Ankle Reconstruction in Fibular Hemimelia: New Approach

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Abstract *Background:* Fibular hemimelia is a congenital disorder that is characterized by the absence of the fibula that could be either partial or complete. Successful management aims to restore normal weight bearing and normal limb length. The introduction of the Ilizarov method of limb lengthening has provided an attractive alternative to amputation. During lengthening, the tight posterolateral soft-tissue structures, the thick fibrous fibular band, and the shortened Achilles tendon become tighter and transfer a valgus force to the talus and calcaneus, further aggravating the deformity. *Questions/Purposes:* We have developed a strategy to address this in patients with Paley type III fibular hemimelia via ankle reconstruction that provides posterolateral stability and buttressing of the ankle and hind foot by reconstructing the lateral buttress. This is achieved through excision of the fibrous fibular anlage, centralization of the ankle, restoring talocalcaneal coronal alignment, and reconstruction of the lateral malleolus by transplanting the cartilaginous remnant of the lateral malleolus or by crafting a bone block autograft taken from the iliac crest or tibia. *Methods:* A prospective non-randomized clinical trial included ten ankles in eight patients with fibular hemimelia Paley type III (two patients had bilateral deformity). The patients' ages ranged from 7 to 36 months. *Results:* After a follow-up ranging from 48 to

96 months, a stable plantigrade foot was achieved in nine ankles; one ankle had residual equinus, five ankles had residual valgus heel, and eight ankles had complete range of motion of the ankle, whereas one patient lost 5° of dorsiflexion. One ankle had equinus deformity. *Conclusions:* To achieve satisfactory results, a stable plantigrade foot and ankle is necessary in patients with fibular hemimelia before attempting to equalize limb length discrepancy. It is important to reconstruct the ankle through an extra-articular soft tissue release, anlage resection, osteotomies, and restoring the abnormal talocalcaneal relationship before any attempt to equalize LLD.

Keywords fibular hemimelia · ankle reconstruction

Introduction

Fibular hemimelia is the most common congenital deficiency affecting long bones. Until recently, the accepted treatment of choice for severe Paley type III fibular hemimelia was Syme's or Boyd's amputation. The option of distraction lengthening using the Ilizarov technique is now available [7].

Fibular hemimelia is a congenital disorder that is characterized by the absence of the fibula that could be either partial or complete [15]. It could be unilateral or bilateral. It is also associated with a wide spectrum of anomalies, which involves the whole lower limb, with defects of the femur, knee, tibia, ankle, and foot [3].

The usual clinical presentation involves limb-length discrepancy (LLD), anteromedial bowing of the tibia, valgus deformity of the knee, equinovalgus deformities of the foot, and ankle instability with absence of the lateral rays of the foot [12].

Successful management aims to restore normal weight bearing and normal limb length so that the patient can walk with a gait that is as normal as possible. In mild cases, treatment includes shoe raises, step-in prostheses, epiphysiodeses or limb-lengthening procedures, and

Level of Evidence: Level IV

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Table 1 Patient information

No.	Sex	Age (ms)	Limb	Side	Classification Paley	Associated deformities	LLD (cm)	Soft tissue release	Talocalc. ost.	Lat. Mal. Reconc	Follow up period (ms)	Comp.
1	M	12	Bilat.	Rt	IIIb	Absent distal phalanges	–	+	+	Anlage	24	Ankle stiff.
2	M	12	Unilat	Lt	IIIb	Absent distal phalanges	–	+	+	Anlage	24	Ankle stiff.
3	F	18	Unilat	Rt	IIIc		–	+	+	Anlage	28	
4	F	18	Bilat.	Lt	IIIc	Absent lat. ray	2	+	+	Anlage	28	Residual valgus
5	F	18	Unilat.	Lt	IIIb	Absent lat. ray	5	+	+	Anlage	22	Residual
6	M	9	Unilat.	Rt	IIIc	HLFC		+	+	Anlage	28	equinus
7	F	36	Unilat.	Lt	IIIc	Absent lat. ray tibial bowing	4	+	+	Bony block	20	Residual valgus
8	M	36	Unilat.	Rt	IIIa	Tibial bowing	5	+	+	Contra-lat. fibular head	21	
9	F	7	Unilat.	Lt	IIIc	Absent lat. ray tibial bowing	4.5	+	+	Anlage	22	
10	F	23	Unilat.	Lt	IIIb	Absent lat. ray	6	+	+	Anlage	22	

correction of foot deformities. For more severe deformities, the management is controversial. Many authors recommend early amputation of the foot and prosthetic rehabilitation [10, 17]. In some countries, parents refuse amputation based on social and religious considerations.

Several classification systems were introduced to describe the spectrum of fibular hemimelia. The most popular was that of Achterman and Kalamchi [2]. Although simple and comprehensive, they lacked the detailed description of the true pathomorphology of the ankle and subtalar joints. Paley's classification addressed the ankle and subtalar morphology and the presence or absence of tarsal coalition. In this classification, fibular hemimelia was divided into four types; a stable normal ankle, type I; a dynamic valgus ankle, type II; a fixed equinovalgus ankle, type III; and a fixed equinovarus ankle (clubfoot type), type IV. Type III was further subdivided into a, b, c, and d according to the

Fig. 1. Preoperative AP weight-bearing hip-to-ankle radiographs of a 3-year-old female child with fibular hemimelia of the right leg, demonstrating complete absence of the fibula, genu valgum, hypoplastic lateral femoral condyle, and limb length discrepancy of 7 cm (5.5 cm tibial, 1.5 cm femoral).

Fig. 2. T1-weighted coronal magnetic resonance image through the ankle demonstrates talocalcaneal coalition (*arrow*) and lateral translation of the calcaneus in relation to the talus.

anatomical anterior distal tibial angle and the talocalcaneal relationship [20].

The introduction of the Ilizarov method of limb lengthening provided an attractive alternative to amputation. Some surgeons prefer the Ilizarov method because it can provide simultaneous correction of LLD and ankle or foot deformities [18]. The high incidence of complications associated with lengthening procedures is well known [1, 21]. During lengthening, the tight posterolateral soft-tissue structures, the thick fibrous fibular band, and the shortened Achilles tendon become tighter and transfer a valgus force onto regenerated bone and the talus and calcaneus, further aggravating ankle instability and deformity. The absence of the

Fig. 3. Intraoperative image depicts exposure of the lateral side of the leg and exposure of the whole fibular anlage (*forceps*) and dissection of the superficial peroneal nerve free of it (*arrow*).

Fig. 4. Intraoperative image demonstrating excision of the fibrous fibular anlage and bulbous cartilaginous lateral malleolus.

lateral malleolus, tight lateral fibular anlage, tight Achilles tendon, and wedge-shaped distal tibial epiphysis results in a loss of posterolateral hindfoot support thus aggravating foot deformity and ankle instability [2, 9]. This has lead some authors to attempt a procedure that releases the tight posterolateral soft tissue tethers and provide a lateral buttress to the ankle, with minor differences in their techniques [11, 14, 20].

The purpose of this study was to evaluate our approach of hindfoot realignment by breaking the talocalcaneal coalition and stabilizing the ankle utilizing three different options to reconstruct the lateral malleolus in Paley type III fibular [20] hemimelia.

Patients and Methods

A clinical trial describing a technique for patients with Paley type III fibular hemimelia was conducted at our institute between January 2008 and December 2014. Signed informed consent was received from the parents of all the patients. Parents were informed that the surgical procedure is a staged procedure necessitating multiple surgical interventions and were made aware of the possible associated complications.

The study included ten ankles in eight patients with Paley type III fibular hemimelia (two patients had bilateral affection). Treatment was ankle and hindfoot reconstruction. Inclusion criteria were the presence of clinical records, age

Fig. 5. Intraoperative image depicting the lateral side of the right leg after complete excision of the fibular anlage and bulbous cartilaginous lateral malleolus; it also shows continuity of the superficial peroneal nerve at the proximal part of the wound.

Fig. 6. Intraoperative image demonstrating talocalcaneal coalition breakage by sharp osteotome passing from superolateral to inferomedial.

before skeletal maturity, and isolated fibular hemimelia (absence of other proximal limb anomalies). Three boys (four ankles) and five girls (six ankles) were included. All patients

Fig. 7. Lateral view obtained intraoperatively with image intensifier depicting calcaneo-tibial transfixing K-wires, which were also advanced proximally intramedullary fixing the tibial corrective osteotomy.

Fig. 8. Schematic diagram explaining operative steps of ankle reconstruction. **a** Excision of the fibrous anlage and bulbous cartilaginous lateral malleolus (dotted lines). **b** Talocalcaneal breakage (red line) from a superolateral to inferomedial direction. **c** Talocalcaneal realignment and fixation by calcaneo-tibial wires; it also shows reimplantation of bulbous cartilaginous lateral malleolus at a more distal level (arrow).

had an equinus deformity of the ankle. All the patients had associated deformities of the tibia and foot (anteromedial bowing and absent lateral rays). Table 1 shows patient's details. Radiographic work up included full limb x-rays (Fig. 1) and preoperative MRI (Fig. 2).

Operative Techniques

With the patient in a supine position and a tourniquet applied, a zigzag longitudinal lateral incision is made with the anterior and posterior skin and subcutaneous tissue flaps developed along a line that runs parallel to the posterolateral cortex of the tibia. This allows for a wider exposure and decreases the tethering effect of a straight longitudinal scar. The incision is extended distally to the calcaneus with care taken not to injure the sural nerve. Proximally, the incision is extended just distal to the knee joint. The deep fascia is incised longitudinally. Proximally, soft-tissue dissection is continued to identify and isolate the superficial peroneal nerve. The fibrous band (anlage or fibular remnant) is

Fig. 9. Image shows excised fibrous fibular anlage with bulbous cartilaginous later malleolus (arrow).

Fig. 10. Intraoperative image showing suturing (*arrow*) of the cartilaginous lateral malleolus onto the posterolateral aspect of the distal tibia after talocalcaneal realignment and fixation.

dissected and completely excised from the common peroneal nerve proximally, to the cartilaginous remnant of the lateral malleolus distally (Figs. 3, 4, and 5).

Distally, a z-plasty lengthening of both peroneal tendons and Achilles tendon was performed. The talocalcaneal coalition was addressed by introducing a sharp osteotome obliquely from superolateral to inferomedial between the talus and calcaneum. With careful and gentle manipulation,

Fig. 11. Lateral radiographs of the leg in a cast taken immediate postoperatively, demonstrating realignment of the calcaneus beneath the talus and corrective of tibial osteotomy and securing positions with calcaneo-tibial intramedullary wires.

Fig. 12. Coronal T1-weighted magnetic resonance image through the ankle at 2-year follow up, demonstrating good talocalcaneal coronal alignment (*arrow*).

the osteotome is levered to effect inferior and medial translation of the calcaneum beneath the talus (Fig. 6). The corrected talocalcaneal position was maintained by two transfixing Kirchner wires (K-wires) introduced from the plantar surface of the calcaneus, to the distal tibia (calcaneo-tibial fixation). These wires are then further advanced proximally to fix the tibial osteotomy done to correct the anteromedial bow. Another technique is to introduce the K-wires intramedullary into the distal tibia osteotomy fragment to exit at the heel fixing talocalcaneal position; the K-wires are then advanced proximally short of the proximal tibial physis to fix the tibial osteotomy (Figs. 7 and 8)

In eight limbs, the cartilaginous anlage of the lateral malleolus was found bulbous (Fig. 9), and it was used to reconstruct the lateral malleolus, by reimplanting it at a more distal level (the level of the lateral malleolus) and securing it by sutures onto the posterolateral aspect of the distal tibia using heavy Ethibond sutures (© Ethicon US) (Fig. 10).

In the two limbs in which the fibula was completely absent and no remnant of the lateral malleolus was present, the lateral malleolus was reconstructed using a harvested contralateral fibular head and neck in one case [14] and by using a bony wedge taken from the corrective osteotomy of the anteromedial bow of the tibia in the other case [22]. Both of which were fixed to the distal tibia by K-wires.

Postoperative management

All patients were put in an above-knee cast for 4 weeks. This is replaced by a below-knee cast for an additional 2 weeks, after which the K-wires are removed and the leg is fitted into an ankle foot orthosis for further 4 weeks. Physiotherapy in the form of passive and active range of motion out of the brace is started immediately after cast removal. Serial radiographs were obtained for all patients during the follow-up period. All patients had an MRI of the ankle 6 months and 2 years mark (Figs. 11 and 12).

Results

The outcome measure was obtained maintaining a stable plantigrade ankle and foot for the whole length of follow up (Figs. 13 and 14). The mean follow-up duration was 53 months (range, 48–96 months). At the latest follow up, nine ankles had a stable plantigrade foot and one ankle had residual equinus. Two ankles had residual valgus heel.

Discussion

The fibular hemimelia remains a complex pathological abnormality that affects different aspects of the limb [5, 6]. Limb reconstruction through staged limb length equalization and deformity correction remains an appealing option.

A review of the literature revealed that most of the unsatisfactory results after these procedures were related to high grades of fibular hemimelia with complex foot deformities.

Naudie et al. and Choi et al. were able to achieve satisfactory results in patients treated by tibial lengthening without ankle reconstruction. After comparing their results with those of amputation, the authors concluded that amputation achieved better functional outcomes than did reconstruction. They explained unsatisfactory results by the residual foot and ankle deformities [8, 19].

Gibbons PJ and Bradish CF lengthened four tibiae with foot incorporation in the frame. The desired length and plantigrade stable ankle and foot were achieved in all patients. However, the authors reported late complications of progressive valgus deformity of the tibia and subluxation of the ankle that required corrective surgery in the form of

Fig. 13. Clinical photograph at 2 years follow up, depicts proper alignment of the right heel on weight bearing.

either soft-tissue or bony procedures in three patients. The authors concluded that the abnormal posterolateral soft tissues could not grow and elongate at the same rate as the lengthened tibial segment, which in turn will lead to posterolateral tethering and subsequent progressive valgus deformity and lateral subluxation of the ankle [13].

Paley reported the outcomes of 38 patients with fibular hemimelia treated by his SUPER ankle technique and subsequent lengthening. He reported satisfactory functional outcomes in 36 patients; however, the complication rate was high, but not related to the final outcome [20].

Fig. 14. Clinical photograph of the lateral side right leg, demonstrating neutral position of the ankle; it also shows the scar of a zigzag incision with no tethering effect of scar tissue.

Changulani et al. reported good outcomes of eight patients treated by soft-tissue release and reconstruction and then lengthening and deformity correction with the Ilizarov frame or the Taylor spatial frame system [6].

Thus, there is an agreement that the unsatisfactory results after the staged limb equalization procedures reported in the literature are because of the recurrence of foot and tibial deformity. Moreover, in the series that reported good results, a stable plantigrade foot was achieved [4, 11, 15, 16].

Advocates of staged limb length equalization procedures without reconstructing the ankle might argue that even if the ankle fails, arthrodesis remains a resort to stabilize a severely deformed hindfoot and ankle [4]. The authors believe that, for younger age with higher grades of fibular hemimelia and severely deformed hindfoot and ankle, multistaged lengthening with foot incorporation in Ilizarov frame then finally having to fuse the ankle will be rather socioeconomically demanding than would a single-stage procedure of below-knee amputation and prosthetic fitting.

The authors of the current study are proponent of staged limb reconstruction. This is dictated by the social and religious beliefs of the studied population, which considers amputation as an undesirable option. This mandates attempting to restore ankle and hindfoot stability and alignment to ensure satisfactory outcomes after future limb length equalization procedures.

In the present study, all deforming elements are addressed; the described zigzag incision, in addition to allowing adequate exposure, helps in decreasing the tethering valgus effect of scar tissue. In addition, as a major deforming structure, excision of the fibular anlage eliminates the lateral valgus deforming forces on the ankle and hindfoot. This is done combined with lengthening of the shortened peroneal and Achilles tendons [11].

Targeting the distal tibial valgus, Paley [20] described realignment of the distal tibial epiphysis through an incomplete supramalleolar osteotomy. The authors' experience suggests this osteotomy may be unnecessary. The rationale behind this is that achieving an aligned plantigrade foot will allow for normal distribution of forces along the distal tibial physis, thus, in theory, encouraging corrective distal tibial remodeling.

Different authors have described several methods to reconstruct the lateral malleolus. If present, the authors prefer using the cartilaginous remnant of the lateral malleolus, as described by Hefny et al. [14], Paley [20], and El-Tayeby and Abdel Razek [11], with acceptable results. This proved to be beneficial as it later ossifies during growth [14, 20]. Weber et al. [22] described using an iliac crest triangular tricortical autograft with the attached apophysis and gluteal fascia establishing good union and stable ankle [22].

In the present study, we had similar results regarding ankle and heel stability and alignment. To date, none of the patients experienced recurrence of valgus deformity, as evident by follow up MRI.

In addition to the small number of patients, short duration of follow up, non-randomization, and absence of control group, the major limitation of this study is that we could not compare our results with the rest of the literature. This is because the scanty literature on the topic lacks unified outcome measure. Moreover, the outcomes reported were after

lengthening procedures. All studies, apart from that of Changulani et al. [22], who used a modified version of the Pediatric Quality of Life Inventory to assess the functional outcome, did not use a unified patient-oriented functional scoring system that is mandatory for evidence-based recommendations.

We consider this to be an initial case series. Future studies are needed using a validated and patient-oriented functional scoring system to compare the functional outcomes between patients who underwent ankle reconstruction with those who did not.

Compliance with Ethical Standards

Conflict of Interest: Hany Hefny, MD; ElHussein M. Elmoatasem, MD; Mahmoud Mahran, MD, MRCS; Tamer Fayyad, MD; Mohamed A. Elgebeily, MD; Ahmed Mansour, MD; and Mamdouh Hefny have declared that they have no conflict of interest.

Human/Animal Rights: All procedures followed were in accordance with the ethical standards of the responsible committee on human experimentation (institutional and national) and with the Helsinki Declaration of 1975, as revised in 2008 (5).

Informed Consent: Informed consent was obtained from all patients for being included in the study.

Required Author Forms Disclosure forms provided by the authors are available with the online version of this article.

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