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Original Article

Pediatric lower limb Ilizarov lengthening with functional evaluation in adulthood: A report on underprivileged patients

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ABSTRACT

Purpose: Ilizarov lengthening, with the principles of Ilizarov, requires a collaboration and supervision of the physiotherapist, nurse, and psychologist, preferably in a group-therapy set-up. We report the mid- and long-term functional outcome of cases that had none of the above listed supporting elements. In addition, we tried to observe the effect of the disease category on the final outcome in the patient.

Method: In this study, 35 children who had undergone Ilizarov lower limb lengthening were evaluated using the following methods: clinical, radiographic, and by four functional scoring systems, and parent/patient satisfaction questionnaires, after an average of 17.2 years (10–25 years).

Results: In this study, 19 boys and 16 girls aged 5–16 years received 18 femoral and 20 tibial lengthening. An average of 6.2 cm lengthening in the femur and 8.4 in the tibia was achieved, with a healing index of 26.5 days. The disease category did not significantly affect the healing index, but the complications, 0.5 per femoral and 0.7 per tibial segment, were more common among congenital, and least among post-traumatic discrepancies. A complete improvement in joint stiffness was observed by 6–12 months post-frame removal in 83% of the cases, following home therapy by parents alone.

Conclusion: The long-term results of Ilizarov lengthening for lower limb discrepancy in children, even without group-therapy or good supportive aids, can improve function and maintain patient satisfaction in two thirds of the cases, over an average period of 17 years.

Levels of evidence: IV.

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1. Introduction

Ilizarov principles revolutionized lengthening and a large correction of discrepancies became possible. Elongation of up

to 40%–70% of the limb segment is now possible, although associated with increased complications.^{1–5} Today, thanks to the Ilizarov principles, several more conditions are deemed suitable for leg preservation and lengthening than ever before.

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The use of Ilizarov in North America was initiated in the 1970s and in Europe in the 1980s.³ Special, well-organized limb lengthening centers have been developed in different parts of the world, with well-organized physiotherapy and psychosocial support for children undergoing major lengthening.

There have been many modifications and advances in the original Ilizarov circular frame fixators, partly with the aim of reducing the associated complications such as joint stiffness, alignment loss, although the basic rules and principles set out by Ilizarov remain in place. Lengthening over an intramedullary rod, maneuvering into lengthening by rotational motion of the leg or with an external magnet, planning the deformity correction through a computer-assisted fixator system (Taylor Special Frame)^{6,7} are some of the newer changes in the use of the Ilizarov principles. We would now like to report our long-term experience with Ilizarov lengthening of lower limbs in children in a general orthopedic setup,

in an environment with challenges accessing secondary support, i.e., physiotherapy, wound care management, regular follow-up care, and social or psychological support. We also reviewed the detailed functional and psycho-emotional status to correlate the cause of discrepancy with the final results.

2. Materials and methods

This is a study of the first series of pediatric cases with limb length discrepancy, who underwent lower limb lengthening by the senior author (GHS) from 1991 to 2006 in accordance with the Ilizarov principles of bone elongation. Only those patients who had reached skeletal maturity or whose planned limb equalization had been achieved were included. Adult patients (over 16 years of age) and those waiting for the second stage of lengthening were excluded from this study.

Fig. 1 – Tibial hemimelia type II. He had undergone tibiofibular synostosis as a small child. (a): X-ray finding at age 2. (b and c): Clinical and X-ray finding at age 7. (d and e): Ilizarov lengthening in progress and 8 cm of tibial lengthening.

Table 1 – Data analysis in 5 groups with lower limb lengthening.

Cause	Frequency (percent)	Age at surgery	Age at follow-up	Length gain	Healing index
Congenital deficiencies	13 (37.1)	9.3 (5–14)	15.7 (10–20)	9.2 (4–14)	24.9 (12.5–50)
Post-poliomyelitis	6 (17.1)	12.8 (12–16)	21.1 (18–25)	5.4 (4–7.5)	30.8 (18–42.1)
Post-infection	5 (14.3)	12.2 (10–14)	18.2 (14–21)	8 (6–13)	25.1 (15–35)
Post-traumatic	3 (8.6)	12.5 (9.5–16)	17.6 (12–23)	7.8 (4–12)	19.4 (11.9–24)
Miscellaneous	8 (22.9)	9.7 (6.5–14)	16.2 (11–22)	8.5 (4–12.5)	30.9 (16.3–65.5)

In addition, patients who underwent surgery for their short stature, and those with upper limb lengthening, were excluded.

The patients were called in for clinical and radiographic evaluations. All the information regarding original joint motions, leg alignment, cause, amount and expected limb discrepancy, were collected from the patient charts. The physical examination included joint motion, limb alignment, neurovascular evaluation, skin scars, and limb length measurements. All the new radiographs included alignment views and scanograms.

Specific questions with regards to receiving any formal physiotherapy, or administered exercise program were asked.

All the patients filled a detailed questionnaire that included: short Musculoskeletal Function Assessment (SMFA), SF36, WOMAC, MACTAR, PODCI (Pediatric Outcome Data Collection Instrument), and parent and patient satisfaction forms.^{8–12}

2.1. Treatment steps

All the 35 cases underwent percutaneous metaphyseal corticotomy, gradual callotasis following the initial 5–7 days latency period. The patients would start ambulation and joint motion exercise from the first post-operative day. The patients were usually discharged 2 days after the beginning of distraction, when sufficient information had been provided to the patient and parents, on lengthening, wound care, and joint exercises.

The patients would then refer back biweekly or monthly (depending on how far away they lived from the hospital) for subsequent clinical and radiographic assessment. The majority of the cases were from small towns or villages with poor access to physiotherapy facilities, and exercise conducted at home by the parents was the only physiotherapy available to them. Following removal of the Ilizarov fixator, 6–8 weeks of a walking cast was also used.

The results were statistically analyzed using the Chi-square test, Fisher's exact test, t-test, Kruskal–Wallis test, and one way-ANOVA.

3. Results

Of the 43 pediatric cases that had the inclusion criteria, only 35 could be reached for evaluation and are included in the present analysis; thus, 81.4% follow-up has been reported. There were 19 males and 16 females, and the average follow-up was 12 years (7–21 years). The mean age at surgery was 10.9 years (5–16 years) and the mean age at final follow-up was 17.2

years (12–25 years). The lengthening included 17 tibias, 15 femurs, and 3 cases of simultaneous lengthening of both bones; 7 tibias and one femur had bifocal lengthening (Fig. 1).

The reasons for lengthening were varied; however, the dominant group was congenital limb deficiencies accounting for 13 (37%) cases (Table 1).

The average lengthening achieved from a single bone was 8.1 cm (3–14 cm): 6.2 (3–10) cm in the femur and 8.4 (3–14) cm in the tibia. Epiphysiodesis of the opposite distal femur and/or proximal tibia were necessary in 5 patients. The healing index was 26.5 days (11.9–50), 29.1 days/cm for unifocal and 19.1 days/cm for bifocal lengthening ($p = 0.005$). No statistically significant difference between the disease types and healing indices was observed (Table 1).

Using Moseley or Paley prediction techniques^{13,14} the LLD had been corrected to be within 1.5 cm in 27 (77%) cases, whereas 8 cases with a remaining discrepancy of 2.5–12 cm had reached skeletal maturity were all satisfied with their original lengthening and did not want a second stage lengthening, included 1 case of osteogenesis imperfecta with loss of length gain following frequent fractures, 1 congenital tibial pseudoarthrosis with healed pseudoarthrosis using a protective brace, 5 cases of congenital bone deficiencies, and 1 case of post Ollier's disease.

The complications included 32 minor ones including pin tract infection, or joint stiffness, and all recovered without any surgery. 12 cases required a second anesthesia during lengthening for frame adjustments, premature consolidations or joint subluxations, with no remaining sequelae, whereas 12 other complications persisted even after lengthening and required additional surgeries. Therefore, with 12 “obstacles” and 12 “complications” (D. Paley¹⁵), the side effects of lengthening were 0.5 per femoral and 0.7 per tibial segment.

The complication of joint stiffness was more specifically looked at, considering the lack of formal physiotherapy or group-therapy. 28 cases (80%) had near complete recovery in joint motion within one year following lengthening, with exercises under parental supervision, and with normal daily activities. In general, 30 feet remained plantigrade and 5 required Achilles tendon lengthening to become plantigrade. Knee stiffness was observed in 4 cases, with 2 cases necessitating quadricepsplasty.

The resolution of joint stiffness was fastest in the post-poliomyelitis cases (an average rate of 2 months) and slowest in patients with congenital deficiencies (an average rate of 5.4 months). In the final follow-up, a residual joint contracture of 10–15° was present in 6 patients, with no functional impairment.

The amount of length gain had no meaningful relation with the encountered complications, except for the pin tract

Table 2 – Frequency of side effect of cases in five groups with Lower limb Lengthening.

Cause	Problem		Obstacle		Complication	
	Case (%)	Issue	Case (%)	Issue	Case (%)	Issue
Congenital limb deficiencies	7 (53.8%)	8	5 (38.4%)	7	6 (46.1%)	8
Post-polio	4 (66.6%)	4	2 (33.3%)	2	1 (16.6%)	1
Post-infection	3 (60%)	3	0 (0%)	0	2 (40%)	2
Post-traumatic	0 (0%)	0	0 (0%)	0	0 (0%)	0
Miscellaneous	6 (75%)	7	3 (37.5%)	3	3 (37.5%)	3
Total	20 (57.1%)	22	10 (28.5%)	12	12 (34.2%)	14

infection ($p = 0.007$), which increased with increased lengthening. The minimum number of complication was observed in post-traumatic, and the maximum in congenital group (Table 2).

The outcome questionnaires showed no deterioration of function following the lengthening procedure. SF36 in 71.4% and PODCI in 74.3% were excellent or good. 31 cases (88.6%) would still have done it, having known the outcome (Table 3).

The 4 unhappy patients (11.4%) included: a congenital pseudoarthrosis (one of the two cases of pseudoarthrosis who gained length but pseudoarthrosis persisted), one fibular hemimelia who did not return for the second stage of his lengthening, a case of osteogenesis imperfect, with over 8 cm remaining discrepancy. The fourth case was a patient with a tuberculosis hip fusion, unhappy with his non-mobile hip despite good length gain.

4. Discussion

In our geographic region, the idea of amputation is very poorly accepted, prosthetic limbs are not only expensive, but also inaccessible to the majority of our patients; making limb lengthening a more appealing procedure. Limb lengthening is an enduring procedure with a high complication rate of 22%–25%.^{3,16–22}

In the present report, the side effects of lengthening (combining obstacles and complications together), were calculated as 65.6% (44.4% in femurs and 82.4% in tibias); these rates are lower than those reported from Hawaii and Spain.^{3,22} However, the tibial lengthening complication report from Canada was 22%.²¹

Studies by Yun et al and Karger et al reported more side effects with the femoral cases,^{22,23} whereas others have reported a similar likelihood of complications in femoral or tibial lengthening.²⁴ Our findings of increased tibial side effects concur with those of Noonan et al.³

Moseley observed increased complications in patients younger than 8 years of age,¹³ whereas Noonan et al, observed

higher complication rates in both femoral and tibial lengthening in patients older than 14 years.³ Although we detected a preponderance of side effects in patients younger than 8 years of age, the difference was not statistically significant. The younger children, who also had more congenital cases among them, often had more length gains (over 10 cm) compared with that of the older children. Therefore, we do not confer with the idea of delaying the lengthening as suggested by other groups.²⁵ The healing index is shorter with a less chance of major joint stiffness among younger patients, and the psychological tolerance may also be better around age 6, as was mentioned by the group in Toronto.²⁶

Pin tract infection was considered as a routine part of the lengthening procedure by Ilizarov.²⁶ The 56% pin tract infections observed in the present study, despite the less than ideal hygienic and medical facilities, mirrors previous literature reports of 10%–100%.^{3,27} The Prophylactic measures such as release of the skin tension at the pin-site, vigilant pin-site care by the parents, helped control the infections and prevent the progression to an “obstacle or complication”.

The reported fracture rate of 1.6%–20% was not observed in our series, although joint stiffness presented in a significant number of our cases. Ankle stiffness is a common side effect of tibial lengthening.^{21–24,28} Catagni reported the need for Achilles tendon lengthening in 35.1% of his cosmetic leg lengthening,²⁹ whereas Stanitski et al,²¹ reported 4.8% and Noonan et al, reported 50%.³ The present series required tendon lengthening in 14% of the patients. Based on the fact that in congenital cases with foot deformity, bifocal lengthening or those requiring more than 6 cm length seem to be more in need of such secondary corrective procedures, we recommend the inclusion of foot in the frame, as was also suggested by Paley.¹⁵

Knee stiffness requiring surgical release or manipulation was observed in 2 femoral lengthening cases, with the congenital short femur and post-infection cases being especially prone to knee contracture. None of the cases in the present series had inclusion of the knee in the frame, but our more recent cases (not included in this review) have had knee joint included, in particular in congenital femoral deficiencies and those with post-infection contractures. This decreases the chance of subluxation of the knee, helps maintain the limb alignment, and does not seem to cause joint stiffness for moderate lengthening of less than 8 cm. Joint stiffness may be a result of the immobilization, and also the effect of lengthening pressure on the joint cartilage.³⁰ Our joint stiffness cases requiring surgical release (2 knees, 4 ankles) may be partly related to the fact that most of our cases were from

Table 3 – Functional outcome.

	Excellent & good	Fair	Poor
SF36	71.4% (25)	22.9% (8)	5.7% (2)
PODCI	74.3% (26)	20% (7)	5.7% (2)
MACTAR	62.9% (22)	25.7% (9)	11.4% (4)

Fig. 2 – (a): 3-year-old boy following diaphysectomy for chronic osteomyelitis. (b): Age 9 after tibiofibular synostosis, and fibulo-talar centralization. (c): During bifocal Ilizarov lengthening, gaining 10 cm of length. (d–f): Clinical and X-ray pictures at 23 years of age.

small towns or villages without access to physiotherapy facilities. However, it is still interesting that self-exercises at home proved quite successful in a good number of our cases. Yun et al reported that 10.3% cases in their study required manipulation or release following knee joint stiffness and Noonan et al, reported 5% of such cases.^{3,22}

Premature consolidations of 1%–33% patients,^{22,31,32} nerve injury in 14% patients,^{20,22} and knee subluxation of 18.7% patients with congenital shortenings²³ have been reported in the literature, and they concur with the present paper. Intensive physiotherapy and splinting helped Yaun et al reduce the number of joint problems.²²

Naudie et al,²⁰ found increased complications in patients who underwent a simultaneous correction of angular or

rotational deformity, compared with that of pure limb lengthening, which was similar to our results.

The evaluation of lengthening outcome has not been adequately addressed in literature. We tried to delve deeper into the real functional results, 12 years later, by employing several different outcome measurement tools. The group from Hawaii commented on their achieved length, alignment, and motion as a functional outcome and reported 78% success.²² In the present paper, the self-evaluated general health (SF36) was good or excellent in only 71.4% patients, and seemed to reflect the frequently associated complex musculoskeletal anomalies. The disease specific functional outcome (WOMAC) that included questions related to pain, joint motion, and psycho-emotional status, was good or excellent in

72% patients and fair in 28.6% patients. Ghoneem, Wright, Cole et al,²⁶ performed a psychological and functional ability assessment on 45 children and observed an 89% functional improvement and a 76% satisfaction with the appearance of leg.

In our series, the patient and parent satisfaction status was 88.6% (Fig. 2). These are the cases who would have accepted the surgery if they had to decide again. Therefore, despite the high complication rate, the achievement of length paired with the elimination of a limp renders this lengthening procedure extremely valuable, with a high level of acceptance. The ability of sitting, cross-legged on the carpeted floor and the use of Persian-type toilet (squatting position) were specific achievements that were highly appreciated by the patients. The patients in this series were often from small towns and villages far away from the place of surgery. The physiotherapy facilities or even a good facility for pin-site and wound cleansing and care were not available for the present group. There was no group-therapy available for these children. The authors realize the merits of access to a good coordinated group treatment; however, even in the absence of such facilities, comparable results can be anticipated.

Conflicts of interest

All authors have none to declare.

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