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Leg lengthening and deformity correction in fibular hemimelia using ring external fixator

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***Abstract*---**Background: The treatment of fibular hemimelia aims to achieve a functional limb nearly equal in length to the normal limb at maturity, with a plantigrade, stable and flexible foot. The aim of this work was to evaluate the functional results of reconstructive lengthening and deformity correction in fibular hemimelia using the ring external fixator. Methods: This observational study was carried out on 41 patients with fibular hemimelia. The cases either prospectively gathered (sixteen cases, 39 %) to start their management plan or retrospectively collected (twenty-five cases, 61 %) and followed up. Foot reconstruction surgery was done in 22 cases while lengthening surgery applied all cases. All patients were subjected to: history taking, clinical examination (local (lower limb), foot and knee), general examination and imaging. Results: regarding overall results, at the end of follow up with an average period of 2 years, 4 patients had excellent results (9.76%), 21 patients (51.2%) had good results, 12 patients (29.28%) had fair results and 4 patients (9.76%) had poor result. Regarding subjective functional assessment, out of 41 patients, 20 of them (48.8%) were from 75 to 100% subjectively satisfied, 11 patients (26.8%) from 50 to 74% and 10 patients (24.4%) from 0 to 49% satisfied. Regarding complications, 2 patients (4.9%) showed no

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complications, pin tract infection occurred in 35 (85.4%), knee joint posterior subluxation occurred in 11 (26.8%), weakness of the regenerate occurred in 9 (22%), translation of the distal segment 2 (4.9%), recurrence appeared in 1 (2.4%), ankle valgus recurrence 15 (36.6%), Patients did not have complication or had pin tract infection only 21 (51.2%), present 20 (48.8%). Conclusions: Limb lengthening and deformity correction using Ilizarov external fixator are effective methods in the management of foot deformities and LLD associated with FH. These reconstructive surgeries yield good subjective and objective functional results, patients and parents' satisfaction.

***Keywords*---**leg lengthening, deformity correction, fibular hemimelia, ring external fixator.

Introduction

Word hemimelial arises from the Greek name —hemi – melos — which means half of a limb [1].Fibular Hemimelia (FH) means partial or complete aplasia (deficiency) of the fibula. It is considered to be the commonest of all congenital longitudinal deficiency of the limb comprising an incidence ranging from 8 to 20 per million of all live births [2].FH comprises a wide range of anomalies and deformities that primarily affect the femoral and tibial bones and also the knee and ankle joints without neglecting its effect on the foot and its joints. The primary clinical presentations of FH are shortening of the ipsilateral femur, anteromedial tibial bowing, genu valgum, equinovalgus deformation in addition to absence of the lateral toes of the foot [3, 4].

The etiology of FH still unknown, and in most cases, it is a sporadic condition, with most of children born with this condition having no family history of other birth defects. Bilatera FH or multiple limb defects can be assumed to be due to autosomal dominant gene disorder (inherited or new mutation) or attributed to teratologic agent as drug or radiation [5].Numerous systems for classification of FH have been proposed. The most known classifications are the Achterman-Kalamchi and Birch [6, 7]. Achterman and Kalamchi classified FH into two types, type 1 in which there is partial absence of the fibula and type 2 in which there is complete absence of the fibula. Over time, the importance of the reconstructability of the foot has raised rapidly, leading into the classification system by Paley which makes treatment recommendations based on foot deformity [8, 9]. The treatment of fibular hemimelia aims to achieve a functional limb nearly equal in length to the normal limb at maturity, with a plantigrade, stable and flexible foot. The treatment of fibular hemimelia is highly dependent on the potential of the foot to be reconstructed and should be held primarily in mind when deciding on amputation versus limb salvage. Treatment of fibular hemimelia should be highly individualized to the patient [10].

As amputation is not recommended socially in our country (due to psychological troubles, loss of the proprioception, periodic prosthetic change) we conducted this research work to evaluate the methods of limb reconstruction regarding deformity correction and lengthening in cases of limb length discrepancy. Also, correction

and reconstruction of the feet to stable, functional and plantigrade ones. Knee deformities and instabilities was attacked by osteotomies, guided growth and ligament reconstruction to obtain stable knees with maximal range of motion. We judged our work regarding the functional and radiological results, patient and his parents' satisfaction. The aim of this work was to evaluate the functional results of reconstructive lengthening and deformity correction in fibular hemimelia using the ring external fixator.

Patients and Methods

This observational study was carried out on 41 patients aged 2 or more years old, both sexes, Achterman and Kalamchi type IA fibular hemimelia, Achterman and Kalamchi types 1B and II fibular hemimelia, and unstable ankle/foot. The cases either prospectively gathered (sixteen cases, 39 %) to start their management plan or retrospectively collected (twenty-five cases, 61 %) and followed up. Foot reconstruction surgery was done in 22 cases while lengthening surgery applied in the remaining cases. The study was conducted at Limb reconstruction and pediatric orthopedic unit of Tanta University Hospital and the time of the study was 2 years from May 2019 to May 2021.

An informed written consent was obtained from the patient or relatives of the patients. The study was done after approval from the Ethical Committee Tanta University Hospitals. Exclusion criteria were parent refusal to do surgical intervention and any comorbidities that endanger surgical intervention.All patients were subjected to: History taking, clinical examination (local (lower limb (the side affected, absence or presence of the lateral malleolus, tibial bowing, the ankle joint), foot, knee and femur).

General Examination

Examination of the other limb was performed to detect bilaterality
Examination of the upper limb was done to exclude associated upper limb Deficiency

Imaging

Plain X-ray films (AP, lateral, oblique views to detect degree of fibular hypoplasia, degree of anteromedial tibial bowing: CORA, talocrural angle, tarsal coalition and mechanical axis alignment. The metaphyseal-epiphyseal angle: For cases with complete deficiency, were the angle between a line bisecting the distal tibial metaphysis and a line joining the tip of the medial malleolus to the most lateral part of the articular surface of the distal tibia was measured

Assessment of limb length discrepancy

- Clinically: tape method was used to determine the length of the femur (from the anterior superior iliac spine (ASIS) to knee joint line), length of the tibia (from the knee joint line to the medial malleolus) and the overall length of the limb (from the ASIS to the medial malleolous), and the standing on block method to determine the overall limb length discrepancy.

- Radiologically: by using full length standing X ray scanogram on both lower limbs with both patellae facing forwards and putting the short limb on a block.

At the end of assessment, a decision was made as to whether the shortening was predominantly tibial, femoral or both.

Prediction of LLD

This was performed using Paley's Multiplier Method of LLD[11] assessment where the predicted LLD at skeletal maturity equaled the current LLD multiplied by a specific multiplier for the age and sex of the patient. The Multiplied application is now available on App Store and can be easily used.

The Management Plan included 3 stages

- Foot reconstruction surgery to achieve plantigrade stable shoeable foot (22 cases, 53.6 %)
- Lengthening surgery: using the Ilizarov method and corticotomy to achieve limb equalization (41 cases, 100 %)
- Corrective osteotomy for correction of residual deformity (4 cases, 9.75%)
- A fourth stage was planned for s which need second dose of lengthening.

Surgical techniques

All surgeries were performed under general anesthesia. After limb elevation for 4 minutes a pneumatic tourniquet was applied to the thigh and inflated. The patient was positioned laterally with the affected side up. Lateral skin incision was utilized midway between the chin of the tibia and TendoAhilles. Subcutaneous dissection was performed with careful protection of the sural nerve and short saphenous vein identification and protection of the sural nerve. TendoAchiliis was identified and lengthened by Z plasty. The deep fascia over the lateral compartment was opened and peroneal muscles were identified and lengthened. The intermuscular septum separating the lateral and posterior compartments (and attached to the fibular anlage) was opened. Now the fibular anlage was fully dissected and cut as proximally as possible and distally followed to the ankle.
- The interosseus membrane was opened.

Supramalleolar osteotomy

An opening wedge osteotomy was performed 1 cm above the physeal plate and was opened posteriorly (to correct the equinus) and laterally (to correct the valgus). The osteotomy was kept open by being fixed with two Kirshner wires introduced from the heel through the ankle and the osteotomy site to the proximal tibia. The two ends of the TendoAchilles were sutured. An above knee posterior slab was placed to allow for postoperative swelling. - One week later, the wound was checked, and the slab was changed into a cast. At four weeks, the K-wires were removed and at six weeks, the cast was removed, and the child was allowed to bear weight (if the child was a walker).

- Lengthening surgery
  Before performing the lengthening surgery, the foot should be reconstructed as discussed before or reconstructed at the time of lengthening surgery. Also, the exact LLD at the time of surgery should be reevaluated and the following rules should be followed up.
- Osteotomy
  Osteotomy was performed through small incision and minimal periosteal stripping. A 3.5 drill pit was inserted through the incision under fluoroscopic imaging to determine the level of the osteotomy. Completion of the osteotomy was performed by an osteotome.

Frame application

- The standard Ilizarov frame was used for all patients. Preassembly of the frame was done prior to surgery to decrease the intraoperative time. Either 2 or 3 rings were applied to the tibia. The first ring in the proximal metaphysis proximal to the osteotomy site and the other ring(s) distal to the osteotomy site. Each ring was stabilized by three items either 2 wires and one Shanz screw or one wire and two Shanz screw. The frame was applied parallel to the axis of the tibia in both AP and lateral planes.

Frame extension

Extension of the frame across the ankle to include the foot were done in 36 cases (87.8%) with one half ring or foot piece that was stabilized by either calcaneal Shanz screw and wires or only olive wires from medial and lateral side

Extension of the frame to the femur across the knee joint:

- When simultaneous femoral lengthening was required as in cases of associated CFD This can be done by applying a complete ring fixed by wire and posteromedial and posterolateral Shanz screws distal to the osteotomy site and another ring proximal to the osteotomy site and more proximal arch that were fixed by Shanz screws only. the frame was connected by oblique supports.
- When more than 5 cm of tibial lengthening was planned the frame should be extended to include the femur from the start for fear of knee subluxation with lengthening (17 cases 41.4%). This was achieved by application of only one ring above the knee.

In these cases, hinges were applied across the knee to allow for knee motion

Distraction phase

- The patient remained in the hospital post operatively for five days to teach the parents how to care about the frame to avoid pin tract infections.
- After a latent period of 7 days distraction at the osteotomy site was begun.
- The rate of distraction was complete circle at the count nut per day which equaled 1mm per day. (in cases of femoral and tibial osteotomies 0.5 mm was carried out for lengthening of for each bone)

Follow up of the patient during distraction phase:

- The patient was seen on weekly basis to ensure proper lengthening and check for any complications, obstacles or problems that may occur during lengthening.

- The frame was checked for stability and proper cleanliness and care
- Serial X-rays every 2 weeks were obtained to check the quality and length of the regenerate.
- If knee subluxation was noted during distraction then the patient was transferred to the operation room to extend the frame above the knee to reduce and prevent further subluxation. (6 cases 14.6 %)
- Lengthening was terminated when the ultimate lengthening goal was achieved or knee subluxation occurred despite o fame extension.

Follow-up through the consolidation phase:
- At the end of the lengthening phase, the knee frame was removed to allow for knee ROM. This was done with cautious manipulation of the knee under general anaesthesia at the time of knee frame removal.
Physiotherapy for the knee was then encouraged. The patients were seen every two weeks and monthly X-rays were obtained to check for maturation of the callus. Any problems, obstacles or complications were noted.

Frame removal

- Criteria of removal of the Ilizarov frame included visualization of at least three cortices in the AP and lateral plain films.
- As general rule consolidation requires the same time for distraction phase. For example, if distraction phase took one and half months then the frame should be retained for another one and half months. i.e. total period of frame application equaled three months
- The frame was removed under general anaesthesia and above knee cast was applied and retained for six weeks and the patient was allowed to bear weight.
-

Patients follow up

The patients were seen every three months in the first year then annually. Follow up visits focused on the following clinical criteria:

- The length measurements (by tape) and the LLD (standing on blocks).
- The lower limb mechanical axis alignment (with particular focus on the detection of valgus drift).
- The ankle position (the ankle valgus angle and ROM).
- The knee ROM.
- The physical activity and degree of patient participation in daily activities or sports.
- The patient and parents' satisfaction.

Methods for evaluation of the results

The results of lengthening were assessed 3 months after removal of the cast with particular focus on:

- The length measurements: the tibia, the femur, the overall limb length measured clinically by the tape method and the residual LLD measured by the standing on block method.
- The mechanical axis alignment: particularly genu valgum angle, or medial tibial bowing (lateral tilt) of the lengthened tibia.
- The knee ROM: Measured in degrees from full knee extension (0°) to full knee flexion.
- The ankle position after lengthening: The ankle valgus angle (angle between the heel bisector line and the distal tibia clinically) and the equinus angle (degree of dorsiflexion range of the ankle).
- The recorded complications

Statistical analysis

Data were fed to the computer and analysed using IBM SPSS software package version 20.0. (Armonk, NY: IBM Corp) Qualitative data were described using number and percent. The Kolmogorov-Smirnov test was used to verify the normality of distribution Quantitative data were described using range (minimum and maximum), mean, standard deviation, median and interquartile range (IQR). Significance of the obtained results was judged at the 5% level. The used tests were: Chi-square test for categorical variables, to compare between different groups. Monte Carlo correction for chi-square when more than 20% of the cells have expected count less than 5. Pearson coefficient to correlate between two normally distributed quantitative variables. Student t-test for normally distributed quantitative variables, to compare between two studied groups. Wilcoxon signed ranks test for abnormally distributed quantitative variables, to compare between two periods.

Results

Error! Not a valid bookmark self-reference.1 shows patient age, sex, consanguinity, side affection, deformity, Aschermann classification, Paley classification, unstable knee, foot rays, LLD, LLD maturity range and previous surgeries of the studied patients.

Table 1: Age, sex, consanguinity, side affection, deformity, Aschermann classification, Paley classification, unstable knee, foot rays, LLD, LLD maturity range and previous surgeries of the studied patients (n = 41)

		Patients (n = 41)
Age (years)	3 – 6.9 years	18(43.9%)
	7- 10 years	15(36.6%)
	More than 10 years	8(19.5%)
Sex	Male	26 (63%)
	Female	15 (37%)
Consanguinity	+ve consanguinity	7(17%)
	-ve consanguinity	34(83%)
Side affection	Right side	31(75%)
	Left side	10(25%)

Deformity	CFD	16(39%)
	Ankle deformities	29(70%)
Aschermann classification	IA	7(17%)
	IB	11(27%)
	II	23(56%)
Paley classification	I	7(17%)
	II	15(36.5%)
	III	19(46.5%)
Unstable knee		17(41.5%)
Foot rays	5 rays	10(24%)
	4 rays	20(48.7%)
	3 rays	11(26.3%)
LLD	3 – 6 cm	26(63.4%)
	6.1 – 10 cm	12(29.2%)
	10.1 – 15 cm	3(7.4%)
LLD maturity range	3 - 6 cm	9(21.9%)
	6.1 – 10 cm	21(51.2%)
	More than 10.1 cm	11(26.6%)
Previous surgeries	Foot	22(53.3%)
	Previous 1 lengthening	10(24.4%)
	Previous 2 lengthening	2(4.8%)
	No	7(17.5%)

Data are presented as frequency (%). LLD: Limb length discrepancy, CFD: Congenital femoral deficiency.

Table 2 shows duration of the Ilizarov during lengthening, lengthened segments, site of tibial osteotomy, extension of the Ilizarov frame to the femur and complications of the studied cases. Table 2: Duration of the Ilizarov during lengthening, lengthened segments, site of tibial osteotomy, extension of the Ilizarov frame to the femur and complications of the studied cases

		N= 41
Duration of the Ilizarov during lengthening		5.5 months
Lengthened segments	Tibia	26(63.4%)
	Femur	2(4.8%)
	Combined	13(31.8%)
Site of tibial osteotomy	Diaphyseal	5(12.8%)
	Metaphyseal	31(79.5%)
	Combined	3(7.7%)
Extension of the Ilizarov frame to the femur	Extension forms the start	17(41.4%)
	Extension during the course of lengthening	6(14.6%)
	No extension at all	18(44%)

Data are presented as frequency (%) or mean. #: Patients do not have complication or have pin tract infection only.

Table 3 shows the overall results, type of fibular anlage (intraoperative), talus (intraoperative), postoperative ankle stability, wound necrosis, postoperative LLD in cm, length gain, and postoperative improvement of the ankle valgus, postoperative limbs with no LLD and subjective functional assessment of the studied cases.

Table 3: Overall results, type of fibular anlage (intraoperative), talus (intraoperative), postoperative ankle stability, wound necrosis, postoperative LLD in cm, length gain, and postoperative improvement of the ankle valgus, postoperative limbs with no LLD and subjective functional assessment of the studied cases

	N= 41	
End results	Excellent	4(9.76%)
	Good	21(51.2%)
	Fair	12(29.28%)
	Poor	4(9.76%)
Type of fibular anlage (intraoperative)	Totally fibrous	26(63.4%)
	Distally cartilaginous	10(24.3%)
	Distally bony	5(12.3%)
Talus (intraoperative)	Present	8(36.3%)
	Absent	14(63.7%)
Postoperative ankle stability	Stable	18(81.8%)
	Unstable	4(18.2%)
Wound necrosis	Present	3(13.6%)
	Absent	19(86.4%)
Postoperative LLD in cm	3.625 cm 0.5 – 6.75 cm	
Length gain	Tibial	26(63.4%)
	Femoral	2(4.9%)
	Combined	13(31.7%)
Postoperative improvement of the ankle valgus	Preoperative number of limbs with normal angle (0≤5°)	0(0%)
	Postoperative number of limbs with normal angle (0≤5°)	18(81.8%)
	Difference	(81.8%)
LLD	Postoperative Limbs with LLD	6(14.6%)
	Postoperative Limbs with no LLD	35(85.3%)

Subjective functional assessment	75-100%	20(48.8%)
	50-75%	11(26.8%)
	0-49%	10(24.4%)

Data are presented as frequency (%) or mean (range). LLD: Limb length discrepancy.

LLD was significantly lower Postoperatively compared to preoperative. Table 4

Table 4: Comparison between preoperative and postoperative LLD

LLD	Preoperative	Postoperative	Z	p
Min. – Max.	3.0 – 15.0	0.0 – 6.0		
Mean ± SD.	5.98 ± 2.70	0.93 ± 1.18		
Median (IQR)	5.50 (4.0 – 7.0)	0.50 (0.50 – 1.0)	5.586*	<0.001*

LLD: Limb length discrepancy. *: statistically significant p value <0.05.
Table 5 shows distribution of the studied cases according to the incidence of different complications.

Table 5: Distribution of the studied cases according to the incidence of different complications

Complications	No complications	2(4.9%)
	Pin tract infection	35(85.4%)
	Knee joint posterior subluxation	11(26.8%)
	Weakness of the regenerate	9(22%)
	Translation of the distal segment	2(4.9%)
	Recurrence	1(2.4%)
	Ankle valgus recurrence	15(36.6%)
	Absent#	21(51.2%)
	Present	20(48.8%)

Data are presented as frequency (%), #: Patients do not have complication or have Pin tract infection only.

The affected side, gender and number of rays has insignificant effect on the final results. No bilateral cases were included so the effect of bilaterality on the final results could not be assessed. The type of FH according to Achterman and Kalamchi had statistically significant effects on the final results. Better results were achieved in milder forms of fibular hemimelia (type IA better than IB better than type II). As one of the cases was unclassified according to Paley system of FH classification then the total number of case series among this classification was 40 cases. Where the P value less than 0.05 then statistically significant results were achieved. So the less grade of FH, the better final results were achieved. The associated CFD showed statistically significant negative effect on the final results. Better results were achieved in cases with tibial lengthening alone than femoral

lengthening alone than simultaneous combined femoral and tibial lengthening. Table 5

Table 6: Relation between final results and affected side, sex, type of FH (Achterman and Kalamchi, type of FH (Paley), number of foot rays, effect of associated CFD and lengthening

		Final Results				MCp
		Poor (n = 4)	Fair (n = 12)	Good (n = 21)	Excellent (n = 4)	
Affected side	Right	3(75%)	10(83.3%)	15(71.4%)	4(100%)	0.775
	Left	1(25%)	2(16.7%)	6(28.6%)	0 (0%)	
Sex	Male	4(100%)	6(50%)	13(61.9%)	3(75%)	0.446
	Female	0(0%)	6(50%)	(8)38.1	1(25%)	
Type of FH (Achterman and Kalamchi)	IA	0	0	4(19%)	3(100%)	0.006 *
	IB	0	3(25%)	8(38.1%)	0	
	II	4(100%)	9(75%)	9(42.9%)	0	
Type of FH (Paley)	I	0	0	4(19%)	3(100%)	0.007 *
	II	0	5(41.7%)	10(47.6%)	0	
	III	3(75%0	6(50%)	6(28.6%)	0	
	IIIa	1(25%)	0	1(4.8%)	0	
	IIIc	0	1(8.3%)	0	0	
Number of foot rays	3	2(50%0	3(25%)	6(28.6%)	1(25%)	0.239
	4	1(25%)	7(58.3%)	11(52.4%)	0	
	5	1(25%)	2(16.7%)	4(19%)	3(75%)	
Effect of associated CFD	no	3(75%)	4(33.3%)	17(81%)	1(25%)	0.012 *
	yes	1(25%)	8(66.7%)	4(19%)	3(75%)	
Lengthening	Tibial	3(75%)	4(33.3%)	17(81%)	2(50%)	0.002 *
	Femoral	0	0	0	2(50%)	
	Combined	1(25%)	8(66.7%)	19	0	

Data are presented as frequency (%). LLD: Limb length discrepancy, CFD: Congenital femoral deficiency, FH: Fibular Hemimelia *: statistically significant difference p value <0.05.

The quality of bone regenerate was better in cases undergoing metaphyseal osteotomy than those with diaphyseal osteotomy than combined osteotomy. Table 6

Table 7: Relation between quality of bone regenerate and site of tibial osteotomy:

Site of Tibial osteotomy	Quality of bone regenerate			p
	Weak (n = 12)	Poor (n = 4)	Good (n = 23)	
Metaphyseal	10 (83.3%)	2 (50%)	18 (78.3%)	0.380
Diaphyseal	2 (16.7%)	1 (25%)	3 (13%)	
Combined	0	1 (25%)	2 (8.7%)	

Data are presented as frequency (%).

More complications occurred in valgus ankle cases than equinovalgus ankle cases than normal ankle cases. The longer the duration of Ilizarov application the more the complications. Table 7

Table 8: Relation between complications and ankle and duration of ILIzarov (months)

Ankle	Complications		MC_p
	Absent (n = 21)	Present (n = 20)	
Normal	8 (38.1%)	3 (15%)	0.406
Valgus	6 (28.6%)	9 (45%)	
Equinovalgus	6 (28.6%)	7 (35%)	
B & S	1 (4.8%)	1 (5%)	0.081
Duration of ILIzarov (months)	4.10 ± 1.27	4.85 ± 1.42	

Data are presented as frequency (%) or mean ± SD.

There was a significant positive correlation between LLD and duration of Ilizarov application through both lengthening and consolidation stages. (Direct relationship). Figure 1

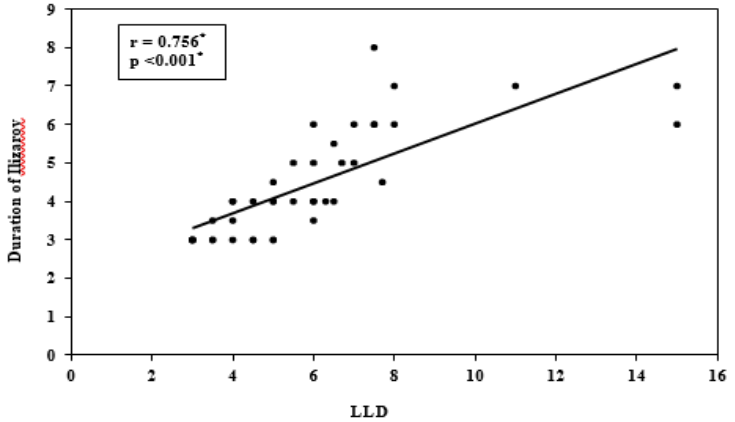


Figure 1: Correlation between LLD and Duration of Ilizarov.

A female patient presented at age 3 years and 9 months, with left sided fibular hemimelia. Type II Actherman and Kalamchi, type I, Paley et al. She had a 5-ray foot, a horizontal mobile ankle, and an unstable knee. A percutaneous Tendo-Achilles tenotomy was done at the age of 1.5 yreas. There were 5˚ of genu valgum, and no anterior tibial angulation. The tibial length discrepancy was 3 cms. The femoral length discrepancy was 3.7 cms, the overall LLD by Block test was 7 cms. This represented a shortening percentage of 15.5%. The predicted LLD at maturity was 12.261 cms. Figure 2

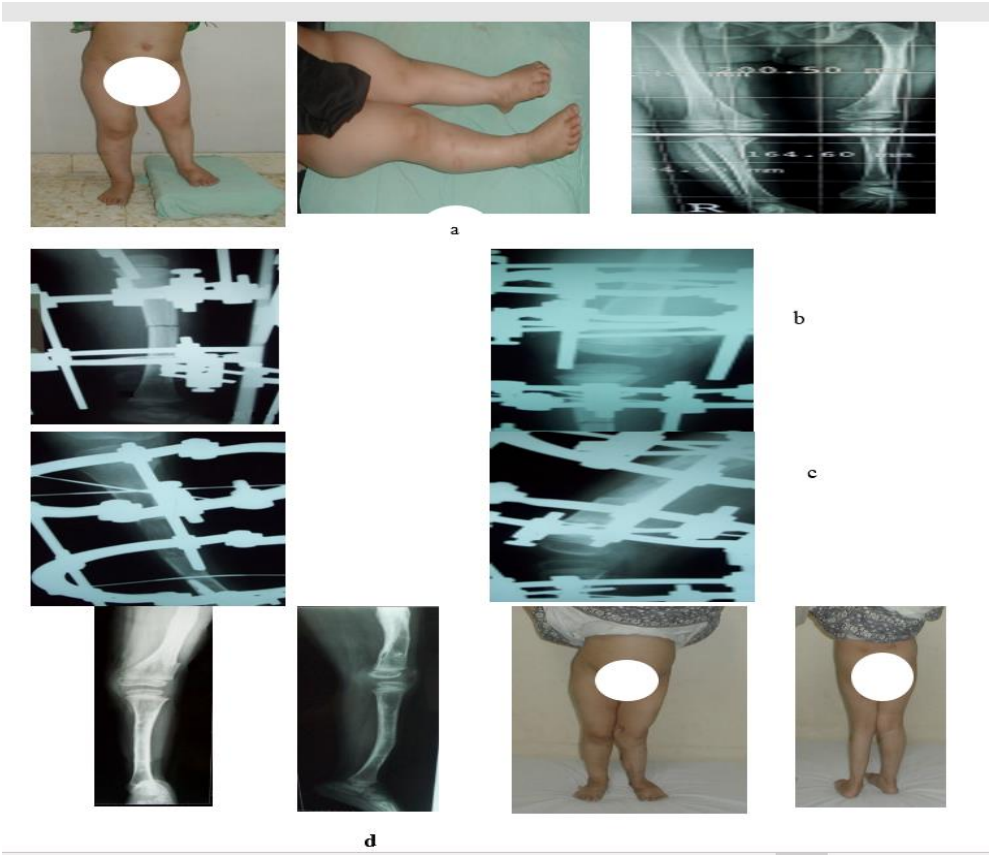
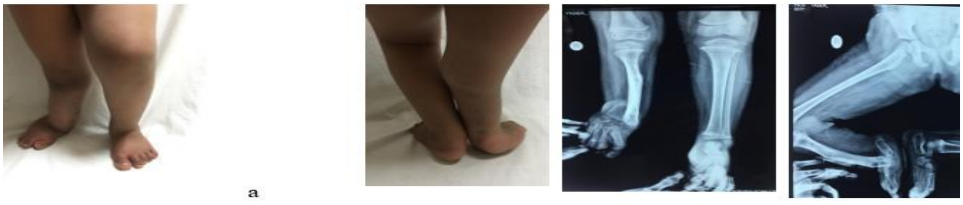


Figure 2: Preoperative clinical photos and scanogram (a), immediate postoperative (b), 2 months postoperative (c) and 4 months postoperative (d)

Female patient 4 years old Rt fibular hemimelia (Achterman II, paley II) 4 rays foot LLD = 3.5 cm LLD at maturity =7 cm . Soft tissue procedures for the foot with TAL + peroneal lengthening was done. Excision of fibular anlagm , supramaleolar osteotomy and correction of valgus and fixation by k wire were performed . Application of the frame Metaphyseal osteotomy for lengthening. Figure 3.



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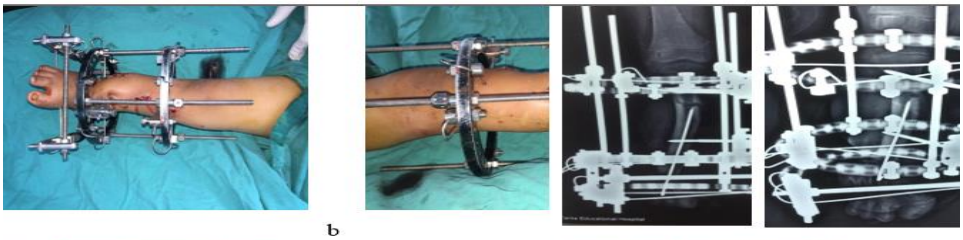

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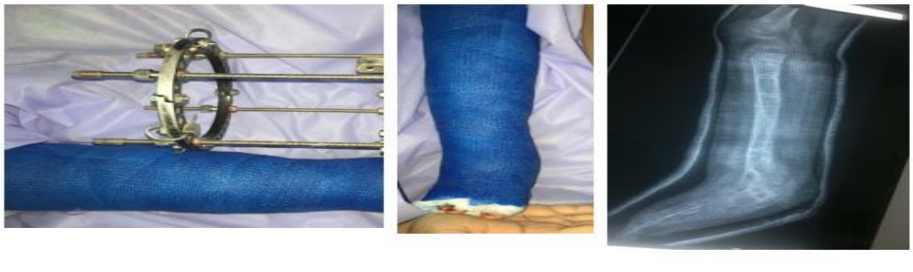

f

Figure 3: Preoprtative clinical photos and x rays (a), immediate postoperative photos (b), 1 month postoperative (c), 2 months postoperative (d), 3 months postoperative(e), removal of the frame and casting(f) , casting for 1 month and one and half year follow up(g)

23 years old female patient presented with type 1A FH, unclassified Paley type (normal ankle and forefoot varus) , Foot rays 3 LLD was 5 cm (tibial origin only) , previous 2 lengthening surgeries at 4 and 9 years . Proximal tibial osteotomy was done and Ilizarov frame was applied and extended by a ring to the femur. Figure 4

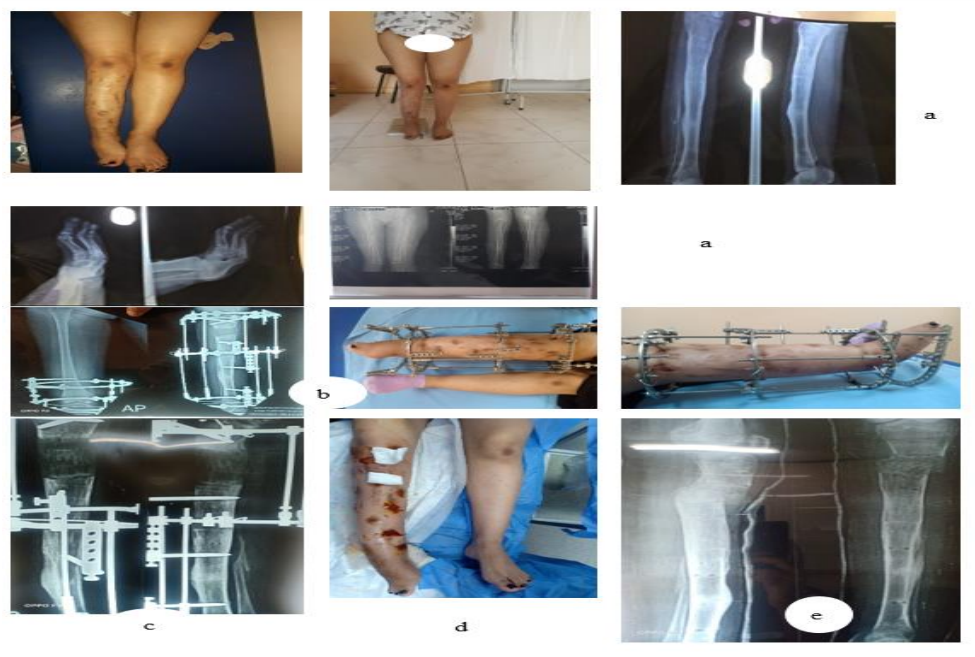

Figure 4: Preoperative clinical photos and x rays and scanogram(a), after 1 month(b), after 5 months (c), removal of the frame with over lengthening of the affected side (d), breaking of the regenerate (e), final equal limbs and good patient satisfaction (casting for two month) (f) and equal limb length (g).

Discussion

Selecting the appropriate and effective method for treating cases with FH is a challenge for orthopaedists. Every case should be tailored according to its conditions and the suitable management of one case may not be necessary suitable for another case.

Discussion of surgical technique

The lateral approach was performed in 16 limbs (51.6%). It allows creation of thick skin flaps with adequate subcutaneous fat and provides the most direct visualization of the peroneal tendons, the fibular anlage, and the lateral tibial surface for performing supramalleolar osteotomy. In this study, the fibular anlage was noted to be completely fibrous in 51.6% of the cases, distally cartilaginous in 35.5% and distally bony in 12.9% .The fibrous type was more associated with less foot rays and more severe deformity.

Gunal et al [12] also identified two types of the fibular anlage by MRI study: Type I: With a well-formed shaped lateral malleolus, cartilaginous distal part and Type II: with a totally fibrous one. They recommended complete excision of the anlage in type II and only partial excision in type I. The argument for preserving the distal cartilaginous part of the anlage is that it contributes to the lateral stabilization of the ankle joint, and it might later on ossify. Arguments for total excision include that the anlage, being a fibrocartilaginous tissue incapable of longitudinal growth, acts as a tethering force, contributing to the ankle deformity, the genu valgum, the anterior tibial bowing and possibly the tibial shortening. Any retained parts of the anlage that are not excised, still do not grow, and retain that tethering effect.

It must be taken into consideration, that though this is a mixed study on both type I and type II fibular hemimelia cases, yet it focuses more on type II cases (56

%), and hence results should be viewed as mainly relating to type II and hence comparable to type II studies (Cheng et al [2], Zarzycki et al [13], Kirienko et al [14]).While the average age of lengthening in this study was 7 years and 6 months, with 39% of cases underwent lengthening before their 5th birthday. In most other studies, the age at lengthening was older. The average age of surgery was 12.5, 7.9, 11.4, 6.5, 10, 7.25, 9.7 years in Choi et al [15], Naudie et al [16], Basbozkurt et al [17], Cheng et al [2], Zarzycki et al [13], Kirienko et al [14], Griffith et al [18] studies respectively.

Foot reconstruction surgery results

As recommended by Paley et al [19], proper restoration of a plantigrade, subtle mobile, stable foot is the most important step for a successful outcome in the salvage management of fibular hemimelia. A limb that has been lengthened to reach the ground yet bears an unstable, deformed, or painful foot is still non-functional, and does not allow proper walking, running or participation in the sports.

Lengthening surgery results

In this study, results of lengthening were assessed by measurement of the length gained in cms, the lengthening indices (including the distraction time – consolidation time – distraction consolidation time – and the healing index), the lengthening complications and post-treatment residuals. Most lengthening surgeries were detected in the same way, as mentioned by Antoci et al [20]

Lengthening Complications

In this study, Paley's scheme [21] for classifying complications in the forms of problems, obstacles, and complications was noted to be very useful and clinically relevant. A complex surgery like lengthening should not be expected to be done without problems or obstacles occurring and being dealt with during the course of the procedure, yet a true complication is, in reality, the lasting problem that affects the final outcome. In this study, the most common problem was pin-tract infection, followed by early contracture of knee, the most frequent obstacle was malalignment, followed by recurrent ankle valgus, followed by subluxation of knee, and the most frequent complication was persistent LLD, followed by late fixed knee flexion deformity. While problems like pin-tract infection and early knee contracture can be considered as an unavoidable event during the procedure, yet every endeavor should be tried to prevent obstacles like malalignment, recurrent ankle valgus, or knee subluxation and certainly complications like fixed knee flexion deformity.

Choi et al [15] recorded lengthening in 11 patients using the Wagner method. There were stress fractures of 4 lengthened femora and 2 lengthened tibia (54%), progressive foot equinovalgus deformity in 5 (45%) cases, delayed union in 4 (36%) cases, pin-tract infection in 4 (36%) cases, malalignment in 2 (18%), wound hematoma in only 1 (9%) case, hip subluxation in 1 (9%), and knee subluxation in 1 (9%).

Thus, the recommendation of this study to extend the frame across the knee and lock it in full extension during the distraction phase of lengthening of all cases of fibular hemimelia due to the underlying knee instability.It is thus the recommendation of this study to perform the corticotomy for lengthening in the metaphysis.Several complications are closely related to each other, including infection, delayed consolidation, fracture regenerate bone and malalignment.It is thus then recommendation of this study to limit the lengthening goal to ≤ 30% of the original bone length for each lengthening session, regardless of the original LLD.It is the recommendation of this study to avoid early removal of the fixator by partially disassembling the frame and mechanically testing the bone before removal. This study supports the saying that "It is better to remove the apparatus one month too late than one day too early". This study also recommends postoperative splinting of the lengthened limb for three months after surgery [22,23].

Most studies report their results in terms of being satisfactory or unsatisfactory, or being fair, good and excellent, without there being an agreement as to what is considered to be satisfactory or not. The criteria set by Choi et al [15] are reasonable and they are ' to achieve equalization of both limb lengths within 1 cm, to be pain-free with resting or walking, to have no (or a mild) limp, to have a cosmetically acceptable plantigrade foot, to participate in sports and be personally satisfied with regard to gait.

In this study, limb reconstruction had not yet been completed in the majority of cases, as it is a continuous procedure starting since the child's birth and proceeding in multiple stages till the patient reaches skeletal maturity. A time limit was set in this study to be able to analyze data and report results at this stage of treatment. A satisfactory result, hence, is considered one in which the goals of management up to the current stage of treatment have been met with, without long term complications that affect the final outcome. The protocol of management set forth by this study is a reliable and valid option, in the management of fibular hemimelia patients that promises good eventual functional and cosmetic results with excellent to good parent satisfaction.Limitations: The sample size was relatively small, and it was a single center study.

Conclusions

Limb lengthening and deformity correction using Ilizarov external fixator are effective methods in the management of foot deformities and LLD associated with FH. These reconstructive surgeries yield good subjective and objective functional results, patients and parents' satisfaction. As amputation is not acceptable in our society, these surgeries are considered good solutions for FH

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