Limb reconstruction for tibial hemimelia

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Background
Tibial hemimelia is a rare congenital anomaly and is usually accompanied by congenital abnormalities in other parts in the body. The treatment is always difficult and challenging. Amputation was the preferred treatment option; however, a reconstructive surgery is sometimes used in the cases of amputation refusal.
Aim
The purpose of this study was to determine the results of limb reconstruction in eight children with Jones type II tibial hemimelia by fibular tibialization, ankle fusion, and lower limb lengthening using Ilizarov external fixator.
Patients and methods
A retrospective study was conducted on eight children presented with Jones type II tibial hemimelia, who were treated by fibular tibialization, ankle fusion, and lower limb lengthening using Ilizarov external fixator. The study had been done at Zagazig University hospitals between March 2007 and October 2014. The mean age of the patients was 3 years (range, 2–5 years). There were five girls and three boys. All had unilateral affection; right side was affected in five children and left in three. All cases had leg-length discrepancy that ranged from 5 to 7 cm, and all feet had equinovarus deformity.
Results
The mean follow-up period was 3 years (range, 2–5 years). In all the cases, the transferred fibular shaft to the distal end of the tibial remnant was united within 2–3 months. Consolidation of the bone at the distraction site was obtained with a gained length of 5–6.5 cm in all patients. The external fixator was applied for an average time of 150 days, and the healing index was 29 days/cm. The mean maximum knee flexion was 100° (range 90°–110°], and instability of less than 5° in varus valgus plane was seen in two cases. The ankle and foot deformity improved by foot repositioning and arthrodesis, achieving a plantigrade foot. The mean calcaneofibular angle was 100° (range, 90°–110°). The parents of all the patients showed full satisfaction at the latest follow-up.
Conclusion
The results of limb reconstruction of the Jones type II tibial hemimelia by fibular tibialization, ankle fusion, and lower limb lengthening using Ilizarov external fixator were acceptable. The limb was saved and reconstructed.

Keywords:
Ilizarov external fixator, limb reconstruction, tibial hemimelia

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Introduction

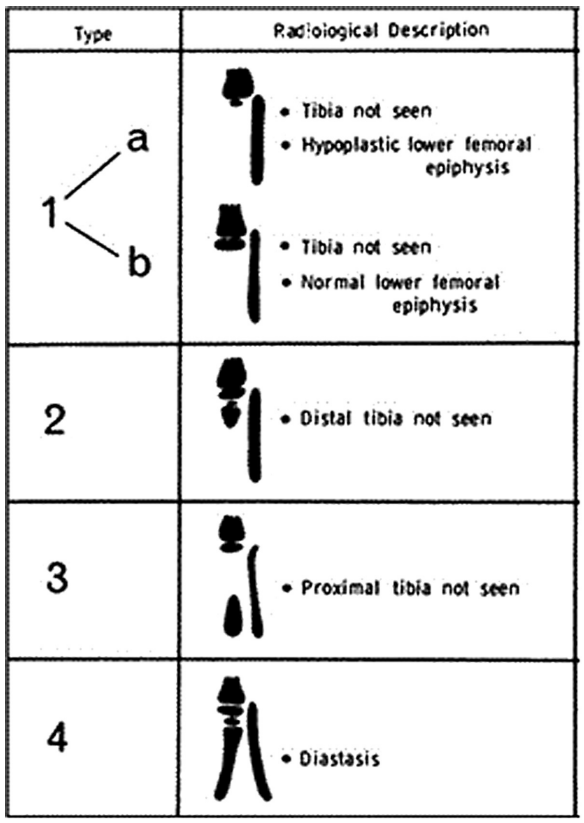
Tibial hemimelia is a congenital condition with a rare incidence of ~1/1 million live births and is usually accompanied by congenital abnormalities in other parts of the body [1]. This preaxial longitudinal deficiency is characterized by the absence of the tibia in variable degrees. There is usually an intact fibula with complete absence or dysplasia of the tibia with marked shortening [2–4]. Knee flexion contracture, instability of the ankle joint, dimpling over the proximal region of the tibia, and foot deformities in the form of rigid varus, supination deformity, and shortening of the first metatarsal ray are common findings in cases of tibial hemimelia [5]. Tibial hemimelia is classified into four types based on the initial radiograph according to Jones *et al.* [6]. In type II, the proximal part of the tibia is present with absence of the distal part, a relative function of the knee joint, and the fibular size is usually normal, with proximal dislocation of the head (Fig. 1).

Early amputation in the knee joint and prosthetic fitting is the standard treatment [1,7–11]. In our Egyptian community, the option of amputation is usually refused and the patient families wish to reconstruct the limb. In contrast to amputation which usually requires one surgical procedure, the reconstructive surgery requires multiple surgical procedures including fibular tibialization, foot

repositioning under the fibula, and lower limb lengthening [1,10,12,13].

The purpose of this study is to determine the results of limb reconstruction in eight children with Jones type II tibial hemimelia by fibular tibialization, ankle fusion, and lower limb lengthening using Ilizarov external fixator.

Figure 1



Jones *et al.* [6] classification of tibial hemimelia.

Patients and methods

This retrospective study was carried out at Zagazig University hospitals from the year 2007 to 2014 on eight children with Jones type II tibial hemimelia treated by fibular tibialization, ankle fusion, and lower limb lengthening by Ilizarov external fixator. Ethical permission for this study was obtained from Zagazig University hospitals, and informed consent was obtained from all guardians before participation in the study. The age of children at the time of reconstruction ranged from 2 to 5 years (average 3 years). There were five girls and three boys. All had unilateral affection; right side was affected in five children and left in three.

All cases had leg-length discrepancy that ranged from 5 to 7 cm. All feet had equinovarus deformity with absence of the medial rays (1–2 rays) in 5 feet. Associated congenital anomalies were present in three cases in the form of a cleft palate in one case, a hand polydactyl in one case, and a contralateral club foot in one case (Table 1).

Operative techniques

Proximal tibiofibular synostosis

Tibialization of distal fibula was performed at the age of presentation. The mean age of the cases at first operation was 3 years. Under general anesthesia with patient in supine position, an above-knee tourniquet was applied. The skin incision begins at the anterolateral aspect of the proximal tibia and ends distally anterior to the middle third of the tibia. The proximal tibial cartilaginous anlage or the bony proximal tibia is exposed by dissection of the muscle of the anterior compartment from the proximal tibia. The atrophic part of the tibia was dissected, and a

Table 1 Patients' data

Case number	Sex	Side	Age at first surgery	Associated anomaly	Maximal flexion (°)	Initial LLD (cm)	Total lengthening (cm)	LLD at final follow-up (cm)	Fixator time (day)	Healing index (days/cm)	Calcaneofibular angle (°)	(follow-up (years)
1	F	RT	2		90	7	5	2	160	32	110	5
2	M	RT	3	Cleft palate	100	6.5	6.5	0	150	25	100	2
3	F	LT	2		110	6	6	0	145	24.16	100	3
4	F	RT	2		90	5	5	0	170	34	110	4
5	M	LT	2.5	Hand polydactyl	90	6	6	0	140	25.45	100	2
6	F	RT	5		110	5.5	5.5	0	135	24.54	90	3
7	M	RT	3	Contralateral club foot	100	5	5	0	140	28	100	2
8	F	LT	5		110	7	5	2	160	40	90	3
Mean			3.06		100	6	5.5	0.5	150	29.14	100	3

F, female; LLD, Leg-length discrepancy; LT, left; M, male; RT, right.

tunnel is created centrally in the medulla at the inferior margin of the bone.

The proximal fibular attachments were left intact while subperiosteal dissection of the fibula was performed. Osteotomy of the fibula was performed at a point opposite the distal end of the proximal tibial anlage and it was transferred in front of the interosseous membrane, behind the anterolateral group of muscles, to be in contact with the cut end of the tibia, leaving the fibular collateral ligament, the fibular head, the proximal physis, and the periosteal sleeve intact. Fixation of the two bones was done by two K-wires of appropriate size.

Ankle fusion

Foot repositioning consisted of a posteromedial soft tissue release by incision from the anterolateral side of the ankle to the posteromedial side. Tapering of the fibular distal end with protection of the fibular physis and a trough in the talus was performed. The fibular distal end was placed into the trough in the talus and fixed by a transcalcaneal K-wire. A long leg cast was applied for 2–3 months until the bone ends were united to each other. All the cases were united in 2–3 months.

Equalization of limb-length discrepancy

The transferred fibula becomes well developed into a tibia like bone with follow-up. At this stage, corticotomy of the tibialized fibula was performed, and Ilizarov apparatus for leg lengthening was applied. Correction of the residual foot deformity by gradual distraction without osteotomy or soft tissue release was done at the same time.

The Ilizarov preassembled apparatus consisting of two equal size rings was applied to the leg, and the proximal ring was applied one finger breadth distal to the tibial tuberosity. The distal ring was applied to the tibialized fibula. When progressive flexion knee deformity is expected during full correction of leg-length discrepancy in cases with limited preoperative extension, addition of distal femoral ring was performed. In one case, the proximal tibial ring was connected to the distal femoral ring by hinges at the level of the knee to promote knee joint motion. Then, low-energy corticotomy was done, and additional two half rings were applied to the foot, one in the calcaneus and the other in the forefoot connected to the lowest ring of the lengthening apparatus with the use of hinges for correction-associated calcaneovarus deformity. Postoperatively, gradual distraction was delayed for 5–7 days. The distraction was done at a rate of 1 mm per day. Follow-up of the distraction site for new bone formation was regularly checked till full consolidation takes place.

Results

The mean follow-up period was 3 years (range, 2–5 years). In all cases, the transferred fibular shaft to the distal end of the tibial remnant was united within 2–3 months. The fibula grew in length and size to be tibialized. Full correction of the limb-length discrepancy was achieved in six cases, and residual shortening was still present in two cases (2 cm) at a mean follow-up of 3 years. This leg-length discrepancy may increase with time, and our patients may need further lengthening procedures. Consolidation of the bone at the distraction site was obtained with a gained length of 5–6.5 cm in all patients. The external fixator was applied for an average time of 150 days, and the healing index was 29 days/cm. Regarding the knee function, the mean maximum knee flexion was 100° (range, 90°–110°), and instability of less than 5° in varus valgus plane was seen in two cases. The ankle and foot deformity improved by ankle fusion; the mean calcaneofibular angle was 100° (range, 90°–110°). Good results were obtained in eight patients, and all patients could walk independently without pain. All the parents of the patients were satisfied.

Complications

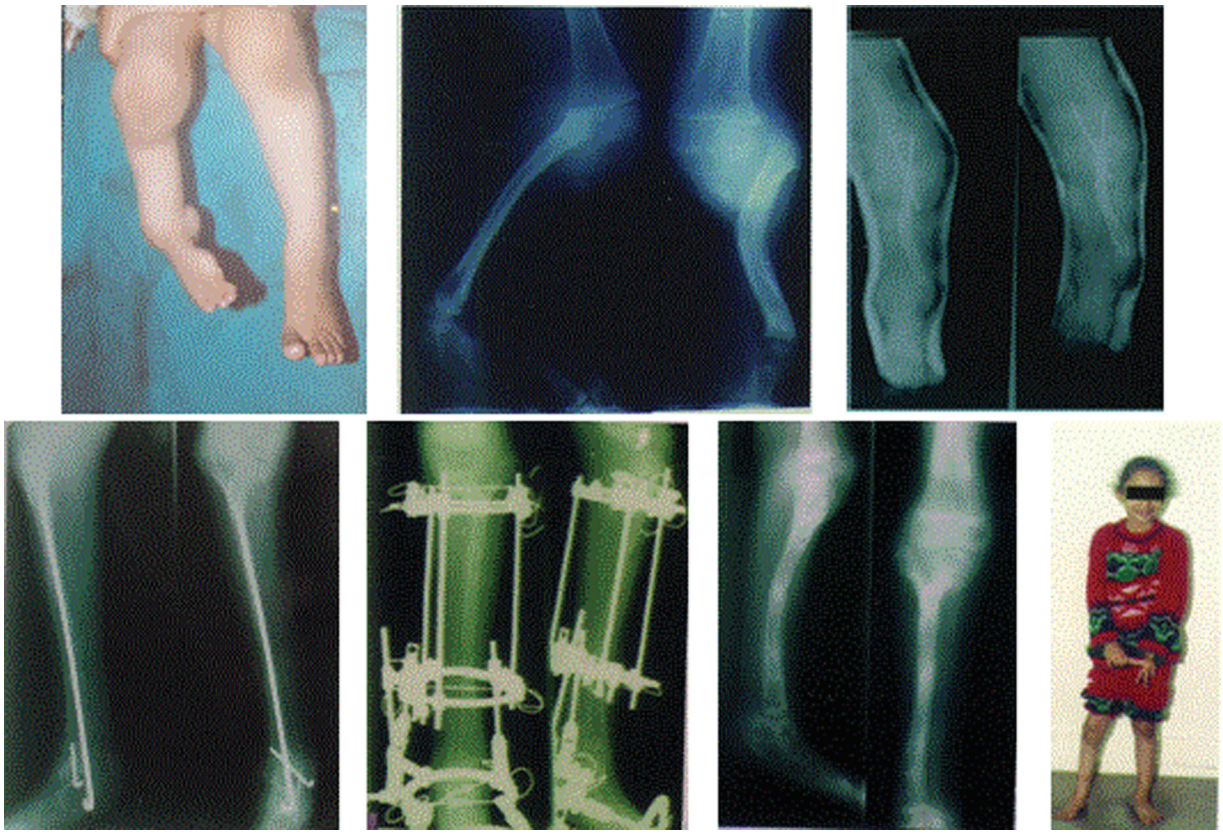
Pin-tract infection was encountered in all patients and was managed by local wound care and oral antibiotics. Knee flexion contracture of 10°–20° occurred during tibial lengthening in all patients, and after removal of the external fixator, this complication improved completely without affecting the final results. Fracture of the fibula at the site of the regenerate after removal of the fixator occurred in one case and was treated by above-knee cast (Figs 2 and 3).

Discussion

The most commonly used procedure for management of tibial hemimelia is amputation and prosthetic fitting [4–10]. In patients with Jones type II deformity, hamstring and quadriceps function is normal and the knee moves normally. The fibular head will be displaced proximally and laterally, and the limb will be in a varus position with significant varus instability. At the ankle joint, the foot is displaced medially relative to the fibula and will also be in varus [12].

Amputation is usually refused in our country, and with reconstructive procedures, amputation of the limb may be avoided, but it needs multiple surgical operations.

Figure 2

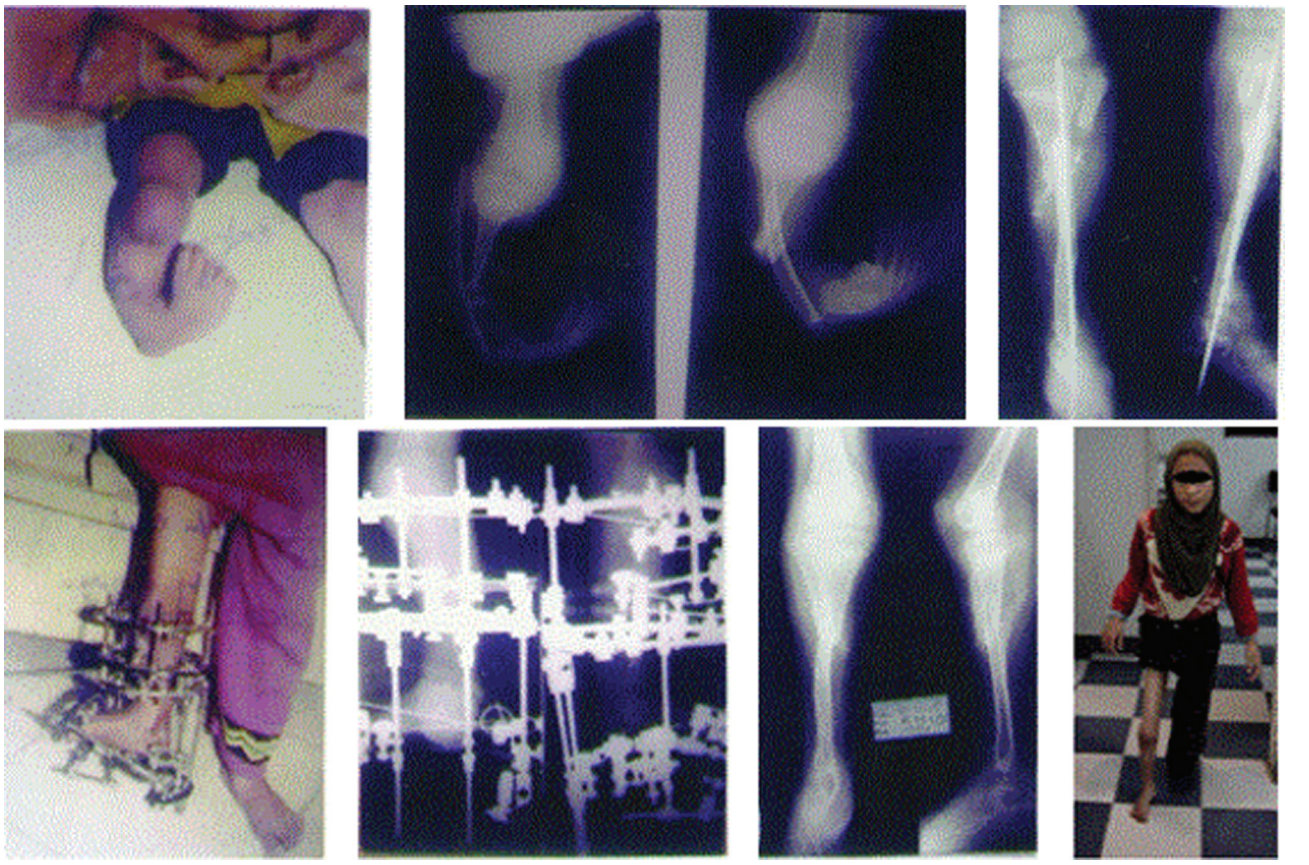


Case No. 4. (a) Preoperative photograph showing right foot equinovarus and right limb shortening. (b) Preoperative radiograph of the same patient. (c) Radiograph after tibiofibular fusion. (d) Radiograph after second stage for ankle fusion. (e) Radiograph after application of the Ilizarov apparatus for tibial lengthening and correction of the foot deformity. (f) Radiograph at the latest follow-up revealed correction of the leg and ankle deformities and bone consolidation of the distraction site. (g) Photograph of the patient at the latest follow-up (4 years) with full correction of the limb deformity and a plantigrade foot.

Kalamchi and Dawe [5] advised that the ideal management for the type II congenital deficiency of the tibia is by tibialization of the fibula by side-to-side technique to give stability to the knee joint and for the leg-length discrepancy, and for the foot deformity is to do modified Boyd amputation, implanting the distal fibula within the body of the os calcis. He also felt that below-knee amputation should be avoided as it may lead to overwhelming difficulties with bony overgrowth and recurrent skin damage at the end of the stump. In the present study, we treated eight patients with type II tibial hemimelia by tibiofibular fusion between the ends of the fibula and tibial remnant with preservation of the proximal fibula to maintain knee stability and form a satisfactory knee joint. The mean range of motion of the knee obtained was 100° (range, 90°–110°) with two cases of instability of less than 5° in varus valgus plane owing to the absence of the central pivot of the knee, ankle fusion, and Ilizarov distraction for correction of the leg-length discrepancy and the residual foot and ankle deformity. According to the study by Wada *et al.* [14], the equinus deformity was determined when calcaneofibular angle was greater than 110° and a talipes calcaneal deformity was determined when the calcaneofibular angle was less than 80°. In the current study, equinovarus foot deformities in the plantigrade feet were corrected by a posteromedial soft tissue release and talofibular arthrodesis, with a mean calcaneofibular angle of 100° (range, 90°–110°).

Limb-length discrepancy was corrected in all patients except in two patients in which there was 2-cm shortening at a mean follow-up of 3 years, and with time, the leg-length discrepancy may increase, and our patients may need further lengthening procedures. Although the healing index of the tibialized fibula in the current study (29 days/cm) differed from some published studies [13,15], as they found that the fibula had delayed new bone formation with healing index of 34.4 days/cm, the findings of the current study are consistent with those of the study by Hosny [16], which treated six cases (four type II and two cases type I a) and obtained good outcome with patients' satisfaction in all cases. Moreover, Wada *et al.* [17] presented a series of nine cases (four type II and five

Figure 3



Case No 6. (a) Preoperative photograph showing right foot equinovarus and right limb shortening. (b) Preoperative radiograph of the same patient. (c) Radiograph of the patient after tibiofibular fusion and fusion of the ankle. (d) Photograph of the patient after application of the Ilizarov apparatus for tibial lengthening. (e) Radiograph of the patient after application of the Ilizarov apparatus for tibial lengthening. (f) Radiograph at the latest follow-up revealed correction of the leg and ankle deformities and bone consolidation of the distraction site. (g) Photograph of the patient at the latest follow-up (3 years) with full correction of the limb deformity and a plantigrade foot.

type I); they applied the same surgical procedures done in this study for limb reconstruction in type II, and they reported that they achieved a satisfactory functional and cosmetic result in all cases, and in cases with type I, none of the five knees managed by fibular transfer obtained a satisfactory functional outcome. Eamsobhana and Kaewpornsawan [18] presented a series of six cases of tibial hemimelia. They obtained a satisfactory functional and cosmetic result in all cases with type II, whereas in patients with type I, none of the three patients managed by fibular transfer obtained a satisfactory functional outcome. Carranza-Bencano and González-Rodríguez [19] presented a case report of a 15-year-old girl with type II tibial hemimelia managed by talofibular fusion and distraction by Ilizarov external fixator, and they obtained a satisfactory outcome.

In our series, there were many complications. Superficial pin-tract infection was encountered in 100% of patients; all improved by local wound care and oral antibiotics. Knee flexion contracture of 10°–20° occurred during tibial lengthening in all patients, and after removal of the external fixator, this complication improved completely without affecting the final results. Fracture of the fibula at the site of the regenerate after removal of the fixator occurred in one case and was treated by above-knee cast. These are comparable to other studies. Wada *et al.* [17] reported on superficial pin-tract infection in all cases, which was managed by antibiotics. Hosny [16] reported that pin-tract infection occurred in all his patients and was managed by local or parenteral antibiotics and drainage under local anesthesia in two cases, cutting through calcaneal wire, replacement of this wire, and extraction of an infected wire in two cases. Knee flexion deformity (5°) persisted in two cases.

Conclusion

The results of limb reconstruction of the Jones type II tibial hemimelia by fibular tibialization, ankle fusion, and lower limb lengthening using Ilizarov external fixator were acceptable. The limb was saved and reconstructed.

Financial support and sponsorship
Nil.

Conflicts of interest
There are no conflicts of interest.

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