Limb sparing reconstructive surgery and Ilizarov lengthening in fibular hemimelia of Achterman–Kalamchi type II patients

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The aim of this study is to evaluate the long-term results of management of fibular hemimelia (Achterman–Kalamchi type-II) using the Ilizarov method. We reviewed 157 consecutive patients (180 limb segments) with a mean follow-up period of 10.7 years (1.2–21 years). The results were favorable. Although, this type of management is technically demanding and entails a lengthy procedure with many complications anticipated, the Ilizarov lengthening after limb reconstruction is still an option for management of this type of limb deficiency. *J Pediatr Orthop B* 19:55–60 © 2010 Wolters Kluwer Health | Lippincott Williams & Wilkins.

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Introduction

Fibular hemimelia is the commonest congenital deformity or absence of long bones, with an incidence ranging from 7 to 20 per million live births [1]. It encompasses a spectrum of anomalies affecting femur, knee, tibia, ankle, and foot. The usual clinical presentation includes limb length discrepancy (LLD), anteromedial bowing of the tibia, valgus deformity of the knee, equinovalgus deformity of the foot, ankle instability, and absence of lateral rays of the foot [2].

Several classifications were proposed to address the deformity, with the Achterman–Kalamchi [3], being the most commonly used. Treatment modalities can be gathered under three main categories: (i) orthoprosthetic care, (ii) operations to improve orthoprosthetic intervention, and finally (iii) reconstructive, limb sparing, and lengthening techniques. Severe type II cases, with complete absence of the fibula, absence of more than one lateral rays of the foot, more than 5 cm LLD at birth and/or more than 8 cm anticipated LLD at skeletal maturity, tarsal coalition and severe unstable valgus ankle constitute a real management challenge for orthopedic surgeons. Early amputation of the foot and prosthetic rehabilitation was reported in the literature, with many advantages. The main disadvantages were: loss of normal sensation and proprioceptive feedback, need for periodic replacement of the prosthetic limb, stump complications, prosthetic failure and unpleasant appearance, and that the procedure is irreversible [4,5].

Recently there has been growing interest in limb salvage and reconstruction for patients with fibular hemimelia. The claimed advantages are: preservation of the foot, and normal proprioceptive feedback mechanism [6,7]. The major drawbacks encountered were: multiple operations needed, unpredictable outcome, long hospital stay, considerably high rate of psychological depression of the patient and/or his/her family, development of foot and ankle deformities, recurrence of anteromedial bowing of lower tibia, knee or hip stiffness, and loss of muscle power [8,9].

This study will present the long-term results of management of this severe deformity in a referral hospital using the reconstructive limb sparing technique and the Ilizarov method. This treatment option is specially considered when the family refuses amputations. Many studies compared the results of limb lengthening with that of amputation, but limb lengthening was sometimes performed using the old Wagner method [10] or the reported number of patients were too small, and/or they were short-term studies [1,9].

Patients and methods

One hundred and fifty-seven patients (180 limb segments) with Achterman–Kalamchi type-II fibular hemimelia were treated between the years 1986 and 2009. There were 96 girls and 61 boys. Mean age at presentation was 2 years (5 months–15 years). The right side was affected in 98 limb segments, the left side in 82 (Fig. 1).

Bilateral cases were 23 whereas unilateral affection was found in 134 patients. The clinical findings at presentation included: (Table 1).

The study was performed at Ascahu Specialized Pediatric Orthopedic Hospital, Germany and The Department of Orthopedics and Traumatology, Tanta University Hospitals, Egypt.

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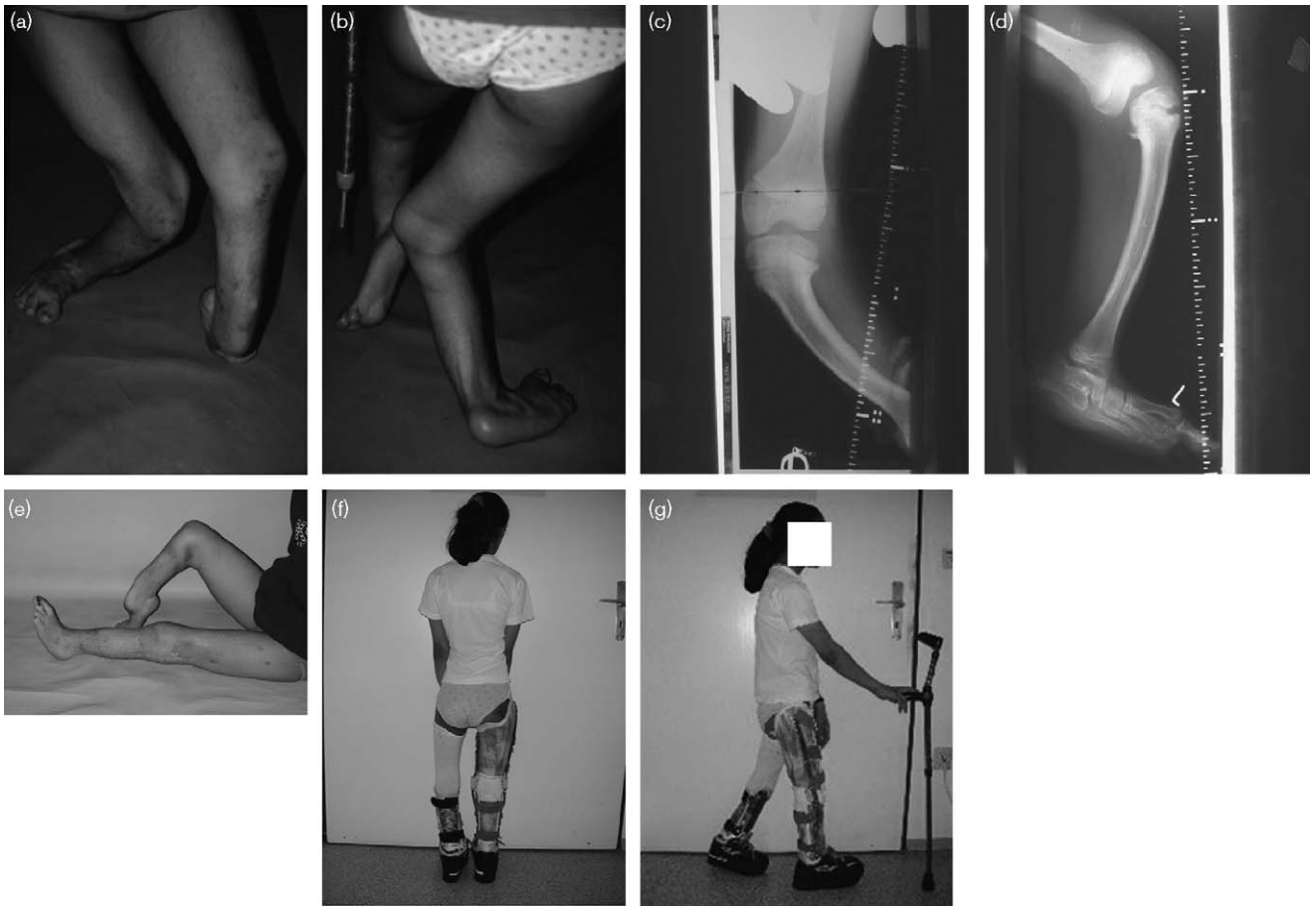
Fig. 1



(a and b) A 14 year-old-female patient with bilateral severe fibular hemimelia type II, at first presentation, (c and d) preoperative anteroposterior and lateral views for the patient, (e) clinical presentation after full correction of the left lower limb, and (f and g) final clinical presentation after bilateral correction of the deformity and independent full weight bearing. (Note that, on the left side only a below-knee orthosis is used after weaning off the knee-ankle-foot orthosis).

Table 1 Preoperative findings at presentation

Preoperative findings	Number of limbs affected
Limb length discrepancy	180
Fibular hemimelia syndrome	15
Tarsal coalition	117
Absent lateral foot rays	
4 rays	3
3 rays	81
2 rays	85
1 ray only	11
Absent anterior cruciate ligament	57
Ankle instability	166
Ankle or knee pain	44
History of previous surgery	37

(1) LLD that ranged from 2 to 15 cm (mean 7.9 cm at presentation). In bilateral cases the LLD ranged between 2 and 5 cm, whereas higher values (3–15 cm) were detected in unilateral cases, specially referred older patients.

(2) Fibular hemimelia syndrome was diagnosed in 15 limb segments (8.3%), with associated shortening and/or deformities of the ipsilateral femora.

(3) Tarsal coalition was detected in 117 feet (65%).

(4) There were three limbs with absent 4 foot rays (1.6%), 81 feet with absent 3 foot rays (45%), 85 feet with absent lateral 2 rays (47.2%), and 11 feet with only absent lateral fifth ray (6.2%).

(5) The anterior cruciate ligament was deficient in 57 limbs (31.66%) with concomitant knee instability and/or valgus deformity. The ankle joint was proven to be subluxed or dislocated (clinically and/or radiographically) in 166 feet (92.2%).

(6) Before the surgery, ankle or knee pain was a major complaint of 44 patients (28%). But it must be mentioned that the majority of patients presented, and were operated upon during the first year of life.

(7) Previous lengthening was performed for 37 patients elsewhere before their first presentation at other institutes.

Limb lengthening sessions

In all cases presented during the first year (104 patients = 66.2%), the first operation was performed by the end of the first year. They were treated with postero-lateral soft tissue release, tendon release and lengthening (s), achilles tendon lengthening, excision of the fibular anlage, correction of the tibial deformity by a closed wedge osteotomy at the point of maximum deformity, and finally centralization of the foot under the tibia. A transcalcaneotibial pin or Kirschner wire was used to stabilize the centralized foot under the tibia at the end of the surgical procedure and was removed by the time of bracing. In all the unilateral case, the anticipated LLD was more than 12 cm at skeletal maturity. The lengthening sessions for this group of patients were planned as follows:

(1) The first lengthening session at preschool age (around 5 years). In addition, the Ilizarov frame was also used to correct the tibial deformity. If severe procurvatum &/valgus of the tibial shaft was found, corticotomy at the point of maximal deformity was simultaneously performed. The rings were assembled to allow correction of the anteromedial angulation first with a distraction hinge and then transformed into a lengthening assembly. At both the proximal and distal ends, three to four 1.5 mm Ilizarov wires were attached to the rings with purchase of six to eight cortices. Extra care was taken to avoid damage to the common peroneal nerve. The wires were tensioned to 110 N. In all cases the position of the foot and ankle were controlled by an orthotic extension connected to the lower tibial ring. Distraction started 5 to 7 days after the surgery. All patients had unifocal lengthening at the proximal tibia at an average rate of 0.25 mm four times a day. Routine lengthening of the achilles tendon was done if the planned lengthening was more than 3 cm at this age.
(2) The second lengthening session was performed around 14 years old to equalize the LLD (particularly in unilateral cases). Moreover, residual or apparent new deformities were corrected at that time.

Cases presented after the first year of life

There were 53 patients (33.7%), referred after the first year of life. In those older cases, a supramalleolar varus and extension osteotomy with or without a subtalar osteotomy, were performed as needed to allow for correction of the valgus and equinus of the ankle joint.

Table 2 Indications for extension of the Ilizarov frame above the knee

Indication for above knee extension	Number of patients
Severe knee instability/deformity	51
$\geq$ 10 cm lengthening procedure planned/session	105
Fibular hemimelia syndrome	12

Please note that, more than one indication could be recorded in the same patient.

Recurrence of the foot deformity was prevented by excision of the fibular anlage and overcorrecting the bone regenerated by distraction towards varus alignment. In this group, the first lengthening session was performed concomitantly with the soft tissue release surgery. The second session was done as in the previous group.

The Ilizarov frame was assembled to allow correction of the anteromedial angulation of the tibia simultaneously with lengthening. The frame was extended above the knee in 143 limb segments (79.5%) using a hinged construct with an anterior extension bar that can lock the knee in extension.

The Ilizarov frame was extended above the knee in the following indications (Table 2):

(1) Severe knee instability/deformities (51 patients).
(2) If planned second lengthening session was estimated to be $\geq$ 10 cm to avoid knee flexion deformity (105 patients).
(3) If simultaneous ipsilateral femoral lengthening is indicated in patients with fibular hemimelia syndrome (12 patients).

Only 37 limb segments (20.5%), in this study were treated using simple tibial frames not extending above the knee joint (late presenting cases with previous lengthening sessions at other institutes).

Postoperative vigorous intensive daily physical therapy that focused on knee extension was used and found to be very helpful in prevention of knee flexion contractures; botox injection was not used for any of the patients in this study. The anterior extension bar was used in long frames (extending above the knee), allowed for locking of the frame in extension after physical therapy. This allowed for supervised mobilization of the patient with a locked construct during the day. A weight-bearing program was maintained throughout the period of treatment. A knee contracture developed in few patients (nine patients), and an early surgical intervention using a limited soft-tissue release was performed for those patients.

At the time of radiological corticalization, the frame was removed and further protection of the leg continued by an external hinged weight-bearing custom-made

knee-ankle-foot orthosis (KAFO). All patients were followed-up with regular radiographs and clinical assessment at monthly intervals in the first 6 months and then every 2 or 3 months as indicated. Special attention was paid to the functional status of the limb, its alignment, and the range of movement of the foot and ankle. The KAFO brace was regularly checked and adjusted during growth and rehabilitation of the patients.

The mean duration of postoperative follow-up was 10.7 years (1.2–21 years). During this period none of the studied 157 patients had an amputation or asked for one.

The Musculoskeletal Function Assessment (MFA) questionnaire was used in this study, at presentation (if applicable), and at the final follow-up visit. It comprises 101 items grouped into 10 categories: self-care, sleep and rest, hand and fine motor skills, mobility, housework, employment and work, leisure and recreational activities, family relationships, cognition and thinking, emotional adjustment, coping, and adaptation [11,12].

Results

All our patients had lengthening of the tibia and 12 of them had ipsilateral femur. The mean lengthening achieved was 13.6 cm (9.9–20.1).

The mean duration of application of the external fixator was 11.7 months/session (7.9–21.8), and the mean lengthening index (total treatment time in months per centimeter of lengthening) was 1.1 (0.82–1.70).

Limb length discrepancy was corrected to within 1 cm in all the treated cases (161 extremities) and in all the 23 bilateral cases (100%). This was owing to weak consolidation of the regenerate in 10 cases, and patients or parents poor compliance in nine patients. In those patients, cessation of the lengthening procedure and early removal of the frame was performed.

At the latest follow-up, the mean number of hospital admissions for the patients was 3.8 (3–8 times) and the mean length of stay at the hospital was 55 days (15–120). Outpatient care included attendances for physiotherapy, regular evaluation for bone consolidation, nursing and rehabilitation care, pin-tract infection control, orthosis follow-up, and management of minor complications.

Hospital admissions during the treatment course were for adjustment of the frame, management of severe or multiple pin-tract infection, removal of the fixator, and management of major complications.

Complications

There were minor postoperative complications including, pin-tract infection in 31 limbs, failure of one or multiple pins posttraumatically in 14 limbs, transient paresthesia in nine limbs, mild depression in four cases, soft tissue contractures in 29 limbs, orthosis failure and complications as pressure sores occurred in 11 limbs, and ankle or knee pain was reported in nine cases after exercise or prolonged walking, but did not affect the daily living activities. All the reported minor complications were treated properly and did not change the final outcome of the procedure (Table 3).

Table 3 Complications list

Types of complications	Number of limbs affected
Minor complications	
Pin-tract infection	31
Broken pins	14
Orthotic complications	11
Transient paresthesia	9
Soft tissue contracture	29
Tibial fractures	3
Ankle/ knee pain	9
Minor psychological disturbance (mild depression)	4 cases
Major complications	
Major psychological disturbance (severe depression)	9 cases
Weak consolidation	10

Three cases had fractures of the tibia after the removal of the frame. This was attributed to early uncontrolled weight bearing (one case), direct trauma (one case), and unfitted orthosis (one case). They were all treated conservatively and this fracture did not affect the final function of the patients.

In only 19 unilateral cases we could not reach the planned lengthening. Those patients were considered to have major complications. LLD in those patients was compensated using our custom-made orthosis. In only nine cases, we had to remove the frame early based on family's request in seven cases and psychiatric consultation in two cases.

Of the studied 157 patients, 141 received the MFA questionnaire, at the final follow-up evaluation and 16 patients could not be reached because they had changed their contact data. One hundred and nineteen patients completed the questionnaire (alone or with their parents' assistance), and 22 either returned an incomplete survey or refused to participate in this study. All the completed forms showed favorable results with patient and family satisfaction. Excellent results (above 85%) were reported by 70 patients and good results (above 75%) in 49 patients. It is of value to mention that the two patients with psychiatric troubles refused to complete this questionnaire. It is of value here to mention that all the parents were happy with the final end results, and independence of their siblings.

Evaluation protocol

The evaluation protocol of the studied patients comprised four major categories: physical examination,

prosthetic assessment, psychological testing, and physical performance testing.

The physical examination of the patients showed good alignment of the mechanical axis of the affected limb by the end of the lengthening procedure in all the studied cases. The foot was plantigrade in 141 limbs (78.3%), and mild residual equinus or valgus ($\leq 20^{\circ}$) was detected in 39 limbs (21.6%). This residual deformity was compensated with the use of the custom-made orthoprosthetic management. All the patients were able to use their lengthened limbs and walk independently and without crutches at the final visit. None of the affected patients showed nonunion of the tibia. Only delayed union was reported in 29 limbs (16.1%), the compression distraction technique (accordion technique) was used and succeeded in management of 19 extremities. In only 10 patients, the regenerate was weak, and the lengthening process was aborted after trials of regenerate stimulation.

Prosthetic assessment was performed regularly and periodically and followed by gait assessment. It was clear that the patient's access to prosthetic care had an influence on the patient's ability to reach his or her goals and interests. Psychological testing was also performed by specialists at the hospital to avoid psychological problems and early consultation was performed for cases that showed early depression symptoms. In only two cases, the frame was removed based on psychiatric recommendations (severe depression).

Physical evaluation of the knee and ankle joints without the brace was performed in all the cases. Weaning of the long-hinged KAFO was performed after about 6 months to 3 years (mean, 9 months) and only a below-knee custom-made orthosis was allowed to control the ankle and support the lengthened leg. At the time of final clinical evaluation, there were 49 patients (31.2%) walking without any orthosis.

Discussion

Management of severe fibular hemimelia has always been a challenge to pediatric orthopedic surgeons. The controversy still persists. Many authors recommended early amputation or disarticulation and prosthetic management to allow weight bearing. The advocates of this procedure consider the foot, in cases of advanced fibular hemimelia, to be nonfunctional and warn against the long treatment time, hospital stay, multiple procedures, and high complication rates in cases of lengthening [13–16]. Although early amputation seems to be an economic procedure, it has many complications and side effects, besides it is an irreversible procedure. We have to consider the cost of the prosthesis and how frequently it might be changed during life. With the introduction of the Ilizarov frames, a bigger interest appeared in limb lengthening and reconstruction [17–20]. The Ilizarov frame provides the option of foot preservation and normal proprioception of the limb. But still, it has many drawbacks. It is a demanding procedure, long operative time with multiple surgeries needed, multiple hospital admissions for surgery and follow-up, expensive frames and custom-made orthotics, long absence periods from schools, and relatively unpredictable duration of management. High intellect parents and very good patient and family compliance are a must for the success of this procedure. But it spares the child from the psychological trauma of an amputation [21].

In this series of 157 patients, amputation was not performed for any of the included cases. We tend to be on the conservative side and this was supported by the cultural background of the community and our decision was also based on the parents' demand, preference, and acceptance.

Independent walking, as well as physical activity (variable), was achieved in all cases of this study, including the complicated and the under-corrected cases.

Lengthening and equalization of the LLD was achievable in the majority of the cases, and only 12.1% did not reach the aimed length.

During the lengthening sessions there were difficulties encountered, the most important is delayed union which was managed by slowing the rate of lengthening, the 'accordion' technique, and regenerate stimulation by low-intensity ultrasound waves. This managed to treat the delayed unions in all but nine cases in this study, in which complete LLD equalization could not be reached.

It was found that, development of knee flexion deformity during lengthening could be controlled by the extension of the frame above the knee as previously mentioned indications. The hinged construct used allowed for active and passive movements during physiotherapy, and the knee was locked in extension for proper gait control during ambulation of the patients. Early soft-tissue lengthening was performed when indicated in all cases developing contractures to avoid knee dislocation, as was recommended by other authors [17,18].

A functional plantigrade foot was obtained in 141 limbs (78.3%) with normal proprioception and anticipated better life style and activity of the patient. Foot and knee subluxation was well controlled by careful external splintage and physiotherapy. This was confirmed and supported by the favorable (MFA) questionnaire results at the final follow-up.

Coexisting deformities added to the complexity of the disease. In cases of fibular hemimelia syndrome or femur fibula ulna syndrome, higher concern and extra care should be provided to avoid complications and the femoral shortening/deformity could be corrected simultaneously.

We agree with the opinion that, the irreversible and destructive nature of amputation surgery raises many ethical issues and makes many parents uneasy about deciding in favor of treatment with amputation [22]. We believe that whenever feasible, efforts should be made to involve the child in the decision-making process, especially when an ablative procedure such as amputation is being considered. If given an opportunity to participate in the decision-making process at an appropriate age and given the option of undergoing limb reconstruction with reproducible results, would the child have made the same decision in favor of amputation? Many older children resent irreversible surgical decisions made on their behalf.

We are of the opinion that, if a limb can be safely and reliably lengthened, reconstructed, and preserved, amputation should not be considered as an option. We believe that every effort should be made to save the limb in severe type II cases of fibular hemimelia. We disagree with the recommendation of Oppenheim [23], that amputation is indicated for a LLD of 7.5 cm or more by the time of skeletal maturity, and fore-foot deformities severe enough to indicate that any surgery to make the foot plantigrade and functional is likely to fail.

We recommend that it should be clearly explained to the family, as well as the patient, when applicable, that the procedure would take a long time. They are expected to be 'frame-friendly', and unless complete family support, preference, and compliance are guaranteed, failure, under correction, psychological problems, and complications are to be anticipated. We conclude that limb lengthening and reconstruction should be the treatment of choice in cases of severe (type II) fibular hemimelia. This option has to be specially considered when the family refuses amputations.

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