Perthes’ disease in the adolescent
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We studied the natural history of Perthes’ disease in 62 children in whom the onset of symptoms was in adolescence.

Three patterns of disease were noted, namely, late-onset pattern, segmental collapse, or destructive with failure of revascularisation. In the late-onset pattern, the disease followed the sequence of healing seen in younger children, but adequate epiphyseal remodelling did not occur. Consequently, the femoral head was never spherical after revascularisation. With segmental collapse, early and irreversible collapse of part of the epiphysis occurred with gross deformation of the femoral head. The destructive pattern was characterised by a failure of revascularisation and repair of the avascular epiphysis.

The radiological outcome was poor in all three patterns. The poorest clinical results were found in the destructive type which was frequently associated with incapacitating pain requiring arthrodesis or excision arthroplasty within three years of onset of the disease.

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In Perthes’ disease the age at onset of symptoms is important in determining the long-term outcome.1-7 Reports, based on data from Caucasian children from several parts of the world, indicate that the mean age of onset of Perthes’ disease is around six years.8-11 In some racial groups the onset is distinctly later.12-14 The clinical presentation and radiological features in these older patients have been reported to be different from those in younger children. Despite this, good results have been obtained with surgical containment in older children who were less than 12 years of age at the onset of symptoms.15,16 Very few reports have appeared on the nature of Perthes’ disease and on the results of treatment in children over the age of 12 years.17,18

Over a period of 25 years, we have treated 674 children with Perthes’ disease, 62 of whom were over 12 years of age at the onset of symptoms. The marked difference in clinical features and radiological appearance of these older children prompted us to analyse the natural history and evolution of Perthes’ disease in adolescence.

Patients and Methods

The medical records and radiographs of 62 patients with Perthes’ disease who were over the age of 12 years at the onset of symptoms were identified and reviewed. Two of the 62 patients (47 boys, 15 girls) had bilateral involvement; of those with unilateral disease the right hip was affected in 35 patients and the left in 25. Haemoglobin electrophoresis was found to be normal in all patients. The clinical features at presentation, the symptoms at follow-up and the outcome of treatment were recorded. Every patient had had anteroposterior and frog-lateral radiographs of the hips. All the radiographs were carefully studied and an attempt was made to apply the Elizabethtown classification19 to identify the stage of evolution of the disease, the Catterall grouping8 to evaluate the extent of epiphyseal avascularity and the classification of Herring et al4 to determine the extent of epiphyseal collapse. Sequential radiographs of anteroposterior and frog-lateral views of the hips during the evolution of the disease were available for 47 of the 62 patients. The radiographs of these 47 patients (49 hips) were analysed in greater detail and the progress of the disease, including metaphyseal, acetabular and dimensional changes, was noted on each radiograph. The final shape of the femoral head was evaluated using Mose’s criteria.20 The size of the femoral head was also measured using the Mose template. In radiographs in which the shape of the head was distorted, the arc of the Mose template which gave the closest fit was taken as the radius of the femoral head. The frequency of premature fusion of the femoral capital physis, the trochanteric physis and the triradiate cartilage was noted. The duration of the disease was calculated for patients who had radiographs from the early stages until either full revascularisation or skeletal

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maturity. The classification of Stulberg et al was applied to all unilateral cases based on the appearance on the final radiograph.

One-way analysis of variance was used to compare differences in the dimensions of the femoral metaphysis, the femoral head and the acetabulum in the different patterns of the disease. The chi-squared test was used to compare the frequency of different Stulberg classes in the three patterns of the disease.

Results

Radiological features. At the time of presentation 17 of the 64 hips were at the initial stage of avascular necrosis, five were at the stage of fragmentation, and 17 showed some radiological evidence of regeneration. One hip had completely reossified by the time the patient reported to hospital. Twenty-four hips had atypical radiological features and could not be staged according to the Elizabethtown classification. Half or more of the epiphysis was involved in every case (i.e., Catterall group II, III or IV). Eight hips were group II, 24 group III, and 31 group IV, in which the entire epiphysis was avascular. Differentiating between Catterall group II and group III was difficult in some instances because of atypical radiological appearances. Only a small proportion of these adolescent patients showed the typical stage of fragmentation where the Herring classification could be applied. In those which did not show the stage of fragmentation, the extent to which the epiphysis had finally collapsed was used: five hips were graded as Herring grade A, 34 as grade B and 25 as grade C. It was also noted that in some patients collapse was more pronounced in the middle pillar rather than the lateral. Neither the Catterall grouping nor the Herring grading correlated with the final outcome.

Osteoporosis of the femoral metaphysis and acetabular roof occurred in all hips during the active stages of the disease. In 32 hips the triradiate cartilage had already fused at the time of presentation. There was premature fusion of the triradiate cartilage in 16 hips and of the trochanteric growth plate and the capital femoral growth plate in 12 hips each.

Radiological patterns. We were able to monitor the evolution of the disease in 47 patients (49 hips), and three distinct patterns based on the radiological appearance were noted.

Late-onset Perthes’ pattern (11 hips). These hips followed the healing pattern seen in younger children. An initial stage of avascular necrosis was followed by fragmentation of the epiphysis and progressive revascularisation, with radiological evidence of formation of new bone leading to final healing (Fig. 1). The sequential radiographs could be classified according to the Elizabethtown classification. The final shape of the femoral head after complete revascularisation was never completely spherical, as judged by Mose’s criteria, and most hips were class III according to Stulberg et al (Table I).

Segmental-collapse pattern (22 hips). In this group, early collapse of an involved segment of the epiphysis had occurred in the weight-bearing area. This was permanent, unlike the transitory loss of epiphyseal height seen in young children with Perthes’ disease. The disease did not follow the Elizabethtown stages of evolution (Fig. 2). In some cases revascularisation remained incomplete. The final shape of the femoral head was markedly distorted in

<table>
<thead>
<tr>
<th>Pattern of disease</th>
<th>Stulberg class</th>
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<tbody>
<tr>
<td></td>
<td>III</td>
</tr>
<tr>
<td>Late-onset (n = 10)*</td>
<td>8</td>
</tr>
<tr>
<td>Segmental-collapse (n = 21)*</td>
<td>0</td>
</tr>
<tr>
<td>Destructive (n = 13)*</td>
<td>0</td>
</tr>
</tbody>
</table>

chi-squared test, 36.56; df 4; p < 0.0001
* only 44 of the unilateral cases were included, as the final radiograph of one patient was not available for analysis
all cases: 19 hips were class IV and two were class V according to Stulberg et al (Table I).

Destructive pattern (16 hips). In this group the avascular area of the femoral epiphysis had undergone progressive resorption, with no evidence of a reparative process and no new bone formation. The hip tended to sublux and severe deformation of the femoral head occurred within a relatively short period of time (Fig. 3).

Table II shows the alterations in metaphyseal width, radius of the femoral head and acetabular radius of the affected hips in different patterns of the disease.

Clinical features and treatment outcome. In those patients with sequential radiographs from the initial stage of the disease, the mean duration of the disease was 31 months. By this time either complete revascularisation of the femoral epiphysis or fusion of all growth plates in the region of the hip had occurred.

Late-onset Perthes’ pattern (11 hips) (mean age at onset 13.72 ± 1.35 years). Three of the children in this group presented early in the course of the disease and underwent varus derotation osteotomy. Based on Mose’s criteria the outcome after surgery was poor in two. The remaining children in this group presented late and were not considered suitable for containment surgery. They were treated symptomatically, with traction if the hip was irritable during the active stages of the disease. They were then advised to avoid weight-bearing until revascularisation was established. When last reviewed they had moderate restriction of hip movement and mild discomfort on exertion. They were able to walk well but all had a mild limp.

Segmental-collapse pattern (22 hips) (mean age at onset 13.82 ± 1.50 years). Three children were considered suitable candidates for containment surgery and underwent varus derotation osteotomy. The final outcome was poor in all three judged by Mose’s criteria. For the remaining children only symptomatic treatment was offered during the active stage of the disease. They had more severe symptoms at follow-up than those with the late-onset pattern. They experienced pain on walking shorter distances and found it difficult to squat. In one patient the pain became sufficiently severe (within two years) to warrant an excision arthroplasty.

Destructive pattern (16 hips) (mean age at onset 14.50 ± 1.55 years). One patient underwent a Chiari osteotomy, but...
severe deformation of the femoral head occurred despite the operation. Eight patients developed incapacitating pain within three years from the onset of symptoms and seven required hip arthrodesis. One patient opted for an excision arthroplasty. The femoral head at the time of surgery showed gross deformation of the articular surface and areas of cartilage destruction and pitting (Fig. 4). The synovium was biopsied in patients undergoing arthrodesis or excision arthroplasty. All showed features of non-specific synovitis with perivascular infiltration of plasma cells and lymphocytes. In one case, lateral subluxation with hinge abduction occurred. A cheilectomy was performed but destruction of the hip progressed with increasing pain. Within two years an arthrodesis was required. In those patients who did not undergo arthrodesis or excision arthroplasty, the hips showed significant restriction of rotation, and pain on walking.

Discussion

Perthes’ disease in adolescence is considered to be a rare occurrence, but was seen in almost 10% of patients presenting to our centre. This appears to be the highest incidence of adolescent Perthes’ disease reported anywhere.

Table II. Mean (± sd) increase in metaphyseal width and the radius of the femoral head and acetabulum in 44 hips with unilateral adolescent Perthes disease, expressed as the percentage increase compared with the contralateral normal hip (95% confidence interval)

<table>
<thead>
<tr>
<th>Pattern of disease</th>
<th>Increase in metaphyseal width</th>
<th>Increase in femoral head radius</th>
<th>Increase in acetabular radius</th>
</tr>
</thead>
<tbody>
<tr>
<td>Late-onset (n = 10)</td>
<td>119.49 ± 7.13 (114.38 to 124.59)</td>
<td>122.36 ± 8.90 (115.99 to 128.72)</td>
<td>112.30 ± 3.41 (109.85 to 114.73)</td>
</tr>
<tr>
<td>Segmental-collapse (n = 21)</td>
<td>117.41 ± 10.64 (112.56 to 122.25)</td>
<td>117.65 ± 9.73 (113.22 to 122.08)</td>
<td>111.23 ± 5.16 (108.87 to 113.57)</td>
</tr>
<tr>
<td>Destructive (n = 13)</td>
<td>102.51 ± 11.92* (95.31 to 109.71*)</td>
<td>100.66 ± 11.56* (93.67 to 107.64*)</td>
<td>109.49 ± 7.17 (105.16 to 113.83)</td>
</tr>
</tbody>
</table>

* statistically significant differences (p < 0.001) are present between values for the destructive pattern and those for the other two patterns of the disease.
The disease in adolescents runs a different course from that in the younger child. The extent of epiphyseal avascularity assessed by Catterall’s grouping \(^1\) and of epiphyseal collapse according to Herring’s classification \(^4\) are accepted as important factors which influence the outcome of the disease in the younger child. Both these classifications should be applied during the stage of fragmentation but only a small proportion of our adolescent patients showed the typical stage of fragmentation. The Catterall grouping was prone to errors of interpretation because of the atypical appearances. The final outcome in our patients with adolescent Perthes’ disease did not appear to be related to either the extent of epiphyseal involvement or of epiphyseal collapse.

We noted three distinct patterns of adolescent Perthes’ disease. The evolution of the disease in the late-onset pattern was similar to that of classical Perthes’ disease seen in younger children, although the outcome was much worse. The segmental-collapse pattern was quite different in its evolution. The destructive pattern was the most atypical in its progression. The extent of bony destruction seen in the destructive pattern might lead one to suspect an infection, but the histological features of the synovium showed changes typical of Perthes’ disease, with perivascular infiltration of immunologically-competent cells \(^{21-23}\) and no evidence of an infective process.

Increase in the width of the metaphysis and enlargement of the femoral head, which are consistently seen in Perthes’ disease, may represent an attempt to facilitate remodelling of the flattened epiphysis. Corresponding alterations in the shape and size of the acetabulum also occur in order to improve the cover of the enlarged femoral head. \(^{24-27}\) Hence the extent of metaphyseal widening, enlargement of the femoral head and changes in acetabular dimensions can serve as indicators of the degree of remodelling of the proximal femur and the acetabulum. We therefore measured the acetabular and femoral radii and the metaphyseal width. The repair and remodelling process in each pattern of the disease was different (Table II). The alterations in the dimensions of the femoral head and neck and the acetabulum suggested that the remodelling process in the proximal femur was most active in the late-onset pattern, although it was seldom sufficient to ensure a spherical femoral head. There was little evidence of remodelling in the destructive pattern. Remodelling of the acetabulum with an increase in the radius of the acetabulum is seen in younger patients, \(^{25-27}\) but in all three patterns of adolescent disease there was very little enlargement of the acetabulum in response to the enlargement of the femoral head (Table II). This may be because the triradiate cartilage had already fused at the onset of the disease in many of the children, and in others premature fusion occurred during the course of the disease. The significance of these dimensional alterations in the three patterns of the disease is summarised in Table III.

The propensity for deformation of the femoral head is very high in all three patterns. The cause of deformation appears to be different in each. In the late-onset pattern

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**Table III.** The extent of collapse and remodelling seen in the different patterns of adolescent Perthes’ disease

<table>
<thead>
<tr>
<th>Pattern of disease</th>
<th>Epiphyseal collapse</th>
<th>Remodelling potential</th>
</tr>
</thead>
<tbody>
<tr>
<td>Late-onset</td>
<td>Moderate</td>
<td>Present but poor</td>
</tr>
<tr>
<td>Segmental-collapse</td>
<td>Severe and localised</td>
<td>Very poor</td>
</tr>
<tr>
<td>Destructive</td>
<td>Severe</td>
<td>Absent</td>
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</tbody>
</table>

<table>
<thead>
<tr>
<th>Epiphysis</th>
<th>Metaphysis</th>
<th>Acetabulum</th>
</tr>
</thead>
<tbody>
<tr>
<td>Present</td>
<td>Present</td>
<td>Absent</td>
</tr>
<tr>
<td>Very poor</td>
<td>Present</td>
<td>Absent</td>
</tr>
<tr>
<td>Absent</td>
<td>Absent</td>
<td>Absent</td>
</tr>
</tbody>
</table>
revascularisation progresses to completion but sufficient remodelling of the deformed epiphysis does not occur. In the segmental-collapse pattern revascularisation does occur but may stop short of completion. In addition, there is no evidence of remodelling of the epiphysis in the area which has collapsed. In the destructive pattern, there is no radiologically demonstrable evidence of revascularisation or repair.

The outcome of the disease in adolescents is poor. Most of the hips show class IV changes of Stulberg et al. Many of the patients with the destructive pattern required salvage surgery to relieve pain. It is likely that patients with the other patterns of the disease will develop degenerative changes in due course.2,17

Current methods of treatment of Perthes’ disease focus on the principle of containment of the femoral head within the acetabulum as a means of preserving the sphericity of the femoral head. The success of this approach depends on resorption of the necrotic avascular bone, replacement with new bone and subsequent remodelling while the femoral head is protected from deformation by adequate containment. The repair process appears to be grossly impaired in the adolescents in this study. As a result they did not benefit from treatment by containment.

Ippolito et al15 noted poor results in 13 patients with adolescent Perthes’ disease. They attributed the poor outcome, in part, to prolonged periods of immobilisation in plaster casts. In our study only one patient, who had a Chiari osteotomy, was in a spica for a period of six weeks. Movement of the joint was not restricted by any form of immobilisation in the other patients. Despite this, the results of treatment were uniformly poor, irrespective of the initial treatment.

It appears that current forms of treatment for Perthes’ disease used in younger children are unsuccessful in adolescence. If any other form of treatment is to succeed it has to be started before any epiphyseal collapse occurs, since there appears to be very little scope for restoring epiphyseal height by remodelling. It may be necessary to counteract early collapse of the epiphysis by reinforcing the subchondral bone of the femoral head and enhancing the process of revascularisation. Such an approach has been used with success by Montella, Nunley and Urbaniak28 in patients with avascular necrosis of the femoral head associated with pregnancy, by performing vascularised fibular grafting. Recently, in three patients (not included in this study) who presented early before collapse of the epiphysis had occurred, we drilled the growth plate and introduced a fibular strut graft from the subtrochanteric area to support the subchondral bone. It remains to be seen if this procedure can prevent deformation of the femoral head in adolescents.

No benefits in any form have been received or will be received from a commercial party related directly or indirectly to the subject of this article.

References