COMPLETE FIBULAR HEMIMELIA : A LONG TERM REVIEW OF FOUR CASES

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Fibular hemimelia is a partial or total aplasia of the fibula, and is usually associated with other anomalies of the tibia, femur and foot. It represents the most frequent congenital defect of the long bones and is the most common skeletal deformity in the lower limbs. A retrospective study was made of four patients (three males and one female) with total aplasia of the fibula. The average follow-up was 15 years. Three patients were treated by successive tibial lengthenings ; in one case amputation was carried out using the Syme technique. Lengthening required numerous operations and led to many complications, whereas the amputated case involved a single surgical operation without complications. The results of our cases and those reported in the literature suggest early amputation as the treatment of choice of Type II fibular hemimelia.

Keywords : fibular hemimelia ; aplasia ; congenital defect.
Mots-clés : hémimélie fibulaire ; aplasia ; malformation congénitale.

INTRODUCTION

Fibular hemimelia or postaxial hypoplasia of the lower limb is a congenital disorder characterized by the partial or total absence of the fibula (16). It is the most common deficiency of long bones (10, 12), and is usually associated with other anomalies such as absence of the lateral rays of the foot, tarsal coalition, absence of the anterior cruciate ligament, tibial and/or femoral shortening, valgus deformity of the knee, and equinovalgus deformity of the foot and ankle (1, 9, 13, 16, 17).

The aim of treatment in such cases is to restore the normal length of the limb, correct the axial malalignment and achieve walking with support on the sole of the foot. The type of treatment varies according to the degree of deformity ; in this context, controversy tends to center on the more complicated cases : some authors advise early amputation of the foot, whereas others favor limb-lengthening procedures associated with corrective osteotomies (12).

The present study describes four patients with fibular hemimelia followed for an average of 15 years and subjected to two different treatment modalities : staged limb lengthening (3 cases) and early amputation (1 case).

MATERIAL AND METHODS

Four patients (three males and one female) were treated for fibular hemimelia in our orthopedic department since 1971, and were followed for an average of 15 years (range 9 to 27 years). The left limb was affected in all four cases. Three patients were treated by limb lengthening, whereas one patient underwent early amputation with the Syme technique.

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Table I. — Results of treatment in the four patients with fibular hemimelia

Case	Gender	Follow-up	Treatment	Associated interventions	Complications	Final LLD	Limp	Pain	Subjective evaluation
1	Male	12 years	Lengthening (9 cm)	Tibial osteotomy Tarsal centralization	Infection Non-union	2 cm	Mild	No	Not assessable
2	Male	11 years	Lengthening (5.5 cm)	Tibial osteotomy Tarsal centralization Femoral varus osteotomy	Unstable genu valgum	3 cm	Mild	No	Satisfactory
3	Female	27 years	Lengthening (13.5cm)	Tibial osteotomy Tarsal centralization	Distal metaphyseal fracture of tibia Infection	2 cm	Mild	No	Satisfactory
4	Male	17 years	Amputation	None	None	4.5 cm	No	No	Satisfactory

LLD = Leg Length Discrepancy.

The Achterman and Kalamchi classification (1) was used : (a) Type I : Partial fibular defect (IA : proximal fibular epiphysis distal to the proximal tibial epiphysis, and distal fibular epiphysis proximal to the talar dome ; IB : 30-50% fibular defect, with no ankle support distally) ; and (b) Type II : total absence of the fibula. Our patients corresponded to Type II, since there was total absence of the fibula in all cases.

Cases 1, 2 and 3 were subjected to limb lengthening. All three patients presented anterior tibial bowing and shortening associated with moderate equinovalgus deformity of the foot and absence of at least one lateral radius. Other associated anomalies were genu valgum (cases 2 and 3), skewfoot (cases 2 and 3), and syndactyly of the second web space of the foot (cases 1 and 3). In addition, case 1 suffered severe mental retardation.

In those patients presenting a fibrocartilaginous remnant of the fibula and equinovalgus deformity of the foot (cases 1, 2 and 3), resection of the fibular residual fragment was performed between the first and second year of life, together with varus osteotomy of the calcaneus and lengthening of the Achilles tendon to correct the foot deformity.

The mean limb length discrepancy of the patients subjected to limb lengthening was 6.1 cm (range 5 to 7.1 cm) at the time of surgery. In all three cases tibial lengthening was carried out, for the older procedures with the Anderson modification of the Abbott technique (18) (an open osteotomy and diastasis control with two pins proximal and two pins distal to the osteotomy with frame fixation), and later with the Wagner approach (2). This involved two-step lengthening with an open osteotomy in the first step followed by daily 1-mm lengthening until the desired length was reached. This was followed by bone graft and osteosynthesis in the event of an insufficient callus, at an average patient age of 4.9 years (range 4 to 5.5 years).

Case 4 suffered complete fibular agenesis as well as the absence of the fourth and fifth ipsilateral lateral radii, tibial antecurvatum, a short femur and a completely dislocated equinovalgus foot. Treatment in this case consisted of Syme amputation associated with realignment osteotomy of the tibia.

The overall results were subjectively classified as either satisfactory or unsatisfactory, according to the following criteria : final limb length inequality less than 2.5 cm, absence of pain, degree of limping (mild, moderate, severe), cosmetic aspect, and patient satisfaction with the resulting walking performance.

RESULTS

Case 1

This patient required three lengthening operations, the first two using Anderson´s technique (at age 5.5 and 8.5 years) and the last using the Wagner procedure (at age 15 years). A lengthening of 3 cm was achieved with each operation (table I).

Following the first lengthening operation, the bone callus bowed anteriorly and required a realignment osteotomy, which healed without difficulty. Deep *Pseudomonas aeruginosa* infection complicated the second lengthening operation and prevented bone consolidation ; as a result, bone

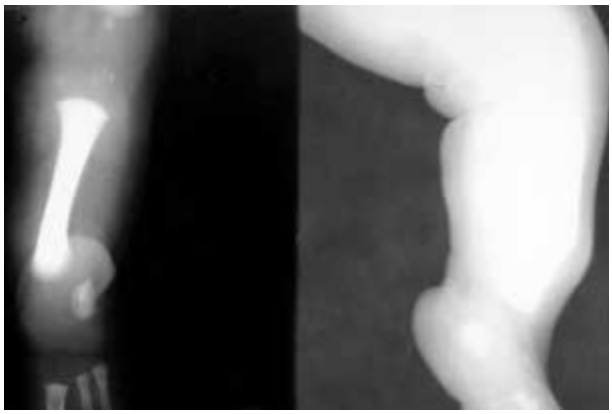

Fig. 1. — Radiological study after birth (case 3), showing absence of the fibula and fourth and fifth lateral radii of the foot, with anterior bend of the tibia.

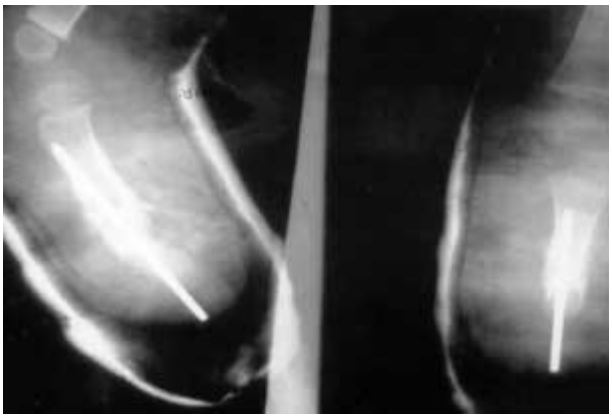

Fig. 2. — Postoperative x-ray follow-up of case 4. The Kirschner pin plays a double role by securing stabilization of the osteotomy and fixation of the plantar cushion.

grafting from the contralateral fibula proved necessary four months later to achieve healing. In the third operation and following the first step of the Wagner technique, local infection of the pin entry sites occurred and was satisfactorily resolved with antibiotics and local treatment.

The patient reported no pain and mild limping, but the final outcome was functionally affected by his severe mental retardation. He presented a final limb-length discrepancy of 2 cm, without requiring an orthosis.

Case 2

This patient required two tibial lengthening procedures : the first at four years of age, which resulted in a lengthening of 2.1 cm (Anderson technique), and the second at age 12 years with a lengthening of 3.4 cm by means of the Wagner technique. Severe knee instability resulted after the first operation, requiring combined tibial and femoral osteotomy ; a knee flexion contracture developed as a result of the second limb lengthening procedure (table I).

This patient reported no pain and a mild limping, and the subjective final evaluation was satisfactory. In the final assessment he presented a limb-length discrepancy of 3 cm, and a shoe elevation was required.

Case 3

Three lengthening operations were performed in this case, at ages 4, 7 and 14 years, using the Anderson (first two interventions) and Wagner (last operation) techniques. The resulting lengthening was 3.6 and 4.5 cm, respectively. After the first operation the patient suffered a distal metaphyseal fracture of the tibia owing to callus flexion, which was treated with a long-leg plaster cast. The second lengthening operation led to pin tract infection that caused frame loosening. Local infection developed during the second step of the third lengthening operation, requiring removal of the plate and screws and placement of an external fixator to ensure healing (table I).

In this case no pain and mild limping were also reported. The final limb-length discrepancy was 2 cm not requiring any orthosis. The final subjective evaluation was satisfactory.

The mean period of hospital stay among the patients subjected to tibial lengthening was 47 days (range 27 to 76 days) not including hospitalisation for complications.

Case 4

In this case there were no postoperative complications. The period of hospital stay was three days,

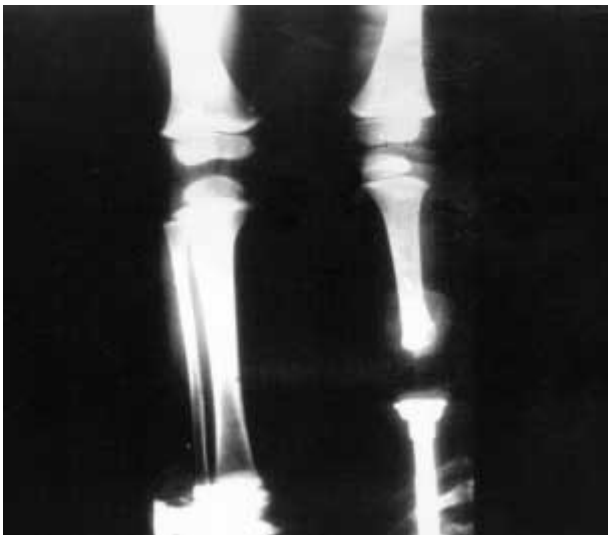

Fig. 3. — Case 4 at 7 years of age : radiological view in the erect position.

and only periodic adjustment of the orthosis was required. The patient reported no pain or limping, and the subjective evaluation was satisfactory.

DISCUSSION

Fibular hemimelia has a wide range of severity, ranging from mild hypoplasia of the fibula with minimal functional disturbance to complete absence and associated deficiency of lateral rays of the foot and tarsal bones (3). A short femur and an anterior cruciate ligament deficiency are frequently associated as well as hip, knee and foot deformities (13, 17) In our series, all patients presented total absence of the fibula and lateral bowing of the tibia. One patient developed a short femur. This was the amputated case and shortening was compensated with the prosthesis.

The treatment of Type II fibular hemimelia remains controversial. Early amputation with prosthetic support offers excellent long-term results with only minimal complications (11). Either Syme or Boyd amputation allows the patients to perform more activities than those who have a lengthening, with less pain and a lower complication rate. It was shown in many studies that the patients who underwent amputations had fewer procedures at a lower cost and were more satisfied that those who had a

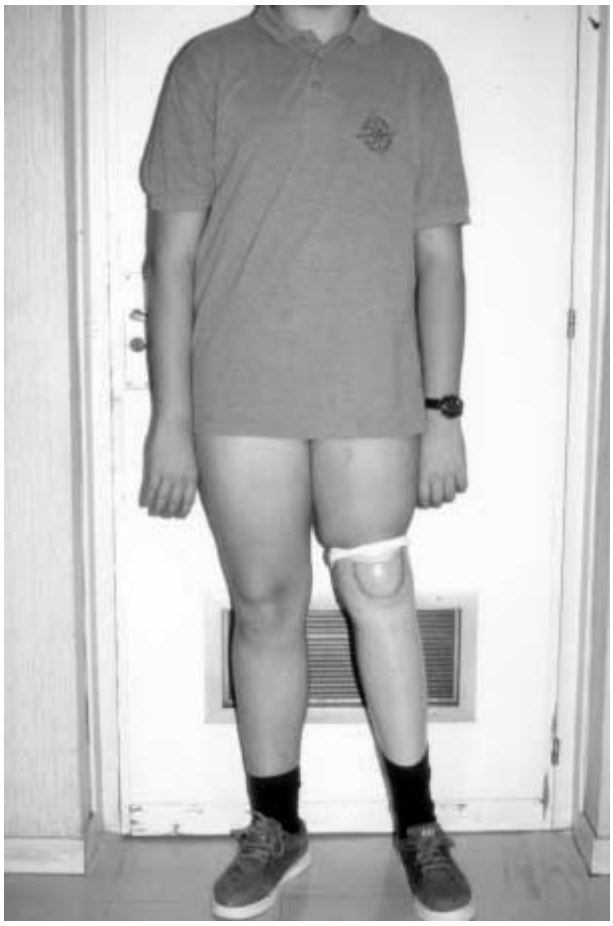

Fig. 4. — Present clinical appearance of the patient treated by early Syme amputation, in the erect position with his orthosis (case 4).

lengthening (4, 6, 11, 19). Amputation is irreversible and in some cases is not accepted by the parents (4, 6, 7).

On the other hand**,** lengthening procedures have been developed that make it possible to preserve the limb and simultaneously correct length and foot deformity. Several authors have shown that limb-length equalizing can be obtained and can result in a functional extremity. Nevertheless the outcome is not always satisfactory, and many complications can arise (5, 12). Few authors compared the results with those of amputations or collected outcome data (4).

Early complications described in the literature following limb lengthening include pin-tract infection or knee contracture and subluxation ; late

complications include progressive valgus deformity and bilateral subluxation of the ankle and foot with antecurvatum deformity of the tibia, stress fractures and delayed unions (3).

It has been stated that epiphysiodesis is a satisfactory solution when it is anticipated that limb length discrepancy will be 7.5 cm or less, but discrepancies greater than that would require tibial or femoral lengthening as well as contralateral epiphysiodesis (19). In our patients we did not perform contralateral epiphysiodesis because it implies surgery in a normal limb, which is difficult to accept by the child's parents.

Another treatment option is conservative with preservation of the foot and the use of an orthosis or prosthesis. This gives a short limb with a deformed foot acting as a stump. The result is a cosmetically unacceptable limb, which is not functionally better than the amputated extremity.

In a patient in whom the heel is in equinus angulation, and it is technically not feasible to center the calcaneus under the tibia, a Syme amputation has been recommended (4). For the severe form before walking age, amputation is the suggested treatment (3, 6).

Syme or Boyd amputations have been reported to give good long-term functional results if performed in early childhood (3, 4, 7). Early amputation and prosthetic fitting is the preferred treatment for the majority of these children (7, 16, 19).

The advantages of a Syme amputation include a more natural feel than transtibial amputation, the possibility of an amputation at a higher level in case of complications and the usefulness of the stump for weight bearing (8) ; and most of all, the fact that it is a single surgical procedure with a short hospital stay that allows immediate walking and equalization of limb length so that the child can adapt quickly and lead a normal life (12).

The major indications for amputation are : a deformity of the foot so severe that any surgery to make the foot plantigrade and functional would be likely to fail (19) and a leg length discrepancy of 7.5 cm or more, actual or predicted at the time of skeletal maturity (12, 19).

In the literature, the most frequently described complications following Syme amputation are wound infection or dehiscence of the stump sutures (5, 6, 7). In some cases the heel pad migrates owing to contraction of the tendinous structures, a problem that can be solved by fixing the pad to the tibia with a thick Kirschner wire. Other possible complications are ossification from the cartilaginous apophysis of the calcaneus due to incomplete excision (19), and phantom or stump pain (8). Syme amputation can result in a conical residual extremity, which compromises prosthetic fitting (7).

Boyd amputation creates a more cosmetically acceptable limb to which the prosthesis can be better fitted (4, 7). It has superseded the Syme procedure because preservation of the heel pad for weight bearing is more easily accomplished by the Boyd procedure, as the calcaneus is left intact. With the Boyd procedure, the function of the heel pad is improved (7).

The major disadvantage of an amputation is the fact that it is irreversible and that prostheses require periodic replacement and cannot provide normal sensation and proprioception (12). Also, parental refusal can be an obstacle (12). Other factors that may adversely affect the outcome are bilateral anomalies or an associated femoral focal deficiency (4).

Other modifications of the treatment have been reported : a modified Boyd amputation to improve the load-bearing characteristics of the residual limb ; a resection of the distal tibial physis at the time of amputation with subsequent proximal tibial epiphysiodesis (7) and a diaphyseal osteotomy of the tibia.

We find especially indicated the resection of the fibular anlage at the time of amputation to eliminate the potential tethering effect of the fibula, and this procedure has been performed in all our patients.

Based on the review of the literature and on the results achieved in our patients, the main indication for limb lengthening would appear to be cases with mild to moderate foot and ankle deformity allowing plantar support, with stable hip, knee and ankle, and an estimated final limb discrepancy at the end of the growth period of less than 7 cm (5, 10). In contrast, amputation appears to be the treatment of choice when the estimated final limb length dis-

crepancy is 7.5 cm or more, and the foot deformity is sufficiently severe not to allow functional support on the sole of the foot (12). The ideal age for amputation is before the child begins to walk, to reduce the psychological impact and the adjustment problems associated with the use of prostheses (4, 12, 19).

Femur and tibia respond differently to lengthening. Growth stimulation after lengthening of the femur appears in congenitally short femur and after lengthening of the tibia. In cases of fibular hemimelia, growth inhibition appears (14, 15). In patients with a lengthened tibia followed to skeletal maturity, a decrease in the postoperative growth rate of the lengthened tibia has been observed, ranging from 80% to 28%, the average decrease being 52% (15).

The long-term course of Type II fibular hemimelia as noted in our series, as well as the results the literature, points to early amputation as the initial treatment of choice.

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SAMENVATTING

J. TOMÁS-GIL, D. VALVERDE BELDA, J. CHISMOL-ABAD, C. VALVERDE-MORDT. Fibulaire hemimelie. Vier gevallen met lange opvolging.

Fibulaire hemimelie bestaat in een partiële of volledige aplasie van de fibula, meestal samengaand met andere misvormingen aan onderbeen, dijbeen of voet. Het is de meest voortkomende congenitale misvorming van de lange beenderen en de frequentste congenitale botafwijking ter hoogte van het onderste lidmaat. Vier gevallen (3 mannelijk en 1 vrouwelijk) van totale fibula aplasie werden gemiddeld 15 jaar opgevolgd. Bij de drie eerste patiënten werden opeenvolgend tibia verlengingen uitgevoerd. Een Syme amputatie werd bij de laatste uitgevoerd. Verlenging vroeg multiple ingrepen met veel verwikkelingen, terwijl amputatie in één ingreep zonder

meer de oplossing bracht. Derhalve menen wij uit de resultaten van deze vier gevallen en deze die voordien zijn gepubliceerd, te moeten besluiten dat een Syme amputatie voor fibulaire hemimelie type II de aangewezen aanpak is.

RÉSUMÉ

J. TOMÁS-GIL, D. VALVERDE BELDA, J. CHISMOL-ABAD, C. VALVERDE-MORDT. Hémimélie fibulaire complète. Présentation de quatre cas suivis à long terme.

L'hémimélie fibulaire consiste en une aplasie partielle ou totale de la fibula et est habituellement associée à d'autres anomalies au niveau du tibia, du fémur et du pied. C'est l'anomalie congéniale la plus fréquente au niveau des os longs et c'est également la déformation osseuse la plus commune au niveau des membres inférieurs. Les auteurs présentent une étude rétrospective de 4 patients (3 de sexe masculin et un de sexe féminin) qui présentaient une aplasie totale de la fibula. Le suivi moyen était de 15 ans. Trois patients ont été traités par des allongements tibiaux successifs ; le dernier a subi une amputation selon la technique de Syme. Les allongements ont exigé des opérations nombreuses et ont donné beaucoup de complications, tandis que le patient amputé n'a subi qu'un geste chirurgical sans complication. Les résultats observés dans ces 4 cas, comme ceux qui sont rapportés dans la littérature, suggèrent que l'amputation est le traitement de choix de l'hémimélie fibulaire de type II.