

The classification and treatment of proximal femoral deficiencies

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Abstract

After a review of previous classifications the authors suggest that cases of congenital longitudinal deficiency of the femur fall into one of two groups, each with specific features. The treatment alternatives and their indications are discussed in detail and a treatment plan suggested.

Introduction

The following paper attempts to present a balanced viewpoint of the management of congenital abnormalities of the femur based on the authors' experience and a review of the literature relevant to the topic as published in major orthopaedic publications.

The paper examines this topic under the headings of Classification, Clinical Problems, Surgical Procedures and Conclusions.

Classification

Standard radiographic techniques have been the basis of most of the classifications published to date, although the use of ultra-sound has recently been examined. One would expect that in the future the patients under discussion will be subjected to Magnetic Resonance (MR) scanning and no doubt further classifications will then arise.

At the present time however, the best known classifications in the English literature are those of Aitken (1968) and Amstutz and Wilson (1962). The original classification of Aitken contained four classes with the least severe being Class A and the most severe being Class D. The classification of Amstutz further subdivided these groups with Aitken's Class A

being subdivided into Amstutz and Wilson Type I and II. However, from the original Amstutz and Wilson classification, further subtypes have arisen.

Hamanishi (1980) defined five types of deficiency (Fig. 1) and further subtyping resulted in the identification of ten groups of patients. More recently publications of Hillman *et al.* (1987) and Stihle *et al.* (1987) have provided further radiographic evaluation of patients with femoral abnormalities.

However, it is apparent in these papers that the radiographic appearance of children born with a congenital femoral abnormality is a continuum from the most minor variance from normal to the most severe deficiency of complete absence of the femur. Taken to its logical conclusion, the number of subtypes one can derive would simply depend on the number of patients that have been examined.

Some questions arise from the radiological classifications. Firstly, at what age should the classification be performed? Secondly, will this classification change with time? Thirdly, and perhaps most importantly, what relevance clinically are these numerous classifications to the patient and their ultimate limb function?

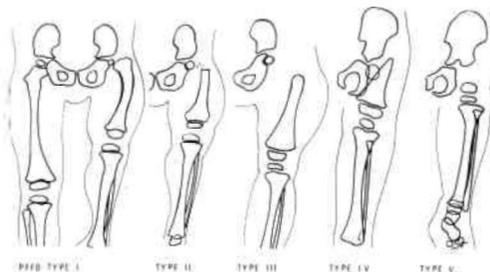
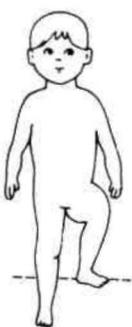


Fig. 1. Morphology, natural history and treatment of PFFD. Amstutz.

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Table 1. Clinical signs of Group I patients

	GROUP I
	CLINICAL SIGNS
	LEG LENGTH DISCREPANCY 20-30%
	VALGUS KNEE
	A-P LAXITY KNEE
	FLEXION DEFORMITIES NOT FIXED
FOOT MID TIBIAL LEVEL	

Of relevance to the first question is the need to provide the parents of an infant born with a congenital abnormality with information that will be the foundation for their relationship with the doctor over the years to come. Indecision and/or incorrect information may well provide a permanent stumbling block in the doctor/patient relationship in the future and may well heighten the anxiety level of the parents who perceive this disability as a disaster for both themselves and their child.

In addressing the second question as to the change of classification with time, some information is gained by examining the expansion of the Amstutz classification with the passage of time. Hillman (1987), showed that with retrospective evaluation, only 17 out of 43 patients were correctly classified. This serves to illustrate that of itself radiographic evaluation may not be the foundation stone for the management of these patients.

The publication of Gillespie and Torode (1983) demonstrates that on clinical grounds patients with congenital abnormalities of the femur can be divided into two groups. The first

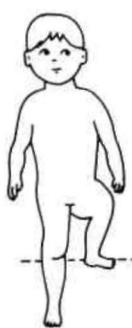
of these groups contains those patients designated as having a congenitally short femur. Radiographically these groups could be classified as Aitken Class A or Amstutz Type I and possibly Type II. The clinical features of the Group I classification are shown in Table 1.

This group contrasts with the Group II patients who have more severe femoral deficiency and different clinical signs. Most of these patients would be classified as Aitken B, C or D groups at birth and show the unstable signs defined by Fixsen and Lloyd-Roberts (1974). The clinical signs of this group are summarised in Table 2.

This clinical classification becomes relevant when attempting to answer the third question posed. For those patients with congenital short femur, classified as Group I, there will be a possibility of performing surgical procedures to equalise limb lengths with the ultimate aim of avoiding the need for prosthetic fitting. This aim contrasts with the goals for those patients in Group II, true proximal femoral focal deficiency, for whom prosthetic fitting will be an inevitable part of the management. Exceptions to this are bilateral cases and where bilateral prosthetic fitting is to elevate the height of the wearer. In this situation the prostheses are often rejected.

Once the examining physician has ascertained into which of the broad clinical groups his patient falls it is necessary to examine the anatomical regions of the whole limb. The radiographic appearances of this femur and the presence of associated anomalies in the rest of the limb will dictate the variation in advice given. The radiographic appearances of the patients of Group I and II are summarised in Table 3.

Table 2. Clinical signs of Group II patients

	GROUP II - PFFD
	CLINICAL SIGNS
	THIGH VERY SHORT
	LEG LENGTH DISCREPANCY 35-50 %
	FLEXED HIP AND KNEE (FIXED)
	ANKLE AT KNEE LEVEL

The limb

The foot and leg

The incidence of anomalies of the foot and leg said to be associated with congenital

Table 3. Radiological signs

GROUP I CONGENITAL SHORT FEMUR	GROUP II PROXIMAL FEMORAL FOCAL DEFICIENCY
FEMUR 40-60 % NORMAL	FEMUR VERY SHORT
NO DEFECT	DEFICIENCY ALWAYS NOTED
COXA VARA	HEAD AND NECK MAY BE ABSENT
LATERAL BOWING SHAFT	SHAFT MAY BE DEFICIENT
HYPOPLASTIC KNEE	HYPOPLASTIC KNEE



Fig. 2. Congenital short femur with associated fibular deficiency.

abnormalities of the femur varies from one report to another, but a reasonable assumption is that some abnormality will occur in approximately 50% of cases. The most common will be a variable degree of fibula deficiency (Fig. 2). This may range from a slightly short fibula in association with a valgus ankle, to the extreme case of a deficient foot with absent rays and a grossly unstable and displaced ankle joint. At either end of this spectrum, the desirability of keeping this foot will be obvious, but in the mid-range of deformities, other factors will be considered in deciding whether or not the foot is to be ablated.

Knee

Clinically the knee in these children will be held in flexion, but the flexion deformity of the Group I patients will become less with time, whereas the flexion deformity of the Group II patients will usually be permanent. This flexion deformity will tend to disguise the valgus attitude of the knee, often associated with a hypo-plastic lateral femoral condyle (Fig. 3). The knee in these children will also be unstable in an antero-posterior direction (Gillespie and Torode, 1983) due to the hypoplasia of the cruciate mechanism which has been documented by numerous authors, (Torode and Gillespie, 1983¹; Johansson and Aparisi, 1983). Instability of the knee will become

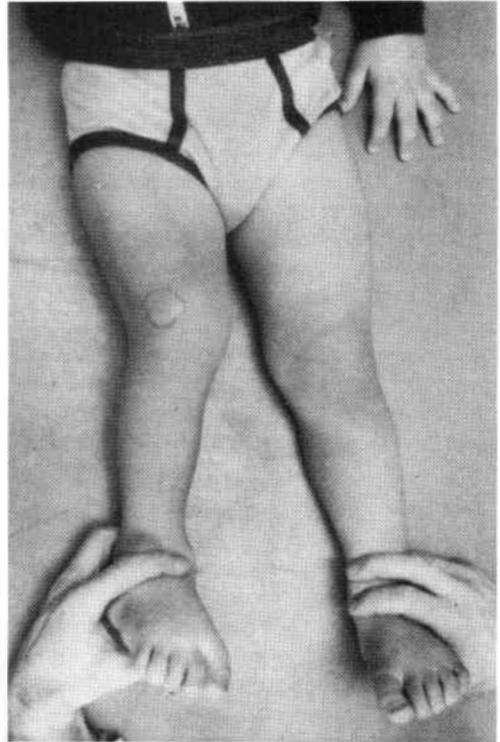


Fig. 3. Patient with right congenital short femur and valgus knee and external rotation deformity of the femur.

important in discussions surrounding lengthening procedures of the limb.

Thigh and hip

The varying degrees of deficiency of the proximal femur will be noted in simple radiographs. This may range from the case with a slightly short femoral neck, through those with a varus femoral neck, or an actual deficiency of the proximal femur distal to an intact femoral head and acetabulum, to a situation of gross deficiency of the proximal femur, femoral head and acetabulum (Fig. 4).

The radiographic appearance may vary with time and as further ossification occurs it will become apparent that the radiographic deficiency may have been a cartilagenous anlage which will go onto full ossification, albeit often in varus, and occasionally with an extreme shepherd's crook deformity. On other occasions this cartilagenous anlage may be absent or fail to ossify resulting in a definite pseudarthrosis in the proximal femur.

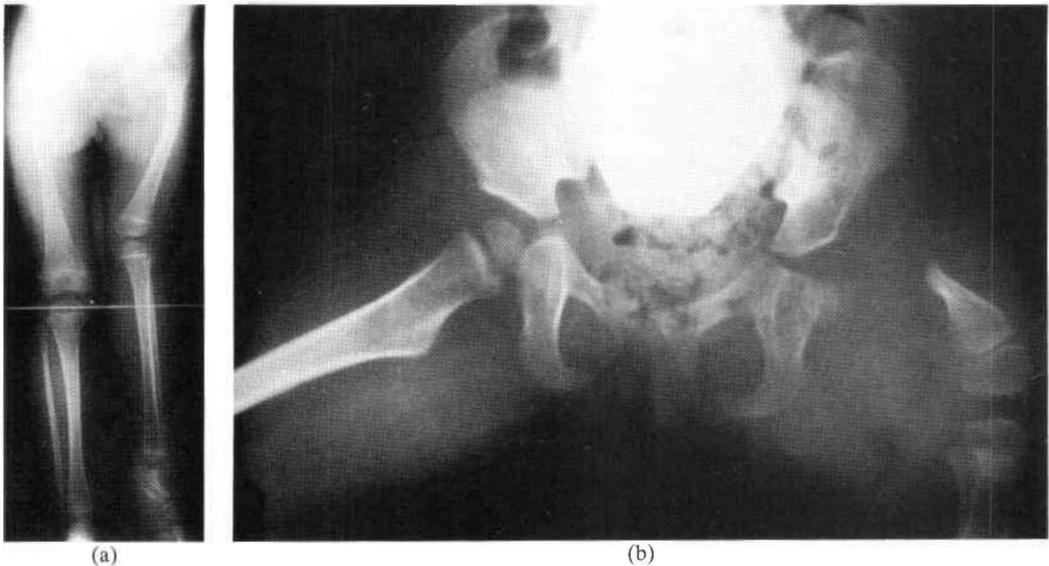


Fig. 4 a) Left congenital short femur (Group I patient).
b) Severe right femoral deficiency (Group II patient).

Computer Tomography (CT) scans and X-ray examinations of the proximal femur also indicate a deficiency of the soft tissues surrounding the hip, and in particular a deficiency of the abductor musculature. One would expect that an MR scan of the hip would help to further delineate the soft tissue deficiencies.

The overall picture of the hip joint will play an important part in deciding whether bony reconstruction of the proximal femur is advisable and further knowledge of the soft tissue deficiency will assist the surgeon in understanding the limitations of the bony reconstructions as pertain to the final function of the hip joint.

Changes of the limb with time and growth

The authors believe it is possible on the basis of clinical signs to ascertain in most cases into which of the two clinical groups the patient should be placed, i.e. either a congenital short femur, or true proximal femoral focal deficiency. Despite the variations with age however, the radiographic appearance of the Group I femur in infancy will show the bulbous stable appearance as described by Fixsen and Lloyd-Roberts (1974), and with time further ossification of the proximal femur will proceed with varying degrees of coxa vara. The varus

deformity however remains static throughout growth unlike true congenital coxa vara.

In Group II patients the actual deficiency of the bony elements will become apparent as ossification takes place. Nonetheless there will be a constant feature of a fixed flexion deformity of the proximal femur, although there may be an extension deformity through a pseudarthrosis in the subtrochanteric region in the femur (Fig. 5).

An important component of the parents' understanding of this clinical problem is the knowledge that the overall limb length deficiency will be relatively constant throughout growth. This predictability of



Fig. 5. Deficient femur with pseudarthrosis in extension in subtrochanteric region of the femur (Group II patient).

growth pattern has been well documented by Hamanishi (1980), Hillman (1987), Gillespie and Torode (1983). It is therefore usually possible to give the families an estimate of the expected limb length discrepancy that the child will have as he or she becomes more mature.

Using this knowledge the families can be advised at an early age of the various prosthetic appliances that can be utilised to assist the developing child in ambulation. However, as regards surgical procedures, particularly ablative procedures, the authors believe that these should be carried out earlier rather than later in the child's life. Clearly some surgical procedures are time dependent and the parents need to be made aware of the factors that the physician is considering in his decision making process for each particular child.

Surgical procedures

It is possible to look at the surgical procedures required in relation to the needs of each of the Groups I and II patients. There will, occasionally, be children who despite a deficiency of the proximal femur may be treated as Group I patients because of their particular anatomy, and there are others who have an intact femur, i.e. Group I, but whose femoral length deficiency is severe and beyond the limit of lengthening procedures. These therefore will be managed as Group II patients. It should be recognised that these are the exceptions rather than the rule and most can be managed utilising the procedures described below.

Surgical procedures appropriate to Group I patients

Syme's amputation

In the presence of an associated foot deficiency, with or without a fibular deficiency, it is usually readily apparent which children should undergo Syme's amputation and in which the foot should be retained. The greater the degree of the associated fibula deficiency and/or tibial deficiency the greater the likelihood of need for foot ablation.

The benefit of foot ablation is that there is a greater tolerance in the limb length equalisation procedures as a prosthesis can make up some of the deficiency in length and any small difference in the height of the knee axes above the ground is of no great consequence. This

procedure should be carried out at about the time the child is attempting to stand and walk so that using a prosthesis becomes second nature early in life.

Proximal femoral osteotomy

Osteotomy of the proximal femur is a most useful procedure in those children who have an associated coxa vara. This procedure adds length to the limb by increasing the neck shaft angle and may also stimulate growth of the femur. Care must be taken not to over-do the valgus correction of the proximal femur, particularly if there is any degree of acetabular dysplasia. This procedure can be carried out in the first few years of life as a "once off" procedure utilising simple fixation techniques.

Distal femoral osteotomy

In children in whom there is significant valgus deformity of the knee, the authors recommend that this be corrected by an open wedge osteotomy, which can be held with cross K-wires and the ipsilateral fibula can be used as a bone graft to stabilise the opening osteotomy. This procedure will also slightly increase the length of the femur both by the opening osteotomy and the realignment of the limb and also by growth stimulation of the distal femur (Fig. 6).



Fig. 6. Radiograph of Group I patient following opening osteotomy of distal femur and fibular graft.

Whilst it is recognised that the valgus attitude of the distal femur may be corrected at the time of femoral lengthening, the authors recommend for two reasons realignment of the limb as a whole prior to performing a femoral lengthening procedure. Firstly, the advantages of the osteotomy can be obtained at an earlier age than would be appropriate for a lengthening procedure. Secondly, it is much simpler to lengthen a straight segment than a deformed bone.

Innominate osteotomy

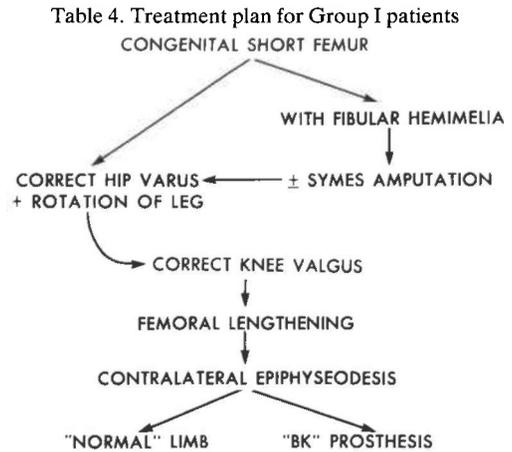
Careful assessment of the acetabulum must be made before performing any surgery on the proximal femur. Any residual dysplasia of the acetabulum can be corrected by a Salter innominate osteotomy, which must be performed before any lengthening procedures on the limb. Some additional gains in length can be obtained by using the Millis and Hall (1979) modification of the Salter innominate osteotomy.

Limb length equalisation procedures

Utilising the knowledge that the proportions of the limb will remain constant throughout growth, one can estimate the expected discrepancy at maturity. The growth of congenitally deficient limbs can be monitored and graphed through the early years of childhood. By around 8 years of age, it will be evident as to whether the discrepancy can be made good by a single lengthening procedure with a contralateral epiphysiodesis, or whether it may be necessary to do two lengthening procedures.

Previously the authors have advocated that femoral lengthening be restricted to an increase of 20% of the original femoral length because of the risk of producing either dislocation or subluxation of the hip and flexion deformities or posterior subluxation of the unstable knee. With better understanding of the biology of bone as applied to bone lengthening and with the use of small wire external fixators, such as the Ilizarov or Monticelli Spinelli frames, the horizon has expanded and the complications reduced. Nonetheless it remains a procedure fraught with difficulties for the patient, parents and surgeon.

Epiphysiodesis may be used judiciously to reduce the femoral length and/or tibial length of



the normal limb. The last few centimetres of femoral lengthening are often difficult to obtain and by reducing the ultimate length of the longer limb it is possible to lessen the complications and heartache. Modifications of the Phemister technique often leave ugly scars around the knee of the normal limb and this can be significantly improved upon by the use of the drill technique as described by Canale (1986). A flow diagram of the suggested plan of management for the Group I patients is shown in Table 4.

Surgical procedures in relation to Group II patients (PFFD)

This group of patients will always need prostheses if they are to achieve optimum function. The exception to this rule may be for these with bilateral deficiencies whose prostheses, used to elevate the patient to a more normal height, often prove unwieldy and are frequently rejected.

Syme's amputation

As discussed previously when the foot is deficient with respect to its lateral rays or tarsal bones, or if the ankle joint is grossly dysplastic in association with a severe degree of leg deficiency a Syme's amputation should be performed in the first few years of life (Fig. 7).

Where the foot is reasonably intact and the ankle joint functional, the families should be made aware of the functional benefits of the tibial rotationplasty and the part that procedure might play in the future management of the child. In such a situation the decision regarding a Syme's amputation must be delayed.

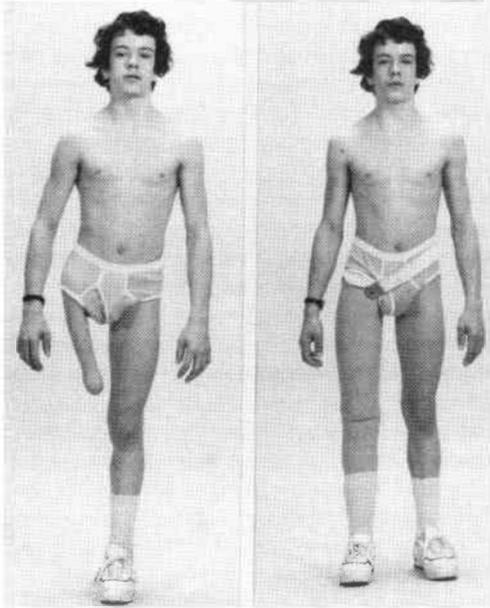


Fig. 7. Group II patient — following conversion to AK fitting by knee fusion and foot disarticulation.

Knee fusion

In cases where the families elect to have a Syme's amputation performed and the patient treated prosthetically as an above-knee amputee, the ablation of the knee joint adds considerably to the ease of prosthetic fitting and of prosthetic wearing. Figure 8 illustrates the difficulties that patients face when wearing an extension or above-knee prosthesis associated with a mobile hip and knee and with flexion deformities of both those joints. It is possible to perform a knee fusion early and preserve either the distal femoral or the proximal tibial growth plate, thereby ensuring further growth of the limb and the maintenance of a long stump. This procedure is performed by excising the appropriate segment of the knee joint and if it is intended to preserve the growth plate, the corresponding cartilaginous portion of the epiphysis is removed down to the cancellous bone of the ossific nucleus. Fixation is obtained by a smooth intramedullary rod passing along the length of the femur into the tibia. Compression devices have not been needed and fusion has been readily attained. This procedure can be performed in association with the Syme's amputation without undue difficulty.

Sometimes the knee fusion has been

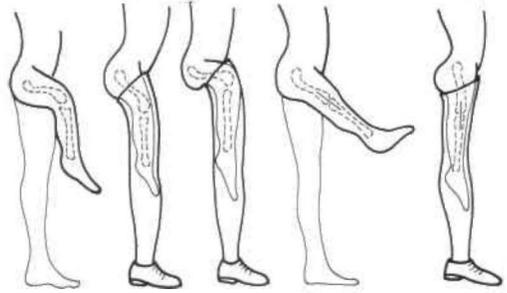


Fig. 8. Diagrammatic presentation of problem that is created by a flexed knee with prosthetic fitting of a short thigh segment.

performed in association with a repair of proximal femoral deficiencies using the intermedullary rod of the knee fusion to provide fixation across the proximal femur. Knee fusion can also be performed as part of the Van Nes rotationplasty (Fig. 9). Patients in whom knee fusion is not to be performed are those for whom a femoro-iliac arthrodesis may be appropriate.

Proximal femoral surgery

The enormous variation in radiographic appearance of the femora of Group II patients suggest an equally vast variety of surgical procedures that might be devised. In reality,



Fig. 9. Radiograph of Group II patient following rotationplasty. Note open proximal tibial growth plate.

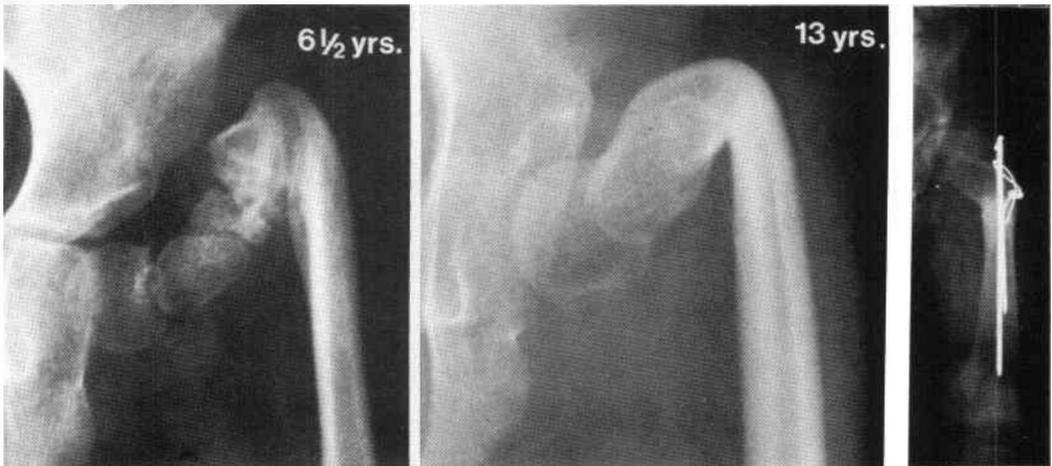


Fig. 10. Radiographs of Group II patient showing "shepherd's crook" deformity corrected by reconstructive osteotomy.

however, there are relatively few which are useful for the proximal femur of these patients.

The authors have not found exploration of the hip joint itself of great value. Exploration to confirm the appearance of the cartilaginous anlage will do little to further the function of these patients.

However, if there is instability or deformity due to a subtrochanteric pseudarthrosis of the femur, instrumentation and bone grafting to provide stability and a solid segment will be beneficial to the patient. Correction and internal fixation of gross varus or shepherd's crook deformity of the proximal femur may also improve the structural integrity of the bone and prevent recurrent stress fractures in the proximal segment of the femur (Fig. 10). A Trendelenburg gait due to the soft tissue deficiency, particularly of the gluteal and abductor musculature will often remain despite correcting the anatomical alignment of the proximal femur.

Tibial rotationplasty

The operative details of this procedure are described by Torode and Gillespie (1983²). However, it is worthwhile considering a few specific points. It is possible to perform the rotationplasty utilising the knee joint to attain either all or most of the rotation necessary. It is also possible to perform this operation at an early age and maintain one or both of the growth plates adjacent to the knee joint to ensure an appropriate length of the new "thigh

segment" (Fig. 9). By rotating through the knee joint rather than through the tibia, the function of the anterior and posterior compartments of the leg are preserved and the tendency to de-rotate is markedly diminished.

The post-operative course is simplified and prosthetic fitting is more readily accommodated if the rotationplasty is performed early in life. There is no doubt that the end result of this surgery produces an unusual appearance, but there is little to differentiate these patients from the rest of the population when the prosthesis is being worn (Fig. 11). Furthermore the

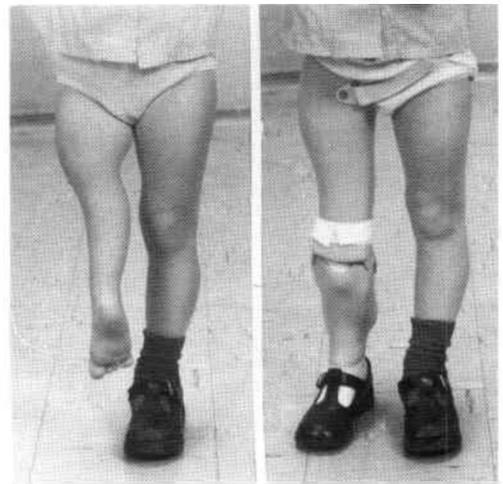


Fig. 11. Group II (PFFD) female patient following rotationplasty and knee fusion. Note long thigh segment to allow for growth discrepancy between legs.

functional advantages are quite significant and must be given due regard in the decision making before surgery is performed.

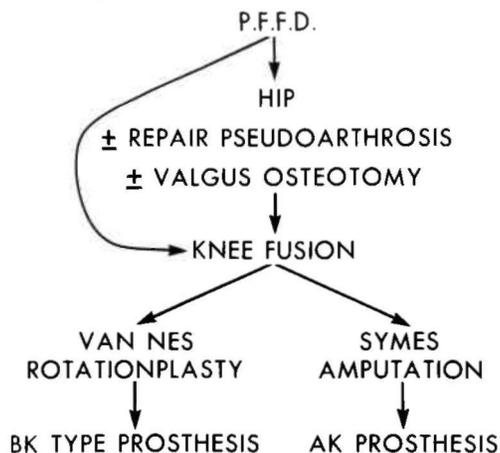
Fixsen (1983) presents the advantages and disadvantages of this procedure. Many of the problems he mentions have been addressed by the modifications of the technique as described by Gillespie and Torode (1983). Fixsen also raises the questions of psychological problems associated with the child having a foot at the end of the leg in a reversed position. However, it is difficult to differentiate between the embarrassment and psychological sequelae of the foot position after rotationplasty and the same sequelae of simply having a foot, often deformed, at the level of the contralateral knee, or if Syme's procedure has been performed, having no foot at all. The authors have interviewed all the children and young adults described in the rotationplasty group of their paper (Gillespie and Torode, 1983) and despite being offered foot removal, no patient has asked for this to be done to alleviate cosmetic or psychological concerns.

Iliofemoral fusion

Some of the patients with a true PFFD, exhibit a grossly deficient acetabulum and an absent femoral head resulting in a major problem of instability at the hip. Some may develop a spindle shaped end to the proximal aspect of the femoral segment which can ride upwards and be palpable just under the skin adjacent to the wing of the ilium. It may be appropriate to perform an iliofemoral fusion on such patients.

There are two important technical points in this procedure. Firstly, the femoral segment must be flexed approximately 90 degrees to its normal anatomical position, so that when the femur is arthrodesed to the ilium, the knee joint in extension will allow sitting and in flexion will allow standing. Secondly, the femoral segment must be reasonably short. If the femoral segment is too long when arthrodesed to the ilium, the weight bearing line of the limb will be at a considerable distance anterior to the coronal plane of the trunk and contralateral limb. However, many patients for whom this procedure appeared attractive have a knee flexion contracture and thus will not be able to extend the joint to allow a sitting position.

Table 5. Treatment plan for Group II patients



A more complete discussion on this procedure is given by Steel *et al.* (1987).

A summary of the surgical procedures described previously is shown in the treatment plan in Table 5. Despite the great variation in radiographic appearance, it should be noted that the number of useful surgical options is relatively limited and a decision needs to be made early in life to aim for below-knee function by use of a tibial rotationplasty or above-knee function by foot ablation and knee arthrodesis.

Summary

This paper has presented an overview of the management of congenital deficiencies of the femur. The two broad treatment patterns have been outlined, i.e. for children with a moderately severe deficiency, the congenital short femur group, and secondly children with a very severe deficiency with either a miniature intact femur, or a true proximal femoral focal deficiency.

Ideally, a child with a congenital short femur will be managed by re-alignment of the femur as a whole, followed by femoral lengthening with contralateral epiphysiodeses where appropriate. A Syme's amputation is reserved for those children in whom the foot is not a reasonably stable and functional component of the limb.

The child with a severe femoral deficiency and a significant foot deformity will be managed by a Syme's amputation and knee fusion and an above-knee prosthetic fitting. If it

is deemed appropriate, the child with a reasonably good foot may be treated by rotationplasty and knee fusion. The rotation being obtained through the former knee joint. The patient is then fitted with a rotationplasty prosthesis. For a very few patients with a grossly deficient femur and an inadequate hip joint, it may be advisable to perform an ilio-femoral fusion with the femur fused to the ilium in a flexed position. The child is then fitted with an above-knee type of prosthesis, with the former knee joint acting as the child's hip joint. It is not necessary to elucidate all the variations of femoral surgery that can be devised in these children, but where appropriate reconstruction of the proximal femur to overcome gross varus or to gain stability through a pseudarthrosis may be necessary.

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