Legg-Calvé-Perthes Disease

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Perthes disease is a condition of unknown etiology in which the vascular supply of the developing femoral head is temporarily interrupted, causing necrosis and collapse of the bony femoral head, followed by spontaneous revascularization and healing of the necrotic bone. It was independently described by four different authors (Legg, Calvé, Perthes, and Waldenstrom) in 1909 and 1910. They identified the condition as an affliction of the hip distinct from trauma and infection. The term "coxa plana" was applied to the characteristic flattening of the femoral head seen in Perthes disease. It was not until 10 years later that the actual pathology of avascular necrotic bone was described.

EPIDEMIOLOGY

The condition has its onset in children ranging in age from 2 to 12 years, with the majority of patients presenting between the ages of 4 and 8 years. Boys are affected four times more frequently than girls, and the condition is bilateral in approximately 15% of affected children.

Certain constitutional factors are frequently found in children with Perthes disease. Skeletal age is delayed in nearly 90% of children affected with it. Affected boys have been found to be an average of 1 in shorter and affected girls 3 in shorter in height than their unaffected peers. Birth weight of affected children is frequently lower than average.

There are some racial variations in incidence; it is most important to note the very rare occurrence of Perthes disease in American Indians and blacks.

CLINICAL FEATURES AND PHYSICAL EXAMINATION

The signs and symptoms of Perthes disease are frequently subtle and gradual in onset, always occurring in the absence of constitutional symptoms. Because it is not unusual for the child to have no overt complaints, a delay in diagnosis is the rule rather than the exception (Table 1).

The most frequently observed clinical sign is a limp; this complaint from a child in the appropriate age group should put Perthes disease at the top of the list of differential diagnoses. The limp is usually painless and may be intermittent for several weeks, often present only after exertion.

Some children present with pain as their chief complaint. The pain is usually located in the groin and inner aspect of the thigh, is aggravated by motion, and is relieved by rest. Due to the phenomenon of "referred pain," some children will present with knee pain as their chief complaint. Occasionally, patients may present with a painful limp.

When knee pain is the presenting complaint, examination of the knee reveals no abnormal findings. However, a thorough, careful physical examination will reveal the hip as the site of the disorder, as motion of the affected hip will frequently elicit pain.

Examination of the hip is best performed with the child relaxed in the supine position, legs extended. Gentle rotation of the leg is usually not resisted unless the hip is irritable. The hip may then be flexed, extended, abducted, and rotated in sequence to determine the presence of pain. These maneuvers will also demonstrate a decrease in range of motion compared with the opposite side. The decrease may be quite subtle and is most frequently noted as a decrease in internal rotation and abduction; differences as slight as 10 degrees are common. The irritability, pain, and decreased range of motion are related to a mild synovitis of the hip joint as well as to involuntary muscle spasm which invariably accompanies the inflammation.

If the condition is more advanced, a slight degree of shortening of the affected femur compared with the contralateral side may be noted.

ETIOLOGY

Perthes disease results from an interruption of the vascular supply to the developing femoral head. The cause of this interruption, however, remains totally elusive. Anatomic variation in the distribution of blood vessels has been suggested as a causative factor, but this seems contributory at best. Occlusion of the vessels by an embolic phenomenon has also been suggested as a cause but has not been well demonstrated pathologically. Attempts to produce an animal model of this condition have only been successful if repeated insults to the blood supply to the head of the femur have been made.

Microtrauma remains a popular theoretical cause of Perthes disease because of the high activity level of young children, especially boys, and because of the increased incidence in boys. However, this theory seems inadequate for a number of reasons, including the preponderantly unilateral occurrence and the absence of a documented correlation between level of activity and incidence of the condition.

Nonspecific synovitis of the hip resulting in excessive synovial fluid and internal pressure on the joint capsule is another possible cause of Perthes disease; the tamponade-like effect on the pericapsular vessels could lead to avascular necrosis. However, although synovitis is a feature of the early stage of Perthes disease, it may well be secondary rather than primary. In addition, patients known to have the condition of "toxic synovitis"

EDUCATIONAL OBJECTIVE

9. The pediatrician should be able to diagnose and develop a program of management for a child with a limp and thigh pain, distinguishing among osteomyelitis, septic arthritis, juvenile rheumatoid arthritis, acute rheumatic fever, Legg-Calvé-Perthes disease, slipped capital femoral epiphysis, postinfectious synovitis of the hip, leukemia, arthritis with inflammatory bowel disease, and osteoid osteoma (Topics, 85/86).

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TABLE 1. Clinical Picture

- Painless limp (or)
- Hip pain
- Decreased range of motion

TABLE 2. Stages of Disease

- Initial (necrosis)
- Fragmentation
- Reparative
- Definitive

rarely progress to full-blown Legg-Perthes disease.

PATHOPHYSIOLOGY

Whatever the cause, in Perthes disease the head of the femur is deprived of its vascular supply, resulting in necrosis of bone. Subsequent events center around the gradual resorption and revascularization of the necrotic bone.

The sequence of events may be conveniently thought of in four stages, well described by Jonsaters (Table 2). The *initial stage* is "necrosis," during which time the epiphysis of the femoral head loses its blood supply. Histologically, the bony trabeculae are necrotic (shown by the absence of living osteocytes) and distorted due to the early structural collapse which is taking place. There are no signs of inflammation in the bone itself, although inflammation of the synovium is frequently present. It is important to remember that in the earliest weeks of the disease, radiographs may not demonstrate any abnormalities.

The *second stage* may be referred to as the "fragmentation" stage. This term aptly describes the radiographic findings, in which the epiphysis frequently appears disrupted. Collapse of the bony trabeculae results in loss of height of the femoral head, as seen on the radiograph. This collapse also results in a "condensation" of trabeculae, seen radiographically as areas of increased bone density. Histologically, increased distortion of the bony trabeculae and some early ingrowth of blood vessels are noted.

TABLE 3. Early Radiographic Findings

- Widened joint space
- Smaller epiphysis
- Increased density of epiphysis
- Subchondral fracture
- Widening of femoral neck

The *third stage* of the disease is "reparative." The hallmark here is vascular ingrowth which promotes not only new bone formation on the surface of dead trabeculae but also resorption and repair of the dead bone. This process has been termed "creeping substitution." As new bone is laid down on the surface of dead trabeculae, radiographs show an increase in density of the femoral head. Other areas of new bone formation are also evident as a homogeneous consistency filling in areas of earlier disruption.

The *final stage* of the disorder may be termed "definitive," by which time the bony architecture has been completely restored. During the earlier phases of the disorder, however, the structural integrity of the femoral head is frequently compromised, allowing its collapse and a distortion of its shape (flattening). As vascular repair proceeds, the flattened bone remains, resulting in a permanent deformity of the femoral head and incongruity of the hip joint. Studies of the natural history of Perthes disease indicate that such a hip is destined to premature degenerative arthritis in the fifth and sixth decades of life.

The total time required for this entire sequence of events is 12 to 24 months. Three months may elapse during the initial avascular stage before collapse and fragmentation of the femoral head are demonstrated radiographically. Three to 6 months more may pass before collapse is complete. The revascularization process may take 6 to 18 months before the femoral head is completely reconstituted.

Despite all the vascular changes taking place in the femoral head, the articular cartilage receives the necessary nutritional support, primarily from synovial fluid and, for this reason, remains healthy. It maintains its integrity and a normal rate of growth throughout the disease process.

RADIOGRAPHIC EVALUATION

Radiographs taken during the first several weeks of Perthes disease may not reveal any abnormalities. However, as the disease progresses, standard radiographs of the hip will demonstrate the characteristic changes and establish the diagnosis. Two views are required: anteroposterior and "frog leg" (90 degrees flexion, 45 degrees abduction). If the diagnosis is established (or suspected), follow-up films using both views should be obtained approximately every 3 months to follow the progress of the disorder.

The earliest radiographic changes include a smaller ossific nucleus of the femoral head on the involved side and a wider joint space than the contralateral side (Table 3). These result from a cessation of bony growth due to interrupted blood supply. The cartilaginous portion continues its growth because of its uninterrupted nutritional support from joint fluid. The ossific nucleus (or a portion of it) on the affected side soon appears slightly more dense than that on the opposite side due to the condensation of bone as the femoral head begins to collapse. This appearance of increased density is accentuated by some demineralization of adjacent bone related to disuse.

The *crescent sign* is also an early radiographic finding, seen only on the flexion/abduction view. It results from a subchondral fracture, allowing separation of the joint cartilage from the rest of the femoral head. It is generally believed that the extent of this fracture is determined by, and is a definite indication of, the amount of the femoral head that is necrotic (Fig 1).

Later in the disease process, the ossific nucleus appears fragmented, with areas of new bone being interspersed. The collapse of the femoral head is evident by its loss of vertical height, and there is almost always an associated increase in width of both the femoral head and neck. This radiographic appearance is the origin of the descriptive term *coxa plana* (Fig 2).

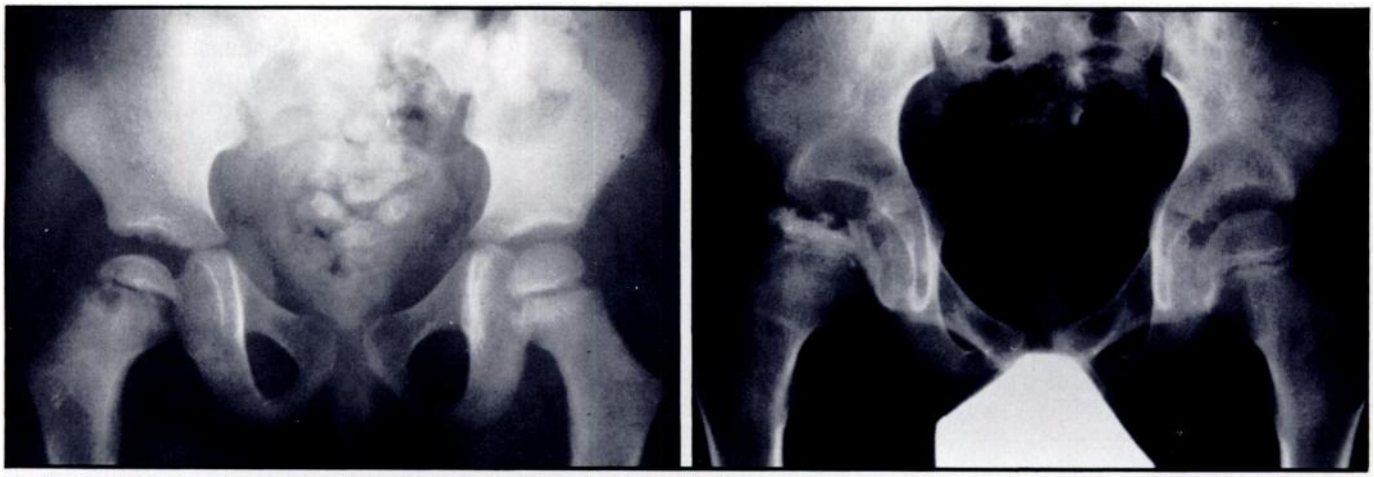

Fig 1. Early Perthes disease of right hip. Note smaller femoral head, wider joint space, and cyst formation in femoral neck. Subchondral fracture line is an early indication of extent of involvement.
Fig 2. Right femoral head has become fragmented, and collapse of vertical height has occurred. Increased density is due to a combination of trabecular collapse and early new bone formation.

The later stages of the disease are characterized radiographically by a return to homogeneous consistency of the femoral head with varying degrees of deformity remaining from the intervening disease process.

Arthrography may be carried out to assess the thickness and the shape of the articular cartilage and may be of use in determining the best useful method of treatment.

Radioactive bone scanning may be of some value in the earliest stages of the disorder to demonstrate avascularity of the femoral head, but special "pin-hole" techniques are required to detect small areas of decreased uptake of the radioisotope. However, because diagnosis this early is unlikely to affect the treatment plan, this study is not routinely recommended.

PROGNOSTIC FACTORS

There are two major clinical factors that influence prognosis (Table 4). The first of these is age at onset, which is very important because children younger than 5 years of age at the time of onset of Perthes disease usually have a much better result. This seems related to the much greater potential for remodeling of the younger hip.

The second major clinical factor influencing prognosis is the range of motion present. Irritability and decreased motion of the hip result from the synovitis sometimes present early in the disease process. Loss of motion seems unrelated to the extent of disease, because some children with total involvement of the femoral head have nearly normal range of motion, whereas others with a small area of involvement of the femoral head have significant loss of motion. This has definite implications for treatment recommendations early in the disease. A hip with persistently limited motion has a much poorer prognosis than a hip that moves freely, regardless of the extent of involvement of the femoral head.

Several radiographic factors are important in gauging prognosis, the most important being the extent of involvement of the femoral head. The classification of Catterall (four groups) is most frequently used to assess this factor. In *group I*, only a small anterior (central) portion of the femoral head is involved, and this is completely surrounded by living bone. Collapse of the femoral head does not occur, and because damage is minor, healing occurs without consequence and the prognosis is excellent. Group I involvement is not common and, in fact, not typical of the usual Perthes syndrome. No treatment is required for patients so affected.

In *Group II*, between 25% and 50% of the femoral head becomes avascular. The radiograph depicted in Fig 3 demonstrates two uninvolved "pillars" of normal bone medial and lateral to the avascular segment. These prevent collapse of the avascular segment, making the prognosis for maintenance of sphericity excellent, frequently without active treatment.

Between 50% and 75% of the femoral head is affected in *group III*. As seen on films taken in the lateral view, the anterolateral portion of the femoral head is completely necrotic and is not surrounded by normal bone. Collapse and extrusion of the femoral head laterally out of the acetabulum occur with regularity. The avascular process extends down to the growth plate, which may also suffer damage. Changes in the femoral neck (metaphysis) are frequently present. Collapse of the femoral head may be quite severe in this group, and its resultant residual deformity may be significant.

In *Group IV*, the entire femoral head becomes necrotic (Fig 4). Collapse occurs early and is often complete. The epiphyseal line is most vulnerable in this group and may be severely damaged, preventing subsequent growth and severely compromising the remodeling potential of the femoral head. The resultant deformity of the upper femur consists of a short femoral neck, a deformed femoral head, and an "overgrown" greater trochanter; the altered anatomy and biomechanics of the hip result in a significant limp (Fig 5).

It may be difficult in the early stages of Perthes disease to determine the precise extent of involvement of the femoral head. Repeating the radi-

TABLE 4. Prognostic Factors

Age at onset
Range of motion
Extent of involvement
Subluxation of femoral head
Metaphyseal cyst
Growth arrest

ograph in 3 months will usually solve this problem; the degree of involvement of the femoral head does not change throughout the remaining course of the disease.

Other radiographic factors help determine a "head at risk" and suggest a poorer prognosis in the absence of treatment. Lateral subluxation of the femoral head may occur as a result of collapse during the avascular process. The epiphysis is soft or "biologically plastic" during this stage, and unprotected positioning and weight-bearing will promote flattening and extrusion of the femoral head from the acetabulum. Calcifications appearing lateral to the growth plate early in the disease predict this occurrence. Cyst formation in the metaphysis, especially in the lateral portion, indicates a greater risk of a poor outcome (Fig 6).

Involvement of the growth plate is the final radiographic prognostic factor and one of great importance. In the more extensively involved hips, the growth plate is exposed to the entire process of avascularity and collapse, and this may result in a premature closure of the growth plate. Complete closure results in an extremely shortened femoral neck and greater trochanteric overgrowth, leading to a significant deformity of the hip and disruption of the biomechanics of the hip joint. A partial arrest of the growth plate may also result in deformity, severely limiting the remodeling potential of the young hip. Unfortunately, closure of the growth plate cannot be predicted early in the course of the disease, nor is there a successful way to prevent its occurrence (20% of cases) once it is detected (Fig 6).

DIFFERENTIAL DIAGNOSIS

The most important diagnoses to be differentiated clinically from Perthes disease include toxic synovitis, septic arthritis, and osteomyelitis of the femoral neck (Table 5). The most difficult of these to distinguish is toxic synovitis, which is a transient, nonspecific synovitis of the hip sometimes associated with a viral infection and which usually responds favorably to a short period of rest. Laboratory and radiographic study findings are usually normal in this condition.

Patients with septic arthritis and osteomyelitis almost always present with systemic complaints typical of an infectious process. These complaints are accompanied by specific laboratory findings. In doubtful cases, an aspiration of the hip joint to obtain fluid for laboratory analysis is mandatory. If no fluid is obtained, radiopaque dye should be injected through the needle into the joint and a radiograph obtained to ensure that the joint was actually entered.

The major diagnosis to be differentiated radiographically is multiple epiphyseal dysplasia, because its appearance is very similar to that of Perthes disease. This condition should always be suspected in patients with bilateral involvement of the hips. It should be confirmed by radiographs of other joints, especially the wrist, hand, knee, and ankle, which demonstrate other abnormal epiphyses in this condition. Sequential films in patients with multiple epiphyseal dysplasia do not demonstrate the typical radiographic course as described for Perthes disease.

The final condition significant in the differential diagnosis is avascular necrosis of the hip from other known causes, the most common of which is a hemoglobinopathy such as sickle cell disease.

PRINCIPLES OF TREATMENT

Because the course of Perthes disease is self-limited with spontaneous healing, the most important principle of treatment is "containment," to maintain the sphericity of the femoral head during its period of vulnerability in the early stages of the disorder. This concept recognizes the biologic plasticity of the femoral head during the early stages of the disease and suggests the need to position it deep within the acetabulum. This can be accomplished either by maintaining the leg in abduction with an orthosis (orthopedic appliance) or by surgically creating containment by increasing the femoral neck-shaft angle or deepening the acetabulum. Motion of the hip during this phase continuously "molds" the femoral head within the acetabulum to maintain its sphericity.

One of the most important implications of the principle of containment is to maintain a full range of motion. In the early stages of Perthes disease, loss of motion is believed to be due to synovitis and muscle spasm, both of which may be improved by rest and/or anti-inflammatory agents such as aspirin. Traction may be useful, particularly as a way to ensure rest. Therapeutic exercises to obtain and maintain as full a range of motion as possible are important. All other treatment modalities should be postponed until a full range of motion has been restored.

Protection against weight-bearing alone is not effective in the treatment of Perthes disease, because it does not necessarily place the hip in a favorable position for containment.

Patients with minimal or moderate involvement of the femoral head (groups I and II in Catterall's classification) rarely require active treatment so long as range of motion is preserved. Because of the small degree of involvement of the femoral head, the prognosis remains excellent.

Patients with more extensive involvement of the femoral head (Catterall's groups III and IV) are more likely to have decreased range of motion and require treatment; efforts to restore motion are mandatory. This may be accomplished with the use of nighttime traction at home, full-time traction at home, or, in some cases, hospitalization for traction. When the range of motion has been restored, containment will be required.

NONOPERATIVE METHODS OF TREATMENT

Containment may be attained by nonoperative or operative methods. The classic nonoperative method involves the use of a Petrie cast. The patient is placed in two long-leg casts, with the hips in approximately 30 to 40 degrees of abduction and

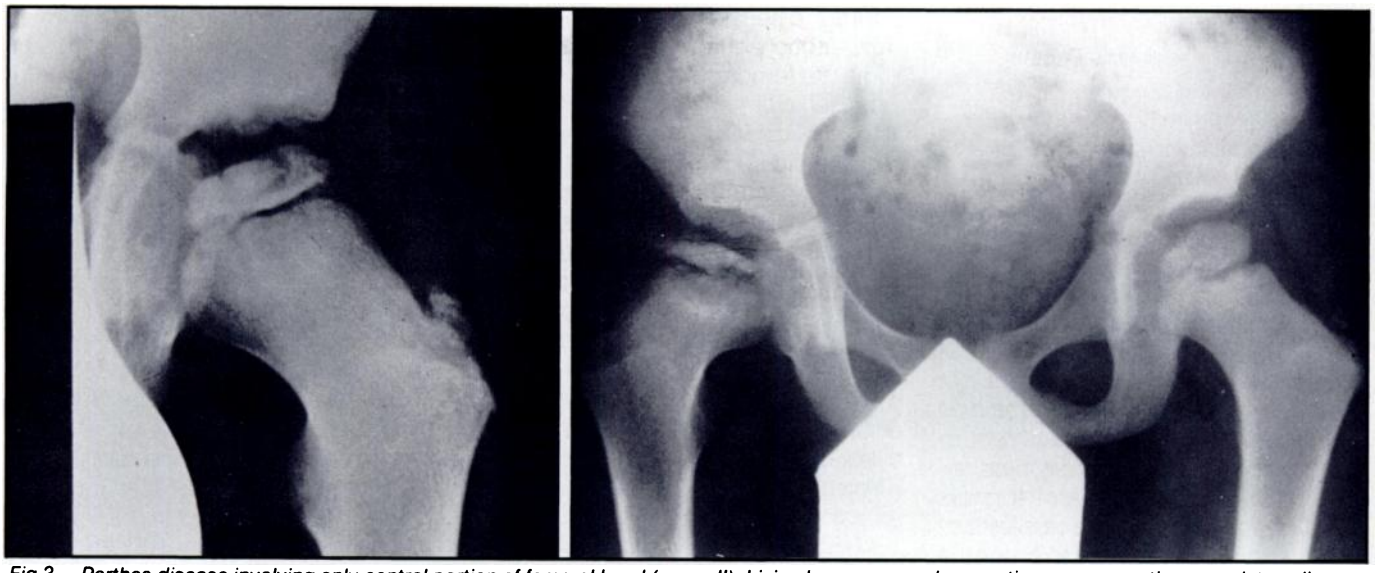

Fig 3. Perthes disease involving only central portion of femoral head (group II). Living bone surrounds necrotic area, preventing complete collapse.
Fig 4. Avascular necrosis of entire femoral head (group IV). Note irregular new bone formation on lateral portion of femoral head. Final shape in this case will be oval rather than spherical.

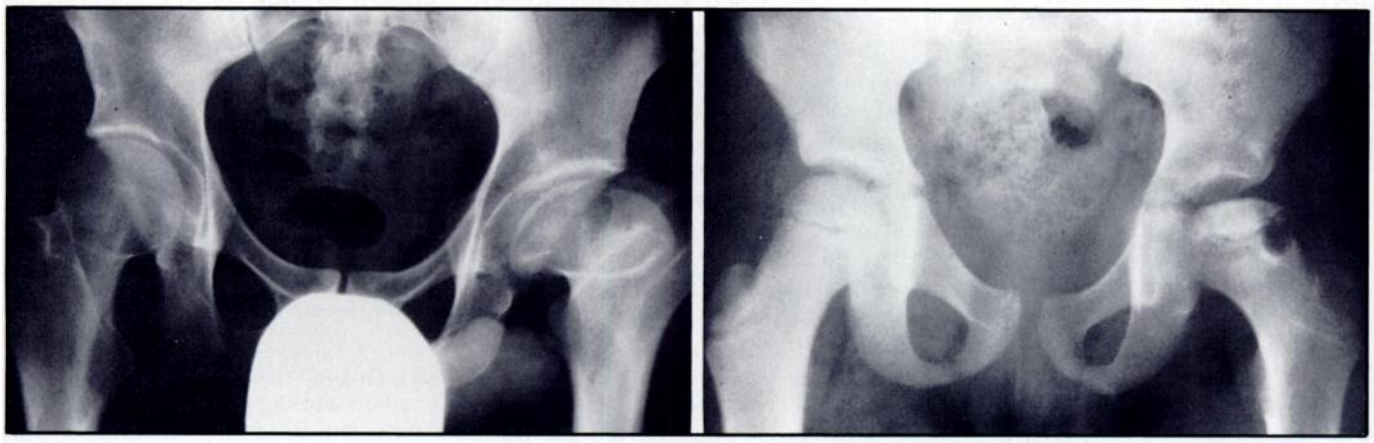

Fig 5. Definitive stage of Perthes disease. Premature closure of growth plate of femoral neck has resulted in a short neck with a superiorly placed greater trochanter.
Fig 6. Approximately three quarters of the femoral head is necrotic (medial portion remains viable) (group III). Note large cyst in femoral neck.

slight internal rotation. This position is maintained with a "broomstick" crossing between the casts. Although cumbersome and inconvenient, the method has proven to be an excellent means of obtaining and maintaining coverage of the femoral head. This method of treatment is prolonged, frequently requiring 6 months in such a cast.

The other nonoperative method of containment involves the use of various abduction orthoses for the hips. Their obvious advantage is the ease of removal for bathing purposes, but they suffer the disadvantage of promoting external rotation and uncovering of the femoral head. As with the plaster method, the length of treatment is frequently prolonged and ambulation with either method is awkward.

There has been extensive debate regarding the appropriate time to discontinue nonoperative treatment. It is now evident that maintaining the abducted position until complete healing is accomplished is unnecessary. Without question, the most vulnerable period of time is during the avascular and early fragmentation stages. As new bone formation takes place, the head becomes strong enough to resist the forces across it. It is generally accepted that nonoperative treatment may be discontinued when a significant amount of new bone can be seen on the radiograph, often within 6 to 12 months of the onset of the disease.

SURGICAL METHODS OF CONTAINMENT

Orthotic methods of containment are at best awkward, cumbersome, and undesirable and, thus, are often completely unacceptable to patients. A surgical procedure, although invasive, may avoid the need for a rigorous and restrictive orthotic treatment plan as well as significantly shorten the period of morbidity.

As in nonoperative methods, the goal of surgery is to promote containment of the femoral head. This may be done with an osteotomy of the

TABLE 5. Differential Diagnosis

- Transient synovitis
- Septic arthritis
- Osteomyelitis
- Multiple epiphyseal dysplasia
- Hemoglobinopathy

proximal femur, increasing the femoral neck-shaft angle to contain the femoral head more deeply within the acetabulum. Conversely, the surgical procedure may be directed toward the acetabulum; an osteotomy of the innominate bone may be carried out to redirect the acetabulum to cover the femoral head more completely.

RESULTS OF TREATMENT

Because Perthes disease is a self-limited condition with spontaneous healing, the final assessment of result is based largely on the final shape of the femoral head as seen radiographically. The ideal result is a perfectly spherical head. Numerous radiographic measurements to assess the shape of the femoral head have been reported. They include measurements of epiphyseal height and width and the center-edge angle of the hip joint. One of the more useful ways to assess sphericity is the use of the Mose template. This superimposes concentric circles over the femoral head and allows a strict measurement of sphericity. A good result requires that the femoral head be within 2 mm of perfect sphericity.

The results in those patients treated by simple weight relief (which does not accomplish containment of the femoral head) compared with those treated by either surgical or nonsurgical containment show a significant advantage for containment. In general, unsatisfactory results occur in nearly 50% of patients treated by any one of a number of noncontainment methods. This figure is reduced to 25% or 30% unsatisfactory results in patients treated by containment. It is also generally accepted that nonweight-bearing status adds little, if anything, to obtainment of a favorable outcome if the hip is properly contained.

The outcome is clearly influenced by the extent of involvement of the femoral head. Nearly 80% of all patients with less than 50% of the femoral head involved will have a satisfactory result. If more than 50% but less than the entire head is involved, 60% will have satisfactory results. If the entire femoral head is involved, results will be satisfactory in only 40% of all patients.

For those patients diagnosed before the age of 6 years, only 32% will have an unsatisfactory outcome; for those between the ages of 6 and 9 years, 44% will be unsatisfactory; and for those more than 9 years of age, 62% will have unsatisfactory results.

Results of operative containment are approximately equal to those obtained with nonoperative containment. A major advantage of operative treatment is its avoidance of cumbersome casts and braces and a significant shortening of intensive treatment. The major complication of surgical treatment seems to be some residual loss of motion.

The long-term prognosis for a hip affected by Perthes disease depends on the final outcome in childhood, when the goal of treatment was to maintain sphericity of the femoral head. Any permanent deformity of the femoral head creates incongruity with the acetabulum, setting the stage for premature degenerative arthritis. Studies of the natural history of the condition have established these relationships, with the risk of arthritis being directly related to the amount of residual deformity of the femoral head.

SUGGESTED READING

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EDUCATIONAL OBJECTIVE

58. Appropriate knowledge of the predictability of esophageal injury after ingestion of caustic substances (Recent Advances, 85/86).

Predictability of Esophageal Injury After Ingestion of Caustics

Predictability of Esophageal Injury From Signs and Symptoms: A Study of Caustic Ingestion in 378 Children. Gaudereault P, et al. *Pediatrics* 1983;71:767.
Pathophysiology and Management of Acute Corrosive Burns of the Esophagus. Haller JA, et al. *J Pediatr Surg* 1971;6:578.
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The ingestion of caustic agents results in direct destruction of tissue. Acid burns usually cause superficial injury as a result of coagulation necrosis. Strong acids are likely to cause damage to the stomach and duodenum, even to the point of perforation. Strong alkalis cause a liquefaction necrosis that penetrates deeply into muscle layers; this may result in esophageal perforation. Such deep burns are often associated with severe scarring and stricture formation. The prediction of the presence and severity of esophageal injury following the ingestion of a caustic substance is a major clinical problem for the pediatrician.

A collaborative retrospective study was designed by Gaudereault and co-workers

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