The long-term outcome of primary osteochondritis of the hip (Legg-Calvé-Perthes’ disease)

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We have previously reported on 57 patients (60 hips) with a past history of Legg-Calvé-Perthes’ disease at a mean of 34 years after the onset of symptoms. From this original group, 48 patients (51 hips) were also available for review after a mean of 50.2 years. We consider that the best prognostic indicator for the hip is the shape of the femoral head at skeletal maturity. Normal or flattened spherical heads present few problems. Irregular or very irregular heads are associated with a poor outcome.

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An understanding of the long-term outcome of Legg-Calvé-Perthes’ disease (LCPD) is important for two reasons. The first is the assessment of the risk that affected hips may develop osteoarthritis in later life and the second is the definition of the prognostic criteria at skeletal maturity which will make it possible to predict this risk. There are few long-term statistics because of the difficulties in finding these patients after a prolonged period. The studies of Ratliff, Mose, Mose et al and others are therefore of considerable value, particularly the repeated updates, as also is the impressive review of published series undertaken by Kohler and Seringe. The findings of a follow-up for 50 years of 51 patients are now presented.

Patients and Methods

In 1987, the findings were reported of 57 patients (60 hips) who had been treated for LCPD more than 30 years earlier. Their mean age was 42 years (33 to 56). The mean follow-up was 34 years (30 to 44) from the onset of the disease.

Examination of these patients 15 years later gave a study population of 48 patients (51 hips) suitable for assessment with a mean age of 57.4 years (47 to 64) and a mean follow-up of 50.2 years (44 to 58). For this longer-term study, eight patients were seen, re-examined, and radiographs taken, 29 (32 hips) replied to a detailed questionnaire and radiographs were sent to the author, nine replied only to the questionnaire but refused to provide radiographs and two had died, although the condition of the hip could still be established from either the family or the general practitioner.

All patients had been treated identically. Children were hospitalised and treated by bed rest, with or without traction. Immobilisation in plaster was not used and no patient underwent surgery as a child. The mean length of hospital stay was 21 months (3 to 45). The total time in hospital was often longer as some children had previously been hospitalised elsewhere. The age at the onset of the disease, functional signs (pain, limp, limitation of movement) including the Merle d’Aubigné and Postel score, subjective signs (the patient’s own assessment of the hip and how it affected everyday life), and clinical examination (range of movement, leg-length inequality, muscle wasting) were recorded.

Radiographs were also taken, the hips being graded using Mose’s system, according to the sphericity of the femoral head as measured by a Curtis coxometer. Patients were divided into four groups as follows: normal head, flattened spherical head (1 to 2 mm), irregular head (2 to 4 mm), and very irregular head (>4 mm). The assessment of minimal defects is difficult and it is therefore likely that the group with a flattened spherical head also includes some with a normal head so that the numbers may be artificially increased. As well as sphericity the radial quotient (RQ), the centre-edge (CE) angle and Edgren’s articulotrochanteric distance (ATD) were recorded. These were expressed as a percentage of the normal side. Finally, signs of osteoarthritis were sought although these were not always obvious with very deformed femoral heads.

Results

34-year follow-up. Of the 60 hips (57 patients) studied, 23 were free from pain, seven were mildly painful, 19 were moderately painful and 11 were very painful. Six patients had a marked limp and 16 a slight limp. Limping without pain was seen in two patients. Eight patients had stiffness
of the hip in everyday activities. A total of 46 hips had not required further medical attention after the initial treatment, although 14 had caused sufficient discomfort to warrant a consultation. Recommended treatments had been simple rest (one hip), medical treatment (six hips), or surgery (seven hips). The mean age of the operated patients was 41.5 years (31 to 47). Surgery had been a Chiari osteotomy (two hips), shelf osteotomy (two hips) or a total hip replacement (THR) in three hips.

Two patients, both free from symptoms, underwent preventive operations at skeletal maturity. One patient had a shelf osteotomy but required a total hip replacement at the age of 44 years. The other had a varus femoral osteotomy, although this did not prevent the subsequent development of osteoarthritis.

On direct questioning of the patients, 33 hips felt normal, 17 caused moderate discomfort and ten severe discomfort. Most hips (38) allowed the patients to lead normal lives although one patient had changed his job as a furniture remover. Nine hips had caused problems during military service. The patients had either been medically discharged or excused from marching. Eight patients had either abandoned sport or had been unable to play at all.

Radiological examination revealed seven normal femoral heads, 26 flattened spherical femoral heads, seven irregular femoral heads and 20 very irregular femoral heads. Four hips showed significant architectural abnormalities although the femoral head was only slightly deformed. There were obvious signs of osteoarthritis in 12 hips and six had severe osteoarthritis.

50-year follow-up. In the 51 hips (48 patients) the situation had clearly deteriorated as 12 had required a THR. The mean age at replacement was 53.8 years (44 to 65) and one patient had died; 39 hips had not required a THR, although one patient from this group had also died. Of the survivors, 21 were free from pain, eight experienced mild pain and nine experienced severe pain. Two patients walked with a stick, 11 walked with a limp, and four had a limited walking distance. Six patients were taking regular analgesia or anti-inflammatory medication. The Merle d’Aubigné and Postel score was used to assess the function of the 38 surviving unoperated hips. The results were excellent (18) in 23 hips, very good (17) in eight, good (16) in one, fair (13 to 14) in four and poor (9 to 12) in two (Table I).

Subjectively, 25 hips were considered to be normal, eight caused moderate discomfort and five caused severe discomfort. Radiological examination revealed signs of osteoarthritis in seven hips but none in 25. Six patients did not wish to undergo radiological examination.

Influence of the shape of the femoral head. The shape of the femoral head at skeletal maturity appeared to determine later function.

Normal heads

34-year follow-up. There were few problems. For these patients (seven hips), the mean age at the onset of the disease was five years (3 to 7). Six patients lived a normal life and one had been excused military service. None had sought medical advice. All considered their hips to be normal. Three had a leg-length discrepancy of between 5 and 10 mm. No patient had radiological evidence of osteoarthritis. As percentages of the normal hip the mean RQ at maturity was 103% (100 to 112), the mean CE angle was 99% (80 to 121) and the mean ATD was 92% (76 to 127).

50-year follow-up. Of the four patients (four hips), one had died but the hip was free from pain at the time of death. The remaining three patients (three hips) had a mean age of 53.4 years and a mean time to follow-up of 47.7 years. Two had occasional discomfort in the hip, but did not take analgesia, did not limp, had an unlimited walking distance and excellent mobility. The Merle d’Aubigné and Postel score was 17 and 18, respectively. The third patient was free from pain.

Radiographs showed no osteoarthritis in any of the affected hips.

Flattened spherical heads

34-year follow-up. For these 26 patients the mean age at the onset of the disease was 6.3 years (3 to 11). Of the 26 hips, 19 were free from pain, three caused mild pain and four moderate pain. Two patients had a slight limp. Although two did not play sport, 20 patients lived a normal life. Two had been exempted from military service (but did play sport) and two had been exempted only from marching. No patient had sought medical advice for the hip. Two hips caused occasional discomfort, but 24 were considered to be normal. Clinical examination revealed nine patients with a leg-length discrepancy (5 to 15 mm) and six with minor limitation of rotation. Radiological examination did not show any osteoarthritis of the hip. As percentages of the normal hip the mean RQ was 105% (93 to 126), the mean CE angle was 100% (46 to 138) and the mean ATD was 79% (26 to 129).

50-year follow-up. For this group of 23 patients, the mean time to follow-up was 50.2 years and the mean age of the patients was 56.2 years. Surgery had not been required for 21 hips; 17 were free from pain and four caused occasional discomfort. All were functionally excellent (Fig. 1). All patients had an unlimited walking distance and no limp. The Merle d’Aubigné and Postel score was 18 (excellent) in 19 hips and 17 (very good) in two hips. Two patients had undergone THR. The first had a THR at 61 years of age on the affected side and at 65 years of age on the unaffected side. The second had a THR at the age of 47 years for a painful hip but without radiological evidence of osteoarthritis. Of the 21 hips which had not required surgery, none had required analgesia and 20 appeared normal. One patient (one hip) had moderate discomfort. No radiograph showed osteoarthritis, although three patients refused to undergo radiological examination.

Irregular heads

34-year follow-up. There were more problems in this group of seven patients. The mean age of the onset of the disease
was 9.5 years (8 to 12). Seven hips caused pain. In four, this was moderate and in three severe. It had begun at a mean age of 32 years (17 to 38). Six patients had a slight limp and all felt some stiffness. Three hips had already necessitated medical advice and one had required a proximal femoral osteotomy in a patient aged 16 years because of radiological signs of degeneration although the hip was painless.

One patient had to change his job as a furniture remover, one had been exempted from military service and two reported a change in athletic ability. No patient felt that they had a normal hip. Five patients considered their discomfort to be moderate and two severe. Five patients had a leg-length discrepancy of between 10 and 25 mm and five limitation of hip movement which was severe in two. There was mild radiological evidence of osteoarthritis in five hips and severe osteoarthritis in one. The mean irregularity of the femoral head was 3.9 mm (3 to 4). Compared with the normal sides the mean RQ was 115% (103 to 128), the mean CE angle 76% (55 to 103) and the mean ATD 47% (5 to 77).

50-year follow-up. For these seven patients the mean follow-up was 49.1 years and the mean age 57.9 years. There had been significant progression in the condition of the affected hip in all patients. By a mean age of 53.2 years, three hips (three patients) had required a THR, including one in the patient who had previously undergone proximal femoral osteotomy. One hip had needed a shelf osteotomy in a patient aged 39 years. Four hips had not required a THR. Of these, all caused pain, severe in one hip, moderate in two and mild in one. Three patients limped, required a walking stick and had a limited walking distance. The Merle d’Aubigné and Postel score was very good (17) in one hip, fair in two and poor in one.

Of those hips which had not needed a THR, one caused moderate discomfort and three severe discomfort. Two patients required regular analgesia. Three hips showed radiological evidence of severe osteoarthritis. The fourth showed no radiological evidence of osteoarthritis.

Very irregular heads

34-year follow-up. For this group of 20 patients, the mean age at the onset of the disease was 8.5 years (5 to 13). Pain was moderate in ten hips, severe in eight and mild in two. The mean age at the onset of pain was 31 years (14 to 41) although for those with severe pain this began after the age of 30 years. Limping was severe in four patients and minimal in ten. Stiffness and limitation of movement were seen in 14 patients. Of the 11 patients who had already sought medical advice for their hip, seven hips had required surgery and four had been treated medically.

Four patients had been exempted from military service, two had been exempted from marching and two had never undertaken any sport. Only two hips were considered to be normal, ten caused moderate discomfort and eight severe discomfort. Of the 14 hips which were examined, eight had severe limitation of movement, four had minimal loss of movement, and two had a normal range of movement.

There was a mean irregularity of 8.1 mm (5 to 14). The mean CE angle was 83.5% (31 to 106) and the mean ATD was 46% (4 to 100) of the normal side. Signs of osteoarthritis were seen in 12 hips, and were severe in five. Six hips did not show evidence of osteoarthritis and for two no radiograph was available.

50-year follow-up. For these 17 patients, the mean follow-up was 51.5 years and the mean age of patient was more than 59 years. The findings confirm the poor prognosis at this stage. Three hips had required conservative surgery (one shelf osteotomy, two Chiari osteotomies). Seven hips (six patients) had required a THR at a mean age of 54.1 years. This group included three patients who underwent surgery under the age of 50 years and one who had previously undergone a shelf osteotomy at the age of 16.
years. Ten hips (ten patients) had not undergone a THR and were available for analysis. Three were free from pain, one caused occasional discomfort and six were painful. Eight patients walked with a limp, one required a walking stick, and two had a limited walking distance. The Merle d'Aubigné and Postel score was excellent (18) in three patients, very good (17) in three, good (16) in one, fair (14 to 15) in two, and poor (12) in one.

Of the ten hips which had not required a THR, four needed analgesia. Two were subjectively normal, six caused moderate discomfort and two severe discomfort. The available radiographs showed four hips with and three without osteoarthritis. Three patients refused to undergo radiological examination.

Discussion

There are clearly limitations to a study such as this, mainly because of a lack of complete data. Despite its length, the series is still too short, and will require updating in ten years or more. Unfortunately, the volume of work required, combined with a steady decrease in the number of patients available for assessment, may make this difficult. Nevertheless the original observation in 1987⁹ can be confirmed in that the long-term prognosis of LCPD is reasonable.

Over 50 years after the onset of the disease, only 12 of 51 hips have been replaced, at a relatively mature age, despite the large number which have deformed femoral heads. Furthermore, 31 hips had excellent or very good function after this long period. The findings agree with those of Perpich, McBeath and Kruse,¹¹ who observed good clinical results, even in patients with non-spherical femoral heads, and also with those of Yrjonen et al¹² who found that only 4% of patients required a THR at a mean follow-up of 35 years. This contrasts with other paediatric hip conditions, for example, congenital dislocation or epiphysiolysis. It was found, however, that significant deterioration had occurred between the follow-up assessments at 34 and 50 years. MacAndrew and Weinstein,¹³ in a series of 35 hips, found the same at a mean follow-up of 47.7 years; 40% of their patients had undergone THR.

The age of onset of the disease is an important prognostic factor. The mean age of onset for patients with a normal head was five years, but was 6.4 years for those with a flattened spherical head, 9.5 years for patients with an irregular head, and 8.5 years for those with a very irregular head. This agrees with the earlier findings of Mose,⁴ Mose et al⁵ and Stulberg and Salter.⁷ It appears to be the severity of the disorder in older children rather than the time left for remodelling which determines the outcome.

Table I. Functional outcome of 51 hips at the 50-year follow-up

<table>
<thead>
<tr>
<th>Number of hips</th>
<th>Excellent or Very Good</th>
<th>Good</th>
<th>Fair</th>
<th>Poor</th>
<th>THR</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>All hips</td>
<td>51</td>
<td>31</td>
<td>1</td>
<td>4</td>
<td>2</td>
<td>12</td>
</tr>
<tr>
<td>Normal heads</td>
<td>4</td>
<td>3</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>3</td>
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<td>Flattened spherical heads</td>
<td>23</td>
<td>21</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>2</td>
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<tr>
<td>Irregular heads</td>
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<td>1</td>
<td>0</td>
<td>2</td>
<td>1</td>
<td>3</td>
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<tr>
<td>Very irregular heads</td>
<td>17</td>
<td>6</td>
<td>1</td>
<td>2</td>
<td>1</td>
<td>7</td>
</tr>
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</table>
Pain occurred regardless of the condition of the femoral head at skeletal maturity, with the exception of patients with a slightly deformed femoral head. These did not have significant pain, except for one who developed the rapid onset of osteoarthritis in the contralateral hip after a THR on the affected side. It appears that severe pain is associated with a very deformed femoral head. An attempt was made to identify factors which may be associated with the later development of pain, particularly for patients with a normal or flattened spherical femoral head. The numbers were small, but the findings suggest that pain is not related to the age of the patient at follow-up, or to the RQ, CE angle, or ATD. One over-riding feature is the shape of the femoral head at skeletal maturity. This confirms the earlier work of Gower and Johnston, Mose, Mose et al and Stulberg and Salter. The more deformed the femoral head, the greater is the risk of osteoarthritis.

Further follow-up is now required to answer outstanding questions. Can LCPD cause subsequent osteoarthritis in the absence of radiological sequelae? Will minor defects, such as a flattened spherical femoral head, always remain well tolerated? Is osteoarthritis inevitable in the presence of a very deformed femoral head? The series by Mose et al is the only long-term study (19 hips at 65 years). All those with an irregular femoral head had osteoarthritis after this period. The findings of this study suggest the same, albeit between follow-up at 34 and 50 years.

Other factors, such as age of onset or at-risk signs, have only an influence inasmuch as they affect the state of the femoral head at skeletal maturity. In themselves they are not formal, prognostic factors. The same observation applies to architectural abnormalities of the femoral head which appear to be well tolerated, except for the resulting limp. Of particular interest is the one patient, a 56-year-old man, with osteochondritis dissecans of the femoral head which was well tolerated after follow-up of 52 years, confirming Ratliff’s opinion, although the femoral head was only slightly deformed and the fragment had not moved (Fig. 2).

Osteoarthritis as a sequel of LCPD has a number of special features. First, it is difficult to identify in the early stages, particularly for very deformed femoral heads with significant architectural defects. Narrowing of the joint space occurs relatively late. Secondly, there is little correlation between the radiological findings and the clinical signs of osteoarthritis. Some authors have stated that this is a feature of LCPD. We do not consider it to be a specific feature of the condition since it may be encountered in all forms of osteoarthritis. We found that most cases of osteoarthritis seen as a late result of LCPD initially involved the medial aspect of the hip and became global later. The relatively good prognosis of osteoarthritis of the medial aspect compared with that of the superior aspect is well known. Thirdly, although the acetabulum can mould to accommodate a deformed femoral head and restore reason-