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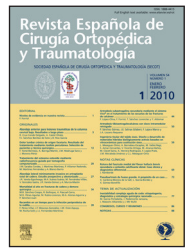

UPDATE

Legg-Calvé-Perthes disease

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KEYWORDS
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Abstract Perthes' disease is an idiopathic avascular necrosis of the proximal femoral epiphysis in children. Plain radiograph is the main diagnosis tool and is also used to classify the phase and the extent of the disease. Although it is a self-limited condition and its natural history is often benign, there are several factors that can worsen prognosis. Those factors are age, extent of the disease and some radiological signs and are useful to identify patients who will benefit the most from treatment. Adequate containment of the femoral head within the acetabulum resulting in a more spherical and congruous joint is the main goal of treatment, which is based mainly on surgical techniques, such as femoral or acetabular osteotomies. It has been shown that orthopaedic containment methods, like abduction orthosis, have not achieved better results.

PALABRAS CLAVE
Enfermedad de Legg-Calvé-Perthes;
Necrosis isquémica;
Epífisis femoral proximal

Enfermedad de Legg-Calvé-Perthes

Resumen La enfermedad de Perthes es una necrosis isquémica idiopática de la epífisis femoral proximal en niños. La prueba diagnóstica más utilizada es la radiografía simple, que además permite clasificar la enfermedad en función de su evolución y de su extensión. Aunque su historia natural, autolimitada en el tiempo, es generalmente benigna, existen una serie de factores asociados a un pronóstico menos favorable. Estos factores son la edad, la extensión de la enfermedad y determinados signos radiológicos, y definen a aquellos pacientes que más se van a beneficiar del tratamiento. La contención de la cabeza femoral en el acetábulo para permitir un óptimo remodelado de la misma es el objetivo del tratamiento que actualmente se basa en métodos quirúrgicos como las osteotomías femorales o acetabulares. Los métodos de contención ortopédicos como las ortesis de abducción no han demostrado obtener mejores resultados.

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Introduction

Legg-Calvé-Perthes disease consists of an avascular necrosis of the proximal femoral epiphysis of unknown aetiology in a child's skeleton. It was described simultaneously and independently by Legg, Calvé, and Perthes in the first decade of the twentieth century.[1] For some years, it was believed to be an infectious process, but it was Perthes who finally described the histological features of the disease, which led to it becoming known as "juvenile deforming osteochondritis."[2] It is a self-limited process that evolves over time in different stages. All patients will reach the phase of healing through re-ossification and remodelling of the necrosed epiphysis, but not all of them will form a spherical, covered, and congruent hip. The orthopaedic surgeon's role lies in knowing how to recognize those cases at risk for an adverse prognosis and in acting to alter the course of the disease.

Epidemiology

Perthes disease usually appears in the first decade of life, although cases in adolescence have also been described.[3,4] Depending on geographical region, its annual incidence varies from 0.2 to 29.4 per 100,000 inhabitants between 0 and 14 years of age. A higher incidence has been observed in the Caucasian race; it is less common among Asians and rare in individuals of the Negro race.[5,6] It is 3 times more common in boys than in girls, and although most of the time it presents as unilateral, it can affect both hips in up to 14% of cases.[3,7,8]

Aetiology

The specific cause of the disease remains a mystery. There are various theories based on the particular factors that are associated with this disorder. Some authors argue a nutritional or environmental origin based on the observation that there is a higher incidence of the disease in more impoverished geographical areas.[7,9] There are also anthropometric differences among the children afflicted with this disease, for they typically present with a delay in skeletal age compared to chronological age. Another observation is that there is a higher incidence of low birth weight among these children and that they are significantly shorter at birth.[3,9-11]

The fact that the pathophysiological phenomenon that occurs is an avascular necrosis has led some researchers to look for a vascular aetiology. Some have attempted to find an association with certain hypercoagulable states, such as the presence of Leiden factor 5 or a protein S or C deficiency, but the results of these studies are contradictory.[12,13] Other authors have observed an association with factors that reduce the oxygen supply, such as the mother using tobacco during the pregnancy.[14] For some years, transitory synovitis of the hip has been considered a possible causative factor because of the damage to epiphyseal vessels resulting from the increased intra-articular pressure. This theory has not been confirmed, however, and it is currently thought that what occurs is a synovial irritation secondary to the epiphyseal necrosis.[15,16]

Finally, a connection has also been found with certain behavioural disorders, such as Attention Deficit Disorder, and with urogenital changes and delays in the ossification of posterior lumbosacral elements.[17-19]

Symptoms

The symptomatology is very similar to that of transitory synovitis of the hip. It usually appears spontaneously as a pain in the hip, thigh, or knee, often accompanied by a limp, with no associated fever. On physical examination, a contracture of the adductor musculature may be found, which manifests as a more or less significant limitation in hip abduction. There may also be a flexion contracture and a limitation of internal rotation. The pain may subside briefly with the administration of non-steroidal anti-inflammatories, but it reappears and persists over the course of several weeks or even months before the diagnosis is made. When a child has been diagnosed with transitory synovitis of the hip that does not abate after a few days, the existence of an incipient Perthes disease must be ruled out.[3,15]

Imaging tests

Diagnosis

Most of the findings described to date, both for establishing the diagnosis and for quantifying the degree of epiphyseal involvement, are based on simple x-rays. Two views of the hip-anteroposterior and axial-are necessary and sufficient to diagnose the disease; depending on the stage of the disease, these will show different radiographic signs.

Waldenström defined 4 progressive radiographic stages: initial, fragmentation, re-ossification, and remodelling (fig. 1). The *initial* stage occurs when the necrosis begins. It is characterised by an increased epiphyseal density, a reduced epiphyseal size, and an increased medial joint space. In the subsequent *fragmentation* phase, resorption of the necrosed bone begins. Lytic and sclerotic areas appear in the region affected by the necrosis. One finding that marks the beginning of this stage and may be appreciated early is the presence of a subchondral fracture. In the *re-ossification* stage, the healing process begins. Areas of resorption are gradually replaced with new bone that has a cotton-like appearance on x-ray. Once re-ossification has been completed, the epiphysis will take on its definitive morphology as bony growth occurs during the *remodelling* phase, which lasts until skeletal maturity.[20,21]

Perthes disease is usually diagnosed in the initial phase or in the fragmentation phase, but there have been cases where it was detected in the remodelling phase. It is important to stress that, in order to alter the course of the process, when necessary, any intervention on the part of the surgeon must be done prior to the beginning of the healing phase.

When the disease involves both hips, the differential diagnosis must be made with dysplasia of the proximal

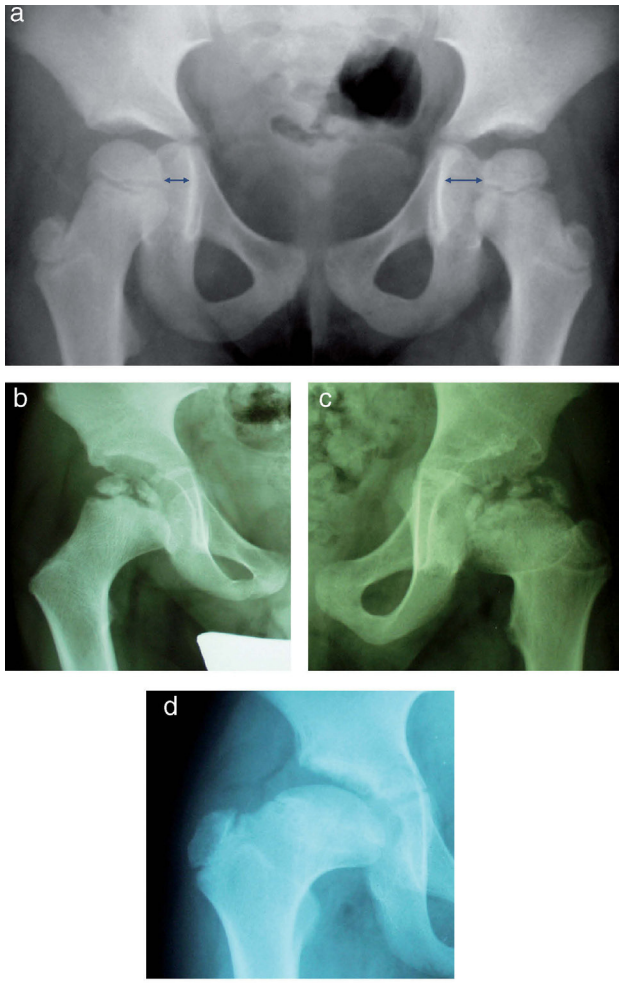

Figure 1 Waldenström stages. a) Initial stage. b) Fragmentation stage. c) Re-ossification stage. d) Remodelling stage.

femoral epiphysis (Meyer dysplasia), the multiple epiphyseal dysplasias, and the spondylo-epiphyseal dysplasias. Perthes disease differs from all of these primarily in that the involvement is usually asymmetrical–that is, each hip is in a different progressive stage. In addition, pelvis and acetabulum growth is altered in the multiple epiphyseal dysplasias and the spondylo-epiphyseal dysplasias, whereas in Perthes disease it is normal. On the other hand, Perthes disease is characterised by an initial deterioration and a subsequent improvement, while in the epiphyseal dysplasias there is a slow progression. However, it is advisable to do a bone series in bilateral cases to rule out the existence of changes in other epiphyses in the skeleton.[22-24]

Other imaging tests such as ultrasound and magnetic resonance are not of much value in diagnosing Perthes disease. The primary finding on ultrasound is an increase in the joint space, which is usually due to a synovial thickening rather than the presence of fluid in the joint, as occurs in transitory synovitis of the hip.[25] Magnetic resonance imaging may be useful in patients who are symptomatic but have normal x-rays. Reduced perfusion may be detected in the femoral epiphysis on the images obtained following contrast administration, so it is very useful for early diagnosis.[26]

Although bone scan has been used in diagnosing Perthes disease, it has not proven to be superior to magnetic resonance imaging for early diagnosis. Moreover, it is the classifications derived from simple x-rays that are the basis for therapeutic decisions. Because of the radiation involved in a bone scan to which the child would be exposed, we do not consider it a useful diagnostic test.[21,27]

Classification

There are 2 types of radiographic classifications: those that define the degree of involvement and those that determine the final result, once skeletal maturity has been reached. There is also a number of radiographic signs that one should be familiar with because, if present, they indicate greater risk of an adverse disease course.

Degree of involvement

Catterall Classification. In 1971, Catterall studied the degree of epiphyseal involvement as a prognostic factor and described 4 groups in relation to the extent of the disease in the anteroposterior and axial views. Group I shows a necrosis of the most anterior and central portion of the epiphysis, without collapse or sequestration. In group II, 50% of the anterior portion is involved, and there is a central collapse, with the height of the medial and lateral regions maintained. In group III, almost the entire epiphysis is involved, with the exception of a small medial and posterior portion. In group IV, the disease extends throughout the epiphysis and results in a total collapse (fig. 2).[21,28]

Salter-Thompson classification. Because the Catterall system is based on the maximum degree of radiographic resorption and, therefore, requires waiting until the fragmentation phase has ended, Salter and Thompson described another classification years later in an attempt to determine the extent of the disease at an earlier point. They based it on the presence of the subchondral fracture at the beginning of the fragmentation phase, distinguishing 2 groups according to the extent of the fracture on the axial x-ray. In group A, the fracture covers less than 50% of the epiphyseal width; in group B, the fracture stretches to more than half of the epiphysis and indicates more extensive disease (fig. 3). The downside of this method is that the fracture is not always detected on x-ray because it is a very early phenomenon; therefore, this method cannot always be applied.[29,30]

Herring lateral pillar classification. Some studies have shown a low reproducibility and reliability for the Catterall classification. For this reason, in 1992, Herring described a simpler system for determining prognosis by the degree of epiphyseal involvement. It is based on division of the femoral epiphysis into 3 sectors called *pillars* on the anteroposterior x-ray. The lateral pillar occupies the lateral 30% of the epiphysis, and according to the degree of its collapse, 3 groups are distinguished. In group A, the lateral pillar is intact and of normal height. In group B, the lateral pillar collapse is less than 50% of its height, and in group C, the collapse is more than 50% of its height (fig. 4).[30,31] Due to the fact that almost all patients fall into group B and group C, a group B/C subtype was subsequently distinguished

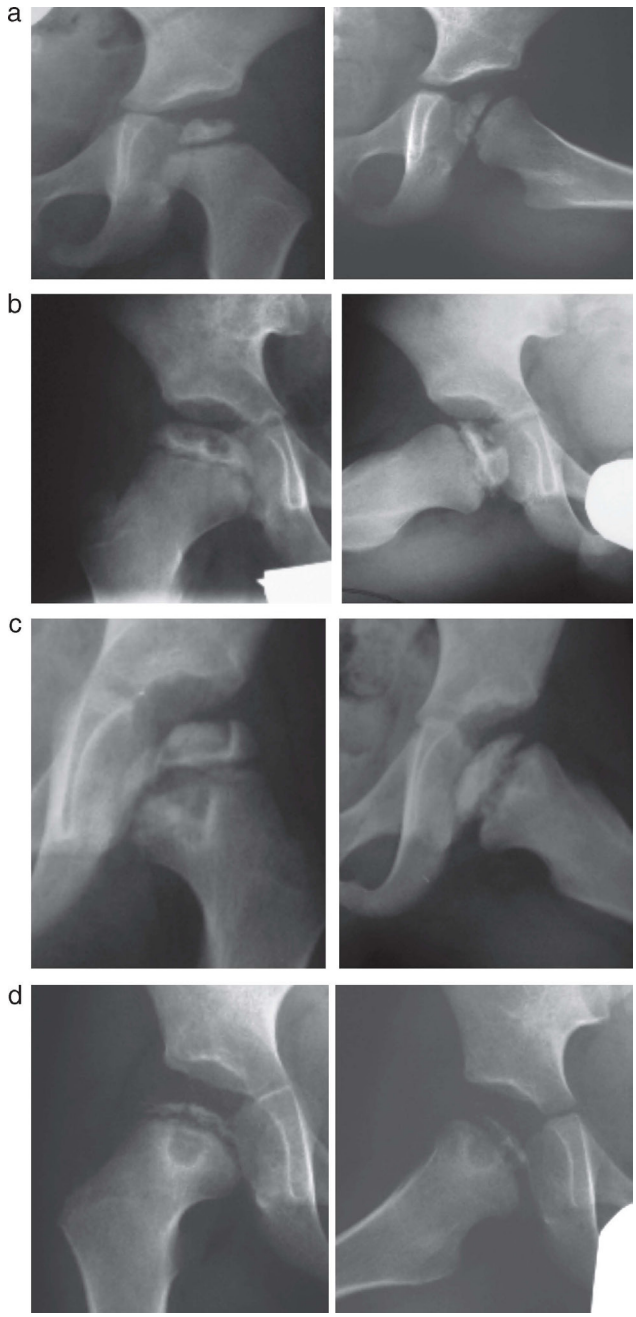

Figure 2 Catterall classification. a) Group 1. b) Group II. c) Group III. d) Group IV.

to describe more precisely the transition between the 2 groups. Group B/C is characterised by a very narrow lateral pillar (2-3 mm) at more than 50% of its height; a lateral pillar with scant ossification but at least 50% of its height; or a lateral pillar with 50% of its height and no collapse of the central pillar.[32]

Sugimoto-Akazawa classification. This classification attempts to complement the lateral pillar system through evaluation of the posterior pillar using the same criteria. The posterior pillar is defined as the posterior 30% of the femoral epiphysis on the axial view. These authors observed

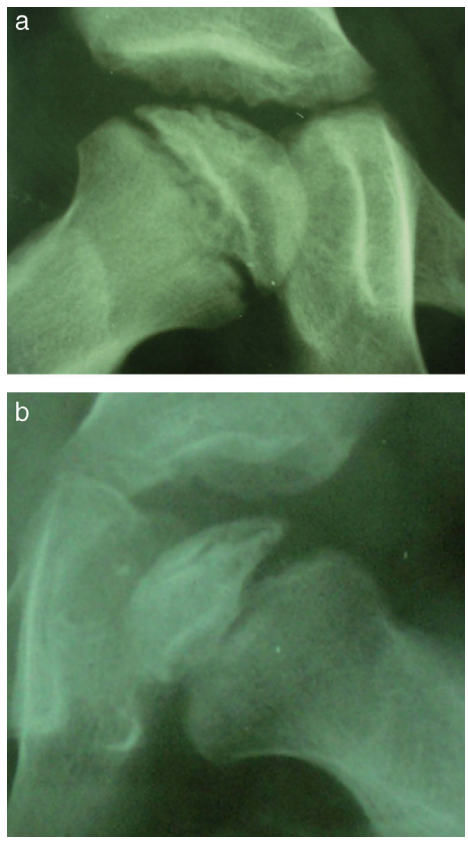

Figure 3 Salter-Thompson classification. a) Type A. b) Type b.

that the combination of the 2 classifications (lateral pillar and posterior pillar) increased the prognostic value because the great majority of cases belong to Herring group B.[33]

Final result

Stulberg classification. Distinguishes 5 hip types. Type I is a normal hip. In type II, the head is spherical but larger than normal, and the neck is shorter. In type III, the head is no longer spherical but has an ovoid shape. The acetabulum has adapted to the shape of the head, resulting in a congruent hip. In type IV, the head has a flattened shape and is still congruent with the acetabulum. In type V, the head is flattened but the acetabulum is normal, so it is not a congruent hip (fig. 5).[34]

Mose system. Determines sphericity of the femoral head by superimposing transparencies with concentric circles on the antero-posterior and axial x-ray films. A head is considered spherical when its surface is enclosed within a single circle, with a variation of not more than 2 millimetres on both x-rays.[35]

Radiographic at-risk signs

Along with his classification, Catterall described a number of radiographic signs that, when present, indicated increased risk of an adverse prognosis. These are referred to as the "head-at-risk" signs and include the following (fig. 6)[28]:

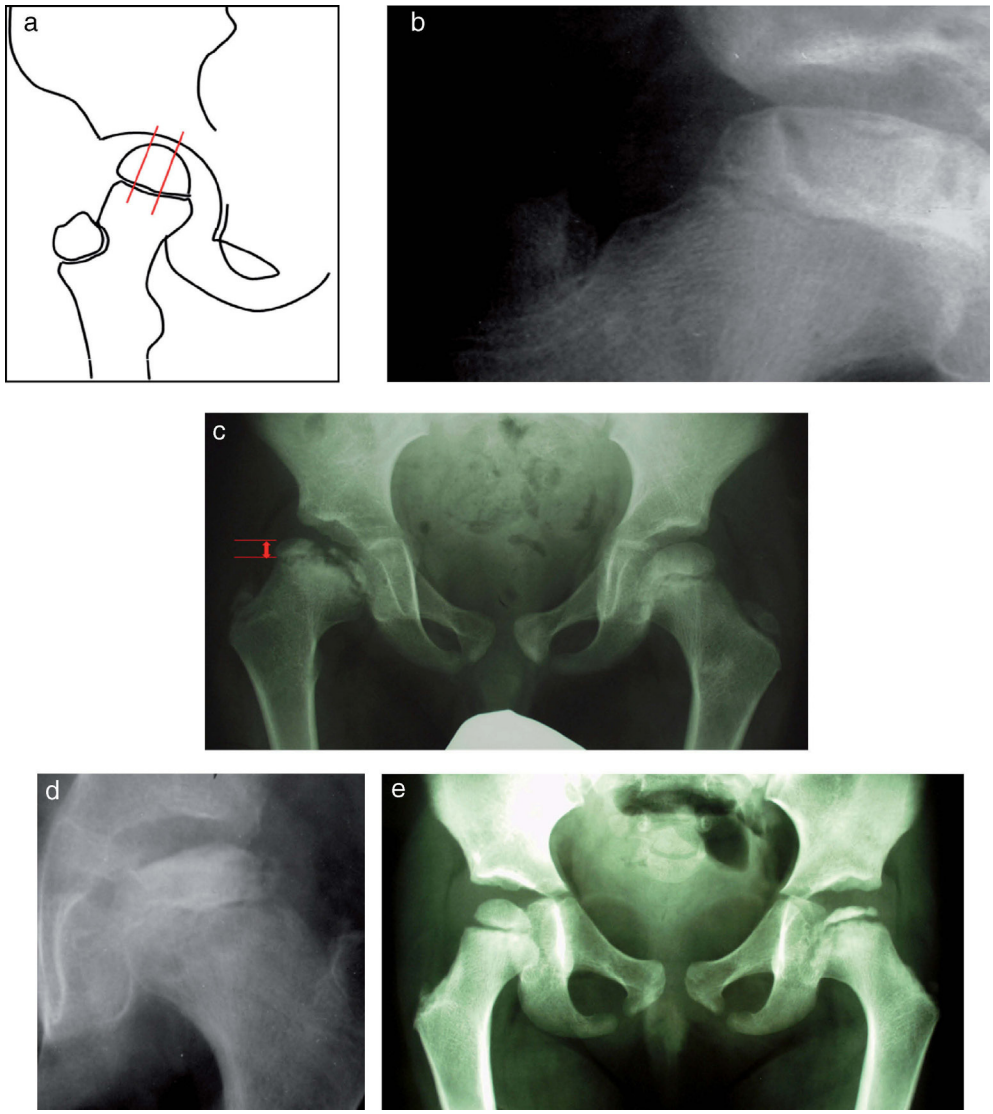

Figure 4 Herring lateral pillar classification. a) The 3 Herring pillars. b) Group A. c) Group b. d) Group B/C. e) Group C.

- Lateral calcification of the epiphysis.
- Horizontalisation of the growth plate.
- Osteolysis in the metaphyseal area, or Gage's sign.
- Lateral subluxation.

When the entire epiphysis collapses, a proximal dislocation may also occur that will manifest as a break in Shenton's line.

Natural history and risk factors

Perthes disease follows a self-limited course, and only those patients who are at high risk for developing an incongruent hip will require treatment. A number of risk factors have been described that enable these cases to be identified.

Age: it has been noted that age at onset correlates to the clinical and radiographic results. Onset at over 6 years of age, and over 8 years, especially, makes the prognosis worse because, beyond this age, the potential for remodelling is reduced.[36,37]

Extent of the disease: the degree of epiphyseal involvement also marks the disease course. Catterall stages III and IV, Salter group B, and Herring classification group C–that is, those with the most extensive involvement–are associated with a worse prognosis.[28-30]

Radiographic at-risk signs described by Catterall.

Adduction contractures: if maintained, these limit range of motion and may condition a lateral dislocation of the epiphysis.[38]

In terms of biomechanics, spherical and congruent hips will have a normal future. Patients who have an incongruent hip at the end of the remodelling phase, however, will be at increased risk for developing osteoarthritis in early adulthood. Thus, in all Stulberg groups, good joint function will last until 30 years of age, but starting in the fourth decade, group V patients, above all, will experience a rapid deterioration.[34,36]

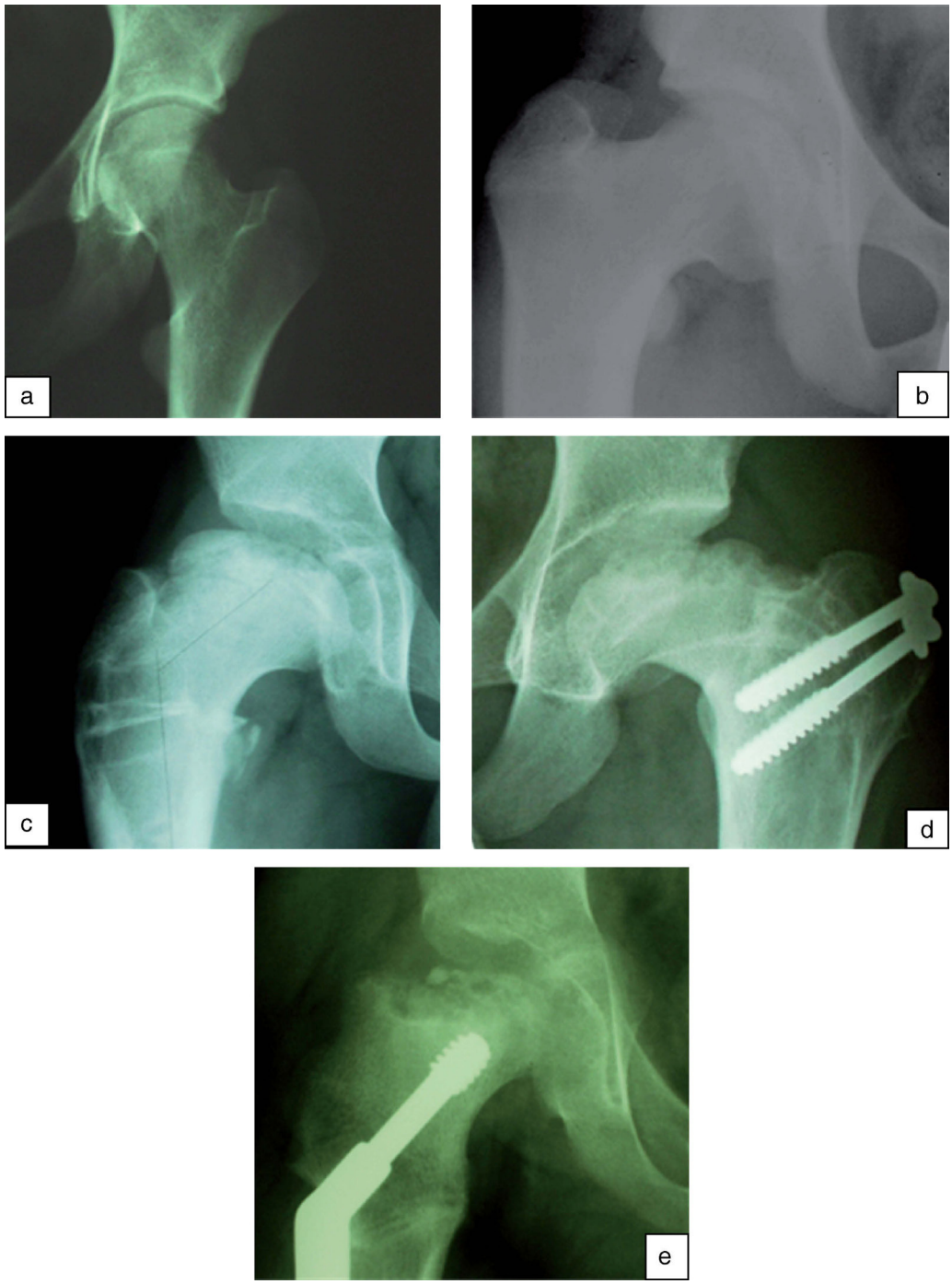

Figure 5 Stulberg classification. a) Type I. b) Type II. c) Type III. d) Type IV. e) Type V.

Treatment

Treatment objectives in Perthes disease are the relief of symptoms; the restoration of joint mobility, if impaired; and the proper remodelling of the femoral epiphysis, which is based on the principle of containment. According to this principle, ensuring proper containment of the femoral head within the acetabulum during the re-ossification process will result in a more spherical and more congruent head, owing to the reciprocal remodelling that occurs between the femoral epiphysis and the acetabulum.

Relief of Symptoms

This consists of functional rest together with administration of non-steroidal anti-inflammatories during the most painful periods of time. Functional rest may be achieved through the use of crutches. Some authors also

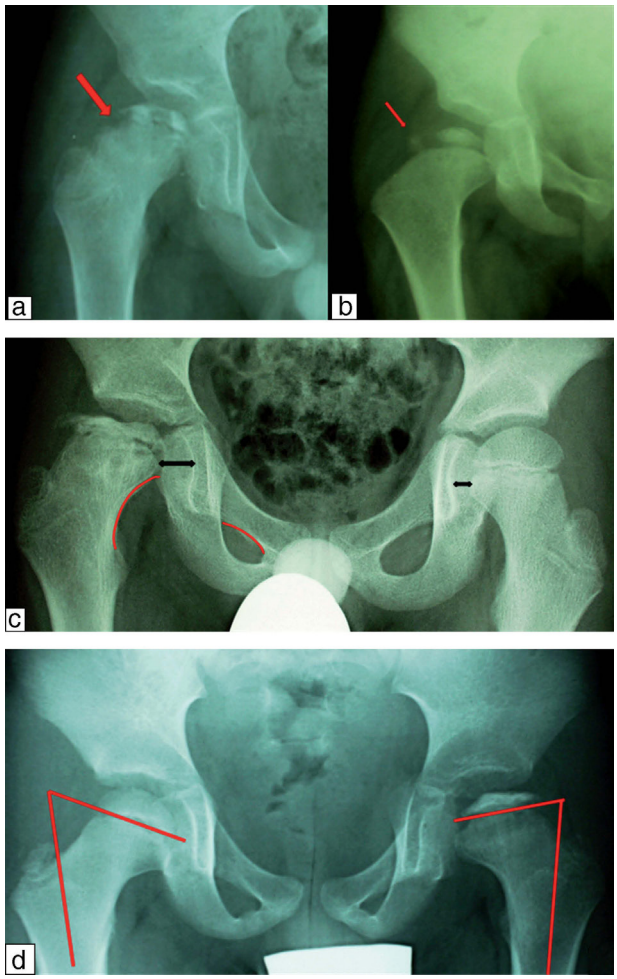

Figure 6 Radiographic At-Risk Signs. a) Gage's sign: an osteolytic area in the lateral portion of the metaphysis that will mean premature epiphyseal closure with relative overgrowth of the trochanter and a leg length discrepancy. b) Calcification lateral to the epiphysis indicates that an area of the epiphysis is ossifying outside the acetabulum, with the attendant risk of developing a coxa magna in the future or even a hinge abduction. c) Increased medial joint space is appreciated, which indicates a lateral subluxation, as well as a break in Shenton's line, which indicates a superior dislocation. d) Horizontalisation of the growth plate.

mention brief periods of traction, although its use is widely debated, especially in patients with extensive involvement.[21]

Restoration of mobility

It is essential to maintain proper mobility in those patients who have an adductor contraction that may condition a lateral dislocation of the epiphysis. For this purpose, physical therapy may be instituted, and in severe or refractory cases with abduction of 20° or less, the patient may undergo an adductor tenotomy followed by application of Petrie casts for 4 weeks.

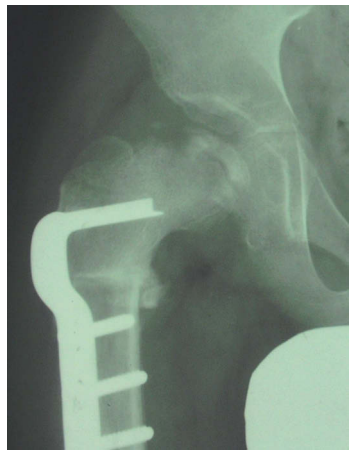

Figure 7 Varus-producing femoral osteotomy.

Containment

Various methods have been described for attempting to achieve good containment: non-surgical containment through the use of an abduction brace and surgical containment through femoral or pelvic osteotomy.

Orthopaedic containment: abduction brace

For many years, this modality was the standard treatment. Many types of braces were used, but they all ensured containment by abducting the hip. There were braces that limited weight-bearing; braces that permitted weight-bearing but limited mobility; and devices, such as the Scottish Rite brace, that permitted weight-bearing as well as a certain degree of mobility while maintaining fixed abduction.[21] In recent years, however, various studies casting doubt on its efficacy have been published, and, currently, more and more authors are advising against using them in Perthes disease.[37,39-41]

Surgical containment

Surgical containment may be achieved by repositioning the head of the femur in the acetabulum through a varus-producing femoral osteotomy or by reorienting the acetabulum through a pelvic osteotomy, or else by a combination of these 2 osteotomies. The classical indications, summarised by Salter, are the following: involvement of more than half of the femoral head (Catterall groups III and IV or Salter group B); age above 6 years at onset; or dislocation of the femoral head.[42]

Varus-producing femoral osteotomy. This technique permits intervention on the affected side of the joint and, because of its simplicity, is very attractive to many authors. According to some studies, the downside of it is an increased risk of leg length discrepancy and coxa vara, with the attendant risk of developing a weakness in the abductor musculature. It is recommended that excessive varus production be avoided to prevent these phenomena. There is no agreement as to the exact angulation that is necessary and acceptable, but it is recommended that calculation of the varus to be produced be such that the head is completely covered by the acetabulum. This can be assessed through

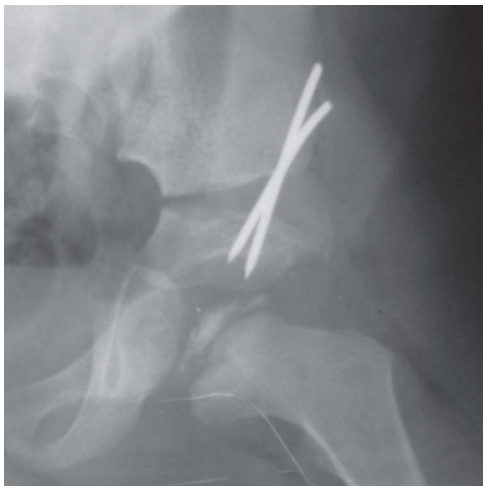

Figure 8 Salter pelvic osteotomy of the left hip.

arthrography, observing the degree of abduction necessary to reposition the head within the acetabulum. If it appears that an extreme angle will be required, another technique should be chosen (fig. 7).[21,43]

Salter pelvic osteotomy. Technically, this is similar to the osteotomy used in developmental dysplasia of the hip. The advantages it affords in comparison with femoral osteotomy are a lower risk of leg length discrepancy and coxa vara; increased articulotrochanteric distance; and greater acetabular coverage. However, it forces intervention on the healthy side of the joint, is more technically demanding, and may increase intra-articular pressure (fig. 8).[42,44,45],

Combined osteotomy. Some authors have argued for the use of combined pelvic and femoral osteotomies in cases with severe involvement and in children who are more than 7 years old, when excessive varus must be produced to achieve good containment of the head. However, other authors have shown that this technique offers no substantial improvement over other techniques in terms of the results, so there is no justification for it.[46,47]

Shelf-type acetabular osteotomy. The "shelf" osteotomy for acetabular coverage has started to be used with older children who have less potential for remodelling; with dislocation of the femoral head; and with children in whom an excessive varus must be produced to achieve satisfactory containment. It appears that the best results are obtained between 9 and 11 years of age. According to some authors, the main drawback of this technique is the possible risk of damage to the lateral acetabular epiphysis and, as a result, inhibition of acetabular growth. However, not only has this never been confirmed but, in recent studies, stimulation of acetabular growth has been detected following this type of osteotomy (fig. 9).[48,49]

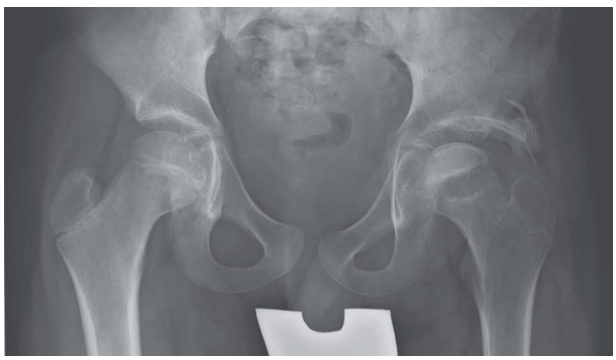

Figure 9 Shelf-type pelvic osteotomy of the left hip.

Triple pelvic osteotomy. As of a few years ago, triple pelvic osteotomy is being performed. The advantage it appears to offer over the classical osteotomies is improved containment with a lower risk of leg shortening. Its primary drawback is the risk of impingement, in the event of excessive containment. The short-term results are encouraging, but there are no long-term studies, as yet, that would corroborate these initial results.[50,51]

Although there are numerous publications on the results of treatment for Perthes disease, in general they are not very conclusive for many reasons, such as lack of controls in the studies, inadequate control of known variables, and the use of different methods to evaluate the results.[52] The first multi-centre, controlled, prospective study comparing the results of different treatments was recently published.[37] The authors observed that the patients who benefited most from surgical treatment, whether by femoral osteotomy or by pelvic osteotomy, were those in Herring groups B and B/C who were more than 8 years old at onset of the disease. For patients in Herring group A and for those under 8 years of age, surgical treatment was not associated with an improved disease course. Likewise, patients in Herring group C did not have a more favourable prognosis following surgical treatment. The most significant predictive prognostic factor was the Herring lateral pillar classification, followed by the patient's age at diagnosis.

Our therapeutic approach in patients with Perthes disease is described below. In children younger than 7-8 years with moderate involvement (Herring A-B), we take a watch-and-wait approach, with clinical-radiological check-ups every 4 months; in the event of an adduction contracture developing, we opt for rehabilitative treatment, if it is not severe, or for an adductor tenotomy, if the hip cannot be abducted more than 30° and does not respond to conservative treatment. In children older than 7-8 years with Herring stage B, or in children with stage C, regardless of age, we usually do an arthrogram to evaluate the degree of varus production necessary to achieve good containment. If it requires abduction and internal rotation, we opt for a varus-producing femoral osteotomy, particularly in patients younger than 8 years, generally in combination with epiphysiodesis of the trochanter. If it requires flexion, abduction, and internal rotation, we opt for a Shelf-type pelvic osteotomy, especially in patients older than 8 years. If excessive varus production is required, we combine the 2 osteotomies.

Complications

The main complications that may appear in Perthes disease are hinge abduction, relative overgrowth of the greater trochanter, and osteochondritis dissecans.

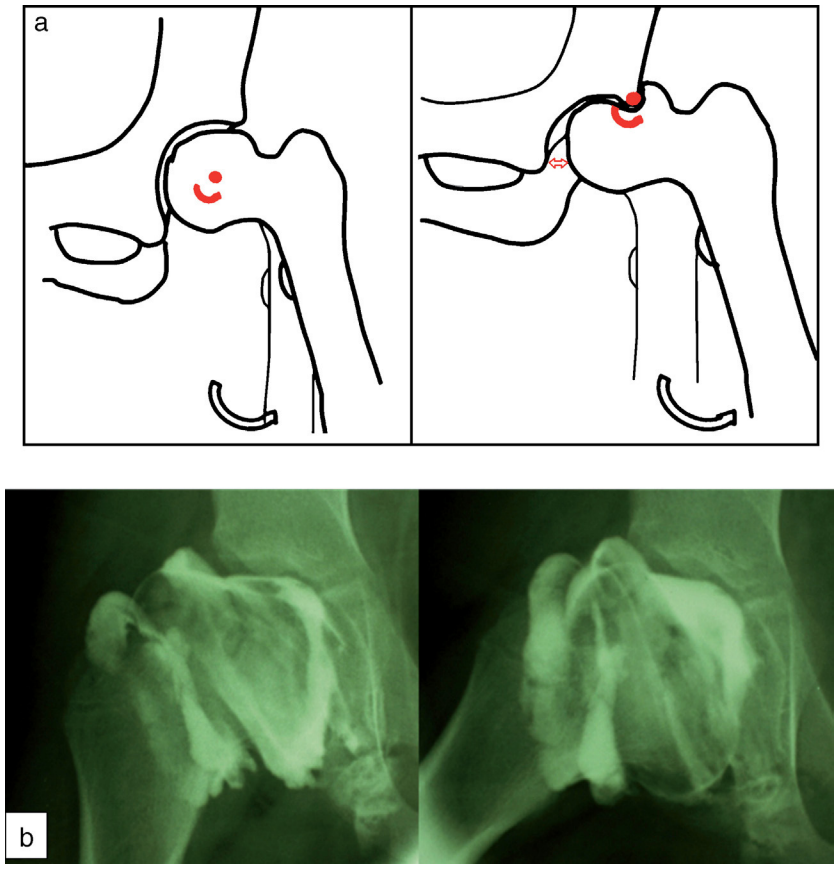

Figure 10 Hinge abduction.

Hinge abduction

Hinge abduction is a term used to describe the anomalous hip movement that occurs when the superolateral portion of the deformed femoral head strikes the lateral edge of the acetabulum, shifting the hip's centre of rotation upon abduction, which is then effected by the head pivoting around the lateral edge of the acetabulum, thus increasing the medial joint space (fig. 10A). Collapse of the central pillar together with a sustained adductor contracture may promote lateral dislocation of the epiphysis in the fragmentation phase. In the re-ossification phase, the inadequately covered lateral portion of the epiphysis will be re-ossified outside the acetabulum, and hinge abduction will gradually develop. This has been associated with onset of the disease at a later age and with more extensive epiphyseal involvement. The first symptom to appear is a limitation of hip mobility in a patient who had been progressing well. Radiographic findings indicating an increased risk of developing this condition are lateral dislocation of the epiphysis and the presence of a lateral calcification. The most reliable test for confirming the diagnosis is arthrography, which will show the hinge effect as evidenced by the medial accumulation of contrast upon abduction of the hip (fig. 10B). The most widely accepted treatment consists of valgus-producing femoral osteotomy as a rescue technique (fig. 11).[53-55]

Relative overgrowth of the greater trochanter

In some patients, growth of the proximal end of the femur may be impaired as a consequence of this disease. While growth in the femoral head growth plate is impaired, growth of the greater trochanter continues, for its vascularisation relies upon metaphyseal vessels that are not affected by the ischaemia, and this results in its relative overgrowth. Most of these impairments result from a slowing of growth

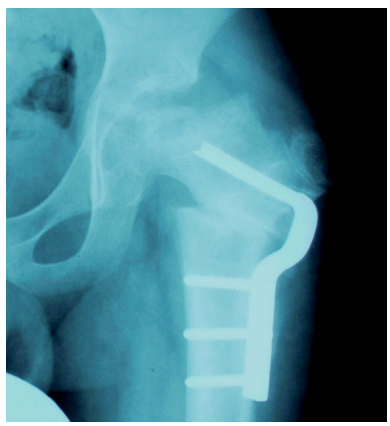

Figure 11 Valgus-producing osteotomy of a hinge abduction.

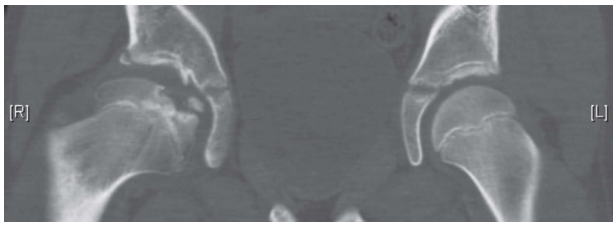

Figure 12 Osteochondritis dissecans in right hip.

rather than from physeal bars. They have been associated with increased pressure at the lateral margin of the epiphysis, which may result from a late varus-producing osteotomy in an already deformed head, and appear to be unrelated to the development of metaphyseal cysts or physeal rarefactions over the course of the disease. Treatment consists of epiphysiodesis to arrest greater trochanter growth in children younger than 8 years, while distal transfer of the greater trochanter is more effective in children over 8 years of age.

Leg length is also affected in these patients, and when we do a distal transfer of the trochanter, we sometimes simultaneously so an epiphysiodesis on the contralateral distal femur.[56-58]

Osteochondritis dissecans

Osteochondritis dissecans is a rare complication seen in 3% of cases. Unlike classical osteochondritis dissecans, in Perthes disease it is due to an impairment in the healing process such that a portion of the epiphysis is not completely reossified. It may manifest as intermittent pain with episodes of joint locking, if the fragment breaks off (fig. 12).[59,60]

Another rare complication that has been described is injury of the acetabular labrum, which also will cause pain with crackling noises and even locking.[60]

Conclusions

Perthes disease is a phenomenon that is self-limited in terms of time but, depending on the patient, can be quite variable in terms of its clinical course. Treatment should be aimed at those patients who have a worse prognosis–that is, those who have a greater degree of epiphyseal involvement or the radiographic signs of risk described by Catterall, especially if they are over 8 years of age. In the consultation with the parents, it is crucial that time be taken to explain the disease process, the clinical course, and the prognosis so that they understand why the approach is to "abstain" in cases with a benign natural history. Due to their limited efficacy, the use of abduction braces as a method of containment has been abandoned in favour of surgical treatment through varus-producing femoral or acetabular osteotomy. Close follow-up is essential, for this permits the development of a hinged hip to be suspected and impairment of growth in the proximal end of the femur to be detected.

Evidence level

Evidence level: V.

Protection of human and animal subjects

The authors declare that no experiments were performed on humans or animals for this investigation.

Confidentiality of data

The authors declare that no patient data appears in this article.

Right to privacy and informed consent

The authors declare that no patient data appears in this article.

Conflict of interest

The authors have no conflict of interest to declare.

References

1. Elly LW. Legg's disease: arthritis deformans juvenilis: osteochondritis deformans juvenilis: ''perthes's'' disease. Ann Surg. 1919 Jan;69:47–51.
2. Morrissy RT, Weinstein SL. Lovell and Winter's Pediatric Orthopaedics. Sixth edition. Filadelfia: Lippincott William and Wilkins; 2006. p. 1039–84.
3. Wiig O, Terjesen T, Svenningsen S, Lie SA. The epidemiology and aetiology of Perthes' disease in Norway. A nationwide study of 425 patients. J Bone Joint Surg Br. 2006;88:1217–23.
4. Joseph B, Mulpuri K, Varghese G. Perthes' disease in the adolescent. J Bone Joint Surg Br. 2001;83:715–20.
5. Rowe SM, Jung ST, Lee KB, Bae BH, Cheon SY, Kang KD. The incidence of Perthes' disease in Korea: a focus on differences among races. J Bone Joint Surg Br. 2005;87:1666–8.
6. Faraj AA, Nevelos AB. Ethnic factors in Perthes disease: a retrospective study among white and asian population living in the same environment. Acta Orthop Belg. 2000;66:255–8.
7. Pillai A, Atiya S, Costigan PS. The incidence of Perthes' disease in Southwest Scotland. J Bone Joint Surg Br. 2005;87:1531–5.
8. Kenet G, Ezra E, Wientroub S, Steinberg DM, Rosenberg N, Waldman D, et al. Perthes' disease and the search for genetic associations. Collagen mutations, Gaucher's disease and trombophilia. J Bone Joint Surg Br. 2008;90:1507–11.
9. Margetts BM, Perry CA, Taylor JF, Dangerfield PH. The incidence and distribution of Legg-Calvé-Perthes' disease in Liverpool, 1982-95. Arch Dis Child. 2001;84:351–4.
10. Harrison MHM, Turner MH, Jacobs P. Skeletal inmaturity in Perthes' disease. J Bone Joint Surg Br. 1976;58:37–40.
11. Lappin K, Kealy D, Cosgrove A, Graham K. Does Low birthweight predispose to Perthes' disease? Perthes disease in twins. J Pediatr Orthop B. 2003;12:307–10.
12. Vosmaer A, Rodrigues Pereira R, Koenderman JS, Rosendaal FR, Cannegieter SC. Coagulation abnormalities in Legg-Calvé-Perthes' disease. J Bone Joint Surg Am. 2010;92:121–8.

13. López-Franco M, González Morán G, De Lucas Jr JC, Llamas P, Fernández de Velasco J, Vivanco JC, et al. Legg-Perthes disease and heritable trombophilia. J Pediatr Orthop. 2005;25:456–9.
14. Bahmanyar S, Montgomery SM, Weiss RJ. Maternal smoking during pregnancy, other prenatal and perinatal factors, and the risk of Legg-Calvé-Perthes' disease. Pediatrics. 2008;122:459–64.
15. Landin LA, Danielsson LG, Wattsgard C. Transient synovitis of the hip. Its incidence, epidemiology and relation to Perthes' disease. J Bone Joint Surg Br. 1987;69:238–42.
16. Kallio P, Ryöppy, Kunnamo Y. Transient synovitis and Perthes' disease. Is there an aetiological connection? J Bone Joint Surg Br. 1986;68:808–11.
17. Loder RT, Schwartz AM, Hensinger RN. Behavioral characteristics of children with Legg-Calvé-Perthes disease. J Pediatr Orthop. 1993;13:598–601.
18. Catterall A, Roberts GC, Wynne-Davies R. Association of Perthes' disease with congenital anomalies of genitourinary tract and inguinal region. Lancet. 1971;1:996–7.
19. Makin M, Meyer S, Gilai AN. Perthes' disease and posterior lumbosacral union. J Pediatr Orthop. 1992;12:607–9.
20. Waldenström H. The first stages of coxa plana. J Bone Joint Surg. 1938;20:559–66.
21. Wenger DR, Ward WT, Herring JA. Legg-Calvé-Perthes disease. J Bone Joint Surg Am. 1991;73:778–88.
22. Crossan JF, Wynne-Davies R, Fulford GE. Bilateral failure of the capital femoral epiphysis: bilateral Perthes disease, multiple epiphyseal dysplasia, pseudoachondroplasia, and spondyloepiphyseal dysplasia congenital and tarda. J Pediatr Orthop. 1983;3:297–301.
23. Guille JT, Lipton GE, Tsirikos AI, Bowen R. Bilateral Legg-Calvé-Perthes disease: presentation and outcome. J Pediatr Orthop. 2002;22:458–63.
24. Rowe SM, Chung JY, Moon ES, Yoon TR, Jung ST, Kim SS. Dysplasia epiphysealis capitis femoris: Meyer dysplasia. J Pediatr Orthop. 2005;25:18–21.
25. Futami T, Kasahara Y, Suzuki S, Ushikubo S, Tsushiya T. Ultrasonography in transient synovitis and early Perthes' disease. J Bone Joint Surg Br. 1991;73:635–9.
26. Dillman JR, Hernández RJ. MRI of Legg-Calvé Perthes disease. Am J Roentgenol. 2009;193:1394–407.
27. Kaniklides C, Lönnerholm T, Moberg A, Sahlstedt B. Legg-Calvé-Perthes disease. Comparison of conventional radiography, MR imaging, bone scintigraphy and arthrography. Acta Radiol. 1995;36:434–9.
28. Catterall A. The natural history of Perthes' disease. J Bone Joint Surg Br. 1971;53:37–53.
29. Salter RB, Thompson GH. Legg-Calvé-Perthes disease. The prognostic significance of the subchondral fracture and a twogroup classification of the femoral head involvement. J Bone Joint Surg Am. 1984;66:479–89.
30. Herring JA, Neustad JB, Wiliams JJ, Early JS, Browne RH. The lateral pillar classification of Legg-Calvé-Perthes disease. J Pediatr Orthop. 1992;12:143–50.
31. Christensen F, Soballe K, Ejsted R, Luxhoj T. The Catterall classification of Perthes' disease: an assessment of reliability. J Bone Joint Surg Br. 1986;68:614–5.
32. Herring JA, Kim HT, Browne R. Legg-Calvé-Perthes disease. Part I: classification of radiographs with use of the modified lateral pillar and Stulberg classifications. J Bone Joint Surg Am. 2004;86:2103–20.
33. Sugimoto Y, Akazawa H, Miyake Y, Mitani S, Asaumi K, Aoki K, et al. A new scoring system for Perthes' diseased based on combined lateral and posterior pillar classifications. J Bone Joint Surg Br. 2004;86:887–91.
34. Stulberg SD, Cooperman DR, Wallensten R. The natural history of Legg-Calvé-Perthes disease. J Bone Joint Surg Am. 1981;63:1095–108.
35. Mose K. Methods of measuring in Legg-Calvé-Perthes disease with special regard to the prognosis. Clin Orthop Relat Res. 1980 Jul-Aug;(150):103-9.
36. Ippolito E, Tudisco C, Farsetti P. The long-term prognosis of unilateral Perthes' disease. J Bone Joint Surg Br. 1987;69:250–3.
37. Herring HA, Kim HT, Browne R. Legg-Calvé-Perthes disease. Part II: Prospective multicenter study of the effect of treatment on outcome. J Bone Joint Surg Am. 2004;86:2121–34.
38. Grzegorzewsky A, Synder M, Kozlowski P, Szymczak W, Bowen RJ. The role of the acetabulum in Perthes disease. J Pediatr Orthop. 2006;26:316–21.
39. Martínez AG, Weinstein SL, Dietz FR. The weight-bearing abduction brace for the treatment of Legg-Perthes disease. J Bone Joint Surg Am. 1992;74:12–21.
40. Meehan PL, Ángel D, Nelson JM. The Scottish Rite abduction orthosis for the treatment of Legg-Perthes Disease. A radiographic analysis. J Bone Joint Surg Am. 1992;74:2–12.
41. 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. J Bone Joint Surg Br. 2008;90:1364–71.
42. Salter RB. The present status of surgical treatment for Legg-Perthes disease. J Bone Joint Surg Am. 1984;66:961–6.
43. Coates CJ, Paterson JM, Woods KR, Catterall A, Fixsen JA. Femoral osteotomy in Perthes' disease. Results at maturity. J Bone Joint Surg Br. 1990;72:581–5.
44. Sponseller PD, Desai SS, Millis MB. Comparison of femoral and innominate osteotomies for the treatment of Legg-Calvé-Perthes disease. J Bone Joint Surg Am. 1988;70:1131–9.
45. Kitakoji T, Hattori T, Kitoh H, Katoh M, Ishiguro N. Which is a better method for Perthes' disease: femoral varus or Salter Osteotomy? Clin Orthop Relat Res. 2005 Jan;(430):163–70.
46. Olney BW, Asher MA. Combined Innominate and femoral osteotomy for the treatment of severe Legg-Calvé-Perhtes disease. J Pediatr Orthop. 1985;5:645–51.
47. Javid M, Wedge JH. Radiographic results of combined Salter innominate and femoral osteotomy in Legg-calvé Perthes disease in older children. J Child Orthop. 2009;3:229–34.
48. Daly K, Bruce C, Catterall A. Lateral Shelf acetabuloplasty in Perthes' disease. A review at the end of growth. J Bone Joint Surg Br. 1999;81:380–4.
49. Domzalski ME, Glutting J, Bowen JR, Littleton AG. Lateral acetabular growth stimulation following a labral support procedure in Legg-Calvé-Perthes disease. J Bone Joint Surg Am. 2006;88:1458–66.
50. Kumar S, Bache CE, O'Hara JN. Interlocking tripple pelvic ostetomy in severe Legg-Calvé-Perthes disease. J Pediatr Orthop. 2002;22:464–70.
51. Wenger DR, Pring ME, Hosalkar HS. Advanced containment methods for Legg-Calvé-Perthes disease: results of triple pelvic osteotomy. J Pediatr Orthop. 2010;30:749–57.
52. Herring JA. The treatment of Legg-Calvé-Perthes disease. A critical review of the literature. J Bone Joint Surg Am. 1994;76:448–58.
53. Quain S, Catterall A. Hinge abduction of the hip. Diagnosis and treatment. J Bone Joint Surg Br. 1986;68:61–4.
54. Reinker KA. Early diagnosis and treatment of hinge abduction in Legg-Perthes disease. J Pediatr Orthop. 1996;16:3–9.
55. Bankes MJK, Catterall A, Hashemi-Nejad A. Valgus extension osteotomy for ''hinge abduction'' in Perthes disease. J Bone Joint Surg Br. 2000;82:548–54.
56. Barnes JM. Premature epiphysial closure in Perthes' disease. J Bone Joint Surg Br. 1980;62:432–7.
57. Sponseller PD, Desai SS, Millis MB. Abnormalities of proximal femoral growth after severe Perthes' disease. J Bone Joint Surg Br. 1989;71:610–4.
58. Schneidmueller D, Carstens C, Thompsen M. Surgical treatment of overgrowth of the greater trochanter in children and adolescents. J Pediatr Orthop. 2006;26:486–90.
59. Bowen R, Kumar P, Joyce J, Bowen JC. Osteochondritis dissecans following Perthes' disease. Arthroscopic-operative treatment. Clin Orthop Relat Res. 1986 Aug;(209):49-56.
60. Grossbard G. Hip pain during adolescence after Perthe's disease. J Bone Joint Surg Br. 1981;63:572–4.