LEGG-CALVÉ-PERTHES' DISEASE

A Study of Late Results

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It is nearly fifty years since Calvé (1910), Legg (1910), Perthes (1910) and Waldenström (1909) independently and within a year of each other described an abnormality of the femoral capital epiphysis in children.

This present study was undertaken in order to gain further knowledge of the natural history of this condition as shown by its late results.

In the extensive literature on Perthes' disease relatively few long-term series have been published. The opinions expressed have been conflicting. Møller (1926) reviewed seventy-five hips with healed Perthes' disease—the number of long-term follow-up cases was not recorded. He emphasised that osteoarthritis might develop early, and 21·6 per cent of the patients had significant symptoms of this disorder. Legg (1927) observed forty-six patients for more than ten years. He concluded that though most of the patients would have minor symptoms of this disorder and more than half would show a persistent limp none would ever be incapacitated. Sundt (1949) reviewed 137 cases, both treated and untreated, with a follow-up of ten to fifty-two years. He agreed with Møller that Perthes' disease was not an "innocent disease," but doubted the value of treatment. Mindell and Sherman (1951) reviewed twenty-two patients after sixteen or more years: most patients had some residual defects, but even those with the worst deformity were not severely handicapped. Branciforti and Montana (1954) described the condition of twelve hips after periods averaging twenty-four years, and found that most of them were deformed. Goff, Shutkin and Hersey (1954) stated that every patient followed up for ten or more years and aged twenty or more had symptoms of joint disorder. Helbo (1954) reported fifty-two patients followed for over twenty-five years. Only eight were symptomless, the remainder having some disability with or without signs of osteoarthritis. Ratliff (1956) reviewed fifty hips followed for periods averaging seventeen years. Of these, one-third appeared normal or nearly so, a third had deformed heads but were symptomless and in about a quarter there was great loss of movement, with marked degenerative changes shown radiographically.

CLINICAL MATERIAL

The patients included in this study were all treated at the Royal National Orthopaedic Hospital.

The criteria for their inclusion were a follow-up of ten years or longer from the initiation of treatment, satisfactory clinical records and, when possible, radiographs of the condition at onset, and details of the method of treatment and after-care. Of over 200 patients with Perthes' disease treated more than ten years before, only a third were traced. This disappointing follow-up was mainly explained by movement of the population and failure to trace changes of address. It may also be ascribed to the innate dislike of the average healthy young adult of attending a hospital, particularly in the absence of symptoms. After all cases not fulfilling the above criteria had been discarded fifty-two patients were available for study. No patient with changes in the upper femoral epiphysis occurring during treatment of congenital dislocation of the hip was included. The patients attending for review were examined by the writer and their hips were radiographed.

GENERAL FEATURES

Thirty-nine patients were boys and thirteen girls—a ratio of three to one (Table I). The right hip was affected in twenty-eight patients, the left in twenty-four.

TABLE I
SEX DISTRIBUTION

Age at onset (years)	Number of hips affected	
	Boys	Girls
3-6	21	6
7-14	18	7

There was radiographic evidence of disease of both hips in at least six patients although in only one were the symptoms and signs present on both sides. Only the hip treated is included in this series.

The age at which treatment began varied from three to fourteen years, the commonest age being between five and eight years (Fig. 1).

FIG. 1
Age distribution and results.

The duration of symptoms before treatment was begun varied from one to thirty-six months, the average being six months.

The routine treatment adopted was rest in bed with simple leg traction, but in four patients a plaster-of-Paris spica was employed and in two patients bed rest alone. Three patients were operated upon early, the epiphysis being drilled or grafted, but were otherwise treated with traction. The period of bed rest varied between two and twenty-four months,

FIG. 2

Case 1—Man aged eighteen. Onset at age four years eight months. Right hip affected. Result good. Shows decreased epiphysial height and slight shortening of femoral neck.

FIG. 3

Case 2—Man aged eighteen. Onset at age five years nine months. Result fair. Shows decreased epiphysial height, slight shortening of femoral neck, posterior displacement of head on neck, and trans cervical line.

the average being ten months. In all patients this was followed by the use of a weight-relieving caliper for an average period of fifteen months. The average duration of treatment was therefore ten months' traction, followed by fifteen months in a caliper—a total of twenty-five months.

The follow-up varied from ten to thirty-six years, with an average of sixteen years. The age at the most recent examination varied from fifteen to forty-two years, with an average of twenty-three years.

EXAMINATION

Symptoms—Most patients were symptomless at the time of the investigation; in the rest symptoms were trivial. Symptoms have been classified under the notation described by

FIG. 4

Lateral radiographs of normal femur showing relation of head to neck and variations in neck-shaft angle in neutral position and in medial and lateral rotation.

Shepherd (1954): "none" (thirty-three cases), "ignores" (fourteen cases), "makes concessions" (five cases), "disabling" (no case) and "crippling" (no case). Only in the last five were activities reduced on account of the hip disorder.

Physical signs—A limp was detected in only five patients. Trendelenburg's sign was absent in all. Shortening was observed in twenty-seven patients and averaged half an inch. Some wasting of the quadriceps was present in thirty-one patients.

Movements—Twenty-five hips were fully mobile; fourteen showed 10 or more degrees of limitation of movement in one direction, three in two directions and ten in three or more directions.

The movements most commonly restricted were abduction (seventeen cases), lateral rotation (seventeen cases), extension (thirteen cases) and medial rotation (ten cases). Although some degree of extension was lost in thirteen cases, only one patient showed a flexion deformity.

An interesting finding was that medial rotation was increased by more than 10 degrees in ten hips with normal or nearly normal movements.

FIG. 5

Case 3—Woman aged twenty-one. Onset at age five years one month. Result poor. Shows distorted head set eccentrically on neck.

FIG. 6

Case 4—Man aged twenty-three. Onset at age eight years four months. Result poor. Shows distorted head, well marked subluxation, marked neck shortening and well developed transcervical line.

RADIOGRAPHIC EXAMINATION

Antero-posterior views of both hips on the same film, and sitting lateral views of each hip, were taken in every case.

Shape of femoral head—The shape of the femoral heads as seen in both projections conformed broadly to three categories, which have been called good, fair and poor.

Good heads (fifteen cases)—The outline was circular in both projections, the head was set

squarely on the femoral neck and the acetabulum was normal (Fig. 2). None of these hips showed any evidence of osteoarthritis, and without comparison with the opposite side would have passed as normal. In each case, however, the comparison revealed at least one of the following stigmata of pre-existing Perthes' disease: a large head, decreased height of the epiphysis, slight subluxation, shortening or broadening of the femoral neck.

Fair heads (twenty-one cases)—The outline was elliptical rather than circular in either one or both projections, but smooth and regular (Fig. 3). The acetabulum was well adapted and the head well covered. In none was there evidence of osteoarthritis.

The lateral projection of a normal head shows it set centrally on the neck whatever the degree of rotation (Fig. 4). A frequent feature of these fair heads was an eccentric position on the neck, the head appearing displaced backwards and its posterior surface even ending in an edge.

Poor heads (sixteen cases)—The outline was deformed in both projections. Though in eleven cases some regularity was preserved (Figs. 5 to 7), five heads were grossly deformed, the head

FIG. 7

Case 5—Man aged thirty-two. Onset at age eleven years ten months. Result poor. Shows distorted head, short neck and osteoarthritis with loose bodies.

appearing square in the antero-posterior and triangular in the lateral projection (Figs. 8 and 9). All showed varying degrees of osteoarthritis and in all the heads appeared displaced backwards on the neck.

Size of head—Four of the fifteen good heads were enlarged, and all misshapen heads were broader than normal.

Subluxation—Waldenström (1938) described subluxation of the hip as an early radiographic feature of Perthes' disease. This had persisted in forty-two hips. Whereas in good and fair hips it was usually minimal and often easier to see than measure, it was more pronounced in poor hips. Jonsäter (1953) concluded from arthrographic studies that increase in the head-socket distance was due not to thickening of the joint cartilage but to an inflammatory swelling of the ligamentum teres and soft tissues in the acetabular fossa which pressed the head laterally.

Shaft-neck angle—This angle was increased in twenty-five and decreased in nineteen hips compared with the normal side. Since the angle can vary with rotation by up to 25 degrees, a difference of less than 10 degrees was considered insignificant. Six hips showed valgus of this amount, but no hip showed this degree of varus.

FIG. 8

Case 6—Man aged thirty-three. Onset at age fourteen years eight months. Left hip: Result poor. Shows gross distortion with squaring in antero-posterior view and triangulation in lateral view, with evidence of osteoarthritis. Right hip: Result fair (untreated).

Anteversion of femoral neck—Anteversion is notoriously difficult to measure. In a lateral radiograph of a normal hip in neutral rotation the angle between the neck and shaft represents the angle of anteversion. Though rotation affects this (Fig. 4), if both hips lie in similar rotation the radiograph will give a qualitative though not a quantitative indication of anteversion. The relation of the greater trochanter to the femoral neck is a useful guide in assessing the similarity of rotation.

Thirty-one out of the thirty-nine hips where rotation was comparable showed an increased angle of anteversion. Though increased anteversion of more than 10 degrees seldom occurred in good hips, it was frequently observed in fair and poor hips. This confirms the finding of Dunlap, Shands, Hollister, Gaul and Streit (1953) that increased anteversion is a common late radiographic finding in Perthes' disease.

Breadth of femoral neck—The breadth of the femoral neck at its narrowest section was normal in seventeen cases, increased in twenty-three, decreased in five and not measurable in the remainder. The greatest increase occurred with the most deformed heads.

Length of femoral head and neck—The frequent deformity of the head and neck precluded measurement of the true neck length. Measurement of the distance from the cortex of the head to that of the shaft in the line of the centre of the neck, however, showed this to be diminished in all but six cases. When the epiphysal scar was still visible the distance between this scar and the shaft cortex was increased in five cases, decreased in seventeen and normal in five. Reduction in head-neck length is thus caused in part by decrease in the height of the

FIG. 9

Case 7—Man aged thirty-three. Onset at age thirteen years 6 months. Left hip affected. Result poor. Shows gross distortion in both views, with evidence of osteoarthritis.

epiphysis—an almost invariable finding—and also frequently by diminished metaphysial growth. The latter, however, was sometimes unaffected and occasionally increased.

Marked shortening of the head-neck segment is shown clinically by elevation of the greater trochanter.

Transcervical line—The transcervical line, a common feature of late radiographs, represents the rim of the distorted head (Fig. 6). It was observed invariably in poor hips, occasionally in fair, and never in good hips.

RESULTS

The results of treatment have been assessed solely on the final shape of the femoral head, but symptoms and mobility have been correlated with this (Figs. 10 and 11).

Good (fifteen cases)—Thirteen patients had no symptoms and two ignored them. Thirteen had full mobility and two had restriction of movement in one direction only.

Fair (twenty-one cases)—Sixteen patients had no symptoms, three ignored them and two made concessions on this account. Eleven patients had full mobility, nine had limitation of movement in one direction and one had limitation in two directions.

Poor (sixteen cases)—Four patients had no symptoms, nine ignored them and three made concessions on this account. One patient had full mobility, three patients had limitation in one direction, one patient in two directions, and eleven patients in three or more directions.

Thus with a well shaped head pain was virtually absent and mobility almost always full. With a fair head most patients were free from pain, and mobility was full or nearly so, whereas with a poor head pain of some degree was usual and mobility was nearly always markedly decreased. Clinical features therefore closely paralleled the radiographic.

FIG. 10
Correlation of pain with shape of femoral head.

FIG. 11
Correlation of mobility with shape of femoral head.

DISCUSSION

Factors influencing the prognosis. *Age*—The age at onset of the disease is important (Fig. 1). Except in one case, a good result did not occur with an age at onset of over six, and with one exception the result was always poor if the age at onset was over eight years. *Sex*—Whereas three-quarters of the boys developed good or fair heads and one-quarter poor heads, less than half the girls obtained good or fair heads and over half had poor heads (Table II).

TABLE II
RESULTS RELATED TO SEX OF PATIENT

	Good	Fair	Poor
Boys	12	18	9
Girls	3	3	7

It thus appears that the prognosis is worse in girls, a finding that may be related to their earlier skeletal maturation.

There were insufficient series of early radiographs to determine the influence of the stage of evolution of the disease at the onset of treatment on the prognosis.

FIG. 12

FIG. 13

Case 8—Man aged twenty-six. Onset at age six years two months. Figure 12—Condition at onset: density of epiphysis, changes in epiphysal line, increased joint space and subluxation. Figure 13—At five months (a week after drilling and insertion of tibial graft). Revascularisation is commencing.

Early radiographic changes—The cycle of epiphysal changes—increased density, flattening, fragmentation and final restoration to normal texture—is a well known characteristic feature, at least in the younger patients.

Certain other features were observed. The first evidence of epiphysal flattening often occurred on the superior and lateral surfaces in the antero-posterior view, and always on the anterior surface in the lateral view. This corresponds to the main weight-bearing area of the head. The earliest evidence of revascularisation always occurred anteriorly, and most commonly laterally, sometimes medially and occasionally superiorly.

FIG. 14

FIG. 15

Case 8. Figure 14—At eleven months (six months after graft). Revascularisation is proceeding proximally. Figure 15—At nineteen months (fourteen months after graft). Revascularisation almost complete.

FIG. 16

Case 9—Man aged twenty-nine. Onset at age six years six months. Right hip: result fair. Left hip: result fair (untreated).

FIG. 17

Case 10—Man aged thirty-three. Onset at age eleven years four months. Left hip: result poor. Right hip: result fair (untreated).

FIG. 18

Case 11—Man aged twenty-six. Onset at age seven years nine months. Result fair. Shows osteochondritis dissecans.

An antero-posterior radiograph did not give a true indication of the changes taking place in the ossific nucleus as a whole. Gross changes visible in the lateral projection were compatible with a nearly normal antero-posterior appearance.

Metaphysial changes were very variable. Though some degree of rarefaction and cystic change was common, marked rarefaction extending well into the neck often occurred. The changes, however, were of short duration; they seldom persisted for more than a year and did not appear to affect the natural progression of the disease or the late result.

Enlargement of the epiphysis and metaphysis was constant in the early stages, giving the appearance of a large head; but this inequality between the two sides usually diminished in the ensuing years.

Effect of operation—Of three patients subjected to operation by drilling and grafting, two were six and one was ten years old at onset. The two former obtained a good result and the latter a bad result. Serial radiographs of one of these showed an alteration in the usual course of the disease (Figs. 12 to 15). After the operation there appeared to be a gradual creeping revascularisation occurring proximally from the epiphysial plate. Howorth (1948) reported that early operation of this type considerably hastened revascularisation.

Affection of both hips—On radiographic examination six patients were found to have had Perthes' disease of the untreated hip. Two were mirror images of the treated hip, one good and one poor (Figs. 16 and 17). Four occurred as the pair to a square type of poor hip and were graded fair (Fig. 8). Sundt (1949) reported that each of his five cases of "quadrangular" head showed minor degrees of subluxation of the opposite side.

Of these affected but untreated hips only one had caused symptoms. Whereas two were radiographically like their treated counterpart, four were much better. There is no evidence, however, of when these contralateral changes occurred or whether the evolution of the disease in these hips was typical.

Osteochondritis dissecans—One case of osteochondritis dissecans was encountered (Fig. 18). The separated fragment was not visible as a distinct entity until at least five years after the onset of the disease, which otherwise passed through the usual changes. The separated fragment was situated on the summit of the head. Ratliff (1956) reported two such cases.

Osteoarthritis—All poor hips showed radiographic evidence of degenerative arthritis of varying severity. The femoral heads were severely deformed, and the diagnosis of osteoarthritis secondary to Perthes' disease was obvious.

The fair heads approximated more closely to good than poor, but they were definitely abnormal. Even good heads bore certain stigmata of Perthes' disease and were therefore imperfect. It seems probable that in time some of these hips will develop degenerative changes, and may well account in later life for some cases of so-called idiopathic or primary osteoarthritis.

SUMMARY

1. Fifty-two patients with Perthes' disease (affecting both hips in six instances) have been reviewed ten or more years after the beginning of treatment.
2. Judged radiographically, approximately one-third developed good, one-third fair and one-third poor femoral heads.
3. The clinical results paralleled the radiographic. Except with the worst shaped heads, function was excellent.
4. Certain constant early and late radiographic features are recorded.
5. Of the factors influencing prognosis, the age at onset of the disease and the sex of the patient appear to be important.

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