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The natural history of Perthes' disease

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Summary. Since 1975, none of our patients with Perthes' disease have received any active treatment, except for managing severe pain. We have studied the functional and radiological results in order to evaluate the consequences of this approach and to demonstrate the natural history of the disease. We have followed up all patients over the age of 19 years; out of 20 only 6 were free of pain and only 2 reached 100 points on the Iowa hip score. Radiographs at follow up demonstrated that no involved hip had a spherical femoral head. We conclude that our passive attitude towards Perthes' disease results in relatively few patients having good hip function. especially when compared with the results of other authors. Active treatment, including operation, is probably more successful.

Résumé. Depuis 1975, aucun de nos patients, atteints de maladie de Legg-Perthes-Calvé, n'a été traité de façon active, sauf en cas d'importantes douleurs. Nous avons étudié les résultats cliniques et radiologiques afin d'évaluer les conséquences de cette attitude et de préciser l'histoire naturelle de la maladie. Nous avons suivi tous les patients au delà de l'âge de 19 ans. Sur les 20, seulement 6 n'avaient aucune douleur et deux seulement atteignaient 100 points sur la cotation de hanche de l'Iowa. Les radiographies montraient qu'aucune des hanches atteintes n'avait une tête fémorale sphérique. Nous en concluons que notre attitude passive vis-à-vis de la maladie de Legg-Perthes-Calvé n'apporte un bon résultat fonctionnel qu'à un relativement petit nombre de malades, surtout si on la compare aux résul-

Introduction

The best treatment for Perthes' disease is still debatable. Many reports advocate operation, especially in older children [4, 7, 9, 11]. There is no reliable way of being sure of the prognosis in any individual because of the great variation in the evolution of one hip compared with another [4, 11]. Catterall's classification [1] may be of some help.

It is important to know the natural history of the condition when discussing the effect of different forms of treatment. To our knowledge there has been only one report on patients who had not been treated; they served as controls with socalled 'uncontained treated controls' in comparison with osteotomies [7]. These untreated controls were not presented separately as far as their results were concerned. Thus there is no modern study of the outcome of unselected strictly untreated patients.

In 1975, we began to study the results after various types of treatment. We felt that there was no convincing evidence that operative treatment was better than nonoperative. Neither did we feel that orthotic treatment produced satisfactory results. We therefore stopped giving any routine treatment to our patients with Perthes' disease.

Material and method

Since 1975, none of our patients with Perthes' disease have had any active treatment, except for the management of severe pain. When the diagnosis was made they were told to live as

tats d'autres auteurs. Un traitement actif, éventuellement chirurgical, est probablement meilleur.

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normally as possible. When pain was severe, traction in an extended position was used for one or two days. This relieved the pain, and was usually needed only once or twice for each patient during the course of the disease.

We decided to study the function of young adults who had been managed in this way. Those included were all born in 1972 or earlier and the diagnosis was made in 1975 or later. There were 22 patients in the group. One had moved out of our district and another was reluctant to come for examination. We can therefore present the results of 20 patients who were managed by the 'nontreatment' approach described.

Both hips were involved in 5 patients. The age at onset was on average 8 years (range from 3.9 to 15.4 years). In the oldest patient only the left hip was affected and this was in Catterall stage 1.

The age at follow up was on average 22.4 years (range from 17.3 to 28.1 years); hence all were skeletally mature.

Subjective symptoms were evaluated with a visual analogue scale. The Iowa hip score [6] was calculated; leg length discrepancy and wasting of the thigh were measured.

The radiographs which had been taken at the onset, then at 6-12 month intervals until remodelling of the head had occurred, were assessed. Anteroposterior and Lauenstein's views were taken of both hips. Finally, radiographs were taken at follow up. All hips were graded according to Catterall's classification, head at risk factors were noted and the tear drop distance measured [3]. In unilateral cases, we measured the height and width of the capital epiphysis, and the radius and height of the head, and then related these to the corresponding measurements of the other hip. This enabled us to calculate the epiphyseal, joint surface and radius quotients according to Mose [10].

An acetabular quotient has also been used which is calculated in the same way as the epiphyseal quotient; the depth and width of the affected acetabulum is compared with the corresponding measurements on the normal side.

Acetabular quotient = $\frac{\text{depth (affected)} \times \text{width (unaffected)}}{\text{width (affected)} \times \text{depth (unaffected)}}$

The sphericity of the head was always measured. All measurements were made by the same person (SH).

Results

The Iowa hip score has 5 parts namely function (maximum 35 points), freedom from pain (35 points), gait 10 points, absence of deformity (10 points), and range of motion (10 points), totalling 100 points.

Thirteen of our patients had full function, but only 6 had no pain; 15 said they walked normally; 7 had a full range of motion in the affected hip, and 4 had a fixed deformity. Only 2 scored a total of 100 points. The average score was 90 (range from 74 to 100).

The circumference of the thighs was measured 10 cm proximal to the patella; 8 had no wasting and 12 showed average wasting of 1.75 cm (range 1 to 3.5 cm).

The Iowa visual analogue pain scale was used, the patient's own evaluation for an average day being marked. No pain or discomfort scored 100

Table 1. Catterall classification at first contact.

Catterall classification	At first contact (number of hips)		
1	14		
2	7		
3	3		
4	1		

points. The average pain score in our patients was 84.8 (range from 55 to 100) which correlated well with the total hip scores.

Radiological evaluation using the Catterall classification is shown in Table 1. Six hips showed one head at risk factor, five showed two factors, and three showed 3 or more.

Patients in category 1 and with no, or only one, head at risk factor did not develop significantly less pronounced changes at the hip at follow up compared to patients with necrosis of the head at the onset and several risk factors. There was no correlation between the hip score and the Catterall classification, or with the head at risk factors.

Figure 1 illustrates the lack of correlation between the hip score and the final radiographs.

Only three patients had limited changes in their hips (category 1 or 2 during the period of observation) and their final result did not differ from the average outcome of the rest.

The tear drop distance was increased in the initial radiograph of every hip; the average was 3 mm (range from 1-7 mm).

The mathematical description of the head of the femur, using the quotients which have been described, can only be calculated in unilateral cases. Some hips showed such severe derangement that these measurements were not meaningful. In all the 14 hips studied, no joint was normal in every respect; no affected hip had a perfect and spherical head at follow up.

Our acetabular quotient, which we designed to compare the shape of the acetabulum and the femoral head, parallels the epiphyseal quotient. The normal value is 1 and in all but 2 hips the values were smaller, on average 0.855 (range from 0.689 to 1.05). There is no correlation between the acetabular and epiphyseal quotients.

Trochanteric overgrowth was measured as the horizontal distance from the upper part of the trochanter to the upper part of the femoral head. The value of the unaffected side was then subtracted from the value of the affected hip. There was overgrowth in all but 2 hips, on average 7 mm (range from -7 to +20 mm).

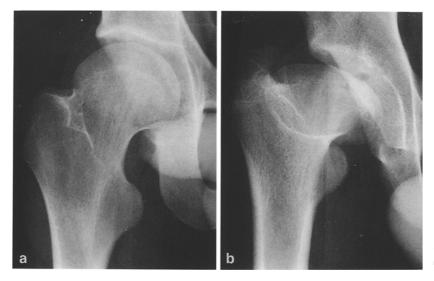


 Table 2. A comparison between our results and those earlier published

	Treatment	Good	Fair	Poor
Salter 1980	Operation	17	77	6
Jani & Dick 1980	Orthosis	42	42	16
Catterall 1971	Orthosis	59	24	17
Karpinski et al 1986	Operation	40	35	25
Lloyd-Roberts et al 1976	Untreated	43	28	29
Eaton 1967	"Treated"	46	18	36
Salter 1980	Orthosis	37	23	34
Mindell & Sherman 1951	"Treated"	56		44
Present study	Untreated	21	11	68
Mindell & Sherman	Untreated	23		77

Discussion

Our patients with Perthes' disease were not treated in any way, apart from brief periods of traction to relieve pain, regardless of the Catterall classification or the head at risk factors. We do not believe that a similar study has been reported. Other series of untreated cases have been selected. We have compared our findings with similar follow up reports on patients of approximately the same age treated either by operation [5, 9] with orthoses [1, 4, 9], or with no treatment [6, 7].

Our approach has provided a consecutive series in which the natural course of the condition can be studied. It has already been noted that only six patients had full activity without subjective symptoms. Since only 24% are free of pain, our passive management is probably less favourable compared to others (Table 2). The terms

Fig. 1. Radiological appearance of two patients with the same Iowa hip score (99 and 100 respectively)

good, fair and poor have been used in the same way as in most of the papers: good = no pain; fair = pain only during activity; poor = pain at rest.

The most striking feature of our radiological evaluation is the total absence of a spherical head at follow up; none of the unilateral cases became normal, and all had some degree of deformity. This may be because of our meticulous measurement of the epiphyseal, joint surface and radius quotients. The absence of correlation between the clinical outcome and the primary Catterall classification shows how difficult it is to give a prognosis for an individual case, and to decide on the best treatment for a particular patient. Usually the age of onset is accepted as being the best parameter to use to make a prognosis [8]. Even if there is a tendency in our series to show a relation between the hip score and the age at onset, there is no statistically significant correlation. Thus the youngest patient shows the lowest score.

Every affected hip showed an increased tear drop distance on the first radiograph, but since we do not have controls the significance of this finding is hard to establish. The sign may be important when radiographs of children are assessed in clinical practice.

The lack of correlation between acetabular and epiphyseal changes is surprising and its significance is difficult to assess. If the acetabular changes were secondary to necrosis of the head, some correlation would be expected. Possibly the acetabular changes develop as a primary disturbance together with the changes in the femoral head, and not secondary to them.

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