The aim of this paper is to determine the late results of idiopathic pseudocoxalgia (Legg-Calvé-Perthes disease) in adult life. It is hoped to obtain some idea of the efficiency of treatment and of the factors that influence prognosis.

In the enormous literature of this condition, only three papers can be found which contain reports of cases which have been observed for long periods. Legg (1927) reviewed forty cases for ten or more years. He divided these cases into two types, the "cap" and the "mushroom." He stated that: "Ultimately the cases of both types will never be incapacitated. In the majority they will tire after long standing and walking, or after movements that require extreme abduction. More than half the cases will probably show a persistent limp." Unfortunately he gave neither details nor case histories. Mindell and Sherman (1951) reviewed seventy-eight hips in seventy-two patients, but only twenty-two hips were observed for sixteen years or more. They stated that most patients had some residual effects, but even those with the worst results were not severely handicapped. They grouped the immediate and late results together, and it is therefore difficult to compare their results with those presented in this paper. On the other hand, the third paper, that of Branciforti and Montina (1954) (a study of twelve hips followed up for an average of twenty-four years) concluded that the fate of these hips is a severe one "leading us to erase from our minds all that we have ever read about osteochondritis of the hip."

A REVIEW OF FIFTY AFFECTED HIPS

The material for this study is based on fifty hips observed for a period between eleven and thirty years (average period of follow-up being seventeen years). Most of the patients studied had been treated at Biddulph Grange Orthopaedic Hospital, a few at the Robert Jones and Agnes Hunt Orthopaedic Hospital, the Ethel Hedley Hospital, the Royal Hospital for Sick Children, Edinburgh, and the Princess Margaret Rose Hospital. In all cases the diagnosis was definite, the radiographic findings in childhood and method of treatment known, and all were seen and radiographed in adult life. Every case that could be obtained has been included and no selection employed. Thirty-eight of the hips had been treated by fixation on frames; three had been immobilised in hip spica plasters. I have not attempted to differentiate between the result of these methods. Nine received no treatment during the active phase of the disease, though a few of these received some treatment too late, when the femoral head was already partly reformed. These are classified together as "untreated" cases.

ASSESSMENT OF RESULTS IN ADULTS

Many patients had no complaints and their hips showed full movement, despite varying degrees of deformity. These cases were difficult to classify, and therefore a point system was employed, based on that used by Muller and Seddon (1953) in assessing cases of congenital dislocation of the hip. Every patient was given a maximum of five points for each of pain, activity, movement, and radiographic appearances, and the points were awarded as shown.

Pain (P)
1. Severe.
2. Severe on movement; little at rest.
4. Slight ache after prolonged standing; does not limit activity.
5. None.

Activity (A)
1. Walks short distances only, with difficulty.
2. Walks half a mile.
3. Walks two miles.
4. Walks long distances, but cannot do heavy work or play games.
5. Normal.

Movement (M)
1. Little or none.
2. Less than 50 per cent; fixed deformity.
3. More than 50 per cent; no fixed deformity.
5. Full.

Radiographic appearance (X)
1. Gross evidence of osteoarthritis, with severe loss of "joint-space."
2. Completely distorted head, only partly contained; no neck; occasionally great trochanter above level of the head; "joint-space" good.
3. Very flattened head, incompletely contained; very short neck; occasionally early sclerosis of the acetabulum.
4. Slightly flattened head; fully contained; slightly short neck.
5. Normal or almost normal appearance.

A normal joint would thus be described as P5, A5, M5, X5, scoring a total of 20 points. Each joint examined was allotted marks in this way, and hence they were finally classified into four groups:

<table>
<thead>
<tr>
<th>Description</th>
<th>Points</th>
</tr>
</thead>
<tbody>
<tr>
<td>Good</td>
<td>18</td>
</tr>
<tr>
<td>Fair</td>
<td>17 to 15</td>
</tr>
<tr>
<td>Poor</td>
<td>14 to 11</td>
</tr>
<tr>
<td>Very poor</td>
<td>10 or less</td>
</tr>
</tbody>
</table>

This classification is demanding. It expects a hip to obtain 18 points out of 20 to be classified as good. But all the patients were less than forty years old, and it was felt that those with appreciably deformed hips could not be placed in the "good" class, even though they had no pain and normal activity.

RESULTS

The general results are illustrated in Table I.

Two general conclusions are seen from examination of these results. First, in cases of pseudocoxalgia followed into adult life, rather more than one-third were good, the same number were fair, and about one-quarter were poor. Second, though three-quarters had no pain and normal activity, only two-fifths had good hips radiologically.

Radiological features in the adult—Legg divided his cases into two types, the "cap" and the "mushroom." In the former type there was simple flattening of the head; in the latter there was spreading out as well. In this series about half the total (twenty-six) showed appreciably deformed hips (X3 or less). The majority were of the mushroom type, and only six were cap-shaped. A characteristic pattern of deformation of the femoral head and neck was found based on the degree of flattening of the head and of shortening of the neck, and this is detailed in the description of the points classification. Noteworthy features, which so far as I am aware have not been detailed previously, are the high incidence of gross neck shortening (see later)
and the frequent early occurrence of sclerosis of the upper lateral aspect of the acetabulum. The cap types were characterised by an early sclerotic appearance of the upper aspect of the head, and marked sclerosis of the adjacent surface of the acetabulum. The numbers observed are not sufficient to merit discussion of the prognostic significance of the two types. In adult hip surgery the diagnosis of "old healed Perthes' disease" is frequently encountered, but this is usually presumed rather than proved. Examples of some of these stages of deformation are shown in Figures 1 to 6.

### TABLE I

**A Review of Fifty Hips in Forty-six Patients**

<table>
<thead>
<tr>
<th>Details</th>
<th>Untreated</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pain</td>
<td>None or slight aches</td>
</tr>
<tr>
<td>Activity</td>
<td>Normal</td>
</tr>
<tr>
<td>Movement</td>
<td>Full or terminal limitation</td>
</tr>
<tr>
<td>Radiographs</td>
<td>Normal or slightly flattened</td>
</tr>
</tbody>
</table>

**THE VALUE OF TREATMENT**

A discussion of the value of different types of treatment is outside the scope of this paper, for in this series (with few exceptions) only one method was employed, namely frame fixation. A comparison of the results of treated and untreated cases is shown in Table II. Only one out of nine untreated cases gave a good result. This series, though small, suggests that treatment is worth while.

### TABLE II

**The Value of Treatment (Fifty Hips)**

<table>
<thead>
<tr>
<th>Result</th>
<th>Treated</th>
<th>Untreated</th>
</tr>
</thead>
<tbody>
<tr>
<td>Good</td>
<td>18</td>
<td>1</td>
</tr>
<tr>
<td>Fair</td>
<td>15</td>
<td>4</td>
</tr>
<tr>
<td>Poor and very poor</td>
<td>8</td>
<td>4</td>
</tr>
<tr>
<td></td>
<td>41</td>
<td>9</td>
</tr>
</tbody>
</table>

**COMPLICATIONS AND SEQUELAE**

This series of cases was free from general complications of prolonged decubitus, such as renal calculi. The following were among the local complications that developed:

- **Osteoarthritis**—The term osteoarthritis is used to describe anatomically degenerate hips (X3 or less) with limitation of movement.

Osteoarthritis may follow pseudocoxalgia in two quite different circumstances. In the adult it may develop years after treatment, being apparently due to incongruity between the articular surfaces of the femoral head and the acetabulum. Both adjacent articular surfaces become flattened and irregular, inviting degenerative change. In this series there were twenty-five hips (one half) with some degree of osteoarthritis. In the majority this was mild and a radiological finding in a joint that was painless. There was gross osteoarthritis with
An almost normal left hip in a woman of twenty-eight after pseudocoxalgia when aged seven. Note the slight elevation of the lesser trochanter (graded X5).

A left hip showing a slightly short neck and a slightly flattened but fully contained head (graded X4). Pseudocoxalgia eighteen years earlier at the age of eight.
loss of joint space, subluxation and pain in five (Fig. 5). The earliest and most common radiologica"
Osteochondritis dissecans—The etiology of osteochondritis dissecans of the hip is obscure. King and Richards (1940) stated that it was due to an aseptic necrosis of a localised segment of subchondral bone. Magnin (1931) had previously stated that no relationship between this condition and pseudocoxalgia existed. He regarded the evolution and treatment of the two affections as entirely dissimilar. In this series there were two cases of osteochondritis dissecans following pseudocoxalgia.

The first was in a boy aged ten, admitted to hospital in 1936 with left pseudocoxalgia. He had previously had twelve months' immobilisation in a hip spica at a local hospital, and then had been allowed to walk freely for a further nine months. Radiographs in 1936 (Fig. 11) showed a healing pseudocoxalgia, with the head slightly flattened, and a localised area of dense sclerosis in the centre of the head towards the supero-lateral aspect. He was treated by a further seven months in a hip spica in hospital. Follow-up radiographs in 1941 and even in
Figure 5—A right hip showing gross osteoarthritis (X1). Pseudocoxalgia twenty-one years earlier at the age of thirteen. Figure 6—A left hip showing the less common "cap" type of deformity with flattening of the femoral head but very little spreading out. Note the sclerosis of the head (X3). Pseudocoxalgia fifteen years earlier at the age of five.

Figure 7—A right hip showing healing pseudocoxalgia in a child aged eleven at the end of twenty-one months' immobilisation in a hip spica. Figure 8—Five years later, the hip already shows some loss of joint space with subluxation. There was gross restriction of all movements and an adduction and lateral rotation deformity. The hip was painful. Movement had never recovered well after immobilisation.
Radiograph of a child aged eight immediately at the end of eleven months' frame fixation. Note the broad, slightly shortened neck.

The same patient seventeen years later, with no femoral neck and the greater trochanter above the level of the head.
FIG. 11
Radiograph showing a healing left pseudocoxalgia in a boy of ten, with the head slightly flattened, and a localised area of dense sclerosis in the centre of the head towards its supero-lateral aspect.

FIG. 12
Antero-posterior and lateral radiographs eighteen years later showing a clearly outlined area of osteochondritis dissecans.
FIG. 13
Radiograph when aged seven, showing a flattened irregular left femoral head with a dense area of sclerosis in its centre.

FIG. 14
Radiographs of the same patient twenty years later showing an area of osteochondritis dissecans.
1954 showed a clearly outlined area of osteochondritis dissecans (Fig. 12). Then, at the age of twenty-eight, he was working as a carpenter.* He suffers an occasional slight ache in the hip, but no interference with work or activity. There was slight wasting of the left thigh, but no limp. There was a full range of painless movement in both hips. No loose body has as yet separated.

The second patient, a boy of seven, was admitted to hospital in 1933 with left pseudocoxalgia. Radiographs (Fig. 13) showed a markedly flattened, irregular head with a dense area of sclerosis in its centre. He was treated by immobilisation on a frame for nine months. When seen recently he was aged twenty-six and was doing work involving long hours and heavy lifting. There were occasional cramp-like sensations in the left hip, but these caused him no disability. He had no limp, and movements were full except for terminal limitation of abduction. Radiographs (Fig. 14) showed a sclerotic "cap" type of head and in the lateral view a clearly outlined area of osteochondritis dissecans. There was no loose body formation.

It seems probable that a localised segment of the femoral epiphysis lost its blood supply for ever in the active phase of the disease. This occurrence of osteochondritis dissecans following pseudocoxalgia is evidence in support of the avascular origin of both conditions.

FACTORS INFLUENCING THE OUTCOME OF PSEUDOCOXALGIA

There is general agreement that the future of a hip afflicted with avascular necrosis of the capital femoral epiphysis will depend upon the amount of deformation which has occurred in that epiphysis by the time the disease is recognised. Perhaps the most important single factor in the outcome of the disease is its natural history in the particular patient afflicted. There appears to be a wide variation between abortive cases in which there is no severe flattening and fragmentation, and severe cases in which the restoration of the shape of the epiphysis appears to take several years. This disease follows no time-table. In most cases its evolution takes about two years, but in many this time may be considerably exceeded.

Apart from such obvious considerations, the future of the hip may depend upon two other factors, the age at the onset of the disease and the effect of the disease upon the growth of the femoral neck. The former was investigated by Eyre-Brook (1936) and by Møller (1926). The age at the onset of the disease—The effect of age upon the outcome of the disease is illustrated in Table III and Figure 15.

It can be seen that in the ten hips in which the disease developed in children aged five or less, all had good results. Conversely, in the nine hips in which the disease developed in children aged ten or more, none had a good result. The younger the child, the better the prognosis.

Failure of femoral neck growth—This has already been discussed. It was quite unpredictable and I was unable to confirm the statement of Mindell and Sherman (1951) that it was more likely to occur when there was evidence of severe femoral neck disturbance. Failure of femoral neck growth is an important cause of the high percentage of poor radiological appearances in the adult.

These factors may be clarified by examining the common features of each class of end-result.

In the hips classified as good, the condition had often developed in a child under seven years of age; fifteen out of nineteen had only slight flattening of the upper femoral nucleus when treatment was commenced; in none was there fragmentation. The cycle was often of short duration, the average period of immobilisation only thirteen months.

In adult life these nineteen patients had no symptoms and good radiological appearances. An example is shown in Figures 16 to 19.

* I am indebted to Mr E. F. West for his kindness in reporting to me the present state of this patient, who has emigrated to Australia.
In the "fair" group the typical picture was that of a more severe disease in an older child. One half of these hips were grossly fragmented at the beginning of treatment. The average duration of immobilisation was longer—sixteen months—and several epiphyses were

TABLE III

<table>
<thead>
<tr>
<th>Age at onset of disease (years)</th>
<th>Results</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Good</td>
</tr>
<tr>
<td>3</td>
<td>1</td>
</tr>
<tr>
<td>4</td>
<td>4</td>
</tr>
<tr>
<td>5</td>
<td>5</td>
</tr>
<tr>
<td>6</td>
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<td>7</td>
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<td>9</td>
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<tr>
<td>11</td>
<td>-</td>
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<tr>
<td>12</td>
<td>-</td>
</tr>
<tr>
<td>13</td>
<td>-</td>
</tr>
</tbody>
</table>

The effect of age upon the prognosis.

not fully reformed at the end of treatment. In the adult these nineteen patients had no symptoms but all these hips had poor radiological appearances (Figs. 20 to 23).

The common feature of the twelve poor and very poor results was that they were all
Figure 16—Photograph on admission to hospital at the age of seven. Note the well marked limitation of abduction of the left hip. Figure 17—Radiograph at the same time showing a typical pseudocoxalgia with only mild flattening of the head and no fragmentation.

Figure 18—Same patient. Radiographs after fifteen months' frame fixation. The femoral epiphysis has reformed fully with only mild flattening. Figure 19—Radiographs at the age of twenty-six showing an almost normal left hip.
Figure 20—Photograph on admission when aged nine. Pseudocoxalgia of left hip. Figure 21—Radiograph at the same time showing gross flattening and fragmentation of the left femoral epiphysis.

Figure 22—Same patient. Radiograph after twenty-two months’ frame fixation, showing a fairly well reformed but considerably broadened femoral head and metaphysis. Figure 23—Radiograph at the age of twenty-five showing a very flattened head, incompletely contained, and a very shortened neck. There was normal activity and no limp, but terminal limitation of all movements.
poor from childhood. Three developed degenerative changes immediately after treatment, and these led rapidly to stiff, painful hips (Figs. 7 and 8). Seven had gross flattening of the upper femoral nucleus when first seen, and four had had no treatment at all. Three showed failure of femoral neck growth (Figs. 9 and 10). In the adult this was the only group which presented symptoms.

SUMMARY AND CONCLUSIONS

A review of fifty cases of idiopathic pseudocoxalgia (Legg-Calvé-Perthes disease) followed into adult life for periods of eleven to thirty years (average seventeen years) after diagnosis shows:

1. In the whole series rather more than one-third of the patients developed hips which were normal or nearly normal.
2. An equal number had hips which could only have been considered “fair.”
3. About one quarter had hips which gave pain and which showed marked loss of movement and gross degenerative changes radiologically.

Judged from the point of view of symptoms, the results were better than the foregoing would suggest. Three-quarters of the patients were fully active and free from pain but only two-fifths had hips which were radiologically good. It is possible that such apparently good results are unlikely to be permanent, and I hope, therefore, to continue this follow-up for another ten or fifteen years. It can, however, be concluded at this stage that an immediate good result is likely to be maintained at least until the age of twenty-five years, even though half of such patients will have radiologically abnormal hips.

There is a characteristic pattern of deformation of the femoral head and neck in the adult resulting from this condition in childhood, based on the degree of flattening of the head and shortening of the neck.

The end-results are better in adequately treated cases.

Two cases are reported in which osteochondritis dissecans developed as a late complication.

My sincere thanks are due to the members of the consultant staffs of the hospitals in which the patients were treated, for permission to study their cases and their records. I am also very grateful to Sir Harry Platt for his constant encouragement, to innumerable colleagues who helped me in tracing patients, and to Professor Sir Walter Mercer, Mr D. Li. Griffiths, and Mr D. L. Savill for their help and criticism in the preparation of this paper.

REFERENCES


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