

Outcome of Segment Transport By Ilizarov In Congenital Pseudoarthrosis of The Tibia

Shujaat Hussain,^{1*} Ghulam Shabbir,¹ Muhammad Javed,¹ Faisal Mubeen¹

ABSTRACT

Objective To find out frequency of bony union and postoperative complications with segment transport by Ilizarov in patients with congenital pseudoarthrosis of the tibia (CPT).

Study design Descriptive case series.

Place & Duration of study Department of Orthopedics, Bahawal Victoria Hospital, Bahawalpur, from June 2009 to December 2016.

Methodology In all patients osteotomy was done and Ilizarov fixator applied for bifocal technique of segment transport. Variables under consideration were leg length discrepancy, associated neurofibromatosis, postoperative hospital stay, bony union, postoperative complications and patients' compliance and patient/attendant satisfaction.

Results Ilizarov external fixator was applied to all seven patients after generous excision of the sclerosed bone (2.5 - 4cm) with thorough debridement of surrounding periosteum. Consolidation (clinical and radiological) was achieved in average 5.86 weeks. Ilizarov fixator was removed followed by application plaster cast for further six weeks. All patients showed bone union. There was no case of re-fracture, nonunion and residual leg deformity during average follow up of two years.

Conclusion Ilizarov fixation not only preserved local circulation, produced adequate compression but also covered the bone loss by segment transport resulting in bone union in all cases.

Key words Congenital pseudoarthrosis, Ilizarov method, Corticotomy, Osteotomy.

INTRODUCTION:

Congenital pseudoarthrosis of the tibia is a rare condition, occurring in only 1 out of 250,000 births.¹ There is a strong association with neurofibromatosis in 50% - 90% of cases and an association with fibrous dysplasia in 10% of cases.² Patients of the congenital pseudoarthrosis of tibia usually present with anterior bowing of the tibia along with defect in lower third, with or without fracture, at birth or before two years

of age. Affected limb is shortened when associated with neurofibromatosis involving the skin. Radiologically medullary canal may be narrow and sclerotic with hour glass constriction or cystic changes in the lower tibia.

Classification according to radiographic findings includes atrophic or hypertrophic pseudoarthrosis.² Pathogenesis of this conditions is still unclear. The fibrous soft tissue found in the pseudoarthrosis and the abnormal periosteum are certainly the key to the pathology, possibly due to decreased osteogenic capacities and impaired local vascularization.^{3,4} Various classification system like Andersen, Boyd, Crawford, and El-Rosasy-Paley-Herzenberg exist.⁵⁻⁸ Classification systems are based upon prognostic radiographic characteristics, natural history, treatment guidelines and other factors that influence the outcome.

¹ Department of Orthopedic Surgery, QAMC / Bahawal Victoria Hospital, Bahawalpur

Correspondence:

Dr. Shujaat Hussain
Department of Orthopedic Surgery
QAMC / Bahawal Victoria Hospital,
Bahawalpur
E mail: drshujaathussainortho@gmail.com

Treatment of CPT is tricky due to high incidence of nonunion and re-fracture.^{6,9} Many treatment strategies have been described and proposed over the years for CPT. Classically resection of disease portion and bone grafting (vascularized or non-vascularized) followed by plating, intramedullary nailing, have been performed. Recently focus on stimulating the healing process through the use of bone grafts, bone morphological protein and fixation techniques are adopted.^{10,11} Most of these treatment modalities result in nonunion, graft failure, hardware failure, limping, pain and difficulty or inability to walk.

The Ilizarov method has been widely used since the late 1980s around the world. It is a useful treatment of CPT. It can address pseudo-arthritis and other components of the complex deformities related to this state. It gives good compaction at the fracture site and is ideal method to cover bone defect by segment transport. This study was done to share the experience of management of this rare condition. It specifically targeted at finding out the bony union with segment transport by Ilizarov in congenital pseudo-arthritis of the tibia.

METHODOLOGY:

This was a case series study conducted at Department of Orthopedics, Bahawal Victoria Hospital Bahawalpur, from June 2009 to December 2016. Patient presenting with pseudo-arthritis of 3-6 year of age, were included in this study. Patients more than 6 year of age or having severe associated deformity were excluded.

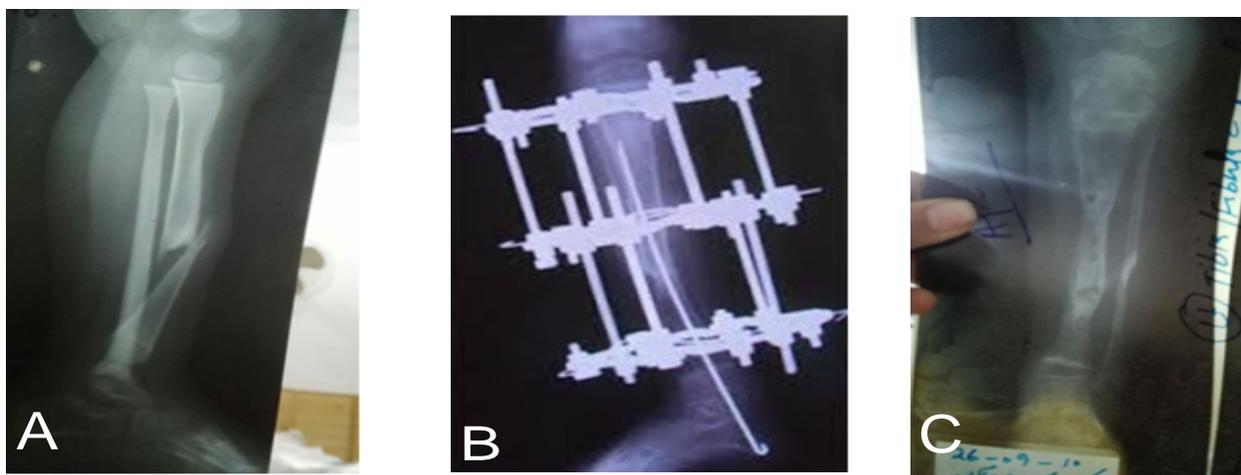
All patients were admitted from outpatient department. Data was collected on predesigned form. It included age, presenting complaint (clinical presentation, age of fracture, site of deformity and clinical shortening) and operative detail (size of

sclerosed end, abnormal periosteum and length of segmental defect). Postoperative details like hospital stay, compliance, satisfaction, time for maturation of consolidation (radiologically and clinically) and complications were recorded.

A formal written permission was taken from IRB prior to study. Patients being minor a consent with explanation of all ethical rights, were taken from Parents/guardians. After detailed preoperative assessment and appropriate investigations, these patients were operated. Ilizarov external fixator was applied to all patients after generous excision of the sclerosed bone. Ilizarov fixator was utilized employing technique of segment transportation after corticotomy (osteotomy). In this technique; the sclerosed, tapered ends of the tibia, hypertrophied abnormal periosteum along with intervening abnormal fibrous tissue, were excised. Corticotomy was done on the proximal normal bone. Three rings were attached with two appropriate sized wires in proximal middle and distal part of tibia in cross fashion. On 5th day of osteotomy segment transport was started at the rate of 1 mm/day.

RESULTS:

Seven patients were included in the study. Three were male and 4 female. Age range was 3-6 year. The most common clinical presentation was fracture in all patients. Evident anterior bowing was found in five patients. Five patients were already treated conservatively in different centers and one patient presented with failed surgical treatment. One patient presented directly to our facility. In 2 out of 7 patients the affected limb was very short (predicted limb length discrepancy >7.5 cm at the time of maturity). Neurofibromatosis was not found in any patient.



Figures: Radiograph before, during and after management with Ilizarov (A): Fractured at presentation (B): Treated with Ilizarov (C): Ilizarov removed after consolidation

Table I: Detail of Clinical and Radiological Assessment

S. No.	No of Patients	Segmental Defect	Period of Union at Fracture Site	Period of Maturation of Consolidation
1	2	< 3 cm	4 weeks	4-6 weeks
2	4	3-4 cm	4 weeks	5-8 weeks
3	1	Less than 3 cm (Re do)	6 weeks	8 weeks
Total	7	2.5-4 cm	4-6 weeks	4-8 weeks

Peroperative segmental defect was found to be less than 3 cm in 2 cases and 3-4 cm in 5 cases. In six patients the segmental bone defect was covered in 23 to 42 days (average 32 days). In one patient, 1.5 months after applying the Ilizarov fixator, there was no sign of bone union and an additional procedure for soft tissue debridement and excision sclerosed end around the fracture site was done.

Once required bone length was achieved, distraction was stopped and the struts of the fixator were locked. Consolidation was achieved at an average of 5.86 weeks. After achieving union on fracture site and consolidation on corticotomy site clinically and radiologically, the fixator was removed and plaster cast was applied for further six weeks. There was superficial pin track infection in two cases and were treated conservatively with antibiotic and local wound care.

All patients except one showed good union at fracture site without further intervention (table I). Patients were followed up three monthly for two years. There was no re fracture during follow up. In our study the patient/attendants compliance and satisfaction was very good in five cases. In two cases the repeated counselling was needed.

DISCUSSION:

Congenital pseudoarthrosis of tibia is a rare disease. Many modalities are employed for the management. No method has famed itself as gold standard in attaining the goals of gaining union and adequate function.¹² Other treatment modalities can be employed if this technique fails.¹³ Different techniques based on different frame configurations have been documented for the Ilizarov method.^{14,15}

Most of the clinical finding in our series correlate with the literature except neurofibromatosis.² Main objective of various techniques used is union of the pseudoarthrosis site, but other factors influencing outcome including leg-length discrepancy, multilevel and multidirectional tibial deformity, foot deformity, associated fibular pseudoarthrosis and subsequent ankle valgus.^{12,16}

CPT is said to be one of the most difficult and challenging diseases for pediatric orthopedic surgeons.¹⁷ Many options have been used by different workers with good results. A study from Japan has shown good results with Ilizarov and fibular vascularized graft.¹⁸ The role of joining surgical technique with biological proteins has been described to enhance healing process. The method is costly and further studies are needed to prove that results are clinically significant.¹⁹

A union rate of 60-100% have been reported in different retrospective case series.¹³ Union was found in all patients in present study which reinforce the usefulness of Ilizarov technique. As reported by other workers union and function after Ilizarov treatment have been seen to be influenced by other factors such as the extent of dysplasia, failure of past treatments and co-existence of deformities.²⁰

It is suggested that the debridement for redundant fibrous tissue, sclerotic bony ends and hyperplastic periosteum should be sufficient. Intramedullary K-wire in our series helped in better alignment but Taylor's spatial frame is superior, though very costly.

CONCLUSIONS:

Congenital pseudoarthrosis of tibia is a rare disease and difficult to treat. Ilizarov fixation provided good results. It preserved local circulation, produced adequate compression and covered the bone loss by segment transport resulting in bone union.

REFERENCES:

1. Hefti F, Bollini G, Dungal P, Fixsen J, Grill F, Ippolito E, et al. Congenital pseudarthrosis of the tibia: history, etiology, classification, and epidemiologic data. *J Pediatr Orthop B*. 2000;9:11-5.
2. Morrissy RT, Riseborough EJ, Hall JE. Congenital pseudarthrosis of the tibia. *J Bone Joint Surg Br*. 1981;63-B:367-75.

3. Lee DY, Cho TJ, Lee HR, Lee K, Moon HJ, Park MS, et al. Disturbed osteoblastic differentiation of fibrous hamartoma cell from congenital pseudoarthrosis of the tibia associated with neurofibromatosis type I. *Clin Orthop Surg.* 2011;3:230-7.
4. Cho TJ, Seo JB, Lee HR, Yoo WJ, Chung CY, Choi IH. Biologic characteristics of fibrous hamartoma from congenital pseudoarthrosis of the tibia associated with neurofibromatosis type 1. *J Bone Joint Surg Am.* 2008;90:2735-44.
5. Andersen KS. Radiological classification of congenital pseudoarthrosis of the tibia. *Acta Orthop Scand.* 1973;44:719-27.
6. Boyd HB. Pathology and natural history of congenital pseudoarthrosis of the tibia. *Clin Orthop Relat Res.* 1982;166:5-13.
7. Crawford AH. Neurofibromatosis in children. *Acta Orthop Scand.* 1986;218:1-60.
8. El-Rosasy M, Paley D, Herzenberg JE. Congenital pseudarthrosis of the tibia. In: Rozbruch SR, Ilizarov S, editors. *Limb Lengthening and Reconstruction Surgery.* New York: Informa Healthcare. 2007: pp. 485-93.
9. Gutmann DH, Aylsworth A, Carey JC, Korf B, Marks J, Pyeritz RE, et al. The diagnostic evaluation and multidisciplinary management of neurofibromatosis 1 and neurofibromatosis. *JAMA.* 1997;278:51-7.
10. Karol LA, Haideri NF, Halliday SE, Smitherman TB, Johnston CE. Gait analysis and muscle strength in children with congenital pseudoarthrosis of the tibia: The effect of treatment. *J Pediatr Orthop.* 1998;18:381-6.
11. Johnston CE. Congenital pseudoarthrosis of the tibia: results of technical variations in the charnley-williams procedure. *J Bone Joint Surg Am.* 2002;84-A:1799-810.
12. Johnston CE, Birch JG. A tale of two tibias: a review of treatment options for congenital pseudoarthrosis of the tibia. *J Child Orthop.* 2008;2:133-49.
13. Vander Have KL, Hensinger RN, Caird M, Johnston C, Farley FA. Congenital pseudoarthrosis of the tibia. *J Am Acad Orthop Surg.* 2008;16:228-36.
14. Toh S, Harata S, Tsubo K, Inoue S, Narita S. Combining free vascularized fibula graft and the Ilizarov external fixator: recent approaches to congenital pseudoarthrosis of the tibia. *J Reconstr Microsurg.* 2001;17:497-508.
15. Gracheva VI, Makushin VD, Shevtsov VI, Kuftyrev LM, Degtiarev VE. Ilizarov's transosseous osteo synthesis in treating congenital pseudoarthroses of the leg. *Ortop Travmatol Protez.* 1981;7:34-8.
16. Ilizarov GA, Gracheva VI. Bloodless treatment of congenital pseudarthrosis of the crus with simultaneous elimination of shortening using dosed distraction. *Ortop Travmatol Protez.* 1971;32:42-6.
17. Rose RE, Wright DE. Treatment of congenital pseudarthrosis of the tibia with the Ilizarov technique. *West Indian Med J.* 2007;56: 294-9.
18. Ohnishi I, Sato W, Matsuyama J, Yajima H, Haga N, Kamegaya M, et al. Treatment of congenital pseudarthrosis of the tibia: a multicenter study in Japan. *Pediatr Orthop.* 2005;25:219-24.
19. Pannier S. Congenital pseudarthrosis of the tibia. *Orthop Traumatol Surg Research.* 2011;97:750-61.
20. Paley D, Herzenberg JE, Tetsworth K, McKie J, Bhav A. Deformity planning for frontal and sagittal plane corrective osteotomies. *Orthop Clin North Am.* 1994;25:425-65.

Received for publication: 09-01-2018

Accepted after revision: 03-03-2018

Author's Contributions:

Shujaat Hussain: Conception ,design, acquisition, analysis and interpretation of data of work, Drafting of work, final approval and agreement for accountability.

Ghulam Shabbir: Design of work, drafting, final approval and agreement for accountability

Muhammad Javed: Analysis, critical revision, final approval and agreement for accountability.

Faisal Mubeen: Interpretation of data, revision, final approval and agreement of accountability

Conflict of Interest:

The authors declare that they have no conflict of interest.

Source of Funding:

None

How to cite this article:

Hussain S, Shabbir G, Javed M, Mubeen F. Outcome of segment transport by Ilizarov in congenital pseudarthrosis of the tibia. *J Surg Pakistan*. 2018;23(1):25-29. doi:<http://dx.doi.org/10.21699/jsp.23.1.7>.