



Review Article

Overview of surgical treatments in Legg-Calvé-Perthes disease

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ARTICLE INFO

Article history:

Received 31 August 2012

Received in revised form

13 September 2012

Accepted 13 September 2012

Available online 9 November 2012

Keywords:

lateral pillar classification

Legg-Calvé-Perthes disease

surgery

ABSTRACT

Since Dr Legg reported the paper entitled “An obscure affliction of the hip joint” in 1910, Legg-Calvé-Perthes disease (LCPD) has been recognized. In the intervening 100 years, our understanding of disease etiology, natural history, treatment options, and factors related to prognosis have expanded, and yet many opinions remain without consensus, especially in treatment decisions. During the past 30 years, containment of the femoral head within the acetabulum by conservative or surgical methods has been popularly accepted as a concept for treatments. Several large and multicenter retrospective studies have noted three factors related to outcome in children treated for LCPD: age at onset, severity of involved femoral head, and type of treatment. In patients with onset over the age of 8 years and greater than lateral pillar B or B/C class, surgical treatment was associated with improved Stulberg outcomes compared with conservative treatments. Moreover, the decision to apply appropriate surgical methods should consider the age at surgery, Waldenström stage, and whether the femoral head was containable in abduction. Relevant studies with evidence-based data regarding the results of different surgical methods for LCPD are reviewed here, and there are valid descriptions of surgical indications, characteristics, and associations with improved radiographic outcome.

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1. Introduction

Legg-Calvé-Perthes disease (LCPD) is a juvenile hip disorder characterized by idiopathic osteonecrosis of the femoral epiphysis that is believed to be related to arterial infarction.¹ According to Waldenström, the disease process commenced with aseptic necrosis, and was followed by a subchondral fracture, fragmentation, revascularization, and remodeling.^{2,3} LCPD predominantly affects boys between the ages of 4 years and 8 years.⁴ If the initial symptoms occur after this age range, the disease is considered late-onset and believed to have a poorer outcome compared with children presenting at a younger age.^{5–7} In the past 100 years, there have been many publications concerning the indications and methods of treatment; however, controversy remains. Current concepts of treatment for LCPD, either nonoperative or operative, are largely based on the principles of containing the vulnerable and biologically plastic femoral head in the acetabulum in the early stage in order to keep a spherical femoral head and congruent joint during the repair process.^{7–9} Nonoperative containment

treatments, which involve different abduction braces, may be appropriate in younger patients, whereas in older children with more severe disease, surgical treatments have been suggested to obtain a better result.^{8,9} There have been various surgical options proposed for management. At the fragmentation stage, the child may present with a subluxated hip due to extruded epiphyseal segment of the femoral head. If the hip is containable in abduction, but subluxated in adduction, containment surgery [e.g., femoral varus osteotomy,^{10–13} Salter and triple innominate osteotomy,^{14–18} or shelf acetabuloplasty (SA)^{19–21}] should be contemplated, taking consideration of the age of onset and the extent of uncoverage of the femoral head. However, once the disease course proceeds to the late reossification or healed stage, the enlarged and saddle-shaped femoral head may become irreducible. In this situation, the surgery should be altered from the containment of the femoral head through remodeling to salvage procedures that improve the acetabular volume. Various surgical techniques have been addressed for noncontainable LCPD hips, such as SA,^{19–21} Chiari osteotomy,²² proximal femoral valgus osteotomy,^{23–25} articulated hip distraction,^{26,27} surgical dislocation, osteochondroplasty,^{28–30} and combinations of these procedures. This article reviews the efficacy of all surgical modalities for LCPD and recommends the appropriate treatment protocols for clinical application.

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2. Surgical indications

The optimal treatment methods for LCPD and its prognosis are still not fully understood. In the past 10 years, some authors have tried to standardize the treatment principles for Perthes' hip. Age at diagnosis and extent of femoral head involvement (lateral pillar classification⁴ or Catterall classification³¹) are the most common classifications used to prognose the outcome following treatments. They also give indication for different types of treatment. In a large prospective and multicenter study, Herring et al compared the results of various conservative and operative interventions.⁹ They summarized a basic treatment plan for LCPD. All lateral pillar Group A and Group B hips whose onset occurred at the age of 6 years or less have an excellent prognosis and require only symptomatic treatments. Surgical treatment is proposed in all Group C, B/C, and B hips whose onset occurred after the age of 8 years. Group B hips whose onset occurs between the ages of 6 years and 8 years have a variable prognosis, which seems unrelated to treatment method.^{8,9} Another large prospective review with medium-term follow-up by Wiig et al⁷ yielded similar findings. They suggested that children aged 6 years and older, with more than 50% femoral head involvement (Catterall Groups 3 and 4) had a better outcome if treated with surgery.

3. Treatment options

3.1. Femoral varus osteotomy

Since the preliminary report by Axer¹⁰ in 1965, femoral varus osteotomy (FVO) has become one of the most popular operative methods for LCPD. Varus osteotomy of the proximal femur aims to center the femoral head deeply within the acetabulum and allows correction of the flexion or rotational deformity simultaneously. The prerequisites for FVO are similar to those for innominate osteotomy, which include a good range of motion, hip congruency, and being able to contain the femoral head in abduction. The surgery is recommended in the early stage of fragmentation, when favorable biological and biomechanical effects may be anticipated. Many studies reported that FVO yields good long-term results.^{11–13}

The average varus angle correction needed is approximately 25° to 35°, which would yield a shortening of 1 cm to 2 cm. The initial shortening caused by osteotomy may gradually correct over a period of years when it was done in children younger than 8 years. The varus angle correction can be limited to 15° in older children to avoid prolonged or permanent limp. Theoretically, this temporary shortening has a positive effect by relieving the pressure over the necrotic head. However, abductor insufficiency, worsened by the varus osteotomy, is a serious problem. It will preclude "dynamic containment" of the femoral head and cause Trendelenburg gait. Intensive physiotherapy to actively strengthen abductors may resolve it. In addition, the greater trochanter must be maintained distal to the level of the femoral head to prevent an abductor lurch at any time. Preventive trochanteric epiphysiodesis or distal transfer is probably required simultaneously or at a later time.¹³

3.2. Salter osteotomy

Salter osteotomy, which redirects the acetabulum and improves the anterolateral femoral head coverage, was introduced as a method for surgical containment of LCPD in 1962.¹⁴ The general indications for Salter osteotomy are the same as for any form of containment.^{15,16} These include: (1) age of onset over 6 years old (perhaps 5 years in girls); (2) more than 50% of the femoral head

affected; and (3) hip subluxation in the weight-bearing position. Salter osteotomy has been reported to produce better long-term results than nonoperative methods with respect to Stulberg classification.^{9,15} Other studies have compared Salter osteotomy and proximal FVO.^{32–34} They found similar results with respect to femoral head sphericity, but increased femoral head coverage by the center-edge angle after Salter osteotomy. Potentially, Salter osteotomy can displace the acetabulum 1 cm medially and distally as well, thereby decreasing the biomechanical stress over the hip joint and improving the generally associated leg length discrepancy.^{35,36} However, cautious clinical examination and radiographic assessment are necessary before surgery. The prerequisites of Salter osteotomy include full range of hip motion preoperatively, especially abduction and reasonable joint congruency.

Occasionally, Salter osteotomy alone may not provide sufficient head coverage, particularly in older children (age >9 years). The use of combined Salter and proximal femoral varus osteotomies (to 110–115° varus angle) have been performed recently to contain a larger and deformed femoral head.^{18,37} In Herring's multicenter study, they concluded that Group C cases tend to have an unfavorable prognosis regardless of age or surgical treatment.⁹ The opinions may be questioned because their treatments lacked combined osteotomies. As Javid and Wedge's claimed,³⁷ the combined surgery may change the otherwise "poor" into "fair" hips and improve the natural history in older children. The other benefits of combined surgery include decreasing the effect of increased intra-articular pressure from innominate osteotomy and compensating the shortening from femoral osteotomy.

3.3. Triple innominate osteotomy

Although FVO and Salter osteotomy have become the most common methods for surgical containments, there are certain practical limitations of these two procedures. The degree of FVO required to contain the femoral head may further shorten the limb and cause prolonged limp, especially in older children. Furthermore, use of Salter osteotomy may not provide enough acetabular rotation to cover the femoral head in severe cases, potentially leading to iatrogenic hinge abduction.³⁸ Because of the above concerns, advanced containment methods (e.g., Salter plus varus osteotomy or triple innominate osteotomy) have been developed for more severe cases.^{17,18}

Some studies have demonstrated that older age and/or extensive femoral head involvement were risk factors for unsatisfactory results. Sponseller et al³² found that patients older than 10 years at onset had a poor outcome regardless of surgical treatment. Herring et al⁹ also noted that lateral pillar C group at any age had a poor outcome, even when treated by femoral varus osteotomy or Salter osteotomy. These prior studies did not include advanced containment methods in severely deformed hips. Triple innominate osteotomy is anticipated to achieve better femoral head containment than could be achieved with Salter osteotomy alone and to avoid the leg length discrepancy associated with femoral varus osteotomy. The retrospective review of Wenger et al¹⁷ included 40 hips of 39 children with onset ranging in age from 5 years to 13 years who were treated by triple innominate osteotomy; a significantly increased CE angle with an average of 17.2° was reported. They achieved predictable head containment in Herring B patients of all ages and Herring C patients of younger than 8 years. There were no poor results in the Herring B groups and 80% of patients older than 8 years attained a good outcome. In Herring C children younger than 8 years, 83% showed either a good or a fair result. Older children of Herring C (>10 years) remain less predictable; triple innominate osteotomy can only be effective in some of them.

3.4. Shelf acetabuloplasty

Lateral SA is indicated for severe cases when redirection osteotomy is thought insufficient to produce optimal coverage of the extruded femoral head. An intraoperative dynamic arthrography is useful for further confirmation. In severe Perthes disease, laterally displaced and enlarged femoral head will preclude normal motion of the hip. This condition, “hinge abduction”, was first proposed by Quain and Catterall³⁹ to describe the abnormal movement of the hip resulting from the impingement of the superolateral portion of a deformed femoral head on the lateral lip of the acetabulum. In case of fixed hinge abduction, the patient would present with pain, restricted hip motion, and permanent gait disturbance.

Several studies have shown that SA is a safe and effective procedure in managing those cases with aspherical congruency or incongruency with hinge abduction.^{19–21} Ghanem et al²⁰ reported a series of 30 severely involved LCPD patients, average age 8.6 years; all patients were pain free and had normal or almost normal hip motion. Age at surgery, severity of femoral head involvement, and presence of “head at risk” signs do not appear to affect the final outcome.²⁰ At the last evaluation, 19 hips (64%) were classified as Stulberg 1 or 2 and six (20%) as Grade 3. A significant improvement was noted in the majority of radiographic parameters. Our retrospective study²¹ also revealed improved abduction, internal rotation, and acetabular coverage after treating Perthes’ patients with an incongruent hip or hinge abduction by using SA. However, despite these favorable results in LCPD, SA should remain a salvage procedure and should be reserved for advanced cases. For containable cases, “anatomical” procedures, such as redirection innominate osteotomies, remain the primary choice.

3.5. Chiari osteotomy

In addition to SA, Chiari osteotomy is another popular salvage procedure for children with insufficient femoral head coverage. The potential advantage of Chiari osteotomy over shelf procedure is the reduction of joint loading by medialization of the hip, which was considered an important factor for improving hip congruency and femoral head remodelling.²² This method has been proposed for severe LCPD cases, similar to the shelf procedure. Great care must be taken, and these salvage procedures should not be performed in Perthes’ patients at the healed stage due to lack of femoral head remodeling ability.

3.6. Femoral valgus extension osteotomy

In LCPD children at the late healed stage, decreased abduction is possibly associated with femoral head overgrowth and saddle-shaped deformity. When an arthrography demonstrates femoral head deformity with unstable movement and hinge abduction but becomes stable in adduction and flexion, a valgus and extension osteotomy may be an effective procedure for unloading of deformed epiphyseal segment, restoration of joint congruity, and alleviation of femoroacetabular impingement. The concept of femoral valgus extension osteotomy depends on redirecting the more congruent and round anteromedial part of the femoral head to the neutral position of weight bearing. This rotational and sagittal correction can improve the gait and hip motion, reduced pain, and femoral head shape.^{23–25} It will also resolve the problems of leg length shortening and provide a more normal abductor mechanism. However, this procedure is contraindicated in the stiff hip.

Choi et al²⁵ reviewed 35 hips, with an average age of 9.4 years at surgery, that underwent valgus osteotomy for hinge abduction. Postoperative pain relief, improvement in limp, and increased

range of hip motion occurred in the majority of cases. The overall Iowa hip score significantly increased from 71.2 before surgery to 95.2 at the final follow-up. The Stulberg classification was II in 4 (11.4%), III in 22 (62.9%), and IV in 9 (25.7%). Favorable remodeling can be anticipated especially when valgus osteotomy is performed before the late reossification/healed stage and at younger age.

3.7. Hip distraction

Arthrodiastasis, or distraction of the joint, has been considered as an alternative treatment in LCPD beyond conventional surgical methods. Articulated hip distraction aims to maintain the head in a properly contained position, reduce the deforming force, encourage synovial circulation, and provide an ideal environment for cartilage repair.^{26,27} This method can be used in late onset LCPD, and cases with a varying degree of femoral head deformity or joint subluxation, and not limited by hip stiffness. The advantages include easy technique, minimal complication rates, and preservation of the original anatomy of the acetabulum and proximal femur. Although some studies reported comparable results with other salvage procedures, the long-term benefits are not conclusive and require further investigation.

3.8. Surgical dislocation and osteochondroplasty

Residual hip deformity secondary to LCPD can be quite complex and may cause hip instability, femoroacetabular impingement (FAI), or combinations thereof, which will ultimately predispose to degenerative joint disease. Dysplastic acetabulum is often caused by improper remodeling in response to the nonspherical head. Impingement in LCPD is possibly due to nonspherical head, over-riding greater trochanter with short neck, or functional retroversion of proximal femur and acetabulum. Although some authors have shown successful treatment of young patients with advanced joint degeneration using hip arthroplasty,^{40,41} others proposed an alternative method, “surgical dislocation and osteochondroplasty”, to deal with those who are symptomatic but do not show severe radiographic degenerative changes.^{28–30}

Recently, Ganz et al⁴² reported that their surgical approach to the hip with dislocation and osteochondroplasty of the head–neck junction can be used in Perthes hip with only a small risk of osteonecrosis. It is important to accurately assess the source of the conflict between the femoral head–neck and acetabular edge to design the surgical plan. In addition to plain film, advanced imaging modalities such as 3D computed tomography and magnetic resonance imaging can help to delineate the detailed configuration of the head–neck junction and the damage pattern of cartilage and labrum after LCPD. Surgical management should start with correction of the proximal femur deformity, first through a surgical dislocation approach. Osteochondroplasty is performed for enlarged femoral head with reduced head–neck offset. Relative neck lengthening osteotomy is then indicated to correct extra-articular impingement from high-riding greater trochanter and short neck. If the osteoplasty is not enough to correct the FAI, an intertrochanteric osteotomy to realign the proximal femur has to be considered. After correction of the femoral deformity, it is critical to evaluate the acetabulum, which may be dysplastic or over-augmented. These conditions can be improved through a redirection periacetabular osteotomy or osteoplasty of the acetabular rim with labrum refixation.^{28–30}

4. Recommended management for children with LCPD

Many treatment protocols depend on the age of onset and radiographic appearances, including Waldenström classification

(necrosis, fragmentation, reossification, healing), extent of involved femoral head (Herring or Catterall classification), and several clinical and radiological “head at risk” signs described by Catterall.³¹ After making the diagnosis, careful clinical examination for limp or limitation of range of hip motion will give further information for consideration. A recommended protocol for treatment based on age, clinical, and radiographic findings is as follows.

Children younger than 6 years – prognosis is generally good.

- Symptomatic treatment and range-of-motion exercise
- The exception is the child with whole head involvement (Catterall 4); the prognosis can be improved by FVO or Salter osteotomy

Children between 6 and 8 years – prognosis is variable – require close observation of signs of “head at risk”, which indicates the need for surgery. Once any signs of head at risk are observed, a dynamic arthrography under anesthesia is valuable before deciding the method of treatment.

- The head is contained and stable, without signs of head at risk – conservative treatment and close monitoring
- Signs of “head at risk” are present, and the head can be contained in abduction – FVO or Salter osteotomy alone should be used in moderate cases, while a combined femoral and Salter osteotomy or triple innominate osteotomy may be necessary in advanced cases
- The head cannot be contained but is uncovered and hinged – SA or Chiari osteotomy will provide adequate support

Children older than 8 years – prognosis is often poor, but advanced or salvage procedures still bring the benefit for improved femoral head coverage.

- The head can be contained in abduction (fragmentation stage) – a combined Salter and proximal femoral varus producing osteotomy, triple osteotomy, or SA is feasible
- The head cannot be contained with unstable movement and hinging (late fragmentation or early reossification stage) – SA or Chiari osteotomy is appropriate
- The established femoral head deformity can cause painful limp, unstable movement and hinge abduction, but is stable in adduction and flexion (early healing stage) – a femoral valgus and extension osteotomy will produce stable movement, improved leg length, and more normal abductor strength
- The deformed femoral head leads to joint subluxation and hip stiffness (any stage) – a hip distraction can help to maintain the head in a proper position, relieve the deforming force and encourage the cartilage repair
- For correction of residual deformities after LCPD, such as joint instability and FAI – a surgical hip dislocation with osteochondroplasty, neck lengthening, and greater trochanter advancement can be used to deal with these sequelae of LCPD and prevent early degenerative joint disease

5. Summary

Despite current nonoperative or operative treatments being shown to produce a favorable outcome effectively with a spherical femoral head in LCPD, there is still much to be investigated. The results of these studies did not answer the question of why treatments produce a satisfactory outcome in some patients but not in

others. One reason is that treatments provide some load-sharing effects on the necrotic femoral head, but they do not directly facilitate the impaired bone healing, especially in older children. Further studies have developed a better understanding of LCPD pathogenesis regarding increased bone resorption and delayed new bone formation in combination with serial mechanical loading. There have been animal studies of femoral head ischemia, which obtained improved head sphericity after the use of bisphosphonates.^{43,44} It will be interesting to see whether these medical strategies could alter the pathophysiology of the disorder and yield a more promising result in the future.

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