Legg-Calve-Perthes Disease

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Benjamin Joseph

Introduction

Legg-Calve-Perthes disease (LCPD) is a self-limiting disorder of childhood characterised by interruption of blood supply to the capital femoral epiphysis [1]. The blood supply is restored spontaneously over a period of 2–4 years. During this period the femoral head is vulnerable to deformation and in a proportion of children the spherical shape of the femoral head is lost. These children are prone to develop premature secondary degenerative arthritis in early adult life. The challenge of treating LCPD is to prevent degenerative arthritis by preserving the spherical shape of the femoral head.

Though the disease was described in 1910 the aetiology still remains elusive. Treatment in the past involved prolonged bed rest and weight relief for the entire duration of the disease [2, 3]. Early reports of surgical methods of treatment, including drilling of the femoral neck [4] and bone grafting of the femoral epiphysis [5], appeared in the literature by the late 1940s and early 1950s. Axer, in 1965, recognised the need to improve the femoral head coverage in LCPD and described the proximal femoral varus osteotomy [6]. Axer's concept, now referred to as "containment", is widely accepted today but controversies abound regarding several aspects of treatment. A greater understanding of the pathology of LCPD and the pathogenesis of femoral head deformation has led to promising experimental trials of biologic treatment with bisphosphonates and bone morphogenetic protein [7–9]. Despite these advances, there is a paucity of high quality research with very few studies with Level I evidence regarding the efficacy of treatment methods.

Pathophysiology

The cause of interruption of blood flow to the capital epiphysis is unknown but it is clear that the vascular insult results in infarction of part or all of the femoral epiphysis. When partial epiphyseal infarction occurs, the medial and posterior parts of the epiphysis are most commonly spared. The vascular occlusion may involve the medial circumflex femoral artery or its lateral epiphyseal branches [10, 11] and the site of vascular occlusion determines the extent and location of the infarcted segment in the epiphysis. There is some evidence to suggest that more than one episode of infarction occurs [12]. Reparative processes begin soon after the bone infarction commences. Initially, there is a robust osteoclastic response to resorb the necrotic bone. However, replacement of the resorbed trabeculae with new bone does not proceed in tandem because of a relatively poor initial osteoblastic response [13]. This

B. Joseph (✉)
Aster Medcity, Kochi, Kerala, India

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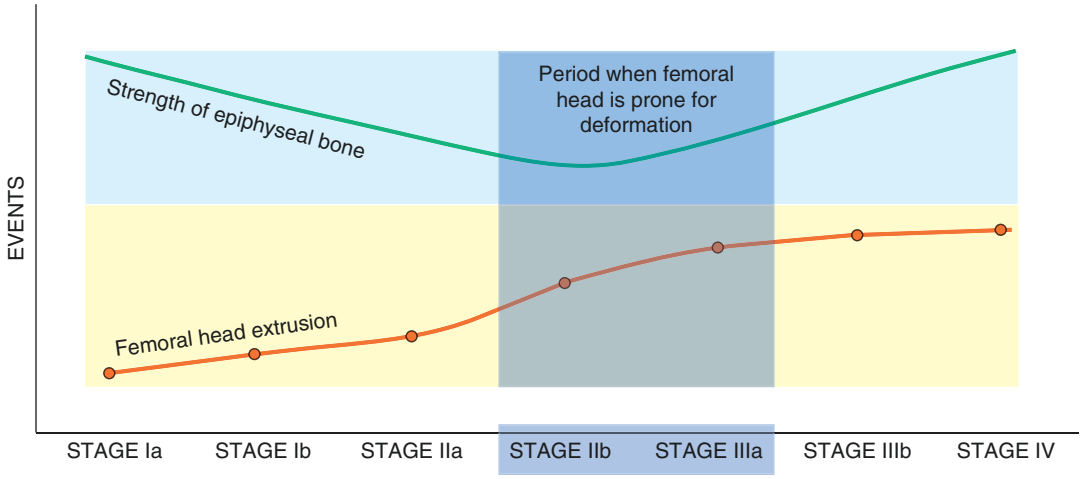

Fig. 6.1 Strength of the epiphyseal bone and femoral head extrusion during the evolution of LCPD by Waldenstrom stage. The femoral head is most vulnerable for deformation when the strength of the epiphyseal bone is least and extrusion is greater than 20%

imbalance between bone resorption and new bone formation renders the bone trabeculae weak and susceptible to collapse. Over a period of time the osteoblasts lay down new bone on the periphery of the infracted epiphysis. This new woven bone is also vulnerable to deformation as the trabeculae are laid down haphazardly and not in directions that resist deformation by weight-bearing stresses seen in lamellar bone. There is thus a period during the evolution of the disease when the femoral epiphysis is inherently prone to deformation. Once the woven bone is replaced by lamellar bone the normal strength of the epiphysis is restored and no further deformation of the epiphysis occurs (Fig. 6.1). The entire process of repair takes 2–4 years [14].

The avascular necrosis associated with LCPD also triggers a chronic synovitis [15] which is characterised by perivascular aggregation of lymphocytes and plasma cells [16] with elevated levels of IgG and IgM [16, 17] in the serum. This suggests that immunological mechanisms may be involved in mediating the synovitis. Elevated interleukin-6 levels in the synovial fluid have also been noted [18]. Muscle spasm induced by the synovitis is partly responsible for the initial reduction in the range of hip motion.

Hypertrophy of the ligamentum teres and femoral and acetabular articular cartilage occurs early in the disease. These soft tissue changes predispose to the femoral head extruding from under the margin of the acetabulum [19, 20]. When the femoral head extrudes, the avascular segment of the epiphysis is exposed to weight-bearing stress and muscular forces across the acetabular margin. Maximal extrusion tends to coincide with the period when the bone is inherently weak due to the imbalance between bone resorption and new bone deposition. The weakened avascular bone of the extruded epiphysis cannot withstand the physiologic stresses of weight-bearing and irreversible deformation of the femoral head ensues (Fig. 6.2). Extrusion in excess of 20% is associated with a high risk of permanent deformation of the femoral head [14, 21]. Extrusion tends to be more severe with more extensive epiphyseal avascularity and it invariably develops in children over the age of seven [14].

> ***"The weakened avascular bone of the extruded epiphysis cannot withstand the physiologic stresses of weight-bearing and irreversible deformation of the femoral head ensues"***

Natural History

The natural history of LCPD can be divided into three parts: the first is from onset until healing of the disease, the second is from healing of the dis-

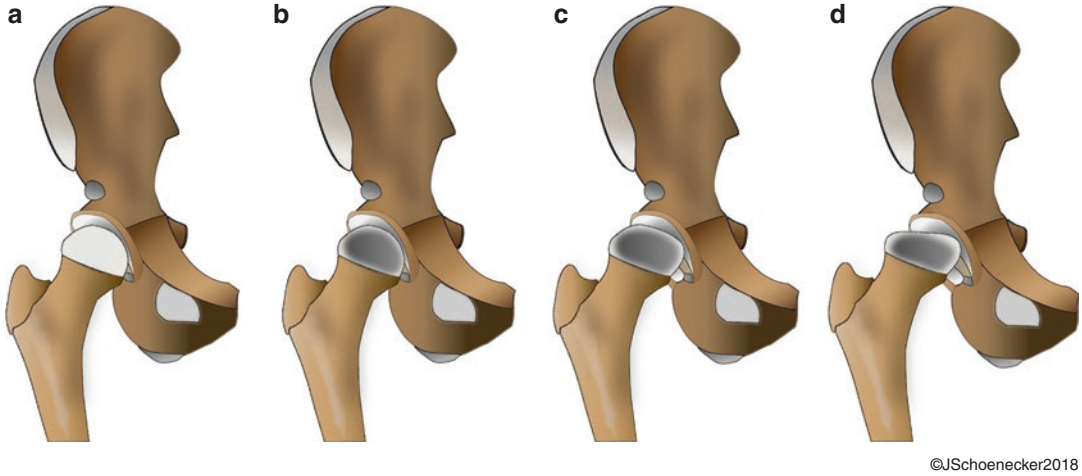

Fig. 6.2 Pathogenesis of femoral head deformation in LCPD. The normal femoral epiphysis (**a**) loses it blood supply (**b**) and the femoral head extrudes outside the acetabular margin (**c**). Weight-bearing stresses and muscular forces passing across the acetabular margin causes the femoral head to deform (**d**)

ease until skeletal maturity, and the third is from skeletal maturity to late adult life.

Onset of the Disease Until Healing

As the disease evolves, from the onset of avascular necrosis until complete revascularisation of the epiphysis, characteristic changes are visible on plain radiographs. On the basis of these radiographic changes, Waldenstrom divided the disease into four stages; the stages of avascular necrosis, fragmentation, reconstitution and the healed stage [22]. The first three of Waldenstrom's stages have been further subclassified into early and late phases for each—Stages Ia, Ib, IIa, IIb, IIIa and IIIb (Fig. 6.3) [14]. The average duration for the avascular necrosis, fragmentation, reconstitution stages is approximately 7, 8 and 18 months, respectively, while the duration of the sub-stages in the modified classification is approximately half of these respective durations. This modified staging of the disease is reproducible [23–25] and of importance in planning treatment.

Extrusion of the femoral head—defined by a lateral migration and loss of containment of the proximal femoral epiphysis—commences early in the disease process and gradually increases as it progresses from Stage Ia to Stage IIa. Thereafter, in Stage IIb, there is an abrupt increase in extrusion. Progressive widening of the femoral metaphysis is another phenomenon that is seen in untreated children. The pattern of progression of metaphyseal widening is almost identical to that seen with femoral extrusion; a modest increase in metaphyseal width occurs between Stage Ia and IIa after which there is a sudden increase in the width of the metaphysis. Metaphyseal width has been shown to accurately reflect the extent of epiphyseal flattening (i.e. "mushrooming") and the ultimate enlargement of the femoral head [14]. The timing of significant metaphyseal widening suggests that irreversible flattening and deformation of the femoral head has already occurred by the late stage of fragmentation or shortly thereafter [23].

Epiphyseal collapse, particularly of the lateral part of the epiphysis (referred to as the "lateral pillar") is another feature of LCPD that is of prognostic significance. The more the lateral pillar has collapsed, the poorer is the prognosis [26]. Treatment planning based on the extent of epiphyseal collapse outlined by Herring has been popular [27] but its value is limited by the fact that the extent of collapse of the lateral pillar can only be determined in Stage IIb. Waiting until stage IIb to apply the Herring grading and then planning

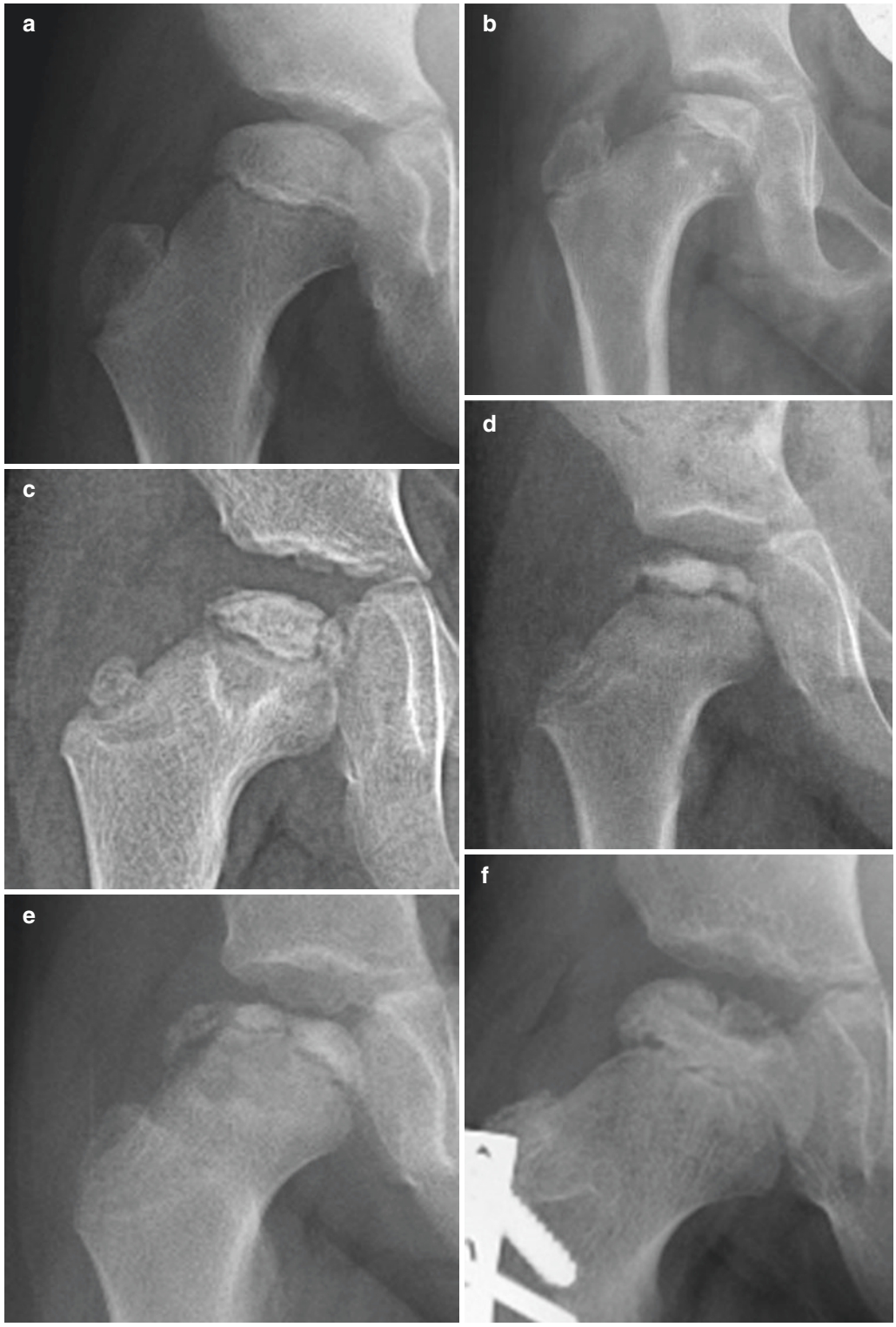

Fig. 6.3 Stages of evolution of LCPD (the modified Waldenstrom classification): (**a**) Stage Ia, (**b**) Stage Ib, (**c**) Stage IIa, (**d**) Stage IIb, (**e**) Stage IIIa, (**f**) Stage IIIb, (**g**) Stage IV

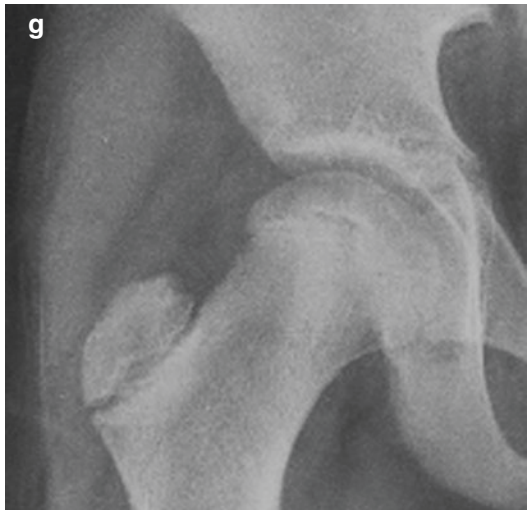

Fig. 6.3 (continued)

treatment especially in the older child is fraught with the risk of intervening too late, thereby missing the opportunity of preventing the femoral head from getting deformed.

The sequence of events described here is not seen if the onset of LCPD is in adolescence [28]. Adolescent LCPD has a very poor prognosis as collapse of the epiphysis occurs early, revascularisation and repair is often incomplete and permanent deformation is exceedingly common [28].

Healed Disease Until Skeletal Maturity

LCPD is considered to be completely healed once no more sclerotic avascular bone is visible in the epiphysis on the radiograph. Though it has been suggested that remodelling of the femoral head may occur between healing phase of the disease and skeletal maturity, it is clear that very little change, if any, occurs in the shape of the femoral head during this period [29]. Hence, it is imperative that appropriate treatment must be instituted early in the disease process to help ensure that the femoral head is spherical at the healed stage.

In children with extensive epiphyseal involvement, premature fusion of the capital femoral growth plate may occur which may become manifest only a few years after healing of the disease. This leads to an impairment in the growth of the femoral neck while the greater trochanteric apophysis continues to grow normally. As a result, at skeletal maturity, the femoral neck is short and the centre of the femoral head is at a level below the tip of the greater trochanter. This is referred to as coxa brevis (Fig. 6.4a) and the resulting altered mechanics of the hip will lead to a Trendelenburg gait due to a relative abductor insufficiency.

Skeletal Maturity to Late Adult Life

At skeletal maturity, the femoral head may be spherical with negligible increase in its diameter; such hips function well through adult life without developing secondary degenerative arthritis [30]. Hips with coxa magna (enlarged femoral head), coxa brevis (short neck) and coxa irregularis (misshapen femoral head) or a combination of these changes are prone to develop arthritis (Fig. 6.4a–c). Stulberg et al. classified hips of skeletally mature persons with healed LCPD into five classes based on the shape and size of the femoral head and congruency of the femoral head and the acetabulum (Fig. 6.5) [30]. Class I and II hips have spherical femoral heads that are congruent with the acetabulum; they do not develop arthritis. Class III and IV hips have non-spherical femoral heads but are still congruent with the acetabulum; they are likely to develop mild or moderate arthritis in late adult life. Class V hips are neither spherical nor congruent; they are likely to develop severe arthritis before the age of 50 years. A modified version of the Stulberg classification with just three classes has become popular recently [31, 32].

Epidemiology

The incidence of LCPD varies quite profoundly both between countries and within countries [33–35] ranging between 0.5 and 15 per 100,000 children under the age of 14 years. In the UK, the

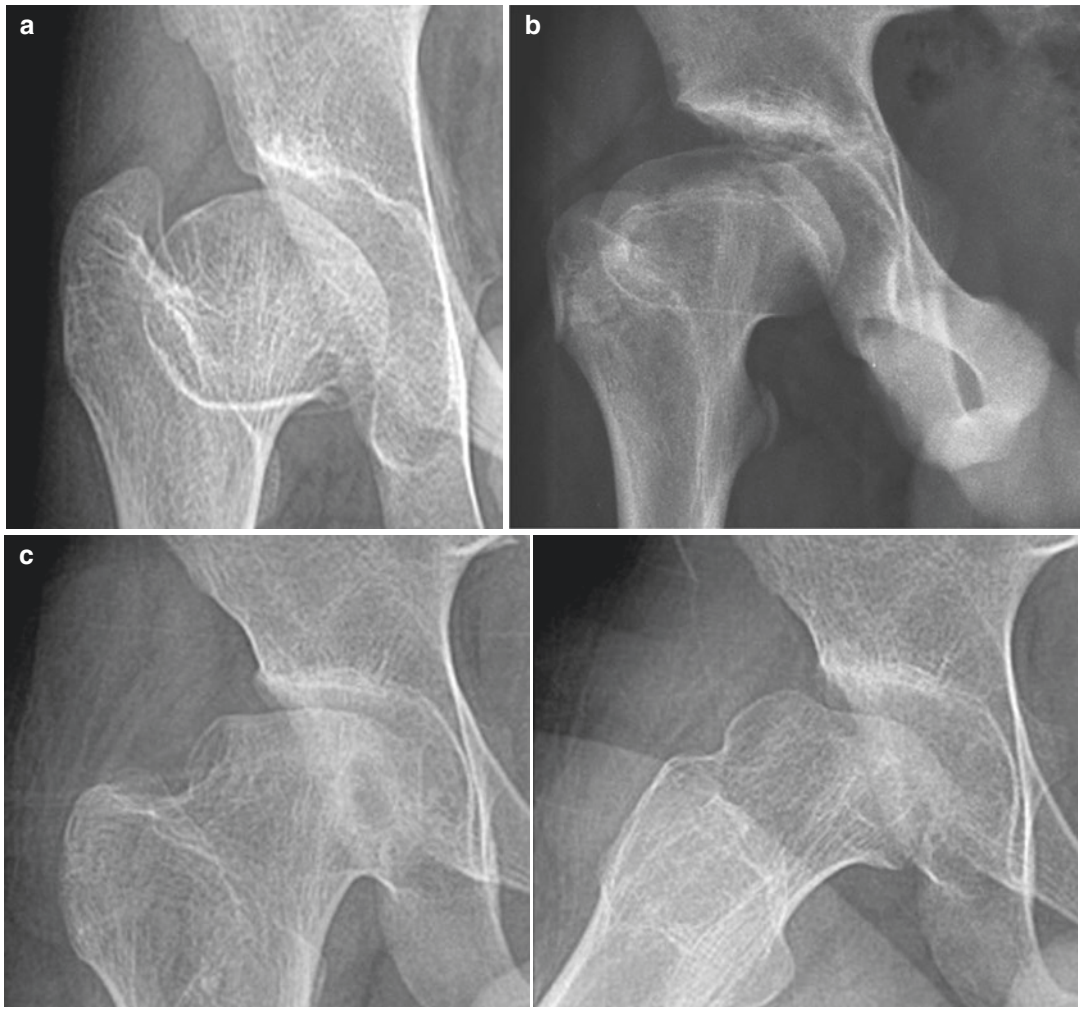

Fig. 6.4 Poor outcomes of LCPD at skeletal maturity; (**a**) coxa brevis, (**b**) coxa magna and (**c**) coxa irregularis

disease is more prevalent in urban, overcrowded and under-privileged regions while in south-west India the disease is relatively common in rural areas. Several studies suggest that LCPD is a disease of social deprivation [36–40]. The incidence of LCPD declined significantly in Merseyside in England and in Northern Ireland over the last three decades [37, 38, 41] as a concomitant improvement in living standards occurred in these regions over the same time period [42].

An association between LCPD and exposure to tobacco and wood smoke has also been demonstrated [43–45]. Maternal smoking during pregnancy, in particular, has also been shown to have a strong association with LCPD [39, 46]. Again, these observations support the association between LCPD and poverty as smoking is more prevalent among the socially deprived.

Some studies have suggested that there is an association between coagulation abnormalities such as thrombophilia and hypofibrinolysis and LCPD [47, 48] however, a definite [49] association has not been established [50].

LCPD occurs three or four times more frequently in boys than in girls. The peak age at onset of LCPD is 6 years in northern Europe while is between 8 and 9 years in India [34, 51]. The disease may affect children as young as 2 years and adolescents up to the age of 16 [28].

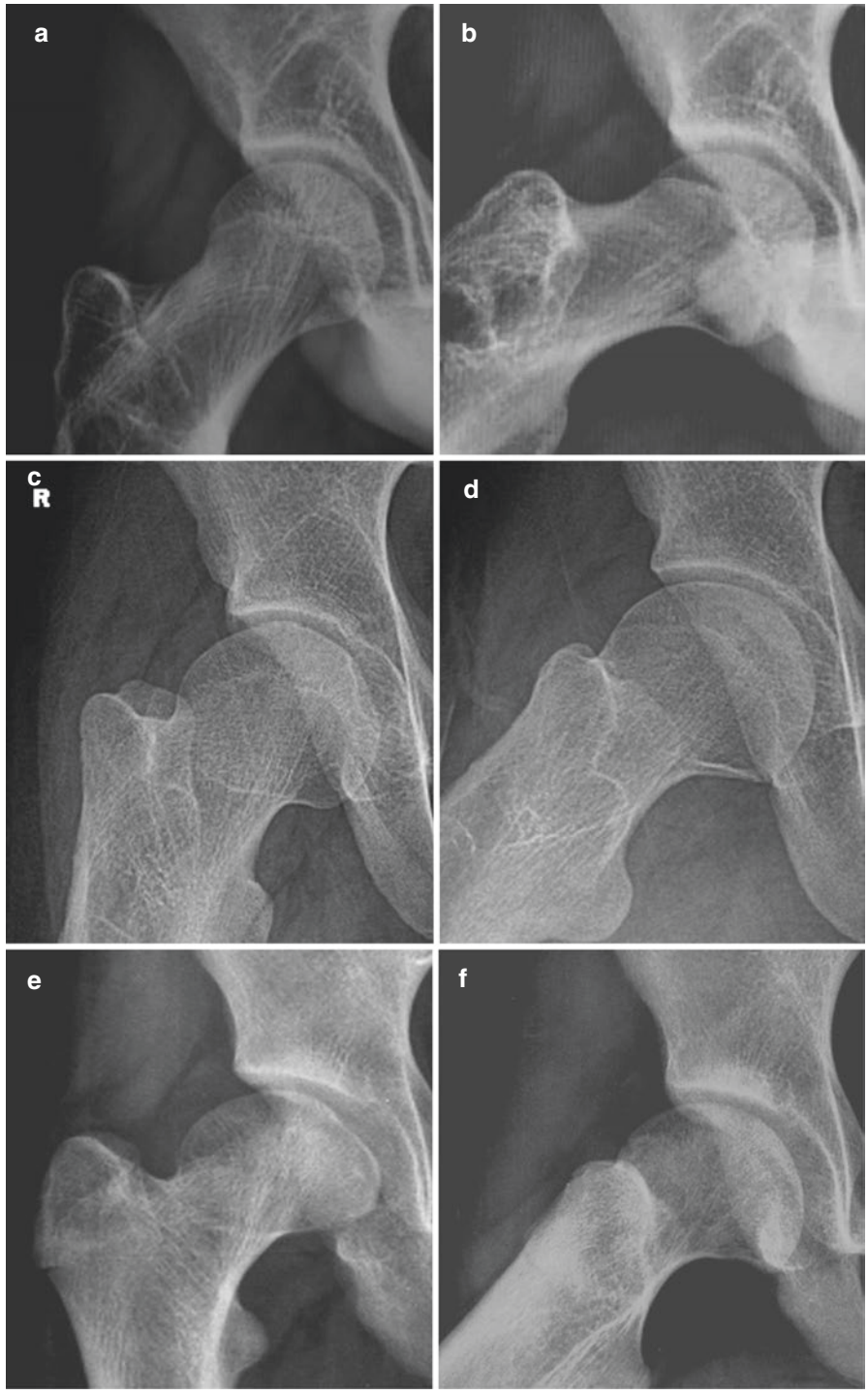

Fig. 6.5 Stulberg classification of the outcome of LCPD. Class I: The femoral head is spherical, the femoral is not enlarged, the femoral neck is not short and the joint is congruous (**a**, **b**). Class II: The femoral head is spherical, the femoral head is mildly enlarged, the femoral neck is slightly short and the joint is congruous (**c**, **d**). Class III: The femoral head is ovoid, the femoral neck is short and the joint is congruous (**e**, **f**). Class IV: The femoral head is irregular in shape, the femoral neck is short and the joint is congruous in the AP view (**g**, **h**). Class V: The femoral head is irregular and the joint is incongruous (**i**, **j**)

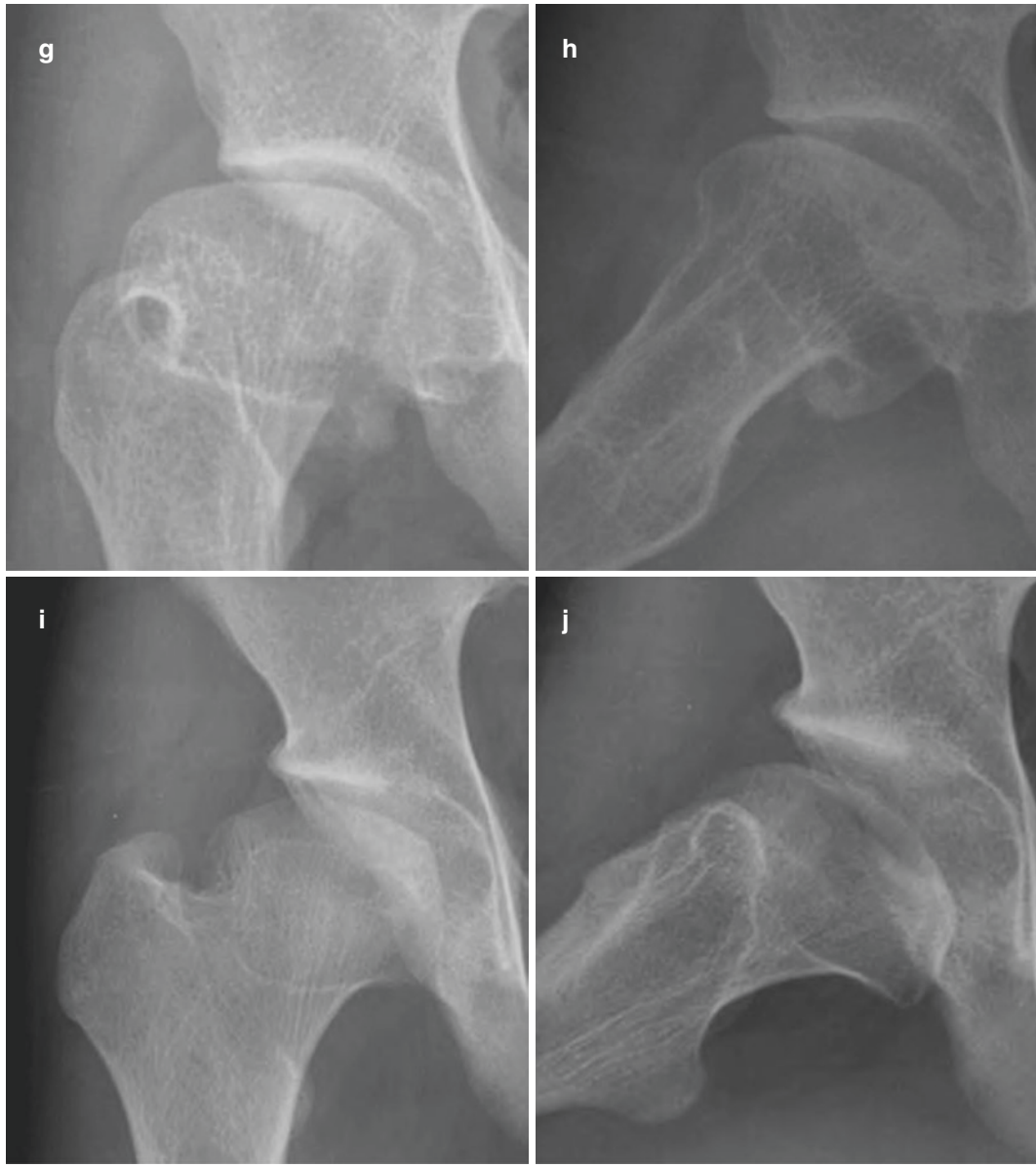

Fig. 6.5 (continued)

Clinical Presentation

The clinical presentation in the vast majority of instances is that of an otherwise healthy child complaining of mild pain in the hip, thigh or knee or having a limp that is noticed by the parents. Very often the symptoms are ignored by the parents for a while on the assumption that the child may have had some trivial injury at play. The pain is most commonly activity-related. There is no fever or any constitutional symptoms. The child may have a history of hyperactivity [52].

Range of motion examination will show moderate reduction in the range of passive abduction and internal rotation of the hip associated with some muscle spasm. Occasionally, all move-

ments may be limited by marked muscle spasm. Early in the disease, if the child is examined under anesthesia, the movement will be normal as the muscle spasm is relieved by the anesthetic. Once the femoral head is deformed limitation of movement may persist even under anesthesia.

A limp is usually present; in the early phase of the disease the gait is usually antalgic, later on when the mechanics of the hip is altered or the epiphyseal growth is affected a Trendelenburg gait or a short-limb gait may develop.

Bilateral involvement is infrequent but when it does occur the disease onset is seldom simultaneous in both hips. When imaging shows features suggestive of bilateral synchronous disease, skeletal dysplasia, Gaucher's disease and hypothyroidism must be excluded as radiographic features of the hips in these conditions may resemble LCPD.

The hip pain tends to reduce as the disease progresses in most of the children. Occasionally, recurrence of pain and sudden reduction in the range hip motion may develop as the femoral head deforms during the advanced stage of fragmentation (Stage IIb). In these cases, attempted hip abduction causes pain and a radiograph taken in abduction may reveal a phenomenon called 'hinge abduction' where the lateral aspect of the femoral head impinges against the acetabular margin.

Essential Clinical Signs

- Antalgic gait initially
- Trendelenburg sign later in the disease
- Reduced range of passive abduction and internal rotation

Imaging

A diagnosis of LCPD can be made on the basis of plain radiographic appearances in the majority of instances. The changes seen in the radiographs will vary with the stage of evolution of the disease, but one consistent feature throughout the course of the disease is the presence of at least some sclerotic bone in the epiphysis (Fig. 6.6). The changes noted on plain radiographs in the femoral epiphysis, metaphysis and the acetabulum are shown in Table 6.1. An AP pelvis radiograph with the hips in abduction and internal rotation can help determine whether the femoral head can be contained or not. The presence of femoral extrusion with inability to achieve complete coverage of the femoral epiphysis on abduction by the acetabular roof and a crescent-shaped joint space in the abduction radiograph (widest medially and narrowest supero-laterally) is suggestive of hinge abduction (Fig. 6.7).

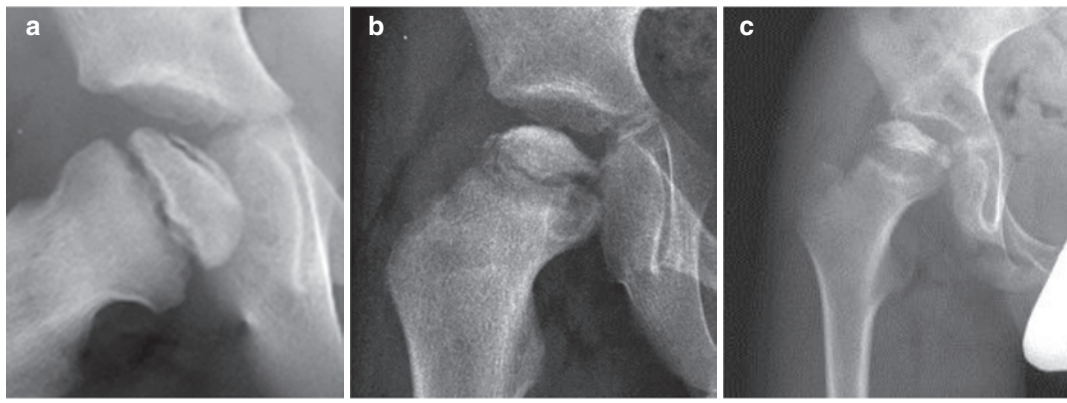

Fig. 6.6 Plain radiographic changes in the epiphysis, metaphysis and the acetabulum in LCPD; (**a**) sub-chondral fracture line, (**b**) metaphyseal cyst, (**c**) diffuse metaphyseal reaction, (**d**) femoral head extrusion, femoral metaphyseal widening and acetabular bicompartmentalisation and widening

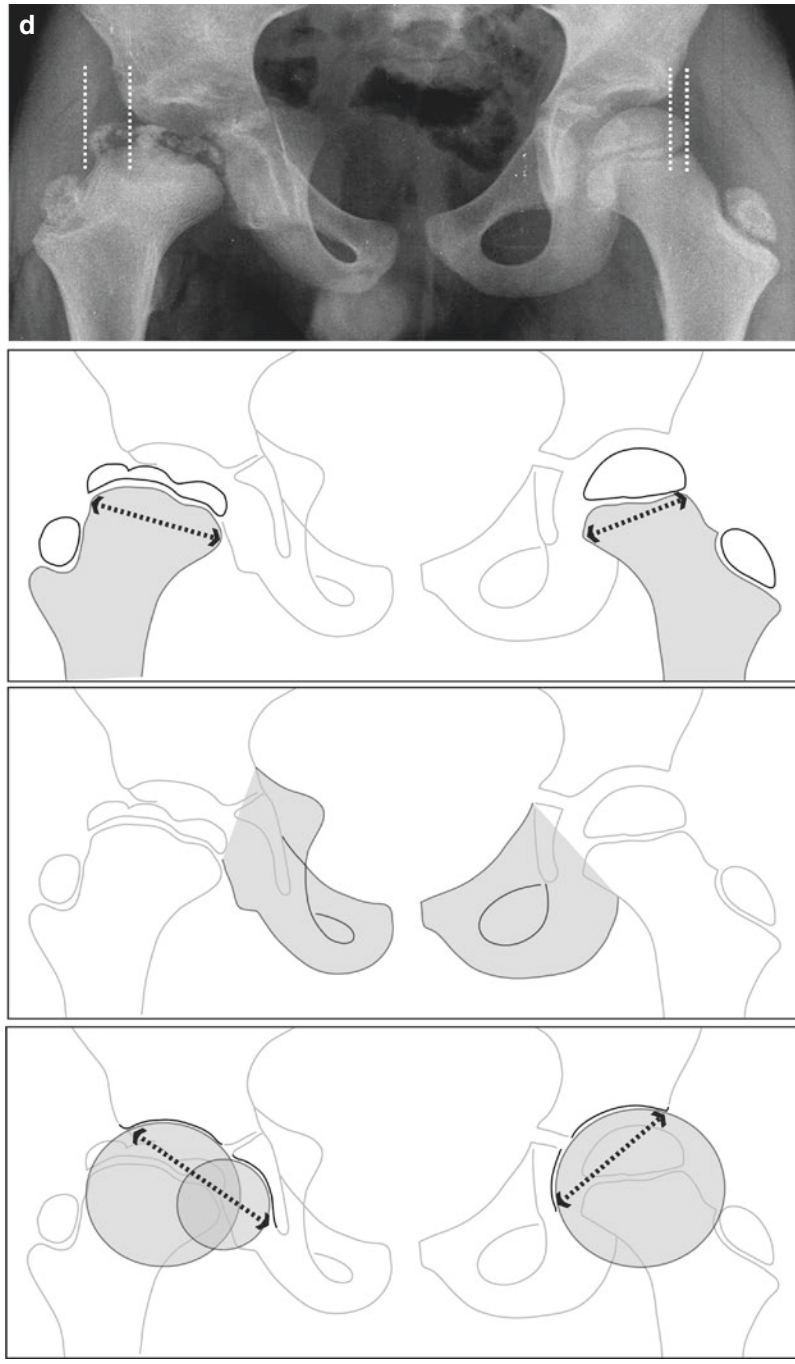

Fig. 6.6 Continued

"Plain radiographs of the pelvis showing both hips must be obtained if a diagnosis of LCPD is suspected"

MRI scans can detect avascular changes in the epiphysis before it is evident on plain radiographs and hence it is useful as a diagnostic modality very early in the disease. MRI perfusion scans with gadolinium enhancement and fat suppression sequencing can quantify the extent of avascularity of the femoral epiph-

Table 6.1 Plain radiographic changes in the proximal femur and the acetabulum in LCPD

Site	Radiographic abnormality	
Femoral epiphysis	Sclerosis	First abnormality to appear Persists till disease heals (though area reduces as disease progresses)
	Sub-chondral fracture[a]	Runs parallel to the articular margin Extent of the fracture line reflects the extent of the avascular segment of the epiphysis Seen very early in the disease (Stage Ia or Ib) but is seen only about one third of children
	Loss of epiphyseal height (collapse)	Often most marked in the lateral part of the epiphysis Collapse increases from Stage Ib until Stage IIb
	Fragmentation of the epiphysis[b]	Occurs as fissures perpendicular to the articular surface Fragmentation increases from Stage IIa to Stage IIb
	New bone formation on the lateral and medial margins of the avascular epiphysis	Initially woolly texture (woven bone); later mature bone of normal texture New bone gradually replaces all the necrotic bone
Femoral metaphysis	Metaphyseal cyst[a,c]	Usually appears once fragmentation has begun Spontaneously resolves by Stage IIIb
	Diffuse metaphyseal osteoporosis[a,c]	Develops when epiphyseal fragmentation is advanced and resolves by Stage IIIb
	Widening of the metaphysis[a]	Starts in Stage Ib and gradually increases till Stage IIb after which widening increases markedly Is a measure of epiphyseal flattening and ultimate size of the femoral head
Acetabulum	Osteoporosis of the acetabular roof[a]	Seen early in the disease
	Bi-compartmentalisation of the acetabulum[a]	Always associated with extrusion
	Premature fusion of the triradiate cartilage[a]	Becomes manifest after the disease has healed
	Dimensional alterations (wider and shallower)	May be seen early Most marked when the femoral head is enlarged
Articular joint space	Increased medial joint space	Starts early and progressively increases particularly after Stage IIa
	Crescent shaped joint space on abducting the hip[a]	Associated with hinge abduction
Femur–acetabular relationship	Femoral head extrusion[b,c]	More frequent in older children and when more of the epiphysis is avascular If it develops it progresses as disease evolves, particularly after Stage IIa

[a]These changes may not develop in all children
[b]These changes may be modified by treatment
[c]Catterall head-at-risk signs which are poor prognostic indicators (in addition to these, he suggested that the Gage sign and a horizontal growth plate are also head-at-risk-signs)

ysis as soon the disease is diagnosed (Fig. 6.8). This is particularly useful as recent studies have based treatment decisions on how much of the epiphysis is devoid of blood supply (e.g. greater or less than 50% involvement) [53–55]. The use of treatment algorithms based on MRI perfusion scans is still under investigation at present.

Arthrography is useful during the course of treatment. In particular, dynamic arthrography performed under anaesthesia can identify alterations in the contour of the cartilaginous portion of the femoral head and congruency of the joint surfaces in different positions of the hip. Arthrography is also useful to confirm the presence of hinge abduction where dye pools medially as the hip is abducted (Fig. 6.9).

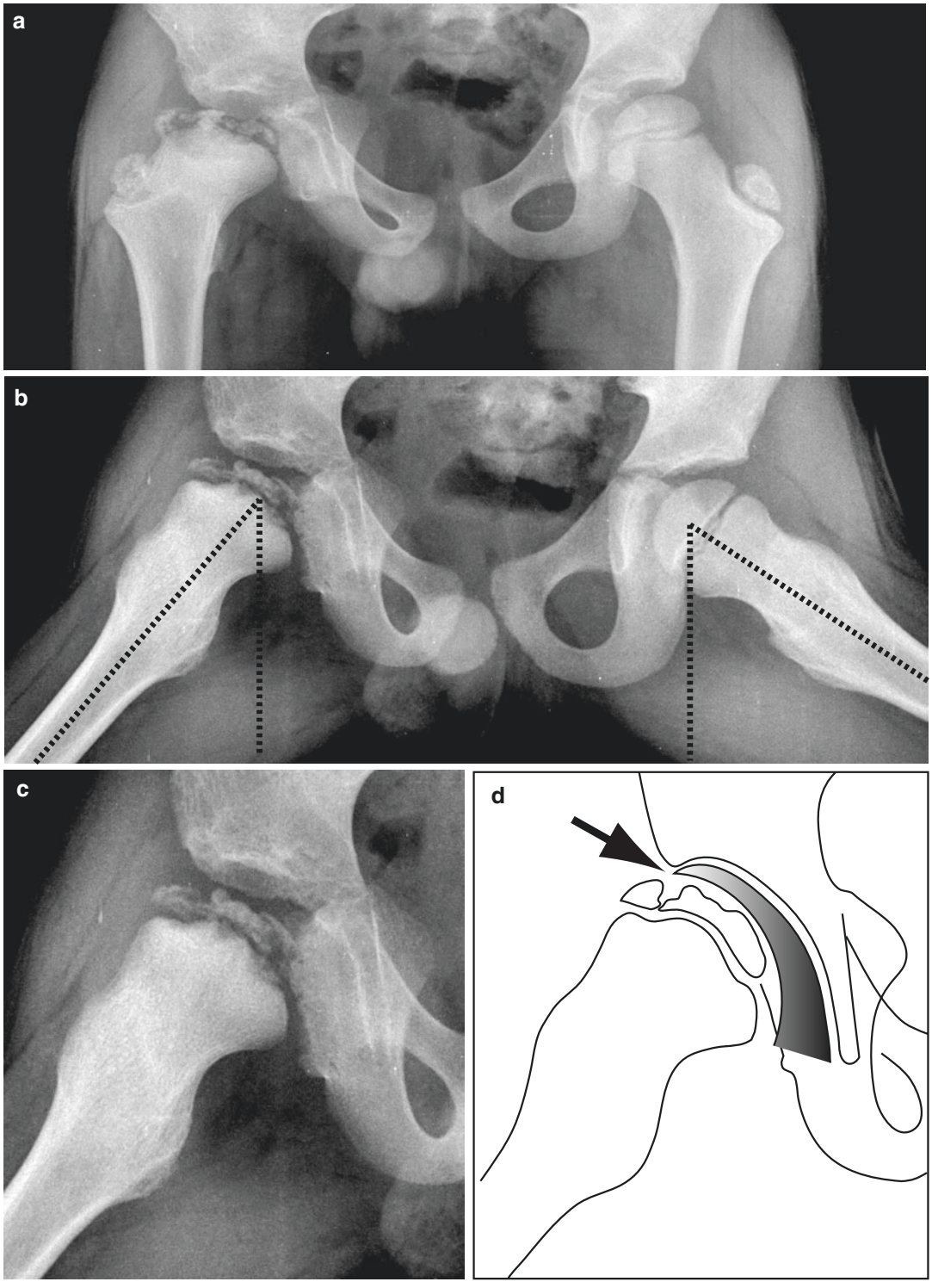

Fig. 6.7 Plain radiographs of hinge abduction of a child with LCPD in Stage IIIa of the disease; (**a**) X-ray of the hips in the neutral position—note the extrusion and the joint space, (**b**) X-ray of the hips in abduction—note the limited abduction in the affected hip. Also note that the joint space is crescent-shaped; wide medially and narrow supero-laterally (**c**, **d**)

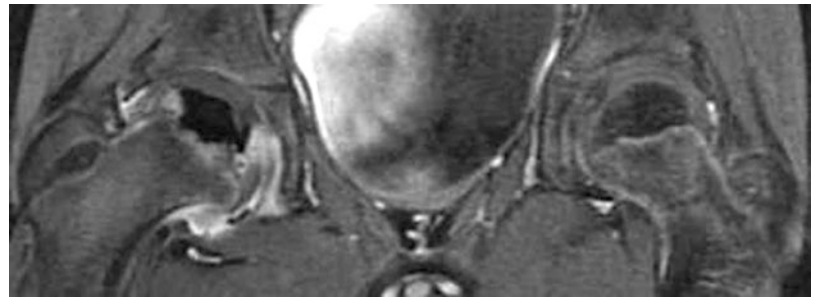

Fig. 6.8 MRI perfusion scan of a child with LCPD. The avascular area (black) in the femoral capital epiphysis is seen in the right hip

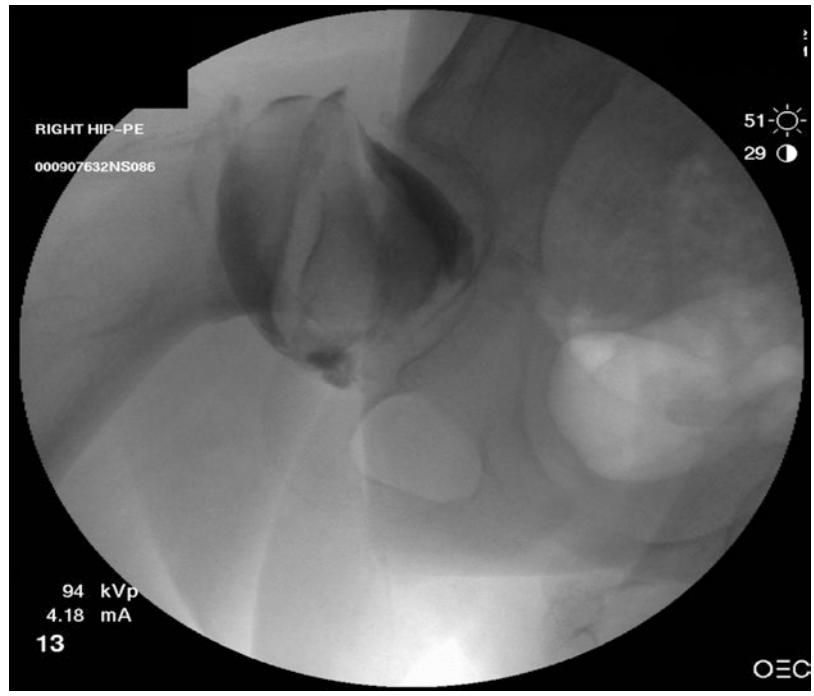

Fig. 6.9 Arthrogram of the hip of a child with LCPD and hinge abduction. Pooling of dye medially in the abducted position (courtesy Prof. In Ho Choi, Seoul)

Essential Imaging

- Plain radiographs of the pelvis (AP and frog-lateral views)
- Perfusion MRI scan
- Arthrogram

Management

If the child presents early in the course of the disease (i.e. before the femoral head has begun to deform), the aim of treatment would be to prevent subsequent femoral head deformation. If the child presents later in the course of the disease, the aim would be to minimise the progression of early femoral head deformation. Once irreversible deformation is established, the aim is to alleviate symptoms caused by the deformity and attempt to delay the onset of degenerative arthritis.

"The management of LCPD will depend on when during the course of the disease the child presents; the aim of treatment and the treatment options will vary accordingly"

Treatment Early in the Course of the Disease (Stage Ia to Stage IIa)

As it is assumed that weight-bearing stress leads to collapse of the epiphysis, for several years, weight relief had previously been advocated. Despite this practice, there is no evidence that weight relief alone prevents epiphyseal collapse. However, there is some clinical and experimental evidence that if weight relief is combined with other forms of treatment it may be beneficial [56, 57].

Containment attempts to ensure that weight-bearing and muscular forces are not imparted from the acetabular margin onto the anterolateral part of the avascular epiphysis that is most vulnerable to deformation during the early stages of the disease process.

> ***"Since the prime factor that leads to femoral head deformation is extrusion, treatment early in the disease is directed towards reversing or pre-empting extrusion by achieving containment of the entire femoral head by the acetabulum"***

There are two strategies for containment; the first is to keep the hip effectively abducted and internally rotated (or abducted and flexed)—either by bracing, casting, or femoral osteotomy—thereby ensuring that the anterolateral part of the femoral head is positioned well within the acetabulum. The second is to either reorient the acetabulum by a periacetabular osteotomy or augment the acetabulum by a shelf acetabuloplasty so that anterolateral part of the femoral head is well covered by the re-aligned acetabulum or by the newly created "shelf".

The factors that need to be considered while planning treatment in the early in the course of the disease are the age of the child at onset of the disease, the extent of epiphyseal avascularity and the presence or absence of femoral head extrusion. The prognosis is very good in children in whom less than half the epiphysis is infracted and consequently they may be treated symptomatically [1]. The prognosis is good in young children (under 8 years of age at the onset); they do well even if more than half the epiphysis is avascular as long as they do not develop extrusion. If extrusion occurs, containment is warranted.

Table 6.2 Indications for containment early in the course of LCPD (i.e. in Stage Ia, Ib or IIa)[a]

Age at onset of the disease	Extrusion of the femoral head	Recommended treatment
Under 8 years of age at onset of the disease	Absent	No containment (4-monthly follow up is needed to ensure that extrusion does not occur)
	Present	Containment indicated as soon as extrusion is recognised
8–12 years of age at onset of the disease[b]	Present or absent (extrusion invariably will occur sooner or later in these older children)	Containment indicated

[a]Catterall's grouping and head-at risk signs and Herring's lateral pillar classification are not considered here for decision-making as these classifications can only be reliably made in Stage IIb

[b]Containment treatment is not recommended if the onset of the disease is in adolescence (over 12 years of age) as it is ineffective

In short, containment may not be required in two groups of children; first, young children with mild disease and no extrusion and second, children who will not benefit from it because it is too late. The indications for containment are outlined in Table 6.2.

> ***"Children over the age of 8 years at onset of the disease almost invariably develop femoral head extrusion sooner or later [14, 58]. In these older children extrusion should be pre-empted by "containing" the hip as soon as the disease is diagnosed since the likelihood of a poor result is 16 times greater if containment is deferred"***

Containment by Non-operative Means

Braces that hold the hip abducted and internally rotated or abducted and flexed can effectively contain the hip [59–62]. To improve the chances of success, the brace needs to be worn constantly till the disease progresses beyond the stage where there is a risk of femoral head deforma-

tion (i.e. until Stage IIIb). Since the brace may have to be worn for as long as 18 months, patient compliance is imperative. Though some surgeons may be sceptical about the efficacy of bracing for this reason, Schoenecker and his colleagues have reported very salutary results with a carefully supervised protocol of physiotherapy to maintain the range of hip motion and abduction bracing [59].

Essential Non-operative Treatment
- Weight-relief
- Abduction bracing

Non-operative Pitfalls
- Poor patient compliance

Containment by Surgery

Femoral varus de-rotation osteotomy is by far the most frequently performed operation to achieve containment in LCPD [6, 63–66]. Moderate varus angulation of 20° and 20–30° of external rotation of the distal fragment is sufficient to achieve satisfactory containment [67]. The osteotomy may be either at the intertrochanteric or subtrochanteric level and the femur can be fixed with any one of the commonly available implants (DCP plate, blade plate, proximal locking plate). An open wedge osteotomy will minimise the extent of shortening caused by the varus angulation (Fig. 6.10a). The potential disadvantages of performing a proximal femoral varus osteotomy with an opening wedge technique include delay in union of the osteotomy, permanent shortening of the limb and compensatory angular deformity at the knee. However, these complications are not commonly seen in practice. The open wedge does not compromise healing of the osteotomy; delayed union is virtually never seen even in older children [67]. The shortening decreases as the child grows and at skeletal maturity the limb length inequality is inconsequential; usually it is around 0.5 cm [68]. Though genu valgum may develop in some other situations where there is coxa vara [69], genu valgum was not observed in children with LCPD who had undergone varus osteotomy of the femur [70].

One of the prerequisites for surgical containment is restoration of the range of motion of the hip.

In some children limitation of internal rotation of the hip may persist for some time. In these children, a femoral varus extension osteotomy should be done rather than a varus de-rotation osteotomy.

"One of the prerequisites for surgical containment is restoration of the range of motion of the hip"

Acetabular realignment with improved containment of the femoral head can be achieved by a Salter osteotomy (Fig. 6.10c) [71] or a triple pelvic osteotomy [72]. Augmenting the acetabulum with a shelf is also an effective way of achieving containment (Fig. 6.10b) [73–76]. The results of containment by operating on the acetabulum is as effective as containment by a femoral osteotomy [77–79].

Some surgeons have advocated combining femoral osteotomy with an acetabular operation anticipating better results than if surgery was done on either the femur or the pelvis alone [80–83]. However, there isn't sufficient evidence to support such an approach [84].

Irrespective of the method of containment employed, it must be achieved by Stage IIa of the disease if it is to be effective in preventing deformation of the femoral head [23, 85].

Preventing Trochanteric Overgrowth

Premature growth arrest of the proximal femoral physis and consequent trochanteric overgrowth appears to be more common in the older child—in whom a major part of the epiphysis is avascular—but this complication cannot be reliably predicted. Prophylactic trochanteric epiphyseodesis by a combination of drilling of the physis and screw epiphyseodesis is effective in reducing the frequency of trochanteric overgrowth (Figs. 6.11 and 6.12) [68]. The effectiveness of this procedure reduces as the child gets older but even in children as old as 10 years at least half of the procedures can be expected to succeed [68].

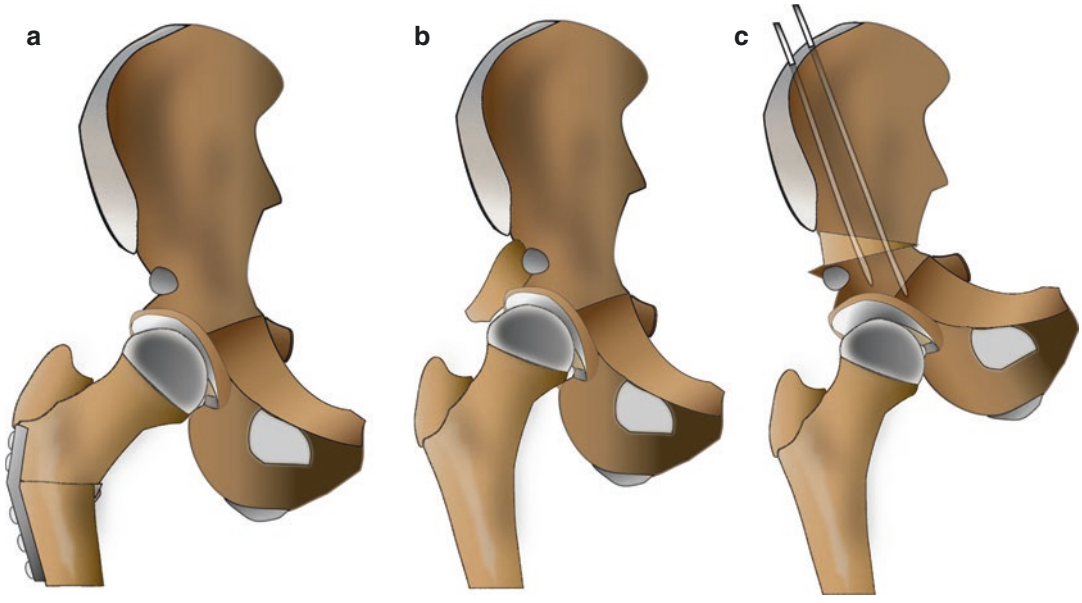

Fig. 6.10 Methods of surgical containment in LCPD include proximal femoral varus osteotomy (**a**), shelf acetabuloplasty (**b**) and re-directional innominate osteotomy (**c**) (e.g. Salter osteotomy)

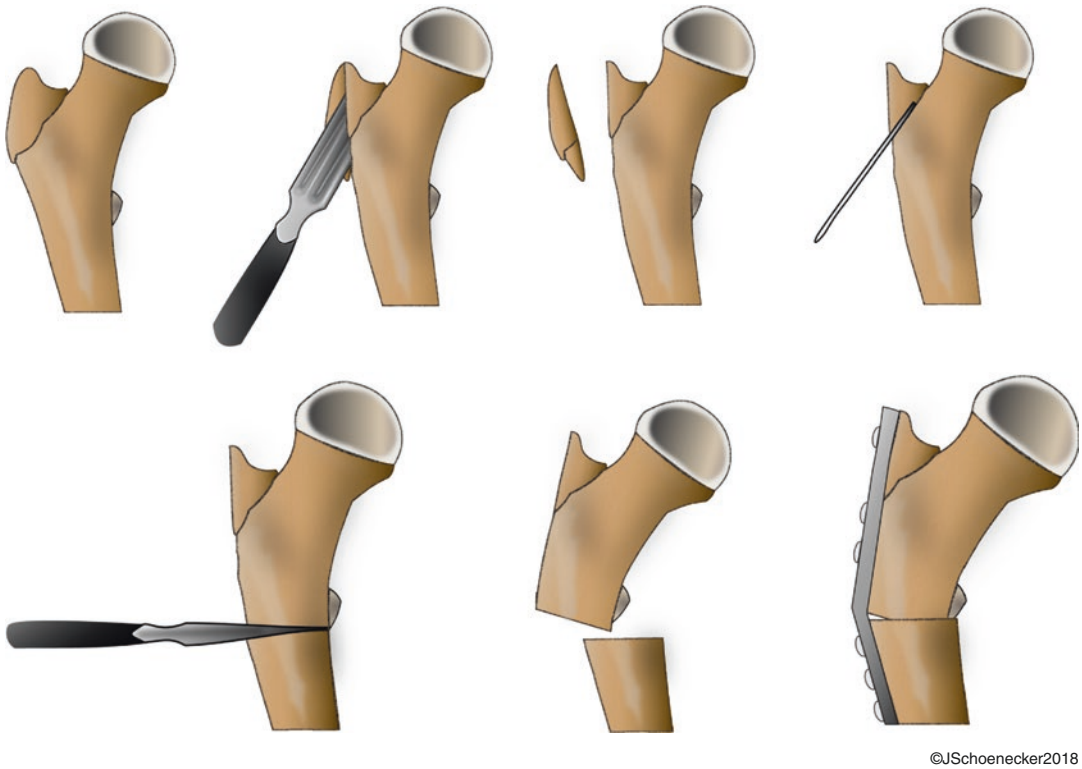

Fig. 6.11 Technique of trochanteric epiphyseodesis combine with a proximal femoral varus osteotomy

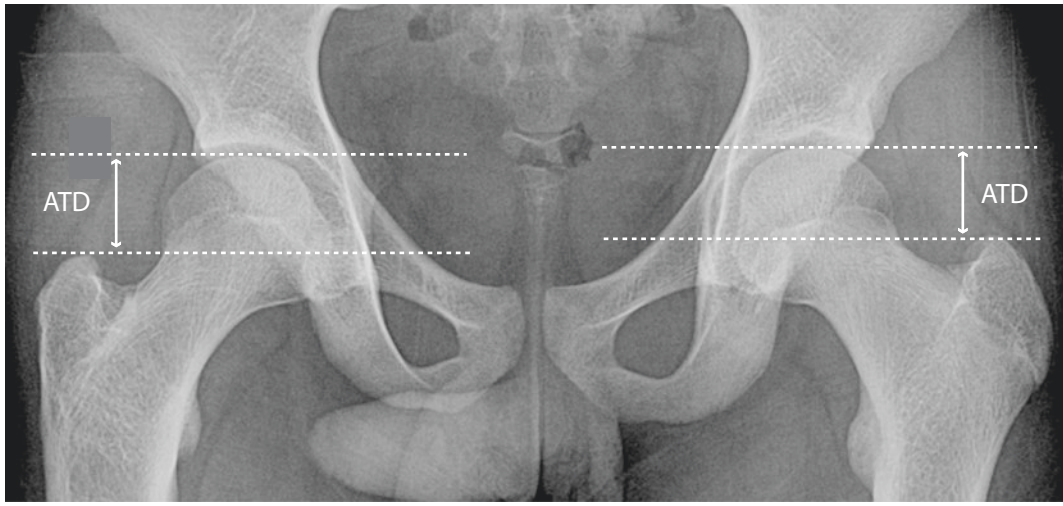

Fig. 6.12 Favourable outcome of a child with LCPD of the right hip treated by a proximal femoral varus de-rotation osteotomy and prophylactic trochanteric epiphyseodesis

"Prophylactic trochanteric epiphysiodesis by a combination of drilling of the physis and screw epiphysiodesis is effective in reducing the frequency of trochanteric overgrowth"

Treatment Late in the Course of the Disease (Stage IIb or Stage IIIa)

By the time the disease reaches Stage IIb the chances of preventing femoral deformation are low and so containment at this point of the disease may have only limited value [23, 86]. If there is exacerbation of pain and reduction in hip motion, hinge abduction should be suspected. Examination under anaesthesia and dynamic arthrography should be done to confirm hinge abduction. If the hip is found to be more congruent in adduction, a proximal femoral valgus osteotomy is indicated. This will overcome impingement of the femoral head on the acetabular margin and also relieve pain [87–89]. The Bernese group have advocated for using a femoral head reduction procedure via a surgical hip dislocation approach (with or without a concomitant acetabular procedure) for the treatment of hinge abduction secondary to an enlarged femoral head even during the fragmentation phase [90]. Though favourable early outcomes have been reported, long term outcomes have yet to be determined.

Treatment After Established Femoral Head Deformation (Stage IIIb and Stage IV)

The patients with greater trochanteric overgrowth and coxa brevis almost invariably have coxa magna and many have frankly aspherical femoral heads; some may have ovoid heads and a few will have spherical femoral heads. The author recommends this operation only for patients who have a spherical or ovoid femoral head since these hips have a lower chance of developing degenerative arthritis prematurely and are the ones that are likely to benefit most from the operation.

Attempts are now being made to reshape deformed femoral heads at skeletal maturity by surgical dislocation of the hips with the hope that late degenerative arthritis may be avoided but such long term results are not yet available [91].

"In children with coxa brevis, greater trochanteric overgrowth and a Trendelenburg gait, advancement of the greater trochanter

distally and laterally can improve the mechanics of the hip, reduce stresses on the hip and abolish the Trendelenburg gait"

Essential Surgical Techniques

- Containment surgery early in the disease (Stage Ia, Ib, IIa)
  - Femoral varus de-rotation osteotomy
  - Femoral varus extension osteotomy
  - Redirectional innominate osteotomy (single or triple)
  - Acetabular shelf operation
- Surgery to prevent trochanteric overgrowth (Stage Ia, Ib, IIa)
  - Trochanteric epiphyseodesis
- Surgery for hinge abduction late in the disease (Stage IIb or IIIa)
  - Proximal femoral valgus osteotomy
- Surgery for established trochanteric overgrowth (Stage IV)
  - Trochanteric advancement

Operative Pitfalls

- Delay in containment—may not be effective in preventing femoral head deformation
- Excessive varus of femoral osteotomy—may result in excessive shortening of the femur, persistent abductor lurch, and failure of varus to remodel
- Excessive rotation of fragment of acetabulum—may cause femoroacetabular impingement

Classic Papers

Axer A. Subtrochanteric Osteotomy in the Treatment of Perthes' Disease: A Preliminary Report. The Journal of bone and joint surgery British volume. 1965;47:489–99. Axer, for the first time, explained the rationale of a proximal femoral varus derotation osteotomy in LCPD and reported early results in a small group of children.

Catterall A. The natural history of Perthes' disease. The Journal of bone and joint surgery British volume. 1971;53 (1):37–53. Catterall classified LCPD in to four groups based on the extent of the epiphysis that was rendered avascular. In Group I hips about one-fourth of the epiphysis is avascular, half the epiphysis is avascular in Group II hips, more than half the epiphysis is avascular in Group III hips, and the entire epiphysis is avascular in Group IV hips. Catterall noted that the outcome was best in Group I hips and poorest in Group IV hips. He also defined "head-at-risk" signs and suggested that treatment is warranted if any of these signs were present.

Stulberg SD, Cooperman DR, Wallensten R. The natural history of Legg-Calve-Perthes disease. The Journal of bone and joint surgery American volume. 1981;63 (7):1095–108. Stulberg and his colleagues followed up patients with healed LCPD for 30–40 years. The patients could be placed into one of five classes based the shape of the femoral head and congruency between the femoral head and the acetabulum. Three types of congruency were recognized: (1) spherical congruency (Class-I and II hips)—for hips in this category, arthritis did not develop; (2) aspherical congruency (Class-III and IV hips)—mild to moderate arthritis develops in late adulthood in these hips; and (3) aspherical incongruency (Class-V hips)—severe arthritis develops before the age of 50 years in these hips.

Herring JA, Neustadt JB, Williams JJ, Early JS, Browne RH. The lateral pillar classification of Legg Calve-Perthes disease. Journal of pediatric orthopedics. 1992;12 (2):143–50. Hips were classified during the fragmentation stage of disease into three groups (A, B and C) based on the extent of collapse of the lateral pillar of the femoral head. At skeletal maturity, the outcome was determined according to the Stulberg classification. Group A hips with no collapse of the lateral pillar had a uniformly good outcome (100% Stulberg I and II); Group B hips with less than 50% collapse had a good outcome in patients who were less than 9 years at onset (92% Stulberg I and II), but a poorer outcome in patients who were older than 9 years at onset (30% Stulberg II). In Group C, the majority of femoral heads became aspherical.

Key Evidence

Evidence for the Timing of Femoral Head Deformation

Joseph B, Varghese G, Mulpuri K, Narasimha Rao K, Nair NS. Natural evolution of Perthes disease: a study of 610 children under 12 years of age at disease onset. Journal of pediatric orthopedics. 2003;23 (5):590–600. Based on 2634 radiographs of 610 children with LCPD, the disease was divided into seven stages (stages Ia, Ib, IIa, IIb, IIIa, IIIb, and IV). The new classification system of the evolution of Perthes disease helped to identify when crucial events occur during the course of the disease. Epiphyseal extrusion and metaphyseal widening were modest in stages Ia, Ib, and IIa but increased dramatically after stage IIb. More than 20% extrusion occurred in 70% of the hips by stage IIIa. Metaphyseal changes were most frequently encountered in stage IIb, while acetabular changes were most prevalent in stage IIIa. The timing of epiphyseal extrusion, metaphyseal widening, and the appearance of adverse metaphyseal and acetabular changes suggest that femoral head deformation occurs by stage IIIa in untreated hips.

Evidence for Optimal Timing of Containment

Joseph B, Nair NS, Narasimha Rao K, Mulpuri K, Varghese G. Optimal timing for containment surgery for Perthes disease. Journal of pediatric orthopedics. 2003;23 (5):601–6. Outcomes following femoral osteotomy of 97 children were analyzed. Univariate and multivariate analyses identified variables that influenced the shape and size of the femoral head at healing. The chances of retaining a spherical femoral head were much higher in children operated on either during the stage of avascular necrosis or in the early part of the fragmentation stage (Stage Ia, Ib, or IIa) than in those operated later. The authors conclude that containment surgery aimed at preventing femoral head deformation in Perthes disease should be performed before the late stage of fragmentation (Stage IIb).

Evidence for the Performing Prophylactic Trochanteric Epiphyseodesis

Shah H, Siddesh ND, Joseph B, Nair SN. Effect of prophylactic trochanteric epiphyseodesis in older children with Perthes' disease. Journal of pediatric orthopedics. 2009;29 (8):889–95. Outcomes of 62 children with unilateral LCPD who underwent trochanteric epiphyseodesis combined with varus osteotomy of the femur during the active stage of the disease (mean age at surgery: 8.4 years) were compared with 20 controls. At skeletal maturity, the articulotrochanteric distance and center-trochanteric distance were greater and the frequency of a positive Trendelenburg sign was less in children who had undergone trochanteric epiphyseodesis than in children who had no surgery ($P < 0.01$). A probability curve plotted on the basis of a logistic regression model suggests that effective trochanteric arrest may be achieved in a high proportion of children operated at or before 8.5 years of age and in half the children operated between the age of 8.5 and 10 years.

Evidence for Performing Containment Surgery (Prospective Studies)

Herring JA, Kim HT, Browne R. Legg-Calve-Perthes disease. Part II: Prospective multicenter study of the effect of treatment on outcome. The Journal of bone and joint surgery American volume. 2004;86-A (10):2121–34. 438 patients between the ages of 6 and 12 were enrolled in a prospective multicenter study who were stratified into five treatment groups consisting of: no treatment, brace treatment, range-of-motion exercises, femoral osteotomy, and innominate osteotomy. 337 patients were available for follow-up at skeletal maturity. There were no differences in outcome among the hips with no treatment, those treated with bracing, and those treated with range-of-motion exercises. The lateral pillar classification and the age at onset of the disease had a strong cor-

relation with outcome. Patients who are over the age of 8 years at onset with lateral pillar B group or B/C border group had a better outcome with surgical treatment than with nonoperative treatment. Group-B hips in children who are less than 8 years of age at onset had favorable outcomes irrespective of treatment. Group-C hips in children of all ages often had poor outcomes.

Wiig O, Terjesen T, Svenningsen S. Prognostic factors and outcome of treatment in Perthes' disease: a prospective study of 368 patients with five-year follow-up. The Journal of bone and joint surgery British volume. 2008;90 (10):1364–71. 152 children with unilateral LCPD over 6 years of age at diagnosis and with more than 50% necrosis of the femoral head were treated by one of three methods of treatment: physiotherapy (55 patients), the Scottish Rite abduction orthosis [26], and proximal femoral varus osteotomy [71]. Proximal femoral varus osteotomy gave a significantly better outcome than orthosis ($p = 0.001$) or physiotherapy ($p = 0.001$). There was no significant difference between the physiotherapy and orthosis groups ($p = 0.36$). The authors recommend proximal femoral varus osteotomy in children 6 and over at onset with more than 50% femoral head necrosis. They also recommend that bracing should not be used.

Take Home Messages

- The aim of treatment of LCPD is to prevent the femoral head from getting deformed
- Weight-bearing forces passing across the acetabular margin onto the extruded femoral head, at a time when the epiphyseal bone is weak, can cause the femoral head to deform
- Reversal of extrusion or preventing extrusion by containment in Stage Ia, Ib or IIa improves the chances of preventing femoral head deformation and retaining the spherical shape of the femoral head
- Any treatment offered after Stage IIa may be ineffective in preventing deformation of the femoral head

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