Perthes' disease

A STUDY OF CONSTITUTIONAL ASPECTS IN ADULTHOOD

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We studied 16 patients suffering from osteoarthritis of the hip who had had Perthes' disease during childhood. They were compared clinically and radiologically with a control group who had not had Perthes' disease, in order to assess whether a generalised, pre-existing constitutional disorder was present. Nine patients with a previous history of Perthes' disease had some other skeletal abnormality, but only three presented with clinical symptoms. Only one patient in the control group was found to have an abnormality but was symptom-free. Our findings provide further evidence that patients with Perthes' disease may have a generalised abnormality related to chondrogenesis which can produce other skeletal anomalies that persist into adult life.

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There is evidence from biochemical^{1,2} anthropometric,³ radiological⁴ and pathological studies⁵⁻⁷ which suggests that a generalised pre-existing condition is present in Perthes' disease. Whether these findings are transient variations of growth or part of a constitutional disorder, the features of which may persist into adult life, is unknown.

We have therefore looked for the presence of musculo-skeletal abnormalities other than those which presented in a group of adults known to have had Perthes' disease in childhood, and compared them with a control group who did not.

Patients and Methods

The study was carried out on two groups of 16 patients admitted to our institute for hip arthroplasty. The patients in group A were known to have had Perthes' disease during childhood, had been treated at our hospital and had medical records and radiographs available for review. Patients with any other form of skeletal dysplasia were excluded from this group. Those in group B were otherwise healthy patients who had sustained a fracture of the neck of the femur and had been admitted for treatment by hip arthroplasty.

Data were obtained from clinical examination and radiographs of the pelvis, hips, thoracic and lumbar spine, hand and chest. When examination suggested that there were other sites of abnormality, further radiographs were taken. Microradiographs were taken of all femoral heads removed at operation and these were then studied histologically.

The data were analysed statistically using an Epi Info V6 program (Centers for Disease Control, Atlanta, Georgia) for studies of frequency, confidence interval (exact limit), odds ratio and Fisher's test.

Results

Clinical examination revealed two patients in group A with a mild thoracic kyphosis. In group B we found one patient with shortening of the left arm by 20 mm. The heights of the patients in the two groups were similar (Table I).

On radiological examination nine of the patients in group A were found to have other skeletal abnormalities (Table II), and one patient in group B had a short, left humerus.

Radiographs of the hips in both groups showed no deformities of the femoral head or neck other than would be expected after either remodelling or fracture. In two of the patients in group A, with unilateral disease, there was evidence of slight contralateral coxa vara. Only three of this group of patients with additional radiological abnormalities, one with spinal stenosis and two with spondylolisthesis, had symptoms related to these, either before or at the time of admission.

None of the patients in group B had symptoms other than those related to their presenting problem. Microradiographs and histological studies of the resected femoral heads gave little additional information. No cartilaginous tissue was

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Table I. Age, height and radiological abnormalities of patients in groups A and B

	Group A	Group B
Total number of patients	16	16
Mean age in years (range)	44.9 (21 to 60)	58.2 (31 to 68)
Mean height in cm (range)	165 (157 to 168)	163 (156 to 172)
Number (%) with radiological abnormalities*	9 (56.25)	1 (6.25)

* 95% confidence interval 1.83 to 899.83; $p = 0.006$ (Fisher's test)

Table II. Distribution of abnormalities in patients in group A

Case	Age at onset (yr)	Gender	Side	Current age (yr)	Radiological findings
1	12	M	R	42	None
2	6	M	L	63	None
3	8	M	R	49	Skeletal lumbar spinal stenosis
4	10	M	R	53	None
5	11	M	L	43	Spondylolisthesis. Aplasia of an epicondyle of an elbow
6	11	M	R/L	21	Sacral asymmetries. Spondylolisthesis
7	9	M	R	40	None
8	5	F	L	59	Pelvic hypoplasia
9	7	F	R	56	Spinal Scheuermann's with mild kyphosis
10	6	M	L	50	Hypoplastic first left rib
11	11	M	R	60	None
12	5	M	R	32	Spondylolisthesis
13	7	F	L	45	None
14	4	M	R/L	30	Spinal Scheuermann's with dorsal kyphosis
15	9	M	L	58	None
16	6	M	R	45	Spinal Scheuermann's

found within the femoral head and this was therefore of no value in assessing constitutional abnormality.

Discussion

It has been suggested that Perthes' disease may form part of a generalised constitutional disorder.³⁻⁸ Catterall et al,⁷ in a post-mortem study, observed several sites of osteochondritis in a patient with Perthes' disease. Ponseti et al⁸ found that lesions of the epiphyseal cartilage in Perthes' disease were similar to those in the vertebral endplates of patients with juvenile kyphosis, and a 'constitutional theory' was suggested for Perthes' disease. Others^{6,9} believe that the disorder of the hip is part of a generalised condition, since children with Perthes' disease may be shorter than those in the normal population.³ They may have hormonal abnormalities,^{1,2} generalised abnormal radiological findings⁹ and bilateral abnormality of the epiphyseal cartilage of the hip.⁵ Our clinical findings did not reveal a difference in height between the two groups, as observed by Burwell et al,³ the sample was, however, small. In unilateral cases, contralateral radiological abnormalities of the femoral head¹⁰ or mild flattening of the head¹¹ were not seen. These findings are probably an expression of transient retardation of growth which subsequently recovers.

The radiological findings in our patients appear to suggest a disturbance of chondrogenesis which possibly co-existed with Perthes' disease at the time of its clinical development. The spine is most commonly affected. Since often there are no clinical symptoms the condition may be overlooked.

In our study nine of the 16 patients (56%) known to have had Perthes' disease (group A) had some other radiological skeletal abnormality which was in the spine in seven (44%). Three had spondylolisthesis and three Scheuermann's disease. Frederickson et al¹² in a follow-up for 25 years of 500 unselected children and the families of children with spondylolysis found that the incidence of spondylolysis at the age of six years was 4.4% which increased to 6% in adulthood. The incidence of spondylolisthesis was 3.0% and 4.2%, respectively. After analysing their cases they believed that their data supported the hypothesis that the spondylolytic defect is the result of a defect in the cartilaginous anlage of a vertebra. A similar incidence of spondylolysis and spondylolisthesis in the general population has been reported from Germany.¹³

The incidence of spondylolisthesis in our group-A patients was 3 of 16 (nearly 19%), significantly greater than that found in the general population.^{12,13} No patient in group B presented with spinal anomalies.

Three of 16 of our group-A patients had Scheuermann's disease. The incidence in the general population is between 0.4% and 8%,¹⁴ although some authors have reported an incidence of up to 60% in males and 23% in females.¹⁵ However, in a large prospective study of 16 104 unselected children in Italy, aged between 6 and 14 years, the incidence of spinal abnormalities was 7.2% which was associated with kyphosis in only 3.12%.¹⁶ The high incidence of Scheuermann's disease in some studies may be because they concentrated on patients with back pain.¹⁷ The criteria for spinal Scheuermann's disease include three of the following radiological features: increased anteroposterior diameter of the vertebral bodies, wedging of the vertebral bodies, irregular-shaped and narrowed disc spaces and kyphosis or loss of lordosis, but not necessarily pain.¹⁷ None of our patients with Scheuermann's disease presented with back pain.

Stoddard and Osborn¹⁷ also found a strong association between Scheuermann's disease and spondylolysis. Half of their patients who sought help for back pain had clinical and radiological evidence of osteochondrosis. They did not, however, study osteochondrosis of the hip.

Our findings provide further evidence that patients with Perthes' disease have a generalised abnormality related to chondrogenesis which may produce other skeletal anomalies that persist into adult life. The possibility of these abnormalities should be borne in mind when investigating a patient with a previous history of Perthes' disease with symptoms other than from the affected hip.

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