The natural history of Legg-Calve-Perthes disease

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The Natural History of Legg-Calvé-Perthes Disease*

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ABSTRACT: **Two groups of patients who had Legg-Calvé-Perthes disease were studied. The first group of patients consisted of eighty-eight patients (ninety-nine affected hips) followed in three hospitals for an average of forty years. The second group consisted of sixty-eight patients (seventy-two affected hips), all of whose radiographs from the onset of disease to maturity were available and all of whom had been treated in one hospital. The patients in this second group were followed for an average of thirty years.**

Each hip in both study groups could be placed into one of five classes of deformity based on its radiographic appearance at maturity. Each class showed a characteristic pattern of involvement during the active stages of the disease and had a specific long-term clinical and radiographic course. The clinical and radiographic course of an involved hip subsequent to childhood was related to the type of congruency that existed between the femoral head and acetabulum. Three types of congruency were recognized: (1) spherical congruency (Class-I and II hips) — in hips in this category arthritis does not develop; (2) aspherical congruency (Class-III and IV hips) — mild to moderate arthritis develops in late adulthood in these hips; and (3) aspherical incongruency (Class-V hips) — severe arthritis develops before the age of fifty years in these hips.

The objective in the treatment of Legg-Calvé-Perthes disease (coxa plana) is to minimize the development of deformities of the hip[2,3,6,8,12-14,17,19,23,28]. Although the deformities themselves do not produce significant discomfort or disability during the active stage of the disease or during adolescence, many believe that they lead eventually to severely disabling osteoarthritis. Many methods have been advocated for characterizing the residual deformities[3,8,11-14,19,21,23]. Each author's method emphasized aspects of the deformity that he considered to be most important in determining the fate of the hip. There is, however, no specific information available by which one can predict the likelihood that a given residual deformity will lead to arthritis. In the long-term follow-up studies that have been reported[1,4,5,7,9,16,18,20,22,24-26], a large proportion of patients who have had the disease, including those with severe residual hip deformities, have relatively mild symptoms as adults. In these long-term studies, the authors pointed out the apparent lack of association between the type and extent of femoral deformity and the development of degenerative joint disease.

In the present paper we report the results of two studies. We undertook the first study to establish the relationship between a residual deformity of the hip joint resulting from the disease and the likelihood of development of osteoarthritis. Because efficacy of treatment generally is defined in terms of success or failure in preventing deformities that lead to arthritis, this relationship must be known before the efficacy of treatment can be evaluated.

The purpose of the second study, using the classification of hip deformities developed in the first study, was to identify the clinical and radiographic features of the hip during the active phase of the disease that are associated with the development of the classified hip deformities.

Methods

The patients evaluated in the first study came from one of three medical centers: The Hospital for Sick Children in Toronto, the University of Iowa, and the Norrbacka Hospital of the Karolinska Institute in Stockholm, Sweden (Table I and Fig. 1). There were eighty-eight patients (with ninety-nine involved hips) in the first study. The average follow-up, from onset of symptoms in the hip to most recent examination, was forty years (range, thirty to sixty years). The results of long-term follow-up studies from The Hospital for Sick Children[27,28] and the University of Iowa[9] have been published previously. For this study, we re-evaluated the original clinical information and radiographs that had been collected by the investigators for those two previous studies and added the data collected from the Norrbacka Hospital.

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TABLE I

STUDY GROUP I

Institution	No. of Patients	No. of Hips	Average Age at Follow-up (Yrs.)	Sex Ratio M/F
University of Iowa	28	29	44.9	24/4
Hospital for Sick Children	27	37	47.1	24/3
Karolinska Institute	33	33	49.6	28/5
Total	88	99	47.3	76/12

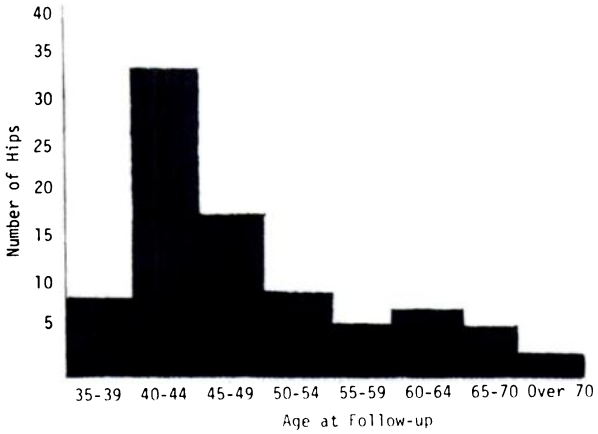

FIG. 1

Age distribution of study group I.

The follow-up examination of this first group consisted of a clinical evaluation of pain, function, and motion of the hip joint, and radiographic assessment of the nature and degree of residual deformity and the extent of osteoarthrosis present. Old medical records were available and made possible an assessment of the clinical course of the disease during its active stage. Detailed histories were obtained at final follow-up examination to establish the nature of the clinical course from childhood to the final examination.

The radiographs made during the active stage of the disease were missing for many of the hips in this group. However, anteroposterior and frog-leg lateral radiographs of the hip were made or were available for all of the patients at the final follow-up examination. There was enough clinical information available on 80 per cent of the patients to permit a rating of hip pain and function, using the Iowa hip-rating system[15], and range of motion of the hip joint, using the Harris hip-rating system. Six patients in this first study group had such severe symptoms that a surgical reconstructive procedure had been performed prior to the period when the follow-up examinations were carried out. The clinical data on these patients are not included in the tabulation of results (Table IV). However, the preoperative radiographs of these patients were available, and their evaluation is included in this study.

Treatment methods varied among the three institutions. However, each institution used a non-operative system which was applied systematically and consistently. The treatment of most of the patients at each medical center was supervised by a single individual who had a special interest in Perthes disease. In all three institutions the patients initially were treated with bed rest until the symptoms associated with the early, active phase of the disease had subsided. A device to prevent weight-bearing, but that did not provide containment, then was used until healing was thought to be complete. No patient was operated on during the active phase of the disease.

At the University of Iowa, after the initial period of bed rest a bilateral spica cast was applied with the hips in 10 to 20 degrees of abduction, full extension, and neutral rotation. The cast was changed every two months. The cast treatment was continued until radiographic evidence of complete healing was present.

At The Hospital for Sick Children, after the period of bed rest an ischial weight-bearing walking caliper (long Taylor walking caliper[29]) was applied. The brace was designed to position the hip in 10 degrees of abduction, full extension, and neutral rotation. The splint was worn until complete regeneration of the femoral head occurred.

At the Norrbacka Hospital, the period of bed rest was followed by the use of crutches and a high cork shoe-lift on the normal side.

Radiographs that had been made at skeletal maturity were evaluated to establish the type and extent of residual deformity that had resulted from the disease before any changes of osteoarthrosis had developed. The radiographs were examined to establish the shape and size of the femoral head, the length of the femoral neck, the slope of the acetabulum, and the adequacy of acetabular coverage of the femoral head (Fig. 2). The radiographs made at the final follow-up examination were evaluated to establish the characteristics and extent of osteoarthritis present.

Osteoarthritis was considered to be present on a follow-up radiograph if *any* of the following findings were present: (1) sclerosis, (2) femoral or acetabular osteophytes, (3) cysts, or (4) narrowing of the joint space. The study previously carried out at The Hospital for Sick Children[28] found that the radiographic sign of osteoarthrosis most closely correlated with clinical symptoms following Perthes disease is narrowing of the joint space. Therefore, a grading system for the extent of osteoarthrosis was established and defined as follows: mild — joint-space narrowing less than 25 per cent of normal; moderate — joint-space narrowing less than 75 per cent of normal; and severe — joint-space narrowing more than 75 per cent of normal.

The second study (Fig. 3) consisted of sixty-eight patients (seventy-two involved hips), all treated at the Norrbacka Hospital between 1921 and 1951. Two hundred and thirty-seven patients were treated for Perthes disease at that hospital during this period. Of these, complete charts and radiographs were available for 171. The following data characterize the sixty-eight patients chosen for

this study: all had been seen early in the disease process (at the fragmentation stage or earlier); had had radiographs made frequently, usually at three-month intervals for two to three years; had been followed to skeletal maturity; and had been treated similarly. There were fifty-six male and twelve female patients. The average age at which the diagnosis was made was 7.5 years (range, three years and eight months to fifteen years and three months). The average age at follow-up was 32.3 years.

two crutches and wearing a cork shoe-lift on the foot of the uninvolved leg. No attempt was made to place the affected hip in abduction. This treatment was continued for an average of nine months, the range being a few weeks to sixteen months.

Radiographs made during the active stage of the disease were examined for the extent and location of epiphyseal involvement and were classified according to the system of Catterall[2]. They also were evaluated to determine

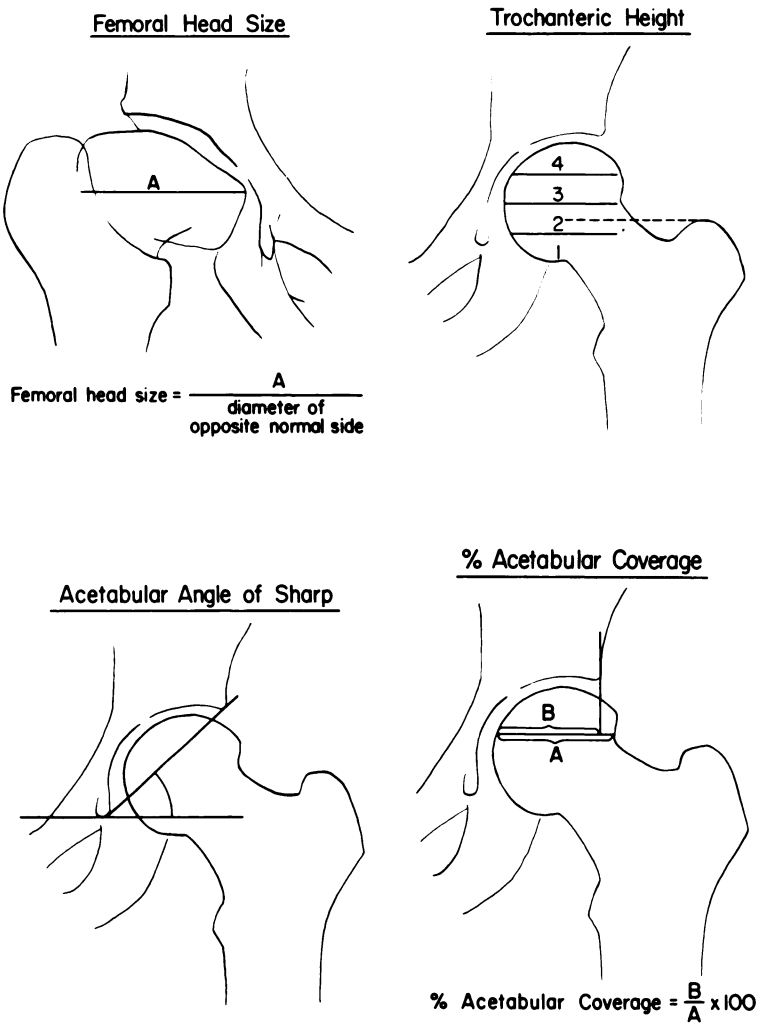

FIG. 2

Radiographic assessment at maturity and measurement of residual deformity.

Femoral head shape — The Mose method[19] was used, employing concentric circular templates.

Femoral head size — A line is drawn on the anteroposterior radiograph, connecting the most medial and lateral points on the femoral head. The diameter of the normal side is measured, and a ratio is formed: involved side divided by normal side. Coxa magna was present if the involved femoral head had a diameter 10 per cent greater than that of the normal side.

Femoral neck length — This was measured by relating the height of the greater trochanter to the femoral head. The head was divided into four quadrants. If the trochanter was at the level of the middle of quadrant 2, it was given the numerical value of 2.0; if it was halfway between quadrants 2 and 3, the value was 2.5, and so on. An imaginary fifth quadrant was above quadrant 4.

Acetabular slope — This was assessed using the acetabular angle of Sharp.

Acetabular coverage — This was expressed as the percentage of the femoral head covered by the acetabulum. A line is dropped perpendicular to a line connecting the teardrop figures from the lateral lip of the acetabulum on the anteroposterior radiograph through the femoral head. The largest extent of the head is measured perpendicular to this line and the percentage of the head medial to the lip of the acetabulum is calculated along this line.

Forty-one of the sixty-eight patients initially were treated with bed rest to alleviate acute spasm of the muscles of the hip (average, two months; range, a few days to five months). Once a patient had a full range of motion, he or she was permitted non-weight-bearing ambulation using

the extent and direction of subluxation of the femoral head (Fig. 4). The presence of lateral epiphyseal extrusion was noted. The location and extent of metaphyseal involvement was determined and was graded as none, mild, moderate, or severe.

TABLE II

CHARACTERISTICS OF THE FIVE RESULT CLASSES

Final-Result Class	Femoral Head Shape	Mose Rating	Study Group I					Study Group II				
			No. of Hips	Coxa Magna Present (Per cent)	Femoral Neck Length*	Acetabular Slope (Degrees)	Acetabular Coverage (Per cent)	No. of Hips	Coxa Magna Present (Per cent)	Femoral Neck Length*	Acetabular Slope (Degrees)	Acetabular Coverage (Per cent)
I	Spherical	Good	20	0	2.97 (2-3.5)	38.3 (37-42)	86 (75-100)	19	0	2.5 (1.5-3)	35.6 (30-42)	85 (73-100)
II	Spherical	Fair	12	83	3.58 (2.5-4.5)	38.1 (32-43)	83 (75-100)	18	72	3.58 (2-5)	36.2 (32-45)	80 (57-100)
III	Ovoid	Poor	17	94	4 (3-5.5)	43 (35-47)	61 (50-100)	11	100	3.9 (3-5)	43.6 (35-51)	75 (63-84)
IV	Flat	Poor	32	93	3.9 (3-5.5)	43.4 (40-52)	64 (50-100)	10	100	3.81 (3-5)	46.9 (40-52)	59 (44-71)
V	Flat	Poor	18	22	3.19 (2-4)	39.5 (30-45)	82 (75-100)	14	36	3.3 (2.5-4.0)	38.9 (35-45)	78 (65-100)

* See Figure 2.

The radiographs made at skeletal maturity were examined to establish the shape and size of the femoral head according to the criteria established (Fig. 2). The patients' records were reviewed to establish the age at onset of symptoms and the clinical course during the active stage of the disease.

We did not carry out a long-term clinical examination of this group of patients.

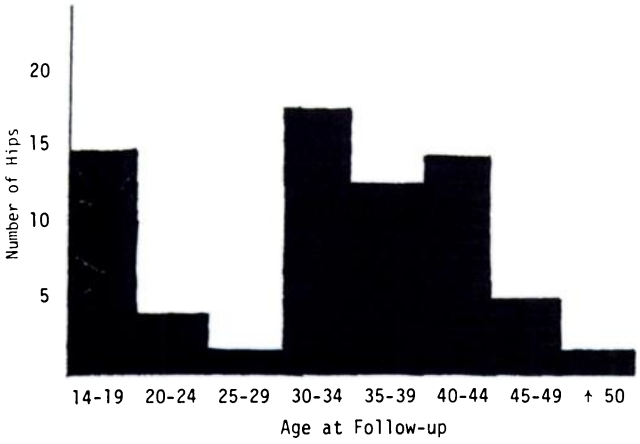

FIG. 3

Age distribution of study group II. There were fifty-six male and twelve female patients; the average age at follow-up was 32.3 years. There were seventy-two hips in this group.

Results

The hips in both groups were placed into one of five result classifications according to the radiographic characteristics and deformities that they shared. The individual radiographs used for classification were those made when the hips became skeletally mature and before any changes of osteoarthritis occurred. The classifications are as follows.

Class I: A completely normal hip joint (Fig. 5-A).

Class II: A *spherical* femoral head (same concentric circle on anteroposterior and frog-leg lateral radiographs), but with one or more of the following abnormal characteristics of the femoral head, neck, or acetabulum: (1) larger than normal (although spherical) femoral head (coxa magna); (2) shorter-than-normal femoral neck; or (3) abnormally *steep* acetabulum (Fig. 5-B).

Class III: A *non-spherical* (ovoid, mushroom-shaped, or umbrella-shaped) but not flat femoral head. Abnormal characteristics of the femoral head, neck, and acetabulum

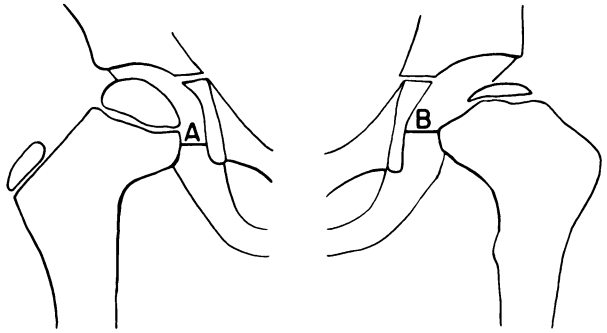

FIG. 4

The extent and direction of femoral subluxation. Superior subluxation is considered to be present if Shenton's line is broken on at least two anteroposterior radiographs made either on separate days or on the same day.

Lateral subluxation is measured by identifying the most medial part of the proximal part of the involved femur and measuring the horizontal distance to the acetabulum. The normal side then is measured at a comparable point and a ratio is formed: involved side divided by normal side.

TABLE III

CLINICAL PROFILE OF PATIENTS IN STUDY GROUP I

Class	No. of Hips	Age at Onset of Symptoms (Yrs.)	Average Age at Follow-up (Yrs.)	Iowa Hip Rating*		Harris Hip Rating for Motion†
				Pain	Function	
I	20	6.0	42.4	35	35	6
II	12	6.41	45.2	33.9	35	6
III	17	8.23	52.2	32.6‡	33‡	4.8‡
IV	32	8.32	47.75	31.1	32.8	4.5
V	18	10.4	47.72	30.0§	32.4§	3.64§

* Out of a possible 35 points.
† Out of a possible 6 points.
‡ Does not include one patient who had surgery prior to follow-up (osteotomy at the age of forty).
§ Does not include five patients who had surgery prior to follow-up (three total hip replacements at the ages of forty-five, fifty-six, and fifty-nine, one cup arthroplasty at the age of thirty-one, and one hip fusion at the age of thirty-two).

(as described for Class II) are present also (Figs. 6-A and 6-B).

Class IV: A *flat* femoral head and abnormalities of the femoral head, femoral neck, and acetabulum (Figs. 7-A and 7-B).

Class V: A *flat* femoral head and a normal femoral neck and normal acetabulum (Figs. 8-A and 8-B).

in all three classes tended to be short, and the acetabula tended to be abnormally steep. The femoral heads of hips in these classes often were poorly covered.

Even though the hips in Class V also all had poor Mose ratings, fewer of the femoral heads than in Classes II, III, and IV were abnormally large. Moreover, the femoral neck lengths and the acetabula of the hips in Class

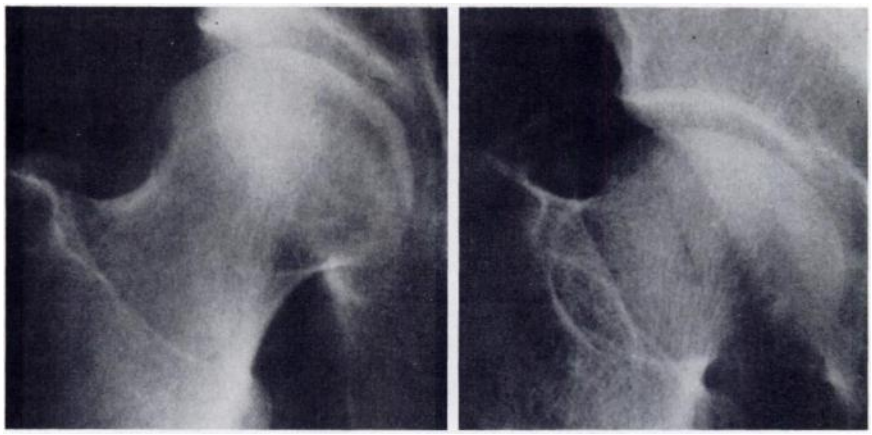

FIG. 5-A

FIG. 5-B

Fig. 5-A: Class I. This sixty-four-year-old man has an essentially normal hip following Perthes disease, with no sign of arthritis. He was completely asymptomatic.
Fig. 5-B: Class II. In this sixty-nine-year-old man, the femoral head is spherical on the anteroposterior and lateral radiographs (not shown) of the hip. The femoral head is larger than normal (coxa magna). The femoral neck is short and there is marked trochanteric overgrowth. The femoral head is well covered, although the acetabulum is steeper than normal. No arthritis is present. The patient has no symptoms.

Analysis of the abnormal characteristics in each class of deformity (Table II) reveals that the Class-I (normal) hips all have good Mose ratings, normal-size femoral heads, normal acetabula, and well covered femoral heads (average, 86 per cent; range, 75 to 100 per cent). All hips in Class I from both study groups conformed to the criteria of the normal hip (Figs. 2 and 4), without exception.

Class-II hips all had fair Mose ratings and the hips in Classes III and IV all had poor Mose ratings. Virtually all of the femoral heads in these three classes were abnormally large. However, the femoral heads of hips in Class II were spherical, while those in Class III were ovoid and those in Class IV were flat. The femoral necks of the hips V were much closer to normal. Virtually all of the hips in Class V had normally covered femoral heads, unlike most of the hips in Classes III and IV.

The clinical profile of the patients in the first study (Table III) reveals an identifiable pattern in each of the five classes. The patients in Class I had the lowest average age at onset of the disease. They were asymptomatic at follow-up examination. Patients in Class II also were functioning essentially normally at follow-up. Patients in Classes III and IV were somewhat older at onset of the disease and were, on the average, more symptomatic at follow-up. However, few had required any restrictions in activity during their years of employment, and many had

worked at heavy labor. In addition, many patients in these groups had been or were athletic. In contrast, patients in Class V, almost without exception, were much older at the onset of their disease. At follow-up, patients in this group had more pain and were more disabled than the patients in previous groups.

Class V were doing less well than the patients in the other groups.

The extent of osteoarthrosis by class (Table IV) shows that the incidence of osteoarthrosis increased from Class I to Class V, as did the radiographic severity of the degenerative joint disease.

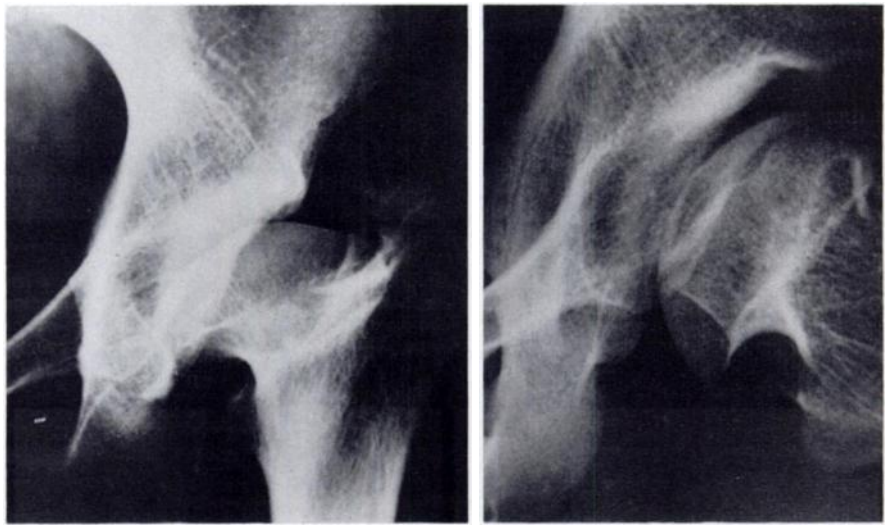

FIG. 6-A

Class III. In this twenty-six-year-old woman the femoral head is not spherical, but it is not flat. It is somewhat mushroom-shaped. The head is larger than normal. The femoral neck is quite short, with considerable trochanteric overgrowth. About 40 per cent of the femoral head is not covered by the acetabulum, which is steeper than normal.

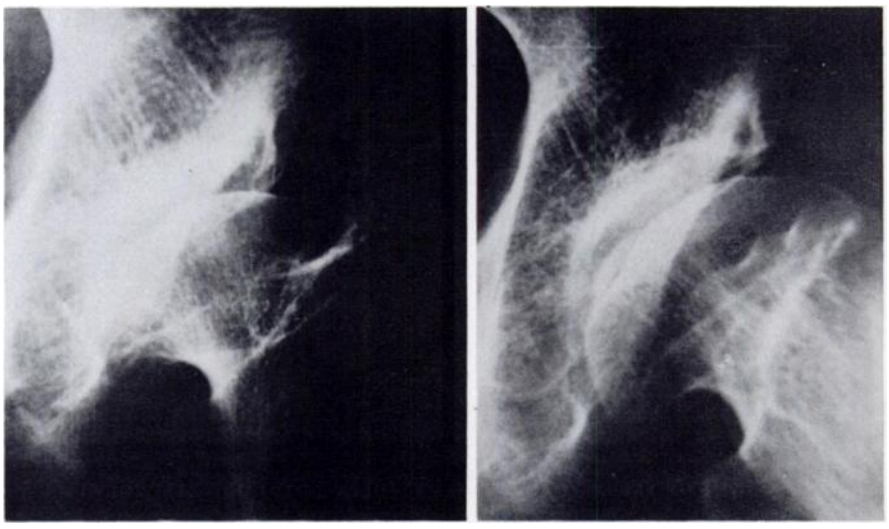

FIG. 6-B

Class III. The same patient seen in Fig. 6-A, at the age of sixty-three years. Minimum signs and symptoms of degenerative joint disease are present. There is some irregularity of the femoral head surface, subchondral sclerosis, and a lateral acetabular cyst. The joint space is well maintained.

In six patients such severe pain in the hip had developed that a surgical reconstructive procedure had to be performed prior to the follow-up examination for this study. Five of these patients were in Class V and one, with an osteochondritis dissecans lesion, was in Class III. Because these patients were excluded from tabulation, Table III significantly understates the extent to which patients in

The clinical and radiographic profiles during the active stage of the disease of the patients in the second study group are given in Table V for each of the five classes. In Class I, the pattern of epiphyseal involvement was partial (Catterall type I or II), with little or no epiphyseal collapse (Figs. 9-A and 9-B). There was little or no lateral subluxation, and no superior subluxation or lateral epiphyseal ex-

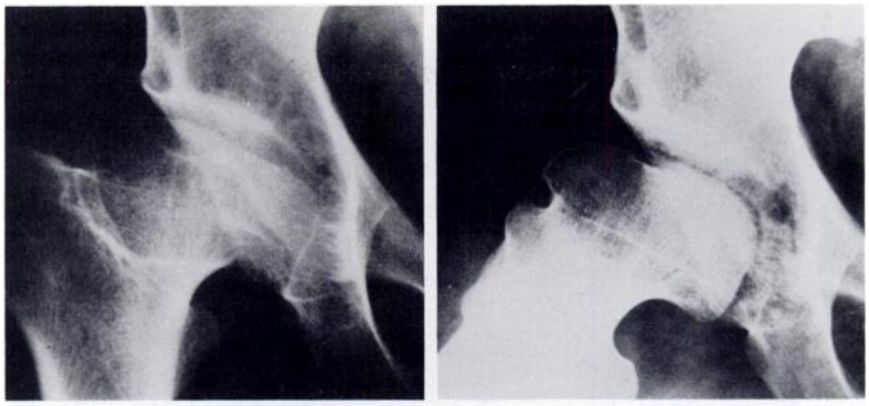

FIG. 7-A

Class IV. In this forty-three-year-old man, marked flattening of the large femoral head is present. The femoral neck is short and the acetabulum is abnormally steep. The femoral head is quite well covered.

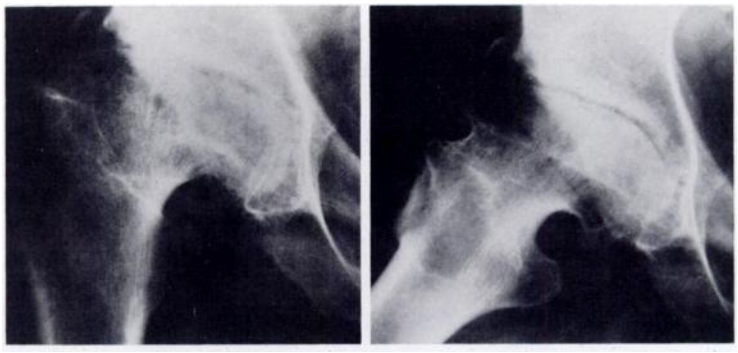

FIG. 7-B

Class IV. The same patient as in Fig. 7-A, at the age of sixty-seven years. Degenerative joint disease is present, with marked joint-space narrowing and osteophyte and cyst formation. The patient retired from a job requiring heavy labor at the age of fifty-eight. He now uses anti-inflammatory agents intermittently and walks with a cane.

trusion. In 25 per cent of the hips there was a slight flattening of the lateral lip of the acetabulum. In the rest, this flattening did not occur during the active phase of the disease.

In Class II (Figs. 10-A and 10-B), partial and whole involvement of the head (Catterall types II, III, and IV) was seen in association with mild to moderate lateral subluxation, no superior subluxation, and mild to moderate metaphyseal involvement. The lateral lip of the acetabulum became flattened or vertically inclined in eight of eighteen hips during the active phase of the disease.

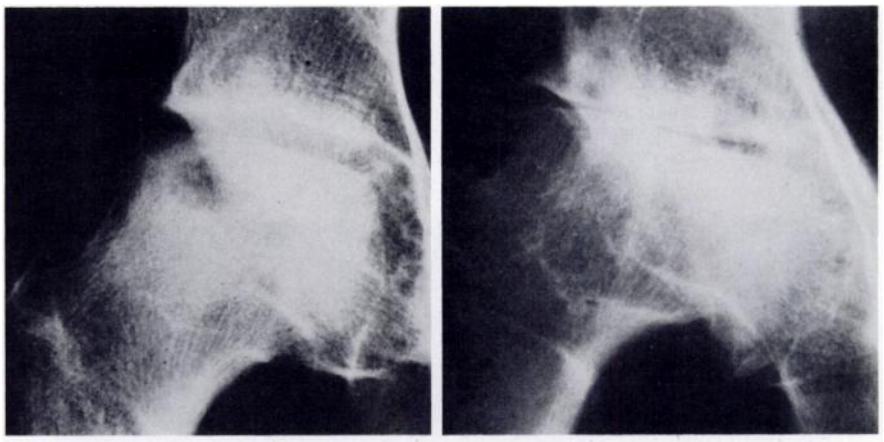

FIG. 8-A FIG. 8-B

Fig. 8-A: Class V. This seventeen-year-old boy has a flat femoral head and essentially normal femoral neck. The normally shaped acetabulum provides normal coverage. There is an unhealed cystic area in the lateral aspect of the femoral head.

Fig. 8-B: Class V. The same patient as in Fig. 8-A, at the age of forty-one years. Severe degenerative joint disease is present, with marked joint-space narrowing and extensive cyst and osteophyte formation. Marked collapse of the femoral head also is present. The patient has significant discomfort and must restrict his activities markedly.

TABLE IV

EXTENT OF OSTEOARTHRITIS AT FOLLOW-UP

Class	No. of Hips	No. of Hips with Osteoarthritis	No. of Hips with Joint-Space Narrowing	Degree of Joint-Space Narrowing				Hips with Femoral and/or Acetabular:		
				None	Mild	Moderate	Severe	Osteophytes	Cysts	Sclerosis
Study group I										
I	20	0	0	20	0	0	0	0	0	0
II	12	2 (16%)	0	12	0	0	0	0	2	0
III	17	10 (58%)	8 (47%)	9	2	3	3	10	9	2
IV	32	24 (75%)	17 (53%)	15	7	7	3	21	18	3
V	18	14 (78%)	11 (61%)	7	1	2	8	13	13	3
Study group II										
I	19	0	0	19	0	0	0	0	0	0
II	18	0	0	18	0	0	0	0	0	0
III	11	0	0	11	0	0	0	0	0	0
IV	10	4 (40%)	2 (20%)	2	0	2	0	3	2	0
V	14	12 (86%)	4 (35%)	10	0	1	3	9	9	5

In Class III (Figs. 11-A and 11-B), involvement of the whole head (Catterall types III and IV), predominated in association with moderate lateral subluxation, mild superior subluxation in one-half of the hips, and moderate to severe metaphyseal involvement. The acetabulum became flattened or vertically inclined in more than 75 per cent of the hips during the active phase of the disease.

In Class IV (Figs. 12-A and 12-B), both partial and whole-head involvement was noted, but involvement of the whole head occurred 70 per cent of the time. Pronounced lateral and superior subluxation occurred, as did extensive and severe metaphyseal resorption. The lateral lip of the acetabulum was flattened or vertically inclined in 75 per cent of the hips.

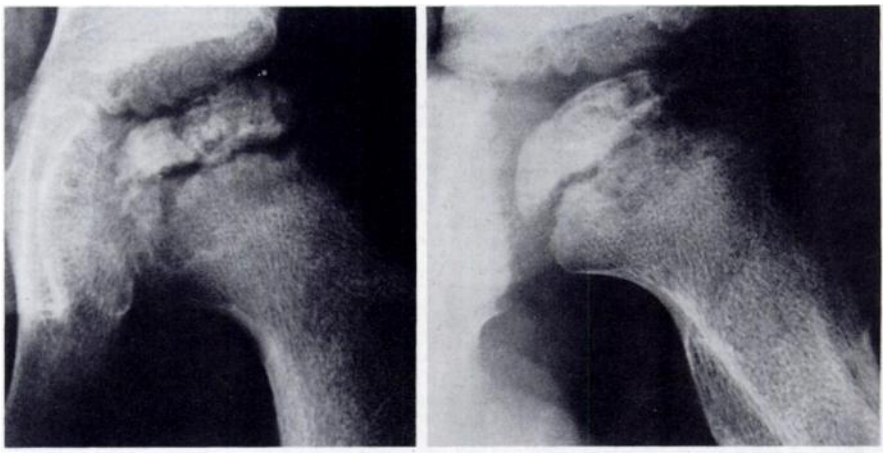

FIG. 9-A

Class I. This boy, six and one-half years old, has partial involvement of the femoral head without collapse. Mild metaphyseal resorption is present. There is no superior subluxation and slight lateral subluxation.

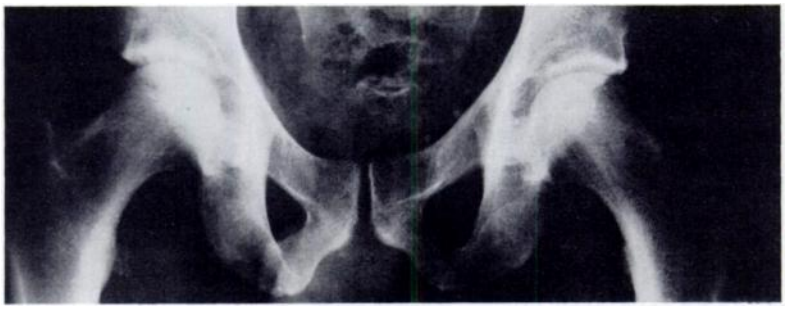

FIG. 9-B

Class I. The same patient shown in Fig. 9-A, at the age of thirty-two. The hip is radiographically normal.

In Class V (Figs. 13-A, 13-B, and 13-C) *partial* involvement of the head was seen almost exclusively. Metaphyseal involvement was limited. Lateral subluxation essentially was absent in six of fourteen hips (average, 1.2 times normal) and marked in the rest (average, 2.5 times normal). Superior subluxation was uncommon. The lateral lip of the acetabulum was flattened in six of fourteen hips during the active phase of the disease. The femoral heads of hips in Group V often did not heal completely, leaving residual cysts in the anterosuperior aspect of the femoral head. Osteochondritis dissecans was seen in three of these fourteen hips.

Discussion

A deformity produced by Legg-Calvé-Perthes disease is significant if it leads to painful and disabling osteoar-

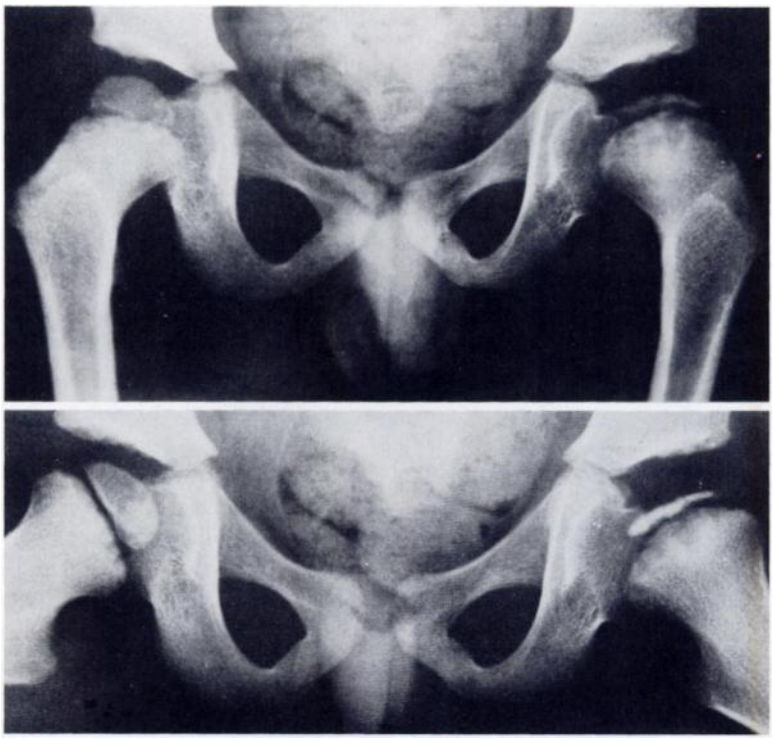

FIG. 10-A

Class II. In this boy, five years and five months old, there is involvement of the whole head and collapse of the epiphysis, with mild lateral and no superior subluxation.

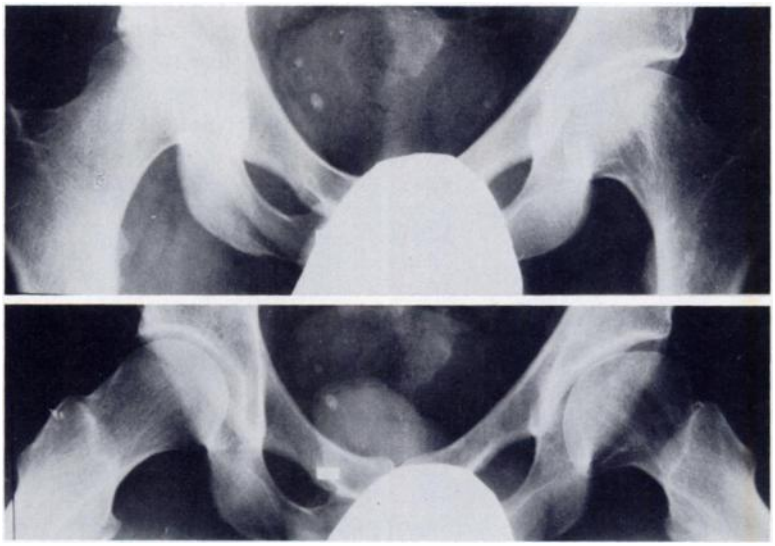

FIG. 10-B

Class II. The same patient shown in Fig. 10-A, thirty-one years old. Femoral neck widening is present, without shortening. Using the epiphyseal scar as a marker, the epiphyseal height is noted to be decreased. The femoral head is spherical and is somewhat larger than on the opposite side. The acetabulum is slightly steeper than normal, but the femoral head is well covered. There is no radiographic evidence of arthritis. The hip joint is spherically congruent.

TABLE V

CLINICAL AND RADIOGRAPHIC PROFILE DURING THE ACTIVE STAGE OF PERTHES DISEASE IN STUDY GROUP II

Class	No. of Hips	Male/Female Ratio	Age at Onset of Symptoms (Yrs.)	No. of Hips of Each Catterall Type	Lateral Epiphyseal Extrusion	Lateral Subluxation Ratio	Shenton's Line
I	19	16/3	6.1 ($3^{8}/_{12}$-$8^{6}/_{12}$)	I — 12 II — 7 III — 0 IV — 0	No — 19 Yes — 0	1.2 (1.0-1.57)	19 intact 0 broken
II	18	16/2	6.5 ($4^{6}/_{12}$-$8^{7}/_{12}$)	I — 0 II — 9 III — 4 IV — 5	No — 13 Yes — 5	1.55 (1.09-2.14)	18 intact 0 broken
III	11	9/2	6.7 ($4^{3}/_{12}$-$8^{7}/_{12}$)	I — 0 II — 1 III — 4 IV — 6	No — 6 Yes — 5	1.75 (1.2-3.0)	6 intact 5 broken
IV	10	6/4	7.6 ($5^{9}/_{12}$-$8^{11}/_{12}$)	I — 0 II — 3 III — 4 IV — 3	No — 5 Yes — 5	2.40 (1.75-3.33)	4 intact 6 broken
V	14	13/1	10.5 ($6^{6}/_{12}$-$15^{3}/_{12}$)	I — 0 II — 13 III — 0 IV — 1	No — 11 Yes — 3	1.85 (1.0-4.0)	12 intact 2 broken

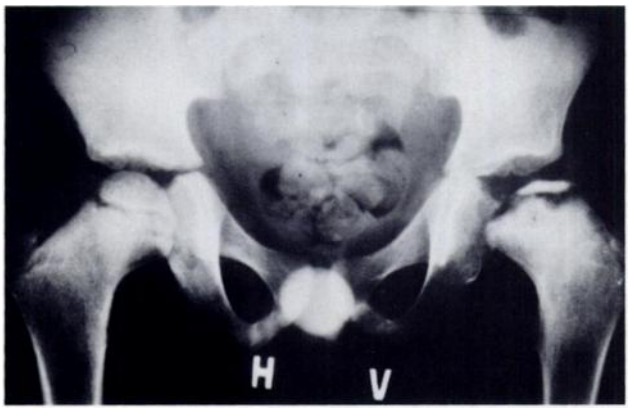

FIG. 11-A

Class III. In this boy, seven years and five months old, the whole femoral head is involved, with complete collapse of the epiphysis. Moderate lateral and mild superior subluxation of the femoral head is present.

thritis in early adulthood. Most studies of this disease have classified the final results of treatment using grading methods similar to that developed by Mose[19]. Those methods equated quality of the result with the degree of sphericity of the femoral head. These classifications have presumed that lack of femoral head sphericity would be associated with the onset of arthritis in early or mid-adulthood. However, the long-term follow-up studies that are available[9,16,20,24,25,27,28] indicated that patients with severely deformed hips may have relatively mild symptoms as adults. Our study also emphasizes that the lack of sphericity of the femoral head, as determined for example by the Mose classification, does not in itself provide an accurate prediction of the long-term course of a hip that has had Perthes disease.

However, the data presented support our thesis that the residual deformity at skeletal maturity that results from

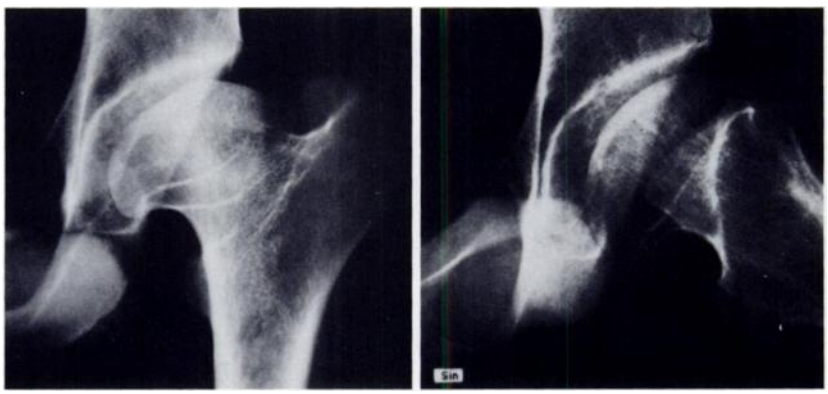

FIG. 11-B

Class III. The patient shown in Fig. 11-A, thirty-four years old. The femoral head is aspherical and somewhat mushroom-shaped. The femoral neck is quite short and there is considerable trochanteric overgrowth. The acetabulum is steep. The hip joint is aspherically congruent. Arthritis is not present.

Legg-Calvé Perthes disease can be accurately assigned to one of five result classes, each of which is associated with a predictable future clinical and radiographic course. In this classification system, three categories of deformity (Classes III, IV, and V) include hips with *poor* Mose ratings. However, only the hips in Class V are associated with the development of painful and disabling osteoarthritis in early adulthood. The hips in Classes III and IV, in contrast, are associated with relatively benign clinical and radiographic courses in spite of significant residual de-

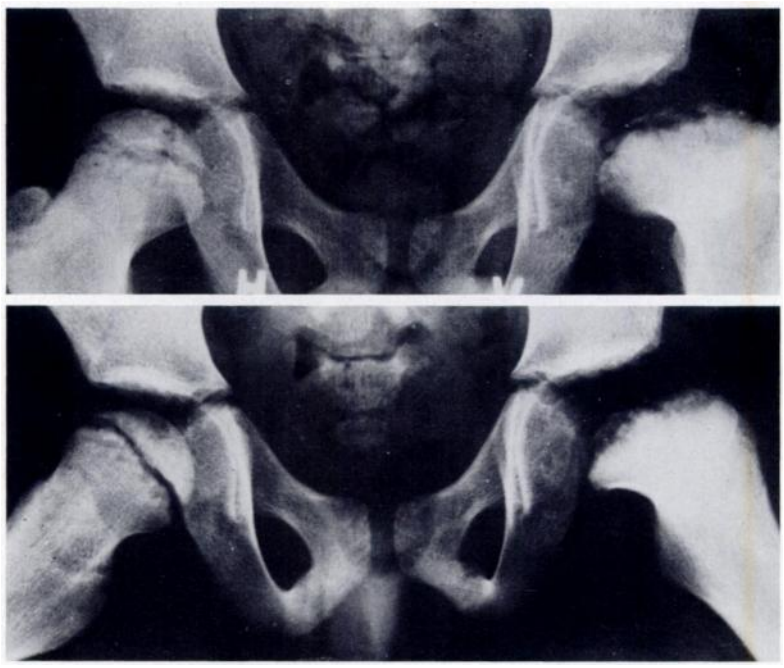

FIG. 12-A

Class IV. This boy, six years and eight months old, has total involvement of the femoral head and intense epiphyseal collapse and resorption, with marked lateral and modest superior subluxation.

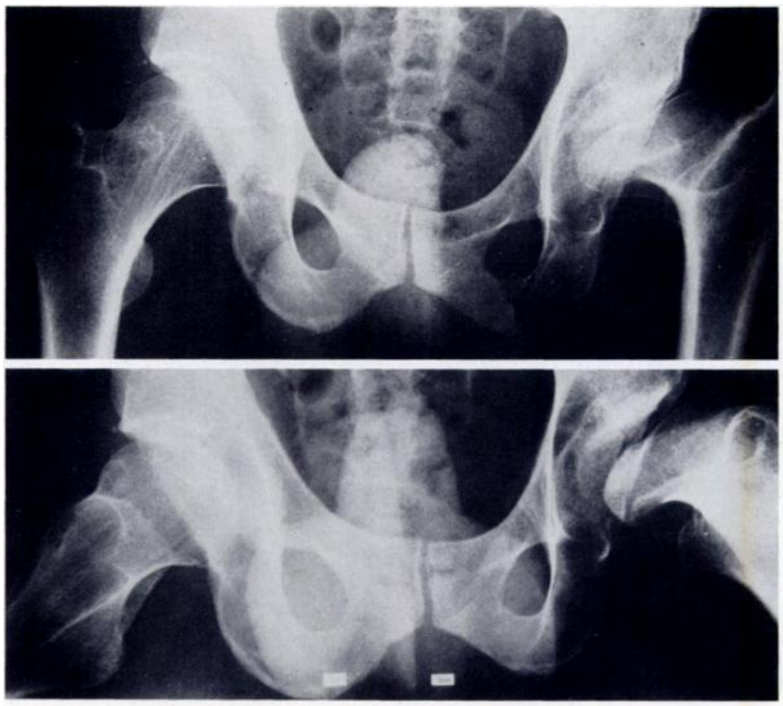

FIG. 12-B

Class IV. The patient shown in Fig. 12-A, at the age of forty-five. Marked flattening of the femoral head and shortening of the femoral neck are present. The very steep acetabulum partially covers the femoral head. However, there are no signs of degenerative joint disease. The hip joint is aspherically congruent.

formities. The Class-V deformity usually occurred in children whose onset of symptoms was after the age of nine years. That deformity is characterized by a flat femoral head and, unlike the deformity of the hips in Class IV (which also have flat femoral heads), a normal acetabulum and a femoral neck of normal length. This deformity is similar to what Mose et al.[20] called the "irregular femoral head". In their long-term follow-up study, this

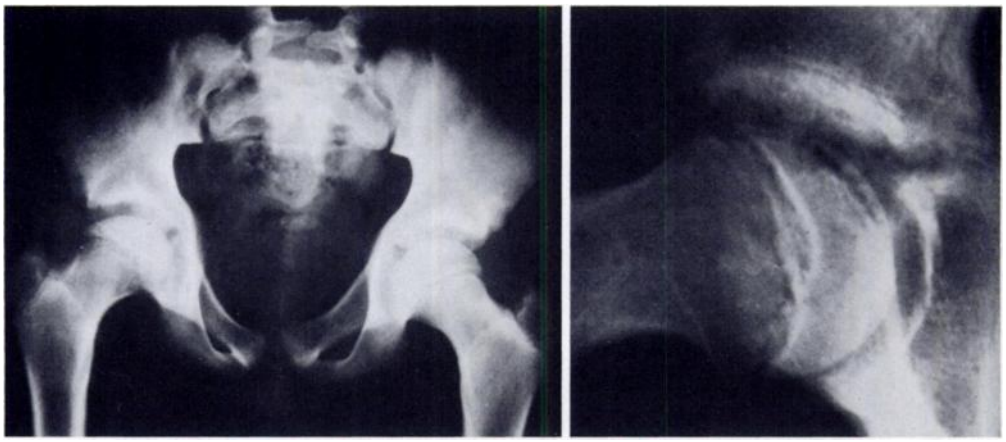

FIG. 13-A

Class V. This thirteen-year-old boy shows early signs of partial involvement of the femoral head. A crescent sign extends across approximately one-half of the epiphysis. There is no significant subluxation.

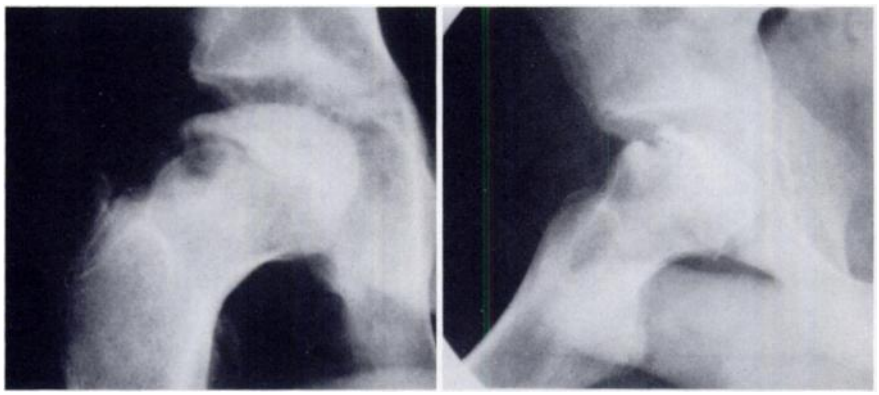

FIG. 13-B

Six months later, collapse of the epiphysis is present. There is a large cyst within the femoral neck. With the exception of the cyst, there is little activity in the metaphysis.

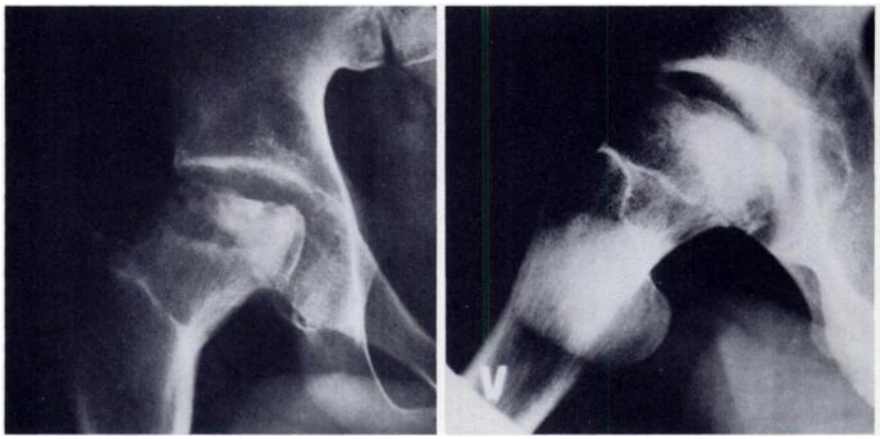

FIG. 13-C

At twenty-three years old, the femoral head is flat. There is nearly complete acetabular coverage, and the acetabular contour is essentially normal. The femoral neck is of essentially normal length. The acetabulum is round and the head is aspherical. The hip joint therefore is asphericallly incongruent.

deformity also was the *only* one associated with a significant incidence of osteoarthritis before the age of sixty years. We believe that it is the late onset of Perthes disease in patients in Class V that leads to the development of a flat femoral head without associated remodeling of the acetabulum. This results in a hip joint that is *aspherically incongruent*. This incongruous relationship leads to the rapid development of discomfort, dysfunction, and arthritis (Figs. 8-A and 8-B).

The pattern of involvement of the femoral head associated with the development of a Class-V deformity was characteristic and was relatively easy to recognize early in the course of the disease. The head was invariably only *partially* involved, usually in the anterosuperior quadrant. Metaphyseal resorption was rare in this group and lateral subluxation was present inconsistently; its degree seemed unrelated to the severity of the residual deformity.

In this study we found, as had others in their long-term follow-up studies, that many patients with deformed hips (Classes III and IV) do quite well, and that radiographic signs of osteoarthritis begin to develop only when the patients are in their forties and fifties. Disabling arthritis with extensive osteophyte formation, multiple cysts, and especially marked joint-space narrowing did not occur in most of these patients. In general, in patients in Class III significant symptoms of arthritis develop more slowly than in patients in Class IV. A study with an even longer follow-up than that of our first study group will be necessary, however, to determine how the clinical courses of hips in Classes III and IV differ after the age of sixty years.

The deformities in Classes III and IV occurred in children whose average age at onset of symptoms was two to three years younger than those in Class V. The acetabulum appears to be able to remodel during the active stage of the disease to conform to the deformed femoral head. This produces a hip joint that radiographically appears approximately congruent (Figs. 6-A through 7-B). This state of *aspherical congruity* seems to protect the hip from arthritis until late adulthood and invariably permits essentially full function through early and middle adulthood.

Patients in Classes III and IV had a pattern of involvement of the femoral head and subluxation that was much different than that seen in Class V. Most of the hips in these groups had extensive involvement of the femoral head (Catterall types III and IV) and severe metaphyseal resorption. However, the radiographic sign that appeared to be most closely related to the extent of flattening was the degree of lateral and superior femoral subluxation. A femoral head with a Catterall type-III or IV pattern of involvement was likely to become large and deformed, but not flat (Class III), if associated with moderate lateral subluxation and only mild superior subluxation. As the degree of lateral subluxation increased, and especially as superior subluxation became more pronounced, a large, *flat* (Class-IV) femoral head tended to develop.

Patients in Classes I and II functioned essentially normally throughout adulthood. Virtually all were pain-free. These patients had the earliest age at onset of the disease, averaging one to two years younger than patients in Classes III and IV.

Deformities in Class II resulted from both extensive (Catterall types III and IV) and partial (Catterall types I and II) involvement of the femoral head. Extensive involvement was associated with the subsequent development of a large femoral head and acetabulum. The hips in Class II often had extensive metaphyseal resorption and involvement of the epiphyseal plate. This appeared to be associated with the development of a short femoral neck. In contrast to hips in Classes III and IV (which also had extensive femoral head and metaphyseal involvement), hips in Class II had only mild lateral subluxation and no superior subluxation whatsoever. This absence of subluxation appeared to be associated with the development of a spherical femoral head. Because the epiphyseal quotients[13,19] of the femoral heads in Class II were less than 60, hips in this category would be classified as fair in the Mose rating system.

The patients in Class I had relatively limited femoral-head involvement (Catterall types I and II) during the active stage of the disease. This particular involvement was associated with mild, if any, metaphyseal involvement and little or no subluxation. The result was a hip without residual hip-joint deformity (classified as good in the Mose rating system).

All of the hips in Classes I and II had a spherical femoral head and correspondingly shaped acetabulum. The hips in these classes were *spherically congruent*. Hips in Class II might have a large femoral head, short femoral neck, and high-riding greater trochanter (Figs. 5-A and 5-B), but these abnormalities do not affect the long-term result, which is essentially full, normal function and no arthritis.

The objective of all of the current containment-treatment programs for Perthes disease is to prevent lateral and superior subluxation. Our study indicates that the likelihood of development of an aspherical femoral head (Classes III and IV) may be directly related to the degree of subluxation that occurs during the active stage of the disease. However, in Class V the development of an aspherical femoral head is *not* related to the degree of subluxation that occurs. Our study emphasizes that it is only the hips in Class V in which severe osteoarthritis will develop in early or mid-adulthood. Thus, the study raises the question of whether current containment-treatment methods are likely to alter the natural course of Perthes disease and its sequelae in patients who are at particular risk of development of arthritis in early adulthood.

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References

1. BRODER, HAROLD: The Late Results in Legg-Perthes' Disease and Factors Influencing Them. A Study of One Hundred and Two Cases. Bull. Hosp. Joint Dis., **14:** 194-216, 1953.
2. CATTERALL, A.: The Natural History of Perthes' Disease. J. Bone and Joint Surg., **53-B(1):** 37-53, 1971.
3. CATTERALL, A.: Coxa Plana. *In* Modern Trends in Orthopaedics, edited by Graham Apley. Vol. 6, pp. 122-147. London, Butterworths, 1972.
4. DANIELSSON, L. G., and HERNBORG, JERKER: Late Results of Perthes' Disease. Acta Orthop. Scandinavica, **36:** 70-81, 1965.
5. EATON, G. O.: Long-Term Results of Treatment in Coxa Plana. A Follow-up Study of Eighty-eight Patients. J. Bone and Joint Surg., **49-A:** 1031-1042, Sept. 1967.
6. EDGREN, WALTER: Coxa Plana. Acta Orthop. Scandinavica, Supplementum 84, 1965.
7. EVANS, D. L.: Legg-Calvé-Perthes' Disease. A Study of Late Results. J. Bone and Joint Surg., **40-B(2):** 168-181, 1958.
8. EYRE-BROOK, A. L.: Osteochondritis Deformans Coxae Juvenalis or Perthes' Disease. The Results of Treatment by Traction in Recumbency. British J. Surg., **24:** 166-182, 1936.
9. GOWER, W. E., and JOHNSTON, R. C.: Legg-Perthes Disease. Long-Term Follow-up of Thirty-six Patients. J. Bone and Joint Surg., **53-A:** 759-768, June 1971.
10. HARRIS, W. H.: Traumatic Arthritis of the Hip after Dislocation and Acetabular Fractures: Treatment by Mold Arthroplasty. An End-Result Study Using a New Method of Result Evaluation. J. Bone and Joint Surg., **51-A:** 737-755, June 1969.
11. HARRISON, M. H. M., and MENON, M. P. A.: Legg-Calvé-Perthes Disease. The Value of Roentgenographic Measurement in Clinical Practice with Special Reference to the Broomstick Plaster Method. J. Bone and Joint Surg., **48-A:** 1301-1318, Oct. 1966.
12. HERNDON, C. H., and HEYMAN, C. H.: Legg-Perthes Disease. An Evaluation of Treatment by Traction and Ischial Weight-Bearing Brace. J. Bone and Joint Surg., **34-A:** 25-46, Jan. 1952.
13. HEYMAN, C. H., and HERNDON, C. H.: Legg-Perthes Disease. A Method for the Measurement of the Roentgenographic Results. J. Bone and Joint Surg., **32-A:** 767-778, Oct. 1950.
14. KAHMI, EDWARD, and MACEWEN, G. D.: Treatment of Legg-Calvé-Perthes Disease. Prognostic Value of Catterall's Classification. J. Bone and Joint Surg., **57-A:** 651-654, July 1975.
15. LARSON, C. B.: Rating Scale for Hip Disabilities. Clin. Orthop., **31:** 85-93, 1963.
16. LEGG, A. T.: The End Results of Coxa Plana. J. Bone and Joint Surg., **9:** 26-36, Jan. 1927.
17. MEYER, J.: Treatment of Legg-Calvé-Perthes Disease. Acta Orthop. Scandinavica, Supplementum 86, 1966.
18. MINDELL, E. R., and SHERMAN, M. S.: Late Results in Legg-Perthes Disease. J. Bone and Joint Surg., **33-A:** 1-23, Jan. 1951.
19. MOSE, K.: Legg-Calvé-Perthes' Disease. Thesis. Copenhagen, Universitetsforlaget, 1964.
20. MOSE, KNUD; HJORTH, LOUISE; ULFELDT, METTE; CHRISTENSEN, E. R.; and JENSEN, ANNA: Legg-Calvé-Perthes Disease. The Late Occurrence of Coxarthrosis. Acta Orthop. Scandinavica, Supplementum 169, 1977.
21. O'GARRA, J. A.: The Radiographic Changes in Perthes' Disease. J. Bone and Joint Surg., **41-B(3):** 465-476, 1959.
22. O'HARA, J. P.; DAVIS, N. D.; GAGE, J. R.; SUNDBERG, A. B.; and WINTER, R. B.: Long-term Follow-up of Perthes' Disease Treated Nonoperatively. Clin. Orthop., **125:** 49-56, 1977.
23. RALSTON, E. L.: Legg-Calvé Perthes Disease — Factors in Healing. J. Bone and Joint Surg., **43-A:** 249-260, March 1961.
24. RATLIFF, A. H. C.: Pseudocoxalgia. A Study of Late Results in the Adult. J. Bone and Joint Surg., **38-B(2):** 498-512, 1956.
25. RATLIFF, A. H. C.: Perthes' Disease. A Study of Thirty-four Hips Observed for Thirty Years. J. Bone and Joint Surg., **49-B(1):** 102-107, 1967.
26. SHARP, I. K.: Acetabular Dysplasia. The Acetabular Angle. J. Bone and Joint Surg., **43-B(2):** 268-272, 1961.
27. STULBERG, S. D.: Legg-Calvé-Perthes Disease: Update. *In* The Hip: Proceedings of the Sixth Open Scientific Meeting of The Hip Society, pp. 263-269. St. Louis, C. V. Mosby, 1978.
28. STULBERG, S. D., and SALTER, R. B.: The Natural Course of Legg-Perthes' Disease and Its Relationship to Degenerative Arthritis of the Hip. A Long-Term Follow-up Study. Orthop. Trans., **1:** 105-106, 1977.
29. WANSBROUGH, R. M.; CARRIE, A. W.; WALKER, N. F.; and RUCKERBAUER, G.: Coxa Plana, Its Genetic Aspects and Results of Treatment with the Long Taylor Walking Caliper. A Long-Term Follow-up Study. J. Bone and Joint Surg., **41-A:** 135-146, Jan. 1959.