



Evaluation of Treatment of Congenital Pseudarthrosis of Tibia by Ilizarov Method

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Abstract

Background: Congenital pseudarthrosis of the tibia, a rare but well-known disorder, has been remarkably resistant to all types of therapy designed to promote healing. Successful treatment consists of the union of the pseudarthrosis and maintenance of that union without malunion, re-fracture, or excessive shortening (<2cm) of the leg. The principle of treatment of congenital pseudarthrosis of the tibia (CPT) with the Ilizarov method corrects all angular deformities and maximizes the cross-sectional area of the pseudarthrosis. Nineteen patients with a total of 19 CPT were treated using the Ilizarov apparatus. CPT is the most perplexing challenging pediatric orthopaedic problem especially when the patient of below years of age and has a history of the previous operations and tibia narrow and osteoporotic. Therefore, the purpose of this study was to evaluate the results of treatment of CPT by the Ilizarov method. **Material & Methods:** The prospective study was done from July 2008 to June 2010 at the National Institute of Traumatology and Orthopedic Rehabilitation, Dhaka. Various forces were used to treat the pseudarthrosis site including compression, distraction, open reduction, resection and shortening, resection and bone transport, and a temporary intramedullary k-wire given through calcaneum and talus into the tibial medullary cavity. **Results:** Lengthening was performed in all except one of the 19 patients. One patient had developed nonunion in both the pseudarthrosis site and proximal corticotomy site. The deformity was tried to correct in all cases. The union rate was 94.73% with one treatment. There were three early re-fracture. Eight patients had a persistent residual deformity of ankle valgus from 5-9 degrees and five patients had residual angular deformity at the pseudarthrosis site from 5-10 degrees. **Conclusions:** One patient's angulation degree required revision surgery, Ilizarov. All patients were given Previous sites, residual angular deformity, and natural history were considered predisposing factors for re-fracture. Two re-fractures united with months (range 10 months). This technique produced initial pseudarthrosis with the correction associated with deformity inequality. angulation and valgus.

Keywords:- Evaluation, Treatment, Congenital Pseudarthrosis, Tibia, Ilizarov method.



INTRODUCTION

Congenital pseudarthrosis of the tibia (CPT) refers to the nonunion of a tibial fracture that develops spontaneously or after trivial trauma on a dysplastic bone segment of the tibia diaphysis. The pseudarthrosis usually develops during the first two years of life; however, there is a strong association between CPT and neurofibromatosis Type (NF-1). CPT develops in about 5.7% of patients with NF-L. On the other hand, 40% of patients with CPT were found to have (NF-1). In my study, out of 19 patients, 11 patients had signs/symptoms of CPT (about 58%). Electron microscopy and histopathological studies showed that the main pathology of CPT is hyperplasia of fibroblasts with the formation of dense fibrous tissue. This invasive fibromatosis is located in the periosteum and between the broken bone ends and surrounds the tibia causing compression, osteolysis, and persistence of pseudarthrosis. CPT remains one of the most perplexing and challenging Orthopaedic problems. This is because of the difficulty of obtaining union and, in cases where none of the proposed classification systems is based on an accurate concept of the cause, natural history and treatment result. The difficulty in treating this condition occurs because of two factors. The first is biological: the poor healing ability of the dysplastic segment of bone. The second factor is mechanical: technical difficulty to fix small and osteopenic bone fragments in children without damaging the distal tibial physis or ankle joint. A multitude of treatment protocols have been described and have varying degrees of success. Most of these treatments focused on stimulating the healing process by using different bone-grafting techniques. The graft materials most

commonly used included osteoperiosteal graft, massive Onlay graft, autogenous iliac crest bone graft and vascularized bone or metal plates, intramedullary (IM) rods of different types, external skeletal fixation, and lastly external skeletal fixation augmented by IM rod. Morrissy, Risborough, and Hall reported union is less than 50% of 40 patients treated with 172 bone grafting procedures.^[1] The most recent and viable option for the treatment of CPT is the Ilizarov technique. In a multicentre study conducted by EOS (Grill et al) the Ilizarov technique is shown to be the optimal method of treatment with a fusion rate of 75.5% with the best correction of additional deformities like LLD, ankle valgus, fibular nonunion.^[2] Paley et al were the western surgeons to report results of treatment of CPT by the Ilizarov method.^[3] Their series consisted of 16 cases with a union rate of 94% with one treatment and 100% with two treatments and five re-fracture. Ilizarov technique is a comprehensive approach to all aspects of CPT. It simultaneously attends to various aspects of the condition, shortening defects, infections, articular function, weight-bearing and ankle valgus. At NITRO, Dhaka, so far no study on CPT has been carried out. The present Study envisaged evaluation of results of CPT treated by the Ilizarov method. One of the important reasons for the selection of this topic was to work on this new revolutionized surgical procedure of CPT which is the viable optimal method of treatment. The study aimed to find out the CPT most perplexing challenging pediatric orthopaedic problem especially when the patient of below years of age and has a history of the previous operations and tibia narrow and osteoporotic. Therefore, the purpose of this study was to evaluate the results of treatment of CPT by the Ilizarov method.

MATERIAL AND METHODS

The prospective study was done from July 2008 to June 2010 at the National Institute of Traumatology and Orthopedic Rehabilitation, Dhaka. Various forces were used to treat the pseudarthrosis site including compression, distraction, open reduction, resection and shortening, resection and bone transport, and a temporary intramedullary k-wire given through calcaneum and talus into the tibial medullary cavity.

Inclusion criteria:

- Congenital pseudarthrosis of the tibia

Exclusion criteria:

- Fracture due to definite trauma (open or close).
- Pathological fracture due to chronic osteomyelitis.

A pretested and predesigned proforma (datasheet) containing history, examination findings, operative procedure and follow-up was used to collect data. Ethical clearance and permission were taken from the thesis grant committee, NITOR, Dhaka to undertake the present research work. In keeping compliance with Helsinki Declaration for Medical subjects 1964, all patients were explained the study design, the underlining hypothesis and the right for the participant to withdraw from the project at any time, for any reason, whatsoever. The study did not involve any additional investigation or economic burden on the patients. All information gathered from physical and biochemical findings was documented in a performed data collection sheet (questionnaire) according to the above-

mentioned criteria. Statistical analyses were carried out by using the Statistical Package for Social Sciences version 20.0 for Windows (SPSS Inc., Chicago, Illinois, USA) and MS-Excel 2016.

RESULTS

This prospective study of 19 patients with CPT was done with Ilizarov external fixator at NITOR, Dhaka. The period of study was from July 2008 to June 2010. Follow up period was between one month to ten months (an average of 5.64 months). Out of my 19 patients, 18 patients' union was achieved. In one case, the union was not achieved at the pseudarthrosis site and nonunion had developed at the corticotomy site. This patient had neurofibromatosis and the limb was short by 8 cm about the healthy limb. The minimum time for the union was 19 weeks and maximum of 52 weeks and an average of 35.61 weeks. In my study, the angular deformity was fully corrected but 7 patients had 5-10 degree angulations deformity and one patient had an unacceptable angular deformity and required revision surgery by the Ilizarov method. Out of 19 patients, 7 patients had residual ankle valgus deformity ranging from 5-9 degrees and one had 30-degree valgus due to fibular proximal migration (fibular pseudarthrosis). Out of my 19 patients, 11 of the 57.88% suffered pin tract infection at some point in the study and after removal of the fixator, all infections were eradicated. In my study 2 cases, 10.52% had limited ROM ankle and in one case 5.26% had limited ROM knee at final follow-up. In my study 4 of my 19 patients, 21.05% had leg length discrepancy between <2cm LLD. In my study one patient, 5.26% had developed refracture through the united pseudarthrosis site and the patient is still now wearing long leg orthosis.



one patient had developed refracture in regenerate following fall from cot 3 weeks after removal of Ilizarov but the union was achieved by long leg full plaster. Now the patient is walking with KAFO. One patient had a fracture in the distal wire insertion site and united conservatively and the patient is given the

advice to use KAFO till skeletal maturity. In my study, the follow-up period after completion of treatment was only 1to10 months (average of 5.64). So, it is very difficult to ascertain the ultimate fate of the union without long term follow-up.

Table 1: Demographical characteristics of study patients.

Characteristics	Frequency	Percentage (%)
Age		
3-Jan	6	31.58
6-Apr	7	36.84
9-Jul	3	15.79
12-Oct	3	15.79
Sex		
Male	8	42.11
Female	11	57.89

Table 2: Showing mode of onset CPT, overall side of involvement, level of CPT and type of CPT.

Variables	Frequency	Percentage (%)
Mode of onset		
Deformity at birth	6	31.58
Gradual deformity after birth	10	52.63
Trauma (minor)	3	15.79
Side		
Right	10	52.63
Left	9	47.37
Level of CPT		
Proximal 1/3	0	0
Middle 1/3	8	42.11
Distal 1/3	11	57.89
Type of CPT		
Type-I	0	0
Type-II	7	36.84
Type-III	2	10.53
Type-IV	4	21.05
Type-V	6	31.58
Type-VI	0	0

Table 3: Showing overall hospital stay of the patients.

Hospital stays	Weeks
Minimum	2
Maximum	9
Average	5.31

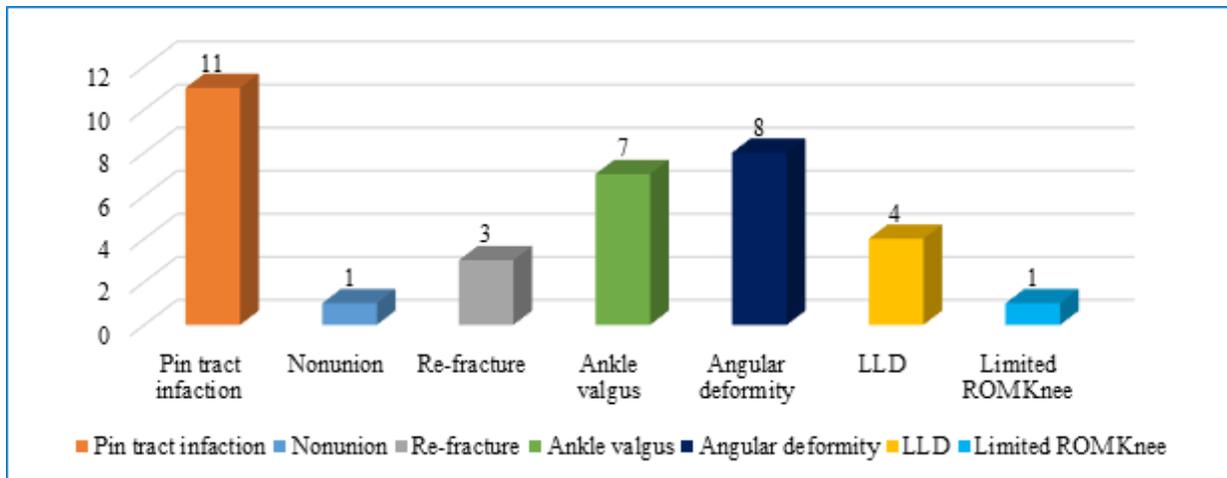


Figure 1: Incidence of complication.

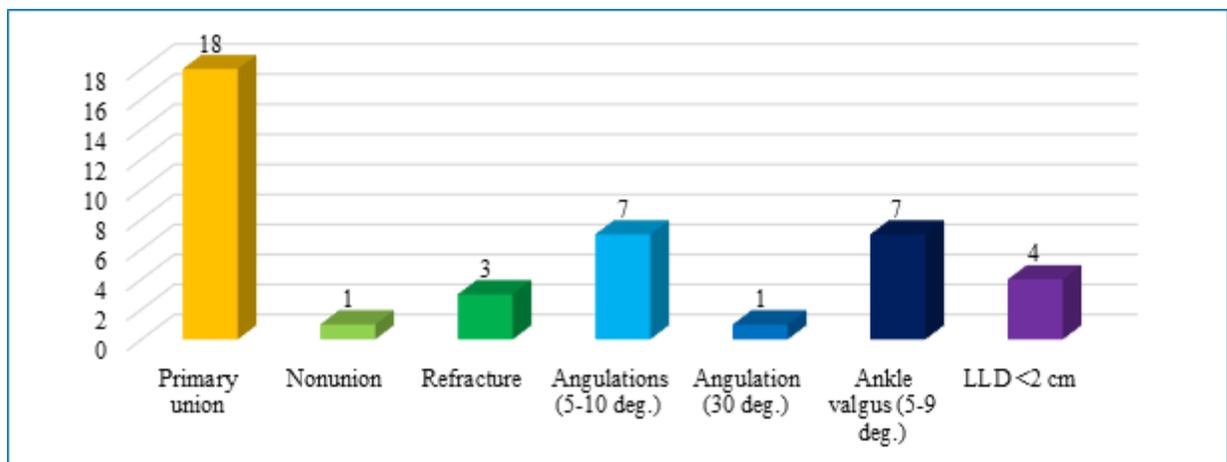


Figure 2: Results of Treatment

Table 4: Showing overall results of treatment

Overall result	Frequency	Percentage (%)
Union	18	94.74
Nonunion	1	5.26

DISCUSSION

Conventional methods of treatment for congenital pseudarthrosis have variable rates of success in achieving union. Morrissey compared different grafting methods: on lay bone grafting had a 12.5% success rate for union; McFarland bypass grafts had a 7% success rate, and the Farmer cross-legged tibial pedicle graft had a 53% success rate.^[4] The increased success of the Farmer graft was attributed to the vascularized biology of the graft. Microvascular techniques allow a more controlled vascularized bone graft transfer, such as the fibula. One of the principles of this type of treatment is to fully resect the bone and its surrounding pathologic soft tissues until normal tissue planes are encountered. This technique relies on hypertrophy of the fibula, which in young patients is reliable. One of the most common problems with this method is nonunion at one end of the fibular transfer. These non-unions are often as recalcitrant as the original CPT. A stress fracture is another potential problem, necessitating prolonged brace protection. Weiland et al. braces patients until skeletal maturity with a long-leg brace.^[5] Donor-site morbidity includes valgus ankle instability. The limb-length discrepancy is not corrected by conventional microvascular fibular transfer (MVFT). Weiland et al recently reported on 19 limbs with CPT treated by MVFT.^[5] This is similar to several patients in the study by Paley D. Catagni M. and others. This is comparable to the number of patients in my study which was 19.^[6,7] Because MVFT is currently accepted as one of the best treatments for CPT, Weiland's study serves as a ready comparison to current alternative methods of treatment. One difference was age distribution:

the mean, was 5.1 years in Weiland's study, eight years in Dror Paley's study and the mean age of my patients was 5.47 years.^[5,8] In the patient's study, Dror Paley had an initial leg-length discrepancy (mean, 4 cm. versus 1.6 cm for Weiland's group and the initial LLD of my study was, a mean of 3.14 (range to 8cm).^[5,8] Dror Paley, the initial union rate of CPT was 100%, compared to 95% in Weiland's series.^[8] There was a 26% (5/19) initial failure rate achieving union in Weiland's series. Four retreated successfully with nine bone-grafting procedures and one with amputation. Dror Paley's early failure rate was 25% (4/16); there was a 6% (1/16) initial failure rate that united on the second treatment and 19% (3/16) early refracture rate, all of which reunited with four further procedures. Weiland et al. had a late refracture rate was 10.5% (2/19) compared with 12.5% (2/16) in Dror Paley's series.^[5,8] The final union rate at follow-up evaluation was 93% in this series and 95% in Weiland et al's study.^[5] In my study, the initial union rate was 94.73% (18/19) and the initial failure rate was 5.26% (1/19) (persistence pseudarthrosis and developed nonunion at corticotomy site). The initial refracture rate of my study was 15.78% (3/18), one at the corticotomy site, one at the distal metaphysis through pin tract site one at the pseudarthrosis union site. But two united with cast except refracture at union site. My only failure case is still under treatment and needs revision surgery and do not know what will be the fate of this patient. The ultimate union, initial, and late failure rates are comparable. However, Prior Paley's series as well as my series has much lower morbidity and complication rate. Of Weiland's cases, 79% (15/19) remained with significant axial malalignment (deformities were 5-45 valgus,

with a median of 25° and a mean of 20° procurvatum).^[5] The Dror Paley's had malalignment of 5-10 in 25 % (5/16) at the lengthening site in four cases and the pseudarthrosis level in only one. These required three procedures for correction. Weiland et al. also had 11% significant donor-site morbidity, including tibial fracture and deformity.^[5] In total, Weiland et al. performed 25 secondary procedures in 14 patients compared to ten secondary procedures in six patients in Dror Paley's series.^[5] My series had malalignment of 5 to 10-degree angulations at pseudarthrosis site in 7 patients (26.13%) and one had 30-degree angulation and required a second procedure for correction. The microvascular fibular transfer is a much more invasive procedure than the application of a circular fixator with or without pseudarthrosis resection. Weiland et al.'s results suggest that it has much higher morbidity with a comparable union rate to the Ilizarov method.^[5] Another currently, used an alternative method of treatment is intramedullary fixation and bone grafting. The results range from 54-to 76% success. The rod is introduced through the calcaneum, talus, and tibia, transfixing the ankle and subtalar joints and the distal tibial physis. Umber et al. recommended dual roding to include the fibula.^[9] Refractures in the unprotected region of the tibia may occur with the growth of the leg as the end of the rod migrates. Stiffness of the ankle and subtalar joints may occur, creating a longer lever arm across the pseudarthrosis site, possibly increasing the risk of refracture. Finally, the distal tibial growth plate, which already has a stunted rate of growth, may be further injured by transfixion. In 1971, Ilizarov and Gracheva first reported the treatment of CPT with circular

external fixation in 16 patients. Union was achieved in all 16 cases in 90-630 days. Between the six- and 12-month follow-up period, five of the 16 refractured. Four were cast and one bone grafted; three were re-healed. In the one- to six-year follow-up period, two additional cases refractured were retreated and healed. The major etiologic factor leading to refracture was residual uncorrected deformity. They conclude that all angular deformities must be eliminated as an essential part of the treatment to avoid refracture. In 1981, Gracheva et al. reported Ilizarov's long-term results with the treatment of 87 cases of CPT.^[10] Seventy-six had been treated with previous surgery. Thirty-seven were stiff (stable) pseudarthrosis and 50 flails (unstable). Seventy-nine cases of these cases (91%) achieved union. There was a 9% initial failure rate to achieve union. At 1.5 to nine-year follow-up evaluation, 59 of 65 cases that were reviewed had persistent union and six patients had stiff pseudarthrosis. There was, therefore, a 9% re-fracture rate. Preoperatively, all had associated deformities ranging from 10-90°. In 74 patients, the deformity was eliminated with treatment, while in 13 cases, a residual of 5-10 cm deformities remained. Eighty-five of the patients had 2-19 cm of limb length discrepancy preoperatively. At follow-up evaluation, 56 patients had no limb length discrepancy, 25 patients had 2-4 cm of limb length discrepancy, four patients had 10 cm. in the cases that failed to achieve union, and the CPT was converted from a flail type of pseudarthrosis to a stiff type of pseudarthrosis, which is more amenable to treatment. Despite careful attention to all of these factors, the natural history of this disease continues to haunt us. There is a subgroup of congenital pseudarthrosis that continues to have re-fractures into adult life. There are several



advantages of the Ilizarov technique for the treatment of CPT. The apparatus used 1.5-mm K-wires and can provide safe fixation of very small bone segments and cross joints without intra-articular fixation. Fixation is even possible to the minute dysplastic fibular characteristic of these limbs. Osteoporosis is not a major obstacle to fixation. The apparatus is also able to apply different forces at different levels of the bone and is, therefore, able to perform a combination of compression and distraction to heal the pseudoarthrosis, correcting of deformity, fill bone defects, and limb lengthening. The Ilizarov method is, therefore, not one technique but a variety of methods that can be used individually or in combinations and that can be modified as the situation demands. One of the most important principles of treatment at the level of pseudoarthrosis that safeguards against re-fracture is the maximization of the cross-sectional area of healing. If closed treatment is chased, the contact area between the ends should already be wide or should be widened by one of several techniques. The simplest method of widening the bone ends is the gradual overlap of two pointed ends with side-to-side compression. This doubles the overall diameter of the union. Alternatively, one or both ends may be spilt, with insertion of one end into the other closed-end. Open or closed reduction with insertion of one end into the other is another means of the wide ring the cross-sectional diameter of healing. A bone graft may have been added at the time of open reduction or after union to augment the volume of bone at the pseudoarthrosis site. If the pseudoarthrosis site is to be respected, then the resection should be radical, including all the surrounding fibrous tissue, until normal soft-tissue planes are encountered. Resection should

extend above and below the level of the sclerosed medullary cavity until a well-vascularized open medullary canal is identified. Computerized tomography and magnetic resonance imaging may be useful in determining the extent of resection. The most advantageous aspect of this methodology is the ability to treat CPT comprehensively. One can address not only the tibial nonunion but also the bone defects, deformity, limb-length discrepancy, fibular nonunion, proximal fibular migration, ankle valgus, and foot contractures. Recalcitrant nonunion, osteoporosis, bone defects, and shortening, all of which are considered indications for amputation by conventional techniques, are not insurmountable problems with Ilizarov's methods. Re-fractures, while they do occur, are retractable using the same technique with equal success as primary cases. Re-fractures may be technically related and, therefore, avoidable if the principles outlined above are followed. The disadvantages of the Ilizarov method are primarily related to the use of an external apparatus. External fixators are less well tolerated by patients than internal fixation devices and are prone to pin-track infections. The pin holes also create a stress riser that may lead to re-fracture. The Ilizarov methods rarely burn bridges. All other conventional treatment modalities are still possible should the Ilizarov method fail. For these reasons, the authors conclude that the Ilizarov methods are currently the most comprehensive yet conservative methods available for the treatment of CPT. Considering the cost-effectiveness of this technique, it is not costly in comparison to other techniques (1/M nail, MVFT). Though these two techniques are recommended for the treatment of CPT but not commonly practised

in our country. So, the only feasible method is the Ilizarov technique. A foreign-made Ilizarov apparatus is costly (around 1, 00000 Taka) but the apparatus made in Bangladesh by a local mechanical workshop is not costly (only 4000-5000 Taka).

Limitations of the study:

The smaller size of the study population with a short duration of follow-up. The low socio-economic condition of our country. So the patient can't come for follow-up frequently. Ignorance of the biomechanics of Ilizarov as well.

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CONCLUSIONS

The Ilizarov method is an effective method of treatment for Congenital Pseudarthrosis of the Tibia. It is a method of choice in achieving union, saving the limb, and correcting the associated deformity. As the outcome evaluation was done for one month to ten months after the operation and the natural history of CPT remains unchanged even after achieving union, the outcome beyond ten months could not be ascertained. So further study should be contemplated considering long-term evaluation. To prevent early re-fracture every patient should be given either orthosis or any kind of intramedullary rod or nail till no maturity is achieved.

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