THE NATURAL HISTORY AND EARLY TREATMENT OF PROXIMAL FEMORAL DYSPLASIA

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Proximal femoral dysplasia (proximal femoral focal deficiency) in early infancy is characterised by considerable shortening in association with a stable hip which is freely mobile from a position of some fixed flexion and lateral rotation (Fig. 1). The radiograph confirms that the femur is short, and the proximal third of the femoral shaft, trochanteric area, neck and head are seemingly absent (Fig. 2). The radiological signs are therefore incompatible with the clinical findings; for were such a defect present the hip would be totally unstable and would lack fixed deformity. The inference to be drawn is that there is continuity between the femoral head and the proximal end of the shortened femoral shaft. The intervening transradiant area is occupied by a cartilaginous model in which ossification is delayed. As growth proceeds two differing patterns of development emerge. When this is favourable, the anlage gradually ossifies and stability is maintained, so that finally the proximal femur resembles the normal though the shaft remains very short (Figs. 2 and 3). The outcome is unfavourable when one or more pseudarthroses develop at the osteocartilaginous junction or within the cartilaginous anlage so that continuity between the hip and femoral shaft is lost. Instability

FIGS. 1 AND 2
Figure 1—The typical clinical appearance of proximal femoral dysplasia of the left femur at 19 months. The femur is short, flexed and laterally rotated. Figure 2—Proximal femoral dysplasia (stable type) at 3 days. Note the bulbous proximal end and angulation of the femoral shaft which lies considerably lateral to a well formed acetabulum.
Proximal femoral dysplasia (unstable type) at 20 months showing upward migration of the femoral shaft relative to the femoral head, which is present in the acetabulum. The proximal end of the femoral shaft is blunt and irregular. Sclerosis is present below the blunted end.

**Fig. 3**
Radiograph of the patient shown in Figure 2 at 18 months. No treatment had been given. Spontaneous ossification of the femoral neck has occurred and stability has been maintained. The sclerosis, not well shown in Figure 2, is at the site of angulation of the femoral shaft, which is correcting spontaneously.

**Fig. 4**
Proximal femoral dysplasia (unstable type) at 20 months showing upward migration of the femoral shaft relative to the femoral head, which is present in the acetabulum. The proximal end of the femoral shaft is blunt and irregular. Sclerosis is present below the blunted end.
is then inevitable because the femoral shaft migrates upwards in relation to the head and neck. Progressive coxa vara is followed by breakdown of the pseudarthrosis, which allows the femoral shaft to displace upwards and away from the proximal segment, which remains in contact with the acetabulum (Fig. 4).

In this paper we will describe certain radiological features which are helpful in distinguishing between those infants in whom a favourable outcome is likely, and those destined to develop pseudarthrosis and instability. This distinction has an important influence upon early management, which will also be discussed.

**RADIOLOGICAL SIGNS IN RELATION TO PROGNOSIS**

Our criteria for distinguishing between those who will proceed to spontaneous healing and those who will not are based on a close study of twenty-five patients, five of whom had bilateral involvement (thirty hips). All have been observed until evolution into a stable or unstable variety has become established. Twelve hips were explored and have provided us with anatomical evidence to correlate with the radiological signs. Three hips in the unstable group were explored more than once. A further nineteen patients were reviewed but excluded for a variety of reasons, such as lack of suitable radiographs in early infancy or incomplete follow-up.

**TABLE I**

**LENGTH OF THE OSSIFIED FEMORAL SHAFT (UNILATERAL CASES)**

<table>
<thead>
<tr>
<th>Number of hips</th>
<th>Stable</th>
<th>Unstable</th>
</tr>
</thead>
<tbody>
<tr>
<td>Shaft length greater than half normal side</td>
<td>6</td>
<td>5</td>
</tr>
<tr>
<td>Shaft length less than half normal side</td>
<td>14</td>
<td>6</td>
</tr>
</tbody>
</table>

**TABLE II**

**DISTANCE OF THE PROXIMAL END OF THE OSSIFIED SHAFT FROM THE ACETABULUM (UNILATERAL CASES)**

<table>
<thead>
<tr>
<th>Number of hips</th>
<th>Stable</th>
<th>Unstable</th>
</tr>
</thead>
<tbody>
<tr>
<td>Distance greater than the normal side</td>
<td>15</td>
<td>10</td>
</tr>
<tr>
<td>Distance equal to the normal side</td>
<td>2</td>
<td>0</td>
</tr>
<tr>
<td>Distance less than the normal side</td>
<td>3</td>
<td>1</td>
</tr>
</tbody>
</table>

The acetabulum—When the acetabulum resembles the normal the femoral head is always present although ossification may be delayed (Fig. 3 and 4). This agrees with the findings of Aitken, Amstutz and King (1969). If the acetabulum is present but less well formed a head may still appear, but should acetabular dysplasia persist dislocation of the hip may follow (Aitken 1969, Amstutz 1969). If there is no apparent acetabulum the head is absent.

**The length of the ossified femoral shaft**—This is not directly related to the presence of a femoral head; when no more than the supracondylar part of the femur is visible a femoral head will appear if the acetabulum is well formed. Nevertheless, the more severe the shaft deficit, the more likely is the acetabulum to be dysplastic. It is difficult to estimate accurately the length of the ossified shaft in relation to the normal because flexion deformity favours apparent shortening in the antero-posterior radiograph. In general, however, the shorter the ossified part of the shaft the less the likelihood of spontaneous healing (Table I).
The distance of the proximal end of the ossified shaft from the acetabulum—Contrary to expectation, a large gap between the proximal end of the ossified shaft and the acetabulum was not necessarily a bad prognostic sign. In ten of eleven hips which became stable spontaneously this distance was greater than on the opposite, normal side (Figs. 2 and 5).

Table II). In nine unstable hips the initial distance was greater than the normal side in five hips, equal to the normal side in two and less than the normal side in two (Table II). Whatever the initial distance, all unstable hips later showed progressive migration of the femoral shaft upwards and sometimes inwards. This is a valuable sign, indicating impending dissolution of a pseudarthrosis (Fig. 4). We may assume that in stable cases the cartilaginous anlage remains intact and prevents the upward and inward movement of the proximal end of the shaft.

**TABLE III**

<table>
<thead>
<tr>
<th>Type</th>
<th>Appearance</th>
<th>Number of hips</th>
<th>Stable</th>
<th>Unstable</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>Bulbous</td>
<td>12</td>
<td>12</td>
<td>0</td>
</tr>
<tr>
<td>II</td>
<td>Tuft or cap</td>
<td>13</td>
<td>3</td>
<td>10</td>
</tr>
<tr>
<td>III</td>
<td>Not bulbous, no cap</td>
<td>5</td>
<td>0</td>
<td>5</td>
</tr>
</tbody>
</table>

The appearance of the proximal end of the ossified shaft—This is a most important feature in establishing prognosis. Three types are recognisable.

**Type I**—If the proximal end of the femur is truly bulbous (Figs. 2, 5 and 9) continuity of the head, neck and greater trochanter always follows (Amstutz 1969). This sign was present in twelve out of thirty hips, all of which became stable proximally (Table III). Nevertheless a pseudarthrosis may develop below the greater trochanter within the ossified shaft. This occurred in three hips. In one, the pseudarthrosis healed spontaneously (Figs. 6 and 7).
Proximal femoral dysplasia (stable type) at 10 days showing an established subtrochanteric pseudarthrosis with an area of sclerosis beneath it. Note that the distance between the acetabulum and the bulbous end of the proximal femoral shaft is greater than on the normal side.

Radiograph of the same patient as in Figure 6 at 17 months. Without treatment ossification of the femoral head has occurred without loss of stability, and spontaneous healing of the subtrochanteric pseudarthrosis has taken place.
Type II—If a tuft or cap of ossification separated from a blunt proximal shaft by an area of transradiance is seen by the age of one year, the hip is likely to be unstable (Figs. 8 and 9). Aitken (1969) made a similar observation in his class B patients which have a good acetabulum but fail to achieve stable continuity. This appearance was seen in thirteen hips. Ten became unstable and three, all in patients with bilateral involvement, became stable (Table III). However, two of these three stable hips developed such severe varus deformity due to untreated subtrochanteric pseudarthrosis that they became dislocated inferiorly.

Type III—Five hips showed no tuft at one year and were not bulbous (Figs. 4 and 9). The proximal end of the ossified shaft was either blunt and irregular or pointed. All five became unstable (Table III).

The site and nature of sclerosis in the femoral shaft—Sclerosis in both stable and unstable defects was related either to the site of angulation of the shaft or to a pseudarthrosis. In the stable defects it occurred well below the proximal end of the shaft and was associated with angulation of the shaft (Figs. 2, 5 and 9). This became spontaneously corrected in ten (Fig. 3) and developed into a subtrochanteric pseudarthrosis in five (Fig. 13). In the unstable defects the sclerosis was always immediately distal to the site of pseudarthrosis or angulation and had the appearance of an inverted V (Figs. 4, 8 and 9).

![Fig. 8](image)

Proximal femoral dysplasia (unstable type) at 2 months, showing a tuft or cap of ossification separated from a blunted proximal femoral shaft by an area of translucency. Sclerosis in the shape of an inverted ‘V’ is present beneath the blunt end of the shaft. At this age upward and inward migration of the shaft has not yet occurred.

![Fig. 9](image)

Diagram from tracings of radiographs showing the typical appearance of the proximal end of the ossified femoral shaft in the stable and unstable types. The shaded area represents the site of sclerosis.

Time of appearance of the femoral head—Early radiographs were usually too infrequent to establish the time of ossification of the femoral head with precision, but this tended to be later in the unstable group.

Thalidomide poisoning—There was a history of thalidomide poisoning in two patients, one in each group. So far as their femurs were concerned they differed in no way from those not
exposed to thalidomide. King (1969) pointed out that the head of the femur and the acetabulum differentiate from a common anlage in the foetus at nine weeks of gestation and postulated some insult to the embryonic lower limb at this time as the cause of proximal femoral dysplasia.

**IMPLICATIONS OF A KNOWLEDGE OF PROGNOSIS**

The rational management of proximal femoral dysplasia is dependent upon the identification of those likely to develop instability, so that measures to prevent this occurring may be taken before continuity is lost.

An untreated pseudarthrosis in the proximal femoral region will become disrupted under the stress of weight-bearing; displacement of the distal component follows, and femoro-pelvic stability will be lost. This is a most undesirable complication when the leg is so short that a prosthesis will inevitably be required as the only practical means of restoring equality of length. Stability and mobility at the hip joint are of the greatest value to the limb fitter, for he may then fashion a prosthesis comparable to that used by an above-knee amputee, whether or not the knee is stabilised or the foot amputated in the future. With an unstable hip, his problems are increased and the patient's disability aggravated. Hitherto our practice has been to recommend exploration of the proximal end of the femur in all patients about one year. Our purpose was to establish the exact anatomy of the defect and to reinforce the potentially weak osteochondral junction or repair a pseudarthrosis by bone grafting before weight-bearing had caused either a fracture at the osteochondral junction or displacement of the pre-existing pseudarthrosis (Lloyd-Roberts and Stone 1963). Knowledge derived from these operations, observations of untreated patients and a study of the literature have led us to recognise that some patients may be expected to retain their initial stability. In these, operation is unnecessary and an expectant attitude is justified. We have also learnt to recognise those who are likely to develop pseudarthrosis and instability. For these, operation remains the logical approach if the undesirable outcome of pseudarthrosis and instability is to be avoided. It is, for example, chastening to confess that of the two patients reported as having successful results from bone grafting in 1963, one (Case 2) was operated upon unnecessarily. It is, however, justifiable to claim that the other (Case 1) gained a successful result from a necessary operation.

**TREATMENT**

Those patients who exhibit the radiological signs which we have described as characteristic of a good prognosis (progressive ossification of the cartilaginous anlage without development of a pseudarthrosis) may be supervised by repeated radiographs alone. If their development is favourable they may be referred to a limb fitting centre at about ten months. If, with time, uncertainty arises it is better to explore the abnormal hip rather than to risk displacement later.

**INDICATIONS FOR OPERATION**

Operation is recommended for those with radiological signs indicating that the outcome will be unfavourable, and for those in whom a trochanteric or cervical pseudarthrosis is known to be present. In addition, delayed incorporation or failure of a stabilising graft demands re-exploration. A dislocated, delayed incorporation or failure of a stabilising graft demands re-exploration. A dislocated Delayed incorporation or failure of a stabilising graft demands re-exploration. A dislocated Delayed incorporation or failure of a stabilising graft demands re-exploration. A dislocated Delayed incorporation or failure of a stabilising graft demands re-exploration. A dislocated Delayed incorporation or failure of a stabilising graft demands re-exploration. A dislocated Delayed incorporation or failure of a stabilising graft demands re-exploration. A dislocated Delayed incorporation or failure of a stabilising graft demands re-exploration. 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signs of incorporation and ossification is proceeding in the pre-existing cartilage. We have also used a medial displacement osteotomy to support the graft and encourage ossification when incorporation of the first or delay in the second is disquieting (Lloyd-Roberts and Stone 1963). We have succeeded in stabilising three of six unstable hips in this manner.

**Impaction of shaft into femoral head** (King 1969)—We have used this method four times in two circumstances. First if there are two pseudarthroses, one at the osteochondral junction and one through the cartilage at the femoral neck, the central fragment seems too mobile and devitalised to be worth preserving. The second indication arose during operations on two children in whom previous grafting operations had failed. Scarring, displacement and deformity permitted no alternative procedure. The femoral shaft is fashioned into a spike which is driven into a pre-drilled hole which transfixes the femoral head. Cancellous iliac bone is placed around the junction and the area stabilised by a Kirschner wire (Figs. 11 and 12).
Figure 13—Radiograph of an established subtrochanteric pseudarthrosis in the stable type of proximal femoral dysplasia at 15 months. Figure 14—Radiograph of the same patient as in Figure 13, showing healing of the subtrochanteric pseudarthrosis three and a half years after excision, onlay bone grafting and intramedullary nail fixation. The nail was removed four months after operation.

Figure 15—Radiograph of a patient two years after pelvi-femoral arthrodesis.
Cancellous onlay bone grafting with intramedullary fixation for persisting subtrochanteric pseudarthrosis—In two patients healing did not occur spontaneously but union was achieved by this method (Figs. 13 and 14). In one patient with bilateral pseudarthrosis the condition was untreated.

Open reduction of dislocated hip—One patient with an otherwise stable femoral defect was operated upon. Reduction presented no great difficulty but unacceptable tension associated with considerable valgus and anteversion deformity dictated femoral osteotomy with shortening at the same time. The early outcome is promising.

Pelvi-femoral arthrodesis—This method was used in two patients. In both, the femoral head had not been found during a previous exploration of the upper end of the femur. In one, pelvi-femoral arthrodesis was performed immediately. The femoral shaft has remained stable but bony union has not occurred. In the other a fibular graft was placed in the cartilaginous upper end of the femur. This resorbed and a femoral head became visible on the radiograph at seventeen months. The hip was re-explored but the femoral head was no longer mobile in the acetabulum. Pelvi-femoral arthrodesis was performed and at three and a half years satisfactory bony union is present (Fig. 15). As shortening progresses we intend to shorten the femur further and perform an extension supracondylar osteotomy. We hope thereby to achieve a stable hinge joint, below which a prosthesis may be fitted.

SUMMARY AND CONCLUSIONS

1. Early recognition of femoro-pelvic instability in proximal femoral dysplasia is essential to rational management.
2. Certain radiological signs helpful in identifying such patients within the first year are described.
3. Early operation is recommended to prevent displacement of a pseudarthrosis and to encourage healing. Alternative methods are described for established defects.
4. Radiological signs are described which indicate that healing without loss of femoro-pelvic stability will occur. Such patients may be managed expectantly.

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REFERENCES


