Perthes’ disease

PROGNOSIS IN CHILDREN UNDER SIX YEARS OF AGE

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Children presenting with Perthes’ disease before their sixth birthday are considered to have a good prognosis. We describe 166 hips in children in this age group. The mean age at onset of the disease was 44 months (22 to 72). Mild forms (Catterall I and II) were treated conservatively and severe forms (Catterall III and IV) either conservatively or operatively. The aim of the former treatment was to restrict weight-bearing. Operative treatment consisted of innominate osteotomy and was indicated by a Conway type-B appearance on the bone scan. All the patients were followed to skeletal maturity with a mean follow-up of 11 years (8 to 15).

The end results were evaluated radiologically using the classifications of Stulberg and Mose. A total of 50 hips were Catterall grade-I or grade-II, 65 Catterall grade-III and 51 Catterall grade-IV. All hips with mild disease had a good result at skeletal maturity. Of the hips with severe disease 78 (67.3%) had good (Stulberg I and II), 26 (22.4%) fair (Stulberg III) and 12 (10.3%) poor results (Stulberg IV and V). Of the Catterall grade-III hips 38 were treated conservatively of which 31 (81.6%) had a good result, six (15.8%) a fair and one (2.6%) a poor result. Operative treatment was carried out on 27 Catterall grade-III hips, of which 21 (77.8%) had a good, four (14.8%) a fair and two (7.4%) a poor result. By comparison conservative treatment of 19 Catterall grade-IV hips led to ten (52.7%) good, seven (36.8%) fair and two (10.5%) poor results. Operative treatment was carried out on 32 Catterall grade-IV hips, of which 16 (50.0%) had a good, nine (28.1%) a fair and seven (21.9%) a poor result.

We confirm that the prognosis in Perthes’ disease is generally good when the age at onset is less than six years. In severe disease there is no significant difference in outcome after conservative or operative treatment (p > 0.05). Catterall grade-III hips had a better outcome according to the Stulberg and Mose criteria than Catterall grade-IV hips, regardless of the method of treatment.

Perthes’ disease occurring in children under six years of age is usually considered to be a benign, self-limiting condition with a good outcome. Salter and Thompson\(^1\) suggested that surgery was not required in such cases and reported a favourable outcome even with major involvement of the femoral head. Herring et al.,\(^2\) and Herring, Kim and Browne\(^3,4\) in their prospective studies specifically excluded patients less than six years of age at the time of onset of the condition. Catterall\(^5\) also reported more favourable results in the younger child. However, this viewpoint was not universally held. Fabry, Fabry and Moens\(^6\) and Schoenecker, Stone and Capelli\(^7\) reported that severe forms with early onset had a potentially poor outcome. This was further supported by Snyder\(^8\) who reported poor results in 43% of children with onset of the disease before the age of five years.

The good outcome in children with early onset of the disease may be explained by the potential for remodelling during the period of remaining growth.

Our aim in this study was to determine the frequency of the severe forms, whether children under six years of age required active treatment and whether a severely-involved femoral head can remodel and potentially return to normal anatomy.

Patients and Methods

Of a group of 480 patients with Perthes’ disease treated at our institution, 238 (49.6%) had the onset of the disease before the age of six years, 185 (38.5%) between the age of six and nine years and 57 (11.9%) after the age of nine years. Of the 238 with early-onset Perthes’ disease, 146 had reached skeletal maturity at the time of the present study. This
included 20 patients with bilateral disease. Therefore 166 hips formed the basis of the study. There were 117 boys and 29 girls with a mean age at the onset of the disease of 44 months (22 to 72). Each patient was followed up and routine records made of the sitting height, standing height and weight, and regular clinical examinations were carried out. Routine anteroposterior and lateral radiographs of the hips together with standard measurements of bone age (anteroposterior radiograph of the left hand, Tanner’s signs, standing and sitting heights) were performed to assess clinical progress and skeletal maturity (Figs 1 to 3).
The radiographs of all the patients were assessed using the classifications of Catterall,\(^5\) Herring et al,\(^2\) Salter and Thompson\(^1\) and Bowen, Soster and Hartzell.\(^10\) However we found it difficult to apply these classifications to each patient in a consistent manner. The severe forms were defined as Catterall grade-III and grade-IV, Herring B and C, Salter-Thompson B and Bowen > 50%. Therefore for simplification we relied only on the Catterall\(^5\) classification.

Patients with mild forms were not actively treated apart from having short periods of traction for limping and loss of movement.

Those with severe forms were investigated by MRI,\(^11,12\) bone scanning\(^13-15\) and dynamic arthrography.\(^5\) Four MRI parameters were studied, namely, the extent of epiphyseal necrosis, lateral extrusion of the femoral head, physeal involvement and metaphyseal changes. The bone scan scintigraphy was classified according to the method of Conway.\(^13\) In this method the A scintigraphic pattern represented the re-canalisation process and the B scintigraphic pattern the process of neovascularisation which is of longer duration and has a poorer prognosis (Fig. 4). Dynamic arthrography was performed in all patients scheduled for surgery in order to assess containment of the hip. These investigations were used to guide decision-making. Patients with Conway type-A changes underwent conservative treatment aimed at restricting weight-bearing. This included traction and the use of a wheelchair with a hip abduction device in order to relieve weight-bearing up to the fragmentation stage. When dynamic arthrography showed a containable femoral head in conjunction with MRI indicating lateral extrusion of the femoral head, and Conway type-B changes were seen on the bone scan, surgery in the form of innominate osteotomy (Salter\(^16\)) was performed.

The results were evaluated radiologically using the classification of Stulberg, Cooperman and Wallensten\(^17\) which identifies the risk of osteoarthritis in the long term. Stulberg grades I and II were considered to be a good result, grade III a fair result, and grades IV and V a poor result. The hips were also rated according to the Mose\(^18\) classification (concentric circles method).

All the patients were treated and followed until skeletal maturity (Figs 1 to 3) by the same group of orthopaedic surgeons at the same institution.

Statistical analysis. This was performed with StatView for Windows (version 5.0; SAS, Cary, North Carolina). Statistical comparison of the results of conservative and operative treatment according to the Catterall group was tested using the chi-squared and Fisher exact tests. The level of significance was set at \(p \leq 0.05\).

Results

A total of 16 hips were Catterall grade-I and 34 were Catterall grade-II, giving a total of 50 hips with mild involvement. In this group there were no poor results and all the hips had a good outcome, being classified as either Stulberg grade I or grade II. All the hips were rated good according to the Mose classification.
Severe disease was found in 116 hips. These included 65 Catterall grade-III hips and 51 Catterall grade-IV hips. Our findings showed that 24 hips (20.7%) were Stulberg I, 54 (46.6%) Stulberg II, 26 (22.4%) Stulberg III and 12 (10.3%) were either Stulberg IV or V. The Mose classification was good in 84 hips (72.4%), fair in 16 (13.8%) and poor in 16 (13.8%). When analysed separately the Catterall grade-III hips had overall better results than those in grade IV (Tables I and II).

Containment was assessed by dynamic arthrography in all patients undergoing surgery. We found that all hips were well-contained and there was no increase in the medial dye pool.

Statistical analysis showed no significant differences between the result of conservative and surgical treatment in Catterall grade-III (chi-squared test, $p = 0.6994$) and Catterall grade-IV hips (Fisher’s exact test, $p = 0.556$).

<table>
<thead>
<tr>
<th>Table I.</th>
<th>Overall results at skeletal maturity independent of the method of treatment in patients with severe disease (Catterall grade-III and grade-IV)</th>
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</thead>
<tbody>
<tr>
<td>Stulberg grade</td>
<td>Catterall grade-III</td>
</tr>
<tr>
<td>I</td>
<td>14 (21.5)</td>
</tr>
<tr>
<td>II</td>
<td>38 (58.5)</td>
</tr>
<tr>
<td>III</td>
<td>10 (15.4)</td>
</tr>
<tr>
<td>IV, V</td>
<td>3 (4.6)</td>
</tr>
<tr>
<td>Total</td>
<td>65</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Table II.</th>
<th>Details (number, %) of the comparison between conservative and operative treatment in patients with severe forms of disease (Catterall grade-III and grade-IV)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Stulberg grade</td>
<td>Conservative</td>
</tr>
<tr>
<td></td>
<td>Catterall III (%)</td>
</tr>
<tr>
<td>I, II</td>
<td>31 (81.6)</td>
</tr>
<tr>
<td>III</td>
<td>6 (15.8)</td>
</tr>
<tr>
<td>IV, V</td>
<td>1 (2.6)</td>
</tr>
<tr>
<td>Total</td>
<td>38</td>
</tr>
</tbody>
</table>
Discussion

We have considered the problems encountered in children who present under the age of six years with Perthes’ disease which represents almost half of the number with Perthes’ disease at our institution. When the disease was mild, the overall outcome was good without surgery. Treatment in this subset was aimed at preserving and maintaining a good range of movement by temporary traction (Fig. 5).

Analysis of the data for severe forms of the disease showed that irrespective of the severity (Catterall grade-III or grade-IV), the method of treatment selected (conservative or operative) did not alter the outcome. At skeletal maturity approximately 67.3% regained their original spherical shape (Stulberg I and II).

Patients with good results were generally asymptomatic with no or mild restriction of movement, whereas those with a poor result had a restricted range of movement, but were not always symptomatic.

Nevertheless, it is very important to differentiate Catterall grade-III from Catterall grade-IV hips. These two groups do not share the same prognosis. In the Catterall grade-III group, regardless of the treatment, 80% regained their spherical shape at skeletal maturity and only two remained deformed (Stulberg III, IV or V). Conservative or operative treatment had no effect.

In the Catterall grade-IV group the prognosis was not as good; 51% regained their spherical shape at skeletal maturity and 49% remained deformed (Stulberg III, IV or V). Again, surgical treatment did not alter this outcome.

Our impression is that this population of children came to our attention at an early stage of the disease and it may explain why the lateral extrusion did not lead to hinge abduction.

The outcome of Perthes’ disease is related to the age of the patient at the time of onset and the amount of involvement of the femoral head. Older age and entire involvement of the femoral head are poor prognostic factors.2,19-22

Kelly, Canale and Jones23 showed good results in 41 of 42 cases, but did not differentiate mild from severe disease. Gigante, Frizziere and Turra24 and Mukherjee and Fabry25 found good results in 70% to 75% of cases of severe disease.

Kamhi and MacEwen26 emphasised the poor prognosis in severe Perthes’ disease, finding good results in only 42% of cases. Clarke et al27 demonstrated that in children who were less than four years of age, better results were seen in mild disease after conservative treatment.

In severe Perthes’ disease it is the extent of the disease rather than the age which is the main factor influencing the outcome. In his original study, Catterall28 studied 97 hips in which the shape of the head and function were evaluated. Of these, 35 were in patients less than four years of age. There were 16 Catterall grade-III and grade-IV hips of which 81% showed a fair or poor result at follow-up. These latter hips were untreated, but had similar results to the cases in our series, suggesting that treatment did not alter the natural history of the disease in this age group.

Schoenecker et al29 reviewed 109 patients with unilateral Perthes’ disease who were under six years of age. Of the patients in Catterall grade-III and grade-IV 24% had a poor
result at the final follow-up. They concluded that despite the early onset of the disease, patients with involvement of more than half of the femoral head were at risk of poor radiological results. However, in this study the patients were not reviewed at skeletal maturity.

Fabry et al. studied 36 hips with Perthes’ disease. Of those with severe involvement, seven were in Catterall grade-III and 16 in grade-IV. At skeletal maturity they found that 44% had good, 22% fair and 33% poor results according to the Stulberg classification. The authors stated that young age did not protect from severe disease. Our results were similar since we found that in Catterall grade-IV 31.4% were Stulberg III and 17.6% were Stulberg IV (Table I).

In their analysis of 190 hips with the onset of Perthes’ disease before the age of six years, Rosenfeld, Herring and Chao showed that all hips with a lateral pillar A and 94.4% with a lateral pillar B had a good outcome (Stulberg I or II). However, they emphasised the severity of the B/C border and C lateral pillar hips in which they found good results in 55% and 48%, respectively. They concluded that for patients with the onset of Perthes’ disease before the age of six years was generally favourable, with 80% having good results.

Our results were similar to those in the literature, but we found that the percentage of poor results was much higher in Catterall grade-IV than in grade-III hips.

In this large series of young patients with Perthes’ disease under the age of six years, conservative or operative treatment had no significant effect on the outcome. When dealing with severe disease in this age group, these treatments achieve the same proportion of good results.

The prognosis is different when considering the different subtypes of severe disease. Our study has shown that Catterall grade-III hips were more likely to have a good radiological outcome than grade-IV hips which had a significantly higher tendency to have a poor result.

However, we conclude that young children are not protected from severe disease and poor outcome simply because of their young age. For this reason, the families of young patients with Perthes’ disease should be informed about the risk of poor results.

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


