The hip in hereditary multiple exostoses
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We defined the characteristics of dysplasia and coxa valga in hereditary multiple exostoses (HME) by radiological analysis of 24 hips in 12 patients. The degree and effect of the ‘osteochondroma load’ around the hip were quantified.

We investigated the pathology of the labrum and the incidence of osteoarthritis and of malignant change in these patients. Coxa valga and dysplasia were common with a median neck-shaft angle of 156°, a median centre-edge angle of 23° and Sharp’s acetabular angle of 44°. There was overgrowth of the femoral neck with a significantly greater ratio of the neck/shaft diameter in HME than in the control hips (p < 0.05), as well as correlations between the proximal femoral and pelvic osteochondroma load (p < 0.05) and between the proximal femoral osteochondroma load and coxa valga (p < 0.01). Periacetabular osteochondromas are related to Sharp’s angle as an index of dysplasia (p < 0.05), but not coxa valga. No correlation was found between dysplasia and coxa valga.

These data suggest that HME may cause anomalies of the hip as a reflection of a generalised inherited defect, but also support the theory that osteochondromas may themselves precipitate some of the characteristic features of HME around the hip.

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Hereditary multiple exostoses (HME) is a common skeletal dysplasia with an incidence of one in 50 000.1 Osteochondromas which develop at the metaphysis of long bones in childhood are often associated with characteristic deformities of the distal radius and ulna, valgus deformities of the knee and hindfoot, and discrepancy in limb length.1-3 The deformities are often more marked within paired bones of the forearm and lower leg.

Children and adolescents with HME often have painful osteochondromas or progressive deformity. Although deep-seated osteochondromas, such as those of the proximal femur or pelvis, rarely cause symptoms or pain, deformities around the hip have been described, with premature osteoarthritis.3-5

Three large studies have been published.1,3,6 Those of Shapiro et al3 and Solomon6 describe the distribution of osteochondromas and associated deformities. An analysis of their combined 84 patients suggests that the incidence of osteochondromas of the proximal femur was 82%, and of the pelvis 62%. Schmale et al,1 however, reported an incidence of 30% involving the proximal femur and 15% involving the pelvis. The difference in the osteochondroma ‘load’ documented in these series is likely to be due to the greater number of individuals with less severe manifestations of HME in the series of Schmale et al,1 which was gathered from the general population.

In these studies, characteristic deformities of the hip include coxa valga in 25%, and occasional features of dysplasia.3 No publication has either quantified these deformities and dysplastic features, or described in detail the association between osteochondromatosis and deformity. Our aim therefore was to assess this association.

Patients and Methods

The clinical details of more than 200 patients with HME were studied. The total numbers of osteochondromas can only be identified from a complete radiological skeletal survey which involves a high dose of radiation. Since the recent elucidation of the genetics of HME suggests that the genes responsible are tumour suppressor genes,7,8 radiation carries the theoretical risk of stimulating malignant transformation in osteochondromas. This risk is greater in the pelvis which is the site where malignant transformation is
Consequently, for ethical reasons, pelvic radiographs were not routinely taken in all patients with HME. We identified patients who had undergone antero-posterior (AP) radiographs of the hip for a defined clinical purpose. We used the first chronological AP radiographs of the hip or pelvis in 12 patients of mean age 17 years (3 to 54) to measure osteochondroma load and the morphology of the proximal femur and acetabulum.

In order to assess the osteochondroma load, the number of osteochondromas and their bony area on AP radiographs were quantified by a single examiner, blind to other parameters. The bony area of each visible osteochondroma was computed by overlaying a transparent grid of 1 mm squares at three anatomical sites: a) the proximal femur; b) the periacetabular pelvis; and c) the extra-acetabular pelvis.

For the morphometry of the proximal femur we measured: a) the femoral neck-shaft angle of Muller; b) the ratio of the diameter of the femoral neck to that of the shaft as an index of overgrowth of the femoral neck; and c) Shenton’s line. The acetabular morphometry was determined using Sharp’s acetabular angle and the centre-edge angle (Fig. 1).

The acetabular morphometry was determined using Sharp’s acetabular angle and the centre-edge angle (Fig. 1). Sharp’s acetabular angle is a measurement of the obliquity of the acetabulum and is the angle subtended by a line joining the inferior point of the tear-drop to that of the opposite hip, and the line joining the superolateral to the inferomedial margin of the acetabulum. The centre-edge angle is the angle subtended by a line parallel to the longitudinal axis of the body through the centre of the femoral head and a line from the centre of the femoral head to the superior rim of the acetabulum.

Radiographs of the hips of 12 patients of a median age of 13 years (4 to 62), were used as controls. These were reported as normal by a specialist radiologist, and had been...
taken either for ‘irritable hip’ syndrome in children or for the assessment of the adjacent hip in adults with disorders of the knee.

Since pairs of hips from the same individual could not be considered independently, statistical analyses were undertaken on the mean value for each pair of hips. Non-parametric statistical tests were used. Comparison between sample values was assessed using the Wilcoxon rank-sum test on unequal samples. Correlation of parameters was determined by Kendall’s coefficient of rank correlation (tau).

**Results**

There was no significant difference in age or gender between the groups. **Individual features.** One patient developed chondrosarcomatous change in a proximal femoral osteochondroma.

Correlation of pelvic and proximal femoral osteochondroma load in paired hips from the same individual.
Table I. The parameters assessed in the paired hips (HME and controls) using the Wilcoxon rank-sum test

<table>
<thead>
<tr>
<th>Parameter assessed in paired hips</th>
<th>Number</th>
<th>Median</th>
<th>Somer’s ‘D’</th>
<th>p value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Femoral neck-shaft angle, Shenton’s line</td>
<td>In HME</td>
<td>4</td>
<td>150°</td>
<td>0.75</td>
</tr>
<tr>
<td></td>
<td>Broken</td>
<td>6</td>
<td>168°</td>
<td></td>
</tr>
<tr>
<td>Sharp’s acetabular angle, Shenton’s line</td>
<td>In HME</td>
<td>4</td>
<td>42°</td>
<td>0.083</td>
</tr>
<tr>
<td></td>
<td>Broken</td>
<td>6</td>
<td>44°</td>
<td></td>
</tr>
<tr>
<td>Centre-edge angle of Wiberg, Shenton’s line</td>
<td>In HME</td>
<td>4</td>
<td>27°</td>
<td>0.333</td>
</tr>
<tr>
<td></td>
<td>Broken</td>
<td>6</td>
<td>28°</td>
<td></td>
</tr>
<tr>
<td>Femoral neck/shaft diameter</td>
<td>In HME</td>
<td>11</td>
<td>1.6</td>
<td>0.515</td>
</tr>
<tr>
<td></td>
<td>In control hips</td>
<td>12</td>
<td>1.23</td>
<td></td>
</tr>
<tr>
<td>Sharp’s acetabular angle, periacetabular exostoses</td>
<td>Absent</td>
<td>7</td>
<td>42°</td>
<td>0.905</td>
</tr>
<tr>
<td></td>
<td>Present</td>
<td>3</td>
<td>48°</td>
<td></td>
</tr>
<tr>
<td>Centre-edge angle of Wiberg, periacetabular exostoses</td>
<td>Absent</td>
<td>7</td>
<td>29°</td>
<td>0.619</td>
</tr>
<tr>
<td></td>
<td>Present</td>
<td>3</td>
<td>21°</td>
<td></td>
</tr>
<tr>
<td>Femoral neck/shaft diameter, periacetabular exostoses</td>
<td>Absent</td>
<td>7</td>
<td>1.64</td>
<td>0.429</td>
</tr>
<tr>
<td></td>
<td>Present</td>
<td>3</td>
<td>1.17</td>
<td></td>
</tr>
<tr>
<td>Femoral neck-shaft angle, periacetabular exostoses</td>
<td>Absent</td>
<td>7</td>
<td>151°</td>
<td>0.048</td>
</tr>
<tr>
<td></td>
<td>Present</td>
<td>3</td>
<td>155°</td>
<td></td>
</tr>
</tbody>
</table>

* not significant

Table II. The first and second parameters for correlation of paired hips (HME and control) showing Kendall’s coefficient of rank correlation (tau)

<table>
<thead>
<tr>
<th>First parameter</th>
<th>Second parameter</th>
<th>Number</th>
<th>Kendall’s tau</th>
<th>p value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Proximal femoral osteochondroma load</td>
<td>Pelvic osteochondroma load</td>
<td>11</td>
<td>0.564</td>
<td>&lt;0.05</td>
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<tr>
<td>Proximal femoral osteochondroma number</td>
<td>Pelvic osteochondroma number</td>
<td>11</td>
<td>0.139</td>
<td></td>
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<tr>
<td>Sharp’s acetabular angle</td>
<td>Centre-edge angle of Wiberg</td>
<td>11</td>
<td>-0.722</td>
<td>&lt;0.01</td>
</tr>
<tr>
<td>Proximal femoral osteochondroma load</td>
<td>Femoral neck-shaft angle</td>
<td>11</td>
<td>0.709</td>
<td>&lt;0.01</td>
</tr>
<tr>
<td>Proximal femoral osteochondroma load</td>
<td>Femoral neck/shaft diameter</td>
<td>11</td>
<td>0.436</td>
<td>&lt;0.05</td>
</tr>
<tr>
<td>Pelvic osteochondroma load</td>
<td>Femoral neck-shaft angle</td>
<td>11</td>
<td>0.527</td>
<td>&lt;0.05</td>
</tr>
<tr>
<td>Pelvic osteochondroma load</td>
<td>Femoral neck/shaft diameter</td>
<td>11</td>
<td>0.455</td>
<td>&lt;0.05</td>
</tr>
<tr>
<td>Sharp’s acetabular angle</td>
<td>Femoral neck-shaft angle</td>
<td>11</td>
<td>-0.165</td>
<td></td>
</tr>
<tr>
<td>Centre-edge angle of Wiberg</td>
<td>Femoral neck-shaft angle</td>
<td>11</td>
<td>-0.018</td>
<td></td>
</tr>
</tbody>
</table>
Although three hips showed evidence of degenerative disease, only one patient required total hip arthroplasty, at the age of 54 years. This hip showed more osteochondromatosis, more coxa valga and more severe dysplasia than the contralateral side (Fig. 2). By contrast, the hip which developed sarcomatous change had a normal neck-shaft angle (Fig. 3) One patient with marked acetabular dysplasia had clinical and radiological features of labral pathology (Fig. 4) and lateral subluxation of the femoral head. A shelf acetabuloplasty was undertaken.

**General features.** All paired hip measurements subjected to statistical analysis are shown in Table I.

Proximal femoral osteochondromas were more prominent medially than laterally in most hips and had a sessile appearance, whereas those in the pelvis were more pedunculated. No intra-articular osteochondromas were identified. The degree of osteochondromatosis in the proximal femur correlated with that in the pelvis (Fig. 5).

There was significant coxa valga (mean angle 156°) in patients whose hips were affected compared with normal subjects (Fig. 6). Shenton’s line was broken in both hips in seven patients. There was a significantly greater neck-shaft angle when Shenton’s line was broken than when intact. By contrast, whether Shenton’s line was broken or not was not related to dysplasia (Table I).

The femoral neck diameter/femoral shaft diameter ratio

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The femoral neck diameter/femoral shaft diameter ratio
was significantly greater in the affected than in the control hips (Table I).

The centre-edge and Sharp’s angles showed a strong inverse correlation (Table II). In patients with HME, the centre-edge angle (mean 23°) was less, and Sharp’s angle greater (mean 44°) than that described in the general population, suggesting a tendency to dysplasia (Fig. 7). The indices of dysplasia did not correlate with coxa valga (Table II).

Increasing osteochondroma load, particularly in the proximal femur, correlated with an increasing neck-shaft angle (Fig. 8) and some correlation with the ratio of the neck shaft/diameter diameter was also seen (Table II).

Only three patients had periacetabular osteochondromas in both hips, which precluded a correlation analysis. Sharp’s angle was significantly greater in those patients than in those with no osteochondroma in either hip (Figs 9 and 10, Table I). By contrast, no significant relationship was found between the presence or absence of periacetabular osteochondromas and either the neck-shaft angle or the neck diameter/shaft diameter ratio (Table I).

**Discussion**

The severity of deformities of the hip identified in this series is likely to exceed that found in the overall population with HME, since only patients with a clinical indication for imaging were studied. These indications included...
pain, a limited range of movement and a leg-length discrepancy. Our data reflect the cohort with more significant hip pathology and may therefore be of greater clinical significance.

The complication of arthritis of the hip was noted in the review of Scarborough and Moreau, but has not been documented or quantified in any community or hospital-based study. Only 1% of our patients with HME have required total hip arthroplasty. Known aetiological factors for premature degenerative arthritis include acetabular dysplasia and coxa valga; both marked features of the hip which required replacement.

The hip with sarcomatous change showed no features of dysplasia. Malignant change occurs as a result of clonal expansion of a single abnormal cell, and its initiation and progression are likely to be independent of the degree of deformity around the hip. Patients who develop chondrosarcoma do not seem to have more severe manifestations of HME.

A symptomatic labral tear and associated cyst were diagnosed in one patient. A torn labrum is often associated with acetalubar dysplasia and it has been proposed that these tears are degenerative, occurring as a result of abnormal stresses imposed by the uncovered lateral portion of the femoral head.

Since the genetic stimulus for the development of an osteochondroma in a patient with HME is identical at each bony site, but differs from individual to individual, the correlation between the osteochondroma load of the proximal femur and that of the pelvis supports the concept that the severity of the disease reflects a greater or lesser stimulus to the formation of osteochondroma inherited by each individual.

The increased ratio of the neck/shaft diameter in patients with HME may simply reflect the presence of sessile osteochondromas of the femoral neck and therefore explain the correlation with the osteochondroma load of the proximal femur.

The presence of coxa valga in almost all patients with HME may be considered to be a developmental consequence of the other major feature of the condition, acetabular dysplasia. Such a relationship has been proposed, for example, in some children with developmental dysplasia of the hip. We were unable to demonstrate this in our series in which the presence of periacetabular osteochondromas related to acetabular dysplasia and the degree of coxa valga, correlated best with the osteochondroma load of the proximal femur.

Shenton’s line is broken in anatomical abnormalities of the hip. This is expected in the presence of osteochondromas of the medial femoral neck and in significant coxa valga, because of the abnormal alignment of the femoral neck in relation to the pelvis. Since clinical manifestations of HME present, on average, at three years of age, usually at sites other than the hip, it is possible that the dysplasia associated with HME reflects a different phenomenon from that of developmental dysplasia, developing more gradually, and at a later age. A physical phenomenon may, in part, be responsible. In several hips there is an anatomical potential for lateral subluxation of the femoral head because of impingement of an osteochondroma of the medial femoral neck on the inferomedial acetabular rim during adduction of the hip (Fig. 10).

There is clinical evidence that the degree of shortening of the forearm in HME may be a consequence of osteochondromatosis at the distal ulnar or radial metaphysis, a concept sympathetic to the molecular aetiology of HME which involves inactivation of a tumour suppressor gene. Since periacetabular osteochondromas relate to acetabular dysplasia, and proximal femoral osteochondromas to coxa valga and overgrowth of the femoral neck, it is possible that local activity of the osteochondroma may precipitate at least a proportion of the local deformities. A minor degree of coxa valga, however, was seen in three hips without radiological evidence of proximal femoral osteochondromatosis, suggesting there may be a role for a generalised effect of HME on deformity of the hip in the absence of the formation of an osteochondroma. Of particular interest is the possibility that HME-associated acetalubar dysplasia may not be a consequence of inadequate infantile location of the femoral head in the acetabulum, but may result from the local effects of growth of periacetabular osteochondromas. Lateral subluxation of the femoral head may then occur as a result of acetabular dysplasia, medial impingement or a combination of both. No benefits in any form have been received or will be received from a commercial party related directly or indirectly to the subject of this article.

References