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A Systematic Review of Ankle Reconstruction with Limb Lengthening in Fibular Hemimelia

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ABSTRACT

Background: fibular hemimelia (FH) is a congenital deficiency where part or all of the fibular bone is hypoplastic, dysplastic or aplastic associated with hypoplasia and dysplasia of the tibia and hypoplasia, dysplasia and aplasia of parts of the foot.

Objective: this systematic review was aimed to review the different method of ankle reconstruction with limb lengthening in patients with Fibular Hemimelia to restore normal weight-bearing and normal limb length so that the patient can walk with a normal gait as possible.

Material and Methods: online search was done using the Medline database on PUBMED, Google Scholar and SCINCEDIRECT from 2001 to 2018; all the English language published studies were identified with the search keywords of, ankle reconstruction with limb lengthening in fibular hemimelia, fibular hemimelia and treatment of fibular hemimelia. Literature search database on PUBMED, Google Scholar and SCINCEDIRECT showed 193 studies.

Results: Our search revealed 6 studies accounting for total of 74 patients included in the final analysis. Numbers of males were 24 patients and females were 30 patients. The mean age is 20.4 month, with average follow up time is 38.6 month. Each study was analyzed, and the following postoperative data were collected: outcomes, recurrence, dorsi-flexion, planter-flexion, and complication.

Conclusion: SUPER ankle procedure is a widely used technique with or without lengthening showing good results in correction of equinovalgus deformity of the ankle. Performing the reconstruction of the ankle at an earlier age plays a significant role in preventing recurrent foot deformities.

Keywords: FH, Ankle.

INTRODUCTION

Fibular hemimelia (FH) is a congenital deficiency where part or all of the fibular bone is hypoplastic, dysplastic or aplastic associated with hypoplasia and dysplasia of the tibia and hypoplasia, dysplasia and aplasia of parts of the foot. The phenotype has a wide spectrum of pathology, ranging from mild to severe limb length discrepancy, ankle/foot deformities with or without subtalar coalition, midfoot coalitions and absent rays. Knee ligament deficiencies and knee valgus deformity as well as associated femoral hypoplasia, dysplasia and partial aplasia are common [(1)].

The incidence of FH is about 1:50,000 in births. Bilateral FH (fibular hemimelia affecting both legs) occurs much less commonly [(2)].

The etiology of FH remains unknown, and in most cases it is usually not an inheritable condition, with the vast majority of children born with this condition having no family history of other birth defects [(3)].

The most commonly used classification of FH is that of Achterman and Kalamchi[(4)], which describes the amount of fibular deficiency.

The **Paley** classification[(6)] is the first classification of FH to be designed with reconstructive surgery options in mind. It is based on the patho-anatomy and deformities of the ankle and subtalar joint. It classifies FH into 4 types; **Type 1:** Stable normal ankle, **Type 2:** Dynamic valgus ankle, **Type 3:** Fixed equino-valgus ankle and **Type 4:** Fixed equino-varus ankle (clubfoot type).

Foot and ankle deformities have been the most challenging and disabling problems with FH. FH foot deformity has many components. At the ankle there is a dysplasia of the distal tibia and of the talus, which ranges from mild valgus of the distal tibia to severe dysplasia with flat malformed, maloriented joint surfaces. The talar neck may be very short and have little concave offset [(6)].

Successful management aims to restore normal weight bearing and normal limb length so that the patient can walk with as normal a gait as possible. In mild cases, treatment includes shoe-raises, prostheses, epiphysiodeses or limb-lengthening procedures, and correction of foot deformities. For more severe deformities, the management is controversial. Many authors recommend early amputation of the foot and prosthetic rehabilitation [(7)].

The introduction of the Ilizarov method of limb lengthening to the world has provided an attractive alternative to amputation [(8)].

The SUPER ankle procedure was developed by **Paley in 2016**. The SUPER ankle procedure is performed in children between 18 and 24 months of age. It involves supramalleolar and/or subtalar osteotomies combined with soft tissue release [(1)]**.**

The aim of the current systematic review was to review the different method of ankle reconstruction with limb lengthening in patients with Fibular Hemimelia to restore normal weight-bearing and normal limb length so that the patient can walk with a normal gait as possible.

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PATIENTS AND METHODS

Search strategy:

Online search was done using the Medline database on PUBMED, Google Scholar and SCINCEDIRECT from 2001 to 2018; all the English language published studies will be identified with the search keywords of, ankle reconstruction with limb lengthening in fibular hemimelia, fibular hemimelia and treatment of fibular hemimelia.

Literature search database on PUBMED, Google Scholar and SCINCEDIRECT showed 193 studies.

1ry screening: 104 studies were excluded due to language other than English and other topics not related to search goals.

2ry screening: We excluded 55 studies due to duplicates and discuss FH associated disease other than ankle deformity.

3ry screening: Full text review was done and 28 articles were excluded due to lack of functional outcome. 6 studies were included.

Inclusion criteria:

Studies which were included in our systematic review met the following guidelines:

1. All cases with ankle reconstruction FH.
2. Surgery or minimal invasive treatment of FH.
3. Pediatric and adolescent.
4. Isolated or associated with other diseases.

Exclusion criteria:

1. Any study not discussing ankle reconstruction FH.
2. Any study discussing conservative methods for FH.
3. Articles with no clinical data.
4. Non English paper.
5. Non human trials.

Three authors used Paley classification, others used Achterman and Kalamchi. There were two studies used the SUPER-ankle surgical correction, and the other four papers used other techniques. Results are summarized in table (1).

Table (1): Showing number of studies, authors, average ages, average follow up, number of patients, average sex, laterality, time of interventions, classifications.

Author	year	Number of patients	Age (average)	sex	laterality	Time of intervention	Classification	Follow up time
Exner[9]	2003	3	7-20 M	M:F (1:2)	Unilateral:2 Bilateral:1	13.5 M	Achterman and Kalamchi type II	(6:42) M
Paley *et al.*[10]	2011	20	18-24 M	Nk	NK	18M	Paley III	NK
El-Tayeby and Ahmed[11]	2012	13	9-26 M	M:F (8:5)	Unilateral:11 Bilateral:2	9-26 M	Achterman and Kalamchi type II	(12:38) M
Cavadas and Thione[12]	2015	1	15Y	F	Unilateral:1	15 Y	Achterman and Kalamchi type Ib	7Y
Hefny *et al.*[13]	2016	8	7- 36M	M: F (3:5)	Unilateral:8 Bilateral:2	18 M	Paley type III	(48:96) M
Kulkarni *et al.*[14]	2017	29	1-9Y	M:F (12:17)	Unilateral:29	4.2Y	Type 1 (8 PT) Type 2 (7 PT) Type 3 (10 PT) Type 4 (2 PT)	(5:14) Y

M = month, Y = year,

Table (2): Showing reported sex data in our study.

	Sex	
	N	**%**
Male	24	44.4%
Female	30	55.6%
Total	54	100%

Table (3): Showing operative data include, number of studies, type of operations, approaches, anlage resections and soft tissue release.

Author	Type	Approach	Lengthening	Anlage	Soft tissue release	Peroneal tendons	Tendo achilles
Exner[9]	Bending osteotomy	Lateral fibular wider exposure	6 weeks later orthoprosthesis	Fully resectio n	+	transposition	lengthening
Paley *et al.*[10]	SUPER ankle	Distal lateral longitudinal excision and proximal lateral at fibular neck	Combined lengthening	Fully resection	+	only lengthen the brevis Z-fashion and never the longus	never
El-Tayeby and Ahmed[11]	Excision of the fibular anlage,act as buttress	Lateral longitudinal excision	Abrace till age of lengthening 3-4 ys	Fibrous anlage	+	Z plasty	Z plasty
Cavadas and Thione[12]	microvascular proximal fibular flap	Lateral longitudinal excision	Not reported	Fully resection	--	--	--
Hefny *et al.*[13]	excision of the fibrous fibular anlage, reconstruction of the lateral malleolus	A zigzag longitudinal lateral incision	later	Fibrous anlage	+	Z plasty	Z plasty
Kulkarni *et al.*[14]	Type 1: lengthenig Type2: hemiepiphysode s or supramalleolar varus osteotomy. Type3: SUPER ankle. Type 4: supramalleolar osteotomy.	Lateral longitudinal excision	later	Fibrous anlage partially resected	+	lengthening	lengthening

Table (4): Showing surgical techniques in our study.

Surgical techniques	N	%
Bending osteotomy	3	4.8%
Super-ankle	30	48.3%
Excision& buttress	21	33.8%
Amicrovascular flap	1	1.6%
Supra-malleolar varus osteotomy	7	11.2%
Total	62	100%

RESULTS

Our search revealed 6 studies accounting for total of 74 patients included in the final analysis. Numbers of males were 24 patients and females were 30 patients. The mean age was 20.4 month, with average follow up time is 38.6 month.

Each study was analysed and the following post-operative data was collected: outcomes, recurrence, dorsi-flexion, planter-flexion, and complication (Table 5).

Table (5): Showing post-operative data, outcomes, recurrence, dorsi-flexion, planter-flexion and complication.

Author	Outcomes	Recurrence	Dorsiflexion	Planterflexion	Complication
Exner[9]	well aligned	Not reported	15-20°	25-30°	Not Reported
Paley *et al.*[10]	Well aligned Painless functional foot	Not reported	Not Reported	Not Reported	Not Reported
El-Tayeby and Ahmed[11]	A stable ankle	Not reported	15-20°	25-30°	Not reported
Cavadas and Thione[12]	acceptable	Not Reported	Not reported	Not Reported	Not Reported
Hefny *et al.*[13]	a stable plantigrade 9 ankles	(1) ankle residual equinus, (5)ankles residual valgus	(8) ankles, complete range	(1) pt lost 5° of dorsiflexion	Late complications of progressive valgus deformity of the tibia and subluxation of the ankle
Kulkarni *et al.*[14]	Excellent result 15 of 27 (55%) Good results 6 of 27 (22%) Fair results 4 pt (14.8%) Poor results 2pt (7%)	(2) type 2patients (4) type 3patients equinovalgus	Not Reported	Not reported	- pin tract infection -recurrent equinovalgus.

Those used SUPER ankle surgical correction include Paley *et al.*[10] and kulkarni *et al.*[14] and reported these results:

Paley *et al.* reported 20 children treated by primary amputation at one institution compared with 22 children treated using the SUPER ankle reconstruction with limb lengthening at another institution, All patients and parents reported satisfaction with treatment method selected and would select the same treatment method again. There were no statistically significant differences in average performance in gait analysis or timed 50-yard dash. Using standardized evaluation tools, both groups showed comparable documented psychosocial adjustment, QoL and physical function. The limb lengthening group will require additional lengthening and/or epiphysiodesis to complete leg length equalization [1, 10].

The results of **Kulkarni and his** associates [14] were evaluated using Association for the Study and Application of Methods of Ilizarov scoring. Excellent results were obtained in 15 of 27 (55%) patients. Six (22%) patients had good results, 4 (14.8%) had fair results, and 2 (7%) had poor results. Mean limb length discrepancy at initial presentation was 3.55cm (range: 2 to 5.5 cm) which significantly improved to 1.01 cm (range: 0 to 3 cm) after treatment (P=0.015).

Results and a review of the literature clearly suggest that limb reconstruction according to Paley classification, is an excellent option in the management of fibular hemimelia. The 2-staged procedure (SUPER ankle procedure followed by limb lengthening) helps in reducing the complications of limb lengthening and incidence of ankle stiffness. Performing the first surgery at an earlier age (below 5 y) plays a significant role in preventing recurrent foot deformities[14].

Other authors have used other techniques for reconstruction of the ankle which included those of **Exner** [9] who reported that feet is well retained in position after a follow up between 6 and 42 months. Furthermore, axis deviations of the tibial antecurvation and valgus deformity partially corrected spontaneously as did the knee valgus. Range of motion in the ankle joint was quite good with dorsiflexion between 15 and 25°and plantar flexion of 25–30°.

El-Tayeby and Ahmed [11] reported that all patients had a stable ankle without tendency to valgus deformity or subluxation. The ankle range of movement was a mean of 27.3° plantarflexion (25–30°) and 18° dorsiflexion (15–20°).

Hefny *et al.* [13] Reported a stable plantigrade foot was achieved in nine ankles; one ankle had residual equinus, five ankles had residual valgus heel, and eight ankles had complete range of motion of the ankle, whereas one patient lost 5° of dorsiflexion. One ankle had equinus deformity. **Cavadas and Thione [12]** using microvascular proximal fibular epiphysis based on the anterior tibial vessels for reconstruction of the fibular malleolus. The results reported were acceptable, although the leg is further shortened with this osteotomy.

Fibular anlage resection:

Some authors prefere fully resection of the fibular anlage and other prefere partially resection. **Paley [15]** underwent fully resection of the anlage with Dissection its borders free anteriorly and posteriorly working from distal to proximal. Separate it from the adjacent calcaneus, it may actually be fused to the calcaneus, cut through the cartilage bridge connecting it to the calcaneus. While **Kulkarni *et al.*[14]** underwent SUPER-ankle with partial resection of fibular anlage.

Hefny *et al.* [13] resect fibular anlage partially, to the cartilaginous remnant of the lateral malleolus distally. In some cases the cartilaginous anlage of the lateral malleolus was found bulbous, and it was used to reconstruct the lateral malleolus, by reimplanting it at a more distal level (the level of the lateral malleolus) and securing it by sutures onto the posterolateral aspect of the distal tibia using heavy Ethibond sutures.

El-Tayeby and Ahmed [11] partially resect of the fibular anlage and the cartilaginous anlage is made to serve as a lateral support to the ankle by fixing it to the distal tibia and talus using two transverse smooth K-wires; it becomes a 'lateral malleolus' buttressing the talus.

Table (6): Fibular anlage resection data in our study.

	N	%
Fully resection	24	43.6%
Partially resection	31	56.3%
Total	55	100%

Tendons lengthening:

Some authors prefer lengthening of peroneal tendons and achillis, others prefer only brevis tendon. Paley prefers lengthening of peroneal brevis only not longus avoid lengthening the Achilles or peroneus longus tendons by the shortening distal tibial osteotomy instead of performing an opening wedge at that level. Paley noticed that in long-term follow up after achillis lengthening there was weak push-off. And many patients developed a supination midfoot deformity with a dorsal bunion due to overpull of the tibialis anterior from a weak peroneus longus tendon (Table 7)[1].

Table (7): Peroneal tendons and Achilles lengthening.

	N	%
Peroneal brievis only	20	37%
Peroneal brevis& longus& achillis	34	73%
Total	54	100%

DISCUSSION

FH presented with the problem of fixed equino-valgus of the foot and ankle. Although, the new lengthening techniques excelled at gaining length, persistent or recurrent foot deformity and recurrent valgus deformity at the ankle, remained unsolved problems despite attempts to solve this problem were performed by many surgeons. The problem again was due to the lack of understanding of the pathoanatomy. The equino-valgus foot deformity was being treated the way club foot deformity was treated, using circumferential surgical release. This approach failed for FH. The reason it failed was that the problem is not contracture of the muscles. The problem is malorientation and dysplasia of the ankle joint and malunion of subtalar coalition. This pathoanatomic problem went unrecognized because the ankle joint is invisible radiographically since it is not ossified and the subtalar coalition is also frequently invisible until a much later age[16].

The absence of the lateral malleolus in fibular hemimelia also causes subluxation and valgus deformity of the ankle joint due to lack of the lateral support at the ankle. Excision of the fibular anlage and centralization of the ankle corrects the valgus deformity but does not prevent the gradual subluxation and valgus deformity that usually recurs with growth and with tibial lengthening, this is illustrated in the study of **El-Tayeby and Ahmed[11] and Hefny *et al.*[13].**

Unsatisfactory results after a salvage procedure are mainly related to recurrent or residual ankle and foot deformities.[1] For example, **Naudie *et al.* [17]** achieved satisfactory results in only 4 of 10 cases after lengthening and the reason for unsatisfactory outcomes involved residual or recurrent foot and tibial deformities. **Cheng *et al.* [18]** reported a similar experience in a small prospective group of 4 cases of lengthening, with unsatisfactory results secondary to recurrent tibial and foot deformities.

Attempts have been described to increase ankle stability in patients with fibula hemimelia. The osteotomy described by Ulrich Exner which altered the convexity of the distal tibia to a concave shape showed aquite good result, the feet were kept well aligned with the tibia. Furthermore axis deviations of the tibial antecurvation and valgus deformity partially corrected [9].

Fibular hemimelia treated by SUPER ankle technique and subsequent lengthening. **Paley** reported satisfactory functional outcomes. Patients were satisfied with their results, were equally and functionally active and had no pain, their function as comparable to normal. The big advantage in using the SUPER ankle is that in addition to normal function, the patient retains a sensate foot that can feel the ground, thereby providing balance and proprioception. No prosthesis provides sensibility or proprioception [1, 15].

The technique of SUPER ankle reported in **Kulkarni *et al.*** [14] study had 2 variations from the original SUPER ankle procedure as described by Paley. **Paley** [15] practiced a combined SUPER ankle procedure with lengthening in a single surgery, with complete resection of the fibular anlage. But Kulkarni et al advocate a 2-staged procedure deformity correction with the SUPER ankle procedure followed by a lengthening procedure. And suggested that the lengthening procedure was performed 1 to 2 years after the first procedure helps in reducing limb lengthening complications and the incidence of ankle stiffness. The second variation is complete resection of the fibular anlage [15].

Paley [1] prefers to perform his procedure when the patient is between 18 and 24 months of age. **Kulkarni *et al.*** [14], revealed a statistically significant relationship between the age at the first surgery and recurrence of foot deformities. Five of 12 patients with an age at the first surgery of more than 5 years had recurrence, while only 1 of 15 patients with an age at the first surgery of less than 5 years had recurrence. Hence, early treatment is important in reducing recurrence and unsatisfactory results.

The major limitation of our study is that we could not compare outcome results. This is because the scanty literature on the topics lack unified outcome measure.

CONCLUSION

SUPER ankle procedure is a widely used technique with or without lenghtening showing good results in correction of equinovalgus deformity of the ankle. Performing the reconstruction of the ankle at an earlier age plays a significant role in preventing recurrent foot deformities.

Using cartilaginous part of the anlage to add to ankle stability is still being experimental and only few case reports showing good result and small case series had published.

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