CONGENITAL HYPOPLASIA OF THE UPPER FEMUR


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The treatment of this rare anomaly is infrequently considered in the literature. We feel justified therefore in reporting the results of surgical treatment in two patients in whom a measure of success has been achieved.

CASE REPORTS

Case 1—This boy was seen four years ago at the age of three months with a short left leg. The radiograph showed that the proximal end of the femur appeared to be absent (Fig. 1). He was admitted two months later. Traction was applied for one week with the leg in suspension and then in full abduction for a further week. The position of the femur remained unchanged. When arthrography was done the needle was felt to enter firm material of cartilaginous consistency in the area normally occupied by the proximal end of the femur. The contrast radiograph showed a negative shadow in the acetabulum similar to the femoral head and continuous with a vaguer shadow which was interpreted as the trochanteric area and upper part of the shaft of the femur (Fig. 2).

Operation—At operation the upper left femur was explored. There was a normal-looking femoral head, neck and trochanteric region formed in cartilage which ended below in a bulbous swelling; this was a pseudarthrosis and it was continuous below with the conical sclerotic bone of the femoral shaft (Fig. 3). An abduction osteotomy was done, a wedge being removed at the level of the pseudarthrosis. A fibular graft was driven from the lateral side of the base of the greater trochanter into the cartilaginous head. Sclerotic bone was removed from the proximal end of the lower fragment, the rawed surface was placed in contact with the protruding fibular graft and cancellous iliac grafts were onlaid. The leg was immobilised in plaster in abduction.

Progress—A radiograph six weeks after operation showed union between the fibular graft and the femoral shaft. New bone was also present between the ilium and the femur (Fig. 4). When the plaster was removed five months later the hip was stiff. The radiograph revealed a bar of ectopic bone bridging ilium and femur, and union at the site of the pseudarthrosis. The ectopic bone was partly excised a year after the first operation and mobility was temporarily restored, but the bone rapidly re-formed and the hip became fixed in 20 degrees of flexion and 20 degrees of abduction. After a further year shortening was three and a quarter inches but femoral lengthening has reduced the discrepancy to two inches (Fig. 5). It is proposed to repeat the femoral lengthening and to excise the now mature ectopic bone.

Case 2—This girl was admitted at the age of four months with shortening of the left femur. The left hip lacked 45 degrees of abduction and 10 degrees of medial rotation. No other abnormality was found. An earlier radiograph had revealed seeming absence of the upper end of the femur (Fig. 6), but arthrography showed the outline of a normal femoral head (Fig. 7).

Operation—At operation the head and neck of the femur were found to be cartilaginous—coxa vara was present—and this cartilage was continuous with the bone of the femoral shaft. A tibial graft was inserted into the femoral head through the trochanteric region (Fig. 8).

Progress—Five months later a radiograph showed the capital epiphysis to be visible. Seventeen months after the operation the hip movements were almost full and a radiograph showed union (Fig. 9). Abduction osteotomy has since been performed to correct the coxa vara.
Case 1. Figure 1—At the age of three months the radiograph showed an apparent absence of the proximal end of the left femur. Figure 2—An arthrogram of the left hip at the age of five months showed that the femoral head was present.

Case 1. Figure 3—A photograph at operation showing the normal femoral head. Figure 4—A radiograph six weeks after operation showing union between the fibular graft and the femoral shaft. Ectopic bone is present between the ilium and the femur. Figure 5—A radiograph eighteen months after operation. The femur has been lengthened, reducing the leg inequality from three and a quarter to two inches.
Case 2. Figure 6—A radiograph showing the seeming absence of the upper end of the left femur. Figure 7—An arthrogram showing the normal-shaped femoral head.

Case 2. Figure 8—A radiograph after operation showing the tibial graft in position. Figure 9—A radiograph seventeen months after operation.
DISCUSSION

Morgan and Somerville (1960) described a case with similar anatomical findings in whom drilling of the cartilaginous fragment failed to promote calcification. They recommended, however, that further attempts should be made to treat this anomaly by surgery.

Van Nes (1950) discussed treatment in older children in whom stabilisation of the hip and arthrodesis of the knee with rotation through 180 degrees allowed the now reversed ankle to act as a substitute for the knee joint.

The expected shortening in untreated cases is between ten and fifteen inches (Amstutz and Wilson 1962). This is due not only to femoral shortening but also to upriding of the femoral shaft. If there is a pseudarthrosis between the shaft and the cartilage of the trochanteric region, upward displacement of the shaft is likely to occur, leaving the femoral head and the neck behind. If continuity exists, severe coxa vara may precede and predispose to pseudarthrosis. In some children coxa vara may be the sole cause of shortening (Ring 1961). It is, therefore, to be recommended that exploration be done early in life, preferably before the child stands, to establish with certainty the anatomy of the anomaly; this may then be treated before weight bearing has caused upward displacement of the femoral shaft. This assessment cannot be made by plain or contrast arthrography, for even the latter fails to distinguish between pseudarthrosis and stability at the osteochondral junction.

In the two children reported here bone grafts appeared to stimulate ossification of the pre-existing cartilage, thus strengthening the femoral neck and preventing severe coxa vara. In the first child the same graft obtained union at the pseudarthrosis. If, in these two children, shortening ultimately dictates amputation, the stability of their hips should make it possible to fit them with a standard above-knee or below-knee prosthesis instead of an extension type. It is hoped, however, that leg lengthening will make this unnecessary.

SUMMARY

1. The surgical management of two children with congenital hypoplasia of the upper end of the femur is described.
2. Early exploration is advocated to establish the nature and extent of the anomaly and to attempt its correction.
3. The preliminary results are sufficiently encouraging for us to recommend that further attempts at surgical treatment be considered in patients with this disorder.

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REFERENCES


