

Legg-Calvé-Perthes Disease

An Overview with Recent Literature

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Abstract

The evolving knowledge on Legg-Calvé-Perthes (LCP) demonstrates the utility of studying a rare disease systematically by piecing together the biology and mechanics of this condition and applying clinical observations to improve patient care. As treatments of less common diseases are hard to randomize and study in meaningful numbers, long-term study groups have been created to provide insight into this entity that remains an enigma in many aspects. These studies permit a more evidence-approached guide to prognosis and treatment. Meanwhile, basic science research contributes to our understanding of pathophysiology of the disruption and repair processes that lead to LCP, with the goal of clinical translation. This review of LCP aims to give an overview of the condition, with specific focus on recent literature.

Legg-Calvé-Perthes disease (LCP) is osteonecrosis of the juvenile hip first described by Arthur Thornton Legg in 1909 as “An Obscure Affection of the Hip Joint,” shortly after roentgen technology was discovered in 1895.¹ His work was published in 1910, which was the same year that the Frenchman Jacques Calvé² and German Georg Perthes³ produced similar descriptions. Interestingly, in 1909 the Swedish Henning Waldenström also gave early insight through patho-histologic evaluation and radiographic staging. Initially, he termed the characteristic lesion as “the upper tuberculous focus in the collum,” however, and was not included in the naming. His contribution to our understanding of this disease, however, would nonetheless prove invaluable.⁴

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Pathophysiology

Despite general consensus that LCP results from the uncoupling of bone metabolism with increased resorption and delayed formation, the exact etiology remains elusive. As few human specimens are available, the use of animal models in the last decade has allowed novel investigation. Suture ties around the femoral neck in a piglet model creates a condition similar to LCP.⁵ When induced vascular insult is combined with the mechanical stress of repetitive loading, the typical progression from osteolysis to flattening is observed. Histologically, osteoclasts around bony trabeculae are present, indicating active bony resorption at the revascularization front. Fibrovascular replacement of normal trabeculae with numerous small vessels and proliferation of mesenchymal and fibroblast-like cells ensues and persists, instead of a reparative phenomenon with new bone formation.

One explanation of pathogenesis involves the large cartilage anlage of the femoral head. As LCP patients tend to have delayed bone age, on average 2 years in girls and 1 year in boys,⁶ their femoral head ossific nuclei are smaller than those in children of similar chronologic age. This makes the cartilaginous component of their epiphysis relatively larger, and the traversing blood vessels are more vulnerable to mechanical compression.⁷

Honing in on vascular phenomena, one study examined super-selective angiography of the medial femoral circumflex artery. The posterior columnar artery branches off the medial femoral circumflex, which then gives rise to the lateral epiphyseal artery (LEA) that extended into the depth of the epiphysis. Angiography demonstrates interruption of the origin of the lateral epiphyseal artery in 68% of LCP patients’ hips in all stages.⁸ Furthermore, perfusion is position-dependent. Abduction slightly decreases perfusion by stretching the posterior circumflex artery despite the LEA remaining patent. The combination of abduction with internal rotation interrupts flow through the LEA as it traverses the capsule.

In addition to decreased vascularity, the concomitant role of repetitive loading has been demonstrated. Spontaneously hypertensive rats, known to have a predilection for femoral osteonecrosis resembling LCP, have been tested in special cages requiring prolonged standing on hind legs to feed. Increased compression from mechanical loading was associated with flattening of the femoral head.⁹ While proximal femoral histology in control rats had discontinuation and thinning of the growth plate as a result of this loading, rats with an inherent predisposition to osteonecrosis had increased physeal depression and distortion as seen in LCP. Of note, in this same model, the bisphosphonate zoledronic acid was found to improve femoral head shape.¹⁰

A technique termed quantitative backscattered electron imaging examines the actual composition of LCP-type bone as it relates to mechanical stress.¹¹ Femora demonstrated increased mineralization density in necrotic calcified epiphyseal matrix and subchondral bone. These changes likely account for the brittle quality of the weakened epiphysis in LCP, seen as increased radiodensity on imaging.

Epidemiology

Clinical onset tends to be between 4 and 8 years of age, though ethnic variations are present. Indian children tend to present later than average.¹² Incidence is lowest in equatorial regions, increasing towards Northern Europe. Incidence is highest in whites and lowest in African Americans.

Boys are diagnosed with LCP five times as often as girls, though bilateral cases are more common in girls. Studies are conflicting with regard to gender differences in prognosis. Physeal closure occurs earlier in girls, leaving less time for femoral head remodeling (3.4 years versus 5.9 years).¹³ Despite this, no difference between genders has been detected in final radiographic outcome.

Twin studies have shown low concordance, pointing more towards environmental causes as opposed to genetic predisposition. The role of nurture over nature is reinforced by the variable distribution in different social classes. Incidence is quoted as varying from 4 to 32 per 100,000 in the highest and lowest social classes, respectively.¹⁴ Lastly, the log-normal age distribution pattern is consistent with a single exposure phenomenon acting during hip development prenatally or early within the first 2 years of life.¹⁵

Associations

Studies associating LCP with delayed bone age and low birth weight have been confounded by factors, such as so-

cioeconomic status, smoking, and steroid use in premature infants.¹⁶ There is an assumed association with small head circumference; however, LCP has been shown in multiple studies to actually demonstrate a phenomenon best termed “rostral sparing dysmorphism.” Head size is average, with proportionately smaller body size distally.¹⁷ The most notable growth restriction is in the feet.¹⁸

The association between LCP and congenital malformations (inguinal hernia, genitourinary abnormalities, undescended testes, and Down’s syndrome) is well accepted, while other connections, such as coagulopathies, are controversial. A 2008 review showed no difference in prothrombotic markers¹⁹; however, a more recent and larger 2010 study revealed a higher incidence of factor V Leiden mutation, protein S deficiency, elevated factor VIII, and prothrombin G20210A mutation in LCP patients, especially males.²⁰

Patient Evaluation

Clinical presentation can range from a painless limp to an “irritable hip.” Children may complain of knee pain that is referred from the hip. Limited range of motion, especially abduction and internal rotation, is present, occasionally with a Trendelenburg gait. Initial workup should include basic labs: CBC, ESR, and CRP, at a minimum. Additionally, an anteroposterior (AP) pelvic radiograph including both femoral heads, along with a frog lateral of the affected side, should be performed.

The differential diagnosis includes many conditions. The painful synovitis causing early symptoms in LCP can be clinically indistinguishable from transient synovitis, and initial radiographs can be negative. Distinct characteristics are described (Table 1), but follow-up radiographs are indicated to check for LCP even after symptomatic improvement. More important is the distinction between the irritable hip and a septic joint since the latter requires emergent treatment. Infectious etiology must be ruled out with reasonable confidence before attributing symptoms to synovitis.

One clue to the diagnosis of LCP is that bilateral involvement almost always demonstrates hips to be in different stages of disease. Symmetric involvement suggests other conditions, such as epiphyseal dysplasia, while atypical presentations should prompt consideration for osteonecrotic conditions, such as hypothyroidism, Gaucher’s Disease,²¹ glycogen storage defects, sickle cell anemia, and Meyer’s dysplasia. A skeletal survey can detect additional epiphyseal involvement. A hand radiograph for bone age

Table 1 Comparison of Transient Hip Synovitis and LCP Characteristics⁸¹

Characteristic	Irritable Hip Syndrome	LCP
Gender	2 male; 1 female	3 males; 1 female
Age	3 years	7 years
Symptom Duration	6 days	6 weeks

determination assists in the differential as well, as LCP patients often have delayed bone age.

Imaging

Radiographs remain the most useful imaging modality for both initial diagnosis and subsequent follow-up. Characteristic changes typically occur after a radiographically silent period the first 3 to 6 months of disease. An effusion or relatively thickened cartilage may widen the medial joint space. The involved hip has a smaller ossific nucleus, often with increased radiodensity. Increase in joint space has been correlated with enlargement of the femoral head.²² Hip pain often correlates with initiation of radiographic resorption when a fracture line and speckled calcification within the femoral head become evident. Increased radiodensity later normalizes during reossification with reconstitution of the subchondral plate. Progressive disease can produce acetabular changes, such as early triradiate closure, bicompartimentalization, and ischium varum.^{23,24}

Several radiographic findings correlate with a poor result and have been termed “head at risk signs” by Catterall.²⁵ The Gage sign, detailed by Courtney Gage in 1933, is an osteoporotic segment visualized on the lateral side of the epiphysis.^{26,27} Speckled calcification appearing lateral to the epiphysis represents the anterior part of the viable lateral fragment that appears with the start of head reformation. Lateral subluxation manifests as an increase in inferomedial joint space. Lastly, a more horizontal physis is seen in those with almost total head involvement.

Arthrography is an adjunct to standard radiographs that aids in the assessment of range of motion and ability to contain the head in the acetabulum. After general anesthesia and strict sterile preparation, contrast is injected anterolaterally with fluoroscopic guidance to examine three features as detailed by Nakamura.²⁸ First, the subluxation index compares the medial clear space to the acetabular width. The normal average is 8.5%, but abduction increases this value in the presence of hinged abduction. As this is a ratio, magnification error is eliminated. Second, the presence or absence of an impingement sign is determined, being positive when the most superior portion of the epiphysis lies lateral to the bony rim of the acetabulum in abduction. Lastly, the range of abduction under general anesthesia is measured in order to eliminate overdiagnosing hinged abduction or epiphyseal extrusion with hip abduction in patients with muscle spasm when awake.

Bone scintigraphy findings can precede radiographic changes by 3 months, potentially allowing for earlier diagnosis and treatment.²⁹ There is a high prognostic value, and correlation has been made between the vascularization patterns seen and the Herring and Catterall classifications. Unfortunately, significant radiation exposure limits practical application of this imaging modality.

Ultrasonography has been reported to detect hip effusion early in disease when radiographs are undiagnostic,³⁰

though this can be nonspecific. Capsular distention caused by synovial effusion in transient synovitis versus synovial membrane thickening in LCP may be differentiated on ultrasound, but this is rarely used in clinical decision making.

Computed tomography (CT) can show early bone collapse, subtle trabecular changes, and sclerotic zones, as well as intraosseous cysts in later stages. Three dimensional CT can be useful in visualizing complex head deformity, but just as with bone scintigraphy, the benefits of the information gained rarely justifies the radiation dose required.

Magnetic resonance imaging (MRI) is an advanced imaging modality that details the extent of bony infarction and the anatomy of the cartilaginous head and labrum.³¹ It can be useful early in the disease’s course in order to differentiate it from other conditions that cause osteonecrosis.³² Perfusion can be evaluated with dynamic gadolinium-enhanced subtraction (DGS) MRI.³³ Apparent diffusion of the femoral neck decreases with worsening pathology.³⁴ Reparative processes in the epiphysis involve increased vascularity, vasodilatation, greater capillary permeability, and diffusion, all of which increase gadolinium uptake. Ischemia and revascularization of the femoral head can be detected early, giving the same perfusion information as scintigraphy without the radiation exposure.²⁹ MRI has taught us that radiographic medial joint space widening only indicates lateral subluxation in the later stages. Initially, apparent medial joint widening actually represents normal joint space filled with overgrown cartilage, not subluxation.³⁵ This is in contrast to true medial joint space widening with an effusion. Apparent medial joint space typically normalizes as the overgrown cartilage ossifies despite the lateral subluxation associated with coxa magna. In hips with healed LCP, MRI demonstrates abnormalities of the acetabular cartilage in 47% and labrum in 75%.³⁶

Classification

Waldenström described the first radiographic staging system for LCP in 1938.⁴ Initially, a small sclerotic epiphysis with radiographic medial joint widening is present, and disease may remain clinically occult at this stage. This progresses to a fragmentation stage, describing the period of resorption that typically lasts 6 months. It is here when collapse occurs in the softened femoral head from the osteoclastic removal of dead bone without reformation. Hips tend to be most symptomatic in this stage, with inflammatory markers at their highest. Reossification is the third stage, characterized by new bone formation proceeding from lateral to medial and posterior to anterior. This can last up to 18 months. Finally, hips enter a healing or remodeling stage when trabecular patterns reform.

Chronologically, Goff was the next to classify LCP in the 1950s.³⁷ Femoral heads were separated into spherical, capped, and irregular shapes. This is of little clinical utility, however, as subsequent schema have been more descriptive and prognostic.

In addition to detailing the head at risk signs, Catterall's 1971 paper classified LCP based on the amount of epiphyseal involvement.²⁵ Group I involved less than 25% of the epiphysis, typically the anterior head. Group II additionally demonstrated central necrosis up to 50%. Group III and IV hips involved 75% and 100% of the epiphysis, respectively. A large Norwegian study group found head involvement greater than 50%, or Catterall III and IV hips, to be the strongest predictor of outcome.³⁸ Caveats of this system include a somewhat poor interobserver reliability, as well as movement between groups if applied prior to the fragmentation stage.

Salter and Thompson differentiated hips based on the extent of subchondral fracture in the superolateral dome of the femoral head.³⁹ Type A hips have fractures involving less than 50% of the head width and generally have good outcomes. Type B hips with greater than 50% involvement are more likely to have fair or poor results. This simply applied system is advantageous in that the extent of subchondral fracture can be seen on radiographs an average of 8 months prior to being able to group heads according to the Catterall system that requires the fragmentation stage. Though this system has been repeatedly shown to be accurate and even more predictive of the extent of necrosis than MRI studies,⁴⁰ it is limited by the absence of a subchondral fracture line in up to two thirds of patients. While seen earlier than the fragmentation stage, it still only present in 23% of initial studies.

The Herring lateral pillar classification is currently the most commonly used system in LCP⁴¹ and is also applied during the early fragmentation stage evident within 6 months of symptom onset. The lateral pillar is defined as the lateral 5% to 30% of the femoral head on an AP radiograph, depending where the lucent line delineates the central necrotic area. When this lucency is not present, the lateral fourth of the head is used. Group A hips do not involve the lateral pillar. Group B hips have limited involvement with maintenance of over 50% lateral pillar collapse, and Group C hips have over 50% collapse of pillar height. Additionally, a B/C border group has been added to describe lateral pillars at the 50% depression level. This group has been additionally stratified into the following three subgroups: B/C1 with over 50% of lateral pillar height but less than 2 mm to 3 mm wide; B/C2 also with greater than 50% height maintenance but minimal ossification; and B/C3 with exactly 50% height but depressed compared to the central pillar. Lateral pillar height loss has been well correlated with the final radiographic outcome and has been found to be more predictive than age of onset.

Stulberg and colleagues studied hips into skeletal maturity and reported on five classes that can be grouped into three shapes.⁴² Spherical heads can be Class I or II depending on the absence or presence of coxa magna, a steep acetabulum, or short neck. Ovoid heads are termed Class III. Flat heads with or without coxa magna or a steep acetabulum are Class IV and V, respectively. With its many classes, this system has variably reported interobserver reliability,⁴³ some stating it to be the best with 92% reliability, and others have

found it only marginally acceptable (below 75% reliability). However, radiographic sphericity and congruence can now be quantitatively measured.⁴⁴ Simplifying this system into spherical (Stulberg I and II) or nonspherical (Stulberg IV and V) heads increases reliability and defines good and poor outcomes, respectively. Class III ovoid heads indicate moderate outcome. One additional facet of this classification is the concept of congruency of the femoral head in the acetabulum. Congruency is present with both spherical and aspherical ovoid or Class IV flat heads. Class V heads demonstrate aspherical incongruency as their irregular flat shape does not match the more normal-shaped acetabulum.

Mature hips were additionally classified by Mose based on their sphericity.⁴⁵ Concentric circular templates of varying radii are imposed onto AP and lateral radiographs to determine conformity to a spherical shape. Good, fair, and poor groups were defined as those with less than 1 mm, 2 mm, or 3 mm and greater deviation from the circular outline, respectively. This system, similar to Stulberg and colleagues', highlights the association between a smooth spherical femoral head at maturity and good outcome.

Prognosis

Determining how to manage LCP patients requires knowledge of its natural history. The best guide we currently have comes from the Perthes Study Group, which involved 39 investigators in a multicenter prospective study of 438 patients.⁶ A non-operative group managed with one of three different protocols and an operative group consisting of two surgical protocols were compared in children with LCP onset between 6 and 12 years of age. Three main prognostic conclusions were drawn, the first of which was that children with symptom onset prior to 8 years of age are likely to have good results regardless of treatment. In children with onset after age 8 with at least 50% lateral pillar height maintenance (Herring Groups B and B/C), operatively managed hips had better outcomes than non-operatively managed hips. Lastly, Group C hips with greater than 50% collapse of lateral pillar height had poor outcome regardless of treatment.

While general prognosis is better for those with early onset, exceptions are common, precluding the ability to reassure parents of a guaranteed good result in almost any case. A cohort of children with symptom onset prior to age 5 showed over half of hips to be in Catterall Group III or IV and Herring Group C.⁴⁶ At skeletal maturity, only 45% of hips had a good outcome according to the Stulberg classification, while the remaining 22% and 33% were fair and poor. Accurately predicting which hips will improve and which will deteriorate remains a challenge.

Treatment

The treatment of LCP depends on the age and stage of presentation. Simple observation is all that is needed in children aged 2 to 3 years. Parents should be advised on the favorable prognosis but monitored with follow-up radiographs. More

commonly, however, children present at 4 years of age and older. In these cases, the first goal is resolution of the synovitis, which can be facilitated by bed rest with or without light (3 pound) skeletal traction for a short time frame (4 to 14 days) and anti-inflammatory medications. Parents should be advised on the natural history of symptom resolution in 24 to 36 months. Long-term outcome will be dictated by the ultimate shape of the femoral head; therefore, the child should be monitored until reossification.

Modalities ranging from exercise to acupuncture⁴⁷ have been recommended. Home physiotherapy with active and assisted range of motion of the hip and knee helps to maintain joint mobility in conjunction with adductor stretching. Short-term Petrie casting in abduction and internal rotation is another modality that can stretch adductors to achieve better range of motion. Bracing follows the same principle but should be part-time so as not to increase intraarticular pressure. Surgical release of contracted adductors may be required to allow adequate range of motion for bracing and exercise. A protocol of release followed by A-frame bracing with daily ranging has been shown to maintain motion and produce congruent hips even in pillar B and C hips.⁴⁸ These recommendations are based on the principle that range of motion and positions of coverage are good for joint health; however, they mainly serve as adjuncts to observation and surgical treatment rather than primary treatments.

Biologics, namely bisphosphonates, are being investigated as their inherent mechanism delays necrotic bone resorption without enhancing anabolism. Numerous trials have been conducted in animals, and a few published reports on use in humans for osteonecrosis have been encouraging. Aledronate has been shown to prevent early collapse of femoral heads and delay total hip arthroplasty in nontraumatic osteonecrosis.^{49,50} Improved function has been shown even when begun in late stage osteonecrosis. Though LCP is a distinct disease, shared features make this an appealing area for further investigation.

Bone mineralization proteins (BMPs) have also been investigated in animal models. BMP-2 displays synergistic behavior when combined with a bisphosphonate in piglets.⁵¹ Compared to controls, there is better preservation of femoral head morphology, increased trabecular thickness and volume, decreased osteoclasts, and increased surface osteoblasts. Heterotopic ossification in the capsule is an adverse effect and must be investigated along with other safety considerations such as oncogenicity⁵² prior to clinical implementation. Other future directions may utilize inhibitors of tumor necrosis alpha factor, cathepsin K, and NF-beta kinase.⁷

Core decompression has been suggested as an initial procedure to remove necrotic bone, reduce venous congestion, and stimulate revascularization prior to collapse. When performed early, it can relieve pain, though epiphyseal height will not improve.⁵³ Techniques may be modified from those used in adults to protect the physis. Localizing the central

head with a small diameter smooth wire is assisted by injecting dye into the joint and using fluoroscopy to ensure subchondral placement without joint penetration. The critical steps include necrotic bone removal and packing of bone graft. Using a small diameter drill minimizes risk of growth arrest,⁵⁴ though other complications, such as femur fracture and head blowout, remain concerns.

More often, procedures are selected to favorably alter hip joint mechanics. The principle of treatment is to protect the weak fragmented head from deforming forces until it reforms, which is accomplished through containment with or without unloading. When the femoral head is subluxating out of the joint laterally, causing a pressure point on the articular surface, a varus derotational osteotomy (VDRO) can increase coverage of the head while allowing for concomitant correction of femoral anteversion. Preoperative planning is paramount⁵⁵ and should involve templating a medial closing wedge osteotomy and stabilization with a fixed angle device. A neck shaft angle (NSA) of less than 105° should be avoided, as progressive varus may occur. Patients must be warned of the creation of a gluteal lurch and mild shortening that are inherent to VDRO, along with the inconvenience of spica casting and need for additional procedures to remove the hardware and further treat the disease. Better results have been achieved when used before age 10, but even older children have shown improvement when compared to non-containment methods.⁵⁶

Trochanteric overgrowth, defined by an articulo-trochanteric distance less than 5 mm, has been reported to occur in 22% of patients treated with containment surgery and correlates with final Stulberg outcome.⁵⁷ Lateral pillar height may be predictive, and in lateral pillar C hips, a greater trochanteric epiphysiodesis can be performed in conjunction with VDRO. Epiphysiodesis is effective for growth arrest in 60% but can lead to overcorrection in 10%.⁵⁸ With premature proximal femoral physeal closure and a shortened neck from a VDRO, an overgrown greater trochanter can have a “gear-stick” type of impingement in abduction. Trochanteric advancement is then indicated to treat this impingement.

Alone or combined with proximal femoral osteotomy, the acetabulum can be reoriented. A Salter innominate osteotomy, first described for treating hip dysplasia, rotates the acetabulum to improve anterolateral coverage. With modification of graft placement, the leg can be lengthened, which can be advantageous if shortening has already occurred through the avascular process or iatrogenically with osteotomy. It has the additional benefit of triggering a biologic reaction that accelerates revascularization of the epiphysis to shorten duration of the active stage. Studies have shown improved painless range of motion maintained at 10 year follow-up.⁵⁹

Both VDRO and innominate osteotomies redirect the weightbearing to improve mechanics of spherically congruent heads. With deformity progression, however, reconstruction is required. Hinged abduction is one such

deformity, described in 1981 by Catterall and Grossbard.⁶⁰ With poor coverage, undue forces pass through the antero-superolateral head, extruding an epiphyseal segment. The earliest signs are loss of abduction and internal rotation. An arthrogram will both diagnose hinged abduction, with an increased subluxation index in abduction, and determine if it is reducible or irreducible. If the head slips under the acetabular roof with abduction, treatment is then aimed at maintaining the hip in this position. This involves release of tight soft tissue constraints, typically the adductor and psoas tendons along with the medial capsule. Once abduction over 45° is achieved, it is maintained with a Petrie cast for 3 to 6 weeks. Alternatively, VDRO or double, triple, or shelf osteotomy of the acetabulum can be used to keep the head more congruently reduced. Occasionally, if containment is difficult to achieve, bed rest with abduction traction for a week can precede surgical treatment.

Hinged abduction can also become fixed, which will lead to progressive subluxation, lateral pillar collapse, and widening of the femoral head. Arthrogram will demonstrate the deformed head hinging and pushing the labrum upwards. This portends a poor prognosis, as the altered center of rotation causes an antalgic or Trendelenburg gait, a short limb, and compensatory rotation of the limb in the body's attempt to avoid impingement. Increased trunk and pelvic motion from ambulating with a stiff hip may cause pain in adjacent motion segments.⁶¹ As remodeling potential is limited at this stage, the treatment goals are to reduce impingement and reposition the most congruent portion of the joint to be load bearing. Four types of operations have been used to achieve this.⁴⁸

The first is a valgus proximal femoral osteotomy (PFO) performed in the subtrochanteric region of the femoral neck. This alleviates the hinge and realigns the leg with the hip for a neutral weightbearing position. With orthogonal arthrography or more advanced imaging, such as 3D CT, the most

congruent portion of the head should be determined, often requiring sagittal and rotational components to the valgus PFO. This can improve functional limb length discrepancy as well as abductor tensioning. Caution must be used not to induce excessive valgus, as this can lead to lateral instability of the hip. Valgus femoral osteotomy in the late fragmentation stage has been shown to improve both femoral head roundness and continuity of Shenton's line in addition to decreasing the medial gap ratio.⁶²

Acetabular modification, alone or in conjunction with PFO, is the second type of procedure. The shelf arthroplasty is appropriate for severe disease (Catterall III or IV) in older children with limited remodeling potential. It increases the size of the acetabulum to better cover the femoral head by taking iliac bone and adding it to the lateral acetabulum. A Chiari osteotomy is another salvage procedure that deepens the acetabulum to improve superolateral femoral coverage. Instead of reshaping the acetabulum, repositioning it with a periacetabular osteotomy can also improve the line of load bearing for pain relief.⁶³ Many studies have detailed improved pain and function; however, osteoarthritis often leads to patients electing for joint arthroplasty at 5 to 6 years postoperatively. The advantage of addressing the acetabulum is that the general joint alignment is preserved for future arthroplasty.

The remaining two procedures described are less often used as their long-term benefit has yet to be demonstrated. Femoral head reshaping is one technique. This involves surgical hip dislocation with osteochondroplasty of the mushroom shaped LCP femoral head to become more spherical.⁶⁴ Resected lateral protuberances can be reimplanted as osteochondral plugs into head defects from impingement. This can be performed with relative head femoral neck lengthening to improve abductor leverage and reduce impingement.⁶⁵ While short-term results and postoperative



Figure 1 AP (A) and lateral (B) pelvic radiographs of an 11-year-old boy with left LCP.

imaging have been favorable, pain and stiffness have been problematic in long-term follow-up.

Arthrodiastasis is another technique for older children with severe and active disease. The principle is to unload and widen the joint while it is weak during the revascularization process. Neutralizing the joint reactive forces across the hip allows for fibrous repair of cartilage defects and prevents further joint damage. Initial nonarticulating constructs fared poorly, as motion is needed for cartilage health. Subsequent articulating external fixation with concomitant release of tight soft tissues demonstrates improved results.⁶⁶⁻⁷² After placement of a monolateral or circular construct, the joint is gradually distracted until the Shenton line is overcorrected by 5 to 10 mm. This is maintained until lateral pillar reossification occurs and has been shown to preserve epiphyseal height in hips that have minimal collapse. Most hips that are indicated for arthrodiastasis end up with Stulberg IV hips, but joint stiffness is still improved.

Figures 1 through 3 demonstrate a case of LCP where arthrodiastasis was used along with core decompression and bone grafting with biologics. An 11-year-old boy with 5 months of left hip pain presented with radiographs that demonstrated LCP with subchondral fracture and collapse already present (Fig. 1). Two months later, he underwent core decompression of the anterolateral head, along with bone grafting to tamp up the collapsed head with allograft and recombinant human BMP-2. Articulated distraction was then performed with three supra-acetabular pins, two

iliac wing pins, and three lateral femoral pins (Fig. 2). The patient was allowed to weightbear throughout the 4 month duration of distraction. After removal, good range of motion was achieved. Figure 3 was taken 8 months after hardware removal, demonstrating persistent collapse of the head despite this intervention.

Hip arthroscopy can be employed to manage painful sequelae of LCP in adolescents and young adults. Treatable findings include labral tears, hypertrophic ligamentum teres, chondral lesions, loose bodies, and cam lesions of the femoral neck. At 2-year follow-up of 22 patients, all had maintained improved in hip scores.⁷³

Eventual need for joint arthroplasty is not uncommon. In addition to traditional total hip arthroplasty (THA), there has been renewed interest in hip resurfacing arthroplasty. With lower dislocation rates and bone stock preservation, this is an attractive option that can be combined with greater trochanteric advancement to address osteoarthritis. Resurfacing is currently not recommended in women of child-bearing age, patients with renal dysfunction, femoral heads with large cysts, or those with less than 75% of a viable femoral head. A retrospective study compared 19 resurfacings in LCP patients to matched total hip arthroplasties.⁷⁴⁻⁷⁶ The resurfacing group had improved range of motion, no impingement, and gained some leg length. Comparing resurfacing in patients with osteonecrosis of various etiologies to patients without osteonecrosis, similar pain relief and function scores result, though activity level is lower.⁷⁷ Unfortunately, LCP



Figure 2 Articulating external fixator applied to the left hip. Note the improved contour of the femoral head after hip decompression and bone grafting with tamping of the collapse femoral head.

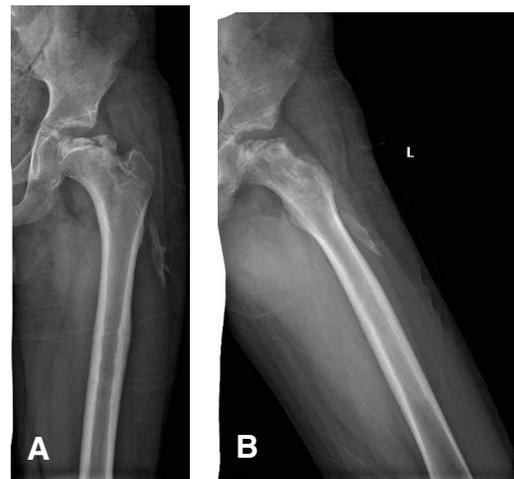


Figure 3 AP (A) and lateral (B) hip radiographs 8 months after hardware removal demonstrating re-collapse of the femoral head.

patients often have factors known to contribute to reduced resurfacing survivorship, such as more femoral defects, small components,⁷⁸ and lower body mass index.

Traditional THA in LCP patients has been studied. Careful preoperative planning is vital to good long-term results, and one can consider preoperative CT scans and navigation to assist in implant positioning. Deformity is common, with acetabular retroversion, wall overhang, and short femoral necks to name a few. Modular implants can help restore femoral version and offset. A large series of over 400 THA for LCP demonstrated no increased in revision rates compared to patients with other indications.⁷⁹ A smaller study with longer 10-year follow-up demonstrated maintained improvement in hip scores.⁸⁰

Conclusion

In summary, LCP is an idiopathic osteonecrosis of the femoral head with uncoupling of bone resorption and formation. In its weakened state, the femoral head deforms with mechanical loading. Much research continues to be dedicated to etiology of the disease in hopes of developing more effective and predictable treatments. Younger onset tends to carry a good prognosis regardless of treatment. In older children with more advanced disease, however, operative management is indicated to try to improve joint mechanics and achieve spherical congruency at maturity. This can be done by addressing the soft tissues, femur, and acetabulum. Biologic treatment strategies are promising adjuncts to preservation procedures. Despite interventions, disease progression does often occur, at which point joint reconstructive can relieve pain.

Disclosure Statement

None of the authors have a financial or proprietary interest in the subject matter or materials discussed, including, but not limited to, employment, consultancies, stock ownership, honoraria, and paid expert testimony.

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