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Staged Soft Tissue, Bony and Ilizarov Procedures for Correction of Leg and Foot Deformities in Tibial Hemimelia

Amin AbdelRazek Youssef Youssef Ahmed*

Lecturer of Orthopedic Surgery, Alexandria University, Egypt

Abstract

Background: The incidence of tibial hemimelia is very rare being about 1 per million in the reported literature.

The deformity varies according to the type of the deficiency, the involved leg is short and the foot is held in equinovarus. In type II, partial development of the tibia eventually occurs, and a relatively functional knee joint is present, the fibula is usually normal in size, but the head is proximally dislocated

Material and methods: The material of this study included 8 patients with tibial hemimelia (Jones type II). Five were boys and 3 girls. The average age of the patients at the time of the first operation was 2.3 years (1-4 years) and the average follow up period was 31 (14-60 months). Right leg was the affected side in 3 while 5 patients had left sided tibial hemimelia.

Operative steps: Soft tissue release and centralization of the fibula in the ankle was done followed by tibiofibular fusion between the proximal tibia and the fibula (side to side) at the age of 3.5-4 years in all patients and to be followed by Ilizarov distraction at the age of 5 years to pull the fibula down and then continue lengthening to equalize limb length inequality.

Results: At the end of follow up period (31 months) good results were achieved in the eight cases with good range of knee movement and plantigrade foot, all patients could walk independently and without pain. All cases showed full satisfaction to the patients and their parents. Bone consolidation of the distraction site was achieved in all cases with 4.5-6.5 cm gained tibial length. There was no limb length inequality in 6 cases with residual shortening in 2 cases (2 and 2.5 cm) which did not affect the final satisfactory results.

Conclusion: Early Soft tissue correction, tibiofibular fusion followed by Ilizarov distraction gave satisfactory results in cases of tibial hemimelia (Jones type II) which were difficult to be treated by the conventional methods.

Introduction

The incidence of tibial hemimelia is very rare being about 1 per million in the reported literature [1].

It represents a spectrum of deformities, ranging from total absence of the tibia (the most severe form) to mild hypoplasia of the tibia (the least severe form).

The most widely used classification scheme for tibial hemimelia is that of Jones et al. [2] which is based on the early radiological presentation, Type II deformity is the most common type in which a proximal tibia of varying size is present at birth. Weber suggested a new classification scheme reflecting the severity the deformity and the importance of any cartilaginous anlage [3]

The deformity varies according to the type of the deficiency, the involved leg is short and the foot is held in equinovarus. In type II, partial development of the tibia eventually occurs, and a relatively functional knee joint is present, the fibula is usually normal in size, but the head is proximally dislocated [4,5].

The treatment, traditionally, has been by amputation and subsequent prosthetic fitting [4-10]. On the other hand, surgical correction of this deformity may avoid the limb amputation but it is difficult and challenging with the need of multiple surgical procedures to correct the foot and knee deformities and to equalize the limbs lengths [11-13]

The aim of this work was to evaluate the results of limb reconstruction in cases of tibial hemimelia (type II) by staged soft tissue and bony procedures followed by Ilizarov distraction osteogenisis.

Material and Methods

Between 2004 and 2012, limb reconstructions were performed in 8 patients with tibial hemimelia (Jones type II). Five were boys and 3 girls. The average age of the patients at the time of the first operation was 2.3 years (1 4 years) and the average follow up period was 31 (14-60 months). Right leg was the affected side in 3 while 5 patients had left sided tibial hemimelia. All 8 feet had equinovarus deformity and absent medial rays (1-2 rays), all cases showed limb length inequality which was assessed before the final stage of callus distraction and it ranged from 4 to 7 cm. There was associated lobster hand in a case, bilateral hand polydactyly in another case and syndactyly between second and third toes of the affected foot in one patient (Table 1).

***Corresponding author:** Amin AbdelRazek Youssef Youssef Ahmed, 33 Bahaa ElDin ElGhatwary st, Smouha, Alexandria, Egypt, Tel: 002031227392464; E-mail: aminrazek@yahoo.com

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Operative Procedure

Ankle centralization

A complete soft tissue release of the foot was done by a curved incision from the anterolateral side of the ankle and directed posteromedially, the distal end of the fibula was tapered with preservation of the distal fibular physis and a trough was done in the posterior facet of the calcaneus. The distal end of the fibula was placed into the trough done in the calcaneus. Ankle centralization was done at the first presentation (age of 1-2 years) in 5 patients while 3 patients were presented to us for the first time at the age 3.5-4 years to whom centralization was done at the same time of the second step, in these 3 cases fibular shortening (1 cm) was required to allow ankle centralization without tension on the neurovascular bundle. The centralized fibula was fixed in its position by a transcalcaneal K wire and a plaster cast for 6 weeks. After removal of the cast the foot was put in a splint till the next operation.

Tibiofibular fusion

At the age of 3.5-4 years tibiofibular fusion between the proximal tibia and the fibula (side to side) was performed with fixation by 1-2 small set screws and immobilization by plaster cast till achieving full union followed by AFK orthosis waiting for the last step.

Ilizarov procedure:

All cases were subjected after that to Ilizarov distraction at the age of 5 years using 2-3 ring-frame with a half ring fixed to the hid foot. Proximal tibial osteotomy was done without fixing the proximal fibula. Distraction started after the latent period (10 days) to lengthen the tibia and pull the fibula down to its normal anatomical site. Then fibular osteotomy was performed with fixation of the pulled down fibular head and continue lengthening to correct limb length inequality, (Figures 1 and 2).

Results

At the end of follow up period (31 months) good results were achieved in the eight cases with good range of knee movement and plantigrade foot, all patients could walk independently and without pain. All cases showed full satisfaction to the patients and their parents. Bone consolidation of the distraction site was achieved in all cases with 4.5-6.5 cm gained tibial length, the average external fixator time is 147.6 days (average 5 months) with healing index of 29.6 days/cm. There was no limb length inequality in 6 cases with residual shortening in 2 cases (2 and 2.5 cm) which did not affect the final satisfactory results.

No complications have been encountered during or after the procedures of ankle centralization and tibiofibular fusion. All the complications reported in our series are those commonly encountered when performing Ilizarov limb lengthening including: pin tract

Patient	Age at the first operation (years)	Sex	Side	Period of follow up (months)	Length gained (cm)	EF time (days)/healing index (days/cm)
1	1	M	R	18	6.5	185/28.5
2	3.5	M	L	48	5	145/29
3	4	F	L	60	4	124/31
4	1	F	R	14	6	182/30.3
5	1.5	M	L	28	4.5	130/28.9
6	2	F	L	20	5.5	162/29.5
7	3.5	F	R	24	4.5	140/31.1
8	2	F	L	36	4	114/28.5

EF=external fixator, M=male, F=female, R=right, L=left

Table 1: Description of patient series.

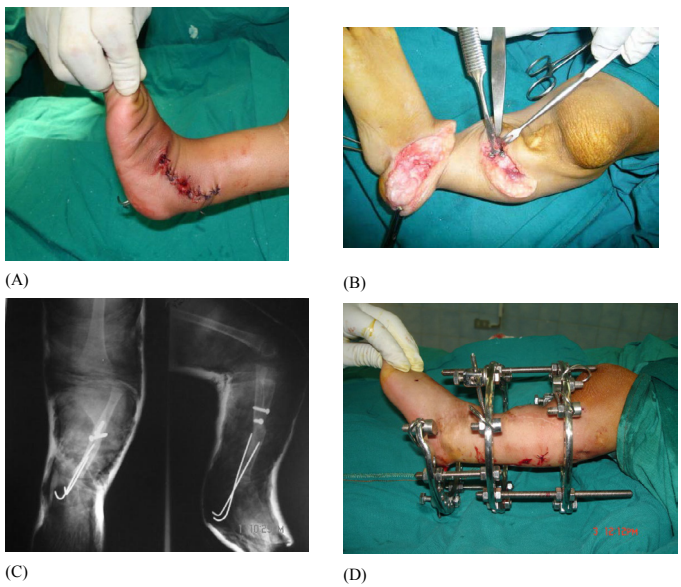

Figure 1: Operative procedure used in management of tibial hemimelia type II. Ankle centralization of the fibula (A). Tibiofibular fusion with internal fixation by 2 small set screws (B). X Ray picture after ankle centralization and tibiofibular fusion (C). Photograph of the patient after application of Ilizarov external fixator (D).

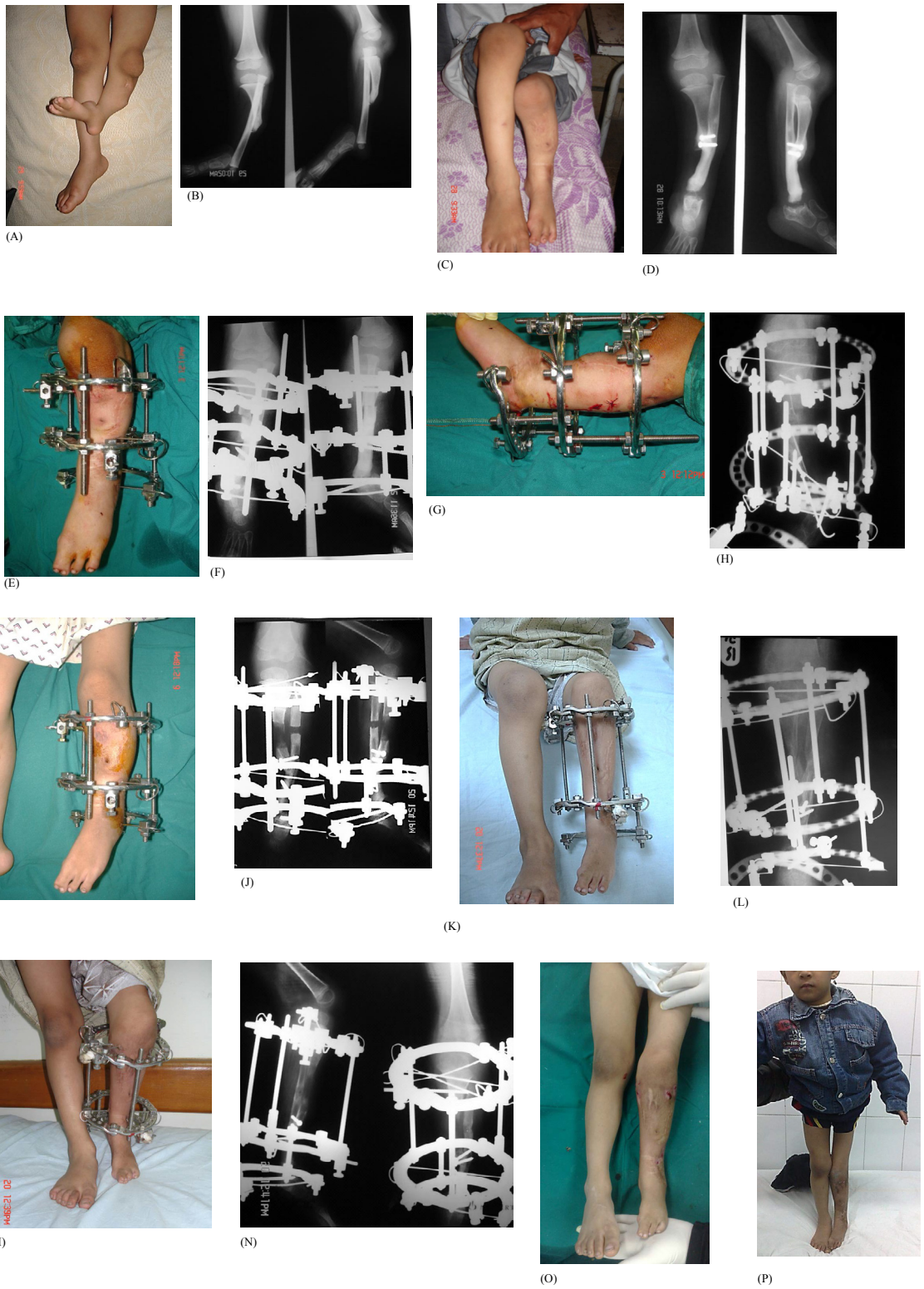

Figure 2: Clinical and radiological presentation of a 3.5 year old boy with type II tibial hemimelia. Preoperative photograph of the patient showing left foot equinovarus and left tibial shortening (A). Preoperative X Ray of the same patient (B). Photograph of the patient after ankle centralization and tibiofibular fusion showing correction of the foot deformity with a plantigrade foot and residual 5.5 cm shortening of the left leg (C). X-Ray picture of the patient after full fusion of the tibiofibular site with centralization of the distal fibula at the ankle (D). Photograph and X Ray of the patient after applying Ilizarov frame for tibial lengthening and to pull the fibula down to its anatomical site (E and F). Photograph and X Ray pictures of the patient after fibular osteotomy and fixation of the fibular head to continue lengthening of the leg (G and H). Photograph and X Ray pictures during limb lengthening (I and J). Photographs and X Ray pictures at the end of distraction showing correction of the leg and ankle deformities and consolidation of the distraction sites of the tibia and fibula (K, L, M and N). Photograph of the patient after removal of the frame (O). Photograph of the patient at the end of follow up (32 months) with full correction of the limb deformity and a plantigrade foot with a residual of 2 cm shortening (P).

Variable	Count (%) or Mean
Consolidation	8 (100%)
External fixation time	147.6 days
Healing index	29.6 days/cm
Limb length inequality	2 (25%)
Transit knee flexion	4 (50%)

Table 2: Final results and complications.

infection in all cases, knee flexion during the tibial lengthening in 4 cases and all were resolved completely during or after removal of the fixator without affecting the final outcome (Table 2).

Discussion

The gold standard of treating tibial hemimelia is amputation and subsequent prosthetic fitting [4-10], the most common approach in patients with a type II deficiency has been foot ablation and proximal tibiofibular synostosis [14]. The current trend for foot ablation in the literature is to perform a Syme or modified Boyd amputation [15]. Federico et al. [8] reported on 22 cases of tibial hemimelia, they treated 13 patients (type I, II and III) by amputation and prosthetic replacement, the only difference in the operative procedure was the level of amputation, and they assumed that the sooner the amputation is performed, the easier and faster the rehabilitation and adaptation to the prosthesis. An early amputation is accepted by the child as a "congenital amputation. Spiegel et al. [14] performed a clinical and radiographic review of 15 patients (19 limbs) with longitudinal deficiency of the tibia, five type II deficiencies were treated by foot ablation and tibiofibular synostosis, they achieved good results but prosthetic problems may arise from varus alignment and prominence of the proximal fibula.

On the other hand, limb salvage may avoid the limb amputation but it is difficult and challenging with the need of multiple surgical procedures. Amputation is not easily acceptable in our community, and the parents always ask for limb salvage and will never be satisfied at all by the amputation whatever the results could be.

The series of this study included 8 patients with type II tibila hemimelia. All patients were treated by the same protocol of treatment by ankle centralization, tibiofibular fusion followed by Ilizarov distraction to correct the shortening and the residual deformity. We succeeded to achieve good results in all cases with patients' parents satisfaction without the need for amputation in any case. On comparing our results of limb salvage with the results of amputation and prosthetic replacements, as regards the functional outcome, the duration of treatment, the incidence of complications and the number of surgical procedures needed, the amputation will be much better. But when parents satisfaction was added to the assessment, our results of limb salvage were very highly satisfied by the parents with the acceptance of any residual deformities or limb length inequality (in 2 cases).

The results of this study are comparable to Hosny [16] who reported on 6 cases (4 type II and 2 cases type I a) and had achieved good results with patients satisfaction in all cases including the more difficult cases of type Ia.

Wada et al. [17] reported on 9 cases (4 type II and 5 type I), they used the same technique performed in this series for limb reconstruction in type II and obtained satisfactory functional and cosmetic results in all limbs, whereas in limbs with type I deficiency, none of the 5 knees treated by fibular transfer achieved a satisfactory functional result.

Eamsobhana et al. [18] treated six cases of tibial hemimelia. They achieved satisfactory functional and cosmetic results in all patients with partial deficiency, whereas in patients with complete tibial deficiency, none of the 3 knees treated by fibular transfer achieved a satisfactory functional result.

Few cases were reported on reconstruction of tibial hemimelia in older literature; De Sanctis et al. [11] followed 3 cases till skeletal maturity, all three patients were very satisfied with the treatment they had received. Cranza et al. [19] reported on a case report of a 15 year-old girl with type II unilateral tibial hemimelia treated by talofibular fusion followed by distraction by external fixator and they achieved satisfactory results.

No complications have been encountered in our series during or after the procedures of ankle centralization and tibiofibular fusion, the same was reported in the series done by Wada et al. [17], who recorded no complication with tibiofibular fusion and ankle centralization except in one patient who showed progressive ankle equinus with the need to repeat the ankle centralization 1.8 year after the initial operation. All the complications reported in our series are those commonly encountered when performing Ilizarov limb lengthening. Pin tract infection occurred in 100% of case, all were superficial and resolved by local care and local antibiotics. Knee flexion contracture happened in 4 cases (50%) during limb lengthening which were resolved by physiotherapy during the consolidation phase in 2 cases and after removal of the frame in the other 2 patients. These are comparable to Wada et al. [17] who reported on superficial pin-tract infection in all patients which was controlled with the administration of antibiotics. In the series done by Hosny [16], pin tract infection occurred in all the cases and was treated by local or parenteral antibiotics with the need of drainage under local anaesthesia in two cases, replacement of a cutting through calcaneal wire and removal of an infected wire was done in two cases and knee flexion deformity (5°) persisted in two cases.

These minor complications which were resolved completely at the end of follow up did not affect the final good results in all patients encountered in this series.

Conclusion and Recommendation

Early soft tissue correction, tibiofibular fusion followed by Ilizarov distraction gave satisfactory results in cases of tibial hemimelia (Jones type II) which were difficult to be treated by the conventional methods. But longer follow up is needed to assess the achieved correction and to detect any recurrence of the deformity by time.

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