DYSPLASIA EPIPHYSEALIS CAPITIS FEMORIS

MEYER'S DYSPLASIA

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We made a prospective longitudinal clinical and radiological study of 18 children diagnosed as having dysplasia epiphysealis capitis femoris. Half the cases were bilateral. Boys were affected five times more often than girls. There were no symptoms or clinical signs in most but some of the bilateral cases had an inconsistent waddling gait.

The imaging studies suggest that the cartilaginous proximal femoral epiphysis is hypoplastic, with delayed appearance of single or multiple ossification centres. Progressive improvement occurred and at an average age of five years and six months, there was complete fusion of all the ossific nuclei and normal density and texture of the epiphyseal bone.

The end result was a round epiphysis with a slightly diminished height. The dysplasia is attributed to focal hypoplasia of the proximal femoral epiphysis.

Though a number of cases had been reported previously (Hilgenreiner 1933; Gickler 1937), Pedersen (1960) was the first to differentiate dysplasia epiphysealis capitis femoris (DECF) from Perthes' disease.

He reviewed 672 patients diagnosed as Perthes’ disease and found 42 (6%) in whom the pattern was atypical. Meyer (1964) estimated that 10% of cases with coxa plana were of the dysplastic rather than the true necrotic type. Among Meyer’s 30 cases, six (20%) converted from the benign course of dysplasia to become typical of necrotic coxa plana. In such combined cases dysplasia occurs first, and Perthes’ disease later.

Our study was undertaken to resolve the conflict of opinions cited in the literature regarding the aetiology and the final outcome of this developmental anomaly which simulates Perthes’ disease in some of its features and may lead to serious misdiagnosis.

PATIENTS AND METHODS

The radiographic criteria for inclusion in this study were smaller or delayed ossification centres in the proximal femoral epiphysis which became progressively normal with advancing age. We used the prediction line derived by Stewart, Patterson and Mollan (1986) as the basis for normal ossification of the capital epiphysis. We excluded all children in whom the capital epiphyses were normally ossified by the age of 15 months. We also excluded those with congenital metabolic abnormalities of the skeleton and those who failed to thrive. None of the patients included had been diagnosed or treated for disease of the hip.

From 1979 to 1989, 18 children with DECF were diagnosed. The average age at diagnosis was two years and eight months (range 16 months to five years and six months). Their ages at the latest clinic visit ranged from three years and nine months to 12 years, giving an average follow-up of five years and nine months (range 32 months to eight years and three months). We reviewed the patients’ clinical records and radiographs. The imaging studies included radiographs of the pelvis and hips of every child, arthograms and CT scans in one, MR imaging in two and technetium-99 MDP bone-scans in three children.

OBSERVATIONS

Epidemiology. There were 15 boys and three girls; eight boys and one girl had bilateral hip involvement (50%). Two children, both with bilateral involvement, came from one family. Extensive questioning revealed that only one patient (a boy with unilateral involvement) had a sibling who suffered from bilateral severe Perthes’ disease. No hip or skeletal pathology was discovered in
Fig. 1

Radiographs of a child with Meyer's dysplasia of the right hip, from 17 months to 12 years of age. Note the late appearance and irregularity of the ossification centres which progressively fuse resulting in a single rounded femoral epiphysis with normal density and texture but slightly flattened.
Radiographs to illustrate the pattern of spontaneous healing which is typical of dysplasia epiphysealis capitis femoris (right side).

Fig. 2

Radiographs of a child with bilateral Meyer's dysplasia. The left hip followed a different healing course from the right, which still showed surface indentations and irregularities at the age of six years and six months.

Fig. 3
any other family member. One girl (with bilateral involvement) had familial mediterranean fever. In five cases retarded skeletal age was associated with short stature.

**Presentation.** The dysplasia was diagnosed by chance in 70% and was diagnosed in patients referred to our hospital as Perthes' disease in 30%. We decided to follow these patients because Meyer (1964) had observed that 20% could convert to Perthes' disease.

Clinical symptoms and signs were absent in most of the children, although some of the bilateral cases had a mild inconsistent waddling gait. This was the main concern of the parents. Pain, limping or limitation of movement were rarely observed and then for a very short period of time. There was no indication for bed rest, admission to hospital or medication against pain. Pain radiating to the thigh or knee was never described.

**Imaging studies.** The first radiographic finding was a reduction in the distance from the upper edge of the metaphysis to the line described by Hilgenreiner (1933). The normal distance at six to seven months of age is said to be 9.3 mm ± 1.5 mm (Harris, Lipscomb and Hodgson 1960). The distance in our cases ranged from 4 mm to 8 mm on the available radiographs from the second year of life, from 4.5 mm to 9 mm and from 5 mm to 10 mm on radiographs from the third and fourth years of life respectively. The distance was 30% to 50% less on the affected side in unilateral cases. These findings suggest that the cartilaginous epiphysis is initially smaller than normal.

The appearance of the ossific nucleus was usually delayed until the age of 18 months to three years (Fig. 1). Two different patterns of ossification were noted. In some, a single granular focus appeared and in others there were from two to six ossification centres. There was no correlation between the number of ossific nuclei, the age of their appearance, unilateral or bilateral involvement or the age at which the centres fused. In all the children the ossification centres enlarged with advancing age. Prior to fusion of the foci the femoral head was irregular in its shape and texture (Figs 1 to 3).

The average age of fusion of the multiple foci was five years and six months (range three years to six years and nine months). In every case, the bony epiphysis attained normal density and texture, though it was always slightly flattened (Fig. 1). The height of the bony epiphysis was less than in normal hips of the same age or less than the height of the normal side in unilateral cases.

Secondary radiological deterioration into true Perthes' disease was not observed in any of our patients; five (27%) of the children showed a central dimple defect in the articular surface of the femoral epiphysis.

Technetium-99 MDP bone scans were performed on three children using a high resolution program with images in the anterior and the frog-leg positions. The three-phase scan demonstrated that the radionuclide angiogram (phase I), the blood pool images (phase II) and the bone-turnover delayed images (phase III) were all normal.

Magnetic resonance imaging (MRI) was conducted on two children aged six years (Fig. 3) and four years respectively, with bilateral involvement. Coronal reconstructions and axial sections were also made. Images were obtained from both hips using a variety of spinecho pulse sequences designed to extract T1- and T2-weighted information. The appearance of the epiphyses was normal except for the 'fragmentation' of the bony nuclei and the reduced height of the cartilaginous epiphysis. In all sections examined there was uniformity of signal intensity, suggesting homogeneous vascularisation of the epiphyseal cancellous bone, the articular cartilage, the physis and the cortical bone.

**DISCUSSION**

Dysplasia epiphysealis capitis femoris appears to be a definite entity, a symptomless developmental disorder of the hip which heals completely and leaves no trace except for slight loss of epiphyseal height. Its recognition may avoid unnecessary treatment.

In most of our patients and those described by others (Meyer 1964; Harrison 1971; Emmerg, Timmermans and Leroy 1983), except those who later developed true Perthes' disease, examination of the hip revealed no physical signs and the gait was normal. Some of our bilateral cases had mild waddling gait. Maroteaux and Hedon (1981) reported 35 cases of isolated bilateral dysplasia of the hip in children under six years. In their experience, the course of Meyer's dysplasia was not as favourable as described here and the prognosis was sometimes difficult to determine. In cases seen for the first time after the third year of life, it was often difficult to distinguish DECF from bilateral Perthes' disease in the active stage or from the localised form of multiple epiphyseal dysplasia. Other conditions that may produce bilateral Perthes-like changes must be considered before the final diagnosis of DECF is made. These include hypothyroidism, spondylo-epiphyseal dysplasia tarda and congenita, and pseudo-achondroplastic dysplasia. The problem gets even more complicated, since Perthes' disease can overlap with DECF and the possibility of transitional forms has been raised (Meyer 1964). However, the appearance of a subchondral fracture line, increased density of the epiphysis and lateral displacement of the femoral head are early characteristics which are present only in Perthes' disease, and they should help to establish the true diagnosis (Catterall 1982).

The possibility that there may be dysplasia of the femoral capital epiphysis in some children who develop Perthes' disease in the first five years of life has been raised by several authors (Pedersen 1960; Meyer 1964; Laurant 1975) and has been confirmed by others (Harrison 1971; Nevelős et al 1977). It is not clear whether the capital dysplasia is congenital or is secondary to ischaemia.
The two main theories of the aetiology of DECF are the congenital vascular theory (Batory 1982a,b) and the ischaemic theory (Meyer 1964) but the evidence does not really support either of them. Congenital vascular anomalies were not demonstrated on an angiogram performed on a four-year-old boy suffering from DECF (Maroteaux and Hedon 1981) nor did it show any deformation of the cartilaginous femoral head. A bone scan performed on the same patient was also normal. The MRI studies done on our patients demonstrated normal bone marrow signal intensity. Bansahel and Bok (1979) have already reported that bone scans in DECF cases are normal. As the radionuclide angiograms, blood pool images and the standard delayed images were all normal in our patients, we believe that the ischaemic theory can be excluded.

Based on the imaging studies, it seems to us that we are dealing with a congenital focal hypoplasia of the proximal femoral epiphysis. The reduced distance from the upper edge of the metaphysis to Hilgenreiner's line suggests that the cartilaginous proximal femoral epiphysis is smaller than normal, as was demonstrated by our MRI studies. The final outcome of this hypoplastic epiphysis, as might be expected, is a small but round femoral head. Our findings disagree with those of Pedersen (1960) who described the end result as a femoral head of normal size.

It is not clear whether Perthes' disease is a single aetiological entity. There are descriptions of children with minimal Perthes-like changes, which may correlate with the segmental blood supply of the femoral head. Almost all such hips lost some epiphyseal height but had good results resembling the final outcome in DECF (Herring, Lundeen and Wenger 1980). On the other hand, Nevels et al (1977) described six boys with unilateral progressive changes in the femoral capital epiphysis, all of whom were asymptomatic when they were examined during the second year of life. These patients later developed radiological changes similar to those of severe Perthes' disease. The possibility of biomechanically induced degenerative joint changes developing later in the spherical but smaller than normal femoral heads cannot be ignored.

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REFERENCES


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