CHAPTER 16

Legg-Calvé-Perthes Disease

Definition

Legg-Calvé-Perthes disease is a syndrome in which an avascular event affects the capital epiphysis (head) of the femur. Following the avascular event, growth of the ossific nucleus stops and the bone becomes dense. The dense bone is subsequently resorbed and replaced by new bone, during which process the mechanical properties of the femoral head are altered such that the head tends to flatten and enlarge. Once new bone is in place, the head slowly remodels until skeletal maturity is achieved.

The disease is of variable severity, and bilateral involvement occurs in approximately 10 to 12 percent of patients. The disorder is most prevalent in children ages 4 to 12 years, but it can be seen in children from 18 months of age to skeletal maturity. It is more common in boys than in girls by a ratio of 4 or 5 to 1. The etiology of Legg-Calvé-Perthes disease is unknown, but the disorder may be due to a silent coagulopathy.

History

Legg-Calvé-Perthes disease was independently recognized as a distinct entity toward the end of the first decade of the twentieth century by *Arthur Legg,* of the United States, *Jacques Calvé,* of France, *Georg Perthes,* of Germany, and *Henning Waldenström,* of Sweden (Fig. 16–1).

In 1910 Legg described the prominent characteristics of the disorder: onset between 5 and 8 years of age, a history of trauma, a painless limp, and minimal or no spasm or shortening of the affected limb.[90,167,168] Calvé noted that affected individuals had minimal atrophy of the leg and no palpable hip swelling.[33,34,90] Illustrations of the radiographic appearance of the diseased hips (Fig. 16–2) were made by Paul Sourdat, who worked for Calvé.[90]

Perthes initially thought the condition was a youthful variation of adult degenerative arthritis (Fig. 16–3).[90,211] Later he defined the disorder as "a self-limiting, non-inflammatory condition, affecting the capital femoral epiphysis with stages of degeneration and regeneration, leading to restoration of the bone nucleus."[212] Schwarz, a pathologist and colleague of Perthes, provided the first illustration of the blood supply to the femoral head (Fig. 16–4).[250] Waldenström reported the radiographic changes associated with the disorder in 1909; however, in this early work, he thought the disease was a form of tuberculosis and not a distinct entity.[281]

EARLY TREATMENT CONCEPTS

In the early years, patients diagnosed with Legg-Calvé-Perthes disease were treated with bedrest, immobilization, and weight relief. To achieve the latter, the patten-bottom brace was frequently employed (Fig. 16–5). The involved leg was hung from an ischial weightbearing caliper, which necessitated the patient's wearing an elevated shoe on the other foot. Although this device appeared to provide relief from direct weightbearing, contemporary biomechanical concepts suggest that the compressive forces of muscles operating across the hip while the leg and brace are suspended actually create greater intra-articular pressure than that produced by ordinary weightbearing.[118]

Despite strong reservations expressed by Calvé, Legg, and Waldenström,[33,167,168,282] relief from weightbearing continued to be a popular therapeutic concept. By the mid-1950s many physicians were using leg calipers to keep their patients in protracted recumbency until there was radiologic confirmation that the head of the femur was completely healed.[218] It was not unusual for patients to be kept in hospitals for 5 years or more for treatment, during which time they would use specially designed carts and gurneys to move about (Fig. 16–6). The Snyder sling (Fig. 16–7) was another popular treatment device during the 1950s.[255]

THE CONCEPT OF CONTAINMENT TREATMENT

Most of today's therapeutic approaches are based on the concept of containment, which, over the years, has evolved to include nonoperative as well as operative treatment methods.

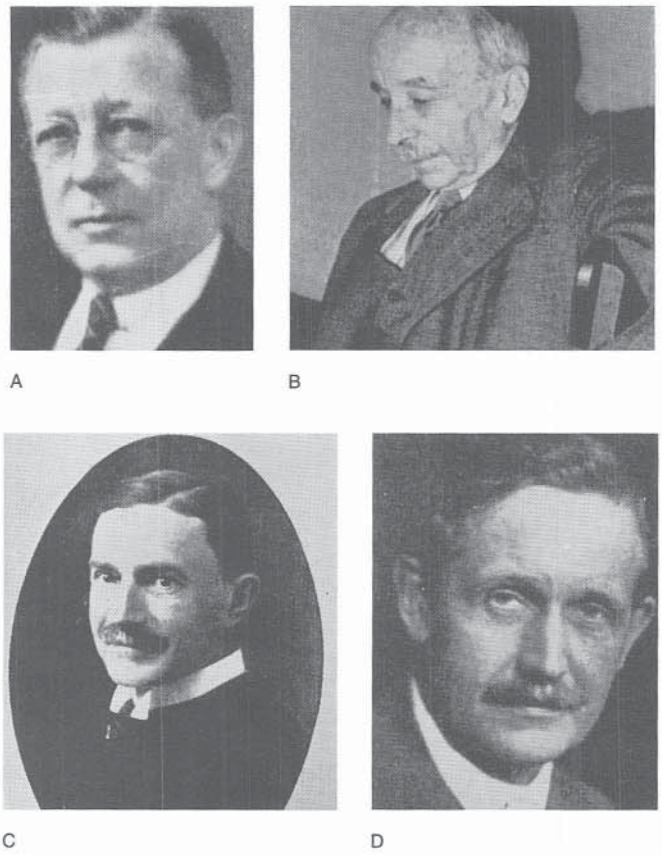


FIGURE 16–1 **A,** Arthur Legg. **B,** Jacques Calvé. **C,** Georg Perthes. **D,** Henning Waldenström.

The use of broomstick plasters (Fig. 16–8) reportedly originated in 1929 with Parker.[109] Harrison and Menon stated that "if the head is contained within the acetabular cup, then like jelly poured into a mold the head should be the same shape as the cup when it is allowed to come out after reconstitution." Shortly afterward, Harrison and associates described using an ambulatory containment brace that was specifically created to position the hip for containment.[111]

Eyre-Brook, in 1936, thought treatment needed to be started while the femoral head was still soft but before it became deformed.[67] Treatment consisted of traction in bed for 18 to 24 months. The primary goals of therapy were to maintain a round femoral head and preserve full range of motion of the hip joint.

In 1966 Salter induced avascular necrosis (AVN) of the head of the femur in a pig model.[242] When the hip was maintained in a neutral or adducted position, the newly modeled bone became deformed. However, when the hip was in flexion and abduction and weightbearing was permitted, the acetabulum acted as a mold and the femoral head did not become deformed. In a subsequent experiment, the pigs were separated into three treatment groups: (1) normal weightbearing, (2) nonweightbearing, with hips maintained in acute flexion, and (3) weightbearing with hips held in abduction. Only the third approach resulted in rounded femoral heads, which again indicated that containment within the acetabulum deterred deformation of the femoral head.[243] This response was referred to as "biologic plasticity."

Later Salter noted that the disorder started with ischemia of the femoral head, followed by revascularization.[245] He also observed the occurrence of a pathologic fracture of the subchondral bone during the early phase of revascularization and believed that at this stage, the femoral head was most vulnerable to deformation (Fig. 16–9).[247] If the fracture did not unite during subsequent resorption of the underlying bone, the head would collapse and flatten, with a portion of it extruding anterolaterally out of the acetabulum. Lateral subluxation of the head of the femur resulted in increased pressure from the edge of acetabulum, which produced stress concentration in the head, leading to progressive deformity. Salter recommended containment to prevent pressure from the edge of the acetabulum.

Despite lack of scientific proof, containment treatment has been clinically accepted for many years and remains popular. Over time, various nonsurgical and surgical methods have been employed in an attempt to contain the femoral head within the acetabulum. These treatments include bracing, Petrie cast wear, femoral osteotomy, innominate osteotomy, and acetabular shelf procedures.

Etiology

In the past the etiology of Legg-Calvé-Perthes disease was considered multifactorial, with the exact cause uncertain (Table 16–1). Some research has demonstrated a consistent relationship to a coagulopathy involving proteins C and S and hypofibrinolysis.* These studies suggested that abnormal lysis of intravascular clots may be the primary cause of a majority of cases of Legg-Calvé-Perthes disease. Subsequent studies, however, have not corroborated these findings,[6,77,181] and additional research is needed to better determine the role of coagulation abnormalities in Legg-Calvé-Perthes disease.

Many studies have identified other factors related to the etiology of the disorder. The pathologic changes seen in the femoral head are likely a result of vascular factors. Various authors have implicated both the arterial and venous systems.† Other theories as to the cause of the disease include the "predisposed child" concept, based on findings of abnormal growth and development;‡ trauma, particularly in the predisposed child;[43,57,90] hyperactivity or attention deficit disorder;[90,174] hereditary influences;§ environmental factors;[97–101] and as a sequela of synovitis.‖

COAGULATION ABNORMALITIES

A number of hematologic abnormalities have been implicated in Legg-Calvé-Perthes disease. Children with hemoglobinopathies such as sickle cell disease and thalassemia

*See references 2, 61, 87, 89, 138, 154, 203, 213, 228, 233, 275.

†See references 37, 43, 55, 75, 94, 114, 130, 134, 171, 201, 240, 265, 271, 273, 290.

‡See references 5, 21, 30, 36, 66, 73, 83, 101, 108, 131, 153, 156, 157, 191, 197, 198, 221, 231, 270, 292.

§See references 29, 102, 103, 106, 206, 292.

‖See references 84, 113, 125, 140, 141, 176, 192, 209, 254, 277, 290.

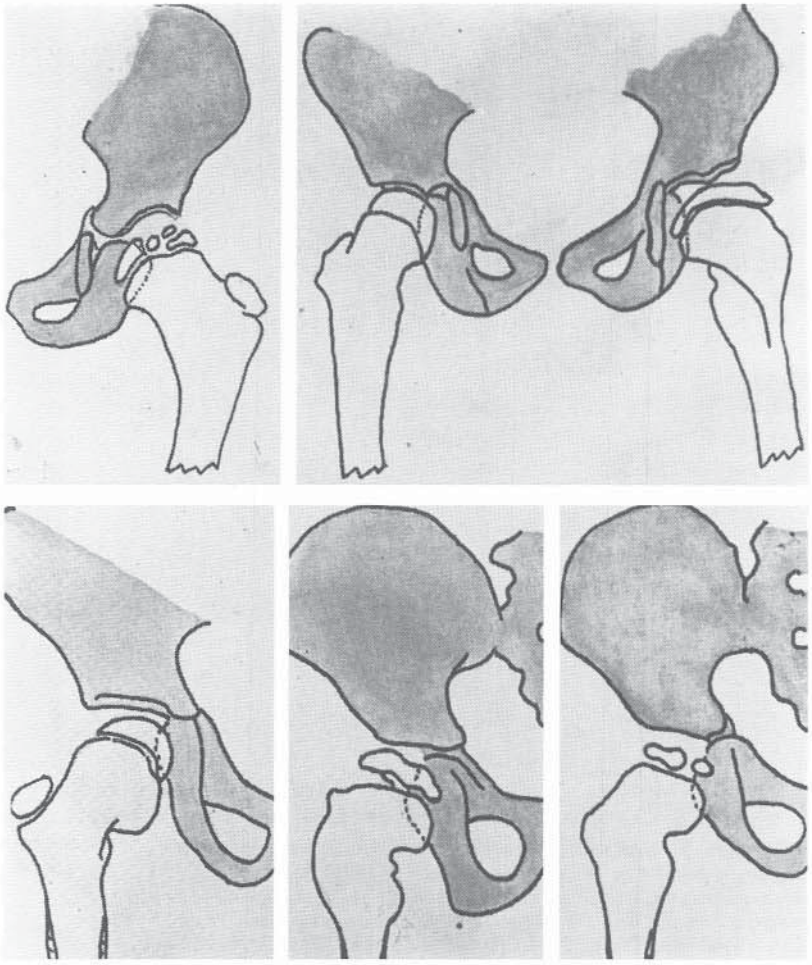


FIGURE 16–2 Drawings by Paul Sourdat showing radiographic changes later associated with Legg-Calvé-Perthes disease. (From Goff CW: Legg-Calvé-Perthes Syndrome and Related Osteochondroses of Youth, 1954. Courtesy of Charles C Thomas, Publisher, Ltd, Springfield, IL.)

commonly suffer AVN of the femoral head.[61,203,228] Avascular changes have also been observed in patients with leukemia, lymphoma, idiopathic thrombocytopenic purpura, and hemophilia.[2,213,233,275] An increase in blood viscosity in patients with Legg-Calvé-Perthes disease has also been reported.[154]

More recently, Glueck and associates reported abnormal venous thrombus formation in five of eight patients with Legg-Calvé-Perthes disease.[88] The specific clotting abnormalities were deficiencies in protein C and S and the presence of hypofibrinolysis. Affected family members also had a history of other thrombotic events. In a second study comprising 44 children with Legg-Calvé-Perthes disease, 23 had thrombophilia, three had hypofibrinolysis, and seven had elevated lipoprotein(a).[86] Of particular significance, of the 23 patients with thrombophilia, 19 had protein C deficiency and four had protein S deficiency. In the general population a deficiency in protein C or S occurs in only one in 15,000 individuals.

These findings were further corroborated in a subsequent study by Glueck and associates in which 50 of 64 children with Legg-Calvé-Perthes disease had coagulation abnormalities.[89] Resistance to activated protein C was the most common thrombophilic trait, with 23 children having a low activated protein C ratio (compared with seven of 160 pediatric control patients). Thrombophilia may cause thrombotic venous occlusion in the femoral head, with venous hypertension and hypoxic bone death resulting in Legg-Calvé-Perthes disease. From this work, it appears that resistance to activated protein C plays a major role in the pathogenesis of the disorder.

Other investigators, however, have not been able to substantiate the findings of Glueck and associates. McDougall and associates did coagulation studies on 49 patients with Legg-Calvé-Perthes disease and did not find frequent abnormalities in protein C or S or lipoprotein(a).[181] The incidence of heterozygotes with factor V Leiden (4 percent) was within the expected normal range. Gallistl and associates reported that of 44 patients with Legg-Calvé-Perthes disease, only one (2.2 percent) had a deficiency in protein C activity and only three (6.8 percent) were positive for activated protein C resistance.[77] These prevalences are slightly higher than the expected normal prevalences but significantly lower than the prevalences reported by Glueck and associates. The authors concluded that inherited thrombophilia is not a cause of Legg-Calvé-Perthes disease. In a study of 61 children with Legg-Calvé-Perthes disease and 296 controls, the only inherited risk factor found by Arruda and associates was mutation in the factor V gene (factor V Leiden).[6] The prevalence of factor V Leiden mutation was 4.9 percent in patients with Legg-Calvé-Perthes disease, compared with 0.7 percent in controls. No patient had a prothrombin gene variant. Clearly, further studies are needed to resolve this issue.

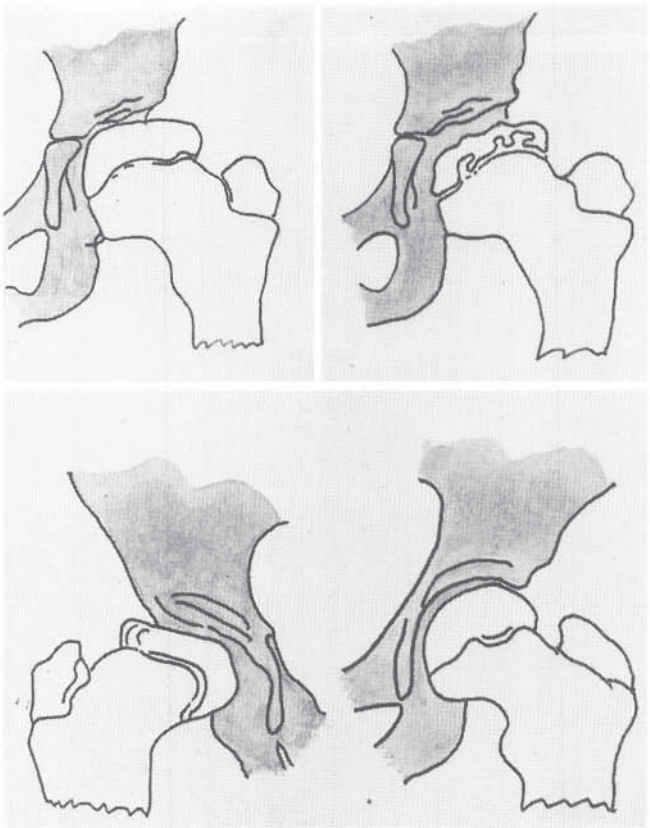


FIGURE 16–3 Drawings by Georg Perthes of radiographic changes describing a disorder he initially thought was a youthful variation of adult degenerative arthritis and which he referred to as "arthritis deformans juveniles." (From Goff CW: Legg-Calvé-Perthes Syndrome and Related Osteochondroses of Youth, 1954. Courtesy of Charles C Thomas, Publisher, Ltd, Springfield, IL.)

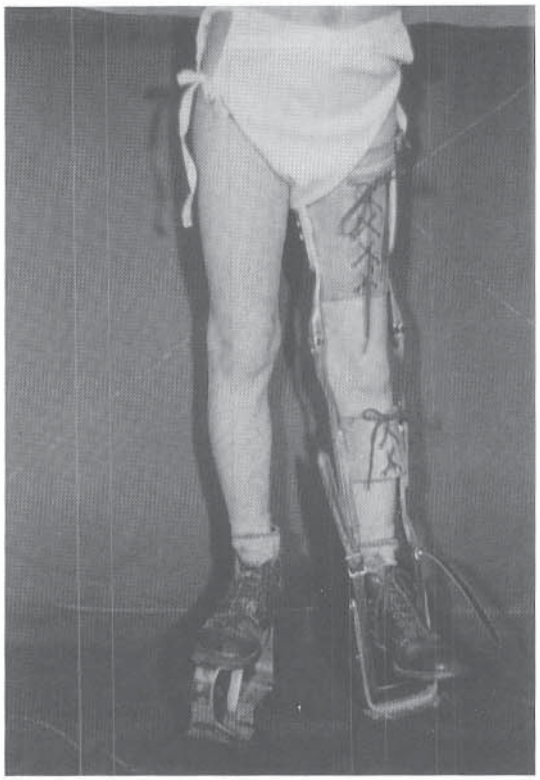


FIGURE 16–5 The patten-bottom brace, designed to relieve the weight across the hip by suspending the limb on an ischial seat. The foot does not touch the end of the brace and the other shoe is elevated to clear the ground.

ARTERIAL STATUS OF THE FEMORAL HEAD

Until recently, the arterial circulation of the femoral head was considered the most likely site of infarction in Legg-Calvé-Perthes disease. The primary blood supply to the capital epiphysis comes from two anastomotic arterial rings located around the neck of the femur (Fig. 16–10).[43] The medial and lateral femoral circumflex arteries form the extracapsular ring, with most of the blood being supplied by the medial circumflex artery. The major arterial supply to

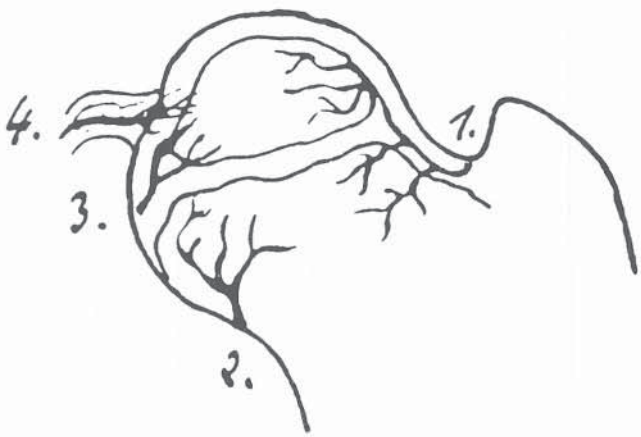


FIGURE 16–4 Drawing by Schwarz of the blood supply to the femoral head. (From Schwarz E: A typical disease of the upper femoral epiphysis. Clin Orthop 1986;209:5–12.)

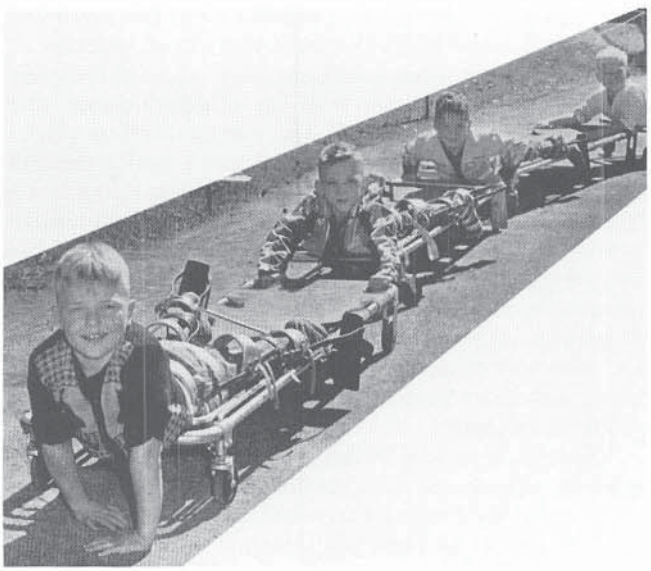


FIGURE 16–6 Carts specially designed to allow children to move about while in the nonambulatory Newington abduction frames. (From Goff CW: Legg-Calvé-Perthes Syndrome and Related Osteochondroses of Youth, 1954. Courtesy of Charles C Thomas, Publisher, Ltd, Springfield, IL.)

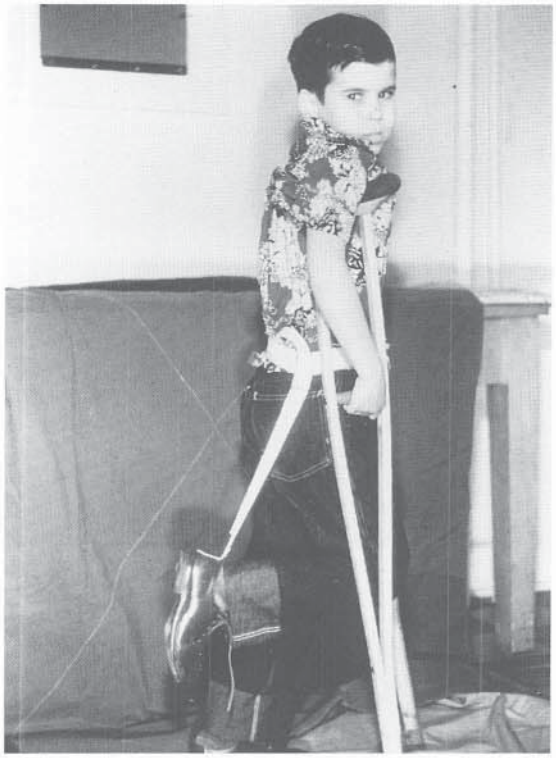


FIGURE 16–7 The Snyder sling being used by a boy. (From Herring JA: Legg-Calvé-Perthes Disease. Rosemont, IL, American Academy of Orthopaedic Surgeons, 1996.)

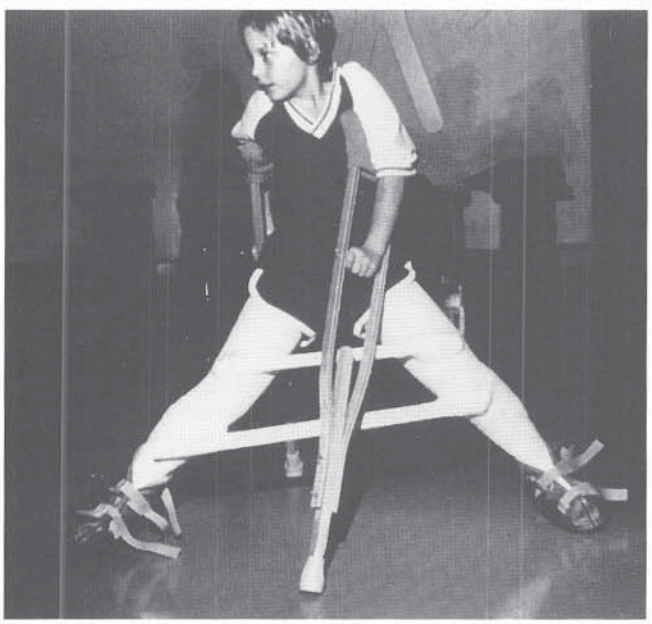


FIGURE 16–8 Broomstick plasters, known as Petrie casts, maintain the hips in approximately 45 degrees of abduction and 5 to 10 degrees of internal rotation with the knees slightly flexed. The patient walks by using crutches in front and back. (From Herring JA: Legg-Calvé-Perthes Disease. Rosemont, IL, American Academy of Orthopaedic Surgeons, 1996.)

the femoral head is the lateral segment of the arterial ring, which is the terminal branch of the medial femoral circumflex artery. After penetrating the lateral capsule in the posterior trochanteric fossa, this blood vessel becomes the lateral

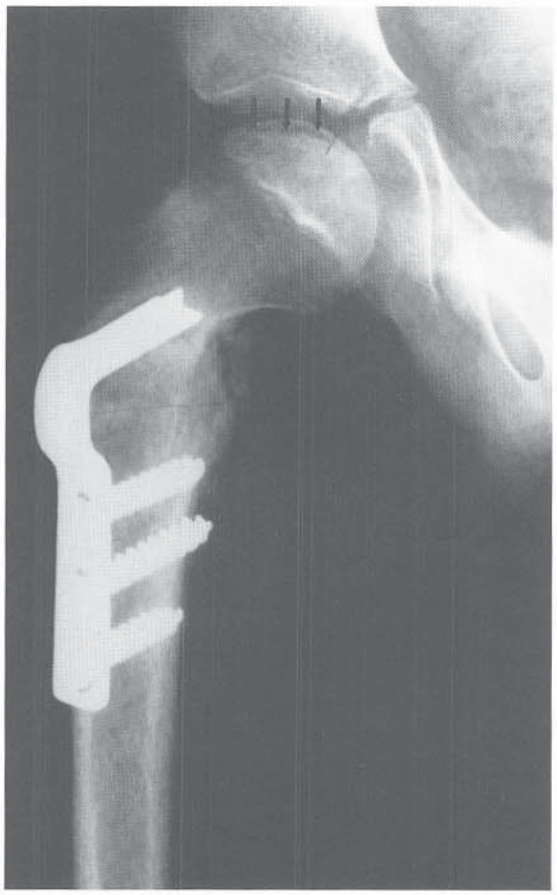
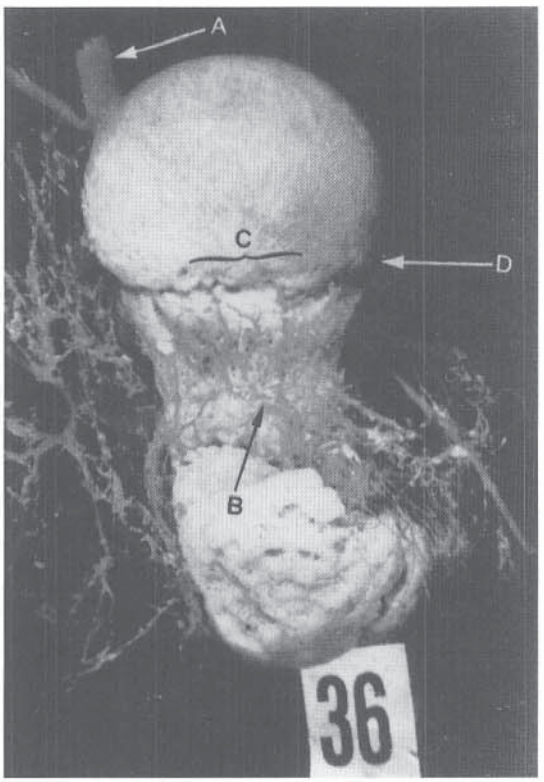


FIGURE 16–9 The subchondral fracture appears during the stage of increased density of the femoral head (*arrows*). The extent or breadth of the fracture is used to predict the degree of head involvement according to the classification system of Salter and Thompson.

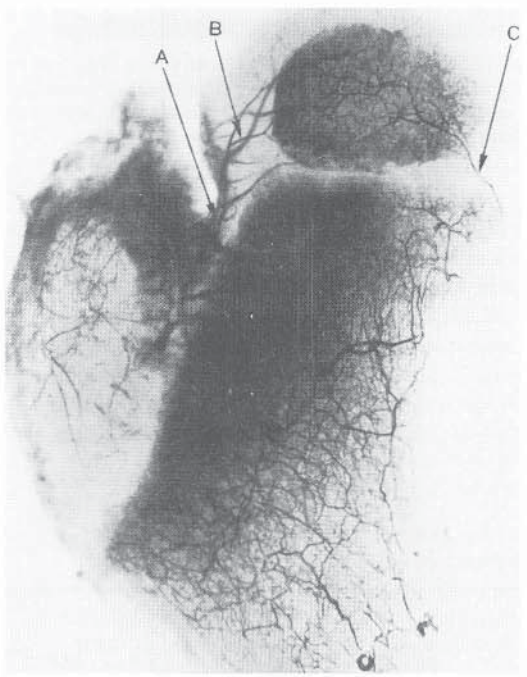
ascending cervical artery. As the vessel advances between the trochanter and the capsule, it courses through a narrow passage, an area particularly constricted in children less than 8 years old. In addition to the extracapsular ring, there is a subsynovial intracapsular ring that joins four ascending cervical arterial groups. This intracapsular ring has been found to be incomplete more often in males than in females.[43] In most cases, no vessels cross the epiphyseal

TABLE 16–1 **Factors Related to the Etiology of Legg-Calvé-Perthes Disease**

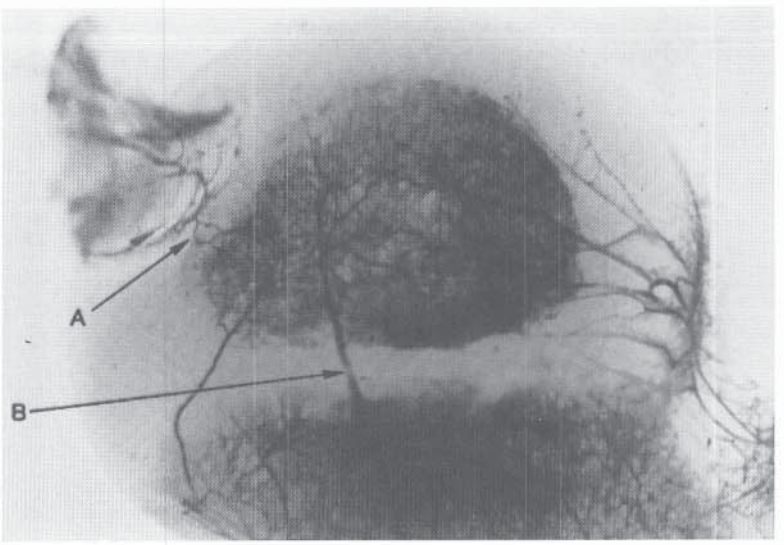
- Coagulation abnormalities involving protein C and S (abnormal lysis of intravascular clots is likely the primary cause in some cases)
- Arterial status of the femoral head (reduction in blood flow resulting in multiple infarcts and avascular necrosis)
- Abnormal venous drainage of the femoral head and neck (venous outflow obstruction)
- Abnormal growth and development (delay in bone age relative to patient's chronological age, and growth hormone abnormalities)
- Trauma, particularly in the "predisposed" child
- Hyperactivity or attention deficit disorder
- Hereditary influences (genetic component)
- Environmental influences, particularly nutritional factors
- As a sequela of synovitis (synovitis may be the first manifestation of the disease but is rarely, if ever, the cause of the disorder)



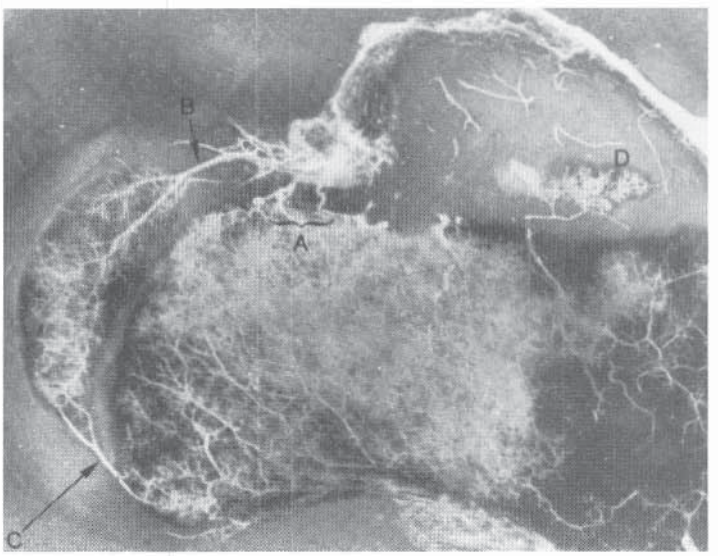
A



B



C



D

FIGURE 16–10 Blood supply to the femoral head. **A,** Superolateral view of the proximal femur showing the femoral artery (*A*), extracapsular ring (*B*), ascending lateral cervical arteries (*C*), and epiphyseal plate (*D*). **B,** Anterior half of the proximal femur showing the lateral ascending cervical artery (*A*) and the epiphyseal branches of the lateral (*B*) and medial (*C*) ascending cervical arteries as they traverse the perichondrial ring rather than the epiphyseal plate. **C,** A single artery (*A*) in the ligamentum teres supplies a portion of the secondary center of ossification. The epiphyseal branch of the posterior ascending cervical artery (*B*) appears to pass through the center of the epiphyseal plate but is actually located at the periphery. **D,** Anterior view of the middle section of the head and neck showing the metaphyseal branches (*A*) of the lateral ascending cervical artery, the epiphyseal branches of the lateral (*B*) and medial (*C*) ascending cervical arteries, and the trochanteric ossification center (*D*) just starting to form. (From Chung SM: The arterial supply of the developing proximal end of the human femur. J Bone Joint Surg 1976;58-A:961.)

plate and minimal blood is supplied by the ligamentum teres.

Angiographic studies have helped demonstrate the blood supply of the proximal femur in patients with Legg-Calvé-Perthes disease. Obstruction of the superior capsular arteries of the femoral head was clearly demonstrated in 11 patients during the first 5 months following onset of symptoms.[273] As the disease process progressed, angiography revealed revascularization. In one case blood flow decreased when the hip was extended. The condition improved, however, when the hip was flexed to 30 degrees. A general reduction in blood flow, with a significant decrease in medial circumflex

artery flow, has also been observed (Fig. 16–11).[55] The major source of blood to the femoral head in black African neonates is the inferior gluteal artery, an anomaly that could explain why the disease is uncommon in black children.[201] In a study comparing the vascular status of the femoral head in miniature dogs (which are prone to spontaneous AVN) with that in normal-sized dogs, the most evident variation was the course of the superior retinacular vessels.[75] In small dogs the vessels coursed through a shallow neck, giving the appearance of a suspended bridge. In the larger dogs the vessels traveled through the deep fossa of the femoral neck and seemed to be more stable.

Technetium scanning has been used to study the vascular status of the femoral head in Legg-Calvé-Perthes disease. Wingstrand and associates used scintigraphy to examine the hips of 25 children with transitory synovitis.[290] Initially, four of the children had decreased uptake of radioisotope (i.e., decreased blood flow). Six weeks later three of the four had normal or increased uptake. The one patient in whom decreased blood flow persisted subsequently was diagnosed with Legg-Calvé-Perthes disease. The investigators concluded that some patients with transient synovitis have coexisting transient ischemia of the femoral epiphysis and that those with more severe cases or recurrent episodes of ischemia are at greater risk of developing Legg-Calvé-Perthes disease. Scans have also demonstrated decreased uptake in the upper pole of the femoral head as well as a reduction in blood flow in the femoral artery on the affected side in the early stages of the disorder.[240]

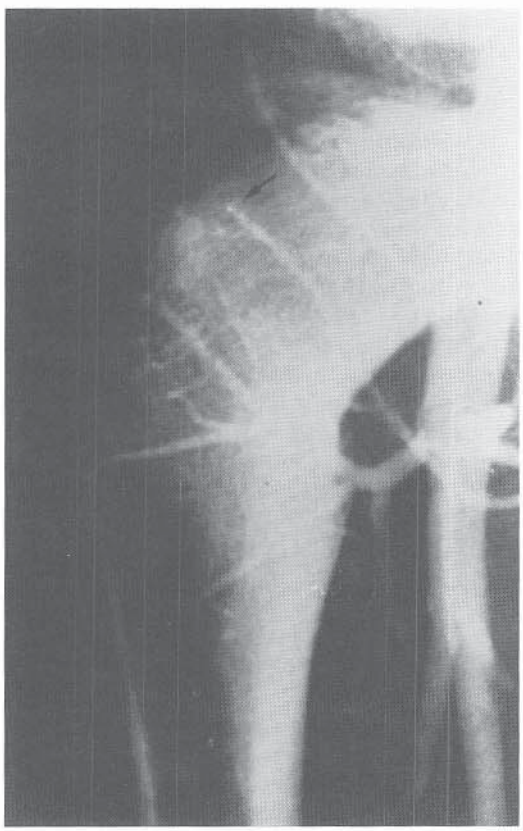


FIGURE 16–11 Arteriogram showing reduced blood flow in the medial circumflex artery (*arrow*). (From de Camargo FP, de Godoy RM Jr, Tovo R: Angiography in Perthes' disease. Clin Orthop 1984;191:216–220.)

Based on two separate investigations, Inoue and associates concluded that two infarcts of the femoral head were required for Legg-Calvé-Perthes disease to develop.[130] In an experiment performed on young dogs, a single induced infarct did not reproduce the changes typically associated with the disorder in humans. However, when a second infarct was created 4 weeks after the first, the resultant changes were comparable to those seen in humans. Additionally, on examining femoral biopsy specimens obtained from 51 patients, Inoue and colleagues found that 51 percent of the specimens displayed changes similar to those seen with double infarctions (Fig. 16–12).

Other experimental studies have also found that a single episode of infarction did not reproduce the anatomic changes seen in Legg-Calvé-Perthes disease[17,18,163,170,194,238] and that multiple infarcts were necessary to produce the characteristic pathologic picture of the disease.[74,248] In an analysis of six femoral heads and five core biopsy specimens obtained from patients with Legg-Calvé-Perthes disease, Catterall and associates found vascular conditions ranging from ischemic arrest without infarction to multiple, complete infarctions of the epiphyseal bone.[39]

VENOUS DRAINAGE OF THE FEMORAL HEAD AND NECK

Abnormal venous drainage of the head and neck of the femur has also been noted in patients with Legg-Calvé-Perthes disease. These studies take on new and added importance in light of the recent findings of frequent coagulopathies and abnormal lysis of clots in children with Legg-Calvé-Perthes disease.

Venous drainage normally flows through the medial circumflex vein. However, in patients with Legg-Calvé-Perthes disease, there is increased venous pressure in the affected femoral neck and associated venous congestion in the metaphysis, and venous outflow has been found to exit more distally through the diaphyseal veins (Fig. 16–13).[94,134,171,265,271] In a study of 55 hips, Heikkinen and associates noted these abnormal venous flow patterns in 46 of the femoral necks during the initial and fragmentation stages and in just less than 50 percent of the necks during the restitution stage.[114] Those hips with the most disrupted venous drainage also displayed the most severe radiographic changes. After the hips had healed, venous drainage returned to normal.

Although these abnormalities in venous outflow are a consistent finding in Legg-Calvé-Perthes disease, it is still not known whether they are a cause of the disorder or the result of some other factor. In a study in which silicone was injected into the femoral neck of a dog model, venous outflow was obstructed, intraosseous pressure increased, and AVN was produced.[171] These results suggest that venous obstruction is a causative factor.

ABNORMAL GROWTH AND DEVELOPMENT: THE "PREDISPOSED CHILD"

Patients with Legg-Calvé-Perthes disease share particular growth and development abnormalities, indicating that some children may be predisposed to developing the disease.[30] A delay in bone age relative to the patient's chronological age is the most commonly observed abnormality and

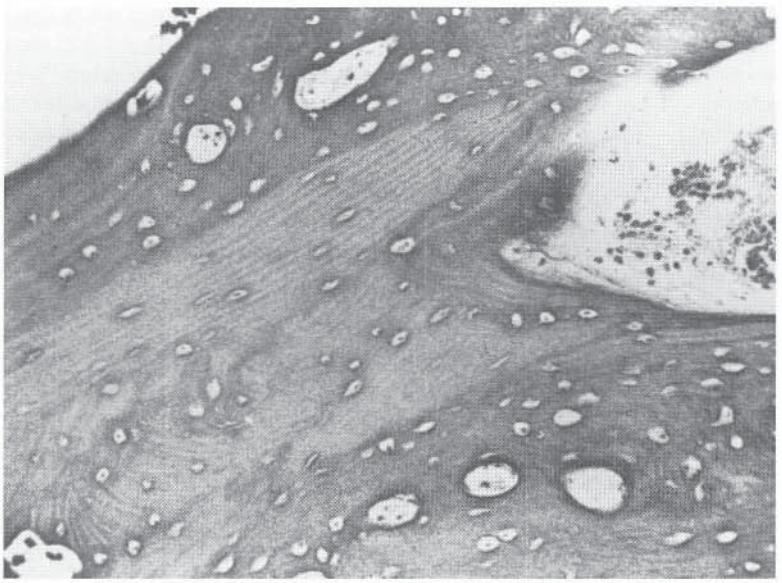


FIGURE 16–12 Femoral biopsy specimen showing dead lamellar bone with a thick surface layer of dead woven bone in a patient with Legg-Calvé-Perthes disease, 14 months after the onset of symptoms. These changes are similar to those seen with double infarctions. (From Inoue A, Freeman MA, Vernon-Roberts B, et al: The pathogenesis of Perthes' disease. J Bone Joint Surg 1976;588:453.)

is seen in the early years of the disorder (Fig. 16–14A).[73,83] For example, the carpal bone age is frequently 2 or more years behind the chronological age. Children diagnosed with the disease before age 5 years have shown an increase in bone-age delay over the following 4 to 5 years, whereas those diagnosed after age 8 years had a decrease in the delay over the ensuing years.[21] A radiologic pause in the bone age has also been observed, with particular carpals affected more than others.[156] This phenomenon had previously been referred to as "skeletal standstill."[110] Although growth of the triquetral and lunate is significantly delayed, the capitate and hamate are not affected. In patients with bilateral disease, there is greater delay in bone maturation of the trapezoid, growth of the carpals is more delayed than growth of the radius and ulna, and the onset of ossification in the carpals occurs later than in patients who have unilateral disease.[156,157] In the later stages of the disease, bone maturation accelerates until the bone age matches the chronological age (Fig. 16–14B). Although it is clear that most patients with Legg-Calvé-Perthes disease have unique bone maturation delays, how this delay relates to the pathogenesis of the disease is still uncertain.

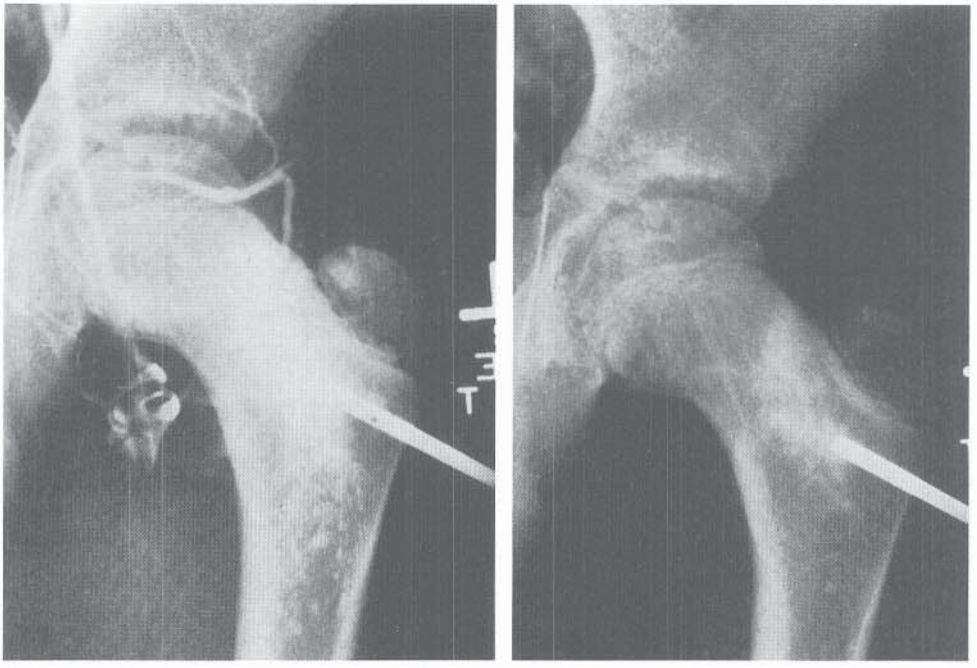
A number of other growth abnormalities have also been reported in children with Legg-Calvé-Perthes disease. The birth weight of children with the disorder was found to be significantly lower than that of unaffected children.[188] Wynne-Davies and Gormley, however, were not able to correlate low birth weight with subsequent short stature.[292] At the time of developing the disorder, some children are shorter than expected for their age, a condition that continues for the rest of their lives.[73,284,292] Cameron and Izatt noted that boys were approximately 1 inch shorter than their peers, while girls were about 3 inches shorter.[35]

After children experience a period of growth arrest, they may undergo a period of accelerated bone maturation.[66] Children with the disorder have also been found to be in the low to normal percentiles when their stature was compared with their chronological age. Growth studies have reported that soon after being diagnosed with Legg-Calvé-Perthes disease, patients had a period of slowed growth velocity followed by a "catch-up" phase. There was normal onset of puberty, and by the time the children were 12 to 15 years old, their stature and bone age were the same as their peers'.[157] Another study found that children diagnosed with the disorder before 6 years of age experienced a premature, unsustained acceleration in pubertal growth.[36] In addition, the hands and feet of children with the disorder are often smaller than those of siblings or peers.[101]

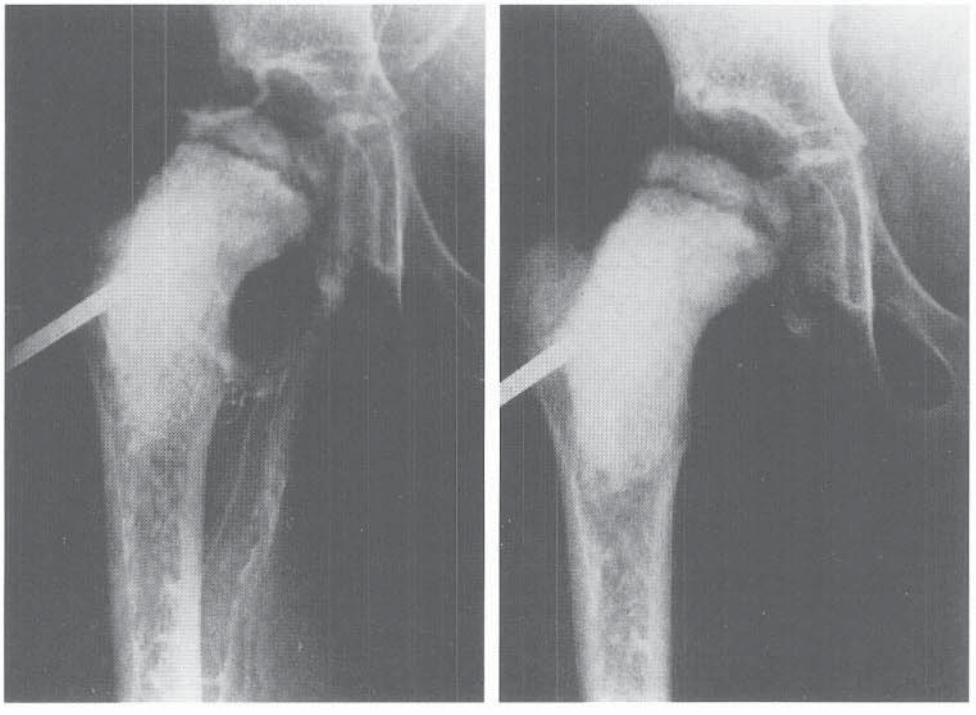
Some explanation for the abnormal growth patterns comes from studies showing growth hormone abnormalities in children with Legg-Calvé-Perthes disease. A marked decrease in somatomedin A has been noted in patients with Legg-Calvé-Perthes disease.[270] Normal somatomedin C activity (as assessed by radioimmunoassay) has been reported,[153] but others have found lower plasma somatomedin C levels in children with Legg-Calvé-Perthes disease (the patients were small in size, with immature bone ages).[191] A possible explanation for this is that plasma somatomedin C increases in normal children as they grow older, but no such increase is noted in children with Legg-Calvé-Perthes disease during the initial stages of the disorder.[199] Previous studies have reported normal thyroid function in patients with the disorder; however, other studies have noted elevated plasma levels of free thyroxin and triiodothyronine in these children, compared with levels in control groups.[197,221,231] Although the hormonal levels were increased in these patients, they were still within normal limits.

TRAUMA

Other authors have suggested that trauma in the predisposed child precipitates AVN of the femoral head and the development of Legg-Calvé-Perthes disease.[43,57] In the developing proximal femur, the major lateral epiphyseal artery must course through a narrow passage, which could make the vessel susceptible to disruption if there was trauma to this area. The risk of vascular interruption secondary to trauma is increased because these vessels must penetrate the thick cartilage of the femoral head.[43] Despite these findings, the premise that trauma is a causative factor of the disease is



A



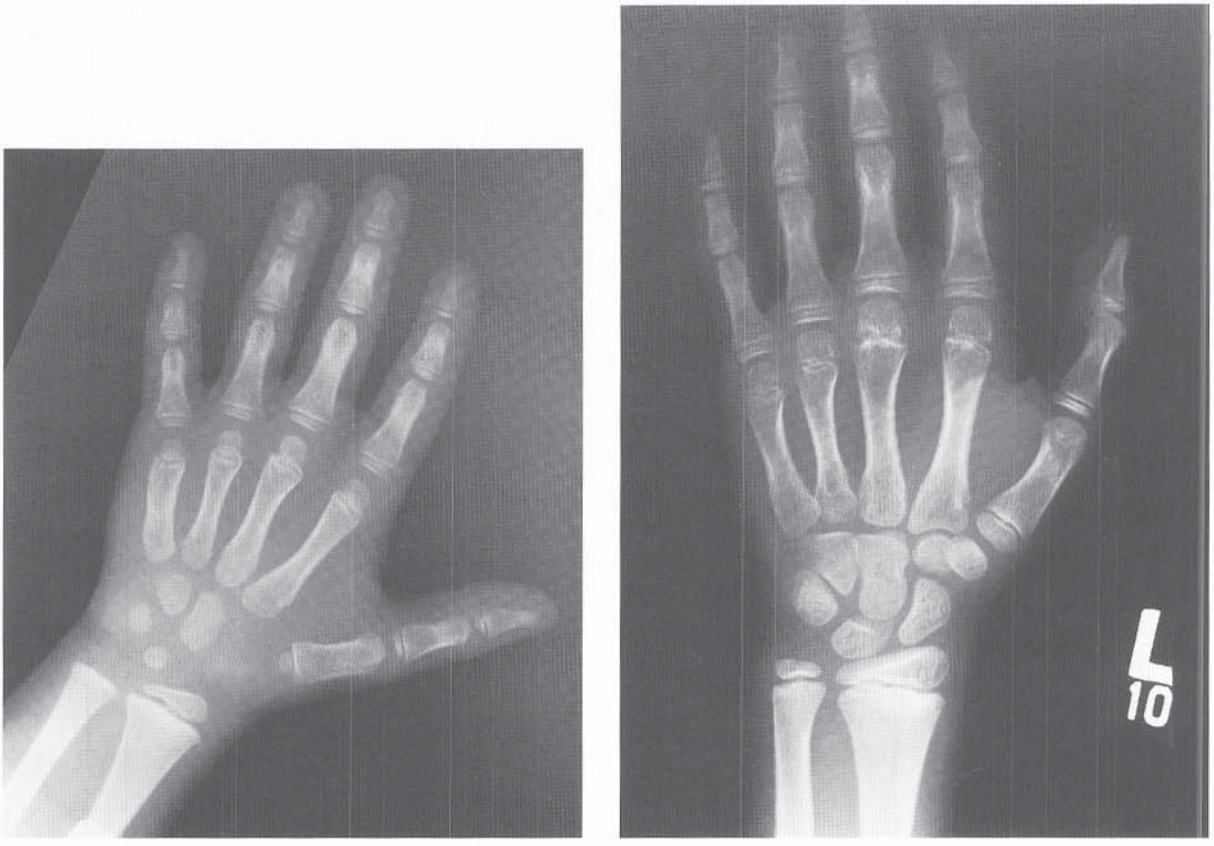
B

FIGURE 16–13 **A,** Venogram of a normal hip. *Left:* Appearance immediately following injection of 5 mL of contrast medium. The gluteal veins are present and normal. The ischial, medial circumflex, and lateral circumflex veins are clearly displayed, and there is no diaphyseal reflux of the contrast agent. *Right:* Five minutes after injection, the contrast agent is almost completely cleared. **B,** Venogram of a diseased hip. *Left:* Appearance immediately following injection of contrast medium. The gluteal and medial circumflex veins are absent. The lateral circumflex vein and numerous nutrient veins are seen. There is significant diaphyseal reflux of the contrast agent. *Right:* Five minutes after injection, a considerable amount of contrast agent remains within the femoral neck and shaft. (From Green NE, Griffin PP: Intra-osseous venous pressure in Legg-Perthes disease. J Bone Joint Surg 1982;64-A:666.)

particularly difficult to substantiate because frequent, mild trauma is a common part of childhood.

HYPERACTIVITY OR ATTENTION DEFICIT DISORDER

Many children with Legg-Calvé-Perthes disease tend to be extremely active physically, and some are pathologically hyperactive or suffer from attention deficit disorder. In a study of 68 children treated with recumbency, 48 became extremely hyperactive as they got older, were in a perpetual state of motion, and were prone to climbing and falling.[90] In their study of the behavioral characteristics of children with Legg-Calvé-Perthes disease, Loder and associates found that one-third of those in the series scored abnormally high on psychological profiles identified with attention deficit–hyperactivity disorder.[174] Because of its high prevalence in this patient population, hyperactivity may



A B

FIGURE 16–14 **A,** Radiograph of the hand and wrist taken at onset of Legg-Calvé-Perthes disease in patient whose chronological age was 6 years 6 months and whose bone age was 4 years. **B,** Radiograph of same patient at a chronological age of 12 years. The bone age was now 11 years 6 months. (From Herring JA: Legg-Calvé-Perthes Disease. Rosemont, IL, American Academy of Orthopaedic Surgeons, 1996.)

have a role in the etiology of the disease, but its precise role remains to be defined.

HEREDITARY INFLUENCES

Some authors have suggested genetic factors as a possible cause of Legg-Calvé-Perthes disease. Stephens and Kerby tracked the disorder through four generations of one family in which 28 of the 63 members investigated were affected.[262] Wansborough and associates reported that approximately 3 percent of siblings born after the index patient developed the disease.[283] A polygenic inheritance was suggested by Gray and associates when they noted that Legg-Calvé-Perthes disease occurred in 0.8 percent of second- and third-degree relatives.[93] After reviewing their own personal series of patients and six other published series, Burch and Nevelos concluded that an X-linked recessive factor and an autosomal homozygous allele was present in each genotype of the disease.[29] A greater number of congenital abnormalities (e.g., hemivertebrae, deafness, Rubinstein-Taybi syndrome, imperforate anus, pyloric stenosis, epilepsy, congenital heart disease, undescended testicle, short tibia) have been observed in patients with Legg-Calvé-Perthes disease.[103] These findings, however, could be associated with genetic or environmental factors, and their significance is unknown. A multifactorial inheritance pattern has also been described in patients with Legg-Calvé-Perthes disease in which the ratio for first-, second-, third-, and fourth-degree relatives was 35 : 4 : 4 : 1, respectively.[102] In one report, four members of one family had Legg-Calvé-Perthes disease, but the authors were not sure whether this incident was due to genetic or to environmental influences.[206]

Other studies, however, indicate that a genetic component is minor and that there is little if any hereditary influence. Although there are a few reports of the disorder in twins,[59,80,128] these cases were selected because of their concordance. In studies of unselected cases, only one of each pair of twins had the disease.[73,292] In a report on the mating of affected persons who had monozygotic twins in accord for Legg-Calvé-Perthes disease, the risk in siblings was less than 1 percent and the risk in children of an affected parent was only 3 percent.[106] In another review of 310 patients with the disorder, there was no indication of a hereditary factor.[292] Within the study population, no parents and only 1.6 percent of siblings had the disease, and the incidence among second- and third-degree relatives was the same as that of the general population. The authors explained the high familial incidence of the disease reported in other studies as possibly due to unintentional inclusion of patients with familial epiphyseal dysplasias.[51,292] When these misdiagnosed cases are eliminated, the true incidence of Legg-Calvé-Perthes disease among first-degree relatives is very low.[73,292]

ENVIRONMENTAL INFLUENCES

Environmental influences have been reported, primarily in Great Britain, as a possible cause of Legg-Calvé-Perthes disease, with an especially high occurrence in particular urban areas. A high incidence of the disease has been reported among children living in the inner city of Liverpool.[98,100,101] Children with the disorder were found to have decreased sitting height and smaller feet compared with a control group. Because the occurrence was also highest among those in the lower socioeconomic groups, the investigators surmised that nutritional factors were involved. Low levels of blood manganese were subsequently found in affected children.[99] Additional common factors included being from a lower socioeconomic group, residing in a Council house, having unemployed parents, and living in a large family. These social findings suggested a nutritional influence in the etiology of the disorder.[97] Environmental factors have not been observed in many other regions of the world, though, and their role in the cause of Legg-Calvé-Perthes disease remains unclear.

SYNOVITIS

Synovitis of the hip occurs early in Legg-Calvé-Perthes disease and may precede radiographic changes. Thus, many investigators have studied the incidence of the disorder following synovitis to evaluate the role that synovitis might play. Findings from these studies clearly demonstrate that while synovitis may be the first manifestation of the disease, it is rarely, if ever, the cause of Legg-Calvé-Perthes disease. In numerous studies, the rate of development of Legg-Calvé-Perthes disease after an episode of synovitis has ranged from less than 1 percent to 4 percent.[113,141,161,176,192,254]

Many children exhibit abnormal femoral head vascular hemodynamics during the acute stage of synovitis. However, in most cases normal circulation returns, AVN does not occur, and the patients do not develop Legg-Calvé-Perthes disease. Houben and associates noted a decrease in technetium phosphate uptake during the early stages of arterial filling of the femoral head in 63 percent of children with synovitis.[125] Eight days later, only 23 percent were abnormal, and 15 days later, only three patients continued to have asymmetry. Of these three, two subsequently developed Legg-Calvé-Perthes disease. In a study of nine children with synovitis, slipped capital epiphysis, hemarthrosis, and post dislocation, Glefand and associates reported that all showed an initial, transient loss of Tc-99m diphosphonate uptake in the femoral head.[84] Complete revascularization subsequently occurred in six of the children, while three developed AVN. Wingstrand and associates performed scintigraphy in 25 children and noted that four had significantly abnormal isotope uptake.[290] Six weeks later, uptake had returned to normal in three of the four patients. The fourth child continued to have abnormal uptake and eventually developed Legg-Calvé-Perthes disease. Magnetic resonance imaging (MRI) has also been used to study children with synovitis.[209] Although low-signal-intensity patterns on T1-weighted images indicated AVN, findings returned to normal in all cases and the patients' symptoms resolved. The MRI results may have been due to transient edema of the bone marrow.

Studies have also been conducted to evaluate the position of the hip and its correlation with intra-articular pressure in patients with synovitis. A slight increase in pressure has been noted during the early stages of Legg-Calvé-Perthes disease.[140] Pressures were moderately increased in patients with idiopathic synovitis and extremely high in those with septic arthritis. The pressure increased markedly when the hip was extended and internally rotated. This positional increase in pressure has been reported by others,[277] which suggests that when a patient is immobilized or placed in traction, the hip should always be in a position of at least 30 degrees of flexion.[140]

A "UNIFYING" HYPOTHESIS

A unifying hypothesis (Fig. 16–15), based on what is known today, would state that lack of thrombolysis in the venous drainage of the femoral neck increases pressure in the femoral head circulation, resulting in AVN. Antecedent trauma might precipitate the avascular event.

Pathology

PATHOLOGIC ANATOMY

Legg-Calvé-Perthes disease is not a terminal condition, and surgery is rarely performed directly on the head of the femur. Nevertheless, over the years a number of pathologic specimens of the femoral head of patients with the disorder have been obtained and examined. Perthes in 1913 was the first

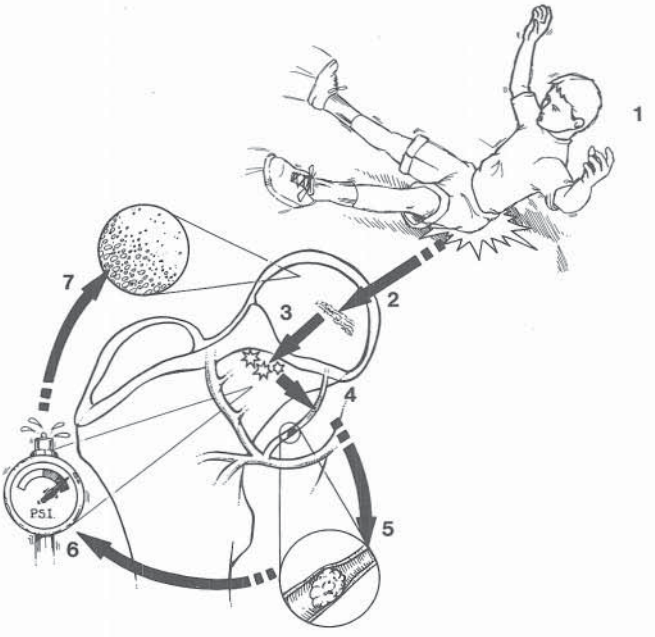


FIGURE 16–15 A "unifying" hypothesis as to the etiology of Legg-Calvé-Perthes disease: *1*, A highly active child falls on the hip. *2*, The fall causes minor trauma to the capital femoral epiphysis. *3*, There is minor injury in the metaphyseal area, inducing an increased tendency to form clots. *4*, The child is deficient in one of the proteins of the clot-lysing system (e.g., proteins C and S). *5*, Clotting occurs in the venous system of the metaphysis. *6*, Venous pressure rises in the femoral neck. *7*, Clotting propagates into the femoral head, where infarction occurs. Over time, the femoral head becomes more radiodense, and the changes associated with Legg-Calvé-Perthes disease ensue.

to describe the pathology of the disorder.[212] His explanation was based on examination of the femoral head of a child who died accidentally while the hip was in the middle stages of disease progression. In 1914 Perthes's colleague, Schwarz, provided his own description of a pathologic specimen in which he indicated that the findings were most likely a result of disrupted blood flow to the femoral head (see Fig. 16–4).[250]

In 1921 Phemister reported his findings from the curettage of a 10-year-old patient who had had symptoms for 8 months.[216] He noted areas of necrotic bone, granulated tissue, new bone formation superimposed on old bone, and osteoclasts. These findings were interpreted as the result of an inflammatory or infectious process. Subsequently, Phemister described the varying degrees of AVN of the femoral head that are found in the different stages of the disease,[217] a finding confirmed by Zemansky.[294] That same year, Riedel described the histologic findings in two cases.[235] He noted that the articular cartilage was thickened, the junction between the bone and articular cartilage was filled with blood, the physeal plate was destroyed, numerous cartilage rests were present, dead bone was surrounded by rich granulation tissue, and there were numerous giant cells. Farther away from the disease process the marrow was fibrotic, with inflammatory infiltrates. Riedel also noted blastic and clastic changes occurring simultaneously in the same bone trabeculae. In his second specimen he reported finding regenerated subchondral cartilage, atrophied cells, and some inflammatory cells.

Jonsäter performed core biopsies of the femoral head in various stages of the disease.[136] The histologic picture in the initial stage (avascular necrotic) was dominated by significant necrosis of bone and bone marrow (Fig. 16–16). Other findings included crushed trabeculae, absent or pyknotic nuclei in the osteocytes, a necrotic mass consisting of dead marrow and pulverized particles of dead bone (*Trümmer-*

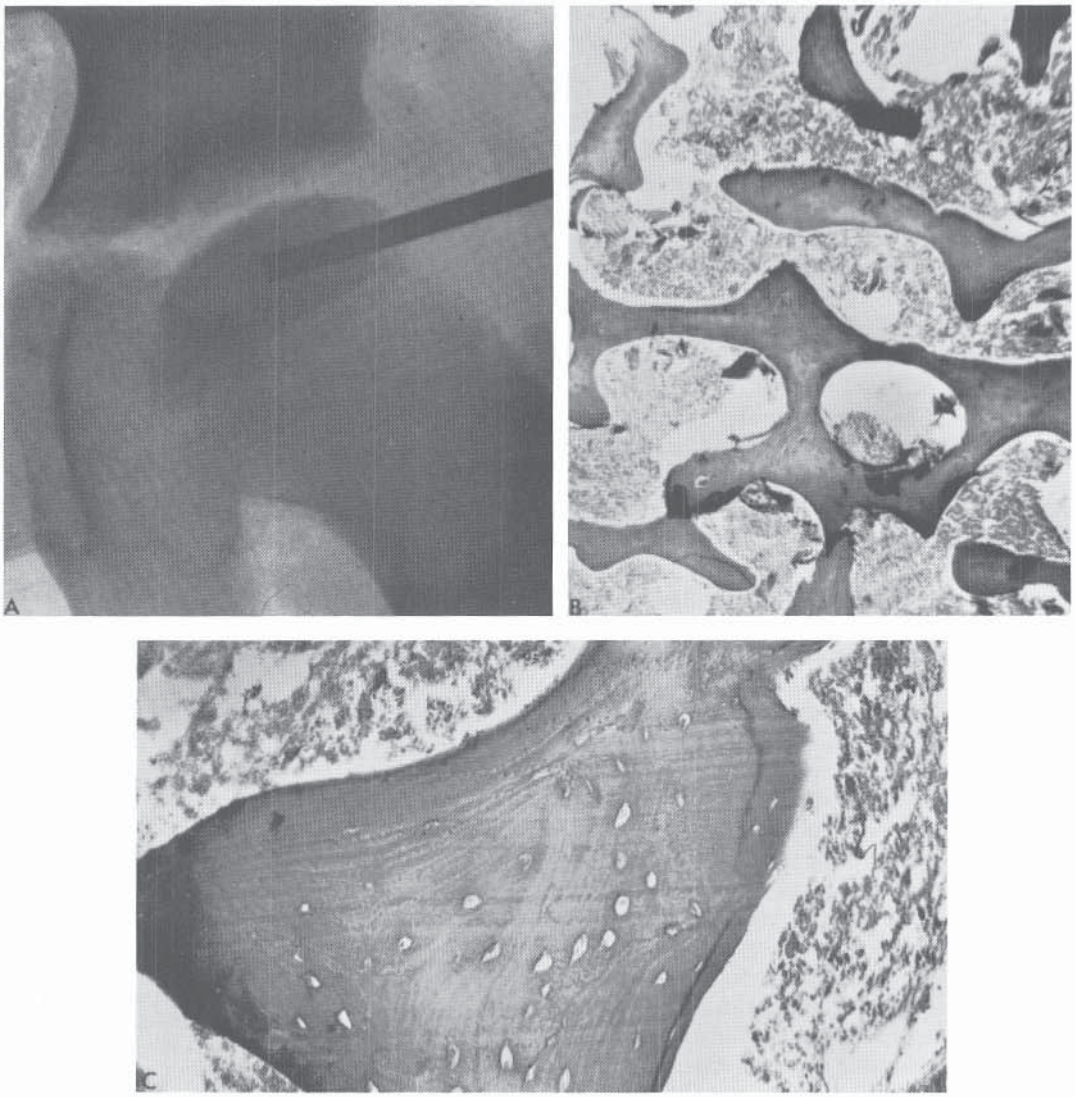


FIGURE 16–16 Histopathologic findings in the avascular or necrotic stage of coxa plana (Waldenström's initial stage). **A,** Radiograph of the hip showing the markedly dense and flattened head. **B,** Photomicrograph (×35). Note the crushed trabeculae, which are totally necrotic. Bone marrow is absent; there is no sign of reaction. **C,** Greater magnification (×200) reveals the absence of nuclei in the dead trabeculae surrounded by amorphous necrotic masses. (From Jonsäter S: Coxa plana: a histo-pathologic and arthrographic study. Acta Orthop Scand 1953;suppl 12:29.)

mehl) accumulated in the marrow spaces, occasional remnants of living bone, no evidence of bone regeneration, degenerative changes in the basal layer of articular cartilage (where the bone necrosis reached as far as the cartilage-bone junction), a thickened peripheral cartilaginous cap, and physeal irregularity in the columnization of the cartilaginous growth cells. Grossly, the bone was softer than normal. Arthrographic studies demonstrated that the cartilaginous head retained its spherical shape during the initial stage.

In the second, or resorption, stage, revitalization of the femoral head occurred through "creeping substitution" and apposition. Histologically, dead bone was infested with vascular connective tissue, which was actively resorbed by osteoclasts and replaced by newly formed immature bone (Fig. 16–17). The stage of repair could vary in different parts of the femoral head because of repeated episodes of infarction.[39] The cartilage changes were similar to those seen in the necrotic stage. There was a loss of epiphyseal height due to collapse of the bony trabeculae and in part to progressive resorption of the fragmented necrotic bone from its deep surface.[39] In the reparative stage, normal bone appeared and replaced the diseased bone (Fig. 16–18).

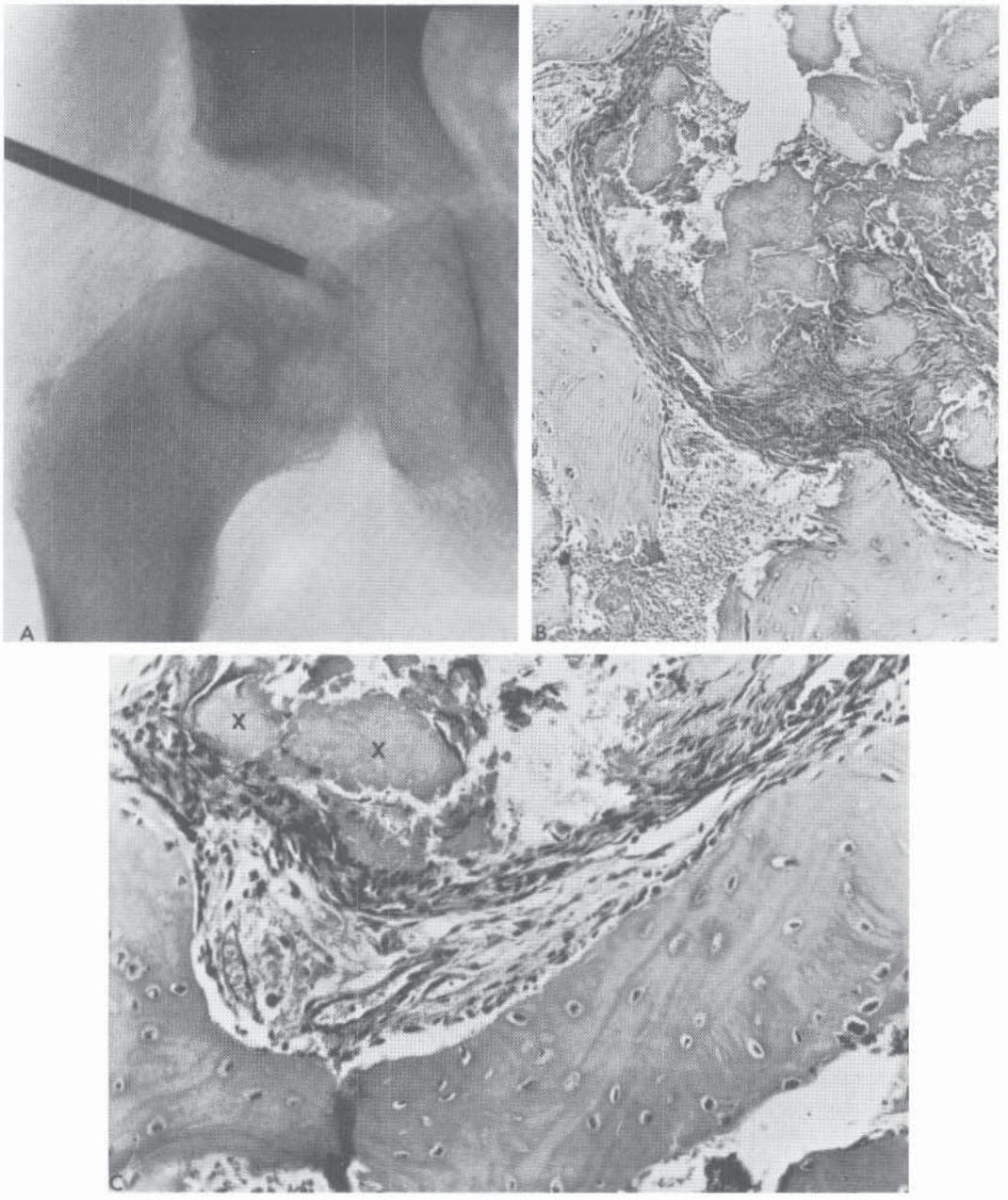


FIGURE 16–17 Histopathologic findings in the regenerative stage of coxa plana (Waldenström's fragmentation stage). **A,** Radiograph of the hip showing the fragmented and flattened femoral head. **B,** Photomicrograph (×100). Note, in the upper right portion, the vascular connective tissue growing into the dead bone. Newly formed bone is seen at lower left. **C,** Photomicrograph (×200). *X*'s mark the remnants of necrotic bone, below which is the markedly vascular connective tissue. Immature bone with large, well-stained nuclei is seen in the lower part of the picture. (From Jonsäter S: Coxa plana: a histo-pathologic and arthrographic study. Acta Orthop Scand 1953;suppl 12:39.)

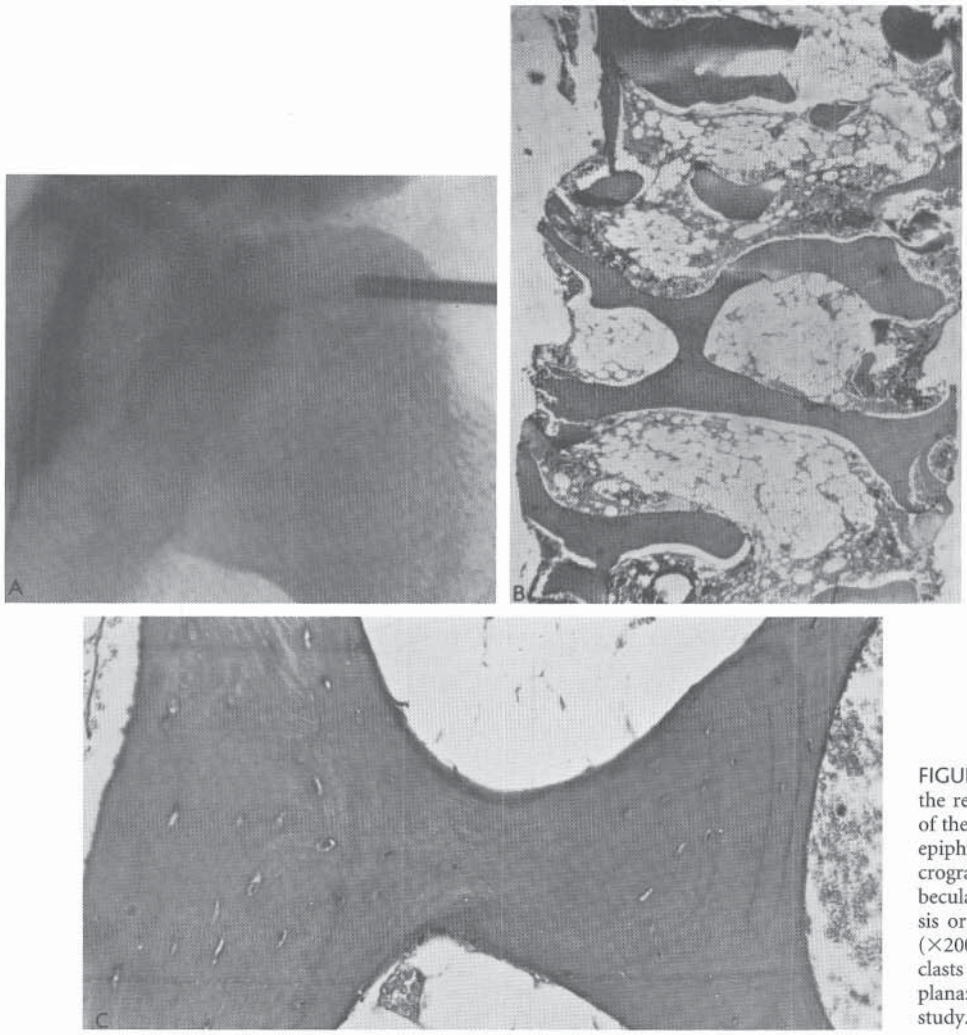


FIGURE 16–18 Histopathologic appearance in the residual stage of coxa plana. **A,** Radiograph of the hip. Note the normal bony structure. The epiphyses are somewhat flattened. **B,** Photomicrograph (×35) showing the normal bone trabeculae and marrow spaces. There is no necrosis or active regeneration. **C,** Photomicrograph (×200). Note the well-stained nuclei of the osteoclasts in the trabeculae. (From Jonsäter S: Coxa plana: a histo-pathologic and arthrographic study. Acta Orthop Scand 1953;suppl 12:51.)

Jensen and Lauritzen described pathologic findings in two children who died of unrelated causes, one age 4 years 10 months and the other age 6 years.[135] Both children had been treated for Legg-Calvé-Perthes disease for 18 months and their disease processes were in the reossification stage. The epiphysis had been replaced by cartilage, fibrous tissue, and granulation tissue, and there was extensive new bone formation (Figs. 16–19 and 16–20). The younger child had changes that indicated multiple episodes of ischemia, and a thrombolytic occlusion of the posterior inferior retinacular artery was found (Fig. 16–21). The authors did not find any evidence of systemic bone disease.

In 1982 Catterall and associates acquired six necropsy specimens and five core biopsy specimens (Fig. 16–22), which they compared with five normal control specimens.[39] The articular cartilage was thicker than normal in both the involved and uninvolved femoral heads. The growth plate of the uninvolved head was thinner than normal, with asymmetric cell columns and primary spongiosa present. Along the growth plate of the involved head there was a greater degree of interference with ossification, and columns of unossified cartilage extended into the metaphysis. These findings were located primarily on the anterior aspect of the femoral head in one group of specimens (Catterall group I).

Catterall noted that the trabeculae and marrow of the bony epiphysis were necrotic, while its subchondral plate was thickened.[39] Fresh bone was found appositionally on the surface of the necrotic trabeculae, and vascular tissue was present in the margin of the femoral head. A greater amount of vascular tissue, comparable to that of immature fracture callus, was found in specimens in which the disease process had been present for a longer period of time. In mildly affected heads (Catterall group I), the trabeculae were thickened and there were continuous layers of bony apposition, but no necrotic bone was present. The radiolucent zones of these specimens consisted of fibrocartilage and dormant ossification.

FINDINGS SUGGESTIVE OF A SYSTEMIC PROCESS

The pathologic changes, particularly those seen in the unaffected femur, may be indicative of a systemic cartilaginous disease process.[39] In the mild form of the disorder (Catterall group I), the disruption of blood supply to the femoral head may be sufficient to interfere with normal growth and ossification but not enough to cause an infarct. Using a dog model, Mickelson and associates compared pathologic

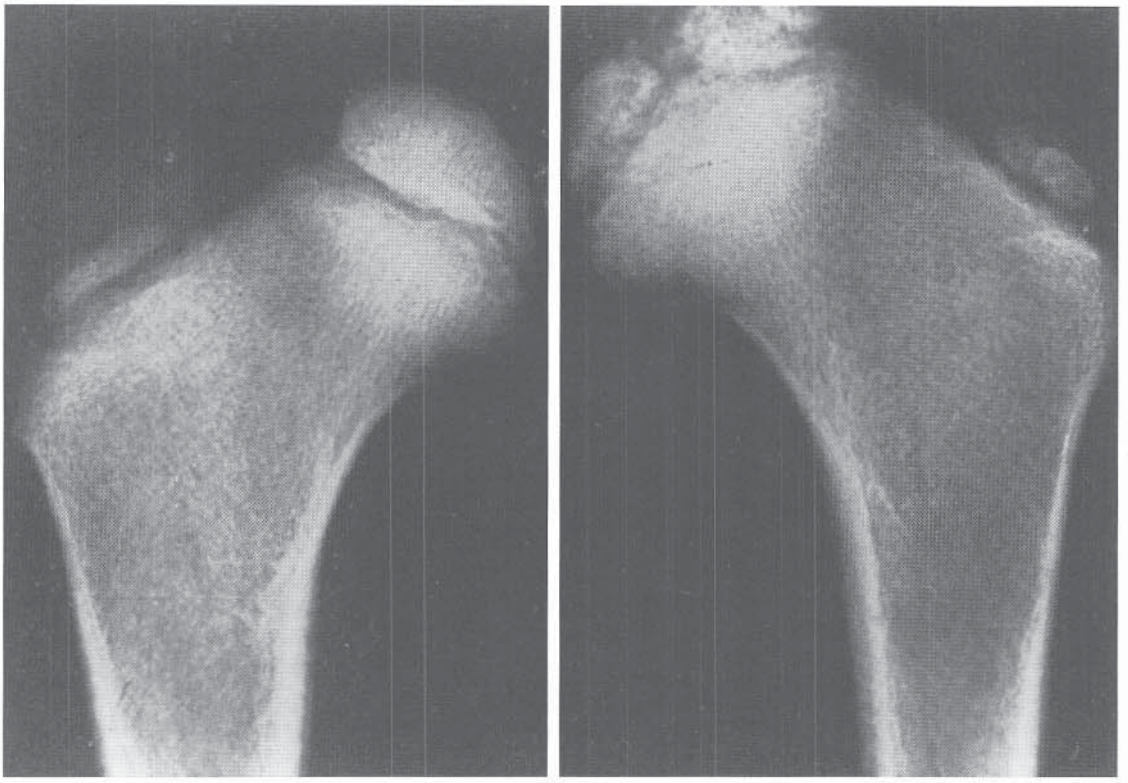


FIGURE 16–19 Postmortem radiographs of both upper femora from a young child. The left epiphysis exhibits fragmentation and irregularity, whereas the metaphysis is composed of normal bone structure. (From Jensen OM, Lauritzen J: Legg-Calvé-Perthes' disease: morphological studies in two cases examined at necropsy. J Bone Joint Surg 1976;58-B:332.)

findings commonly associated with Legg-Calvé-Perthes disease with those seen in naturally occurring AVN in canines.[185] They determined that flattening of the femoral head was caused by mechanical collapse, irregular growth, and disrupted endochondral ossification at the growth plate.

In a histochemical and ultrastructural examination of the epiphyseal cartilage and physis of the lateral portion of the femoral head and neck, Ponseti and associates found a thick region of hyaline (epiphyseal) cartilage under the normal articular cartilage (Fig. 16–23).[222] This zone of hyaline cartilage was composed of clearly separated sections of hypercellular and fibrillated cartilage with pronounced blood vessels. There also were numerous asymmetrically oriented large collagen fibrils and varying amounts of proteoglycan granules. In the fibrillar area there were a number of distinct findings: (1) increased amounts of proteoglycan, (2) a reduction in structural glycoproteins, and (3) collagen fibrils that differed in size from that of normal epiphyseal cartilage. The border of the lateral physis, which frequently was asymmetric with a significant decrease in collagen and proteoglycan granules, contained profuse amounts of large lipid inclusions. The investigators speculated that these findings represented a localized manifestation of a generalized, transitory epiphyseal cartilage disorder, comparable to changes observed in the vertebral end-plates in juvenile kyphosis. Ponseti and associates concluded that the epiphyseal change was primary, whereas femoral head collapse and necrosis could be caused by deterioration and disarray of the matrix of the epiphyseal cartilage and subsequent abnormal ossification.[222]

The manifestation of discrete abnormalities in the patient's opposite femoral head is another potential sign of systemic abnormalities in cases of Legg-Calvé-Perthes disease. Findings include heads not as round as controls (with anterior and lateral flattening),[5] and flattening or dimpling of the head in almost half of patients.[108] On examining biopsy specimens of the femoral head, Ippolito noted that occluded nutrient vessels of the head were surrounded by abnormal cartilage, which he regarded as part of a systemic abnormality of cartilage metabolism (a theory supported by Ponseti).[131,220]

FINDINGS SUGGESTIVE OF DOUBLE INFARCTION

A disorder similar to Legg-Calvé-Perthes disease was created in a dog model by inducing two separate infarcts, the second 4 weeks after the first.[130] A subsequent examination of 57 human biopsy specimens revealed findings associated with double infarction sustained at separate times in more than 50 percent (see Fig. 16–12). The primary finding was dead trabecular bone lying underneath dead woven bone. The investigators concluded that Legg-Calvé-Perthes disease was typically caused by repetitive infarctions occurring over a certain time period, a finding supported by other investigators.[74,248] Catterall and associates also proposed that moderate

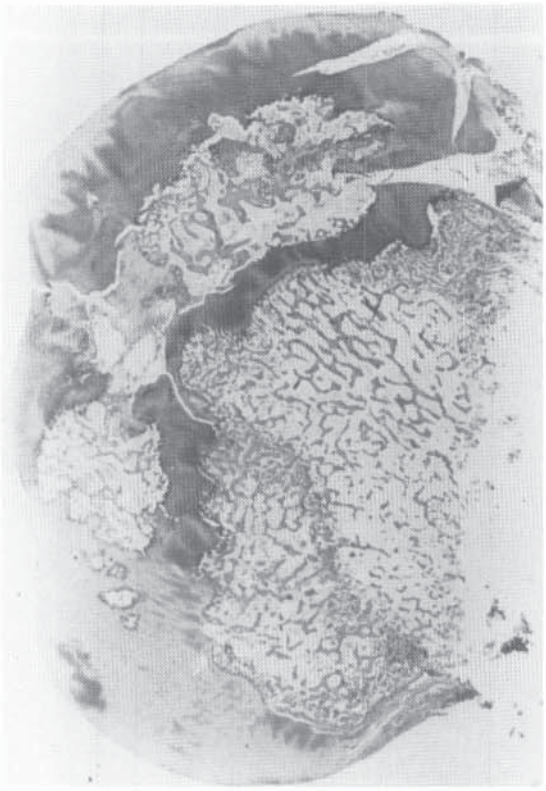


FIGURE 16–20 Low-power (×3) frontal plane view of the left femoral head showing strands and islands of cartilage and fibrous tissue in the epiphysis, and irregular bone structure. The pale area in the metaphysis indicates where bone marrow has been replaced by fat. (From Jensen OM, Lauritzen J: Legg-Calvé-Perthes' disease: morphological studies in two cases examined at necropsy. J Bone Joint Surg 1976;58-B:332.)

disease (Catterall groups II and III) involved two incidents of infarction, and that in severe disease (Catterall group IV) there were repeated episodes of infarction.[39]

METAPHYSEAL CHANGES

Four types of metaphyseal changes have been observed: (1) an overabundance of fatty marrow, (2) circumscribed osteolytic lesions with a sclerotic border, (3) a wide growth plate with disarrayed ossification and columns of unossified cartilage coursing down into the metaphysis, and (4) extension of the growth plate down the side of the neck of the femur.[39] The metaphyseal lucencies seen on radiographs may represent unossified cartilage from the growth plate.

Clinical Presentation

In 1920 Sundt described the clinical manifestations of Legg-Calvé-Perthes disease; his description still holds true today (Table 16–2).[264] He noted that the onset of the disorder occurred between ages 2 and 12 years (with a peak between 6 and 8 years), that boys were four times more likely to develop the disease than girls, and that 10 percent of patients had bilateral disease. Observations made by Catterall in 1982 support Sundt's findings. He reported that the mean age at onset was 6 years (82 percent of patients were between 4 and 9 years old) and that boys were 3.7 times more likely than girls to be affected.[37] Sundt also noted that because symptoms were frequently mild, considerable time often passed before the child was seen by a physician.[264] On presentation the child usually had a limp, and some complained of pain in the hip, thigh, or knee. The primary findings on physical examination were limited range of motion of the affected hip (particularly in abduction and internal rotation) and slight atrophy of the femoral muscles.

SYMPTOMS

Patients most often present with a limp, which is usually first noticed by a parent. The limp is exacerbated by strenuous physical activities and alleviated with rest. The limp may be more pronounced later in the day if the child has been involved in extended periods of ambulation. The second most frequent complaint is pain, which may be located in the groin, anterior hip region, or laterally around the greater trochanter. There often is referred pain to the knee, which may obscure the true nature of the disorder. The pain is aggravated by increased physical activity and is usually worse

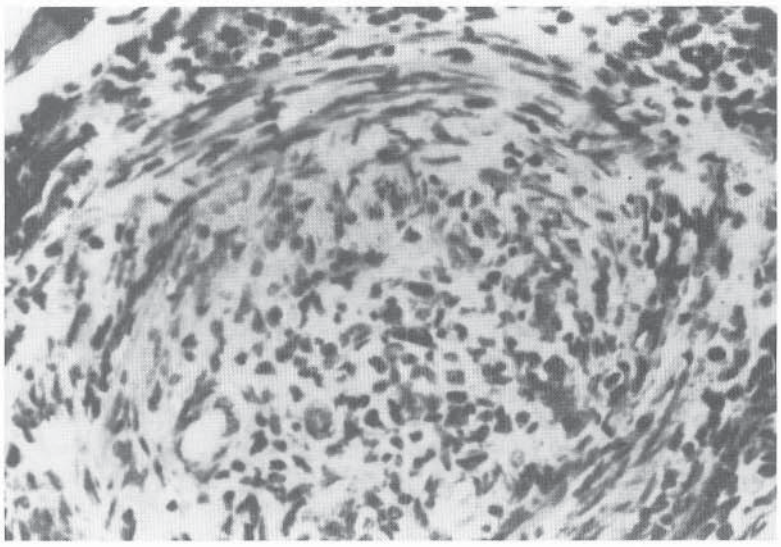
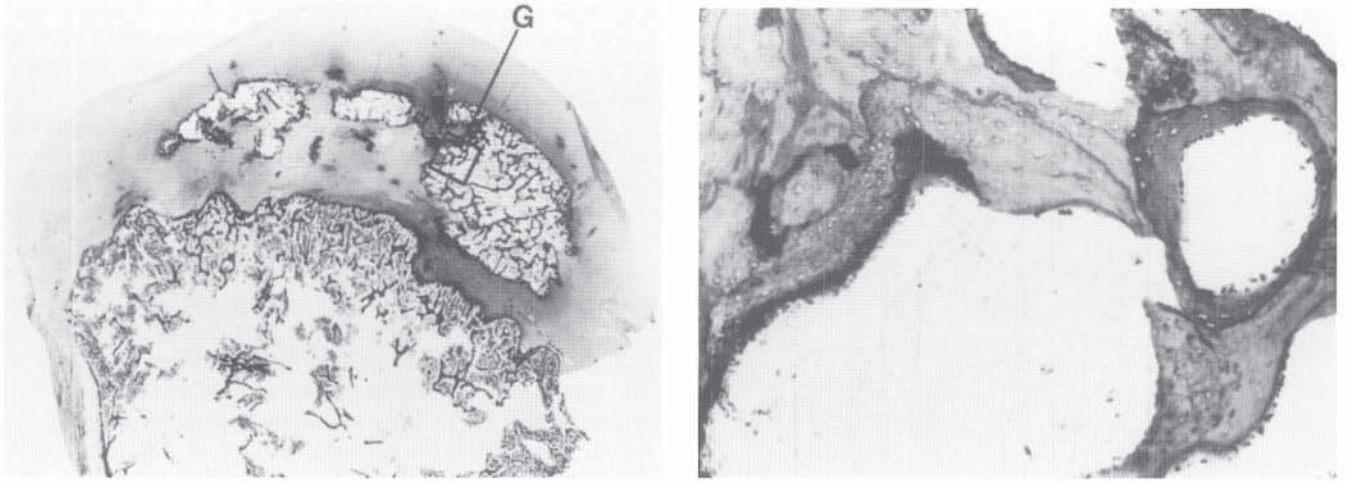


FIGURE 16–21 Magnification (×100) of specimen in Figure 16–20 showing the occluded posterior inferior branch of the retinacular arteries. There is a weak inflammatory reaction in the occluded lumen and around the vessel. The lumen is partially recanalized. (From Jensen OM, Lauritzen J: Legg-Calvé-Perthes' disease: morphological studies in two cases examined at necropsy. J Bone Joint Surg 1976;58-B:332.)



A B

FIGURE 16–22 Pathologic specimen of diseased hip. **A,** Central sagittal section of femoral head showing substantial thickening of the articular cartilage, which is perforated by a vessel supplying the epiphysis. The central dense area has been completely replaced by a fibrocartilaginous material. The trabecular bone on the medial and lateral aspects shows extensive avascular necrosis, as does the bone formed by endochondral ossification on the deep surface of the articular cartilage. **B,** High-power view of area *G* (*left*) shows grossly thickened trabeculae with extensive necrosis. The numerous cement lines suggest recurrent episodes of remodeling. The marrow is viable, and there is appositional new bone formation on the surface of the avascular trabeculae. (From Catterall A, Pringle J, Byers PD, et al: A review of the morphology of Perthes' disease. J Bone Joint Surg 1982;64-B:269.)

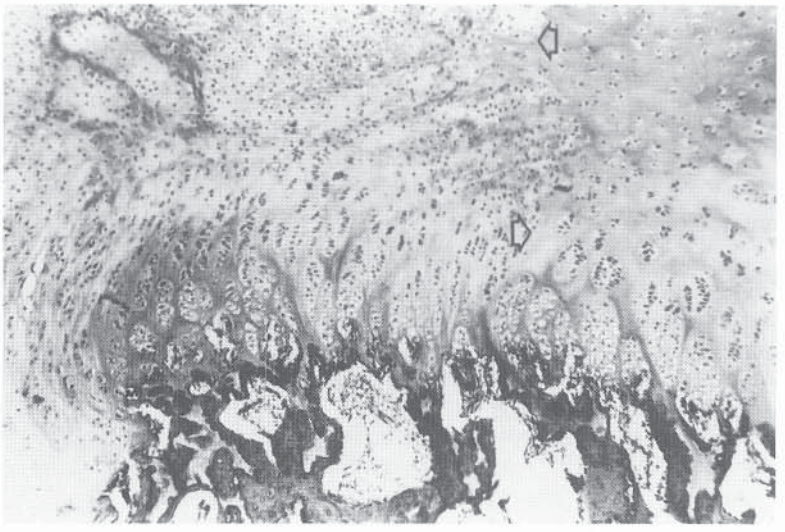


FIGURE 16–23 High-magnification view of the periphery of the physis. In the lateral (left) two-thirds of the figure, the resting zone is markedly abnormal, with profuse chondrocytes and a fibrillated matrix. Under the abnormal cartilage the physis is poorly organized and the ossification is irregular. The border between abnormal and normal cartilage is well defined (*arrows*). (From Ponseti IV, Maynard JA, Weinstein SL, et al: Legg-Calvé-Perthes disease: histochemical and ultrastructural observations of the epiphyseal cartilage and physis. J Bone Joint Surg 1983;65-A:797.)

TABLE 16–2 **Clinical Presentation of Legg-Calvé-Perthes Disease**

- Onset: between age 18 months and skeletal maturity (most prevalent between 4 and 12 years of age)
- Male sex prevalence: boys four or five times more likely to develop the disease than girls
- Involvement: bilateral in 10%–12% of patients
- Symptoms: limp that is exacerbated by activity and alleviated with rest; pain, which may be located in the groin, anterior hip region, or laterally around the greater trochanter; history of antecedent trauma
- Signs: abductor limp; decreased range of motion of the hip, especially on abduction and internal rotation (decreased range of motion transient early in disease, persistent later on); flexion/extension less affected

late in the day. The child will occasionally have night pain.

The patient or parents may recollect an isolated incident of trauma (often a fall or twisting injury) several months earlier, followed by the onset of a limp and hip pain. After a few days, these initial symptoms normally resolve completely. The patient often goes through periods of exacerbation and alleviation, with the symptoms waxing and waning. To relieve the pain, the child may deliberately decrease his or her normal level of activity. If the symptoms are mild, several months may pass before the child is seen by a physician.

The parent will often describe the child as being extremely active—in perpetual motion, running and jumping more than other children. Some patients may exhibit attention deficit disorder or may have been diagnosed as pathologically

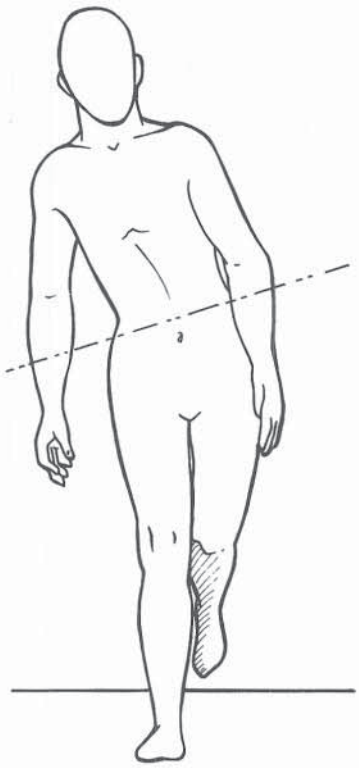


FIGURE 16–24 Illustration of a typical Legg-Calvé-Perthes limp, which is a combination of an antalgic gait and a Trendelenburg gait. In the stance phase of gait, the patient will lean the body over the involved hip to decrease the force of the abductor muscles and the pressure within the hip joint. (From Herring JA: Legg-Calvé-Perthes Disease. Rosemont, IL, American Academy of Orthopaedic Surgeons, 1996.)

hyperactive. The child may also be smaller in stature than peers. All of these details paint the classic portrait of the child with Legg-Calvé-Perthes disease—small, often thin, extremely active, constantly running and jumping, and limping after strenuous physical activities. Occasionally, however, a youngster with the disorder will be just the opposite—overweight and physically inactive.

Sometimes a relative of the patient may also have the disorder; however, a family history of Legg-Calvé-Perthes disease is rare. A strong family history of hip problems (particularly bilateral) may indicate a diagnosis of familial epiphyseal dysplasia.

SIGNS

The physical examination should include observation of the child's activity level. Some children will show other signs of injuries, such as scars or old fractures (sometimes a child will be wearing a forearm cast for a recent wrist fracture).

The child will have a limp, which he or she will often attempt to conceal during the examination. When possible, the physician should indirectly observe the child as the child is walking to the examination room. The child's limp is normally a combination of an antalgic and a Trendelenburg gait (Fig. 16–24). In the stance phase of gait, the child will often lean the body over the involved hip to decrease the force of the abductor muscles and the pressure within the hip joint. The Trendelenburg test will be positive on the involved side (Fig. 16–25). Depending on the duration and severity of the condition, the examiner may note varying degrees of atrophy of the gluteus, quadriceps, and hamstring muscles.

During the early phase of the disorder, reduction of hip motion (which is caused by muscle spasm) will vary from patient to patient. By examining the hip very gently, the clinician can usually elicit a greater range of motion because muscle spasm is avoided (the same holds true if the child is examined following a night's rest). Early in the disease process, only slight loss of motion is noted at maximum abduction and internal rotation. In these patients, the log roll test will be positive. Hip abduction is best examined in extension with the patient supine. Dropping the uninvolved leg over the side of the examining table helps stabilize the pelvis (Fig. 16–26). When the examination is done properly, the true abduction is less than originally appreciated. The degree of rotational loss is best assessed with the patient both prone and supine. Normally, at this stage of the disease, there is no flexion contracture.

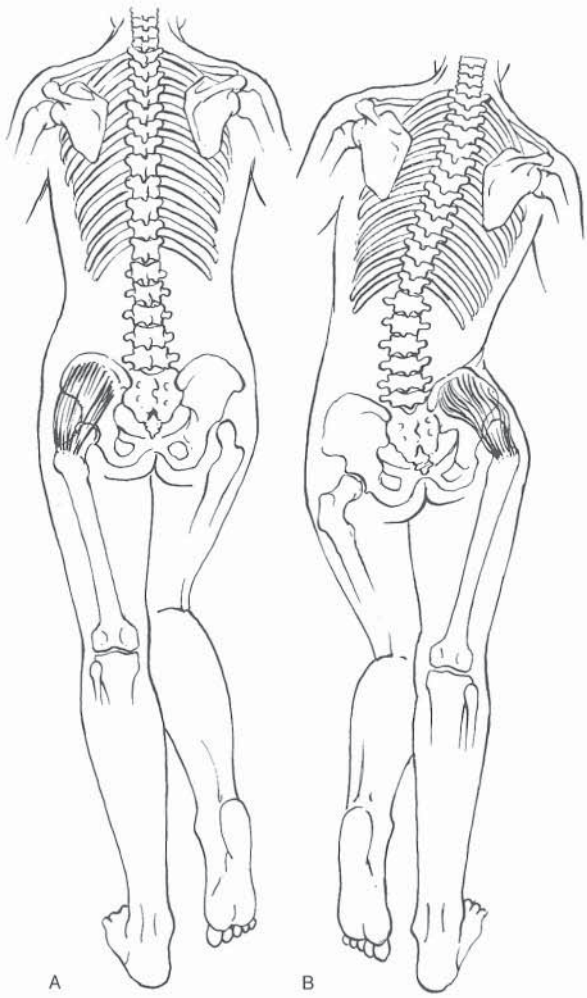


FIGURE 16–25 Positive Trendelenburg's test. **A,** As the patient stands with the weight on the normal hip, the pelvis is maintained in the horizontal position by contraction and tension of the normal hip abductor muscles. **B,** As the patient stands with the weight on the affected hip, the pelvis on the opposite normal side drops owing to weakness of the hip abductor muscles.

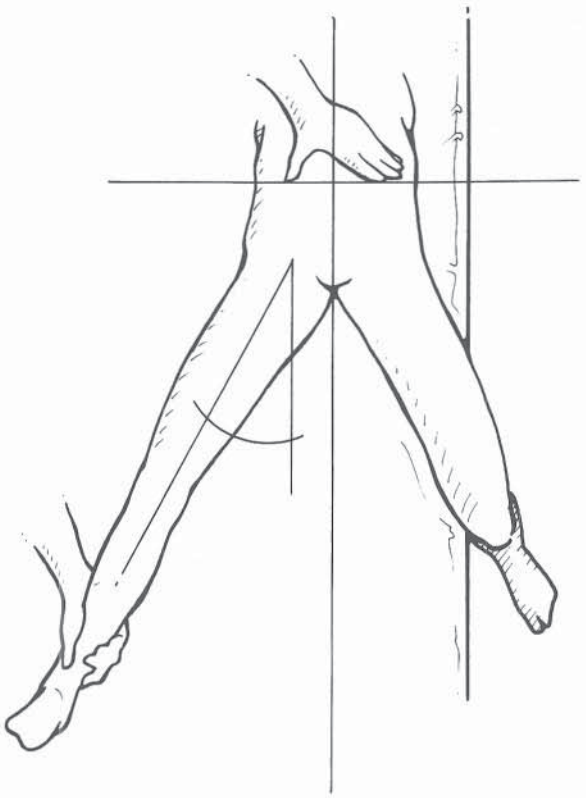


FIGURE 16–26 Hip abduction is best examined in extension with the patient supine. Dropping the uninvolved leg over the side of the examining table helps stabilize the pelvis. (From Herring JA: Legg-Calvé-Perthes Disease. Rosemont, IL, American Academy of Orthopaedic Surgeons, 1996.)

As the disorder progresses, findings on the clinical examination may be significantly different. Patients with mild disease may experience only slight loss at the extreme ranges of motion and may rapidly regain normal mobility of the leg. Those with more severe disease will have greater loss of motion, particularly of abduction and internal rotation. When the hip is flexed, it may go into obligatory external rotation. In severe cases adduction contractures develop and the child may lose all rotation of the hip. The range of flexion and extension, though, is seldom affected.

CLINICAL COURSE

Waldenström noted that the clinical course of the disease was quite variable.[282] He observed that while some children experienced only minor symptoms and minimal changes in the shape of the femoral head, most had a more severe course, resulting in pain while walking and greater loss of limb motion. Waldenström defined the stages of the disease as shown in Table 16–3. His classification has been modified by most authors to the four stages of initial, fragmentation, healing (reossification), and residual phases. In a retrospective study, we found that the time period from first radiographic evidence of disease to the start of fragmentation was a mean of 6 months (range, 1 to 14 months), the fragmentation phase lasted 8 months (range, 2 to 35 months), and the healing stage occupied 51 months (range, 2 to 122 months).[120]

Clinical findings correspond to some degree with the radiographic stages of the disease (Table 16–4). During the early stage of the disorder, radiographs show only increased density of the femoral head, and the patient may experience recurrent aggravation and alleviation of symptoms and signs. There may be only mild limp and pain for a period of time, interrupted by episodes of moderate discomfort lasting a couple of weeks. During this latter phase, a subchondral fracture is frequently noted on radiographs (Salter's sign), and the patient's clinical status may worsen.[247]

At the beginning of the fragmentation stage, the femoral head starts to collapse and may extrude from the acetabulum. The patient's limp and pain will be more pronounced, and there will be a greater loss of range of motion of the affected limb. Because the femoral head is deformed, resting the hip usually does not return normal motion to the joint. In mild cases, where there is minimal change in the shape of the femoral head, symptoms and signs may be limited. Patients who have a very brief fragmentation stage will be asymptomatic. In more severe cases, though, clinical symptoms and signs progressively worsen throughout the fragmentation phase.

The beginning of the healing stage is characterized radiographically by the development of new bone in the subchondral regions of the femoral head. By this time pain and limp have usually started to resolve, but there is still some limitation of motion. The degree of motion restriction is

TABLE 16–3 **Waldenström's Classification of the Stages of Legg-Calvé-Perthes Disease, Based on Radiographic Changes in the Femoral Head**

Stage	Features	Radiographic Changes
I	Evolutionary period (3–4 yr)	
	a. Initial stage (6–12 mo)	Femoral head density gradually increases; dense epiphysis with lucencies seen; density changes in metaphysis appear and later resolve.
	b. Fragmentation stage (2–3 yr)	
II	Healing period (1–2 yr)	Femoral head gradually reossifies and epiphysis becomes homogeneous.
III	Growing period (to conclusion of growth)	Femoral head reforms completely.
IV	Definite stage	

From Waldenström H: The definite form of the coxa plana. Acta Radiol 1922;12:384.

TABLE 16–4 **Association Between Clinical Findings and Radiographic Stages of Disease**

Stage	Clinical Findings	Radiographic Changes
Early initial stage	Recurrent aggravation and alleviation of symptoms and signs; mild limp and pain, interrupted by episodes of moderate discomfort lasting a couple of weeks	May show only increased density of femoral head
Latter initial stage	Clinical status may worsen.	Subchondral fracture (Salter's sign)
Fragmentation stage	Limp and pain more pronounced; greater loss of range of motion (depending on severity of disease)	Femoral head starts to collapse and may extrude from acetabulum.
Healing stage	Pain and limp usually start to resolve, but still some limitation of joint motion; child gradually resumes normal activities; symptoms normally absent as femoral head completely reossifies.	Development of new bone in subchondral regions of femoral head

directly related to the extent of change in the shape of the femoral head. Usually the child gradually resumes normal activities without complaints. Symptoms are normally absent as the femoral head becomes completely reossified.

If reossification in the central segment of the femoral head is significantly delayed, the patient may begin to experience pain after a number of years without symptoms. A loose fragment or osteochondritis dissecans lesion may subsequently develop in this soft portion of the head. The child will complain of locking and popping of the joint, and crepitus may be present on physical examination.

NATURAL HISTORY OF THE DISEASE

Because treatment programs were started as soon as Legg-Calvé-Perthes disease was identified, no definitive natural history studies exist of persons who were completely untreated. Thus, the natural history of the disorder has been difficult to accurately ascertain. We have been forced to surmise the likely natural history by reviewing groups of patients managed by programs that are now regarded as ineffective, with the presumption that these often intense and long-term treatment methods (e.g., nonweightbearing, cast immobilization, use of patten-bottom braces) did not affect the course of the disorder either favorably or adversely. It is improbable that these therapies had no effect whatsoever on patients' hips. However, we must accept, with some reservations, the findings of these reviews to gain knowledge of the disease process, because better alternatives are lacking.

We have learned that Legg-Calvé-Perthes disease varies considerably from patient to patient (Table 16–5). Over the course of the disorder, most children will experience moderate symptoms and endure 12 to 18 months of difficulty, followed by gradual resolution of the symptoms and a return to normal physical activities. At one extreme, some children will have minimal, transient symptoms, while patients with more severe involvement will have severe symptoms that persist into late adolescence or early adulthood.

The age of the patient at onset of disease is the most consistent and most frequently reported factor affecting the course of the disorder.[27,38,65,124,129,187] With a few exceptions, generally, the younger the child, the better the outcome. Those with early onset (particularly before age 6 years) usually experience mild disease, those with onset between 6 and 9 years of age normally have moderate symptoms, and children age 9 years or older at onset have the most severe course and worst outcome.

The extent of radiographic changes also varies, with the poorest results seen in hips with the greatest degree of involvement. A number of classification systems have been developed to estimate severity of disease based on radiographic findings. Legg described a "cap" head and a "mushroom" head, with the cap being more common and less severe than the mushroom.[168] Waldenström recognized three types of radiographic changes, with the most severe type occurring least often but resulting in the poorest outcome.[280] In Catterall's four-part classification system, coexisting risk factors increased the chances of a poor outcome.[38] In the lateral pillar classification system, the degree of involvement of the lateral segment of the femoral head is graded as A, B, or C, with C being the most severe.[120]

Outcome is also affected by how long a time period there is from onset of the disorder to complete resolution. The shorter the duration of disease, the better the final results.[118,120,121,172] In a series of patients with disease classified according to the lateral pillar system, group A hips healed in 37 months, with a 100 percent good/excellent outcome; group B hips healed in 50 months, with a 79 percent good/excellent outcome; and group C hips healed in 67 months, with a 29 percent good/excellent outcome.[120]

TABLE 16–5 **Natural History of Legg-Calvé-Perthes Disease**

- Disease severity: varies from mild to severe, with the majority of children experiencing moderate symptoms for 12 to 18 months, followed by complete resolution of symptoms and a return to normal physical activities.
- Patient's age: most consistent factor affecting course of disease. Usually those with early onset (before 6 yr) have mild disease, those with onset between 6 and 9 years have moderate symptoms, and those with onset at 9 years or later have the most severe course and worst outcome.
- Extent of radiographic changes: varies, with the poorest results seen in those hips with the greatest degree of involvement.
- Outcome: affected (in addition to patient's age at onset) by duration from onset of disease to complete resolution: the shorter the duration, the better the final results.

Imaging Studies

RADIOGRAPHIC STAGING OF DISEASE EVOLUTION

Waldenström's classification of the stages of Legg-Calvé-Perthes disease based on radiographic changes is summarized in Table 16–3.[279] Commonly, four radiographic stages—initial, fragmentation, reossification, and healed—are attributed to Waldenström.[286] A modified version of Waldenström's classification—initial, fragmentation, reossification (healing), and residual stages—is presented in Table 16–6. The more severe the disease, the longer is the duration of each stage, particularly the healing phase.[121]

Initial Stage. Slight lateralization of the femoral head in the acetabulum[31,280] and a slightly smaller ossific nucleus due to cessation of growth of the capital epiphysis[71] are the initial radiographic signs of Legg-Calvé-Perthes disease. The apparent widening of the medial joint space may be caused by synovitis and hypertrophy of articular cartilage (Fig. 16–27A). Hypertrophy of the cartilage has been verified in pathologic specimens.[39] Enhanced computed tomography (CT) has also demonstrated early lateralization secondary to swelling of the ligamentum teres.[142] Another sign, capsular swelling, was also reported as an early radiographic change. However, this finding was later shown to be a result of the position of the hip (abducted and laterally rotated) while the radiograph was made, rather than a pathologic condition.[28]

During the first stage of the disorder, other radiographic changes can be observed. In approximately one-third of cases, in the earliest phase of the disease a linear fracture is noted in the subchondral area of the femoral head. Known as Waldenström's sign, it is usually best seen on the frog-leg lateral view.[31,280] On rare occasions the fracture will fill with gas on this radiographic projection (presumably due to a vacuum phenomenon that generates intra-articular gas during forced frog-leg positioning). Another indication of early disease is a small increase in the density of the ossific nucleus that can be seen only on lateral views.[31]

Subsequently the femoral head appears progressively more radiodense. This increase in density is secondary to accumulation of new bone on the dead bone trabeculae in the head.[39,71] This occurs regardless of the extent of any ensuing femoral head collapse.[31,282] Additional radiographic findings include clearly delineated cysts and indistinct lucencies in the metaphysis. The end of this stage occurs when lucencies are seen within the ossific nucleus. This radiographic phase normally lasts a mean of 6 months, with a maximum time period of 14 months.[121]

Fragmentation Stage. The second radiographic stage is fragmentation (Fig. 16–27B).[282] Lucencies develop in the ossific nucleus (evolving rapidly within several months), while other sections remain sclerotic. Often a central dense fragment becomes demarcated from the medial and lateral segments of the femoral head.[31] In more severe disease there is no separation between the central and lateral portions, and there may not be any between the central and medial portions. The end of the fragmentation stage is marked by the appearance of new bone in the subchondral sections of the femoral head. This radiographic stage lasts a mean of 8 months (range, 2 to 35 months).[121] In mild disease fragmentation may be observed only on a frog-leg lateral view, while on the AP view a slightly mottled density may be noted. This indicates that only the anterior segment of the epiphysis is necrotic. In the mildest cases there is no actual fragmentation phase, and healing of the femoral head with gradual resolution of the dense areas follows the second radiographic stage.

Reossification (Healing) Stage. The third radiographic stage is the healing phase, which starts with the manifestation of new subchondral bone in the femoral head (Figs. 16–27C and D).[282] Reossification frequently starts in the center of the head and expands medially and laterally. Normally, the last areas to reossify are the anterior segment of the femoral head (demonstrated on the frog-leg lateral view) and the center of the head. The lucent portions of the femoral head gradually fill in with woven bone and, over time, the new bone remodels into trabecular bone. The healing phase concludes when the entire head has reossified. This radiographic stage lasts a mean of 51 months (range, 2 to 122 months).

In the past it was believed that the shape of the femoral head was minimally altered during the healing stage. We now know that the majority of heads gradually improve during this period, regaining their roundness (Fig. 16–28).[121] A few femoral heads, however, gradually flatten, primarily in children with disease onset before age 5 years and whose femoral head is totally affected by the disease.

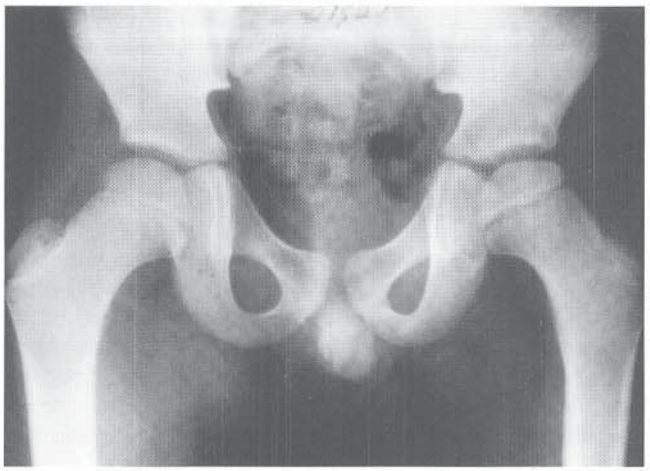
Residual Stage. The fourth radiographic stage is the residual stage, during which there are no additional changes in the density of the femoral head.[282] However, the shape of the head may continue to evolve, and only at the completion of skeletal growth is its permanent contour established (Figs. 16–27E and F).[121] After healing, the shape of the femoral head may vary from completely normal to extremely flat and aspherical. If the disease has disrupted growth of the capital physis, gradual relative overgrowth of the greater trochanter may occur during the residual stage.

TABLE 16–6 **Modified Waldenström Classification of Radiographic Staging of Disease Evolution**

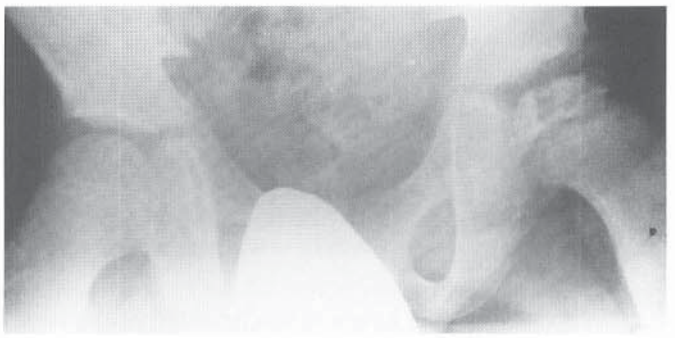
Stage	Radiographic Findings
Initial	Early signs include lateralization of the femoral head and smaller ossific nucleus. Later signs include subchondral fracture, increased density of the femoral head, and metaphyseal lucencies.
Fragmentation	Lucent areas appear in the femoral head; segments (pillars) of femoral head demarcate; increased density resolves; acetabular contour is more irregular.
Reossification (healing)	New bone formation occurs in the femoral head; lucencies are replaced by new (woven) bone.
Residual	Femoral head is fully reossified; gradual remodeling of head shape occurs until skeletal maturity; acetabulum also remodels.

OTHER RADIOGRAPHIC FINDINGS

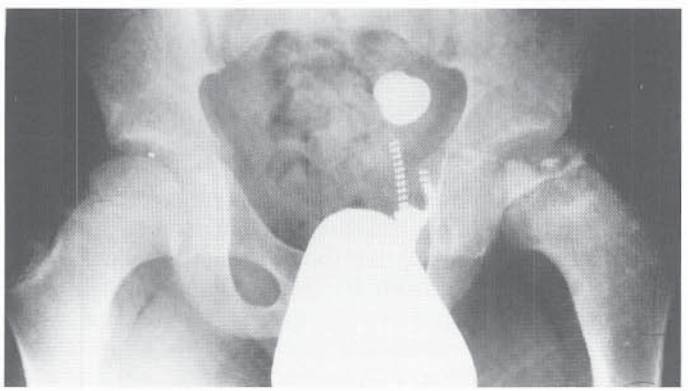
Changes in the Metaphysis. Debate continues as to the nature and importance of radiographic changes observed in the metaphysis proximate to the affected capital epiphysis.



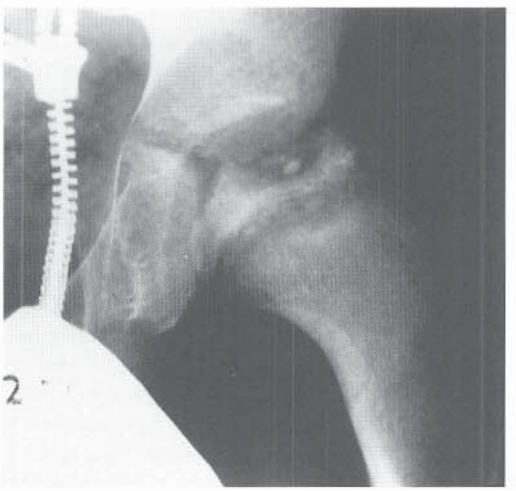
A



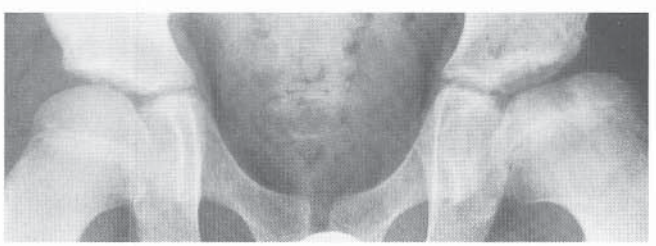
B



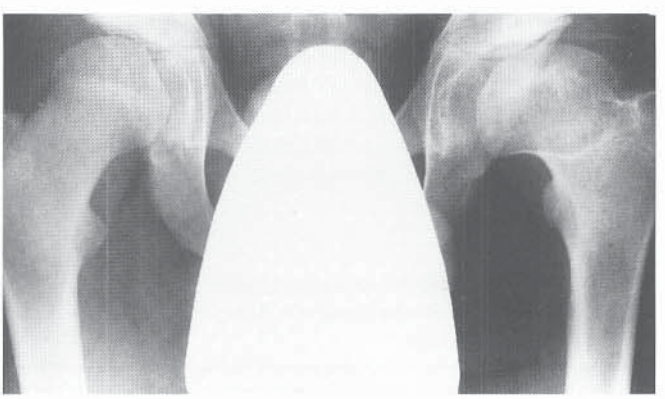
C



D



E



F

FIGURE 16–27 Radiographic evolution of Legg-Calvé-Perthes disease, with onset in a boy at age 10 years 11 months. Despite the late age of onset, the femoral head remodels well as the patient approaches skeletal maturity. **A,** AP radiograph obtained at onset of the disorder shows increased density in the femoral head and apparent widening of the joint space (Waldenström's initial stage). **B,** AP radiograph obtained 9 months after onset shows the head entering the fragmentation stage. The central fragment remains dense and has collapsed relative to the lateral portion (lateral pillar) of the femoral head. The lateral pillar is lucent but has not collapsed, and the hip is classified as type B in the lateral pillar classification system. The joint space has widened further. **C,** AP radiograph obtained 17 months after onset shows early reossification of the femoral head (the healing stage). **D,** A closer view of the femoral head at 22 months after onset of disease. There is still widening of the joint space, and the acetabulum has a bicompartmental appearance. **E,** AP radiograph obtained 4 years after onset. The femoral head is healed and in the residual state. There is still widening of the joint space and incongruity of the head with the acetabulum. **F,** AP radiograph obtained 6 years after onset shows improved roundness of the femoral head and better joint congruity.

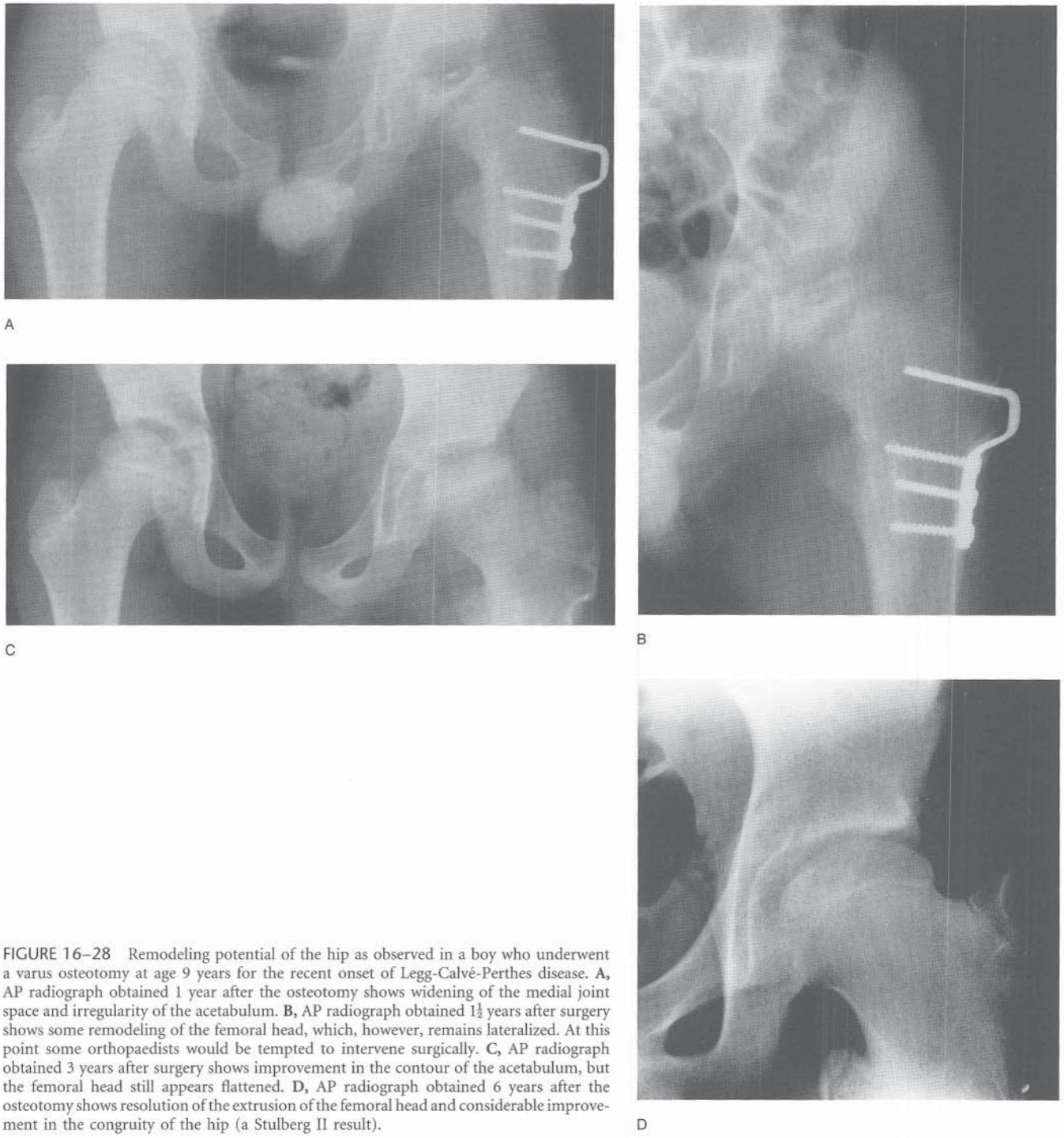
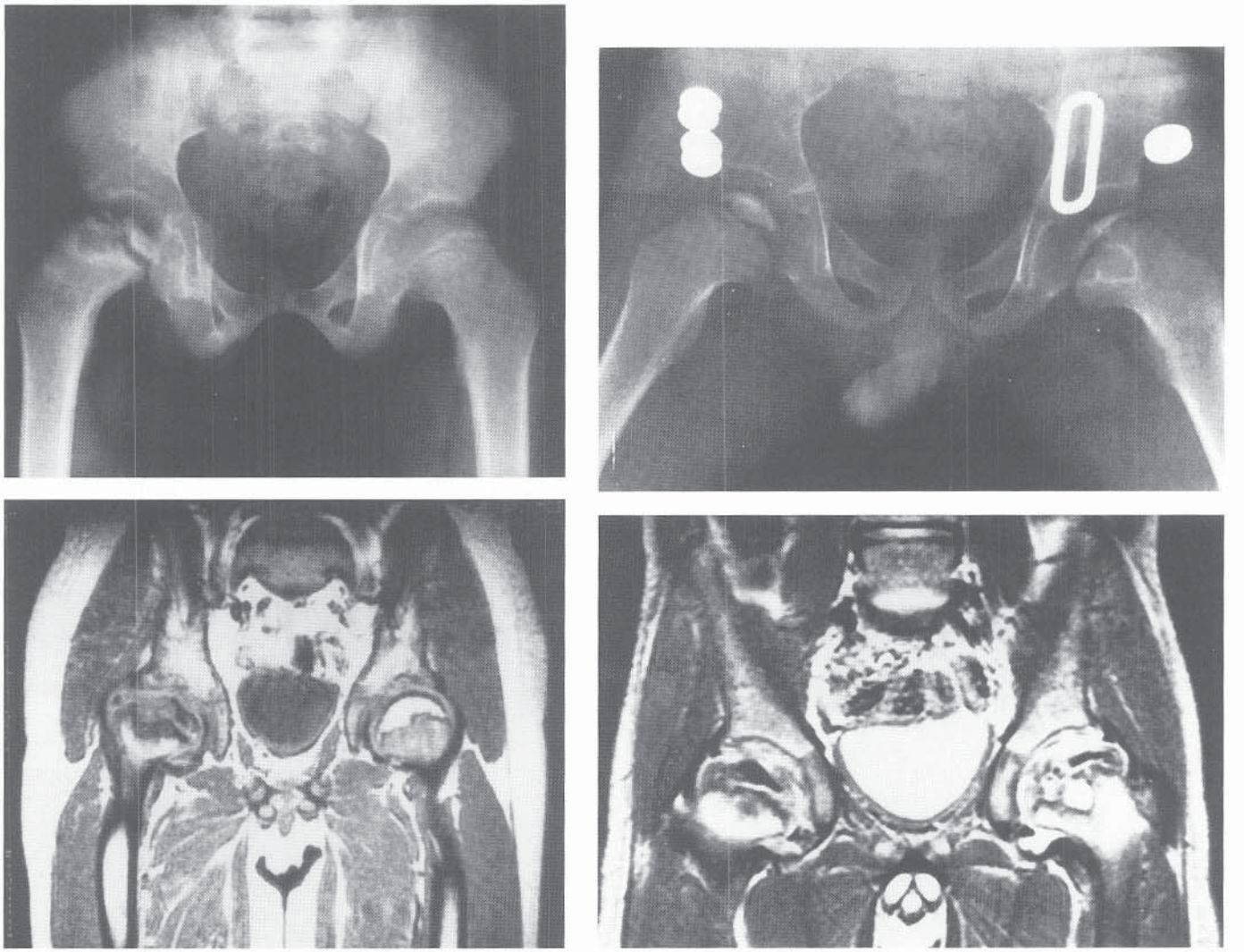


FIGURE 16–28 Remodeling potential of the hip as observed in a boy who underwent a varus osteotomy at age 9 years for the recent onset of Legg-Calvé-Perthes disease. **A,** AP radiograph obtained 1 year after the osteotomy shows widening of the medial joint space and irregularity of the acetabulum. **B,** AP radiograph obtained 1½ years after surgery shows some remodeling of the femoral head, which, however, remains lateralized. At this point some orthopaedists would be tempted to intervene surgically. **C,** AP radiograph obtained 3 years after surgery shows improvement in the contour of the acetabulum, but the femoral head still appears flattened. **D,** AP radiograph obtained 6 years after the osteotomy shows resolution of the extrusion of the femoral head and considerable improvement in the congruity of the hip (a Stulberg II result).

In 1940 Gill reported that changes in the metaphysis were apparent very early in the disease process (the findings were referred to as "holes of decalcification"), and he thought that metaphyseal necrosis caused the changes seen in the femoral head.[81] Ponseti described the presence of cystic changes in the femoral neck, which he believed were caused by tongues of fibrillated cartilage stretching deep into the neck.[220] Katz and Siffert, however, thought the cysts were a result of resorption associated with revascularization as healing occurred.[148] They believed that the metaphyseal changes were of prognostic value, noting that hips with cystic changes were twice as likely to have poor outcomes as hips without cysts.

Studies by Hoffinger and associates corroborate Ponseti's opinion that the lucencies are due to physeal cartilage extending into the metaphysis and challenge the idea that the cysts are located entirely within the metaphysis (Fig. 16–29).[122,123] The investigators compared 24 MRI scans with plain radiographs that indicated the presence of "cysts" in the metaphysis. On MRI, 11 hips had no metaphyseal



A B

FIGURE 16–29 **A,** AP radiograph (*top*) and corresponding MR image (*bottom*) showing extension of physeal cartilage into the metaphysis, with the changes resembling those of a metaphyseal cyst. Most metaphyseal changes are of this nature. **B,** AP radiograph (*top*) and corresponding MR image (*bottom*) showing a true metaphyseal cyst of the left hip. (From Hoffinger SA, Henderson RC, Renner JB, et al: Magnetic resonance evaluation of metaphyseal changes in Legg-Calvé-Perthes disease. J Pediatr Orthop 1993;13[5]:602–606.)

changes, 12 had changes in the anterior portion of the metaphysis where it adjoins the physis, and 1 had a discrete metaphyseal cyst. These findings led to the conclusion that the majority of metaphyseal "cysts" seen on plain radiographs are actually located within the epiphysis or physis and that their appearance in the metaphysis is probably a projection artifact. Others have also concluded that few cysts are actually located strictly within the metaphysis.[1]

The "sagging rope" sign, a radiodense line overlying the proximal femoral metaphysis (Fig. 16–30), is a result of growth plate damage associated with significant metaphyseal response.[4]

Changes in the Physis. Patients with Legg-Calvé-Perthes disease often experience abnormal growth of the proximal femoral physis (Fig. 16–31). However, true epiphyseal bridging is usually not evident, and there are no reliable early indications of growth interference in patients with Legg-Calvé-Perthes disease. A number of radiographic findings associated with abnormal physeal formation have been described, though. Premature physeal closure has been reported in one-fourth of patients with Legg-Calvé-Perthes disease[151]; however, this prevalance is considerably greater than that observed in other studies. Early closure was assumed based on greater trochanteric overgrowth, physeal shape changes, lateral extrusion of the capital nucleus, and medial bowing of the femoral neck.[151]

In patients with evident premature closure of the epiphysis, coexisting overgrowth of the trochanter has not been associated with a Trendelenburg gait.[11] Several deformities related to early femoral neck physeal closure have been described.[26] With central closures, the femoral head was round, the neck was short, and there was greater trochanteric overgrowth. With lateral arrests, the femoral head tilted

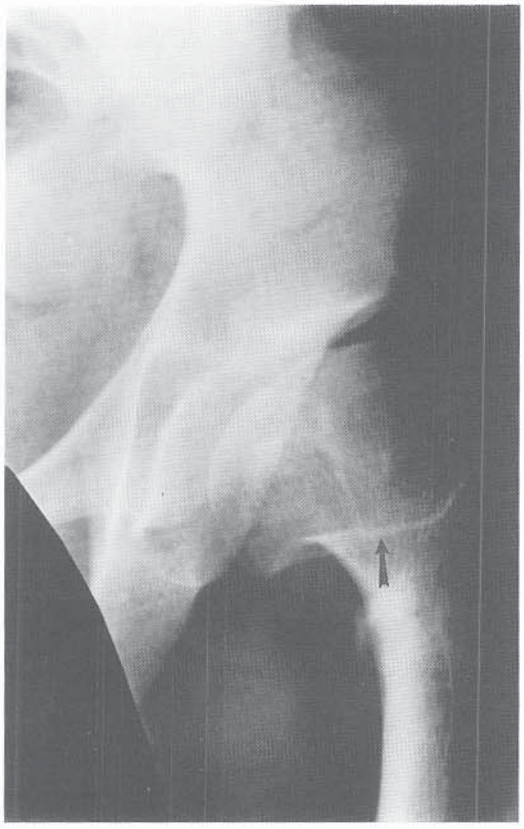


FIGURE 16–30 The "sagging rope" sign, a ropelike, radiodense line across the intertrochanteric area (*arrow*), represents the edge of the enlarged femoral head, or coxa magna.

laterally, the medial neck was longer, and there was overgrowth of the trochanter.

In a study of 52 hips, Sponseller and associates reported that only three had evidence of obvious early physeal closure.[258] Although many of the hips were deformed because of a change in growth velocity, true physeal bar development was not observed. Langenskiöld noted on a lateral radiograph a bulge in the metaphysis (a step-shaped irregularity) that was comparable to changes observed in patients with Blount's disease.[162] This finding was sometimes associated with premature closure of the physis.

Changes in the Acetabulum. In most cases, whenever there are changes in the shape of the femoral head, there will be accompanying acetabular changes.[137] When the femoral head protrudes from the acetabulum, the medial wall may form what looks like a second compartment for the head. Referred to as "bicompartmentalization" by Yngve and Roberts, the authors felt it was an indication of a poor outcome.[293] They observed this sign in 19 of 61 patients, appearing as early as 3 months after the onset of Legg-Calvé-Perthes disease. Bicompartmental changes have also been associated with early closure of the triradiate cartilage.[137] Osteoporosis of the roof of the acetabulum has been noted during the initial phases of the disorder, peaking in the middle phases. The bone returned to normal when the patient reached skeletal maturity. The position of the femoral head, rather than its shape, has also been proposed as the most significant factor in the growth and remodeling of the acetabulum.[143] Bicompartmentalization of the acetabulum usually resolves during the healing stage of Legg-Calvé-Perthes disease.

Limited Radiographic Changes. In 1964 Meyer reported a series of cases in which limited changes of the femoral head were noted on incidental radiographs.[184] The changes were not permanent, and he felt the patients had primary dysplasia of the hip rather than AVN. Although the condition was named *Meyer's dysplasia*, it may be that this entity is asymptomatic AVN in very young children. In a series of 24 cases we found minimal radiographic changes, which were limited to specific segments of the femoral head—ten in the anterior portion, seven in the posteromedial portion, four in the central portion, and three in the lateral portion (Fig. 16–32).[119] These changes developed over time, with all of the femoral heads losing some height compared with the opposite femur. These changes were most likely the result of a minimal form of Legg-Calvé-Perthes disease, with segmental patterns of blood supply to the femoral head dictating which locations were affected. In patients with unilateral disease, small changes have been reported in the opposite hip, such as the femoral head being less round than normal with flattening of the anterior segment.[5] A central notch in the femoral head, which is an expectation of the normal fovea, is often mistaken for avascular change (Fig. 16–33).

Bilateral Changes. Density changes seen in both femoral heads may or may not represent Legg-Calvé-Perthes disease. Four distinct patterns of bilateral hip disease have been reported.[200] In type 1, the radiographic changes and their evolution are exactly the same for both hips. This pattern is most likely indicative of a form of multiple epiphyseal dysplasia, and the physician should examine other joints to verify the diagnosis. In type 2, changes are seen at the same time bilaterally, but fragmentation occurs only in one hip. In type 3, characteristic initial changes are noted in one hip, while radiographs show healed changes in the contralateral hip. In type 4, changes occur sequentially in both hips, with a more severe course seen in the second one. True bilateral Legg-Calvé-Perthes disease appears sequentially in the two hips, as in types 3 and 4.

MAGNETIC RESONANCE IMAGING

MRI is an accurate imaging modality for the early diagnosis of Legg-Calvé-Perthes disease and for visualizing the configuration of the femoral head and acetabulum (Figs. 16–34 and 16–35). However, to date, MRI has not resulted in definite clinical advantages.

Early on, there was some equivocation regarding the efficacy of MRI. Elsig and associates reported a case in which technetium scanning showed early AVN when the MRI study was normal.[63] Ranner and associates found MRI to be as effective as bone scintigraphy for early diagnosis, but thought that bone scintigraphy was a better follow-up modality for showing the start of revascularization.[229] However, other studies and reports have clearly shown the effectiveness of MRI in the diagnosis and evaluation of patients with Legg-Calvé-Perthes disease.

In 1984 MRI was being used to create arthrogram-like images to aid in the diagnosis of the disorder.[251] Using MRI, Grimm and associates evaluated the congruity of articular surfaces, femoral head containment, joint effusion, and sy-

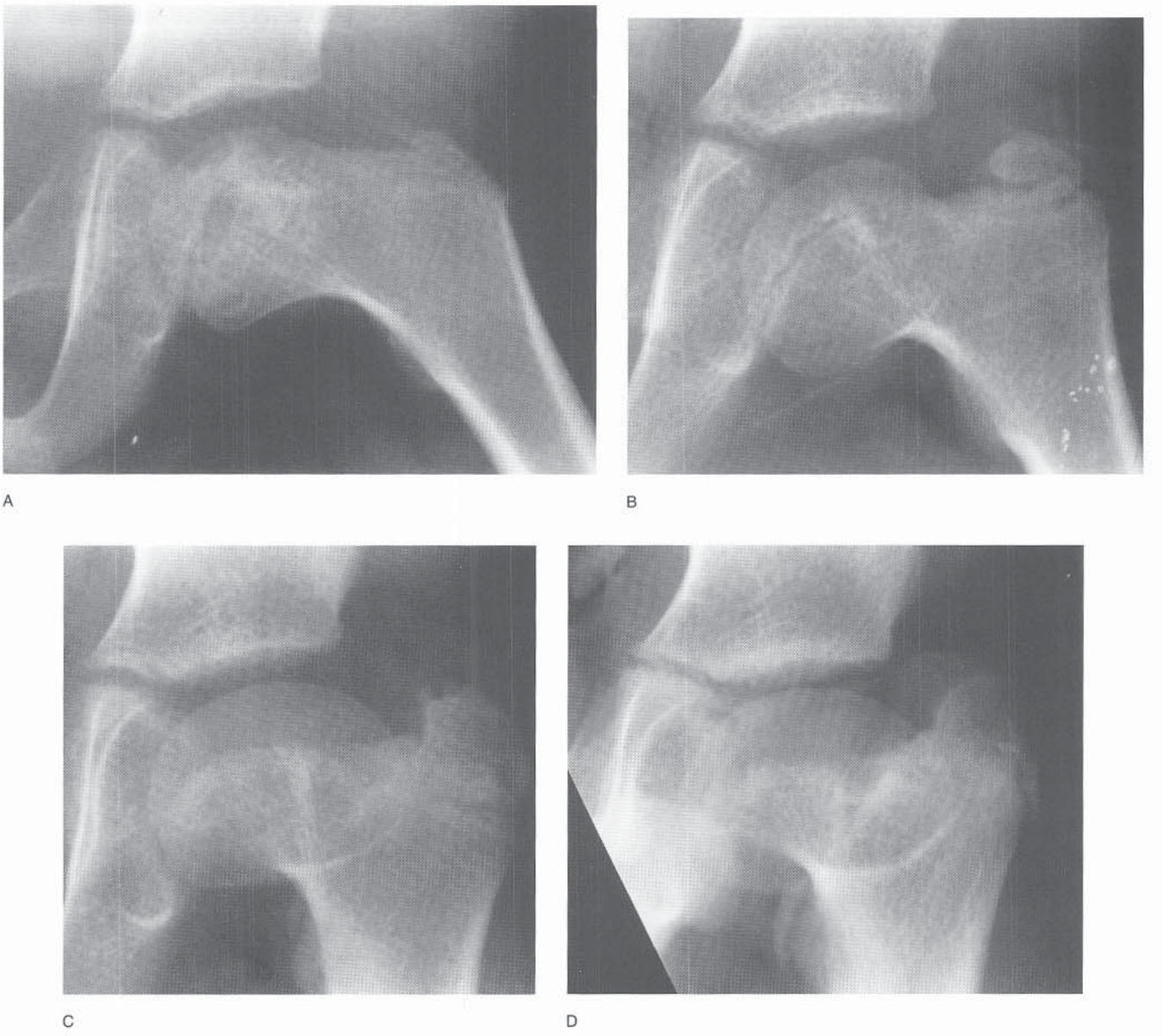


FIGURE 16–31 Growth arrest in patient with Legg-Calvé-Perthes disease. **A,** AP radiograph of a 6-year-old boy with mild involvement of the left hip. There is a dense area laterally in the metaphysis and epiphysis. **B,** AP radiograph of the boy at age 8 years shows an apparent bridge across the physis and growth disturbance of the femoral neck. **C,** AP radiograph at 10 years of age shows further loss of growth of the upper femoral neck, with a valgus tilt of the femoral head. **D,** AP radiograph at 13 years of age shows trochanteric overgrowth and a short femoral neck, with a valgus tilt to the femoral head.

novial hypertrophy.[95] Lateralization of the femoral head was shown to be secondary to medial hypertrophy of the head's cartilage. The authors were also able to determine the extent of revascularization with MRI. MRI can also provide better delineation of affected sites and the degree of involvement during the early phases of the disorder.[117] Epiphyseal involvement is more clearly visualized on MRI 3 to 8 months after the first symptoms.[160]

MRI has also been found to be more accurate in the early diagnosis than other imaging modalities. Theissen and associates reported a diagnostic accuracy of 97 to 99 percent for MRI, compared with 88 to 93 percent for radiography and 88 to 91 percent for scintigraphy.[272] In a study comparing MRI with arthrography, Egund found that MRI showed more detail of the medial and lateral aspects of the cartilaginous capital epiphysis.[62] In two cases the early diagnosis of Legg-Calvé-Perthes disease was rendered on the basis of MRI findings when radiographs and bone scans were normal.[219] MRI has also been reported to provide earlier and more reliable information about the true extent of femoral head necrosis than radiography or scintigraphy.[145,204] Revascularization was also more clearly visualized than on technetium scanning. Subtraction MRI correlates well with bone scintigraphy in delineating ischemia of the femoral head early in the disease process.[252] The affected area appears as a widespread absence of enhancement. Subtraction MRI also allows recognition of early reperfusion patterns.

Sequential MRI studies of Legg-Calvé-Perthes disease

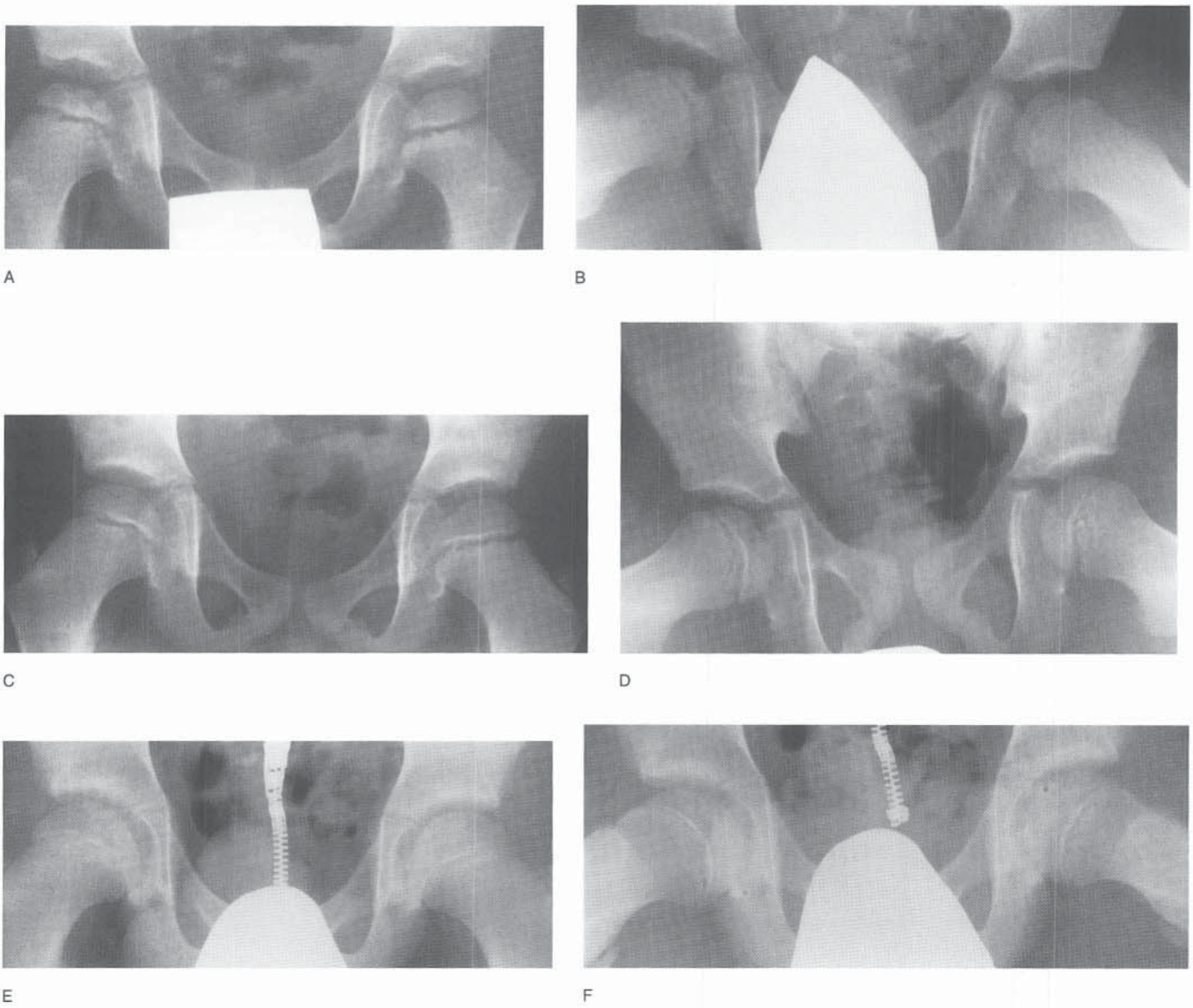


FIGURE 16–32 Minimal femoral head involvement in a 4-year, 9-month-old boy with Legg-Calvé-Perthes disease. **A,** AP radiograph obtained at onset of the disease. The right femoral head has a lucent area centrally and is smaller than the contralateral femoral head. **B,** Frog-leg lateral radiograph obtained at onset of the disease shows central lucency in the right femoral head. **C,** AP radiograph obtained 3 years after onset shows partial resolution of the radiolucency. **D,** Frog-leg lateral radiograph obtained 3 years after onset shows a central depression of the ossific nucleus. **E,** AP radiograph obtained 9 years after onset shows complete healing of the femoral head. The height of the head is reduced compared with the contralateral hip. **F,** Frog-leg lateral radiograph obtained 9 years after onset shows a round femoral head (a Stulberg II result). (From Herring JA: Legg-Calvé-Perthes Disease. Rosemont, IL, American Academy of Orthopaedic Surgeons, 1996.)

have established a correlation with the Catterall classification system.[23] Images showed substantial amounts of viable bone medially, laterally, and posteriorly in group II hips, considerable necrosis in group III hips, and growth plate involvement with repair extending into the metaphysis in group IV hips.

SCINTIGRAPHY

Technetium scanning is an effective means of diagnosing Legg-Calvé-Perthes disease in its early stages, before associated radiographic findings are apparent (Fig. 16–36).[266] Scintigraphy has also been used to classify the severity of the disease, with grade I representing one-fourth epiphyseal involvement and grade IV complete involvement.[69] Others have reported that bone scintigraphy provides more accurate information about the extent of the necrosis than initial radiography, and that the scans are able to reveal revascularization and consequently the stage of the disease.[13,15,22,40,70,145,274] Care must be taken, however, as early bone scans may at times suggest a more severe condition than really exists (Fig. 16–37).

Scintigraphy has also been used to classify revascularization as either recanalization or neovascularization.[47,274] The investigators also noted that a hip could change from an "A" track to a "B" track in a few cases. They defined an A pathway as an indication of uncomplicated revascularization

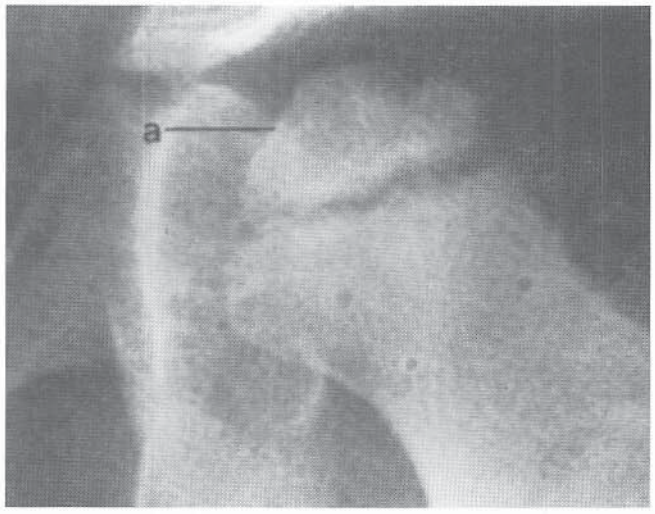


FIGURE 16–33 A central notch, or "dimple effect," on the lateral side of the dome of the femoral capital epiphysis in the "normal hip," opposite a hip with Legg-Calvé-Perthes disease. The letter *a* denotes the normal medial flattening of the epiphysis at the insertion of the ligamentum teres. (From Harrison MHM, Blakemore ME: A study of the "normal" hip in children with unilateral Perthes' disease. J Bone Joint Surg 1980;62-B:31.)

of the femoral head and a B pathway as representing a slower rate of revascularization and healing (with healing possibly occurring through neovascularization rather than recanalization). Patients on pathway A had an average Mose classification of 1.2, an average Catterall score (an averaging of several patients) of 2.4, and a favorable prognosis. Those on pathway B had an average Mose classification of 5.2 and an average Catterall score of 3.5, with 18 of 20 hips showing head-at-risk signs (particularly lateral extrusion). This group had a poorer prognosis, often requiring surgical intervention. Within the study population, the healing pattern of seven patients changed from the A to B pathway, a finding that makes early prognostication difficult.[274]

ARTHROGRAPHY

Arthrography shows well the configuration of the femoral head and its relationship to the acetabulum. An increase in the medial joint space, with concomitant lateralization of the femoral head, has been frequently observed on plain radiographs.[9] Arthrographic studies have shown that the apparent widening in the joint space is due to thickening of the articular cartilage.[79] Changes in the shape of the femoral head also result in lateral shifting of the femoral head. Arthrography can also provide reliable information regarding containment of the femoral head within the acetabulum.[50] The major advantage of arthrography is that the examiner can assess the congruity of the hip in many different positions.

Despite the positive aspects of arthrography, its use in the diagnosis and evaluation of Legg-Calvé-Perthes disease is controversial. Gallagher and associates did not believe that the routine use of arthrography was necessary for managing the disease.[76] They reported that plain radiography yielded adequate information to evaluate femoral head protrusion (particularly if the acetabulum-head quotient was used).

Currently, arthrography is most often used in the early diagnosis of hinge abduction of the hip, in which the femoral head "hinges" out of the acetabulum when the hip is abducted (Fig. 16–38).[232] Hinge abduction sometimes occurs early in the course of Legg-Calvé-Perthes disease, and the longer it remains untreated, the worse is the final outcome. Traction is often used initially to relieve hinging, and surgery is performed later to contain the head.

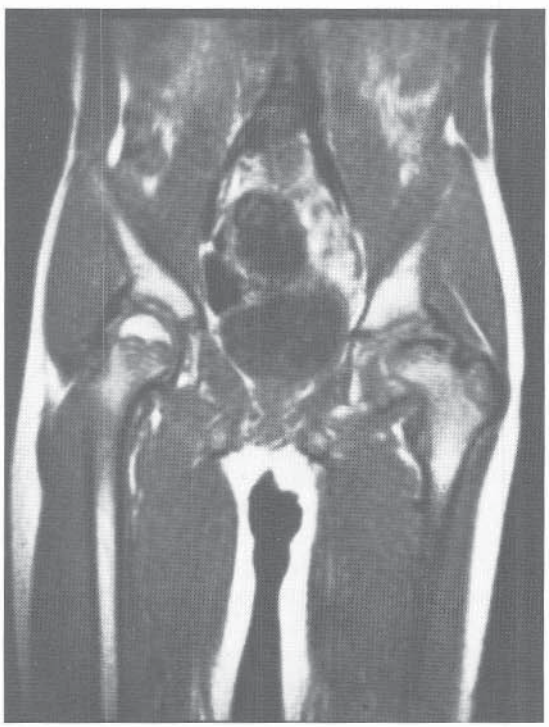


FIGURE 16–34 MRI appearance in Legg-Calvé-Perthes disease. The distorted shape of the cartilage model of the femoral head is evident.

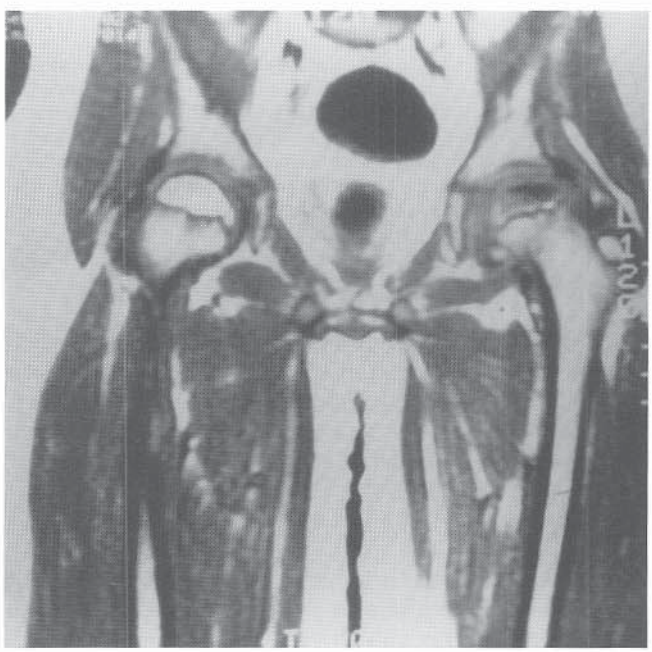


FIGURE 16–35 MRI of a 9-year-old boy with Legg-Calvé-Perthes disease showing extensive involvement of the left hip.

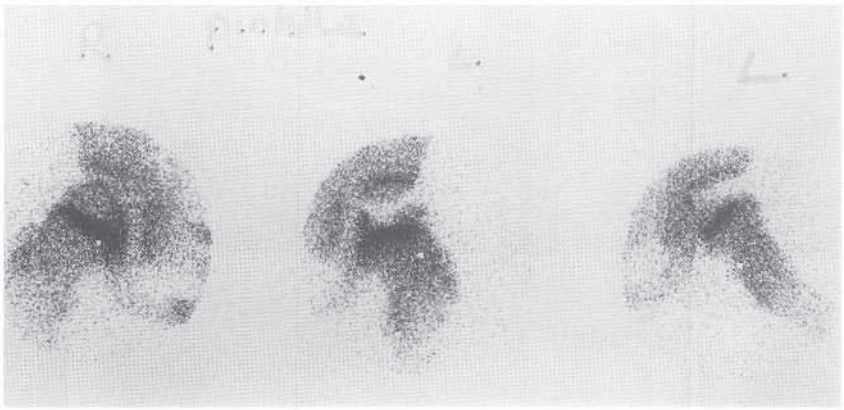


FIGURE 16–36 A technetium scan with pinhole collimation. The right hip (*left*) is normal. The AP view of the left hip (*middle*) shows lack of uptake in the lateral two-thirds of the femoral head, and the frog-leg lateral view (*right*) shows lack of uptake anteriorly.

ULTRASONOGRAPHY

Ultrasonography is sometimes used in the early stages of Legg-Calvé-Perthes disease to demonstrate joint effusion and in later stages to assess the shape of the femoral head. Ultrasound can provide a good profile of the cartilaginous femoral head (comparable to that of arthrography) and allows subsequent observation of deformation of the head without the need for radiographs.[267] A four-stage classification system based on ultrasound and sonoanatomic criteria has been suggested for patients with Legg-Calvé-Perthes disease.[196]

COMPUTED TOMOGRAPHY

CT is able to provide accurate three-dimensional images of the shape of the femoral head and acetabulum.[165,189,287] A classification system has been developed based on CT findings.[189] In group A, only the periphery of the femoral head is affected; in group B, there is considerable necrosis of the central portion of the head but no posterior involvement; and in group C, the entire head is affected.

Despite these reports, CT is not typically used on a routine basis to evaluate patients with Legg-Calvé-Perthes disease, and clinical decisions are usually not made on the basis of early CT findings. CT may be of benefit, however, in the later stages of the disease to evaluate pain, locking of the joint, and other mechanical symptoms. CT of the hip may help the clinician distinguish the cause as an area of incomplete reossification within the femoral head or as a true osteochondritic lesion.

Classification Systems Based on Radiographic Findings

Because the clinical course and final results vary considerably among patients with Legg-Calvé-Perthes disease, investigators have attempted to predict the severity of the disorder based on early radiographic findings. One of the earliest descriptions was that by Legg, who identified two types of femoral heads—a "cap" and a "mushroom."[168] The latter was less common but associated with more severe disease. Waldenström classified the heads into three categories.[279] Type 1 and 2 hips correlated with good results. The type 3 hip was associated with a poor prognosis, with the final result being a conical head and acetabulum that limited the patient's range of motion to flexion and extension. Goff also identified three types of femoral heads—spherical, cap, and irregular—that correlated with disease outcome.[90]

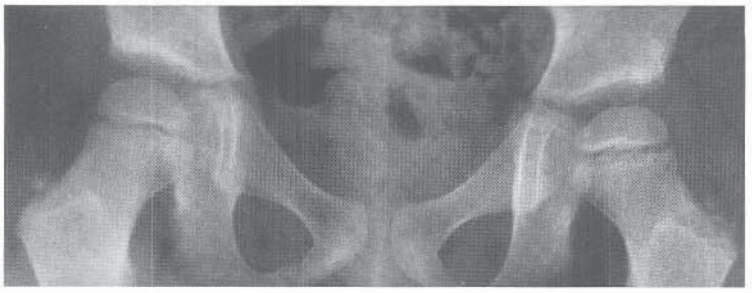
THE CATTERALL CLASSIFICATION

The Catterall classification system, introduced in 1971, represented a milestone in the treatment of patients with Legg-Calvé-Perthes disease.[38] At that time, most patients underwent very rigorous therapeutic programs, regardless of the severity of the disease. Catterall used radiographic findings to identify four groups of patients in which treatment decisions could be individualized based on radiographic appearances.

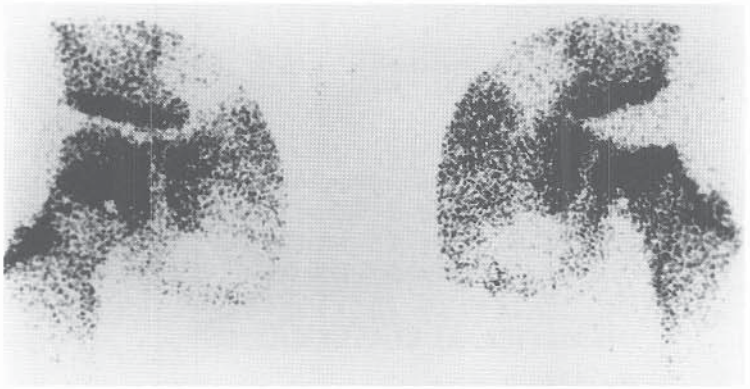
In Catterall group I (Fig. 16–39A), only the anterior portion of the epiphysis is affected. In group II (Fig. 16–39B), more of the anterior segment is involved and central sequestrum is present. Although the affected segment may collapse, epiphyseal height is preserved. According to Catterall, patients in groups I and II had benign prognoses and did not need medical intervention. In group III (Fig. 16–39C), most of the epiphysis is "sequestrated" (Catterall's term), with the unaffected portions located medial and lateral to the central segment. In group IV (Fig. 16–39D), all of the epiphysis is sequestrated. Patients in groups III and IV required treatment.

Catterall also described four "head-at-risk" factors that he believed could be used to predict patient prognosis.[38] The risk factors were lateral subluxation of the femoral head, a radiolucent V in the lateral aspect of the epiphysis (Gage's sign), calcification lateral to the epiphysis, and a horizontal physeal line. The presence of these signs increased the chances of a poor outcome.

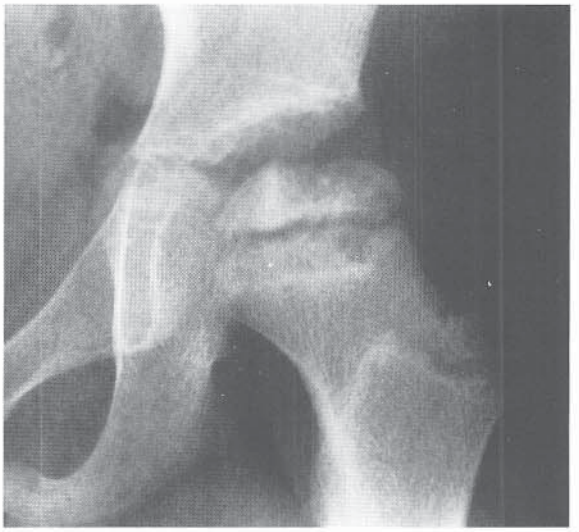
Catterall's groupings are difficult to define, and several interobserver studies have shown a low degree of reproducibility in differentiating the groups by means of test radiographs.[42,105] Interpreting the head-at-risk signs has also proved difficult.[105] In addition, it has been shown that the classification changed in only 6 percent of cases if applied during the fragmentation stage, compared with 40 percent when applied earlier in the disease process.[276] This finding, though, can be accounted for by the fact that Catterall developed his classification system based on radiographs made during the fragmentation stage.



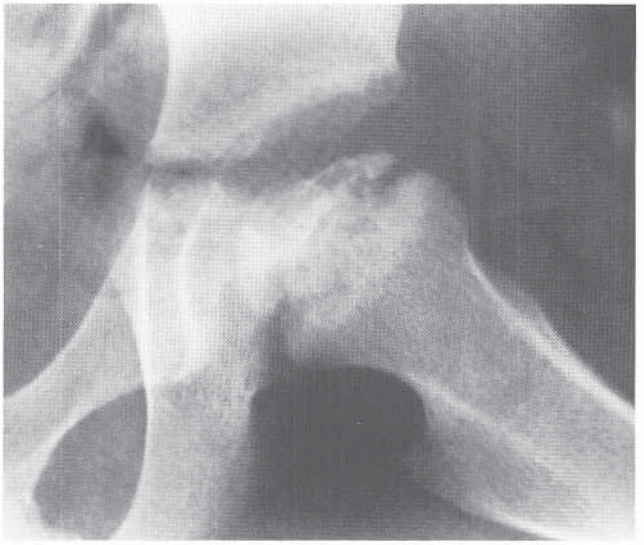
A



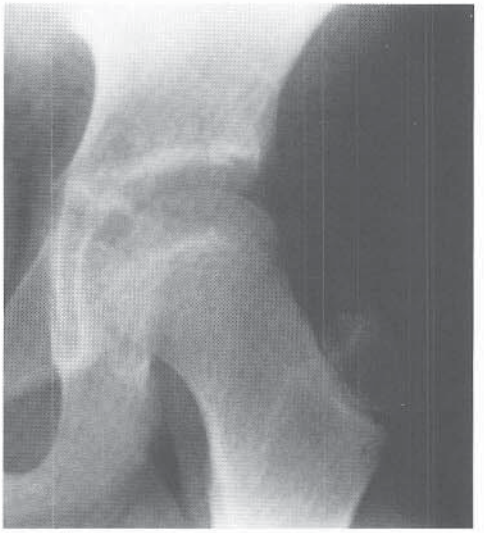
B



C



D



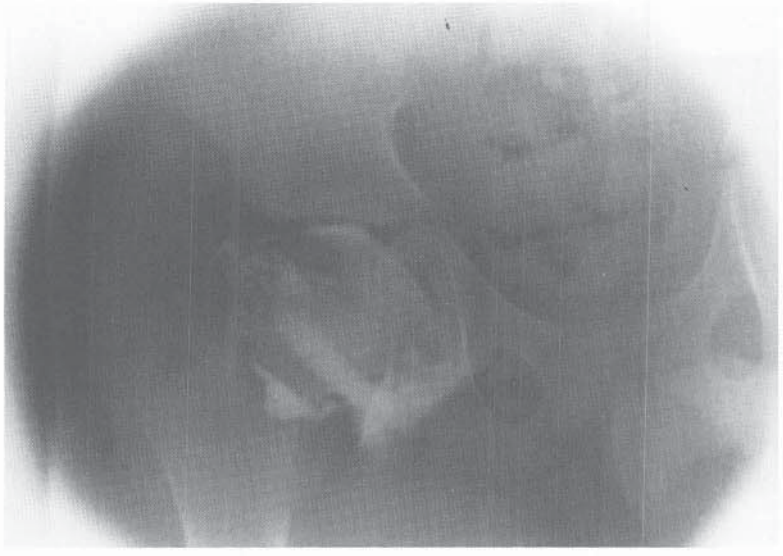
E

FIGURE 16–37 Bone scans may be misleading. **A,** AP radiograph of a 6-year-old boy with a limp of 1 month's duration. The left femoral head is slightly smaller than the right head. **B,** A technetium scan made at the same time shows lack of uptake throughout most of the left femoral head. **C,** AP radiograph obtained 15 months later shows central lucency in the femoral head and a diffuse metaphyseal reaction. **D,** Frog-leg lateral radiograph obtained at the same time shows anterior involvement of the femoral head. This is classified as a lateral pillar B hip. The patient was able to maintain excellent range of motion and was not treated. **E,** AP radiograph obtained 5 years after onset of disease shows a round femoral head (a Stulberg II result).

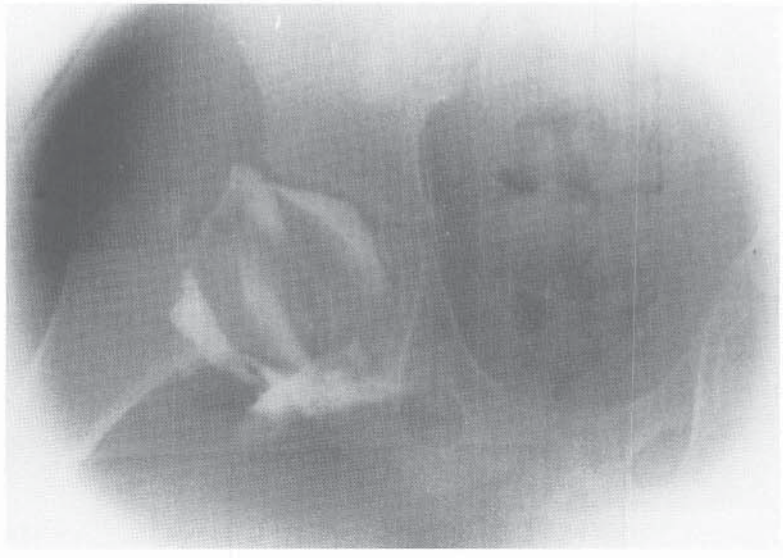
Despite these criticisms, Catterall's work was instrumental in furthering the search for more reliable prognostic indicators and underscored the fact that many patients with Legg-Calvé-Perthes disease are able to achieve full recovery even if untreated.

THE LATERAL PILLAR CLASSIFICATION

The lateral pillar classification system is based on radiographic changes in the lateral portion of the femoral head when it enters the fragmentation stage as seen on the antero-



A



B

FIGURE 16–38 Arthrograms illustrating hinge abduction. **A,** In adduction the joint is congruous, with flattening of the femoral head and matching flattening of the acetabulum. **B,** In abduction the joint "hinges" open and the femoral head does not roll into the acetabulum. Instead, the joint space widens and is filled with contrast medium.

posterior (AP) view.[120] At the beginning of fragmentation, there is frequent separation between the central, medial, and lateral segments ("pillars") of the femoral head. When the lateral pillar remains intact, it acts as a weightbearing support to protect the central avascular segment.[72]

The classification consists of three groups—A, B, and C (Fig. 16–40, Table 16–7). In group A (Fig. 16–41) there is minimal density change in the lateral pillar and no loss of height. In group B (Fig. 16–42), lucency is observed in the lateral segment, and there is subsequent height loss up to, but not exceeding, 50 percent of the original height of that segment of the epiphysis. Collapse of the central fragment beneath the level of the lateral segment is often an early manifestation of this group. As the fragmentation stage evolves, the lateral pillar may lose height and protrude laterally, but the segment does not collapse beyond half its original height in group B. In group C (Fig. 16–43), early lucency is noted in the lateral pillar, there is minimal or no separation between the lateral and central segments, and the lateral pillar collapses to less than half its original height. The lateral pillar is frequently lower in height than the central pillar early in the fragmentation stage.

A strong correlation exists between the lateral pillar classification and subsequent outcome, with group A hips faring the best, group B having an intermediate outcome, and group C the worst.[120] A mechanical basis may account for the good results seen when the lateral pillar is maintained.[227] If only the central part of the femoral head is necrotic, the remaining lateral rim of bone stress shields the central core and prevents collapse. However, when necrosis is widespread, this shielding effect is lost and the femoral head collapses.

Compared with the Catterall classification system, the lateral pillar classification system has been reported to have

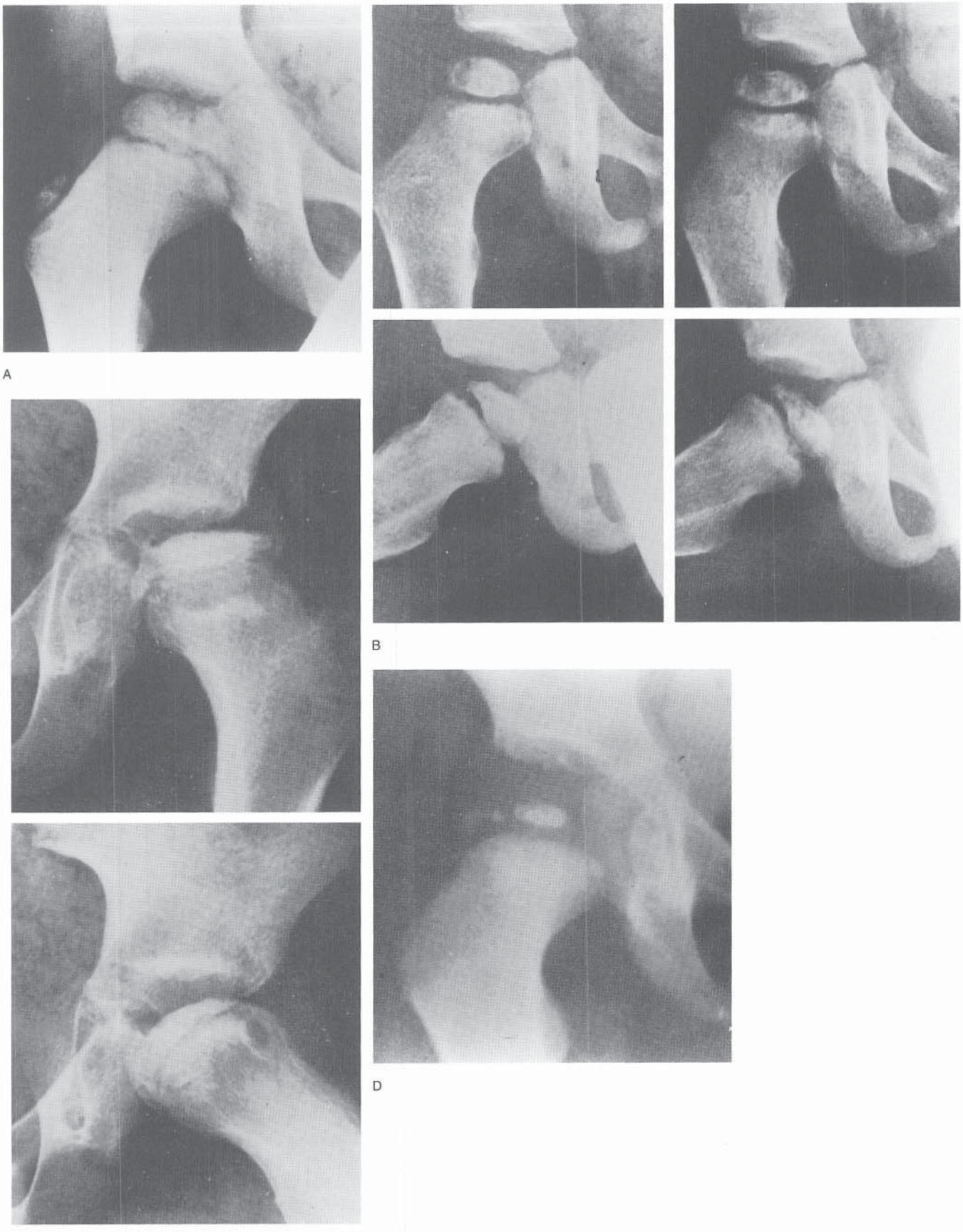


FIGURE 16–39 Catterall's classification. **A,** Group I. **B,** Group II. **C,** Group III. **D,** Group IV. (From Catterall A: The natural history of Perthes' disease. J Bone Joint Surg 1971;53-B:37.)

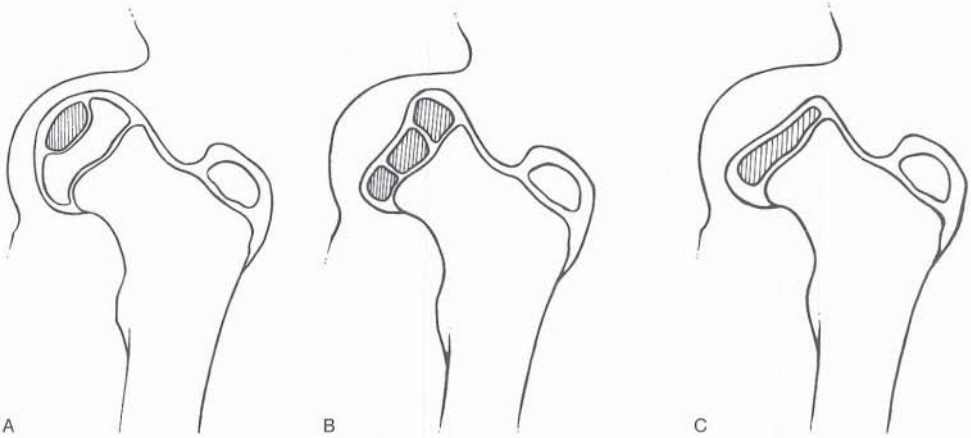


FIGURE 16–40 Lateral pillar classification. **A,** Group A: no loss of height in the lateral pillar. **B,** Group B: partial collapse (less than 50 percent) of the lateral pillar. **C,** Group C: more than 50 percent collapse of the lateral pillar.

greater interobserver reliability and to better predict final outcome.[236] Other investigators have noted that the lateral pillar system is relatively easy to apply and interpret (requiring only an AP radiograph of the hip during the fragmentation stage), reliable, highly reproducible, and useful for formulating a valid long-term prognosis in Legg-Calvé-Perthes disease.[68,133,268]

CLASSIFICATION OF END RESULTS

Along with classifications to help determine the severity of disease, several systems have been developed to address final outcomes. Because the femoral head continually changes during a child's growth, these classifications are best used to evaluate the skeletally mature patient.

The Mose Classification. The Mose classification system is based on fitting the contour of the healed femoral head to a template of concentric circles.[190] In good outcomes, the shape of the femoral head deviates no more than 1 mm from a given circle on both AP and frog-leg lateral radiographs. If the shape falls within 2 mm, it is considered a fair outcome. If the deviation is greater than 2 mm, it is a poor outcome. Although this system is easily reproducible, it is very limited and does not cover the myriad possible outcomes.

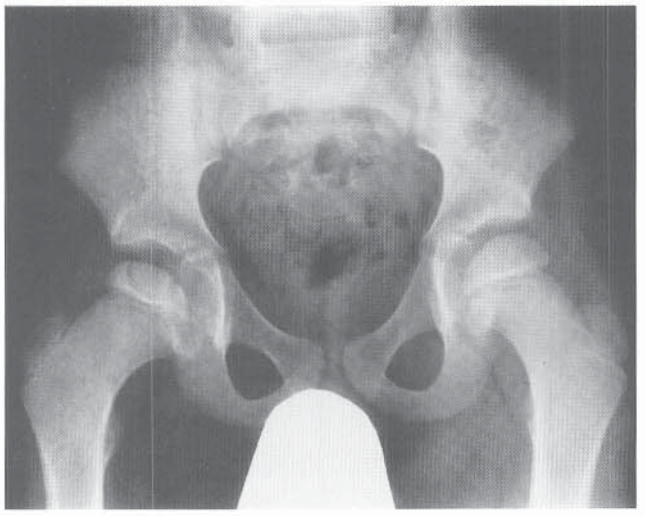
The Stulberg Classification. The Stulberg classification is a reproducible system in which hips are separated into five groups (Table 16–8).[263] The shape of the femoral head is basically normal in group I hips (see Fig. 16–41C). Loss of head height is seen in group II hips, but the head's contour conforms to a concentric circle within 2 mm on the AP and frog-leg lateral radiographs (see Fig. 16–42D). The femoral head is more elliptical in group III hips, deviating from a circle by more than 2 mm (Fig. 16–44). In group IV hips, the femoral head is flattened; however, the degree of flattening required to qualify for this group is not specified. We have used the group IV classification when there is a flattened area greater than 1 cm in length (see Fig. 16–43E). Hip range of motion changes to match the cylindrical shape of the femoral head, resulting in almost normal range of flexion and extension but a minimal arc of rotation (Fig. 16–45). Often the hip rotates externally whenever the joint is flexed and resumes a neutral position with extension. This motion has been likened to the motion of a cow's hip, which is flattened and more cylindrical than round. In groups III and IV, the contour of the acetabulum matches that of the femoral head (referred to as "congruous incongruity"). In group V hips (Fig. 16–46), there is collapse of the femoral head but the acetabular contour does not change (referred to as "incongruous incongruity"). These hips are similar to those seen in adult AVN in which there is collapse of the central portion of the head without acetabular change. This classification system has been found effective in predicting subsequent arthritic changes. Patients with group II hips have a good long-term prognosis, those with group III or IV hips develop mild to moderate degenerative changes in late adulthood, and those with group V hips develop painful arthritis in early adulthood.

TABLE 16–7 **The Lateral Pillar Classification System**

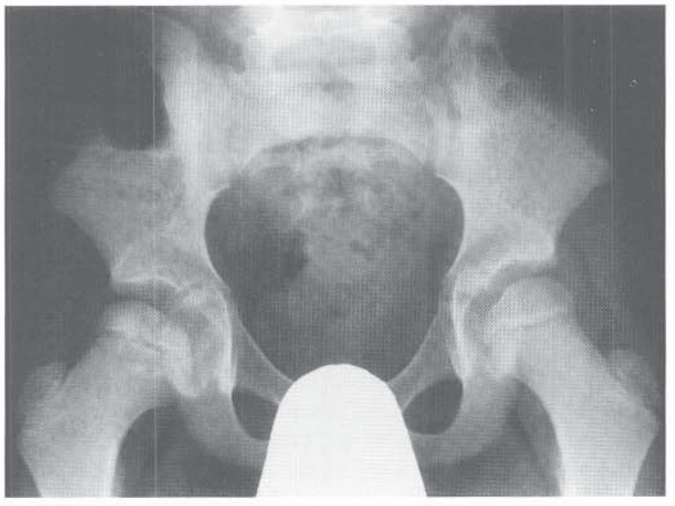
Group	Radiographic Findings (Anteroposterior Radiographs)
A	Minimal density changes in lateral pillar and no loss of height.
B	Lucency observed in the lateral segment, and there is subsequent height loss up to but not exceeding 50% of the original height of that segment of the epiphysis. Collapse of central fragment beneath level of lateral segment is often an early manifestation of this group. As the fragmentation stage evolves, the lateral segment does not collapse beyond half its original height.
C	Early lucency noted in lateral pillar. Lateral pillar frequently is lower in height than central pillar early in the fragmentation stage. There is minimal or no separation between lateral and central segments, and the lateral pillar collapses to less than half its original height.

PROGNOSTIC RISK FACTORS

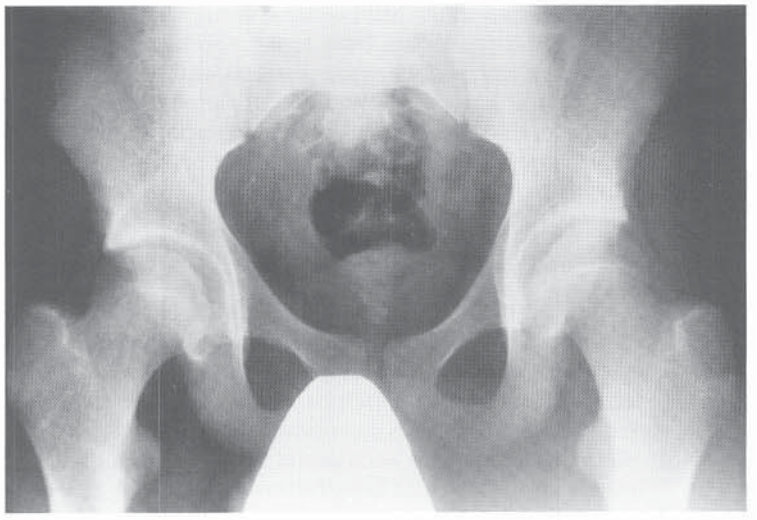
A number of different risk factors have been studied to determine their prognostic significance (Table 16–9), with agreement on some and disagreement on others. Although some authors have reported that the only factor associated with a poor prognosis is lateralization of the femoral head,[193] others have described a variety of risk factors that have an impact on outcome. These factors include the extent of uncovering of the femoral head, the Catterall classification, lateral calcification, lateral head displacement (using the head-to-teardrop distance), widening of the femoral head before fragmentation, the Saturn phenomenon (a sclerotic



A



B



C

FIGURE 16–41 Radiographic appearance of lateral pillar group A. **A,** AP radiograph showing demarcation between the lateral pillar and the central dense fragment of the ossific nucleus. There is no change in the density of the lateral pillar, and no loss of height. **B,** AP radiograph obtained 9 months later shows reossification and no loss of lateral pillar height. **C,** AP radiograph obtained 5 years later shows a hip that cannot be distinguished from normal (a Stulberg I outcome).

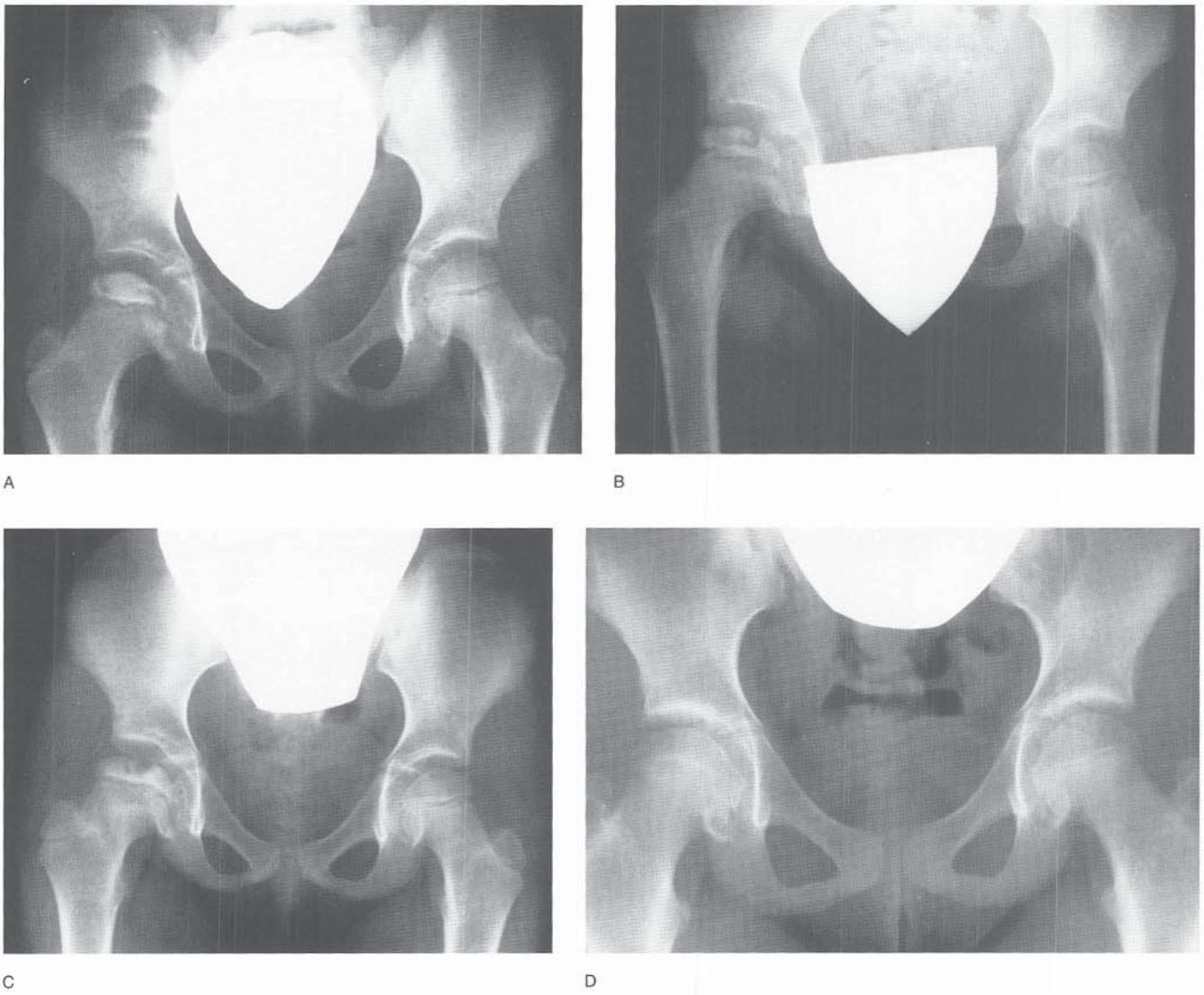


FIGURE 16–42 Radiographic appearance of lateral pillar group B. **A,** AP radiograph showing early demarcation between the lateral and central segments of the femoral head. There is some loss of height of the lateral pillar. **B,** Well into the fragmentation stage, the lateral pillar has not collapsed any further. **C,** Reossification stage. The shape of the ossific nucleus is partially restored. **D,** The femoral head has now healed and is more round in shape (a Stulberg II result).

epiphysis surrounded by a ring of lucency) (Fig. 16–47), and widening of the femoral neck in the early stages of the disorder; all are associated with a poor prognosis.[56,223]

Differential Diagnosis

Certain conditions share features with Legg-Calvé-Perthes disease (Table 16–10). AVN can be caused by a variety of disorders, including hemoglobinopathies (e.g., sickle cell disease, thalassemia),[61,203,228] leukemia, lymphoma, idiopathic thrombocytopenic purpura, and hemophilia.[2,213,233,275] A thorough history and physical examination can exclude these conditions in most patients. Legg-Calvé-Perthes disease is rarely seen in black children, and appropriate studies should be ordered to rule out hemoglobinopathies in this patient population. Patients with hypothyroidism may have radiographic changes that resemble those of Legg-Calvé-Perthes disease[239] (Fig. 16–48); however, the changes are bilaterally symmetric. Patients with hypothyroidism may develop progressive ossification areas in the femoral head known as pseudo-fragmentations, which can deform the head and produce coxa plana.[54]

If there is a strong family history of hip abnormalities or if bilateral disease is present, multiple epiphyseal dysplasia (Fig. 16–49), spondyloepiphyseal dysplasia, and Morquio's disease should be ruled out.[178] These conditions cause symmetric flattening, fragmentation, and uniform mild sclerosis of the femoral head, but no metaphyseal changes.[3] In contrast, Legg-Calvé-Perthes disease is associated with asymmetric involvement of the head with areas of increased density, and the metaphysis is affected.[51,178] Children with epiphyseal dysplasia are short in stature and have abnormalities of other epiphyses, particularly flattening of the distal femoral epiphysis. In epiphyseal dysplasias, both hips are affected at the same time. When there is bilateral involvement in Legg-Calvé-Perthes disease, the disorder normally

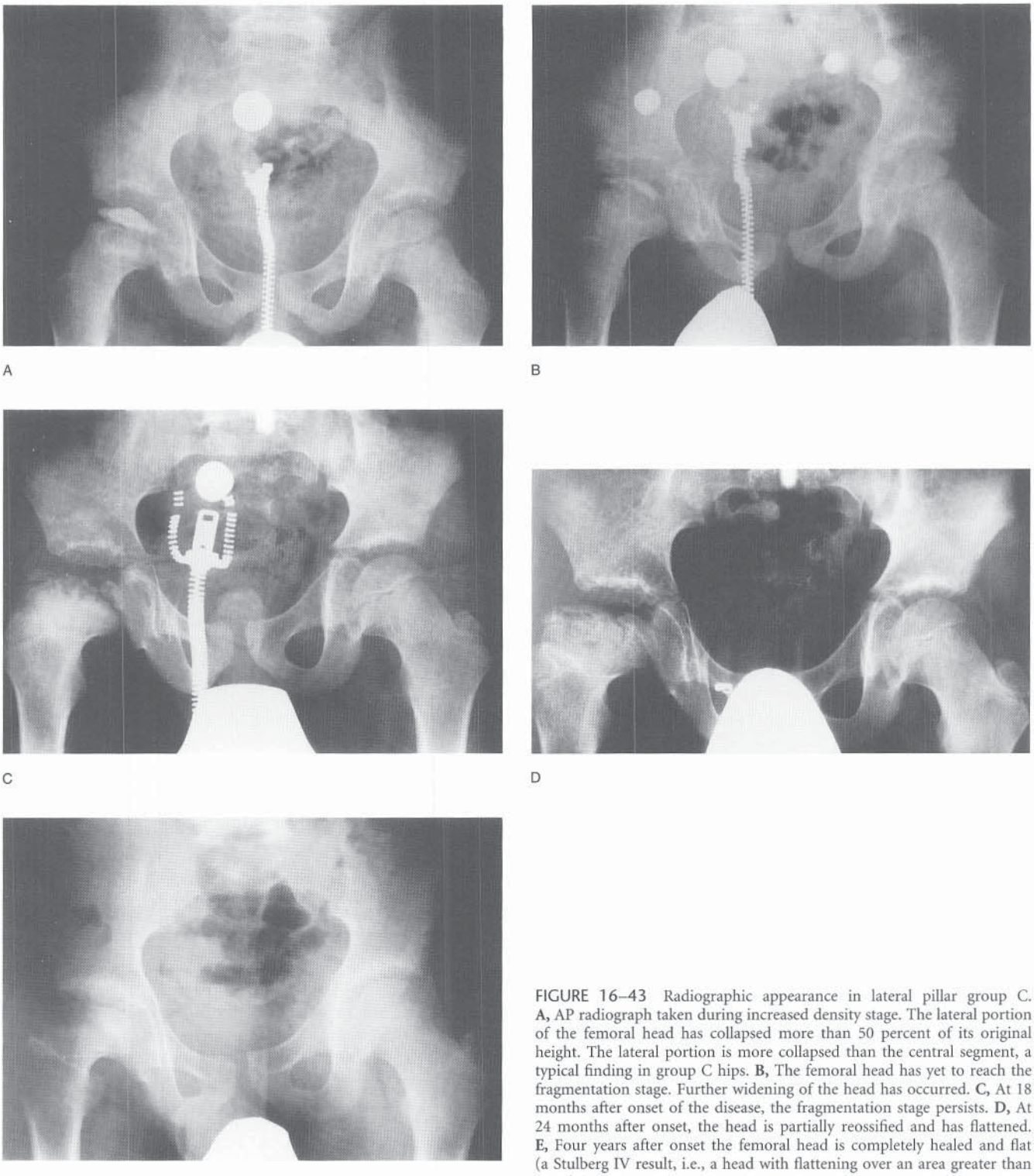


FIGURE 16–43 Radiographic appearance in lateral pillar group C. **A,** AP radiograph taken during increased density stage. The lateral portion of the femoral head has collapsed more than 50 percent of its original height. The lateral portion is more collapsed than the central segment, a typical finding in group C hips. **B,** The femoral head has yet to reach the fragmentation stage. Further widening of the head has occurred. **C,** At 18 months after onset of the disease, the fragmentation stage persists. **D,** At 24 months after onset, the head is partially reossified and has flattened. **E,** Four years after onset the femoral head is completely healed and flat (a Stulberg IV result, i.e., a head with flattening over an area greater than 1 cm).

develops sequentially, with one hip affected a year or more before the other.

AVN may complicate the changes of epiphyseal dysplasia.[127] Mandell and associates described 10 patients with multiple epiphyseal dysplasia who also had AVN changes of the femoral head, including sclerosis and subchondral fissuring superimposed on ossification centers that were already irregular.[177] The avascular features of these changes were corroborated by MRI and scintigraphy.

In younger children, AVN following traumatic dislocation of the hip may have a similar presentation as Legg-Calvé-Perthes disease.[12] AVN is a known sequela of the treatment of developmental dislocation of the hip and may appear several years after treatment is completed.

Other radiographic findings have been mistaken for the disorder, such as a femoral head notch, which is a normal variant.[207] Findings comparable to those seen in Legg-Calvé-Perthes disease have also been noted in a number of

TABLE 16–8 **The Stulberg Classification System for End Results**

Group	Radiographic Findings (Anteroposterior and Frog-leg Lateral Radiographs)
I	Shape of femoral head is basically normal.
II	Loss of head height occurs, but head's contour conforms within 2 mm to a concentric circle on AP and frog-leg lateral radiographs.
III	Femoral head is more elliptical and deviates from a circle by more than 2 mm. Contour of acetabulum matches that of the femoral head ("congruous incongruity").
IV	Femoral head is flattened, with the flattened area greater than 1 cm in length. Contour of acetabulum matches that of the femoral head (same as group III).
V	Collapse of femoral head, but acetabular contour does not change ("incongruous incongruity"). Appearance is similar to that seen in adult avascular necrosis in which there is collapse of central portion of the head.

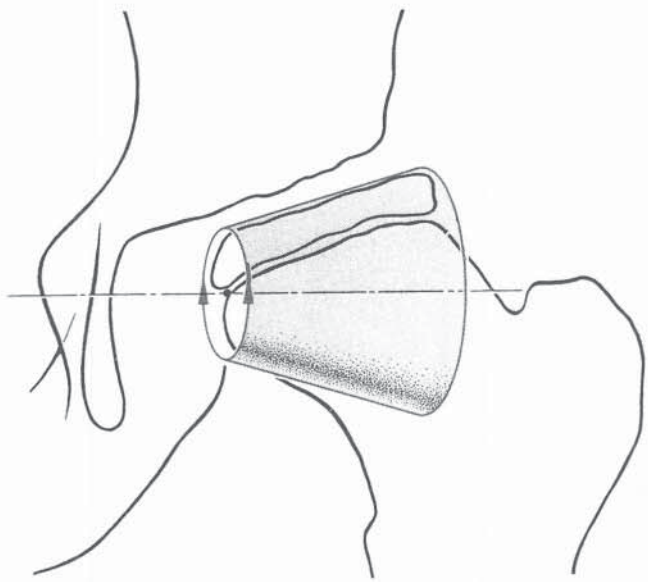


FIGURE 16–45 An illustration of the concept of "congruous incongruity" in Stulberg's group IV hips, in which the femoral head assumes a cylindrical shape. There will be almost normal range of flexion and extension, but no available rotation or abduction. Often the hip externally rotates with flexion and returns to a neutral position with extension. This motion has been likened to the movement of a cow's hip, which has a similar shape. (From Schultz KP, Dustmann HO: Morbus Perthes: Ätiopathogenese, Differentialdiagnose, Therapie und Prognose. Berlin, Springer-Verlag, 1991.) Copyright © 1991 by Springer-Verlag.

uncommon conditions, including Maroteaux-Lamy syndrome,[208] osteochondroma of the femoral neck,[25] multiple osteochondromatosis,[147] synovial osteochondromatosis,[186] metachondromatosis,[150] as well as the Schwartz-Jampel syndrome.[215] Perthes-like changes of the hip have also been reported in patients who have trichorhinophalangeal syndrome.[126,278] In these cases, radiographs will demonstrate distinctive cone-shaped epiphyses, and the patient will have abnormal hair and facies.[261]

Treatment

Because the severity of Legg-Calvé-Perthes disease can vary significantly from one patient to another, treatment decisions continue to be difficult, and the therapeutic methods applied to the disorder differ greatly between centers. Some centers prefer to limit treatment only to interventions that

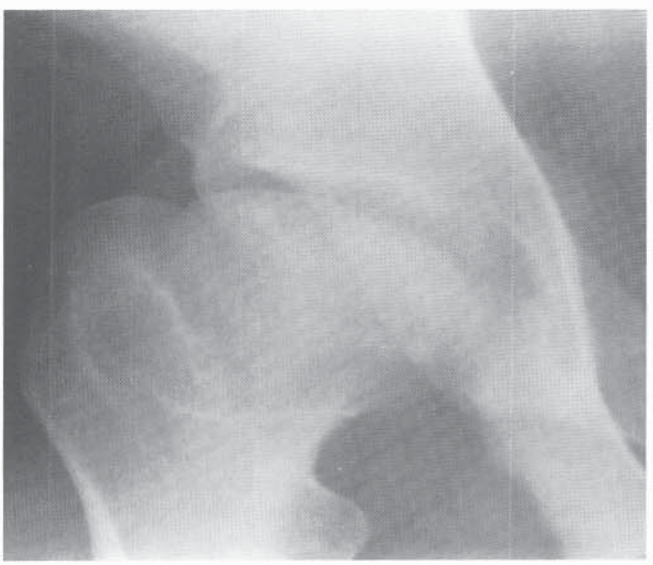


FIGURE 16–44 A hip with a Stulberg III result. AP radiograph obtained 11 years after onset of the disease shows the femoral head fully reossified and ovoid. (From Herring JA: Legg-Calvé-Perthes Disease. Rosemont, IL, American Academy of Orthopaedic Surgeons, 1996.)

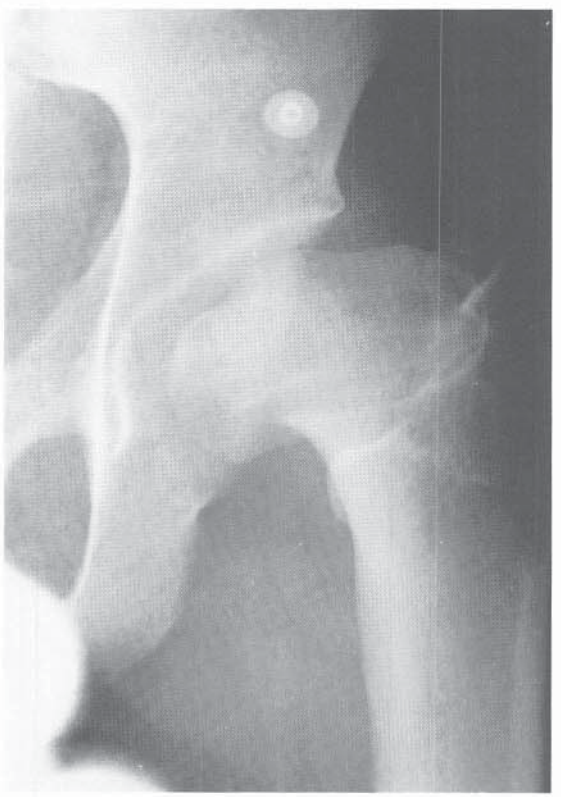


FIGURE 16–46 An example of a Stulberg V result. The femoral head is irregular, with hills and valleys, while the acetabulum has a smooth contour.

TABLE 16–9 **Prognostic Risk Factors Associated with a Poor Outcome**

Lateralization of the femoral head
Extent of uncovering of femoral head
Lateral calcification
Lateral head displacement (using head-to-teardrop distance)
Widening of femoral head before fragmentation
Saturn phenomenon (a sclerotic epiphysis surrounded by a ring of lucency)
Widening of femoral neck in early stages of disease

TABLE 16–10 **Conditions That Have Features in Common with Legg-Calvé-Perthes Disease**

- Conditions producing avascular necrosis, such as hemoglobinopathies, leukemia, lymphoma, idiopathic thrombocytopenic purpura, or hemophilia
- Hypothyroidism (patients may have radiographic changes resembling those of Legg-Calvé-Perthes disease)
- Multiple epiphyseal dysplasia, spondyloepiphyseal dysplasia, and Morquio's disease if there is a strong family history of hip abnormalities
- Traumatic dislocation of the hip resulting in avascular necrosis
- Uncommon conditions in which radiographic findings are similar to those of Legg-Calvé-Perthes disease, among them Maroteaux-Lamy syndrome, osteochondroma of the femoral neck, multiple osteochondromatosis, synovial osteochondromatosis, metachondromatosis, Schwartz-Jampel syndrome, and trichorhinophalangeal syndrome

maintain joint range of motion. Other centers apply nonsurgical and surgical containment methods to children believed to be at risk for developing significant deformation of the femoral head. Still others use surgical containment to treat most children who develop the disorder after 6 years of age, in the belief that aggressive treatment provides these patients with the best chance of a good outcome.

My current approach is based on a review of the early results of a prospective study of Legg-Calvé-Perthes disease that was started in 1984 (the results will be finalized and published as soon as the majority of patients have reached skeletal maturity). We have found, as have several other investigators,[68,133] that the lateral pillar classification system coupled with the age of the patient at onset of the disease provide useful prognostic information.

Our current treatment philosophy is to treat symptomatically those patients with group A hips and those with group B hips in whom onset of the disease occurred at a skeletal age of 6 years or less. Children with group A hips can usually be recognized and spared treatment, as they rarely experience persistent loss of joint motion or major symptoms. Initial management should focus on pain relief, with a reduction in activities and the use of anti-inflammatory medications, and short periods of bedrest for major episodes of pain or loss of joint motion.

Patients with group B hips in whom disease onset occurred after skeletal age 6 and all children with group C hips are treated by surgical containment once joint range of motion has been achieved by symptomatic means. Our current preferred surgical method is the Salter innominate osteotomy. Although femoral osteotomy produces similar results in terms of femoral head shape, this surgical procedure results in elevation of the greater trochanter and a subsequent tendency toward abductor dysfunction. In addition, removal of hardware is more problematic in patients who have undergone a femoral osteotomy. On the other hand, patients with more severe disease are more likely to develop postoperative stiffness after a Salter osteotomy than after a femoral osteotomy. Older patients (i.e., those over 9 years of age) with B and C hips may benefit from combined femoral and pelvic osteotomies, but the success rate is unpredictable in this patient population regardless of which treatment methods are applied (Table 16–11).

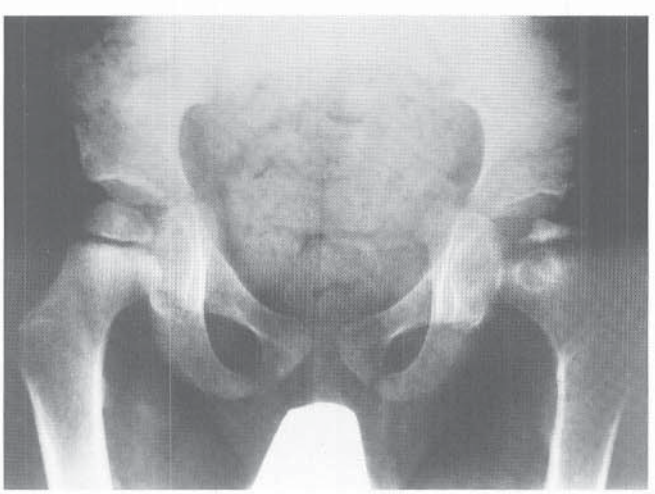


FIGURE 16–47 The Saturn phenomenon, a radiolucent ring occasionally seen in young children with Legg-Calvé-Perthes disease. When this pattern is present, the outcome is often poor. Note flattening of the epiphysis medially and laterally, with the central fragment having the shape of a flying saucer.

SYMPTOMATIC THERAPY

The two primary means of treating symptoms related to Legg-Calvé-Perthes disease are bedrest and traction. The use of nonsteroidal anti-inflammatory drugs (NSAIDs) for pain and discomfort and crutches to reduce weightbearing may also be of benefit.

Having the child rest in bed for a day can alleviate pain and restore range of motion of the affected hip, particularly during the early stages of the disorder (presumably due to reduction of synovitis). The beneficial effects of bedrest are greatest around the time of development of the subchondral fracture. As the disease progresses into the fragmentation and reossification stages, impaired hip motion is often due to deformity of the femoral head, and bedrest will not restore motion.

Traction of the affected leg can be implemented in a number of different ways. In the child's home, simple longitudinal traction using 5 pounds of weight can be attached to his or her bed. It may be necessary to use the traction only at night while the child is sleeping (every night, or only when the child experiences pain and spasms), or it may need to be applied for the entire day for several days or weeks until range of motion is regained. If the child is hospitalized, traction may also be used to gradually abduct the affected leg. Various approaches include simple longitudinal traction with the leg on the bed, balanced suspension and traction, and "slings and springs" (Fig. 16–50).

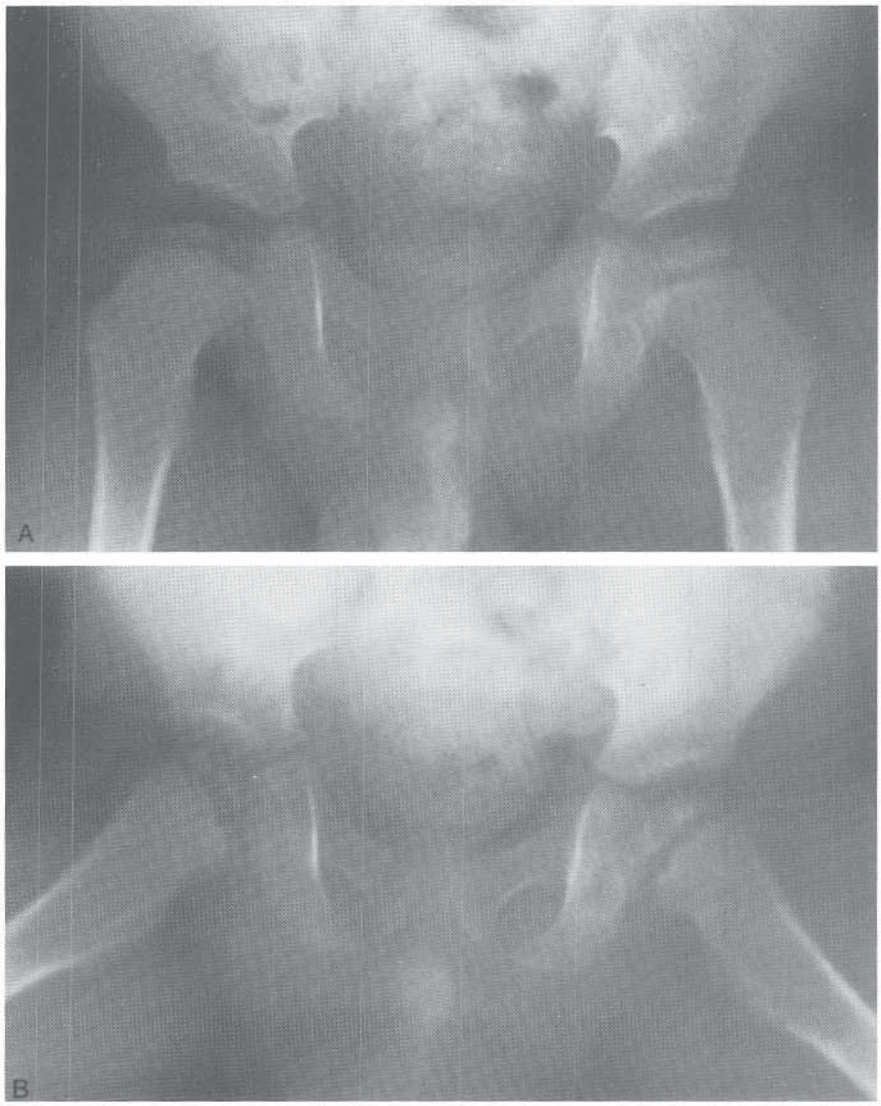


FIGURE 16–48 Epiphyseal changes in hypothyroidism that may be mistaken for Legg-Calvé-Perthes disease. **A,** AP and **B,** frog-leg lateral radiographs of both hips of a 4-year-old boy with hypothyroidism. Note the delay in and irregularity of ossification of the femoral heads.

Findings from a number of studies support the use of traction and provide guidance regarding the best hip position during traction. Elevated intra-articular hip pressure secondary to synovitis has been reported to be highest when traction was applied with the hip fully extended and lowest when the hip was flexed 30 to 45 degrees and rotated slightly externally.[140] Seven to 14 days of Russel's traction has also been reported to significantly reduce intra-articular pressure associated with Legg-Calvé-Perthes disease, to restore range of motion of the hip by a mean of 34 degrees, and to markedly decrease venous congestion of the femoral head.[253] In a canine study, investigators found that traction equal to one-half body weight did not adversely affect blood flow to the femoral head; however, traction equal to body weight resulted in marked compromise of blood flow.[195]

Partial weightbearing or nonweightbearing can help alleviate pain and increase range of motion. Crutches are frequently used for this purpose, even though there are no specific studies validating their use. In addition, it can be difficult to keep a child on crutches from weightbearing—a problem that led to the development of the Snyder sling (see Fig. 16–7).[255]

EXPERIMENTAL RATIONALE FOR CONTAINMENT TREATMENT

To assess the dynamic effects of various containment devices, Rab conducted gait analyses in children with Legg-Calvé-Perthes disease.[226] The use of Petrie casts resulted in an increase in anterior and lateral coverage of the femoral head and a reduction in posterior and medial coverage. By establishing hip flexion and external rotation of the limb, the Atlanta brace provided greater posterior coverage than lateral coverage. Using a "containment index," Rab reported that containment increased from a normal of 64 percent to 72 percent with the Atlanta brace, but that the index was unaffected with the Petrie casts and the Toronto brace.

In a subsequent finite element analysis of femoral head stresses, Rab and associates reported that if only the central part of the femoral head was necrotic, the unaffected lateral rim of bone would protect the central core from stress and

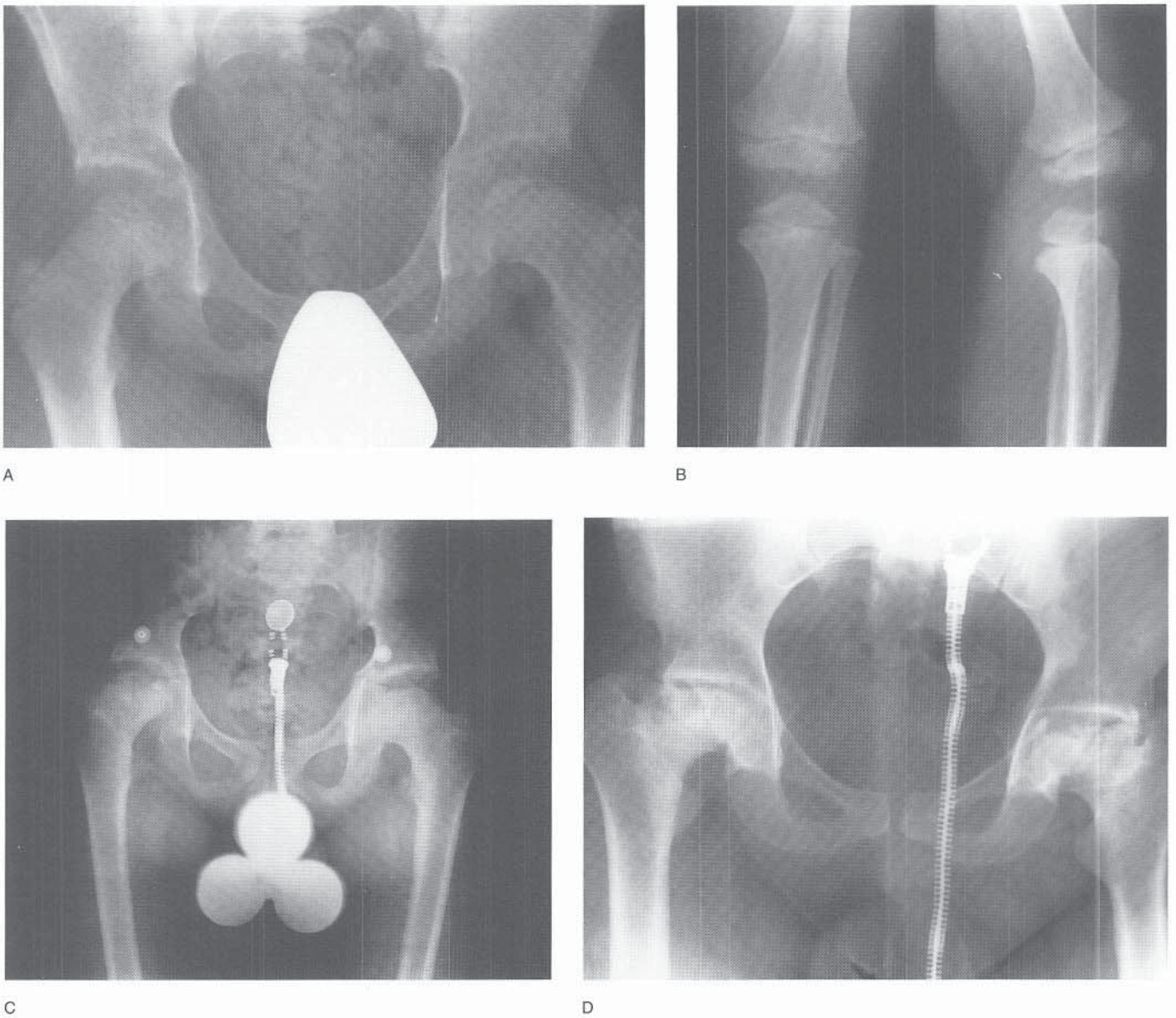


FIGURE 16–49 Multiple epiphyseal dysplasia (MED) simulating Legg-Calvé-Perthes disease in a 9-year, 7-month-old boy presenting with a painful left hip. The family indicated that the boy's father had had bilateral "Perthes" and was in need of hip replacements. **A,** AP radiograph showing increased density of the left femoral head and a right femoral head that is smaller than normal. **B,** Radiograph of the knees showing irregularity of the femoral and tibial epiphyses. **C,** AP radiograph of the boy's hips at age 13 years 7 months showing bilateral, symmetric changes. The femoral heads have not gone through the framgentation stage, as would be expected in Legg-Calvé-Perthes disease. **D,** AP radiograph of the hips of the boy's father showing bilateral flattening of the femoral heads and degenerative changes. (From Herring JA: Legg-Calvé-Perthes Disease. Rosemont, IL, American Academy of Orthopaedic Surgeons, 1996.)

prevent collapse.[227] However, when considerable necrosis of the femoral head was present, this shielding effect was lost and the head collapsed. The investigators noted that only 63 percent of the femoral head is in contact with the acetabulum at any one time, because the head represents 120 percent of a hemisphere whereas the acetabulum is only 75 percent of a hemisphere. Which segment of the head is covered by the acetabulum depends on the phase of gait as the joint moves. By examining varus osteotomy and innominate osteotomy configurations, the authors found that stresses were not reduced by the procedures, and they argued that no mechanical reason existed to justify containment osteotomies if considerable femoral head necrosis was present.

NONSURGICAL CONTAINMENT USING ORTHOTIC DEVICES

A number of different orthotic devices have been developed for patients with Legg-Calvé-Perthes disease to nonsurgically contain the femoral head. All braces abduct the affected hip, most allow for hip flexion, and some control rotation of

TABLE 16–11 **Author's Preferred Treatment Approach (Based on Lateral Pillar Classification System and Age of Patient at Onset of Disease)**

- Only symptomatic treatment for patients with group A involvement and patients with group B involvement whose onset was at skeletal age of 6 years or less. Initial management focuses on pain relief, with reduction in activities, anti-inflammatory medications, and short periods of bedrest for major episodes of pain or loss of joint motion.
- Patients with group B involvement whose onset occurred after skeletal age of 6 and all patients with group C involvement are treated with surgical containment once joint range of motion has been achieved. The author's current preferred surgical method is the Salter innominate osteotomy.
- Patients more than 9 years old with group B or C hips may benefit from combined femoral and pelvic osteotomies, but the success rate is unpredictable in this patient population, regardless of the treatment method used.

the limb. Before starting containment therapy, though, it is important to restore normal range of motion to the "irritable" hip. Bedrest, traction, and reduced weightbearing are beneficial in this regard. Throughout containment treatment, it is vital that range of motion be preserved, although this may be difficult in cases of severe disease.

In 1971 Petrie and Bitenc reported the results of treating 60 patients by placing them in casts with bars between the legs so that the hips were abducted to 45 degrees and rotated internally 5 to 10 degrees (these casts are referred to as Petrie casts; see Fig. 16–8).[214] This treatment program was achieved through traction or bedrest, with adductor tenotomy performed when required. Every 3 to 4 months the casts were changed, with the patient's knees and ankles mobilized between changes. Cast containment continued until the femoral head was well into the healing stage, with the average length of treatment being 19 months. Using the criteria from Mose's end-result classification system, the authors reported that good results were achieved in 60 percent of patients, fair results in 31 percent, and poor results in 9 percent. Petrie casts continue to be employed, particularly when other treatments are not effective. They have been reported to be useful in older patients whose range of motion is too restricted and painful to allow proper positioning of the femoral head for brace wear.[234]

A variety of braces (named for the city of origin or for the developer) have been designed and used over the years for containment therapy. Some of them, such as the Toronto brace and the Birmingham brace, were extremely complex devices that are not used much today. The Toronto brace (Fig. 16–51), which was very heavy and complicated, was created to keep the hip abducted while allowing hip and

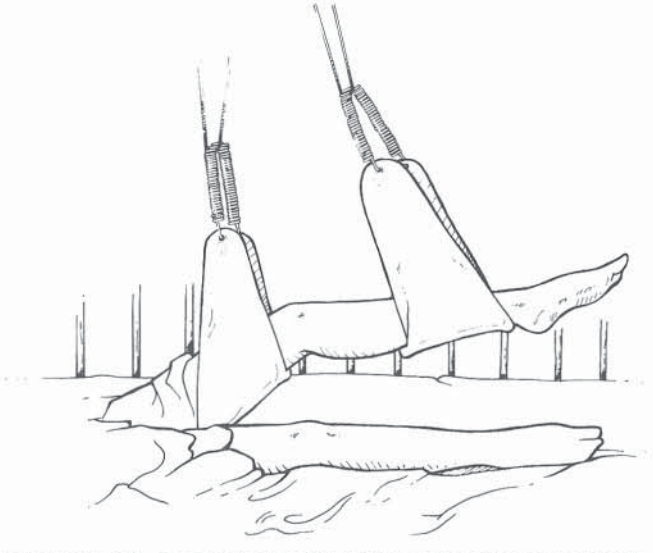


FIGURE 16–50 Longitudinal traction achieved with "slings and springs," a simple and useful method of gradually abducting the affected leg and restoring range of motion to an irritable hip. (From Herring JA: Legg-Calvé-Perthes Disease. Rosemont, IL, American Academy of Orthopaedic Surgeons, 1996.)

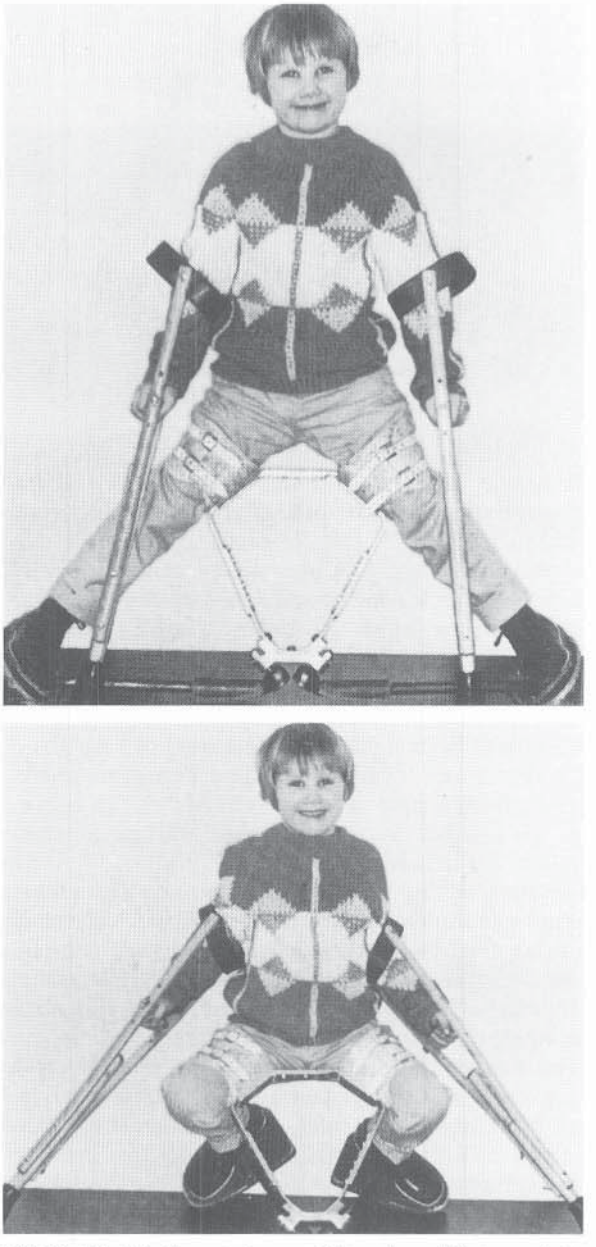
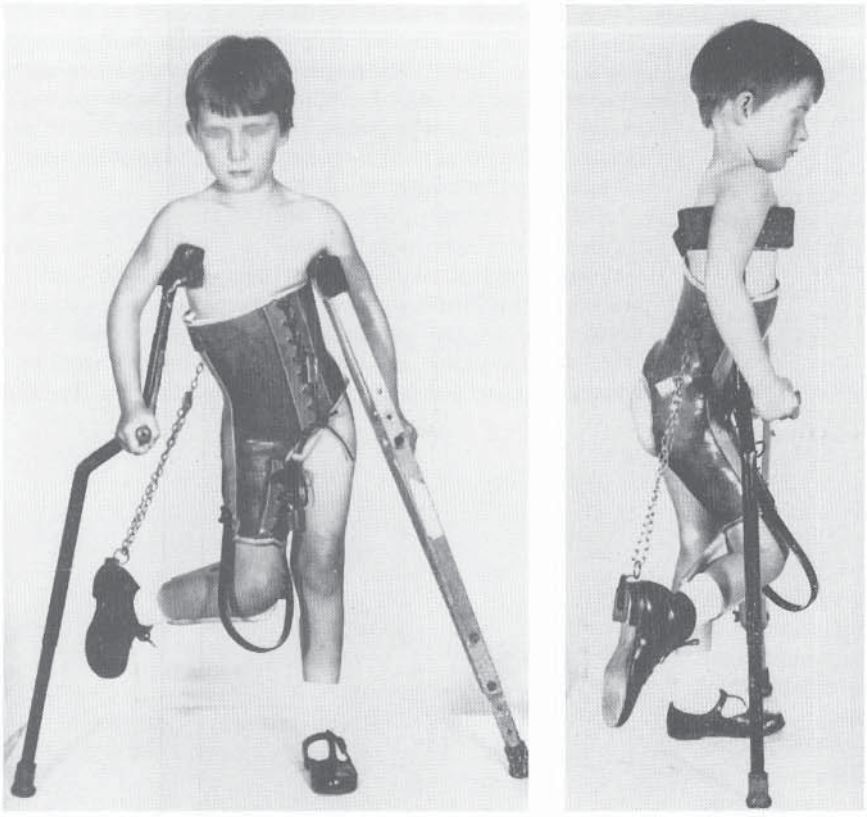


FIGURE 16–51 The Toronto brace, with its universal joints, was designed to keep the hip abducted while allowing hip and knee flexion. Thus, the patient can both sit and stand. (From Bobechko WP: The Toronto brace for Legg-Perthes disease. Clin Orthop 1974;102:115–117.)



A B

FIGURE 16–52 **A,** Frontal view of the Birmingham brace, which has a kneeling bar and a chain to keep the foot off the ground, while a specially altered crutch allows the abducted, internally rotated limb to clear the body when the patient walks. **B,** Side view of the Birmingham brace showing one of the three padlocks that keeps the child in the orthosis and ensures compliance. (From Harrison MH, Turner MH, Smith DN: Perthes' disease: treatment with the Birmingham splint. J Bone Joint Surg 1982;64-B:3.)

knee flexion (thus, the patient could both sit and stand). The universal joint at the base of the brace often wore out during patient use.[19,20] The Birmingham brace (Fig. 16–52) was created with the goal of attaining perfect immobilization.[111,112] The brace incorporated a leather corset that extended from the nipple line to just above the knee of the affected leg. A kneeling bar and a chain kept the foot off the ground, while a specially altered crutch permitted clearance of the abducted, internally rotated limb when the patient walked. Several padlocks were used to keep the child in the device and ensure compliance. Tachdjian and Jouett created a fairly simple unilateral orthosis with an ischial seat (Fig. 16–53), but the device lacked enough purchase to maintain the irritable hip in proper position.[269] A leather brace designed by Roberts replicated the abduction and internal rotation maintained by Petrie casts.[237] The Newington brace (Fig. 16–54), another unwieldy and complicated device, incorporates a metal A-frame with a central support for the thighs.[53,58] It also reproduces the hip position obtained with Petrie casts.

The Atlanta Scottish Rite brace (Fig. 16–55A) is commonly used today. The device consists of a metal pelvic band, hip hinges, thigh cuffs, and an extensile bar between the thigh cuffs that permits abduction of the limb but restricts adduction. With the hips abducted, the legs are usually flexed and externally rotated when the patient walks. With this brace, many patients quickly regain the ability to walk and run, and are able to return to normal physical activities (Fig. 16–55B). As with all containment treatments, obtaining and preserving hip range of motion is essential. In a 1980 description of the use of the Atlanta brace, Purvis and associates reported that 1 to 2 months of preliminary treatment was necessary to regain range of motion in severely affected hips.[224] If motion was lost during containment treatment, the patient was put back in traction. The brace was worn 24 hours a day until there was evidence of new bone formation on both AP and lateral radiographs. At that point, the patient was weaned from the orthosis, with the average length of treatment being 19 months.

Present treatment protocols with the Atlanta brace are usually not as demanding. At our institution, treatment programs are individualized to the patient's particular condition. In cases of mild disease (as determined by radiographic findings and by the patient's ability to maintain range of motion), the brace is used only during the day. If there is no collapse of the femoral head and if the lateral pillar is intact when the disease process enters the fragmentation stage, the brace is employed on a part-time basis as long as the patient maintains adequate range of motion. Treatment is stopped when there is radiographic evidence on the AP view of new subchondral bone formation. The average length of treatment ranges between 9 months and 1 year. In cases of severe disease, it is necessary for the patient to wear the brace full time, with range of motion preserved during the entire treatment program.

SURGICAL CONTAINMENT

Femoral Osteotomy. Since the 1960s, varus derotational femoral osteotomy has been successfully used to contain the

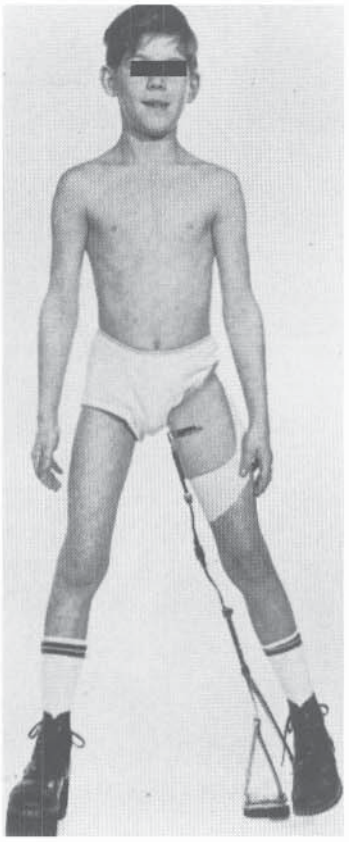


FIGURE 16–53 Frontal view of the trilateral socket hip abduction orthosis (the Tachdjian brace), a unilateral brace with an ischial seat.

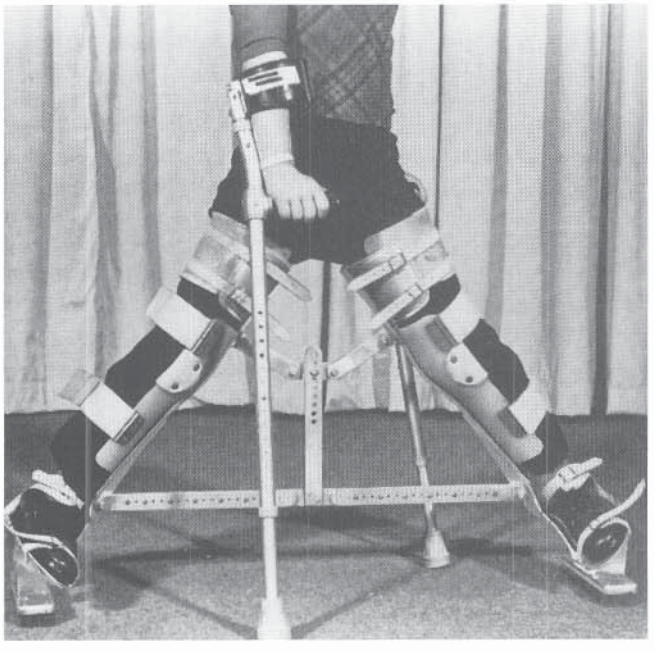


FIGURE 16–54 The Newington brace incorporates a metal A-frame with a central support for the thighs. It also reproduces the hip position obtained with Petrie casts. (From Drennan JC: Orthotic management of Legg-Calvé-Perthes disease. In Leach RE, Hoaglund FT, Riseborough EJ (eds): Controversies in Orthopaedic Surgery, p 315. Philadelphia, WB Saunders Co, 1982.)

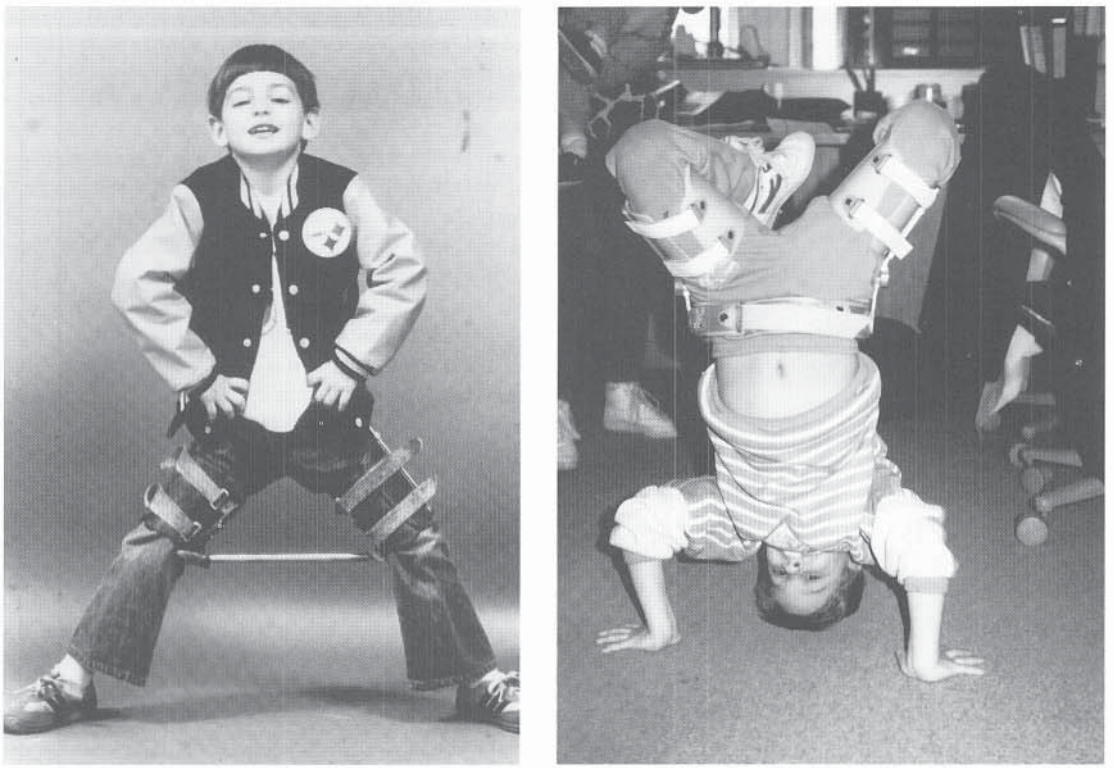
femoral head in patients with Legg-Calvé-Perthes disease (Fig. 16–56).[7,49] The two most common indications for femoral osteotomy in this patient population are onset of disease after 6 years of age and a hip with a poor prognosis based on radiographic findings. Surgery has not been found to improve outcome in children younger than 6 years at onset of the disease,[173] although some surgeons have operated on patients as young as 3 years 6 months.[182] Femoral osteotomy has been recommended for hips that have "at-risk" radiographic signs, even if the head is not severely deformed.[173]

Before femoral osteotomy is performed, it is important that the patient regain a reasonable range of motion. Petrie casts can be used to achieve this goal. Others have employed preoperative traction, using skeletal traction when skin traction did not restore sufficient motion to contain the head within the acetabulum. Using this approach to distract the joint and seat the head in the acetabulum, the surgeons were able to contain subluxated hips and those that were "hinge abducted."[152] Some have refuted the need for preoperative traction prior to femoral osteotomy based on the finding that hip motion improved under anesthesia.[173]

Whether femoral osteotomy accelerates the healing of the femoral head is questionable. Although some investigators have suggested this possibility, others have found no difference in healing when comparing hips treated surgically with those treated nonsurgically.[179] In addition, when "biologic" osteotomies were performed in which there was no displacement of bone fragments, there was no effect on the rate of healing.[44,149]

There also is debate as to when femoral osteotomy should be performed and in which patients it is appropriate. Some authors have recommended performing the operation within 8 months of onset of symptoms.[173] One report found that premature physeal closure was more likely if femoral osteotomy was performed very early or very late.[11] In general, femoral osteotomy should be performed before reossification of the femoral head begins. The extent of remodeling of the varus femoral neck depends on the growth of the capital physis, a factor that is difficult to predict. Certain patients develop an abductor limp following surgery, and trochanteric epiphysiodesis has been recommended when trochanteric overgrowth is likely (Fig. 16–57).[146]

Some aspects of the surgical technique of femoral osteotomy itself are also controversial. There is a difference of opinion regarding the degree of varus and derotation necessary to contain the femoral head. In one series the preoperative angle of the neck shaft averaged 137 degrees, the immediate postoperative angle was 116 degrees varus, and at final follow-up the angle was 129 degrees.[65] A postoperative neck-shaft angle of 100 to 110 degrees varus has also been recommended.[115] This was not considered excessive, based on the fact that the femoral neck tends to grow back into valgus. Laurent and Poussa reported adding 30 degrees of varus regardless of the neck-shaft angle.[164] Others, however, believe that the amount of postoperative varus should be limited to that needed to position the femoral head just under the lateral rim of the acetabulum, and that the angle should never be less than 105 degrees.[285] Extension derotational osteotomy has been used to decrease residual deformity of the femoral neck.[183] To decrease the amount of trochanteric overgrowth, some surgeons perform an epiphysiodesis of



A B

FIGURE 16–55 **A,** Frontal view of an Atlanta Scottish Rite brace, which consists of a metal pelvic band, hip hinges, thigh cuffs, and an extensile bar between the legs that permits abduction but restricts adduction. With the hips abducted, the legs are usually flexed and externally rotated when the patient walks. **B,** This view of a patient in the Atlanta brace shows one of the activities possible in the orthosis. The brace allows considerable mobility and enables many patients to quickly regain the ability to walk, run, and return to normal physical activities. (From Herring JA: Legg-Calvé-Perthes Disease. Rosemont, IL, American Academy of Orthopaedic Surgeons, 1996.)

the greater trochanter at the time of osteotomy or at plate removal.[65,183]

A number of complications are associated with femoral osteotomy. These include excessive postoperative varus, failure of the varus to remodel, persistent external rotation of the limb following rotational osteotomy, shortening of the extremity, increased abductor lurch resulting in an abductor limp, trochanteric overgrowth, the need to remove the fixation device, fracture after removal of the plate, and delayed union or nonunion.[65,146] Compared with innominate osteotomy, femoral osteotomy has been found to cause greater shortening of the limbs and to more frequently result in abductor limp.[259] Further collapse of the femoral head, with severe loss of range of motion, has been occasionally noted postoperatively (requiring rigorous treatment with traction, muscle releases, and abduction casting or bracing).[288] Loss of external rotation and abduction has also been reported.[65] A reduction in articular trochanteric distance associated with a Trendelenburg gait may occur following surgery, particularly in older patients. As a result, some surgeons do not recommend femoral osteotomy for children over 8 years old.[169] In some cases a second osteotomy is required when the first operation does not provide adequate containment of the femoral head.[8]

Innominate Osteotomy. The first innominate osteotomy for patients with Legg-Calvé-Perthes disease was performed by Salter in 1962.[246] His indications for the procedure included onset of disease after 6 years of age, a moderately or severely affected head, and loss of containment. Preoperative prerequisites were minimum deformity of the femoral head (as determined by arthrography), no irritable hip, and no significant restriction of range of motion. In addition, the hip had to be able to abduct to 45 degrees and the femoral head had to be contained in that position.[245] To regain joint motion before surgery, many patients required bedrest, traction, use of slings and springs, and, at times, surgical release of the adductor muscles with abduction casts for several weeks.[246]

The originally described surgical technique was modified with the recommendation that the capsule should not be opened and that the iliopsoas should always be lengthened.[246] If the adductor muscles were tight, they too should be lengthened. In most cases postoperative casting was not necessary if the osteotomy was secured with two or three heavy threaded pins or screws (screws are preferred in older children) (Fig. 16–58).

Some surgeons favor the Kalamchi version of the Salter procedure, in which the distal pelvic fragment is repositioned into a notch created posteriorly in the proximal side of the transected ilium.[139] By doing so, the acetabulum is repositioned without lengthening the pelvis, thereby preventing increased pressure on the femoral head.

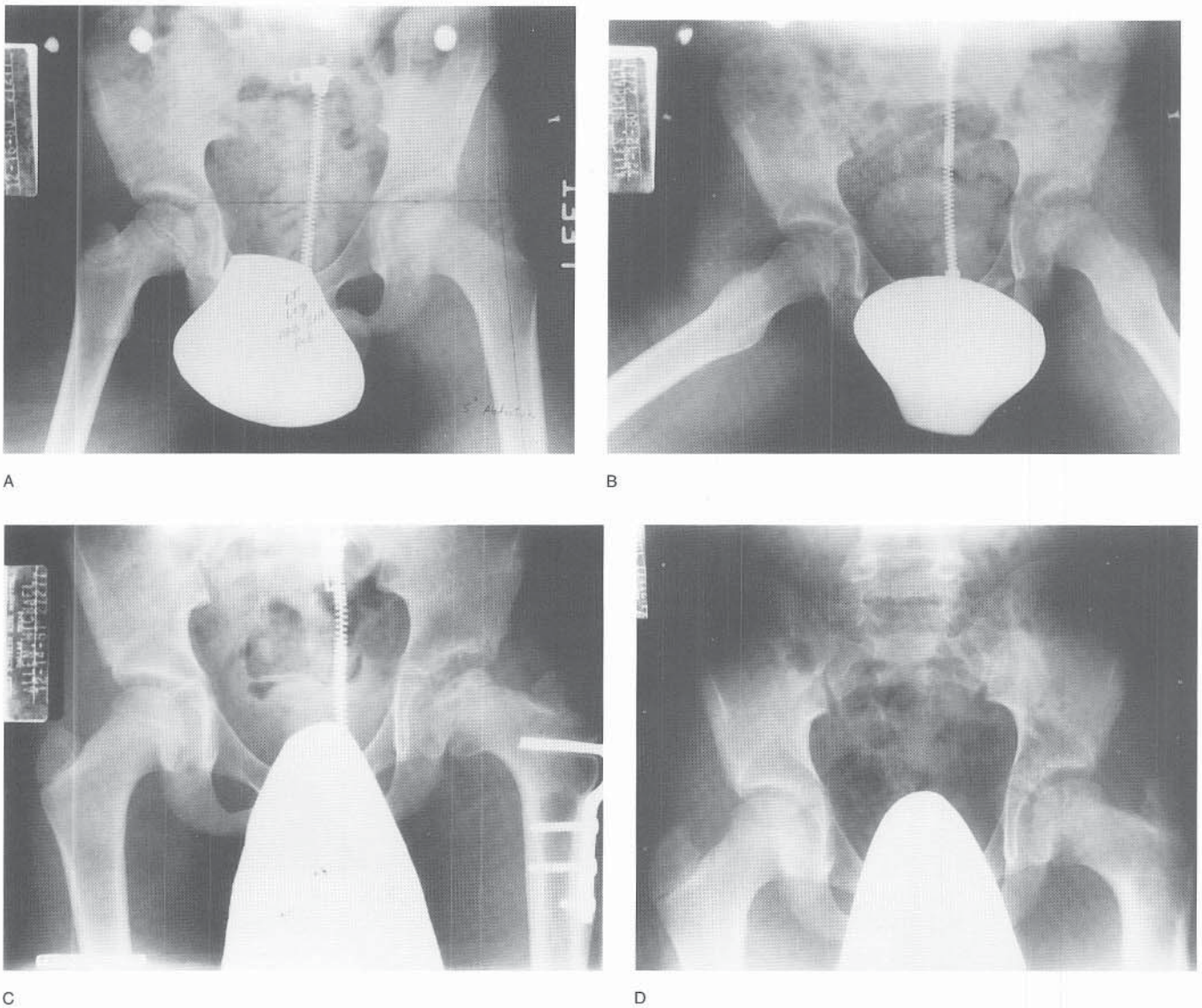


FIGURE 16–56 A 9-year, 8-month-old boy treated with a varus femoral osteotomy. **A,** AP radiograph showing early disease changes with a subchondral fracture. **B,** Frog-leg lateral radiograph showing a subchondral fracture extending over one-half the femoral head. **C,** AP radiograph obtained 1 year after onset of the disease and 6 months after the varus femoral osteotomy. The femoral head is well covered and the greater trochanter is at the level of the head. **D,** AP radiograph obtained 4 years after onset of the disease shows a round femoral head (a Stulberg II result). The blade plate has been removed. (From Herring JA: Legg-Calvé-Perthes Disease. Rosemont, IL, American Academy of Orthopaedic Surgeons, 1996.)

Some of the complications associated with innominate osteotomy include loss of fixation with displacement of the distal fragment, lengthening of the leg, decreased hip flexion, and joint stiffness.[46] Hinge abduction can also occur postoperatively (Fig. 16–59).

Combined Femoral and Innominate Osteotomy. Combining a femoral osteotomy with an innominate osteotomy has also been recommended for severely affected hips at high risk of a poor outcome.[52,202] Indications for the procedure are the presence of lateral subluxation, lateral calcification, and considerable changes in the metaphysis. This dual surgical approach may provide greater coverage of the femoral head than that achieved with either procedure alone. To maintain containment of the femoral head after surgery in more severe cases, the hip may have to be braced or casted for an extended period of time.[52]

Valgus Osteotomy. For Legg-Calvé-Perthes disease, valgus osteotomy was initially recommended for treating hinge abduction, in which the flattened femoral head interferes with abduction.[37,244] The operation is performed if the head and acetabulum are congruent when the joint is adducted but are incongruent in a neutral or abducted position. The most noticeable result following abduction of the limb is improvement in gait. Valgus osteotomy has also been reported to improve roundness of the femoral head.[37] Other reported positive results include healed central head fragmentation; improved joint space, joint motion, and leg length; reduced subluxation; and lessened pain.[225] These

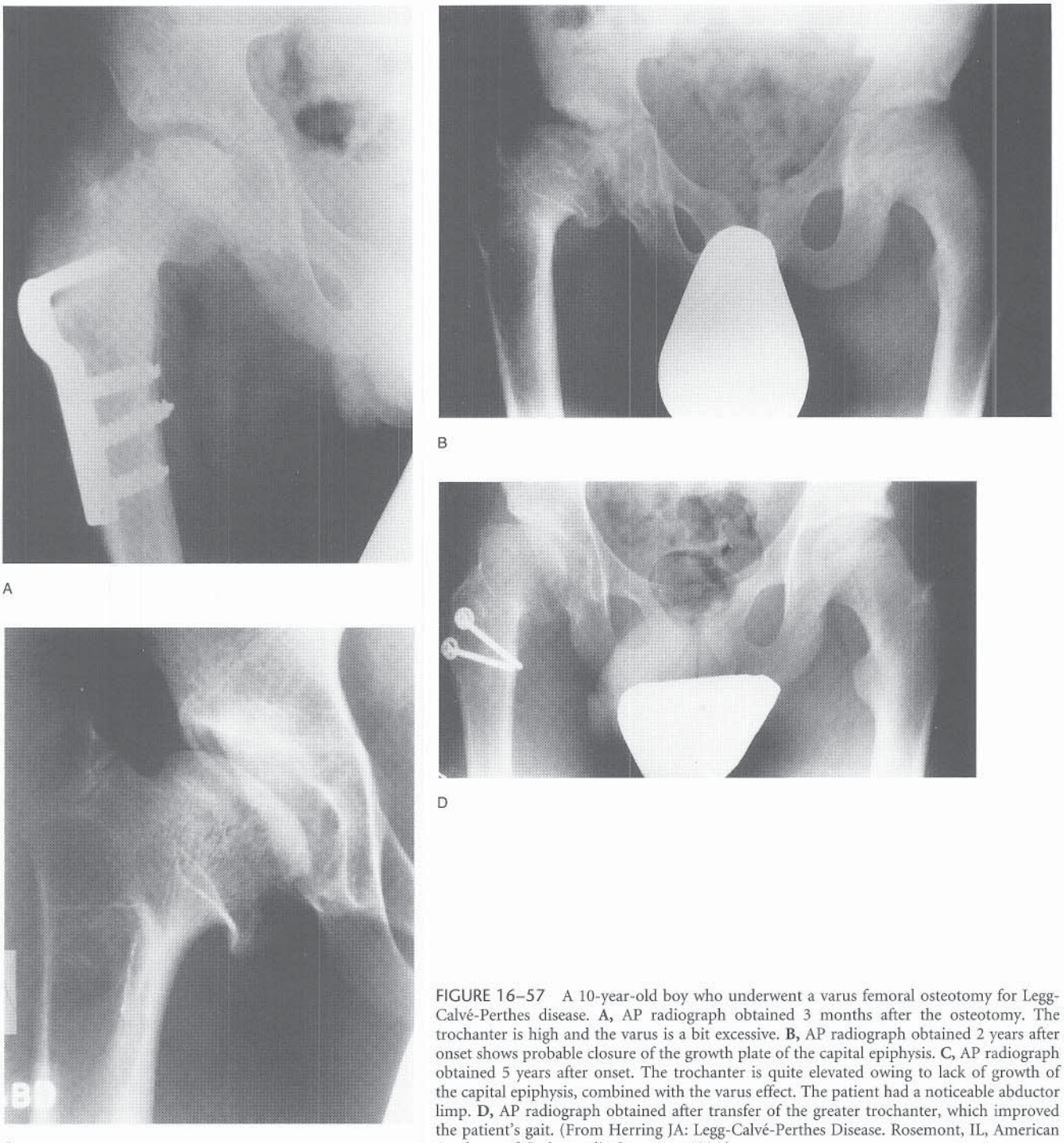


FIGURE 16–57 A 10-year-old boy who underwent a varus femoral osteotomy for Legg-Calvé-Perthes disease. **A,** AP radiograph obtained 3 months after the osteotomy. The trochanter is high and the varus is a bit excessive. **B,** AP radiograph obtained 2 years after onset shows probable closure of the growth plate of the capital epiphysis. **C,** AP radiograph obtained 5 years after onset. The trochanter is quite elevated owing to lack of growth of the capital epiphysis, combined with the varus effect. The patient had a noticeable abductor limp. **D,** AP radiograph obtained after transfer of the greater trochanter, which improved the patient's gait. (From Herring JA: Legg-Calvé-Perthes Disease. Rosemont, IL, American Academy of Orthopaedic Surgeons, 1996.)

are anecdotal accounts, though, and no long-term studies have been published regarding the procedure's effectiveness.

Shelf Arthroplasty. Shelf arthroplasty has been used to treat patients with Legg-Calvé-Perthes disease since 1940.[158] Indications for the operation include lateral subluxation of the femoral head, insufficient coverage of the femoral head, or hinge abduction of the hip. The procedure is often performed after other nonoperative treatments have failed.[158] Some surgeons, however, believe that shelf arthroplasty should be considered a primary means of containment and have performed the operation within 6 months of the onset of symptoms.[289]

For Legg-Calvé-Perthes disease, the shelf procedure has been performed as described by Gill, in which a deep notch is made above the acetabulum to permit the acetabular roof to be "pried down."[82] Others prefer the Staheli technique.[260] Postoperative complications include loss of hip flexion due to an excessively wide augmentation graft, and inadequate

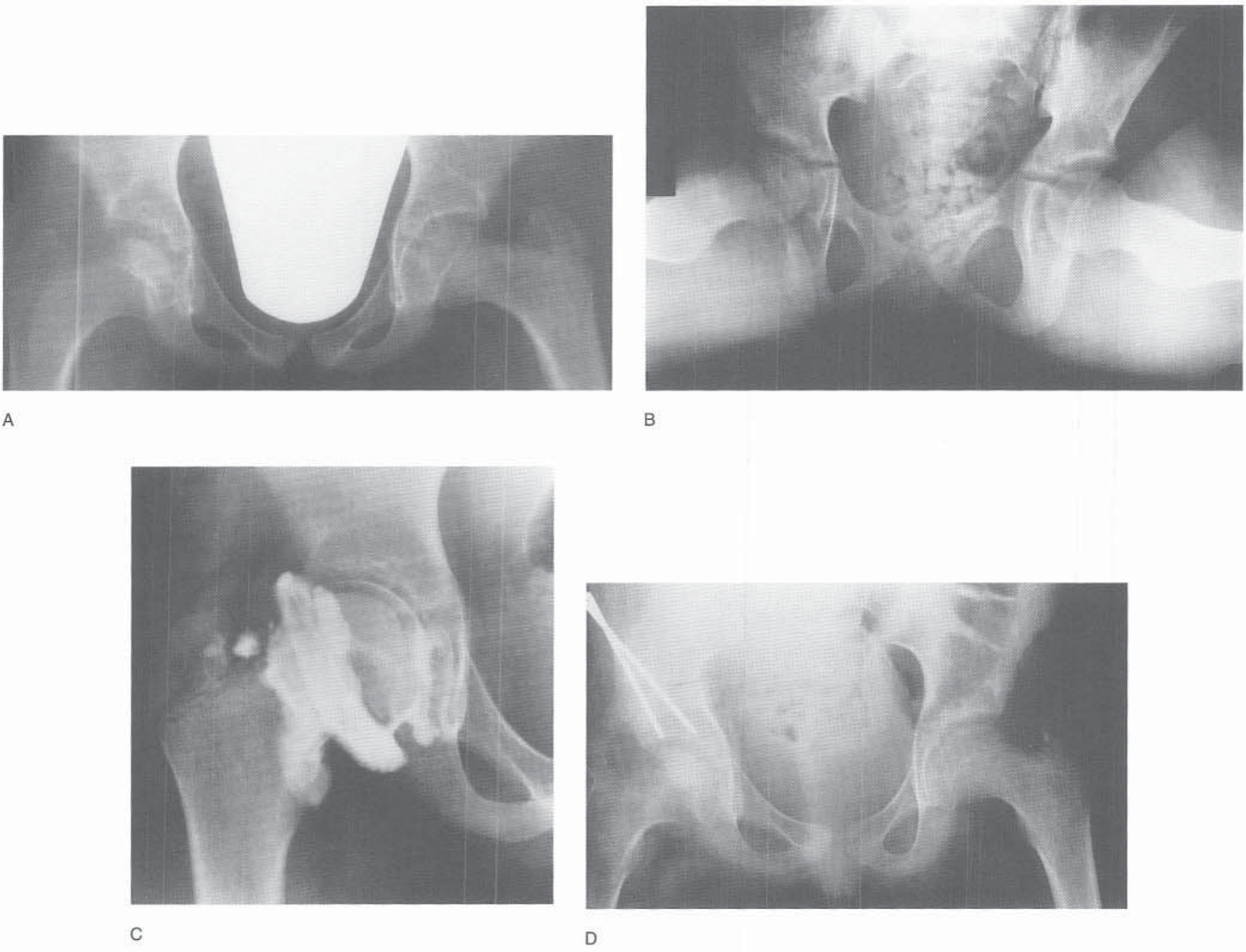
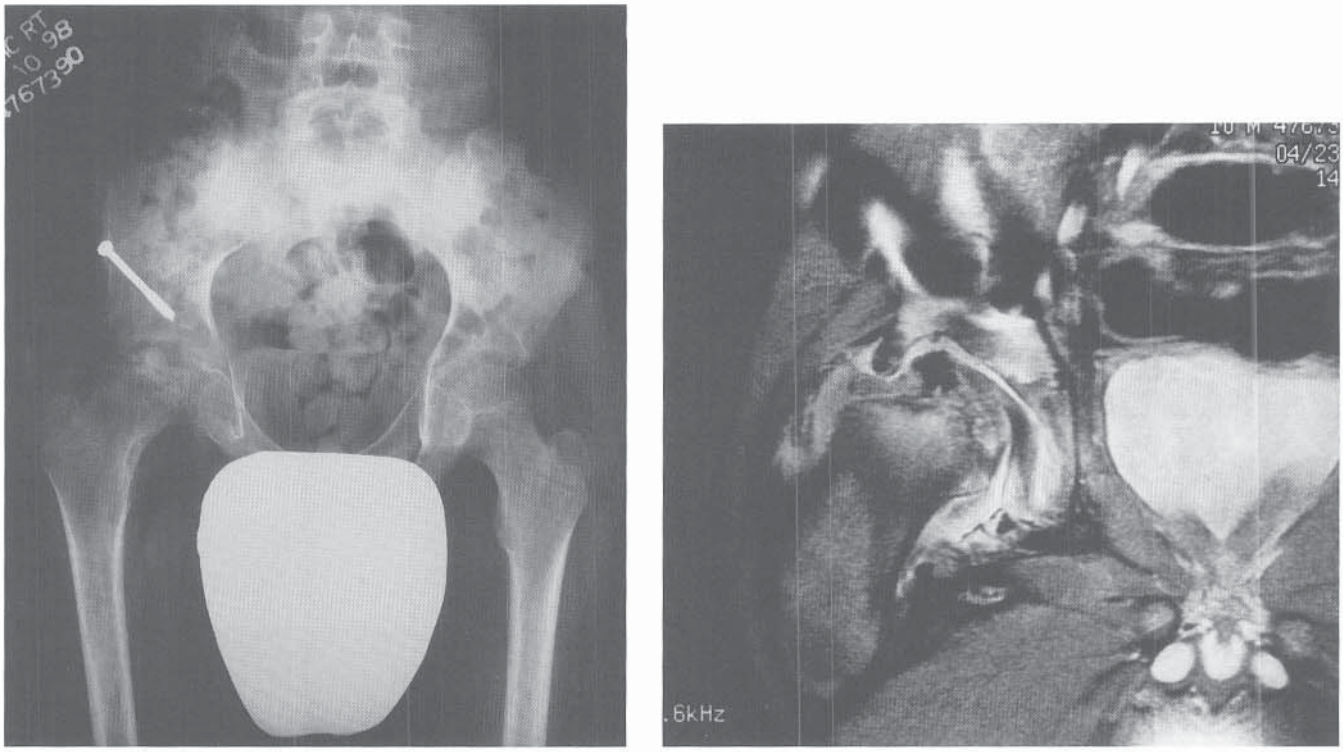


FIGURE 16–58 A 10-year-old girl treated with a Salter innominate osteotomy. **A,** AP radiograph showing a subchondral fracture. **B,** Frog-leg lateral radiograph showing the fracture extending over one-half of the femoral head. **C,** An arthrogram showing that the joint is congruous. **D,** AP radiograph obtained after the innominate osteotomy was fixed with threaded pins. We often use screw fixation to gain stability and avoid postoperative immobilization. **E,** AP radiograph obtained 8 years after the osteotomy shows a rounded femoral head (a Stulberg II result). The trochanter is high, suggesting early closure of the capital physis. (From Herring JA: Legg-Calvé-Perthes Disease. Rosemont, IL, American Academy of Orthopaedic Surgeons, 1996.)

hip coverage because the graft was too thin. As with any anterior hip procedure, patients may also experience dysesthesia of the lateral femoral cutaneous nerve following surgery.[260]

Chiari Osteotomy. Although Chiari's pelvic osteotomy has been used by some surgeons as a primary procedure for patients with Legg-Calvé-Perthes disease,[32,41,159] it has been described by others as difficult, demanding, and even unsafe.[10,16,175] Chiari's osteotomy is usually reserved for surgically treating the healing femoral head that remains lateralized.[14,155] It has also been recommended for use in older children who present with a painful hip, significant femoral head deformity, and incongruity between the head and acetabulum as demonstrated on arthrography (Fig. 16–60).[14] The actual benefits of this surgery are unknown, though, because roundness of the femoral head often improves gradually until the patient reaches skeletal maturity, even when untreated.[121]

Cheilectomy. Surgically removing protruding fragments of the femoral head (usually the anterolateral segment) was performed in the past,[78] but excision is seldom, if ever, carried out today because of potentially serious complications. The procedure weakens the attachment of the femoral head to the neck, which can result in epiphyseal slippage. In addition, most patients experience varying degrees of postoperative joint stiffness.[246] If the procedure is done, it should always be performed after closure of the capital physis to avoid producing a slipped epiphysis.



A B

FIGURE 16–59 A case of postoperative hinge abduction in a 10-year-old boy 9 months after a Salter osteotomy, resulting in severe limitation of abduction. **A,** AP radiograph showing healed osteotomy and apparent loss of joint space, with lateralization of the femoral head. **B,** MR image showing indentation of the lateral acetabular lip into a depression in the femoral head. A medial joint space is evident in this imaging study.

TREATMENT OF OSTEOCHONDRITIC LESIONS IN THE FEMORAL HEAD

In a few cases of Legg-Calvé-Perthes disease, hip pain will appear late in adolescence after the patient has been asymptomatic for several years. Complaints of locking, catching, or crepitation can indicate the presence of an osteochondritic lesion in the femoral head.[96,246] Radiographs may demonstrate a lucent area in the central part of the head (Fig. 16–61A); however, it is often difficult to differentiate a loose body from softened cartilage and fibrous tissue. To help establish the diagnosis, arthrography should be performed. If the contrast material surrounds the defect, it is most likely a loose fragment.[91,104] A normal arthrogram does not exclude the presence of a loose body, and the defect may need to be evaluated by arthroscopy or arthrotomy of the hip.[85]

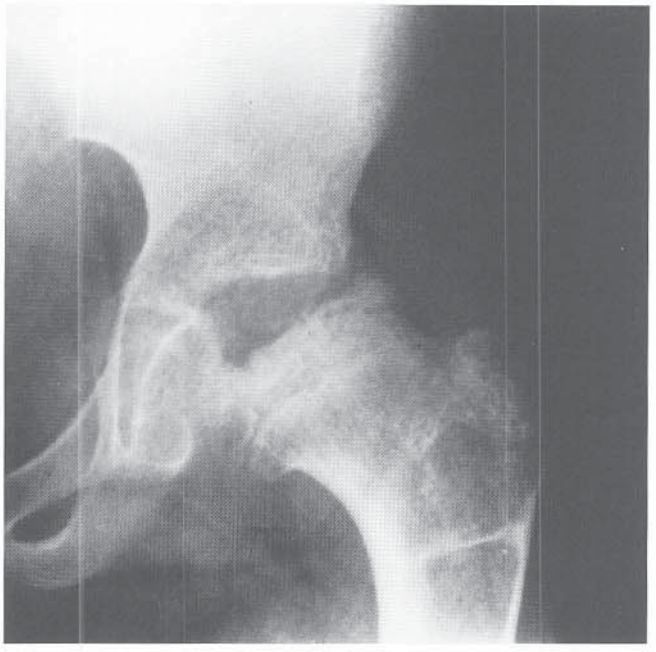
When the symptoms are not severe, nonsurgical treatment is often successful in managing the condition.[24,144,205] Conservative therapy includes rest, refraining from activities that exacerbate symptoms, and the use of NSAIDs for pain. When excision of the lesion is deemed necessary, an attempt should be made to remove it arthroscopically and the bed of the lesion should be debrided.[24,249] The hip may also be approached via an arthrotomy; however, to see the central fragment, the surgeon may need to dislocate the joint.[104,291]

If the lucent area represents a softened area of the femoral head and not a loose body, treatment may or may not be effective (Fig. 16–61B). Drilling the affected area may help trigger ingrowth of blood vessels and healing, but results are mixed.

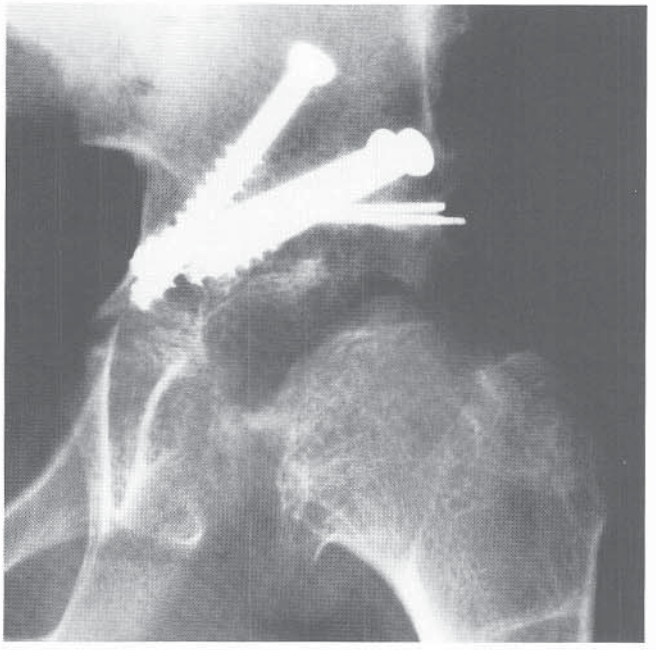
Long-Term Prognosis

The most important predictor of long-term outcome of Legg-Calvé-Perthes disease is the shape of the healed femoral head and its congruency with the acetabulum.[92,116,264] In a classic description by Stulberg and associates, hips were placed into one of five classes of deformity based on their radiographic appearance at maturity.[263] Long-term outcome was associated with the type of congruency that existed between the femoral head and acetabulum. Class I and II hips had spherical heads, there was congruency between the head and acetabulum, and the patients did not develop arthritis over the long term. The heads in class III hips were neither spherical nor truly flat, while the heads and acetabula in class IV hips were both truly flattened. Class III and IV hips had what was referred to as "aspherical congruency," and the patients developed mild to moderate arthritis in late adulthood. The heads in class V hips had a loss of shape without any concomitant change in the acetabulum, resulting in "aspherical incongruency." These patients developed severe arthritis before the age of 50 years.

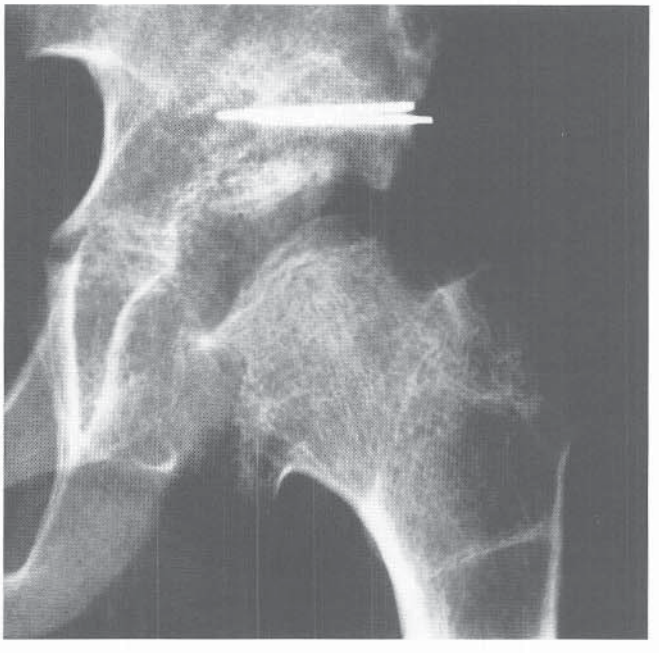
Earlier studies of the natural history of the disorder described very promising results.[60,92,230] A number of patients who were not treated or who were treated for less than



A



B



C

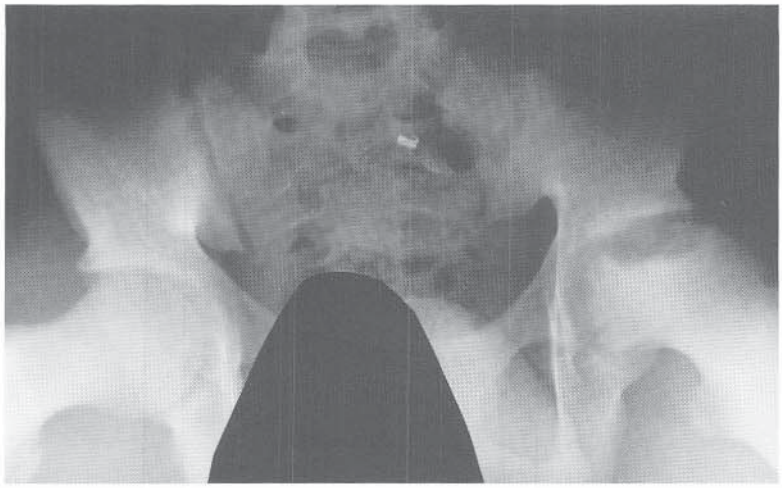
FIGURE 16–60 **A,** AP radiograph of an 11-year-old boy initially treated with a varus osteotomy. At the time the imaging study was performed, he had pain and persistent extrusion of the femoral head. **B,** AP radiograph obtained after a Chiari osteotomy. **C,** AP radiograph of the boy at age 15 years. There is improved coverage of the femoral head, which remains severely deformed. The patient's symptoms improved, but the prognosis for this hip remains guarded.

6 months with bedrest had good to excellent results.[60] Even patients who did not have good radiographic findings were fully active and pain free.[230] Subsequent reports, though, have noted an increased incidence of late degenerative arthritis in this patient population.[64,180,210,241]

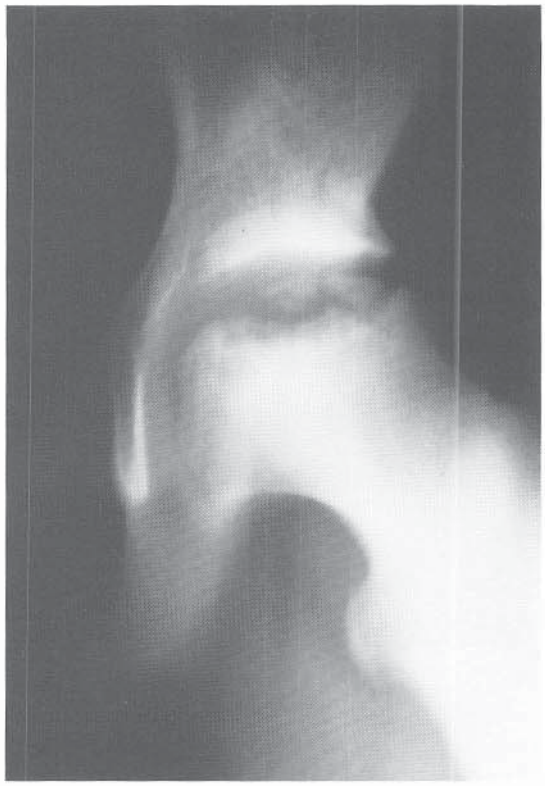
Retrospective studies of patients with degenerative arthritis of the hip have also revealed a high incidence of previous hip joint disease, including Legg-Calvé-Perthes disease, old slipped epiphysis, acetabular dysplasia, other epiphyseal dyplasias, juvenile rheumatoid arthritis, and postreduction AVN in congenital hip dislocation.[48,107,257]

Factors associated with late degenerative disease include the patient's age at onset of the disease, lateral calcification, loss of sphericity of the femoral head,[64] and the degree of steepness of the lateral edge of the acetabulum.[241] Better outcomes were associated with diagnosis before 9 years of age, a congruous joint, and minimal involvement of the head and neck.[210]

In a reevaluation in 1984 of 35 patients (average age, 48 years) originally treated between 1920 and 1940 with spica cast changes every 2 months for 1 to 23 months, it was found that almost 50 percent of these patients had subsequently undergone arthroplasty for degenerative disease.[180] Only 40 percent of the hips had good function. Degenerative disease



A



B

FIGURE 16–61 A 13-year-old boy who sustained the onset of Legg-Calvé-Perthes disease at age 8 years. Shortly before the imaging study he began experiencing pain in the left hip, with locking and popping. **A,** Frog-leg lateral radiograph showing a persistent lucent area in the center of the femoral head. **B,** Tomogram showing failure of reossification in the head. Arthroscopic evaluation and drilling might relieve some of the patient's symptoms, but large lesions like this one may be difficult to treat.

was associated with the presence of two or more Catterall risk factors, disease onset after age 8 years, and the degree of coxa magna. A spherical head was associated with good results. The conclusion that the only assurance of a good outcome is a spherical head and that 50 percent of patients will eventually develop degenerative arthritis significant enough to require hip replacement challenged the concept that Legg-Calvé-Perthes disease is a benign disorder. However, the findings are limited by the small number of the patients in the study (whose disease may not represent the true spectrum of the disorder) and by the fact that all of the patients underwent substantial treatment early in the disease process, which may have changed the results for better or worse.

Final outcome is also significantly affected by the age of the patient at onset of the disease and by the duration of the condition. In general, the younger the patient is at onset, the milder is the course of the disease.[27,38,65,124,129,132,256] Patients less than 6 years old normally fare the best. Those diagnosed with the disorder in adolescence have the worst outcomes. Final results also are better when the length of time from onset of the disorder to complete resolution is short. The longer it takes for the hip to heal, the less likely is a good outcome.[120]

Studies have also addressed the influences that other factors might have on final outcome. If the central portion of the head does not heal properly, patients may experience persistent symptoms due to osteochondritis dissecans.[24,205] Premature closure of the capital physis and a mushroom head have also been associated with a poor prognosis.[45] Finally, in a study of patients treated with varus osteotomy, five early radiographic signs were identified that predisposed patients to a poor outcome.[223] These signs were calcification lateral to the femoral head and extending toward the greater trochanter, deformation and widening of the femoral head prior to the fragmentation stage, deformation and widening of the femoral neck early in the disease process, the Saturn phenomenon (a sclerotic epiphysis surrounded by a ring of looser bony tissues), and early wide metaphyseal sclerotic changes.

Because long-term outcome is directly related to the shape of the healed femoral head and its congruency with the acetabulum, the emphasis in managing patients with Legg-Calvé-Perthes disease is careful containment of the head so that it heals properly. In many cases, this can be accomplished nonsurgically. However, if conservative treatment fails to achieve this goal, surgical intervention should be considered.

It is also important to keep in mind that the shape of the femoral head often continues to evolve from early reossification to skeletal maturity.[121] It may become more spherical or it may progressively flatten during this period. Thus, patients need to be continuously assessed until they reach skeletal maturity before any conclusions can be made regarding the effect of the disease or the results of treatment. The long-term prognosis can then be more accurately determined based on the shape of the femoral head at that time.

REFERENCES

1. Aguirre M, Pellise F, Castellote A: Metaphyseal cysts in Legg-Calvé-Perthes disease. J Pediatr Orthop 1992;12:405.

2. Alabi Z, Durosinmi M: Legg-Calvé-Perthes disease associated with chronic myeloid leukaemia in a child: case report. East Afr Med J 1989;66:556.
3. Anderson P Jr, Schantz K, Bollerslev J, et al: Bilateral femoral head dysplasia and osteochondritis: multiple epiphyseal dysplasia tarda, spondylo-epiphyseal dysplasia tarda, and bilateral Legg-Perthes disease. Acta Radiol 1988;29:705.
4. Apley AG, Wientroub S: The sagging rope sign in Perthes' disease and allied disorders. J Bone Joint Surg 1981;63-B:43.
5. Arie E, Johnson F, Harrison MH, et al: Femoral head shape in Perthes' disease: is the contralateral hip abnormal? Clin Orthop 1986;209:77.
6. Arruda VR, Belangero WD, Ozelor MC, et al: Inherited risk factors for thrombophilia among children with Legg-Calvé-Perthes disease. J Pediatr Orthop 1999;19:84.
7. Axer A: Subtrochanteric osteotomy in the treatment of Perthes disease. J Bone Joint Surg 1965;47-B:489.
8. Axer A, Gershuni DH, Hendel D, et al: Indications for femoral osteotomy in Legg-Calvé-Perthes disease. Clin Orthop 1980;150:78.
9. Axer A, Schiller MG: The pathogenesis of the early deformity of the capital femoral epiphysis in Legg-Calvé-Perthes syndrome (LCPS): an arthrographic study. Clin Orthop 1972;84:106.
10. Bailey T Jr, Hall JE: Chiari medial displacement osteotomy. J Pediatr Orthop 1985;5:635.
11. Barnes JM: Premature epiphyseal closure in Perthes' disease. J Bone Joint Surg 1980;62-B:432.
12. Barquet A: Natural history of avascular necrosis following traumatic hip dislocation in childhood: a review of 145 cases. Acta Orthop Scand 1982;53:815.
13. Beneke G, Deutschle N: Frühveränderungen in der proximalen Femurepiphyse nach experimenteller Blutkreislaufstorung. Virchows Arch 1967;344:125.
14. Bennett JT, Mazurek RT, Cash JD: Chiari's osteotomy in the treatment of Perthes disease. J Bone Joint Surg 1991;73:225.
15. Bensahel H, Bok B, Cavailloles F, et al: Bone scintigraphy in Perthes disease. J Pediatr Orthop 1983;3:302.
16. Benson M, Evans D: The pelvic osteotomy of Chiari: an anatomical study of the hazards and misleading radiographic appearances. J Bone Joint Surg 1976;58-B:164.
17. Bentzon PGK: Experimental studies on the pathogenesis of coxa plana. Acta Radiol 1926;6:155.
18. Bergman E: Theoretisches, Klinisches und Experimentelles zur Frage der aseptischen Knochennekrosen. Dtsch Z Chir 1927;206:12.
19. Bobechko W: The Toronto brace for Legg-Perthes disease. J Bone Joint Surg 1976;58-B:115.
20. Bobechko WP, McLaurin CA, Motloch WM: Toronto orthosis for Legg-Perthes disease. Artif Limbs 1968;12:36.
21. Bohr H: Skeletal maturation in Legg-Calvé-Perthes disease. Int Orthop 1979;2:277.
22. Bok B, Cavailloles F, Lonchampt MF, et al: Bone scintigraphy in the diagnosis, prognosis, and follow-up of Legg-Calvé-Perthes disease. Ann Radiol 1983;26:665.
23. Bos CF, Bloem JL, Bloem RM: Sequential magnetic resonance imaging in Perthes' disease. J Bone Joint Surg 1991;73-B:219.
24. Bowen JR, Kumar VP, Joyce JD, et al: Osteochondritis dissecans following Perthes' disease: arthroscopic-operative treatment. Clin Orthop 1986;209:49.
25. Bowen JR, Schmidt T: Osteochondroma of the femoral neck in Perthes disease. J Pediatr Orthop 1983;3:28.
26. Bowen JR, Schreiber FC, Foster BK, et al: Premature femoral neck physeal closure in Perthes' disease. Clin Orthop 1982;171:24.
27. Brotherton BJ, McKibbin B: Perthes' disease treated by prolonged recumbency and femoral head containment: a long-term appraisal. J Bone Joint Surg 1977;59-B:8.
28. Brown I: A study of the capsular shadow in disorders of the hip in children. J Bone Joint Surg 1975;57-B:175.
29. Burch PR, Nevelos AB: Perthes' disease: a new genetic hypothesis. Med Hypotheses 1979;5:513.
30. Burwell RG: Perthes' disease: growth and aetiology. Arch Dis Child 1988;63:1408.
31. Caffey J: The early roentgenographic changes in essential coxa plana: their significance in pathogenesis. Clin Orthop 1968;103:620.
32. Cahuzac JP, Onimus M, Trottmann F, et al: Chiari pelvic osteotomy in Perthes disease. J Pediatr Orthop 1990;10:163.
33. Calvé J: Sur une forme particuliere de pseudo-coxalgie greffée sur des deformations carateristiques de l'extremite duperieure du femur. Rev Chir 1910;30:54.
34. Calvé J: On a particular form of pseudo-coxalgia associated with a characteristic deformity of the upper end of the femur. Clin Orthop 1980;150:4.
35. Cameron JM, Izatt MM: Legg-Calvé-Perthes disease. Scott Med J 1960;5:148.
36. Cannon S, Pozo J, Catterall A: Elevated growth velocity in children with Perthes disease. J Pediatr Orthop 1989;9:285.
37. Catterall A: Legg-Calvé-Perthes Disease. London, Churchill Livingstone, 1982.
38. Catterall A: The natural history of Perthes' disease. J Bone Joint Surg 1971;53-B:37.
39. Catterall A, Pringle J, Byers PD, et al: A review of the morphology of Perthes' disease. J Bone Joint Surg 1982;64-B:269.
40. Cavailloles F, Bok B, Bensahel H: Bone scintigraphy in the diagnosis and follow-up of Perthes' disease. Eur J Nucl Med 1982;7:327.
41. Chiari K: Medial displacement osteotomy of the pelvis. Clin Orthop 1974;98:55.
42. Christensen F, Soballe K, Ejsted R, et al: The Catterall classification of Perthes' disease: an assessment of reliability. J Bone Joint Surg 1986;68-B:614.
43. Chung SM: The arterial supply of the developing proximal end of the human femur. J Bone Joint Surg 1976;58-A:961.
44. Clancy M, Steel HH: The effect of an incomplete intertrochanteric osteotomy on Legg-Calvé-Perthes disease. J Bone Joint Surg 1985; 67-A:213.
45. Clarke NM, Harrison MH: Painful sequelae of coxa plana. J Bone Joint Surg 1983;65-A:13.
46. Coleman S, Kehl D: An evaluation of Perthes' disease: comparison of non-surgical and surgical means. Presented at a meeting of the American Academy of Orthopaedic Surgeons, Las Vegas, 1981.
47. Conway JJ: A scintigraphic classification of Legg-Calvé-Perthes disease. Semin Nucl Med 1993;23:274.
48. Cooperman DR, Emery H, Keller C: Factors relating to hip joint arthritis following three childhood diseases—juvenile rheumatoid arthritis, Perthes disease, and postreduction avascular necrosis in congenital hip dislocation. J Pediatr Orthop 1986;6:706.
49. Craig W, Kramer W, Watanabe R: Etiology and treatment of Legg-Calvé-Perthes syndrome. J Bone Joint Surg 1963;45-A:1325.
50. Crawford AH, Carothers TA: Hip arthrography in the skeletally immature. Clin Orthop 1982;162:54.
51. Crossan JF, Wynne-Davies R, Fulford GE: Bilateral failure of the capital femoral epiphysis: bilateral Perthes disease, multiple epiphyseal dysplasia, pseudoachondroplasia, and spondyloepiphyseal dysplasia congenita and tarda. J Pediatr Orthop 1983;3:297.
52. Crutcher JP, Staheli LT: Combined osteotomy as a salvage procedure for severe Legg-Calvé-Perthes disease. J Pediatr Orthop 1992;12:151.
53. Curtis B, Gunther S, Gossling H, et al: Treatment for Legg-Perthes disease with the Newington ambulation-abduction brace. J Bone Joint Surg 1974;56-A:1135.
54. Daudet M, David M, Aimard P: Lesions of the hip in congenital myxedema in children. Chir Pediatr 1986;27:94.
55. de Camargo FP, de Godoy RMJ, Tovo R: Angiography in Perthes' disease. Clin Orthop 1984;191:216.
56. Dickens DR, Menelaus MB: The assessment of prognosis in Perthes' disease. J Bone Joint Surg 1978;60-B:189.
57. Douglas G, Rang M: The role of trauma in the pathogenesis of the osteochondroses. Clin Orthop 1981;158:28.
58. Drennan J: Orthotic management of Legg-Calvé-Perthes disease. In Leach R, Hoaglund F, Riseborough E (eds): Controversies in Orthopedic Surgery. Philadelphia, WB Saunders Co, 1982.
59. Dunn AW: Coxa plana in monozygotic male twins. J Bone Joint Surg 1960;42-A:178.
60. Eaton G: Long term results of treatment in coxa plana: a follow-up study of eighty-eight patients. J Bone Joint Surg 1967;49-A:1031.
61. Ebong WW: Avascular necrosis of the femoral head associated with haemoglobinopathy. Trop Geogr Med 1977;29:19.
62. Egund N, Wingstrand H: Legg-Calvé-Perthes disease: imaging with MR. Radiology 1991;179:89.
63. Elsig J, Exner G, von Schulthess G, et al: False negative MRI in the early stage of Legg-Calvé-Perthes disease. J Pediatr Orthop 1989;9:231.
64. Englehardt P: Die Spätprognose des Morbus Perthes: welche Faktoren bestimmen das Arthroserisiko. Z Orthop 1985;123:167.
65. Evans IK, Deluca PA, Gage JR: A comparative study of ambulation-

abduction bracing and varus derotation osteotomy in the treatment of severe Legg-Calvé-Perthes disease in children over 6 years of age. J Pediatr Orthop 1988;8:676.
66. Exner G, Schreiber A: Wachstumsretardierung und Aufholwachstum bei Morbus Perthes. Z Orthop 1986;124:192.
67. Eyre-Brook A: Osteochondritis deformans coxae juvenilis or Perthes' disease: the results of treatment by traction in recumbency. Br J Surg 1936;24:166.
68. Farsetti P, Tudisco C, Caterini R, et al: The Herring lateral pillar classification for prognosis in Perthes disease: late results in 49 patients treated conservatively. J Bone Joint Surg 1995;77-B:739.
69. Fasting OJ, Bjerkreim I, Langeland N, et al: Scintigraphic evaluation of the severity of Perthes' disease in the initial stage. Acta Orthop Scand 1980;51:655.
70. Fasting OJ, Langeland N, Bjerkreim I, et al: Bone scintigraphy in early diagnosis of Perthes' disease. Acta Orthop Scand 1978;49:169.
71. Ferguson A Jr: The pathology of Legg-Perthes disease and its comparison with aseptic necrosis. Clin Orthop 1975;106:7.
72. Ferguson A Jr: Pathology and treatment of Legg-Perthes disease. Pediatr Ann 1976;5:113.
73. Fisher RL: An epidemiological study of Legg-Perthes disease. J Bone Joint Surg 1972;54-A:769.
74. Freeman MAR, England JPS: Experimental infarction of the immature canine femoral head. Proc R Soc Med 1969;62:431.
75. Fujikawa K: Comparative vascular anatomy of the hip of the miniature dog and of the normal-size mongrel. Kurume Med J 1991;38:159.
76. Gallagher JM, Weiner DS, Cook AJ: When is arthrography indicated in Legg-Calvé-Perthes disease? J Bone Joint Surg 1983;65-A:900.
77. Gallistl S, Reitinger T, Linhart W, et al: The role of inherited thrombotic disorders in the etiology of Legg-Calvé-Perthes disease. J Pediatr Orthop 1999;19:82.
78. Garceau G, Rapp G, Lidge R: Coxa plana (a surgical approach). J Bone Joint Surg 1973;55-A:1313.
79. Gershuni DH, Axer A, Hendel D: Arthrographic findings in Legg-Calvé-Perthes disease and transient synovitis of the hip. J Bone Joint Surg 1978;60-A:457.
80. Giannestras N: Legg-Perthes disease in twins. J Bone Joint Surg 1954;36-A:149.
81. Gill A: Legg-Perthes disease of the hip: its early roentgenographic manifestations and its cyclical course. J Bone Joint Surg 1940;23-A: 1013.
82. Gill AB: Plastic construction of an acetabulum in congenital dislocation of the hip—the shelf operation. J Bone Joint Surg 1935;17-A:48.
83. Girdany BR, Osman MZ: Longitudinal growth and skeletal maturation in Perthes' disease. Radiol Clin North Am 1968;6:245.
84. Glefand MJ, Ball WS, Oestreich AE, et al: Transient loss of femoral head Tc-99m diphosphonate uptake with prolonged maintenance of femoral head architecture. Clin Nucl Med 1983;8:347.
85. Glick JM: Hip arthroscopy using the lateral approach. Instr Course Lect 1988;37:223.
86. Glueck C, Crawford A, Roy D, et al: Association of antithrombotic factor deficiencies and hypofibrinolysis with Legg-Perthes disease. J Bone Joint Surg 1996;78-A:3.
87. Glueck C, Crawford A, Roy D, et al: Familial protein C and S deficiency and hypofibrinolysis: common causes of Legg-Perthes disease. Presented at the annual meeting of the Pediatric Orthopaedic Society of North America, Memphis, 1994.
88. Glueck C, Glueck H, Greenfield D, et al: Protein C and S deficiency, thrombophilia, and hypofibrinolysis: pathophysiologic causes of Legg-Perthes disease. Pediatr Res 1994;35:383.
89. Glueck CJ, Brandt G, Gruppo R, et al: Resistance to activated protein C and Legg-Perthes disease. Clin Orthop 1997;338:139.
90. Goff C: Legg-Calvé-Perthes Syndrome. Toronto, Charles C Thomas, 1954.
91. Goldman AB, Hallel T, Salvati EM, et al: Osteochondritis dissecans complicating Legg-Perthes disease: a report of four cases. Radiology 1976;121:561.
92. Gower WE, Johnston RC: Legg-Perthes disease: long-term follow-up of thirty-six patients. J Bone Joint Surg 1971;53-A:759.
93. Gray IM, Lowry RB, Renwick DH: Incidence and genetics of Legg-Perthes disease (osteochondritis deformans) in British Columbia: evidence of polygenic determination. J Med Genet 1972;9:197.
94. Green NE, Griffin PP: Intra-osseous venous pressure in Legg-Perthes disease. J Bone Joint Surg 1982;64-A:666.
95. Grim J, Haist J, Higer H: Diagnosis of Perthes disease using magnetic resonance tomography. Z Orthop 1991;129:151.
96. Grossbard GD: Hip pain during adolescence after Perthes' disease. J Bone Joint Surg 1981;63-B:572.
97. Hall A, Barker D, Lawton D: The social origins of Perthes disease of the hip. Paediatr Perinat Epidemiol 1990;4:64.
98. Hall A, Barker DJ: Perthes disease in Yorkshire. J Bone Joint Surg 1989;71-B:229.
99. Hall A, Margetts B, Barker D, et al: Low blood manganese levels in Liverpool children with Perthes disease. Paediatr Perinat Epidemiol 1989;3:131.
100. Hall AJ, Barker DJ, Dangerfield PH, et al: Perthes' disease of the hip in Liverpool. Br Med J 1983;287:1757.
101. Hall AJ, Barker DJ, Dangerfield PH, et al: Small feet and Perthes' disease: a survey in Liverpool. J Bone Joint Surg 1988;70-B:611.
102. Hall DJ: Genetic aspects of Perthes' disease: a critical review. Clin Orthop 1986;209:100.
103. Hall DJ, Harrison MH, Burwell RG: Congenital abnormalities and Perthes' disease: clinical evidence that children with Perthes' disease may have a major congenital defect. J Bone Joint Surg 1979;61-B:18.
104. Hallel T, Salvati EA: Osteochondritis dissecans following Legg-Calvé-Perthes disease: report of three cases. J Bone Joint Surg 1976;58-A:708.
105. Hardcastle PH, Ross R, Hamalainen M, et al: Catterall grouping of Perthes' disease: an assessment of observer error and prognosis using the Catterall classification. J Bone Joint Surg 1980;62-B:428.
106. Harper PS, Brotherton BJ, Cochlin D: Genetic risks in Perthes' disease. Clin Genet 1976;10:178.
107. Harris WH: Etiology of osteoarthritis of the hip. Clin Orthop 1986; 213:20.
108. Harrison MH, Blakemore ME: A study of the "normal" hip in children with unilateral Perthes' disease. J Bone Joint Surg 1980;62-B:31.
109. Harrison MH, Menon MP: Legg-Calvé-Perthes disease: the value of roentgenographic measurement in clinical practice with special reference to the broomstick plaster method. J Bone Joint Surg 1966;48-A:1301.
110. Harrison MH, Turner MH, Jacobs P: Skeletal immaturity in Perthes' disease. J Bone Joint Surg 1976;58-B:37.
111. Harrison MH, Turner MH, Nicholson FJ: Coxa plana: results of a new form of splinting. J Bone Joint Surg 1969;51-A:1057.
112. Harrison MH, Turner MH, Smith DN: Perthes' disease: treatment with the Birmingham splint. J Bone Joint Surg 1982;64-B:3.
113. Haueisen DC, Weiner DS, Weiner SD: The characterization of transient synovitis of the hip in children. J Pediatr Orthop 1986;6:11.
114. Heikkinen E, Lanning P, Suramo I, et al: The venous drainage of the femoral neck as a prognostic sign in Perthes' disease. Acta Orthop Scand 1980;51:501.
115. Heikkinen E, Puranen J: Evaluation of femoral osteotomy in the treatment of Legg-Calvé-Perthes disease. Clin Orthop 1980;150:60.
116. Helbo S: Morbus Calvé-Perthes. Odense, Denmark, 1953.
117. Henderson R, Renner J, Sturdivant M, et al: Evaluation of magnetic resonance imaging in Legg-Calvé-Perthes disease: a prospective, blinded study. J Pediatr Orthop 1990;10:289.
118. Herring JA: Legg-Calvé-Perthes disease: a review of current knowledge. Instr Course Lect 1989;38:309.
119. Herring JA, Lundeen MA, Wenger DR: Minimal Perthes' disease. J Bone Joint Surg 1980;62-B:25.
120. Herring JA, Neustadt JB, Williams JJ, et al: The lateral pillar classification of Legg-Calvé-Perthes disease. J Pediatr Orthop 1992;12:143.
121. Herring JA, Williams JJ, Neustadt JN, et al: Evolution of femoral head deformity during the healing phase of Legg-Calvé-Perthes disease. J Pediatr Orthop 1993;13:41.
122. Hoffinger SA, Henderson RC, Renner JB, et al: Magnetic resonance evaluation of metaphyseal changes in Legg-Calvé-Perthes disease. J Pediatr Orthop 1993;13:602.
123. Hoffinger SA, Rab GT, Salmon PB: Metaphyseal cysts in Legg-Calvé-Perthes disease. J Pediatr Orthop 1991;11:301.
124. Hoikka V, Lindholm TS, Poussa M: Intertrochanteric varus osteotomy in Legg-Calvé-Perthes disease: a report of 112 hips. J Pediatr Orthop 1986;6:600.
125. Houben J, Godart S, Abramovic J, et al: Vascular disorders in irritable hip disclosed by dynamic scintigraphy with radioactive colloids. Chir Pediatr 1982;23:309.
126. Howell CJ, Wynne-Davies R: The tricho-rhino-phalangeal syndrome: a report of 14 cases in 7 kindreds. J Bone Joint Surg 1986;68-B:311.
127. Ikegawa S, Nagano A, Nakamura K: A case of multiple epiphyseal

dysplasia complicated by unilateral Perthes' disease. Acta Orthop Scand 1991;62:606.
128. Inglis A: Genetic implications in coxa plana. J Bone Joint Surg 1960; 42-A:711.
129. Ingman AM, Paterson DC, Sutherland AD: A comparison between innominate osteotomy and hip spica in the treatment of Legg-Perthes' disease. Clin Orthop 1982;163:141.
130. Inoue A, Freeman MA, Vernon-Roberts B, et al: The pathogenesis of Perthes' disease. J Bone Joint Surg 1976;58-B:453.
131. Ippolito E: Legg-Calvé-Perthes (LCP) disease in the light of recent findings. Ital J Orthop Traumatol 1982;8:77.
132. Ippolito E, Tudisco C, Farsetti P: Long-term prognosis of Legg-Calvé-Perthes disease developing during adolescence. J Pediatr Orthop 1985; 5:652.
133. Ismail AM, Macnicof MF: Prognosis in Perthes disease. J Bone Joint Surg 1998;80-B:310.
134. Iwasaki K, Suzuki R, Okazaki T, et al: The haemodynamics of Perthes' disease: an intraosseous venographic study combined with measurement of the intramedullary pressure. Int Orthop 1982;6:141.
135. Jensen OM, Lauritzen J: Legg-Calvé-Perthes disease: morphological studies in two cases examined at necropsy. J Bone Joint Surg 1976; 58-B:332.
136. Jonsäter S: Coxa plana: a histo-pathologic and arthrographic study. Acta Orthop Scand 1953;12(suppl):1.
137. Joseph B: Morphological changes in the acetabulum in Perthes' disease. J Bone Joint Surg 1989;71-B:756.
138. Kahle WK: Association of antithrombotic factor deficiencies and hypofibrinolysis with Legg-Perthes disease. J Bone Joint Surg 1997;79-A: 1114.
139. Kalamchi A: A modified Salter osteotomy. J Bone Joint Surg 1982; 64-A:183.
140. Kallio P, Ryoppy S: Hyperpressure in juvenile hip disease. Acta Orthop Scand 1985;56:211.
141. Kallio P, Ryoppy S, Kunnamo I: Transient synovitis and Perthes' disease: is there an aetiological connection? J Bone Joint Surg 1986; 68-B:808.
142. Kamegaya M: Comparative study of Perthes' disease treated by various ambulatory orthoses. Nippon Seikeigeka Gakkai Zasshi 1987;61:917.
143. Kamegaya M, Shinada Y, Moriya H, et al: Acetabular remodeling in Perthes' disease after primary healing. J Pediatr Orthop 1992;12:308.
144. Kamhi E, MacEwen GD: Osteochondritis dissecans in Legg-Calvé-Perthes disease. J Bone Joint Surg 1975;57-A:506.
145. Kaniklides C: Diagnostic radiology in Legg-Calvé-Perthes disease. Acta Radiol 1996;406(suppl):1.
146. Karpinski MR, Newton G, Henry AP: The results and morbidity of varus osteotomy for Perthes' disease. Clin Orthop 1986;209:30.
147. Katz JF: Osteochondroma of the neck of the femur in Legg-Calvé-Perthes disease: report of two cases. Clin Orthop 1970;68:50.
148. Katz JF, Siffert RS: Capital necrosis, metaphyseal cyst and subluxation in coxa plana. Clin Orthop 1975;106:75.
149. Kendig RJ, Evans GA: Biologic osteotomy in Perthes disease. J Pediatr Orthop 1986;6:278.
150. Keret D, Bassett G: Avascular necrosis of the capital femoral epiphysis in metachondromatosis. J Pediatr Orthop 1990;10:658.
151. Keret D, Harrison MH, Clarke NM, et al: Coxa plana: the fate of the physis. J Bone Joint Surg 1984;66-A:870.
152. Killian JT, Niemann KM: Preoperative skeletal traction in Legg-Perthes disease. South Med J 1985;78:928.
153. Kitsugi T, Kasahara Y, Seto Y, et al: Normal somatomedin-C activity measured by radioimmunoassay in Perthes' disease. Clin Orthop 1989; 244:217.
154. Kleinman RG, Bleck EE: Increased blood viscosity in patients with Legg-Perthes disease: a preliminary report. J Pediatr Orthop 1981; 1:131.
155. Klisic P, Bauer R, Bensahel H, et al: Chiari's pelvic osteotomy in the treatment of Legg-Calvé-Perthes disease. Bull Hosp Jt Dis Orthop Inst 1985;45:111.
156. Kristmundsdottir F, Burwell RG, Hall DJ, et al: A longitudinal study of carpal bone development in Perthes' disease: its significance for both radiologic standstill and bilateral disease. Clin Orthop 1986;209:115.
157. Kristmundsdottir F, Burwell RG, Harrison MH: Delayed skeletal maturation in Perthes' disease. Acta Orthop Scand 1987;58:277.
158. Kruse RW, Guille JT, Bowen JR: Shelf arthroplasty in patients who have Legg-Calvé-Perthes disease: a study of long-term results. J Bone Joint Surg 1991;73-A:1338.
159. Lack W, Feldner-Busztin H, Ritschl P, et al: The results of surgical treatment of Perthes' disease. J Pediatr Orthop 1989;9:197.
160. Lahdes-Vasama T, Lamminen A, Merikanto J, et al: The value of MRI in early Perthes' disease: an MRI study with a 2-year follow-up. Pediatr Radiol 1997;27:517.
161. Landin LA, Danielsson LG, Wattsgard C: Transient synovitis of the hip: its incidence, epidemiology and relation to Perthes' disease. J Bone Joint Surg 1987;69-B:238.
162. Langenskiöld A: Changes in the capital growth plate and the proximal femoral metaphysis in Legg-Calvé-Perthes disease. Clin Orthop 1980; 150:110.
163. Langenskiöld A, Sarpio O, Michelsson JE: Experimental dislocation of the hip in the rabbit. J Bone Joint Surg 1962;44-B:209.
164. Laurent LE, Poussa M: Intertrochanteric varus osteotomy in the treatment of Perthes' disease. Clin Orthop 1980;150:73.
165. Lee DY, Choi IH, Lee CK, et al: Assessment of complex hip deformity using three-dimensional CT images. J Pediatr Orthop 1991;11:13.
166. Legg A: End results of coxa plana. J Bone Joint Surg 1927;9-A:26.
167. Legg A: An obscure affection of the hip joint. Boston Med Surg J 1910;162:202.
168. Legg A: Osteochondral trophopathy of the hip joint. Surg Gynecol Obstet 1916;22:307.
169. Leitch JM, Paterson DC, Foster BK: Growth disturbance in Legg-Calvé-Perthes disease and the consequences of surgical treatment. Clin Orthop 1991;262:178.
170. Lemoine A: Vascular changes after interference with the blood flow of the head of the rabbit. J Bone Joint Surg 1957;39-B:763.
171. Liu SL, Ho TC: The role of venous hypertension in the pathogenesis of Legg-Perthes disease: a clinical and experimental study. J Bone Joint Surg 1991;73-A:194.
172. Lloyd-Roberts GC: The management of Perthes' disease. J Bone Joint Surg 1982;64-B:1.
173. Lloyd-Roberts GC, Catterall A, Salamon PB: A controlled study of the indications for and the results of femoral osteotomy in Perthes' disease. J Bone Joint Surg 1976;58-B:31.
174. Loder RT, Schwartz EM, Hensinger RN: Behavioral characteristics of children with Legg-Calvé-Perthes disease. J Pediatr Orthop 1993; 13:598.
175. Malefut de Wal M, Hoogland T, Nielsen H: Chiari osteotomy in the treatment of congenital dislocation and subluxation of the hip. J Bone Joint Surg 1982;64-A:996.
176. Mallet J, Rigault P, Padovani J, et al: Transient synovitis of the hip in childhood: "observation hip." Rev Chir Orthop 1981;67:791.
177. Mandell G, MacKenzie W, Scott C, et al: Identification of avascular necrosis in the dysplastic proximal femoral epiphysis. Skeletal Radiol 1989;18:273.
178. Mandell GA, Harcke HT, Kumar SJ: Avascular necrosis and related conditions. Top Magn Reson Imaging 1991;4:31.
179. Marklund T, Tillberg B: Coxa plana: a radiological comparison of the rate of healing with conservative measures and after osteotomy. J Bone Joint Surg 1976;58-B:25.
180. McAndrew MP, Weinstein SL: A long-term follow-up of Legg-Calvé-Perthes disease. J Bone Joint Surg 1984;66-A:860.
181. McDougall PA, Hresko MT, Kasser JR, et al: Prospective re-evaluation of the association between thrombotic diathesis and Legg-Calvé-Perthes disease. Presented at the Pediatric Orthopaedic Society of North America 1998 Annual Meeting, Cleveland, May 1998.
182. McElwain JP, Regan BF, Dowling F, et al: Derotation varus osteotomy in Perthes disease. J Pediatr Orthop 1985;5:195.
183. Menelaus MB: Lessons learned in the management of Legg-Calvé-Perthes disease. Clin Orthop 1986;209:41.
184. Meyer J: Dysplasia epiphysealis capitis femoris: a clinical radiological syndrome and its relationship to Legg-Calvé-Perthes disease. Acta Orthop Scand 1964;34:183.
185. Mickelson MR, McCurnin DM, Awbrey BJ, et al: Legg-Calvé-Perthes disease in dogs: a comparison to human Legg-Calvé-Perthes disease. Clin Orthop 1981;157:287.
186. Milgram JW: Synovial osteochondromatosis in association with Legg-Calvé-Perthes disease. Clin Orthop 1979;145:179.
187. Moller P: The clinical observations after healing of Calvé-Perthes disease compared with the final deformities left by that disease and the bearing of those final deformities on ultimate prognosis. Acta Radiol 1926;5:1.
188. Molloy MK, MacMahon B: Birth weight and Legg-Perthes disease. J Bone Joint Surg 1967;49-A:498.

189. Moreno P, Cahuzac J, Pasquie M: Topographic and developmental study of primary osteochondritis of the hip by x-ray computed tomography. Rev Chir Orthop 1986;72:173.
190. Mose K: Methods of measuring in Legg-Calvé-Perthes disease with special regard to the prognosis. Clin Orthop 1980;150:103.
191. Motokawa S: The effect of serum factors on skeletal growth in Perthes disease. Nippon Seikeigeka Gakkai Zasshi 1990;64:790.
192. Mukamel M, Litmanovitch M, Yosipovich Z, et al: Legg-Calvé-Perthes disease following transient synovitis: how often? Clin Pediatr 1985; 24:629.
193. Mukherjee K, Fabry G: Evaluation of the prognostic indices in Legg-Calvé-Perthes disease: statistical analysis of 116 hips. J Pediatr Orthop 1990;10:153.
194. Nagura S: Das Wesen und die Entstehung der Osteochondritis dissecans Konigs (bzw der Perthes, Köhler II- und ähnlichen Krankeiten und Veränderungen an wachsenden Knochenden). Zentralbl Chir 1937;64:2049.
195. Naito M, Schoenecker PL, Owen JH, et al: Acute effect of traction, compression, and hip joint tamponade on blood flow of the femoral head: an experimental model. J Orthop Res 1992;10:800.
196. Naumann T, Kollmannsberger A, Fischer M, et al: Ultrasonographic evaluation of Legg-Calvé-Perthes disease based on sonoanatomic criteria and the application of new measuring techniques. Eur J Radiol 1992;15:101.
197. Neidel J, Boddenberg B, Zander D, et al: Thyroid function in Legg-Calvé-Perthes: cross-sectional and longitudinal study. J Pediatr Orthop 1993;13:592.
198. Neidel J, Schonau E, Zander D, et al: Normal plasma levels of IGF binding protein in Perthes' disease: follow-up of previous report. Acta Orthop Scand 1993;64:540.
199. Neidel J, Zander D, Hackenbroch MH: No physiologic age-related increase of circulating somatomedin-C during early stage of Perthes' disease: a longitudinal study in 21 boys. Arch Orthop Trauma Surg 1992;111:171.
200. Nevelos AB: Bilateral Perthes' disease. Acta Orthop Scand 1980;51:649.
201. O'Hara JP, Dommisse GF: Extraosseous blood supply to the neonatal femoral head. Clin Orthop 1983;174:293.
202. Olney BW, Asher MA: Combined innominate and femoral osteotomy for the treatment of severe Legg-Calvé-Perthes disease. J Pediatr Orthop 1985;5:645.
203. Orzincolo C, Castaldi G, Scutellari PN, et al: Aseptic necrosis of femoral head complicating thalassemia. Skeletal Radiol 1986;15:541.
204. Oshima M, Yoshihasi Y, Ito K, et al: Initial stage of Legg-Calvé-Perthes disease: comparison of three-phase bone scintigraphy and SPECT with MR imaging. Eur J Radiol 1992;15:107.
205. Osterman K, Lindholm TS: Osteochondritis dissecans following Perthes' disease. Clin Orthop 1980;152:247.
206. O'Sullivan M, O'Rourke SK, MacAuley P: Legg-Calvé-Perthes disease in a family: genetic or environmental? Clin Orthop 1985;199:179.
207. Ozonoff MB, Ziter F Jr: The femoral head notch. Skeletal Radiol 1987;16:19.
208. Paterson DE, Harper G, Weston HJ, et al: Maroteaux-Lamy syndrome, mild form—MPS vi b. Br J Radiol 1982;55:805.
209. Pay NT, Singer WS, Bartel E: Hip pain in three children accompanied by transient abnormal findings on MR images. Radiology 1989; 173:881.
210. Perpich M, McBeath A, Kruse D: Long-term follow-up of Perthes disease treated with spica casts. J Pediatr Orthop 1983;3:160.
211. Perthes G: Über arthritis deformans juveniles. Dtsch Z Chir 1910; 107:111.
212. Perthes G: Über osteochondritis deformans juveniles. Arch Klin Chir 1913;101:779.
213. Peterson H, Wingstrand H, Thambert C: Legg-Calvé-Perthes disease in hemophilia: incidence and etiologic considerations. J Pediatr Orthop 1990;10:28.
214. Petrie JG, Bitenc I: The abduction weight-bearing treatment in Legg-Perthes' disease. J Bone Joint Surg 1971;53-B:54.
215. Pfeiffer R, Bauer H, Petersen C: The Schwartz-Jampel syndrome. Helv Paediatr Acta 1977;32:251.
216. Phemister DB: Perthes' disease. Surg Gynecol Obstet 1921;33:87.
217. Phemister DB: Repair of bone in the presence of aseptic necrosis resulting from fractures, transplantations and vascular obstruction. J Bone Joint Surg 1930;12:769.
218. Pike MM: Legg-Calvé-Perthes disease: a method of conservative treatment. J Bone Joint Surg 1950;32-A:663.
219. Pinto M, Peterson H, Berquist T: Magnetic resonance imaging in the early diagnosis of Legg-Calvé-Perthes disease. J Pediatr Orthop 1989; 9:19.
220. Ponseti I: Legg-Perthes disease: observations on pathological changes in two cases. J Bone Joint Surg 1956;38-A:739.
221. Ponseti I, Cotton R: Legg-Calvé-Perthes disease: pathogenesis and evolution. Failure of treatment with L-triiodothyronine. J Bone Joint Surg 1961;43-A:261.
222. Ponseti IV, Maynard JA, Weinstein SL, et al: Legg-Calvé-Perthes disease: histochemical and ultrastructural observations of the epiphyseal cartilage and physis. J Bone Joint Surg 1983;65-A:797.
223. Poussa M, Hoikka V, Yrjonen T, et al: Early signs of poor prognosis in Legg-Perthes-Calvé disease treated by femoral varus osteotomy. Rev Chir Orthop Reparatrice Appar Mot 1991;77:478.
224. Purvis JM, Dimon JD, Meehan PL, et al: Preliminary experience with the Scottish Rite Hospital abduction orthosis for Legg-Perthes disease. Clin Orthop 1980;150:49.
225. Quain S, Catterall A: Hinge abduction of the hip: diagnosis and treatment. J Bone Joint Surg 1986;68-B:61.
226. Rab GT: Determination of femoral head containment during gait. Biomater Med Devices Artif Organs 1983;11:31.
227. Rab GT, Wyatt M, Sutherland DH, et al: A technique for determining femoral head containment during gait. J Pediatr Orthop 1985;5:8.
228. Rand C, Pearson TC, Heatley FW: Avascular necrosis of the femoral head in sickle cell syndrome: a report of 5 cases. Acta Haematol 1987;78:186.
229. Ranner G, Ebner F, Fotter R, et al: Magnetic resonance imaging in children with acute hip pain. Pediatr Radiol 1989;20:67.
230. Ratliff AH: Perthes' disease: a study of thirty-four hips observed for thirty years. J Bone Joint Surg 1967;49-B:102.
231. Rayner PH, Schwalbe SL, Hall DJ: An assessment of endocrine function in boys with Perthes' disease. Clin Orthop 1986;209:124.
232. Reinker KA: Early diagnosis and treatment of hinge abduction in Legg-Perthes disease. J Pediatr Orthop 1996;16:3.
233. Renowden S, Fitzgerald EJ, Kemp AM: Non-Hodgkin's lymphoma of bone causing avascular necrosis of the femoral head. Postgrad Med J 1988;64:68.
234. Richards BS, Coleman SS: Subluxation of the femoral head in coxa plana. J Bone Joint Surg 1987;69:1312.
235. Riedel G: Pathologic anatomy of osteochondritis deformans coxae juvenilis. Zentralbl Chir 1922;49:1447.
236. Ritterbusch JF, Shantharam SS, Gelinas C: Comparison of lateral pillar classification and Catterall classification of Legg-Calvé-Perthes disease. J Pediatr Orthop 1993;13:200.
237. Roberts J, Meehan P, Counts G, et al: Ambulatory abduction brace for Legg-Perthes disease. Presented at the First International Symposium on Legg-Calvé-Perthes Syndrome, Los Angeles, 1977.
238. Rokkanen P: Role of surgical intervention of the hip joint in the aetiology of aseptic necrosis of the femoral head. Acta Orthop Scand 1962;58:1.
239. Roy DR: Perthes'-like changes caused by acquired hypothyroidism. Orthopedics 1991;14:901.
240. Rutskii A, Kovalenko I, Kriuchok V, et al: Dynamic angioscintigraphy and static scintigraphy of the hip joint in the complex diagnosis of Legg-Perthes disease. Orthop Traumatol Protez 1989;10:35.
241. Saito S, Takaoka K, Ono K, et al: Residual deformities related to arthrotic change after Perthes' disease: a long-term follow-up of fifty-one cases. Arch Orthop Trauma Surg 1985;104:7.
242. Salter R: Experimental and clinical aspects of Perthes' disease. J Bone Joint Surg 1966;48-B:393.
243. Salter R, Bell M: The pathogenesis of deformity in Legg-Perthes disease: an experimental investigation. J Bone Joint Surg 1968;50-B:436.
244. Salter R, Willis R, Malcolm B: The treatment of residual subluxation and coxa vara by combined innominate osteotomy and abduction femoral osteotomy. Ann R Coll Phys Surg Canada 1978;11:63.
245. Salter RB: Legg-Perthes disease: the scientific basis for the methods of treatment and their indications. Clin Orthop 1980;150:8.
246. Salter RB: The present status of surgical treatment for Legg-Perthes disease. J Bone Joint Surg 1984;66-A:961.
247. Salter RB, Thompson GH: Legg-Calvé-Perthes disease: the prognostic significance of the subchondral fracture and a two-group classification of the femoral head involvement. J Bone Joint Surg 1984;66-A:479.
248. Sanchis M, Zahir A, Freeman MA: The experimental simulation of Perthes disease by consecutive interruptions of the blood supply to

the capital femoral epiphysis in the puppy. J Bone Joint Surg 1973; 55-A:335.
249. Schindler A, Lechevallier J, Rao N, et al: Diagnostic and therapeutic arthroscopy of the hip in children and adolescents: evaluation of results. J Pediatr Orthop 1995;15:317.
250. Schwarz E: A typical disease of the upper femoral epiphysis. Clin Orthop 1986;209:5.
251. Scoles PV, Yoon YS, Makley JT, et al: Nuclear magnetic resonance imaging in Legg-Calvé-Perthes disease. J Bone Joint Surg 1984;66-A: 1357.
252. Sebag G, Ducou Le Pointe H, Klein I, et al: Dynamic gadolinium-enhanced subtraction MR imaging: a simple technique for the early diagnosis of Legg-Calvé-Perthes disease. Preliminary report. Pediatr Radiol 1997;27:216.
253. Serlo W, Heikkinen E, Puranen J: Preoperative Russell traction in Legg-Calvé-Perthes disease. J Pediatr Orthop 1987;7:288.
254. Sharwood PF: The irritable hip syndrome in children: a long-term follow-up. Acta Orthop Scand 1981;52:633.
255. Snyder C: A sling for use in Legg-Perthes disease. J Bone Joint Surg 1947;29-A:524.
256. Snyder CR: Legg-Perthes disease in the young hip: does it necessarily do well? J Bone Joint Surg 1975;57:751.
257. Solomon L: Patterns of osteoarthritis of the hip. J Bone Joint Surg 1976;58-B:176.
258. Sponseller P, Desai S, Millis M: Abnormalities of proximal femoral growth after severe Perthes' disease. J Bone Joint Surg 1989;71-B:610.
259. Sponseller PD, Desai SS, Millis MB: Comparison of femoral and innominate osteotomies for the treatment of Legg-Calvé-Perthes disease. J Bone Joint Surg 1988;70-A:1131.
260. Staheli L, Chew D: Slotted acetabular augmentation in childhood and adolescence. J Pediatr Orthop 1992;12:569.
261. Stearns ZR, Lacassie Y, MacEwen GB: Perthes-like disease and the tricho-rhino-phalangeal syndromes: the first black patient. Orthopedics 1990;13:468.
262. Stephens FE, Kerby JP: Hereditary Legg-Calvé-Perthes disease. J Hered 1946;37:153.
263. Stulberg SD, Cooperman DR, Wallensten R: The natural history of Legg-Calvé-Perthes disease. J Bone Joint Surg 1981;63-A:1095.
264. Sundt H: Malum coxae: Calvé-Legg-Perthes. Zentralbl Chir 1920; 22:538.
265. Suramo I, Vuoria P: Cineangiographic study of the venous drainage of the femoral neck in children. Ann Clin Res 1976;8:8.
266. Sutherland AD, Savage JP, Paterson DC, et al: The nuclide bone-scan in the diagnosis and management of Perthes' disease. J Bone Joint Surg 1980;62-B:300.
267. Suzuki S, Awaya G, Okada Y, et al: Examination by ultrasound of Legg-Calvé-Perthes disease. Clin Orthop 1987;220:130.
268. Synder M, Dorman T: Remarks on classification of Perthes disease. Chir Narzadow Ruchu Ortop Pol 1997;62:115.
269. Tachdjian M, Jouett L: Trilateral socket hip abduction orthosis for treatment of Legg-Perthes disease. J Bone Joint Surg 1968;50-A:1271.
270. Tanaka H, Tamura K, Takano K, et al: Serum somatomedin A in Perthes' disease. Acta Orthop Scand 1984;55:135.
271. Tao S: Hemodynamic changes in proximal femur of patients with femoral head necrosis. Chung Hua Wai Ko Tsa Chih 1991;19:452.
272. Theissen P, Rutt J, Linden A, et al: The early diagnosis of Perthes disease: the value of bone scintigraphy and magnetic resonance imaging in comparison with x-ray findings. Nuklearmedizin 1991;30:265.
273. Theron J: Angiography in Legg-Calvé-Perthes disease. Radiology 1980;135:81.
274. Tsao AK, Dias LS, Conway JJ, et al: The prognostic value and significance of serial bone scintigraphy in Legg-Calvé-Perthes disease. J Pediatr Orthop 1997;17:230.
275. Ura Y, Hara T, Mori Y, et al: Development of Perthes' disease in 3-year-old boy with idiopathic thrombocytopenic purpura and anti-phospholipid antibodies. Pediatr Hematol Oncol 1992;9:77.
276. Van Dam BE, Crider RJ, Noyes JD, et al: Determination of the Catterall classification in Legg-Calvé-Perthes disease. J Bone Joint Surg 1981; 63-A:906.
277. Vegter J: The influence of joint posture on intra-articular pressure: a study of transient synovitis and Perthes' disease. J Bone Joint Surg 1987;69-B:71.
278. Verbruggen LA, Van Laere C, Lamoureux J, et al: Tricho-rhino-phalangeal syndrome type I in a Belgian family. Clin Rheumatol 1987;6:185.
279. Waldenström H: The definite form of coxa plana. Acta Radiol 1922; 1:384.
280. Waldenström H: The first stages of coxa plana. J Bone Joint Surg 1938;20-A:559.
281. Waldenström H: Der obere tuberkulose Collumherd. Z Orthop Chir 1909;24:487.
282. Waldenström H: On coxa plana. Acta Chir Scand 1923;55:577.
283. Wansborough RM, Carrie AW, Walker NG, et al: Coxa plana, its genetic aspects and results of treatment with the long Taylor walking caliper. J Bone Joint Surg 1959;41-A:135.
284. Weiner DS, O'Dell HW: Legg-Calvé-Perthes disease: observations on skeletal maturation. Clin Orthop 1970;68:44.
285. Weiner SD, Weiner DS, Riley PM: Pitfalls in treatment of Legg-Calvé-Perthes disease using proximal femoral varus osteotomy. J Pediatr Orthop 1991;11:20.
286. Weinstein S: Legg-Calvé-Perthes disease. In Morrissy R (ed): Pediatric Orthopaedics, vol 2, p 851. Philadelphia, JB Lippincott Co, 1990.
287. Weisz I, Bialik V, Adler O, et al: Some observations on the use of computerized tomography in Legg-Calvé-Perthes disease. Z Kinderchir 1988;43:402.
288. Wenger DR: Selective surgical containment for Legg-Perthes disease: recognition and management of complications. J Pediatr Orthop 1981; 1:153.
289. Willett K, Hudson I, Catterall A: Lateral shelf acetabuloplasty: an operation for older children with Perthes' disease. J Pediatr Orthop 1992;12:563.
290. Wingstrand H, Bauer GC, Brismar J, et al: Transient ischaemia of the proximal femoral epiphysis in the child: interpretation of bone scintimetry for diagnosis in hip pain. Acta Orthop Scand 1985;56:197.
291. Wood J, Klassen R, Peterson H: Osteochondritis dissecans of the femoral head in children and adolescents: a report of 17 cases. J Pediatr Orthop 1995;15:313.
292. Wynne-Davies R, Gormley J: The aetiology of Perthes' disease: genetic, epidemiological and growth factors in 310 Edinburgh and Glasgow patients. J Bone Joint Surg 1978;60-B:6.
293. Yngve DA, Roberts JM: Acetabular hypertrophy in Legg-Calvé-Perthes disease. J Pediatr Orthop 1985;5:416.
294. Zemansky AP: The pathology and pathogenesis of Legg-Calvé-Perthes disease (osteochondritis juvenilis deformans coxae). Am J Surg 1928; 4:169.