight transmitted forces during remodeling and ossification stages containment is the widely accepted principle of treatment. Although the Petrie cast still has a role in the short-term treatment of LCPD before and during the reossification stage, available data does not support additional benefits from braces during the course of LCPD.

Key words: Brace, Lateral Pillar, Legg-Calvé-Perthes, Osteotomy, Stulberg classification

Introduction

Over one hundred years have pass since the first three reports on Legg-Calvé-Perthes disease (LCPD) were published. It is a disease mostly affecting children between 2 to 12 years old. The interruption in femoral head blood supply is attributed to its formation and progression. It consequently leads to changes in the femoral head, metaphysis, growth plate, and acetabulum. One early presenting sign is the subluxation and lateral displacement of the femoral head out of the acetabulum (1). The femoral epiphysis is soft and susceptible to deformity by loading. Lateral migration will lead to femoral head deformity because of resting on the edge of the acetabulum and uneven transmitting of the loading force (2).

Etiology

The reported incidence is between 0.2 to 19.1 per 100,000. One important predisposing factor is race, with the East Asian race being affected the least and the white race being affected the most. Interestingly, latitude also has an influence on susceptibility of being affected with LCPD (3). It has been shown that the incidence increases with the increase in latitude. Genetics is also a well-described factor. The mutation of the COL2A1 gene of the 12q13 chromosome has been shown in two generations with four male members in a family being affected (4). Other studies suggest repetitive trauma, abnormalities of the blood supply, and coagulation disorders as causative factors (5).

It has been shown that acetabular retroversion and LCPD can coexist. However, the true correlation of cause and effect is not known yet (6). Obesity can also play a role in the initiation of LCPD. It has been shown that circulating leptin, which is a regulator of the adipose tissue mass, is higher than normal in patients with LCPD (7). Another reason of necrosis in the femoral head is attributed to hyper coagulopathy, and it has been shown a significant relationship between LCPD and mutation in factor V Leiden (8).

Clinical manifestation

The prevalence is four times more in boys and nearly 10-15% are affected bilaterally. The first presenting complaint is limping that is usually notice by parents. The second common complaint is pain mostly in the
Table 1. Stulberg and Mose Classification System for LCPD at skeletal maturity

<table>
<thead>
<tr>
<th>Stulberg</th>
<th>Description</th>
</tr>
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<tbody>
<tr>
<td>1</td>
<td>Normal Spherical Head (spherical congruency → No OA)</td>
</tr>
<tr>
<td>2</td>
<td>Spherical head with coxa magna/breva or steep acetabulum (Spherical Congruency)</td>
</tr>
<tr>
<td>3</td>
<td>Non-spherical head (aspherical congruency → OA late adulthood)</td>
</tr>
<tr>
<td>4</td>
<td>Flat head and flat acetabulum (aspherical congruency → OA late adulthood)</td>
</tr>
<tr>
<td>5</td>
<td>Aspherical Incongruence (aspherical incongruency → OA before 50 years)</td>
</tr>
</tbody>
</table>

Imaging

Simple radiographs can diagnosis LCPD. The size and shape of the femoral head is of importance when followed by radiographs. Prognostic radiographic signs rarely appear until LCPD is established and it usually takes over six months after the onset of the disease. Other modalities such as magnetic resonance imaging (MRI) and pneumoarthrography can give more comprehensive information regarding the stage of the disease (9). The application of dynamic pneumoarthrography is used to evaluate the relationship between the femoral head and acetabulum when surgical treatment is indicated.

Moreover, gadolinium-enhanced MRI can reveal femoral head perfusion early in the course of the disease and help in early prediction of the prognosis. Lower magnetic resonance perfusion index is correlated with greater femoral head deformity (10). Although the perfusion index is variable in the early stage of the disease, it can serve as a prognosticator early in the disease course. To help decide the proper treatment in the early stage of LCPD, a predictor of outcome prognosis is needed. Before and after gadolinium-enhanced MRI, two different components of stage I of LCPD are detectable; however, MRI with contrast better reveals the affected area of the femoral head (11).

In bone scanning, there is a strong correlation between the size of the uptake defect on the femoral head and prognosis. The indication is limited to patients who are suspected of being affected by LCPD, which further serves as a prognosticator as well (12). The results of a study by Maranho et al. showed that there is a significant correlation between hip deformity and labral and cartilage abnormalities about the hip on MRI, and a main predisposing factor was the loss of sphericity of the head and decrease in femoral head-neck offset (13).

Classification

In order to predict the prognosis and decide on the proper treatment, a classification with prognostic properties would be a great help (14). There are three classifications in use that mostly take the involvement size and region into account. Interobserver reliability ranges from poor to fair for the Salter-Thompson classification, fair to moderate for the Catterall classification, and moderate to good for the lateral pillar (Herring) classification (15). Stulberg classification is used after skeletal maturity, in which prognosis is almost predictable by observing the deformity of the femoral head and congruity in relation with acetabulum (Table 1).

Herring first described the lateral pillar classification in 1992. Based on plain radiography in the fragmentation phase, the degree of lateral pillar involvement of the femoral head is classified into three stages. When correlating with the Stulberg classification after skeletal maturity, type A of lateral pillar classification had uniformly good results ending in Stulberg I and II in 100% of the cases. Type B of lateral pillar involvement had a good result if the age was below eight years (92% Stulberg I and II, 8% Stulberg III results), but less favorable if the age was above eight years (30% Stulberg II, 50% Stulberg III, and 20% Stulberg IV results). In type C, at large the results were unfavorable regardless of the treatment (29% Stulberg II, 52% Stulberg III, and 19% Stulberg IV results) (16). A moderate type of B/C in between types B and C has also been described, which is used in cases with a poorly ossified lateral pillar or when the loss is exactly 50% of the lateral pillar. The treatment recommendations for type B/C are the same as type B (17). Lateral pillar classification is now widely used in decision-making regarding treatment options in and after the fragmentation phase (18).

Prognosis

The most important determinant of long-term outcome is the femoral head shape. Extrusion of the head is the only factor that determines future degenerative arthritis and influences on treatment options. In children older than eight years, extrusion of the femoral head out of the acetabulum should be addressed to obtain containment (19). The surgical treatment for this purpose should be performed in the fragmentation stage as soon as the diagnosis is made (20). In the absence of lateral extrusion, most of the studies proved spontaneous healing without remaining functional impairment (21). Rosenfeld showed more than 80% favorable results in affected children less than six years old (22). Also Canaves et al. confirmed the good results with children less than six years of age. On the other hand, in severe cases, the prognosis did not differ between operative and non-operative treatment (23). A modified classification
of lateral pillar also has been described by Lee et al, in which they classified type C into C1 and C2. Based on the Stulberg classification, they showed that the results of C1 were significantly better than C2. This modified classification has a significant value in predicting the outcome of LCPD (24).

One other major complication of LCPD is trochanteric overgrowth. Kitoh et al. showed that there is a significant correlation between trochanteric overgrowth and the Stulberg classification at maturity. The only significant predictor of trochanteric overgrowth was the lateral pillar height with much greater risk of trochanteric overgrowth development in type C (44%) rather than types B and B/C (10%). In patients with more advanced stages of the disease, close follow-up is recommended to be able to detect and decide on early prophylactic epiphyseal arrest of the greater trochanter (25).

As a result of femoral head conditions including coxa magna deformity (large femoral head and neck), coxa brevis deformity (shortened femoral neck) and flattened femoral head (coxa plana), the cam effect will be produced, which results in femoroacetabular impingement during hip flexion. In case of limitation of motion and pain during extreme motion, femoroacetabular impingement should be considered and treatment be initiated with proper physical therapy (26).

One other complication of LCPD is leg length discrepancy (LLD). The most important predictor of LLD is the extent of lateral pillar involvement. It is the only predictor of LLD and no other factor including age, sex, and treatment modality is correlated with LLD at skeletal maturity (27).

**Conservative treatment**

Containment is considered in order to hold the femoral head inside the acetabulum during the plasticity period of the head while the necrotic bone is resorbing and the living bone substitutes through the creeping substitution phenomenon. The choice of treatment to contain the head inside the acetabulum depends on the surgeon’s preference and experience, as well as the psychosocial level of the patient and the family. Failure is not because of the treatment method, but usually is because of technical errors, inappropriate patient selection, and delayed treatment (28).

When reviewing the articles and present practice of bracing in LCPD, a major controversy exists in terms of weaning and discontinuation of bracing and even if bracing itself would have any influence on the outcome. The present literature does not support bracing in the treatment of LCPD. Petrie cast and other braces are rarely used in the salvage stage with hip subluxation or hinged abduction. However, they may be of some help in a deformed femoral head before reossification completes (29).

In a large study using the Thomas splint, 143 patients were followed. The age at diagnosis was 6.6 years and patients were followed up to an average age of 16. The center-edge angle at the time of diagnosis was 18 degrees in the control group and 10 degrees in the affected hips. At the skeletal maturity, the center-edge angle decreased and the acetabular index angle increased in both affected and non-affected hips with subsequent Stulberg type III, IV, and V compared with the control group. The radiographic changes were initially visible on the affected side, but after maturity, both hips showed the changes (30).

Scottish-Rite orthosis is another conservative treatment, but there is no sufficient data to support its efficacy. It is believed that Scottish-Rite orthosis is not only unable to improve containment, but also does not unload the hip joint during walking (31).

Larson et al. in a prospective study assessed the level of pain and function among LCPD patients treated non-operatively. They concluded that in lateral pillar type B, B/C and C, pain and hip dysfunction is common. Also, clinical signs of femoroacetabular impingement and radiographic signs of hip osteoarthritis were found to be correlated as a source of pain in non-operatively treated patients (32).

**Surgical treatment**

It is believed that prior to any osteotomy, range of motion should be regained. However, the effect of osteotomy on increasing the range of motion is not clear. It has been shown and suggested that osteotomy makes the head more spherical in lateral pillar type B than the other types, but in patients with lateral pillar type C or in those older than eight years, the outcome is still unfavorable. Rich et al. reported 240 involved hips that were managed in the necrotic or the fragmentation stage to restore hip abduction with adductor tenotomy and abduction cast followed by daily hip range-of-motion exercises and an A-frame orthosis to facilitate the concentric position of the epiphysis within the acetabulum. This treatment protocol resulted in sphericity of the femoral head in a high proportion of patients regardless of the extent of involvement. In lateral pillar type B and C, 87% of involved hips were congruent and in total, 93% of the hips were congruent at the final follow-up (33).

In a study with pooled data to calculate the odds of improved results, Saran et al. showed that children older than six years benefit more from varus or innominate osteotomy compared to non-operative treatments (34). The main advantage of the Salter or innominate osteotomy is its effect on femoral head remodeling during remaining growth. This osteotomy alone is usually indicated for younger children with recent clinical onset and no femoral head deformity or subluxation. (35).

Arthrodiastasis of the hip joint with soft tissue release is accounted as a surgical method when other treatment options are contraindicated. It also improves the range of motion, reduces superior and lateral subluxation, and provides better radiographic sphericity of the femoral head. Treatment with distraction can be done even in stiff hips and hips with deformity (36).

Varus femoral osteotomy was first introduced in 1965 and became a popular surgery in the treatment of LCPD (37). It allows realignment and identification of the best-fit position of the hip, while restoring the joint congruity and reducing the femoroacetabular impingement. Valgus femoral osteotomy is done to reduce the hinged
abduction during remodeling and improves substantially the symptoms and range of motion (38). With valgus femoral osteotomy in severely deformed femoral heads, greater congruency is obtained in abduction rather than in abduction. In a study to evaluate the effectiveness of valgus osteotomy on femoral head roundness, femoral head subluxation and function, Kim et al. found that it helped to keep the deformed femoral head inside the acetabulum during the fragmentation phase to be remodeled and fit to the acetabulum (39). Also, valgus osteotomy is beneficial in relieving the hinge abduction after skeletal maturity (40).

Shelf acetabuloplasty showed improvement in femoral head coverage; however, the available literature does not support this procedure to prevent late osteoarthritis and warrants improved long term function (41). Recently, the labral support technique that is a sort of shelf arthroplasty has been proposed to maintain containment. This new technique includes a minimal-incision variation of the labral support shelf arthroplasty, which uses arthroscopic visualization and an allograft buttress on the shelf support. The outcome of this minimal incision technique is compared with the traditional technique and both showed similar results when compared with a Petrie cast, a varus femoral osteotomy, or an innominate osteotomy of Salter. They believe that the labral support shelf arthroplasty technique is simple to perform and does not induce a permanent deformity in the proximal femur or acetabulum (42). Another advantage of the labral support procedure is the additional lateral growth of the true acetabulum to create more coverage following surgery, whereas varus osteotomy lacks this advantage. Thus, the labral support technique can prevent subluxation, stimulates the lateral acetabular growth, and restores a shelf after femoral epiphysseal reossification (43).

Triple innominate osteotomy is another option in achieving containment in LCPD. It is one of the most efficient techniques in femoral head containment in all cases. However, over coverage can lead to pincer impingement. To prevent pincer impingement, correction beyond 44 degrees of enter-edge angle is not recommended (44).

Intra- and extra-articular deformities of the hip can lead to premature osteoarthritis of the hip. Impingement and instability can result in joint damage as well, which are usually addressed by trochanteric advancement. Surgical dislocation of the hip along with the retinacular flap allows lengthening of the femoral neck and reduction of the femoral head in deformities such as coxa brevia and coxa vara with Stulberg type II, III, IV, and V sequel. Leuing and Ganz reported 14 patients with a minimum follow-up of three years who have been treated by this technique with no major complication. Hip mobility, pain, and gait improved greatly in this patient group (45). The surgical dislocation of the hip joint allows visualization and identification of the residual deformities and further allows correction of deformities by dynamic evaluation of the hip. It has shown promising results in the treatment of femoral head deformities following LCPD (46). Transtrochanteric rotational osteotomy is a new technique for patients with the onset of LCPD after nine years of age. It is an effective procedure to salvage late-onset LCPD in affected hips. Moreover, amount of head involvement and lateral pillar influence surgical outcomes (47).

Recent techniques are focusing on reshaping the femoral head, both in size and congruency, to match with the acetabulum and decrease the impingement, as well as restoring the normal cartilage in the weight-bearing zone of the head (48). Total hip arthroplasty is a salvage procedure for the complications and subsequent osteoarthritis. Cementless THA showed 90% survival rate in an eight year follow-up. However, in spite of promising results, nerve injury and intraoperative fracture is common. Care should be taken to avoid excessive limb lengthening (49).

Conclusion
The reviewed studies showed three factors influencing the results in patients with LCPD including the age at onset, severity of the femoral head involvement, and the type of treatment. Patients over eight years old benefit from surgical intervention, whereas children less than six years of age often have a good prognosis, except in the group with lateral pillar type C involvement of the head (50). Other meta-analysis studies showed that the probability of developing the spherical head is more likely with operative rather than non-operative treatments in children older than six years. However, in children younger than six, operative and non-operative treatments did not yield any difference in prognosis and both resulted in good outcomes. When patients older than six were operated on, there were no differences between the femoral or pelvic procedure in radiographic outcomes and both techniques had good results. However, in children younger than six, pelvic procedures were more likely to result in a good radiographic outcome rather than femoral osteotomy. (51). Herring et al. in a prospective multicenter study concluded that the lateral pillar classification and age at the time of onset of the disease strongly correlate with the outcome in patients with LCPD (52). These two factors (age and lateral pillar classification) can be used to decide on operative versus non-operative treatments. Future studies need to focus more on treatment options for each complication and the choice of arthroplasty and resurfacing after osteoarthritis (53, 54).

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