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Fibular deficiency and the indications for Syme's amputation

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Abstract

Recent literature on the subject is reviewed, and the role of Syme's amputation, reconstructive surgery and prosthetic management is discussed in relation to the severity of the condition. Amputation, which should be performed between 18 months and two years old is specifically recommended for total fibula absence with ankle instability. The operative technique is detailed.

Introduction

Congenital fibular deficiencies are frequently a source of frustration and anxiety for the physician charged with meeting both the psychosocial and functional needs of these otherwise normally functioning children. The birth of such a child can engender feelings of guilt, blame, and helplessness in the parents. The concept of ablating the foot in order to facilitate prosthetic fitting is initially not well accepted as this organ may be quite normal. The parents may question why the same technology that sends men into outer space cannot be called upon to salvage their child's limb. The physician faced with this scenario must improvise, based on his expertise in several areas including orthotics and prosthetics, reconstructive ankle surgery and leg lengthening. Considering the variability of the condition, it is little surprise that over the years many methods have evolved to address the situation. Unfortunately, for the profoundly involved limb, few methods, other than amputation and prosthetic fitting, have stood the test of time. The purpose of this discussion is to review the subject of fibular deficiency, to examine the treatments currently available, and to put the subject of amputation into proper perspective.

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Background

The fibula serves an integral purpose in the lower limb as a lateral buttress to the talus as weight is transmitted across the tibial plafond during plantigrade activity. In addition, strain determinations of the fibula, imply that it maintains a weight-bearing function during stance (Lambert, 1971). Congenital malformations of the fibula result from an alteration of musculoskeletal organogenesis that probably occurs in humans at approximately the fourth to sixth week of embryologic life. Because these malformations are not associated with classic modes of genetic transmission it is likely that they result from some embryonic insult perhaps at variable times during the development of the limb bud. Experimental evidence suggests that the earlier the insult, the more the involvement of the proximal femur and fibula. Later insults involve the fibula and foot to a greater degree (Pappas *et al.*, 1972). Whatever the timing, the range of severity is from simple hypoplasia to a total absence of the fibula often associated with proximal femoral focal deficiency, shortening and/or bowing of the tibia, general limb growth retardation, delayed epiphyseal ossification, absence of the lateral or other rays of the foot, tarsal coalitions, residual fibrous bands, deficiency of muscle, genu valgum, and loss of ankle integrity. Upper limb anomalies are seen in a significant proportion of cases (Achterman and Kalamchi, 1979; Coventry and Johnson,

1952; Herring *et al.*, 1986; Hootnick *et al.*, 1977; Westin *et al.*, 1976). Although rare, the fibula is the most commonly congenitally absent long bone (Coventry and Johnson, 1952).

Classification

From a clinical standpoint the most important issues are the stability and function of the ankle and foot and the overall length of the limb. To better characterise the variability, a number of classifications have evolved. Coventry and Johnson (1952) subdivided the condition into three types based on severity. In Type I only one limb is affected and the degree of deficiency ranges from a shortened fibula to partial absence of its upper portion. In Type II, the involvement is unilateral but absence of the fibula is complete. In Type III, the involvement may be bilateral, and the deformity may occur in association with other malformations such as proximal femoral focal deficiency. Recently Achterman and Kalamchi (1979) refined Type I into Type I-A and Type I-B. In Type I-A, the entire fibula is present, but is shortened with the proximal fibular epiphysis distal to the upper physis of the tibia and the distal fibular physis proximal to the dome of the talus. In Type I-B there is partial absence of the upper fibula, and distally the lower fibular epiphysis is elevated above the talar dome and thus not buttressing the ankle. Generally, in Type I deformities the foot remains plantigrade but may be associated with a ball and socket ankle joint; equinovalgus or equinovarus may also be present. While some surgeons have commented that the degree of ankle instability is related to the absence of rays (e.g., a four toed foot can be salvaged, but a three toed foot should be sacrificed), the author has found that ankle stability can be directly assessed and that a three toed foot can often be quite serviceable. Thus, most Type I feet will not be converted to amputations. In Type II, however, the entire fibula is absent, the incidence of tibial bowing frequent, and ankle instability is the rule. In classifying these anomalies, the degree of tibial shortening increases with advancing staging, though associated femoral shortening is maximal in the Type I category (Achterman and Kalamchi, 1979).

In Type II deformities due to the lack of lateral malleolar buttress and lateral ligamentous support of the ankle joint, the foot very often tends to become subluxed into a valgus and equinus position. This can be accentuated by a fibrous anlage of bone that extends from the upper end of the tibia to the calcaneus, exerting a tethering force on the tibia. It is in these cases that surgical salvage of the limb is most difficult. In areas where cultural concerns do not allow amputation, or in lesser developed areas where surgery may not be readily available, it is both possible and permissible to fit a prosthesis around the foot. Surgically, from the turn of the century until the 1950's attention was directed toward stabilisation of the ankle followed by leg length equalisation. Stabilisation of the ankle was first described by Braun (1886). Since then a number of different methods have come and gone, many of which are now recognised as counter-productive. For example, stabilisation by arthrodesis was complicated by delayed ossification of the epiphyses, as well as by inadvertent injury to the distal tibial physis. The Bardenhauer procedure (Rincheval, 1895) involved inserting the talus into a sagittal split in the tibia, clearly breaching the growth plate. The Albee (1921) procedure involved the substitution of autogenous bone grafted into the tibial metaphysis in an effort to replace the fibular malleolus. With growth of the tibia, the buttress so created rose above the level of the ankle. More recent attempts at ankle stabilisation have centred on the Gruca procedure (Gruca, 1959; Serafin, 1967). In a review of this technique by Thomas and Williams (1987) at the Royal Children's Hospital in Melbourne patients were considered for the procedure only if they exhibited minimal shortening of the tibia, and a foot that comfortably reached the ground with no gross deformity. The operative technique consisted of an oblique sectioning in the sagittal plane at a point between the lateral and middle thirds of the lower articular surface of the distal tibial epiphysis and extending obliquely and medially for about 7 cm. The medial tibial fragment was then displaced upward and medially 1.5 cm, and the gap between the fragments above the physis packed with cortical bone. The space at the site of the growth plate was filled with adipose tissue to prevent physeal bridging and allow further growth. The tibial fragments were transfixed by two screws to maintain positioning during consolidation. A

primitive ankle mortise was thus formed. Of seven attempts reported, three have been converted to an amputation, three were awaiting amputation, and the remaining patient had embarked on a programme of leg length equalisation. The study concluded that although in some patients a satisfactory ankle may be achieved, this procedure was not suitable for every patient, and in most cases the operation can only be regarded as an interim procedure to be followed by a Syme's amputation due to the progressive leg-length inequality that develops.

Ankle disarticulation and prosthetic fitting

In approaching the subject of conversion to a prosthetic situation, Syme's amputation should be regarded as a reconstructive rather than as an ablative procedure. Kruger and Talbott (1961) pointed out that while the goal of preserving the foot might be laudable, in fact many children underwent repetitive operations culminating eventually in Syme's amputation, and that upon honest review there was good evidence that the procedure should have been offered as a primary treatment rather than for secondary salvage. Similarly, the author (Anderson *et al.*, 1984) found a higher incidence of complications in a series of 61 patients at the Los Angeles Shriner's Hospital when Syme's amputation was performed as a salvage procedure in multiply operated limbs rather than as a primary procedure. In this series with the exception of one patient who died of unrelated causes, all patients were ambulatory, and patient satisfaction was excellent. Patients participated in sports activities including bicycling, swimming, football, soccer, and roller skating. Patients did report occasional problems such as callouses and rashes, but on closer inspection these problems seemed to be related to prosthetic fit rather than to the stump *per se* and were easily addressed with minor prosthetic accommodations in the socket. Posterior heel pad migration was commonly encountered though it rarely required surgical intervention. Hypertrophy of the skin over the distal tibia, and prosthetic adjustment were usually enough to compensate for changes in heel pad position. Forty percent of the patients felt that they had no functional restriction at all and all of the adults reviewed were employed. In another study evaluating the physical and psychological function in young patients after a Syme's amputation, the results demonstrated a surprisingly easy adjustment process (Herring *et al.*, 1986.) Patients were able to compete in most athletic endeavours, and the prosthetic device provided a cosmetically acceptable appearance. These authors noted, however, that the age of amputation was of major importance, and that it was preferable that the procedure be done between the ages of 18 months and two years since at this time the infant has an incompletely developed body image and adapts to the new physical status quite quickly. It seems that a missing foot compensated by a functional prosthesis is more acceptable to a child or teenager than a significantly deformed foot which compromises activities and gait. Further enhancing the activities of these amputees are recent advances in energy storing prosthetic feet utilising newer materials which are now available and which have further extended their abilities in terms of athletic competition; they also report a more lively feel to the leg (Burgess *et al.*, 1983; Wagner *et al.*, 1987). In brief, particularly in the severely involved child, it is more definitive and simpler to lengthen the prosthesis each year rather than lengthen the child. Young children growing up with such a conversion are fully active and functional (Anderson *et al.*, 1984; Herring *et al.*, 1986).

Indications and technique of ankle disarticulation in children

James Syme first described ankle disarticulation with preservation of the heel pad in 1843, writing that "the risk to life would be small, that a more comfortable stump would be afforded, and that the limb would be more seemly and useful for progressive motion". Since then the procedure has been modified, particularly for the child, though the basics remain.

The indications for a Syme's amputation are (1) a deformity of the foot so severe that any surgery to make the foot plantigrade and functional is likely to fail (Wood *et al.*, 1965) and (2) a leg-length discrepancy of 7.5 cm or more, actual or predicted, by the time of skeletal maturity (Thomas and Williams, 1987). This value of 7.5cm is somewhat arbitrary and tends to vary between physicians (Pappas *et al.*,

1972; Thomas and Williams, 1987). The author now makes his decision more based on the ankle situation than the leg length discrepancy, provided the discrepancy at growth is not projected to exceed a 15 cm combined femoral and tibial discrepancy.

The procedure of Syme's ankle disarticulation begins by marking out the anterior and posterior skin flaps and carrying the dissection down through the subcutaneous tissue to the level of the medial and lateral collateral ligaments of the ankle. These ligaments are identified and divided so that the talar dome can then be pulled forward away from the distal tibia. In the interval between the talus and tibia the extensor hallucis longus tendon is identified as a key to the location of the neurovascular bundle posteromedially. By protecting this tendon and drawing it medially with a retractor, the dissection can be continued with the neurovascular bundle well-protected. The calcaneus is excised from the heel pad in a *subperiosteal* fashion so that the periosteum remains thus maintaining the hydraulic structural function of the fat pad. The entire calcaneal apophysis is excised so that it will not persist as an ossicle later in growth. Two centimeters of the Achilles tendon is resected so that there will be no tendency to reattach and pull the heel pad posteriorly. Once the talus and calcaneus and the remainder of the foot have been excised, the distal end of the tibia is then shaved with the use of a knife creating a broad base for weightbearing with or without a prosthesis but taking special care not to injure the distal tibial physis. This is done only in older children since smoothing and remodelling occur spontaneously in the younger child once the talus no longer occupies the mortise. The heel pad is then stabilised underneath the tibia utilising a K-wire and the skin is closed over a Penrose drain with interrupted nylon stitches for the skin. In approximating the heel pad to the tibia care is taken to maintain the neurovascular bundle all the way to the tip of the flap and to avoid crimping. After skin repair a spica cast is applied for short stumps (as when the amputation is combined with a knee fusion for PFFD) and a long leg cast applied when the knee can be effectively bent to prevent the cast from coming off. The drain is removed at two days, and six weeks is allowed for soft tissue healing. By eight weeks the stump is generally ready for prosthetic fitting.

Some authors have reported problems with stabilisation of the heel pad and have preferred the Boyd amputation in which the calcaneus is trimmed and displaced anteriorly in an effort to fuse it into the distal tibia or at least to stabilise it under the tibial plafond (Blum and Kalamchi, 1982; Boyd, 1939). The author has some experience with this procedure, and feels it is a reasonable alternative. There is danger to the distal tibial physis in attempting to expose the epiphyseal ossification centre, especially in this group of children in which the ossification can be delayed and the nucleus, at the time of surgery, quite small.

Alternate approaches

In cases of Kalamchi Type I hypoplasia in which ankle stability is not a problem, or in which the ankle can be made stable and plantigrade either by fibula lengthening or supramalleolar osteotomy, the secondary problem of equalising leg lengths becomes paramount. For mild discrepancies no treatment, a lift, or contralateral epiphysiodesis will suffice. Several authors have documented that growth inhibition was constant throughout childhood and that the relative difference in leg lengths remains constant (Hootnick *et al.*, 1977· Moseky, 1977; Ring, 1959; Westin *et al.*, 1976). Thus a 10% discrepancy at birth will translate to 10% discrepancy at growth, but the absolute amount of shortening will of course increase. For an average 37 cm tibia, this would translate to 3.7 cm. It is important to remember that the femur is also short, and that the total amount of shortening in the limb is the sum of all of the regions of shortening. The foot itself may also be short and this should be taken into account as well in planning future treatment. In general, total discrepancies predicted to be over 5 cm at growth are usually addressed by leg lengthening as opposed to shortening techniques. Although the newer methods of lengthening — Wagner, Ilizarov, DeBastiani — are reporting extended lengthenings in areas previously thought to represent contraindications (Bjerkreim and Hellum, 1983; Dal Monte and Donzelli, 1987; DeBastiani *et al.*, 1986; Paley, 1988), a word of warning is in order. In congenital conditions, as opposed to acquired conditions, the limb is programmed to be short. Bringing such a limb out to the length of the opposite member amounts to over-lengthening not only the bone but the soft tissues as well. Under these

circumstances small associated deformities such as genu valgum, or ankle subluxation may be accentuated by the act of lengthening itself. Several lengthenings may have to be done throughout growth to "keep up" and the average time of disability and recovery may be in the order of 12–18 months per segment lengthened. This is particularly true in the tibia where leg-lengthening is frequently tardy in healing, and may require supplemental autogenous grafts and expensive prolonged and repeated hospitalisations. The economic and psychologic burden on both the family and the child undergoing the lengthening procedure must also be considered. The complication rate for lengthening approaches 100%, and a marginal foot or tenuous union may require chronic bracing and preclude an active childhood (Aldeghiri *et al.*, 1985; Bjerkreim and Hellum, 1983; Dal Monte and Donzelli, 1987; Mosca and Moseley, 1986). For Kalamchi Type I the author believes that lengthening is a reasonable and attainable goal in selected cases. In Type II deformities with grossly unstable ankles, in view of the prolonged course in preserving a less than satisfactory ankle and the speculative nature of extended lengthening in congenital situations in general, the alternative of conversion to ankle disarticulation is still the primary treatment to be considered. Furthermore, if an amputation is to be done, it should be done early so the child experiences the least psychologic disturbance (Anderson *et al.*, 1984; Herring *et al.*, 1986).

Conclusion

A Syme's amputation in a young patient is compatible with athletic and psychological function closely approaching that of a non-handicapped child of the same age. It should be considered as a primary reconstructive procedure rather than a last resort in patients with total congenital fibular absence. With lesser degrees of fibular involvement each patient must be individually assessed so that the treatment best suited to his or her personal needs can be selected. In spite of the newer methods of lengthening, ankle disarticulation and/or prosthetic fitting remains the standard treatment against which the newer methods of reconstruction and lengthening must be functionally compared. Although a number of procedures are now being proposed as alternatives to a Syme's amputation and prosthetic fitting, none have yet duplicated the degree of long-term success that the Syme's procedure has enjoyed.

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