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Treatment of Congenital Pseudarthrosis Tibia by Ilizarov Principle

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ABSTRACT

Background: Congenital pseudarthrosis of the tibia (CPT) presents significant treatment challenges due to its complex pathology and high complication risk. The Ilizarov technique, based on the principles of distraction osteogenesis, offers a promising approach to manage this condition. **Objective:** To assess the effectiveness of Ilizarov technique in resecting congenital pseudarthrosis and lengthening the tibia, with evaluations of union rates, mechanical axis deviation, range of motion in ankle and knee joints, and complications.

Patients and Methods: In a prospective, retrospective cohort study at Benha University Hospitals, 30 patients with CPT underwent treatment using the Ilizarov technique. Clinical and radiological evaluations were conducted to assess union, while mechanical axis deviation and joint range of motion were measured.

Results: The mean age of the participants was 11.3 ± 3.65 years, with a male predominance (60%). The Ilizarov technique, involving corticotomy and bone transport in 83.33% of patients, showed union in 93.33% of cases, with treatment duration averaging 9.9 ± 4.06 months. Radiological outcomes were rated as excellent in 20%, good in 46.67%, fair in 26.67%, and poor in 6.67% of patients. Complications included pin tract infections in all patients, pain in 13.33%, non-union in 6.67%, refracture in 20%, ankle stiffness in 10%, and valgus deformity at the ankle in 16.67%. The final leg-length discrepancy was corrected to a mean of 2.7 ± 1.04 cm.

Conclusion: The Ilizarov technique demonstrates a high efficacy in treating CPT, with significant improvements in bone lengthening, union rates, and mechanical alignment.

Keywords: Congenital Pseudarthrosis; Tibia, Ilizarov, Complications.

INTRODUCTION

Congenital pseudarthrosis of the tibia (CPT) is a rare orthopedic condition characterized by nonunion of the tibia observed at birth or developing in early childhood. This disorder poses significant challenges in treatment due to its complex etiology, which is often associated with neurofibromatosis type 1 (NF1) and results in a high rate of complications including refractures, leg length discrepancies, and joint deformities [1]. The pathophysiology of CPT involves abnormal periosteal and endosteal bone formation, leading to segmental weakness and a predisposition to fracture and nonunion [2]. Traditional management strategies for CPT have included bracing, surgical resection of the pseudarthrosis site, intramedullary fixation, and bone grafting. Despite these efforts, the treatment outcomes have often been unsatisfactory, with high rates of refracture, nonunion, and significant functional disability. These challenges have necessitated the exploration of alternative treatment modalities that can provide better outcomes for patients suffering from this debilitating condition [3].

The Ilizarov technique, based on the principle of distraction osteogenesis, has emerged as a promising approach for the management of CPT. This method involves the surgical application of an external fixator to gradually lengthen and straighten the bone through mechanical distraction at a controlled rate. The technique stimulates new bone formation at the distraction site and promotes the healing of nonunions by improving the biological and mechanical environment of the bone. It offers the potential for correcting leg-length discrepancies, improving alignment, and achieving bone union without the need for extensive bone grafting [4,5].

However, the application of the Ilizarov technique in the treatment of CPT is not without challenges. The process requires prolonged use of the external fixator, which can be associated with complications such as pin tract infections, joint stiffness, and psychological impact on the patient due to the cumbersome nature of the device. Additionally, the success of the technique is highly dependent on the expertise of the treating team and the compliance of the patient and their family [6,7].

Given the complexities associated with the management of CPT and the potential benefits of the Ilizarov technique, there is a pressing need to systematically evaluate its effectiveness and safety in this patient population. The aim of this study is to provide a comprehensive assessment of the Ilizarov technique for the treatment of CPT, focusing on clinical and radiological outcomes, correction of deformities, and the incidence of complications, thereby contributing valuable insights to the existing body of knowledge on this challenging orthopedic condition.

PATIENTS AND METHODS

In this prospective, retrospective cohort study, thirty patients admitted to the Orthopedic Surgery Department at Benha University Hospitals with congenital pseudarthrosis of the tibia (CPT). Patients were deemed ineligible if they met the following criteria: age greater than eighteen, failure to complete a minimum follow-up of one-year, reduced foot circulation, or severe injury to the posterior tibial nerve.

All individuals who were part of the study underwent the following evaluations:

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Clinical: Full history taking from the patient's guardians, including any prior operations, age at first fracture, and demographic data (age, sex).
A complete medical history detailing the fracture site, the extent of the tibia with aberrant bone involvement, and any fibula involvement. Screening for neurofibromatosis was also conducted. The Crawford classification was used to examine all patients [1].

Radiological Evaluation:
Standard radiographs, including anteroposterior and lateral views. Total bone scintigraphy using a long film.

Surgical Treatment by Ilizarov:

The surgical treatment of CPT using the Ilizarov technique involved precise preoperative planning and execution. Before surgery, patients were assessed through clinical and radiological evaluations to customize the Ilizarov apparatus for each case, considering the extent of pseudarthrosis, tibial deformity, and leg-length discrepancy. Under general anesthesia, the Ilizarov frame, consisting of circular rings connected by threaded rods, was applied. The surgery included resection of the pseudarthrotic tissue, application of wires and half-pins for rigid fixation, and corticotomy for bone lengthening, when necessary, all under fluoroscopic guidance to ensure optimal alignment and avoid damage to neurovascular structures. Postoperatively, the process of distraction osteogenesis commenced after a latency period, with gradual bone lengthening at a controlled rate, closely monitored through clinical and radiological follow-ups. The comprehensive postoperative care regimen emphasized infection prevention at pin sites, pain management, and early mobilization to prevent joint stiffness. Weight-bearing was gradually introduced based on the strength of the regenerate bone. The Ilizarov frame was removed once satisfactory bone union was confirmed radiologically, marking the end of a complex but effective treatment process aimed at restoring function and improving the quality of life for patients with CPT.

Ethical considerations: The Ethics Committee of the Faculty of Medicine at Benha University in Egypt approved the study. All participants' legal guardians provided written informed consent before their children could participate in the study. The consent form explicitly outlined their agreement to participate in the study and for the publication of data, ensuring protection of their confidentiality and privacy. This work has been carried out in accordance with The Code of Ethics of the World Medical Association (Declaration of Helsinki) for studies involving humans.

Statistical analysis

For the statistical analysis, SPSS v26 was utilised (IBM Inc., Armonk, NY, USA). The statistics were shown using the mean and standard deviation (SD) for the quantitative variables. The qualitative factors were displayed using frequency and percentage (%).

RESULTS

This flowchart shows that 33 individuals were assessed for eligibility, all were allocated to receive the Ilizarov principle intervention, 30 completed the treatment and follow-up, and all 30 were included in the final analysis with none excluded **(Figure 1)**.

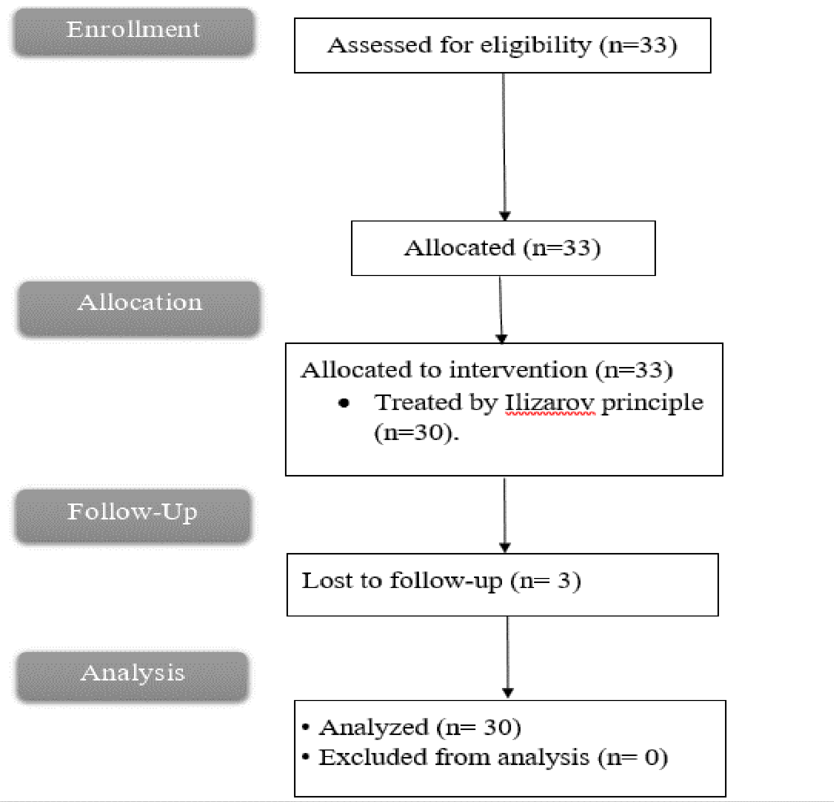

Figure 1: Flowchart of the studied patients

The age of the patients who were studied varied from 5 to 17 years. Eighteen percent were males. Thirteen patients, or 43.33% of the total, had symptoms in their right leg, whereas fifteen patients, or 50% of the total, had symptoms in their left leg (Table 1).

Table 1: Demographic data of the studied groups.

		n=30
Age (years)	Mean ± SD	11.3 ± 3.65
	Range	5 - 17
Sex	Male	18 (60%)
	Female	12 (40%)
Side	Right	15 (50%)
	Left	15 (50%)

SD: standard deviation.

Type II Crawford classification was seen in all thirty individuals (100 percent). Concerning neurofibromatosis, four patients exhibited this condition. The variability in leg length differed by an average of 7.4 ± 3.95 cm (Table 2).

Table 2: Clinical history of the studied patients

		n=30
Crawford classification	Type II	30 (100%)
Neurofibromatosis	Yes	4 (13.33%)
	No	16 (53.33%)
LLD (Cm)	Mean ± SD	7.4 ± 3.95
	Range	1 - 17

LLD: leg-length discrepancy.

In terms of the Illizarov procedure, 25 patients underwent a corticotomy and bone transport. The Ilizarov therapy lasted an average of 9.9 ± 4.06 months (Table 3).

Table 3: Treatment data of the studied patients

		n=30
Illizarov Technique	BT	25 (83.33%)
	CD	5 (16.67%)
Duration of the Ilizarov treatment (months)	Mean ± SD	9.9 ± 4.06
	Range	4 - 16
Bone graft used	No	30 (100%)

Radiological data showed that 14 patients (46.67 percent) had good outcomes (Table 4).

Table 4: Radiological results of the studied patients

		n=30
Radiological results	Excellent	6 (20%)
	Good	14 (46.67%)
	Fair	8 (26.67%)
	Poor	2 (6.67%)

Complications in the studied patients are shown in table 5.

Table (5): Complications in the studied patients

		n=30
Pain	Yes	4 (13.33%)
	No	26 (86.67%)
Non-union	Yes	2 (6.67%)
	No	28 (93.33%)
Refracture	Yes	6 (20%)
	No	24 (80%)
Final LLD (Cm)	Mean ± SD	2.7 ± 1.04
	Range	1 - 4.4
Ankle stiffness	Yes	3 (10%)
	No	26 (86.67%)
Angulation (> 10°)	Yes	7 (23.33%)
	No	22 (73.33%)
Valgus deformity at the ankle	Yes	5 (16.67%)
	No	24 (80%)

LLD: leg-length discrepancy.

DISCUSSION

Regarding demographic data, **Enemudo *et al.*** [8] included five patients, predominantly males (2:3 ratio), aged 6 to 18 years (mean 11.6) with a mean leg-length discrepancy (LLD) of 7.2 cm, reduced to zero post-treatment. All patients had unilateral limb involvement, café au lait spots, and were from low-income backgrounds, with varied non-union types observed. **Zayda *et al.*** [9] retrospectively analyzed sixteen CPT patients treated with segmental resection or the Ilizarov method, noting a mean age of 5.4 ± 2.8 years at the index operation. There was a slight majority of right-leg (56.2%) involvement over the left (43.8%), with patients having a history of 0 to 7 unsuccessful prior surgeries. Intervention occurred; an average of 2.8 ± 2.0 years after the initial surgery.

According to the classification by Paley, seven patients had type I CPT, seven had type II, and two had type III, with one case of segmental tibial dysplasia not fitting any category due to the absence of atrophic or sclerotic bone ends before fracture. The study highlighted the lack of a universally applicable CPT classification that encompasses its pathological, clinical, and prognostic aspects. **Enemudo *et al.*** [8] reviewed 18 CPT cases treated with the Ilizarov technique and intramedullary fixation, noting an average patient age of 6.2 years, with a gender distribution of 38.9% males and 61.1% females, and unilateral engagement with a slight preference for the left side (52.6%).

Our study observed that 83.33% of patients had distal third tibia fractures, and 16.67% had mid-shaft fractures, with a significant portion (50%) also having fibula involvement. All patients were classified as type II Crawford, with 13.33% showing neurofibromatosis and an average leg length discrepancy of 7.4 ± 3.95 cm. **Zayda *et al.*** [9] reported an average limb shortening of 3.6 ± 1.9 cm, classifying patients according to the El-Rosasy-Paley classification, and noted a high incidence of fibula dysplasia (81.3%). Preoperative gaps at the

CPT site averaged 1.3 ± 1.0 cm, expanding to 6.1 ± 1.0 cm post-resection, with preoperative AOFAS scores ranging from 38-65. **Enemudo *et al.*** [8] found that most patients had lower third tibia involvement, were classified as Crawford type IV, with half showing fibula pseudarthrosis and a significant portion (72.2%) having NF1, and an average preoperative LLD of 3.2 cm. Four patients had undergone previous unsuccessful surgeries, while fourteen had no prior surgical attempts.

Ankle valgus was a common postoperative complication, observed in 38.9% of patients with an average deformity of 10.3° (5-20°), attributed to inadequate lateral support from the distal fibula [10,11]. **Andersen *et al.*** and **Dobbs *et al.*** noted that despite over 50% of patients with fibula nonunion not showing ankle valgus, the condition often developed in those with fibular pseudarthrosis, evident in five of the seven individuals with ankle valgus in their study [12,13]. Given that recurrent pseudarthrosis can occur in nearly half of the patients after supramalleolar distal tibia osteotomy for ankle valgus, this method is considered least favorable [14].

Stevens *et al.* recommended medial hemi-epiphysiodesis for timely treatment of ankle valgus [15]. Despite eligibility for surgery to correct ankle valgus, all patients declined, with most having an angle under 13°. Postoperatively, 22.2% developed proximal tibial valgus, averaging an angle of 12.1° (5-25°), potentially due to proximal tibial dysplasia [16]. The average post-surgery LLD was 1.4 cm (0.6-3.1 cm) in 11 patients, with minor LLDs treated non-surgically. Recent research has shown fibular pseudarthrosis's impact on refracture rates and ankle stability [10], with **Tudisco *et al.*** linking worse outcomes to fibular pseudarthrosis [17]. Persistent fibular pseudarthrosis was associated with tibial nonunion and increased ankle valgus [18].

Four patients (25%) developed knee valgus deformity (10-20 degrees) within 1-3 years, with two undergoing corrective hemiepiphysiodesis. A 15-year follow-up revealed one case of knee varus deformity. Pin tract infections were common but manageable with oral antibiotics and dressing changes, except for three severe cases requiring intervention [9]. **Cho *et al.*** reported one non-union and 19 refractures among 43 patients treated with the Ilizarov technique [19]. **Agashe *et al.*** observed non-union, delayed union, and one amputation recommendation in their cohort [20]. Our study recorded no surgical complications, with 44.4% developing pin-tract infections, and 38.9% showing ankle valgus at the latest follow-up. Additionally, proximal tibial valgus was noted in 12.1° on average, and LLD averaged 1.4 cm. Refractures occurred in 27.8% of patients, partly due to non-adherence to bracing instructions [8].

Of the five patients with refracture, two cases were healed by cancellous bone graft and acute compression and 3 cases were healed by cancellous bone graft combined with plate fixation. According to **Liu *et al.*,** refracture is the most severe consequence following CPT healing. Refracture prevention should be a primary objective of CPT treatment.[21].

The maintenance of intramedullary rod fixation may be a viable strategy for mitigating the risk of refracture. Generally, it is required to alter the position of the intramedullary rod in order to restore ankle joint function once the tibial pseudarthrosis has healed. It is not advisable to remove the intramedullary rod until the tibial pseudarthrosis has fully healed. In most cases, ankle fixation lasts no longer than two years, and ankle fixation for less than two years has minimal effect on ankle functioning [1,22].

This study had some limitations, as it was a single center study with a modest sample size.

CONCLUSION

The Ilizarov technique demonstrates a high efficacy in treating CPT, with significant improvements in bone lengthening, union rates, and mechanical alignment. Despite the high occurrence of manageable complications, its ability to correct leg-length discrepancies and promote union supports its use as a viable option for CPT treatment.

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REFERENCES

1. **Shah H, Rousset M, Canavese F (2012)**: Congenital pseudarthrosis of the tibia: Management and complications. Indian J Orthop., 46:616-26.
2. **Xu J, Zhang Y, Zhu K *et al.* (2022)**: Clinical characteristics and in silico analysis of congenital pseudarthrosis of the tibia combined with neurofibromatosis type 1 caused by a novel NF1 mutation. Front Genet., 13:991314.
3. **Eisenberg K, Vuillermin C (2019)**: Management of congenital pseudoarthrosis of the tibia and fibula. Curr Rev Musculoskelet Med., 12:356-68.
4. **Malkova T, Borzunov D (2021)**: International recognition of the Ilizarov bone reconstruction techniques: Current practice and research (dedicated to 100th birthday of G. A. Ilizarov). World J Orthop., 12:515-33.
5. **Xie L, Huang Y, Zhang L *et al.* (2023)**: Ilizarov method and its combined methods in the treatment of long bone defects of the lower extremity: systematic review and meta-analysis. BMC Musculoskelet Disord., 24:891.
6. **Fahad S, Habib A, Awais M *et al.* (2019)**: Infected non-union of tibia treated with Ilizarov external fixator: Our Experience. Malays Orthop J., 13:36-41.
7. **Li J, Li M, Wang W *et al.* (2022)**: Evolution and development of Ilizarov technique in the treatment of infected long bone nonunion with or without bone defects. Orthop Surg., 14:824-30.
8. **Enemudo R, Edomwonyi E, Obumse A *et al.* (2023)**: Management of congenital pseudoarthrosis using Ilizarov Device–Delsuth, Oghara Experience. Niger J Clin Pract., 26:23-34.
9. **Zayda A, Mesregah M, Zalalo S *et al.* (2022)**: Functional and radiological outcomes after treatment of congenital pseudarthrosis of the tibia using the Ilizarov

technique: a retrospective single-center study. J Orthop Traumatol., 23:48-55.
10. **Yalikun A, Yushan M, Hamiti Y *et al.* (2022)**: Combination of the Ilizarov method and intramedullary fixation for the treatment of congenital pseudarthrosis of the tibia in children: A retrospective observational study. Front surg., 9:901262-9.
11. **Vanderstappen J, Lammens J, Berger P *et al.* (2015)**: Ilizarov bone transport as a treatment of congenital pseudarthrosis of the tibia: a long-term follow-up study. J Child Orthop., 9:319-24.
12. **Andersen K (1973)**: Radiological classification of congenital pseudarthrosis of the tibia. Acta Orthop Traumatol Turc., 44:719-27.
13. **Dobbs M, Rich M, Gordon J *et al.* (2004)**: Use of an intramedullary rod for treatment of congenital pseudarthrosis of the tibia: a long-term follow-up study. JBJS., 86:1186-97.
14. **Horn J, Steen H, Terjesen T (2013)**: Epidemiology and treatment outcome of congenital pseudarthrosis of the tibia. J Child Orthop., 7:157-66.
15. **Stevens P, Kennedy J, Hung M (2011)**: Guided growth for ankle valgus. J Pediatr Orthop., 31:878-88.
16. **Zhu G, Mei H, He R *et al.* (2016)**: Combination of intramedullary rod, wrapping bone grafting and Ilizarov's fixator for the treatment of Crawford type IV congenital pseudarthrosis of the tibia: mid-term follow up of 56 cases. BMC Musculoskelet Disord., 17:1-8.
17. **Tudisco C, Bollini G, Dungl P *et al.* (2000)**: Functional results at the end of skeletal growth in 30 patients affected by congenital pseudoarthrosis of the tibia. J Pediatr Orthop., 9:94-102.
18. **Pannier S (2011)**: Congenital pseudarthrosis of the tibia. Orthop Traumatol Surg Res., 97:750-61.
19. **Cho T, Choi I, Lee S *et al.* (2008)**: Refracture after Ilizarov osteosynthesis in atrophic-type congenital pseudarthrosis of the tibia. J Bone Joint Surg Br., 90:488-93.
20. **Agashe M, Song S-H, Refai M *et al.* (2012)**: Congenital pseudarthrosis of the tibia treated with a combination of Ilizarov's technique and intramedullary rodding: 15 cases followed for mean 4.5 years. Acta Orthop., 83:515-22.
21. **Liu Y, Yang G, Zhu G *et al.* (2021)**: Application of the "telescopic rod" in a combined surgical technique for the treatment of congenital pseudarthrosis of the tibia in children. J Orthop Surg Res., 16:1-6.
22. **Shah H, Doddabasappa S, Joseph B (2011)**: Congenital pseudarthrosis of the tibia treated with intramedullary rodding and cortical bone grafting: a follow-up study at skeletal maturity. J Pediatr Orthop., 31:79-88.