HIP DYSPLASIA WITHOUT DISLOCATION IN ONE-YEAR-OLD BOYS

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From Leeds/Bradford Orthopaedic Training Scheme

Six boys were examined during the second year of life, each with symptoms in one hip. The affected femoral capital epiphysis was smaller or more irregular (or both) than that of the unaffected hip but was neither subluxed nor dislocated. The subsequent radiographs revealed changes similar to those in severe Perthes' disease. Nevertheless, we give reasons for believing that this disorder in boys under two years of age should be distinguished from Perthes' disease.

Pelvic radiographs of healthy children taken during the second year of life usually show well-defined femoral capital epiphyses and symmetry of development between left and right. Symptoms suggesting hip disease and leading to radiological examination rarely occur at this age. Hilgenreiner (1933) and Gickler (1937) reported that radiographs of children examined because of hip symptoms during the second year of life showed delay in development of the affected femoral capital epiphysis but no evidence of dislocation. The subsequent changes observed radiologically were thought to be similar to those of Perthes' disease and Hilgenreiner (1933) called the condition "Frühperthes" (early Perthes). Meyer (1964) and Harrison (1971) also studied patients with similar initial radiological changes which, they felt, were distinct from those of Perthes' disease; they called the disorder "dysplasia epiphysealis capitis femoris". At healing the shape of the affected femoral heads in these children was normal.

In this paper we report the findings in six patients with pain in the hip or with a limp, who were first examined before the age of two years. From the radiographs taken at first attendance they were all seen to have dysplasia or hypoplasia of the affected femoral capital epiphysis.

CLINICAL MATERIAL

The six patients in this study, all boys, had been treated in Leeds and Nottingham between 1953 and 1976. None had received any previous treatment for congenital dislocation, subluxation, or instability of the hip.

Case 1. Figure 1—Radiograph of a boy aged twenty months. History of pain in the left leg for two days. No previous complaints. Figure 2—Four months later; fragmentation of the left femoral capital epiphysis. Figure 3—Eight months later; fragmentation is still evident. Figure 4—Eight months later; the epiphysis is re-forming. Figure 5—One year later; although the epiphysis has re-formed, its density clearly differs from that on the right. Figure 6—One year later; the difference between the two sides is still evident. Figure 7—One year later; the density has still not returned to normal. Figure 8—One year later; healing has taken place.

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Case 1. At the age of twenty months this boy developed a limp on the left side. Two days later his left hip was painful. The radiographs taken after admission to hospital showed a smaller capital epiphysys and slight metaphysial changes on the affected side, but these were not considered significant (Fig. 1). As the child continued to limp, he was placed on a Jones’ abduction frame a week after admission. Subsequent radiographs (Figs. 2 and 3) showed fragmentation of the femoral capital epiphysis and a diagnosis of “early and severe Perthes’ disease” was made. Eighteen months after his first admission to hospital he was fully mobilised and weight bearing. The progress of the condition was followed radiologically (Figs. 4 to 7) until five and a half years after his first admission, when healing was considered to have occurred (Fig. 8).

Case 2. This boy developed a limp on the left side at the age of seventeen months. After admission to hospital he was placed on Pugh’s traction. The radiographs taken at this time (Fig. 9) showed a small and irregular left femoral capital epiphysis, but this was not thought significant. The child’s limp settled and he was discharged ten days after admission. He remained well until three and a half years later when he again began to limp on the left leg. The radiographs now (Fig. 10) showed changes thought to be typical of “advanced and severe Perthes’ disease”. He was prescribed patterned calipers for two years and the progress of the condition was followed radiologically (Fig. 11). He was believed to be healed seven years after his first admission to hospital and was then discharged (Fig. 12).

Case 3. This boy was one of dizygotic twins who developed a limp on the right side at the age of eighteen months. The radiographs taken on admission to hospital five weeks later showed the right femoral capital epiphysis to have developed a large “defect” (Fig. 13). Treatment was by a Jones’ abduction frame for two and a half years. The radiological course of the condition is illustrated in Figures 14 and 15 and although fragmentation of the epiphysis did not develop, the child was thought to have had “early and atypical Perthes’ disease”. Three and a half years after his first admission to hospital the condition was considered to have healed (Fig. 16).

Case 4. At the age of nineteen months this boy suddenly developed pain in the right thigh. On admission to hospital radiography showed a small and irregular femoral capital epiphysis on the right, with broadening of the femoral neck (Fig. 17). “Early Perthes’ disease” was diagnosed and after an initial period on traction he was placed in “broomstick” plaster casts. He remained in these for a year and was then allowed to walk normally. Subsequent radiographs showed fragmentation of the femoral capital epiphysis. Four years after his first admission to hospital he again developed a limp on the right side and on this occasion he spent three months in hospital on Pugh’s traction (Fig. 18). Five years after his first admission to hospital the condition was considered healed.

Case 5. At the age of twenty-three months this boy developed a limp on the right side. Three weeks later he was admitted to hospital and radiographs showed a hypoplastic and irregular femoral capital
epiphysis on the right, with changes in the femoral neck (Fig. 19). A diagnosis of "early Perthes' disease" was made. He was placed on a Jones' abduction frame for a year and then mobilised in patten-ended calipers. He wore these until the condition was thought to have healed, four years after his first admission to hospital. Radiographs at that time showed no fragmentation of the capital epiphysis.

Fig. 19
Case 5. Radiograph of a twenty-three-month old boy who developed a limp on the right three weeks before this film was taken. "Early" Perthes' disease was diagnosed.

Case 6. This boy was admitted to hospital at the age of seventeen months because of pain in his right leg. The smaller right capital epiphysis was noted on the radiographs but this was not regarded as significant (Fig. 20). With rest in bed his symptoms settled rapidly and he was discharged ten days later. Two and a half years later he again complained of pain in the right hip. Radiographs revealed "advanced and severe Perthes' disease" (Fig. 21). He was treated in "broomstick" plaster of Paris casts for a year and a half. The condition was considered healed five years after his first admission.

Figs. 20 and 21
Case 6. Figure 20—Radiograph of a boy aged seventeen months. The smaller right capital epiphysis was noted but thought insignificant. Figure 21—Two and a half years later. Recurrence of pain in the right hip. "Advanced and severe" Perthes' disease diagnosed.

RADIOLOGICAL STUDY
The initial radiographs were studied and Wiberg's (1953) CE angle was measured (Table I) to determine whether subluxation or dislocation of the hips was present.

Healing was considered to have occurred when the radiological density and texture throughout the femoral capital epiphysis, femoral neck and pelvis had returned to normal. In Case 2 the "final" radiograph was taken before these features had developed and the boy was discharged at this stage. This "final" radiograph was included in the present study as it was assumed that little further change in the shape of the femoral head would have occurred.

At healing the hips were assessed quantitatively using the techniques of Heyman and Herndon (1950). The following were measured: the epiphysial quotient, the acetabular quotient and the acetabulum–head quotient. The mean of these three was called the "comprehensive quotient".

RESULTS
The mean age of the patients in this study was one year and seven months at the time when they were first seen. The presenting symptoms were pain around the hip, a limp, or both. Most of the patients were seen within a week of the onset of the symptoms. All the children were treated but none had a surgical operation.

The radiographs obtained at the first attendance showed that, on the affected side, in every case the femoral capital epiphysis was smaller, or irregular, or both. In two children the initial radiographs were not thought to show any significant abnormality and these children were discharged. They were not seen again until several years later, when they came back with symptoms in the same hip; the radiographs then were thought to show "severe and advanced Perthes' disease". Radiographs of the other four children were made at regular intervals and changes characteristic of "severe or atypical Perthes' disease" gradually became evident; in two patients fragmentation was never seen.

Table I shows the CE angle (Wiberg 1953) of the affected and unaffected hips as measured on the initial radiographs. On the affected side the lowest angle was 25 degrees which, according to Wiberg, is well within normal limits.

The results of the radiological measurements at healing are summarised in Table II. These findings indicate that severe lasting damage occurs not only to the upper end of the femur but also to the acetabulum.

Table I. CE angle at first attendance (in degrees)

<table>
<thead>
<tr>
<th>Case number</th>
<th>Affected side</th>
<th>Unaffected side</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>25</td>
<td>25</td>
</tr>
<tr>
<td>2</td>
<td>40</td>
<td>40</td>
</tr>
<tr>
<td>3</td>
<td>25</td>
<td>25</td>
</tr>
<tr>
<td>4</td>
<td>30</td>
<td>35</td>
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<tr>
<td>5</td>
<td>40</td>
<td>40</td>
</tr>
<tr>
<td>6</td>
<td>45</td>
<td>40</td>
</tr>
</tbody>
</table>

Table II. Radiological data at healing. Fragmentation was present in all but Cases 3 and 5.

<table>
<thead>
<tr>
<th>Case number</th>
<th>Epiphysial quotient</th>
<th>Acetabular quotient</th>
<th>Acetabulum–head quotient</th>
<th>Combined quotient</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>51.4</td>
<td>71.8</td>
<td>92.2</td>
<td>71.8</td>
</tr>
<tr>
<td>2</td>
<td>56.6</td>
<td>78.2</td>
<td>97.0</td>
<td>77.2</td>
</tr>
<tr>
<td>3</td>
<td>67.0</td>
<td>72.1</td>
<td>73.5</td>
<td>70.9</td>
</tr>
<tr>
<td>4</td>
<td>41.1</td>
<td>72.3</td>
<td>78.3</td>
<td>63.9</td>
</tr>
<tr>
<td>5</td>
<td>80.0</td>
<td>94.7</td>
<td>80.0</td>
<td>84.9</td>
</tr>
<tr>
<td>6</td>
<td>38.9</td>
<td>72.0</td>
<td>87.0</td>
<td>66.0</td>
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(mean epiphysial quotient 55.8; mean acetabular quotient 76.8; mean acetabulum–head quotient 84.7). This damage is seen even in the absence of fragmentation of the capital epiphysis.

Delay in the institution of treatment had little effect on the final outcome. The children who were treated from the time of their first attendance did not have an appreciably better final result than those who were untreated for several years (Table III).

Table III. Comparison of radiological quotients between patients treated immediately and those in whom treatment was delayed.

<table>
<thead>
<tr>
<th></th>
<th>Treated from first admission (4 patients)</th>
<th>Not treated until later (2 patients)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Epiphysial quotient</td>
<td>59.9</td>
<td>47.7</td>
</tr>
<tr>
<td>Acetabular quotient</td>
<td>77.7</td>
<td>75.1</td>
</tr>
<tr>
<td>Acetabulum–head quotient</td>
<td>81.0</td>
<td>82.0</td>
</tr>
<tr>
<td>Combined quotient</td>
<td>72.9</td>
<td>71.6</td>
</tr>
</tbody>
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At no stage did any of these children show signs of inflammatory joint disease and in all cases the unaffected hip remained normal.

DISCUSSION

In a series of eighty-seven healthy children, observed because of a “clicking hip” at birth, Fitton and London (1977) found four whose radiographs at the age of twelve to fourteen months showed one femoral capital epiphysis to be smaller than the other. These children never had symptoms related to the hip, had no evidence of hip displacement and within the following year the epiphyses on the two sides became similar in shape and size. In our six patients, clearly recognised symptoms led to the first radiological examination. In every case the epiphysis on the affected side was small and irregular, but in no case was there evidence of dislocation or subluxation of the hip. Despite the relatively minor and transient clinical symptoms at first presentation, severe radiological changes followed and persisted for several years. This suggests that the pathological process leading to the long-lasting progressive changes in the capital epiphysis precedes the onset of symptoms by a considerable interval and might even be congenital.

In Perthes’ disease the age distribution of onset is strikingly narrow; thus in a series of 351 patients which we studied, no single child at onset was aged less than two years. Similar findings have been reported extensively. The group of children described in this present paper had symptoms which started during the second year of life; that is to say, their age distribution was completely different from the series with classical Perthes’ disease. The radiological changes seen in our six patients resemble those seen in severe Perthes’ disease but the condition took much longer to heal. The final results, as measured on the radiographs taken at healing, were poor. Thus the mean comprehensive quotient for all patients in the present series is 72.4, with a standard deviation of 7.7 and a standard error of the mean of 3.1. In our earlier study of patients with Perthes’ disease (Nevelos et al. 1977) we found that at healing the mean comprehensive quotient in patients with the non-fragmenting type of the disease, in the group aged two to four years, was 95.8 (standard error 1.8). In the same age group patients with the fragmenting type of Perthes’ disease had a mean comprehensive quotient of 83.7 (standard error 2.5). It appears, therefore, that at least some of the aetiological or pathogenetic factors affecting the patients in this present series differ from those of classical Perthes’ disease.

Congenital dislocation or subluxation of the hip leads to delay in development of the affected femoral capital epiphysis. Correction of the displacement leads eventually to normal development: the affected femoral head “catches up”. Occasionally avascular necrosis of the femoral head occurs with radiological changes resembling those observed in our patients, but rarely lasting longer than two years. When our patients were admitted to hospital, three were placed on Jones’ abduction frame, two on traction and one on bedrest. None of these forms of treatment has been shown to contribute to avascular necrosis of the femoral head. Apart from the significant difference between the initiation of the condition seen in our patients and in that of patients with congenital dislocation of the hip complicated by avascular necrosis, the radiological changes were overt and continuous in our patients, the final results at healing were poor and the radiological changes took, on average, some five years to heal.

Meyer (1964) and Harrison (1971) in describing “dysplasia epiphysealis capitii femoris” stated that the condition which they had observed in children under the age of four appeared to start with a smaller and flattened femoral capital epiphysis on the affected side; it was bilateral in 50 per cent of cases and always healed with a perfect end-result. Our patients were all under the age of two when they were first seen, all had unilateral disease and the end-results were less than perfect. There was no evidence of an infective process nor of generalised joint disease.

We conclude that, in a small proportion of children, a disease process attacks the growing upper femoral epiphysis and the child develops symptoms during the second year of life. Initial radiographs show a small and irregular capital epiphysis without evidence of subluxation or dislocation of the hip. Although the ensuing radiological changes resemble those of severe Perthes’ disease, differences in both the age of onset and in the outcome distinguish the disorder observed in these six patients from Perthes’ disease. The aetiology remains to be elucidated.
ACKNOWLEDGEMENT
Our thanks are due to Emeritus Professor J. M. P. Clark and Mr C. L. Colton for allowing us to study their patients, and to the Department of Medical Illustration at St James’s University Hospital, Leeds, for the preparation of the illustrations in this paper.

REFERENCES