

Poor prognosis in atypical Perthes' disease

Radiographic analysis of 19 hips after 35 years

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We analyzed clinical and radiographic long-term results in 96 patients (106 hips) with Perthes' disease 35 years after nonoperative treatment. 19 hips in 17 patients had early radiographic signs indicating atyp-

ically severe course of the disease and poor prognosis. None of them was radiographically good at skeletal maturity; 2 cases were fair and 17 poor. At the final follow-up 12 hips had radiographic arthrosis.

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In our recent study (Poussa et al. 1991) of osteotomy for Perthes' disease, we identified a set of early radiographic changes in the proximal femur indicating a poor prognosis (Table 1). The aim of this study was to re-evaluate these changes in a subset of nonoperated patients.

Patients and methods

Long-term clinical and radiographic follow-ups were carried out for 96 patients (106 hips) treated nonoperatively at our hospital between 1946 and 1958 (Yrjönen 1992).

19 hips in 17 patients had early radiographic signs indicating a poor prognosis (Table 2). 3 boys and 1 girl had bilateral disease. This girl and one boy showed only mild radiographic changes in the other

hip, so those hips were not included in this series. 5 of the hips were Catterall (1971) Group III and 14 Group IV. In addition 14 hips showed two or more signs of head-at-risk. Bicompartimentalization of the acetabulum was found in 6 hips, and osteopenia was observed in 14 hips in the early stages of the disease.

A Thomas splint was used in 13 cases and crutches or no treatment in 6 cases.

For the analysis of data in the whole material the Cochran-Mantel-Haenszel chi-square test was used for testing and measuring linear association between an ordinal-scaled response and covariables. The significance level of the *P*-value was 0.01.

Results

No patient had a good result at skeletal maturity; 2 hips were fair and 17 poor. The 2 patients with fair results were boys who were 4 and 6 years old at the time of diagnosis, both being Catterall (1971) Group IV, and both having two or more signs of head-at-risk. At follow-up radiographic arthrosis was found in 12 hips, 4 were graded as severe. None of them had undergone total hip replacement, but all these cases had severe symptoms. The two hips that were fair at skeletal maturity had no radiographic signs of arthrosis.

Discussion

In our previous study of the results of operative treatment for Perthes' disease (Poussa et al. 1991), we

Table 1. Early radiographic signs of poor prognosis in Perthes' disease (Poussa et al. 1991)

1. Lateral calcification extending far laterally outside the epiphysis towards the greater trochanter at the beginning of the disease.
2. Deformation and widening of the femoral head before the fragmentation stage.
3. Deformation and widening of the femoral neck in the initial phase of the disease.
4. Early sclerotic changes in the metaphysis.
5. The Saturn-phenomenon; a sclerotic epiphysis surrounded by a ring of looser bony tissue.

Table 2. 17 patients (19 hips) with early radiographic signs of poor prognosis in Perthes' disease

A	B	C	D	E	F	G	H	I	J	K	L	M	N
1	b	6	IV	-	+	+	-	-	+	-	-	3	0
2	b	9	IV	+	+	+	+	-	+	+	-	3	3
3	b	4	IV	-	+	+	-	-	+	-	+	2	0
	b	6	IV	-	+	+	+	-	+	-	+	3	0
4	b	6	IV	+	+	+	+	-	+	+	+	3	0
5	b	6	IV	+	+	+	-	+	-	+	-	3	0
6	b	10	IV	-	+	+	-	+	-	-	-	3	2
7	b	6	IV	-	+	+	-	-	-	+	+	2	0
8	b	8	IV	+	+	+	+	-	-	+	-	3	1
9	b	7	IV	+	+	+	+	+	-	+	-	3	0
10	b	9	III	-	-	+	-	-	+	-	-	3	1
11	b	12	III	-	+	-	-	-	+	+	-	3	3
12	b	8	IV	+	+	+	-	-	+	-	-	3	1
13	g	12	III	-	-	-	-	-	+	-	-	3	3
14	g	9	III	-	+	+	-	-	+	+	-	3	1
15	b	6	IV	-	-	-	-	+	-	-	-	3	1
16	b	6	IV	-	+	-	-	+	-	+	-	3	1
17	b	7	IV	-	-	-	-	-	-	-	+	3	3
	b	7	III	-	-	-	-	-	-	-	+	3	2

A Case	G Subluxation, lateral	M Final result
B Sex	H Subluxation, superior	2 fair
C Age	I Wide lateral calcification	3 poor
D Catterall group	J Deformation, head	N Arthrosis
E Bicompartimentalization of the acetabulum	K Deformation, neck	0 no
F Two or more head-at-risk signs	L Metaph. sclerosis	1 mild
		2 moderate
		3 severe

found early radiographic signs of poor prognosis in 20 percent of cases. In a study of the results of nonoperative treatment in 106 hips (Yrjönen 1992), we found 19 hips with the same radiographic signs. This supports our previous findings of an atypical course of Perthes' disease.

In the whole material the radiographic result at skeletal maturity was poor in 60 percent of the hips (Yrjönen 1992), which may have been due to the selection of more severe cases in our hospital. In the present atypical series the result was poor in 17/19 cases. The radiographic changes were found early, and the course of the disease was unexpectedly severe in all 19 hips. In analyzing the whole material of 106 hips, 46 percent of the hips were graded as Catterall (1971) Group III and 50 percent as Group IV; 67 percent of the femoral heads had two or more radiographic signs of head-at-risk. In the present material of 19 hips, the incidence of Catterall Group IV (14 hips) and two or more signs of head-at-risk (14 hips) was increased. However, in the statistical analysis of the whole material, Catterall Groups III and IV (P 0.50) or concept of head-at-risk (P 0.15) did not appear to be of prognostic value.

The patients' age at the onset of the disease did not differ in the present series (mean 7.6 years) from the whole material (mean 7.3 years).

Catterall (1982) has evaluated radiographic changes that might indicate deterioration in the shape of the femoral head. Radiographic hallmarks of head-at-risk signs are difficult to identify and often appear too late to be used as a criterion for the decision about treatment. Specks of lateral calcification are common and represent ossification in the enlarged cartilaginous part of the femoral head. However, this change is most often evident only at later stages of the disease and is frequently reversible. Lateral calcification, extending far laterally towards the greater trochanter in the initial phase of the disease, seems to be irreversible (Figure 1) in contrast to the milder type of lateral calcification described by Catterall. This more severe type, found in five hips, led in all cases to a poor radiographic result.

Widening of the epiphysis occurs as the result of ossification, and apparent flattening of the epiphysis is a consequence of increased width of the femoral head without an associated increase in height. This change also is not often noted until the later phases of the disease, but we have described deformation of the femoral head before, or at an early stage of, fragmentation (Figure 2). This might be due to extraordinary softness of the proximal femur because of crushing of epiphyseal bone trabeculae at the onset of the disease. This change, found in 10 hips, led in all but one to a poor radiographic result at skeletal maturity.

Deformation and widening of the femoral neck at the initial phases of the disease (Figure 2) were noted in 9 hips, and in all cases except one these changes were combined with other early signs of a poor prognosis. This variation may be due to exceptionally severe local or general growth disturbance because of metaphyseal and growth plate lesions. However, deformation of the femoral neck may occur so early that growth disturbance cannot explain the whole phenomenon. The main reason may be wide collapse of bone structure because of Perthes' disease. Diffuse metaphyseal reactions are among Catterall's signs of head-at-risk and are thought to be associated with growth disturbances of the femoral head and neck (Catterall 1982). However, metaphyseal changes are often also found in cases with good results. In the present series we found sclerosis of metaphyseal bone (Figure 3) in 6 cases.

The Saturn-phenomenon, a sclerotic epiphysis surrounded by a ring of looser bony tissue (Poussa et al. 1991), was not found in this series.

Bicompartmentalization of the acetabulum has been thought to be a sign of poor prognosis (Yngve and Roberts 1985). This was not confirmed in this series or our other series of Perthes' disease (Hoikka et al. 1991, Poussa et al. 1991, Yrjönen 1992).

In general, age at the onset of Perthes' disease is considered to be a highly important prognostic factor (Brotherton and McKibbin 1977, Stulberg and Salter 1977). In our series this could not be seen; atypically severe cases seem to lead to poor results irrespective of the patient's age at the onset of the disease.

We had two bilateral cases. Their other joints were normal at follow-up, and there was no clear reason to classify any case in this series as epiphyseal dysplasia, which might have explained the poor results (Monty 1962, Herring et al. 1987, Andersen et al. 1988).



Wide lateral calcification in the initial phase of Perthes' disease.



At the age of 7.5 years.

Figure 1. Case 5. 6-year-old boy.



Figure 2. Case 2. 9-year-old boy. Early deformation and widening of the femoral head and neck.



Figure 3. Case 3. 4-year-old boy. Early metaphyseal sclerosis.

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