CLASSIFICATION AND MANAGEMENT OF CONGENITAL ABNORMALITIES OF THE FEMUR

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Sixty-nine patients with congenital abnormality of the femur were reviewed. Their manifestation of femoral dysplasia ranged from an intact femur approximately 60 per cent of the length of the normal leg to a subtotal absence of the femur in which only the femoral condyles remained, often with a congenital fusion of the knee joint. Two groups were defined: Group I consisted of those with congenital hypoplastic femur in which the hip and knee could be made functional and where, in some patients at least, leg equalisation was possible; Group II consisted of those with true proximal focal femoral deficiency where the hip joint was never normal and the knee joint was always useless. The patients in each group were examined and evaluated with respect to clinical signs, surgical procedures performed, and prosthetic requirements and function. A protocol of treatment for both groups is suggested.

The established classifications of congenital abnormalities of the femur concentrate on detailed examination of radiographs. The best known classifications in the English literature are those of Aitken (1959), and Amstutz and Wilson (1962). Aitken’s classification concentrates on the presence or absence of cartilaginous continuity of the proximal femur and the development of the hip joint. The classification of Amstutz and Wilson attempts a detailed breakdown of all the anatomical types from a mild degree of femoral hypoplasia to subtotal absence of the femur. Hamanishi (1980) defined five types which with subtypes gives 10 groups of patients.

A review of our patients reveals that a purely radiographic classification of these patients at birth will produce a collection of types limited only by the number of patients seen. Furthermore this classification must change with the varying degree of ossification that occurs with growth and in essence the radiographic abnormalities seen represent a spectrum of abnormal development from mild to severe in deficiency, a finding also documented by Hamanishi (1980).

The purpose of this paper is not to provide a classification to supplant those of Aitken (1959), Amstutz and Wilson (1962), Hamanishi (1980) and others; these we recognise as essential in the attempt to understand the range of pathological anatomy that may exist. Our purpose is to suggest that such classifications do not include all possible variations because they are all inevitably based on small series of cases, and to show that the great majority of patients fall into two major groups which can be differentiated on clinical grounds.

CLASSIFICATION

Sixty-nine patients were reviewed. All had attended the Amputee Clinic at the Ontario Crippled Children’s Centre and their surgical operations had been performed at The Hospital for Sick Children, Toronto. The patients, their surgical and prosthetic histories and their radiographs were examined. All the patients included in this study had severe degrees of femoral dysplasia. Those patients with relatively minor degrees of femoral shortening and normal anatomical configuration of the femoral head and neck were excluded.

We are, therefore, discussing that group of patients whose mildest manifestation of femoral dysplasia is an intact femur that is approximately 60 per cent of the length of the normal side, and whose most severe manifestation is a subtotal absence of the femur in which only the femoral condyles remain, often with a congenital fusion of the knee joint. In the classification of Amstutz and Wilson (1962) and in the minds of many referring orthopaedic surgeons the above extremes and the intermediate degrees of dysplasia are considered to represent varying degrees of proximal focal femoral deficiency. This concept of the problem is unnecessarily complex in view of the great number of types that it is possible to create in a classification based on the radiographic appearance (Amstutz and Wilson 1962; Hamanishi 1980).

We found that in spite of the variable appearance of radiographs, especially in the more severe forms of proximal focal femoral deficiency, when the patients
were studied from the clinical point of view the vast majority could be placed in only two groups. These two groups differed both clinically and radiographically and were ultimately quite different functionally and therefore in their surgical and prosthetic requirements.

In our view much of the previously published material on this subject fails to assess surgical results in relation to final prosthetic function. The relationship between surgical procedures and prosthetic fitting and function cannot be overstressed.

At birth all infants with femoral deficiency present with similar appearance of an extremely short femur with hip and knee held in flexion (Figs 1 and 2). The appearance of the radiographs varies from one in which the entire shaft and proximal femur is missing to an appearance of a short femur in which the proximal end looks fairly normal but lies laterally and proximally displaced, suggesting a lesser degree of cartilaginous defect or sometimes a congenital dislocation of the hip.

The importance of accurate diagnosis in the neonatal period cannot be overstressed. If a blanket diagnosis of proximal focal femoral deficiency is made then major errors of management may be committed—such as early amputation of a potentially functional foot, and inappropriate arthrodesis of the knee or exploration of the hip to repair a non-existent cartilaginous defect or an irreducible congenital dislocation of the hip.

Our plea is for the recognition of two major groups. Group I or congenital hypoplastic femur in which the hip and knee are or can be made functional and where, in some patients at least, leg equalisation may be possible; and Group II or true proximal focal femoral deficiency where the hip is never normal and the knee always useless. This group always needs prosthetic management and all operations should be designed to facilitate the fitting of the prosthesis.

**Group I (congenital short femur).** The 26 patients in this group presented at birth as described. However, important physical signs were present that allowed differentiation from Group II patients.

Clinically, the involved leg is not as short as in true proximal focal femoral deficiency; the foot is at approximately midtibial level in relation to the uninvolved leg. It is in lateral rotation and abduction with the hip and knee flexed. The flexion deformities are not severe and will decrease over the first year of life.

There is a constant finding of anteroposterior laxity of the knee and a valgus deformity which can be hidden with the chubby leg in lateral rotation and flexion. The laxity of the knee joint becomes more evident as the normal infantile knee flexion contracture resolves.

The radiographic appearance of the proximal femur in infancy is bulbous ("stable" appearance described by Fixsen and Lloyd-Roberts in 1974) and there may be sclerosis at the midshaft level. The proximal end of the femur lies proximal and lateral to a normally developed
acetabulum. The cartilaginous head, neck and greater trochanter are present but are radiolucent. The head and neck are in varus and retroversion; this explains the proximal and lateral relationship of the proximal end of the shaft which is visible radiographically, simulating the appearance of a congenital dislocation of the hip (Fig. 3). However, there is no defect in the cartilage and therefore the term proximal focal femoral deficiency is inappropriate.

By the age of two years the proximal femur ossifies revealing a well-developed greater trochanter and a variable degree of coxa vara. The varus remains static throughout growth, unlike that in some forms of true proximal focal femoral deficiency where great progression of varus occurs through a fragile or defective cartilaginous bridge. The affected femur is 40 to 60 per cent of the length of the normal femur, with a lateral bow of the femoral shaft.

It is the contention of this paper that this type of case represents a totally different clinical problem from that in true proximal focal femoral deficiency as seen in Group II. These children do not have significant fixed flexion contractures of the hip and knee and by the age of two years they walk with the hip and knee in full or nearly full extension.

The most typical Group I patient then represents a clinical problem of leg length, the affected femur being approximately 40 to 60 per cent of the length of the normal side. The hip is stable, has a full range of flexion and extension but lacks medial rotation (Figs 4 and 5). The latter feature and a mild Trendelenburg gait are both caused by the varus and retroverted position of the femoral neck.

The knee joint shows a valgus deformity of 20 degrees or more with somewhat flattened femoral and tibial condyles (Fig. 6) and marked anteroposterior laxity. If the leg below the knee is normal then the overall discrepancy in leg length will be characteristically about 20 per cent or 15 to 20 centimetres at the end of growth. This clinical picture is markedly different from that in the true proximal focal femoral deficiency syndrome where the knee is non-functional and the discrepancy in leg length is approximately 40 to 50 per cent.

The most severe Group I patient may have a femur only 40 per cent the length of the contralateral normal femur but the essential diagnostic features of hip and knee remain. We consider that the Group I patients with relatively long femora are potential candidates for surgical equalisation of leg length. An extension prosthesis will be functionally and cosmetically quite adequate early in life (see Figs 24 and 25). However, with skeletal growth the distance between foot and floor and between knee axes increases so that the prosthetic fitting becomes more difficult, is cosmetically unappealing and is less functional with an extremely long and clumsy lower segment.
Previously we have treated many of the patients by Van Nes' tibial rotation-plasty combined with leg shortening and fusion of the knee. This allowed approximation of the ankle joint to the contralateral knee and with rotation of 180 degrees allowed the ankle to simulate knee function, with subsequent fitting of a modified below-knee prosthesis.

The advent of the Wagner technique of leg lengthening has now altered the surgical possibilities for those patients in whom there are predicted final leg-length discrepancies of 20 centimetres. Wagner (1971) has demonstrated the feasibility of equalising leg-length deficits of the order of 20 centimetres, and has thereby re-opened the issue of lengthening the congenital short femur after a period of surgical history in which it has been justifiably closed by the high incidence of complications. Wagner has also shown that delayed union can be avoided by early rigid internal fixation and bone grafting (Wagner 1971).

In our experience with this technique it has been possible to lengthen the femur up to between 15 and 20 per cent with reasonable safety, but beyond that posterior subluxation of the knee and dislocation of the hip have occurred—with subluxation of the knee especially likely in this group of patients because of the previously described anteroposterior instability. The goal of equality of leg lengths is attainable in Group I patients by means of multiple lengthenings and contralateral epiphysiodesis but is fraught with a high incidence of complications. A compromise solution used in two cases has been to lengthen the femur 20 per cent and combine this with a disarticulation at the ankle, allowing a below-knee prosthesis to be fitted. This is an option for patients with associated fibular hemimelia and deformity of the foot.

A secondary problem is the Trendelenburg gait with abductor weakness associated with the coxa vara deformity. Osteotomy of the proximal femur and transposition of the greater trochanter have improved both the gait and the radiographical appearance.

Group II (true proximal focal femoral deficiency). The cases included in this group are those where a true deficit exists in the proximal femur whether or not the femoral head and acetabulum are present, and also those cases in which a tenuous cartilaginous bridge exists between the proximal shaft and the femoral head and which is undergoing late ossification in a markedly varus position.

The clinical features of this group are as follows: first, the thigh segment is extremely short; secondly, the leg is held in abduction and lateral rotation; thirdly, the flexion contractures at both knee and hip are fixed and severe in degree and do not resolve as the child matures (Figs 7 and 8); and fourthly, the overall discrepancy in leg lengths is between 35 and 50 per cent with the foot at the level of the contralateral knee.

Radiographically, these cases demonstrate a great variety of deficiencies. Most lie in the Aitken B, C, or D groups at birth (Aitken 1959) and show the "unstable signs" defined by Fixsen and Lloyd-Roberts (1974). A
significant minority have a stable hip and develop a fully ossified "mini femur"; however, the development of the greater trochanter is abnormal and the knee is extremely hypoplastic (Table I).

Table I. Radiographic characteristics of the hips of Group I and Group II patients

<table>
<thead>
<tr>
<th>Group I</th>
<th>Group II</th>
</tr>
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<tbody>
<tr>
<td>Femur 40 to 60 per cent of normal length</td>
<td>Femur very short</td>
</tr>
<tr>
<td>No defect</td>
<td>Deficiency always noted</td>
</tr>
<tr>
<td>Coxa vara</td>
<td>Head and neck may be absent</td>
</tr>
<tr>
<td>Lateral bowing of shaft</td>
<td>Shaft may be deficient</td>
</tr>
<tr>
<td>Hypoplastic knee</td>
<td>Hypoplastic knee</td>
</tr>
</tbody>
</table>

The hypoplasia of the knee, not previously described in the literature, varies from the moderate condylar flattening with marked anteroposterior instability of the Group I patients to the congenital knee fusion of the severe Group II patients. Most of the Group II patients will have marked condylar flattening with absence of the cruciate ligament although the flexion contracture may disguise the anteroposterior instability.

The aim of the surgeon in the Group II patients should be to modify the leg to assist the prosthetist to fit the best possible functional and cosmetic prosthesis. The leg is always too short to consider lengthening in relation to the normal leg. The hip joint may appear inadequate radiographically but nonetheless is mobile and painless. The flexed knee is functionless and a significant problem to prosthetic fitting.

It has been our experience that these patients may show a Trendelenburg lurch, due to the lack of pelvic femoral stability and because the prosthetists have difficulty in getting a socket around the anteriorly protruding flexed knee to achieve true ischial weight-bearing. Radiographs showing the patient in the prosthesis demonstrate that true ischial bearing does not occur and that the small hip and knee are being forced into even more excessive flexion in the stance phase of gait.

The standard management of Group II patients in our clinic has been early fusion of the knee, at or around the age of two years, as recommended by King (1969) to create a solid straight tibiofemoral lever, with either an ankle disarticulation done at the same time or preservation of the foot to retain the option of a Van Nes rotation-plasty. In all these cases except one, fusion resulted in much earlier prosthetic fitting with subsequent improvement in the hip flexion contracture to a significant degree by the effect of the leverage on the long tibiofemoral segment. The exception was a case where knee fusion was carried out at the age of eight years in a boy who had severe flexion deformity through a subtrochanteric pseudarthrosis. This was corrected and fused surgically to reduce the flexion of the tibiofemoral segments.

The Van Nes tibial rotation-plasty which has formed an integral part of treatment of many of the patients with proximal focal femoral deficiency was initially described by Borggreve (1930) and later popularised by Van Nes (1950).

In Group II, 18 patients had operations around the hip either to repair a pseudarthrosis or to correct gross degrees of coxa vara in those patients (Aitken B or C) where a femoral head was present in the acetabulum. Regardless of many very satisfactory radiographic results, the results in terms of gait were less satisfactory. The reason for these disappointments seems to lie in a persistent Trendelenburg gait due to inadequacy of the abductor mechanism. The reason for inadequacy of the abductor mechanism is a short femoral neck, a posteriorly positioned or absent greater trochanter, tissue hypoplasia of the abductors, or a combination of these factors.

RESULTS

Group I (congenital short femur)

In the early years of our clinic the aim was to achieve a functioning Van Nes rotation-plasty; knee fusion was carried out late with appropriate excision of bone to bring the new "knee joint" (that is, the ankle joint of the short limb) approximately to the level of the contralateral knee. Before the advent of the Wagner apparatus the techniques of leg-lengthening had proved hazardous with a high incidence of complications and difficulty in consistently obtaining a desirable increase in bone length. Over the past seven to eight years the aim of treatment of these cases has been to correct the angulatory and rotational deformities of the affected limb and then attempt to equalise limb length by appropriate lengthening and contralateral epiphysiodesis (Table II).

Table II. Operations performed on the 26 Group I patients

<table>
<thead>
<tr>
<th>Operation</th>
<th>Number</th>
</tr>
</thead>
<tbody>
<tr>
<td>Valgus osteotomy proximal femur</td>
<td>6</td>
</tr>
<tr>
<td>Knee arthrodesis</td>
<td>5</td>
</tr>
<tr>
<td>Varus osteotomy distal femur</td>
<td>4</td>
</tr>
<tr>
<td>Distal femoral epiphysiodesis</td>
<td>5</td>
</tr>
<tr>
<td>Van Nes rotation-plasty</td>
<td>5</td>
</tr>
<tr>
<td>Syme's amputation</td>
<td>6</td>
</tr>
<tr>
<td>Femoral lengthening</td>
<td>8</td>
</tr>
</tbody>
</table>

Hip operation. The patients in Group I can be expected to have satisfactory hip function. Operative procedures for these patients aim to correct the coxa vara and
external rotation deformities. This also results in a gain in limb length of up to two centimetres and improvement in gait and abduction strength.

Six patients have undergone successful valgus (abduction) osteotomy and rotation osteotomy of the femur in the subtrochanteric region.

Knee operation. As described, the initial plan of management in Group I patients was to perform a knee arthrodesis following Van Nes rotation-plasty and shorten the limb appropriately at the same operation. Five patients underwent this successfully. In all cases a solid one-piece tibiofemoral thigh segment was obtained which allowed fitting of a modified below-knee prosthesis. There were no significant complications; in particular the problem of flexion at the fusion site did not occur with excision of both epiphyses.

Our current plan of management is to correct the consistent valgus deformity of the knee of the affected limb (Figs 9 and 10). This deformity can be easily overlooked as the affected limb presents in lateral rotation thus allowing the valgus knee to be disguised as slight flexion.

Four patients have had an opening wedge osteotomy of the distal femur using a segment of fibula as a graft. No complications have been seen in these patients and correction of the alignment of the lower limb has been good.

Van Nes rotation-plasty. Although the tibial rotation-plasty is no longer performed routinely in Group I patients, five patients did undergo this procedure (subject of a previous report; Kostuik et al. 1975). Three patients continue to function well. However, the other patients, all girls, underwent revision of the rotation-plasty and eventually came to Syme's amputation; two of the girls, who had had knee fusions, used above-knee prostheses and the two who had not had knee fusions were converted to satisfactory below-knee stumps by femoral lengthening and they function well with below-knee prostheses.

The reasons for revision with Syme's amputations were the combination of derotation and poor function together with the realisation that the original procedure would result in less satisfactory function than could be obtained by retention of the knee joint, femoral lengthening and use of a standard below-knee prosthesis.

Ankle disarticulation. There were six Syme's amputations performed in the Group I patients. Four of these were performed in girls who had previously had a tibial rotation-plasty. The two other patients had associated fibular hemimelia.

Femoral lengthening. With the advent of the Wagner technique the emphasis on the management of Group I patients has shifted from shortening legs to equalisation of leg lengths.

Appropriate leg-length projections are made using the knowledge that the femoral proportions will remain constant through to the completion of growth if they are not altered surgically. If hope of leg equalisation exists then femoral lengthenings and contralateral epiphyseodeses are planned for the appropriate ages.

Eight patients from Group I have undergone femoral lengthening to date. Two of these patients were converted after Van Nes rotation-plasty to below-knee stumps by lengthening the femur to align the knee and amputation of the rotated foot. One patient with a left congenital short femur and right proximal focal femoral deficiency had a left femoral lengthening to bring the left knee axis to the right ankle after having had a successful Van Nes rotation-plasty performed in the limb with proximal focal femoral deficiency.

In Group I patients with hypoplasia of the knee and laxity in an anteroposterior direction (Figs 11 and 12), there is a higher risk of posterior subluxation of the tibia on the femur than in the normal knee. Posterior dislocation was noted in five out of eight knees during femoral lengthening. Two of these patients required surgical release of the knee and others were managed with traction and casts.

Posterior subluxation appeared to be precipitated by a femoral lengthening beyond 20 per cent (Figs 13 and 14). One patient also had a dislocated hip treated by closed reduction.

Other complications included fracture of the lengthened femur in two patients and a transient foot drop in one patient.
Group I patient. Radiographs showing instability of the knee in the sagittal plane in a patient aged 16 years.

Group I patient. Figure 13—Anteroposterior radiograph of femur during lengthening. Figure 14—Lateral radiograph showing the posterior dislocation of the tibia on the femur.
**Group II (true proximal focal femoral deficiency)**

This group of 43 patients contained 10 patients with varying degrees of bilateral involvement. Of the bilateral cases two patients have been fitted with bilateral extension prostheses which they selectively use to achieve a more cosmetic standing height; in other respects they function well with their natural limbs. Of the other eight patients with bilateral involvement, six had a combination of proximal focal femoral deficiency and congenital short femur, and two had combinations of proximal focal femoral deficiency and amelia. Treatment in such patients must be tailored individually to obtain maximal function and to facilitate prosthetic fitting.

The 33 unilateral cases have all been fitted prosthetically following the surgical procedures shown in (Table III).

**Table III. Operations performed on the 43 Group II patients (10 of these patients had bilateral deformities)**

<table>
<thead>
<tr>
<th>Operation</th>
<th>Number</th>
</tr>
</thead>
<tbody>
<tr>
<td>Knee arthrodesis</td>
<td>23</td>
</tr>
<tr>
<td>Valgus osteotomy</td>
<td>11</td>
</tr>
<tr>
<td>Repair of pseudarthrosis</td>
<td>7</td>
</tr>
<tr>
<td>Syme’s amputation</td>
<td>8</td>
</tr>
<tr>
<td>Van Nes rotation-plasty</td>
<td>21</td>
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</tbody>
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**Knee arthrodesis.** Fusion of the knee of the leg with proximal focal femoral deficiency has several beneficial effects. First, it corrects the knee flexion contracture which provides a single lever arm between the pelvis and ankle joint, leading to resolution of the flexion contracture of the hip. Secondly, it stabilises this segment of the limb allowing easier prosthetic management with an above-knee fitting or the Van Nes prosthesis.

The patients who do not benefit from knee arthrodesis are those with a congenital knee fusion or with a femoral segment so short and a knee joint so close to the pelvis that instability and knee flexion contractures are not great problems.

There have been 23 fusions of the knee performed in Group II patients. Most have been performed at an early age, under three years, as part of an elective management decision although several have been performed at a later age with good results. The patients operated on early have had either the distal femoral or proximal tibial epiphyses preserved and a central intramedullary Rush rod inserted for internal fixation (Fig. 15). There have been no non-unions and in all patients marked improvement in vertical alignment of the limbs has resulted with elimination of flexion contractures of the knee and hip.

The only complications recognised have been two temporary partial sciatic nerve lesions due to sudden extension of the knee, and one infection which required early removal of the Rush nail (after 10 weeks). The infection subsided without further problem.

Two patients suffered minor falls resulting in Type I epiphyseal injuries to the proximal tibial epiphyses which were still open; these injuries healed without problem (Fig. 16). When the knee fusion is done late the epiphyses can be sacrificed as desired to obtain an appropriate leg length for prosthetic fitting and function. Only one of the early knee fusions showed a tendency to develop a significant flexion deformity through the fusion area as observed by King (1969), and in all cases, parents and prosthetists remain pleased with the greatly increased ease of prosthetic fitting and improved cosmesis.

**Hip operation.** In describing the hip operations of these patients it should be appreciated that there are three anatomical subgroups of patients.

First, patients who lack a femoral head and acetabulum and thus in whom no actual hip joint exists. We have not found exploration of the hip of benefit and have not performed any femoropelvic fusions, preferring to depend on soft tissues to provide stability and a painless joint.

Secondly, there is a group of patients in the Aitken B type who have a femoral head visible in the acetabulum and a “defect” shown radiographically between the head and the proximal end of the shaft. With maturity this cartilaginous defect may eventually ossify leaving a gross degree of coxa vara (“shepherd’s crook” deformity) or a true gap may persist with a well-developed and highly mobile pseudarthrosis which can be confirmed by push-pull radiographs (Figs 17 to 19).

Eleven patients of this type underwent valgus osteotomy with or without repair of the pseudarthrosis. In spite of satisfactory radiographic results these patients continued to have abductor weakness with a marked Trendelenburg gait. We feel these poor functional results can be explained by the deficiency of the abductor musculature as noted on scans using computerised tomography and that they can be inferred by the absence, hypoplasia or posterior position of the greater trochanter. We have one case that illustrates this point extremely well. This patient had a classical proximal focal femoral deficiency of the Aitken A variety and went on without operation to develop a completely ossified “mini-femur”. She had a congenital fusion of the knee without significant varus, and a stable hip joint radiographically (Fig. 20). In spite of her intact proximal femur she walks with a marked Trendelenburg gait. Scans of her hip using computerised tomography revealed a hypoplastic greater trochanter lying posteriorly (Fig. 21).

Thirdly, a small subgroup of patients have a well-formed greater trochanter and head and neck but a subtrochanteric pseudarthrosis which creates a biplane deformity in both varus and flexion. We feel that this group represents the least involved form of true proximal focal femoral deficiency and represents the best indication for a corrective valgus and extension osteotomy.
Group II patient. Figure 15—Radiograph of a patient with a solid knee fusion using an intramedullary rod. Note the open epiphysial plates. Figure 16—Radiograph of a patient with a solid knee fusion and epiphysial injury.

Group II patient. Figures 17 and 18—Push-pull radiographs of patient showing movement through the area of femoral deficiency. Figure 19—Radiograph after operation showing solid union of the pseudarthrosis and fusion of the knee.
which offers the opportunity to convert such cases into stable hips similar to those in Group I.

**Ankle disarticulation (Syme's amputation).** In Group II patients Syme's amputation of the foot greatly facilitates cosmesis and prosthetic fitting where a decision has been made not to perform a Van Nes rotation-plasty. This applies to those patients with associated fibular hemimelia, found in six of this subgroup of patients, two of whom had previously had a Van Nes procedure.

To date, eight Group II patients have undergone Syme's amputation without complication.

**Van Nes rotation-plasty.** Twenty-one patients from Group II were treated by tibial rotation-plasty. This allowed fitting of the limb with a Van Nes prosthesis using the patient's foot to power the below-knee portion of the prosthesis. The rotation-plasty was performed through the shaft of the tibia using a compression plate fixation in most cases in the older children and an intramedullary pin in the younger children.

In two cases full rotation was not attempted as the maximal range of movement for flexion-extension of the foot on the tibia was through combined movement of the ankle and subtalar joints (Figs 22 and 23). In all other cases full rotation of the ankle and foot through 180 degrees was attempted to allow the ankle joint to function as a knee with the foot in a reversed position.

Ten patients required further operation to correct
significant derotation including two patients who underwent Syme’s ankle disarticulation because of poor function.

Previously with the midtibial rotation-plasty the timing of the procedure was important. Rotation at an early age provided a mobile ankle joint but a longer time for the spiralled muscles to derotate the open epiphysial plates. Rotation later ensured a stable limb but the ankle was less mobile from wearing an extension prosthesis for the earlier years. We feel that if an adequate amount of bone is excised at the time of rotation-plasty the incidence of derotation is greatly reduced, whereas previously only enough bone was removed to allow rotation of the foot and maintain its blood supply.

The rate of revision indicates the significance of the problem of derotation; however, most patients in this group have maintained a satisfactory range of movement with good power by developing a ball-and-socket ankle joint and increased rotatory movement through the subtalar joint.

In spite of the problems described, the Van Nes rotation-plasty retains a functional joint and a potentially useful segment of the limb that would otherwise be sacrificed or functionless and a major problem in prosthetic fitting. The function obtained after a rotation-plasty is markedly superior to that of an above-knee type of fitting following knee fusion and ankle disarticulation.

Our current practice is to perform the knee fusion and rotation-plasty in one procedure. This technique involves rotation of the tibia through the knee fusion to the extent allowed by soft tissues (usually 140 degrees or over) with excision of the distal femoral epiphysis and final rotation through a diaphysial tibial osteotomy. Fixation is with an intramedullary Rush rod.

**Prosthetics**

A discussion on the type, fabrication and modification of prostheses needed in these children is beyond the scope of this paper. The reader is referred to Bochmann (1980) for more complete information. Nonetheless the authors wish to emphasise the fact that the patient with true unilateral proximal focal femoral deficiency will always require a prosthesis for walking and furthermore any operation must be embarked upon with the intent of improving function of the patient wearing a prosthesis.

The devices used are shoe lifts, below-knee extension prostheses and knee-disarticulation prostheses. Shoe lifts are for the young child with modest leg-length discrepancy. Below-knee extension prostheses are used for Group I patients and Group II patients before definitive surgery (Fig. 24). It should be noted that long-term use of this prosthesis results in loss of movement at the ankle joint and development of an equinus contracture. Knee disarticulation prostheses are used for Group II patients after Syme’s amputation and knee fusion. Van Nes prostheses are used for Group II patients after knee fusion and tibial rotation-plasty. The device resembles a below-knee prosthesis in design and function (Fig. 25).

![Fig. 24 Group I patient in an extension prosthesis. Figure 25 Group II patient in a prosthesis.](image)

**DISCUSSION**

A baby with a short leg can be differentiated into one of two groups by the clinical signs described in the text. Radiographically, Group I infants show the appearance described by Fixsen and Lloyd-Roberts (1974) as a round and bulbous proximal end and a normal looking acetabulum, indicating cartilaginous continuity of the proximal femur. Radiographically, Group II infants include those babies showing the “unstable” signs of Fixsen and Lloyd-Roberts (1974) and which are included in Types B, C, and D of Aitken (1959).

We recognise that this syndrome covers a spectrum of femoral deficiency and that some cases may be difficult to categorise because they contain features of both groups. We had two patients who had moderate hip and knee flexion contractures, severe coxa vara and femora approximately 40 per cent of the length of the normal side. These were converted to fairly typical Group I patients by abduction osteotomy. We also recognise a group with short femora and flexion contractures but with a subtrochanteric pseudarthrosis, thus being in Group II. These patients may have a well-developed greater trochanter and by appropriate operation can be converted functionally from Group II to Group I and, in fact, represent the best subgroup of patients in Group II for operations around the hip.

In Group II the most useful surgical intervention is early fusion of the knee as described by King (1969);
however, the literature overstates the value of repeated surgical attempts to improve the anatomical status of the hip. In the congenital short femur we believe that a place for early osteotomy exists to correct coxa vara and that Wagner lengthening should be considered for femora that are 60 per cent or more of the normal length.

In conclusion patients with a clinical appearance of gross femoral deficiency can be differentiated on clinical grounds into two groups: a milder form, the congenital short femur, where in most cases the goal should be limb length equalisation; and a severe form, the true proximal focal femoral deficiency, in which a prosthesis is always necessary and function can be optimised by early knee fusion and tibial rotation-plasty.

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REFERENCES