Treatment of congenital pseudoarthrosis of tibia with Ilizarov external fixator

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Abstract:
Objectives: To determine the outcome of congenital pseudoarthrosis of tibia (CPT) with ilizarov external fixator in terms of stable union.
Study Design: Descriptive study.
Settings: This study was performed on 10 patients in Orthopaedic and Trauma unit, Khyber Teaching Hospital and Khyber Medical Center Dabgari Garden, Peshawar from March 2007 to March 2011.
Material & Methods: Patients of either gender with age below 15 years and with supple ankle and knee joints having congenital pseudoarthrosis of tibia were included in the study excluding those with other associated congenital anomalies and those with acquired pseudoarthrosis of tibia. These patients were followed up to 2 years. Outcome was recorded in terms of solid union at the pseudoarthrosis site by radiography and clinically at time of removal of the ilizarov ring fixator.
Results: Out of 10 patients, six (60%) were female and four (40%) were male. Mean age was 8.5 years (3 to 14 years). The right tibia was affected in 7 cases (70%) and left one in 3 cases (30%). All patients were treated with resection of the pseudoarthrosis site and compression + fixation with ilizarov fixator initially with union achieved in 8 patients (80%). The over all success rate was 90%.
Conclusion: Ilizarov fixator is an excellent treatment modality for the treatment of Congenital Pseudoarthrosis of tibia regarding bone union, deformity correction, limb lengthening procedures and especially in situations when healing has failed to occur despite many previous surgeries.

Keywords: Congenital pseudoarthrosis of tibia, ilizarov, external fixator.

Introduction:
Congenital pseudoarthrosis of tibia is one of the challenging problems in paediatric orthopaedics. Congenital pseudoarthrosis of tibia is not a homogenous entity but is composed of several disease types, with different prognoses. The precise cause of CPT is unclear, even though it appears to be related to neurofibromatosis-1 (NF-1). The natural history of CPT varies and is unpredictable and no surgical and medical option appears to be capable of altering its natural history or pathobiology.

The treatment goals are to achieve bony union without axial and rotational malalignment, stabilization of ankle mortise by fibular stabilization and lower limb length equalization. In atrophic type CPT, these goals are extremely difficult to accomplish. However, primary union appears to have been improved markedly by modern treatment methods such as, intramedullary fixation, microvascular transfer of fibular graft, and external fixation using ilizarov technique.

The Ilizarov method was introduced in the western world during the late 1980s, and has been
widely popularized and applied to the treatment of CPT, because it can address pseudarthrosis as well as all components of the complex deformities associated with this condition. Further more, in contrast to other treatment modalities, the Ilizarov technique is not precluded by previous surgery, and the Ilizarov device can be reapplied in the event of refracture.4,6,16 This technique is particularly beneficial when other methods have failed or in patients with angulation threatening refracture, shortening exceeding 5 cm, late symptomatic ankle or proximal tibial valgus, proximal fibular migration, or procurvatum producing a severe calcaneus-type gait.20 In addition, because the Ilizarov technique rarely burns bridges,21 all other conventional treatment modalities are still possible if this technique fails.2 If necessary, a combination of two or three techniques can be used sequentially or simultaneously in an effort to obtain union. For example, a free vascularized fibular graft can be combined using the Ilizarov technique, thereby offering the advantage of addressing both bone loss and leg length inequality.1,22 For these reasons, many surgeons agree that the Ilizarov method is a viable surgical alternative. However, it is not the ultimate solution for all types of CPT.1

A variety of techniques based on several different frame configurations and strategies have been reported for the Ilizarov method.1,5-7,18,19,23,24 Although the primary aim of treatment is union of the pseudarthrosis site, the therapeutic considerations should include other associated problems, such as leg-length discrepancy, multilevel and multidirectional tibial deformity, foot deformity, associated fibular pseudo-arthritis and subsequent ankle valgus.1,23,25,26 According to the literatures produced by retrospective case series, the overall union rate for the Ilizarov technique ranges from 60 to 100%.1,21 However, the union rate and function after an Ilizarov treatment can be confounded by a variety of factors, such as the severity of dysplasia, failed previous treatments, and co-existing deformities.1

Materials and methods:
This study was conducted in the department of orthopedic and trauma unit of khyber teaching hospital and khyber medical center Dabgari Garden, peshawar from March 2007 to March 2011. All the patients of age below 15 years and either gender with supple ankle and knee joints presented with congenital pseudoarthrosis of tibia were included in the study. Patients with age more than 15 years, with other associated congenital anomalies and those with acquired traumatic pseudoarthrosis of tibia were excluded from the study. The patients were enrolled from out-patients departments. After informed consent, patients were admitted and pre-operative investigations including baseline investigations, plain x-ray (anterioposterior and lateral views) were performed. All the patients with congenital pseudoarthrosis of tibia were treated with ilizarov external fixator under general anesthesia. In supine position under strict aseptic measures the tourniquet was applied and incision was made on the anterior tibia directly over the pseudoarthrosis site. After incision the sclerotic bone at pseudoarthrosis site was resected to the point of bleeding bone on both ends. The ends were then closely opposed to each other and the first transverse wire was passed proximal to the fracture site and then ring was fastened after tensioning the wire either with wire tensioner or manually with spanners at both ends simultaneously on plain wires and only opposite end on olive tip wires and the other wires were passed at least 45° to the first wire. Then second construct was made distal to the pseudoarthrosis site. The usual distance between constructs proximal and distal to fracture was 2 – 3 cm. Then another constructs were made proximal and distal to the previous constructs respectively and tensioned similarly and then fastened with threaded rods and compression was applied to both fragments. Sometimes, we also used drops wires and attached them with the help of posts to rings. All the rings were larger of 2-finger breaths to diameter of leg over anterior aspect and 3-finger breaths to posterior aspect of leg. For each ring, minimum of 2 wires were used. While inserting the wires, they were first gently pushed upto the bone through skin and then drilled with power drill. As soon as they come out through other cortex, they were hammered gently to get out
to other side. Muscles were at their maximum length while inserting the pins and all the wires were passed through safe zones. The wire sites were dressed with hydrogen per oxide and pyodene solution soaked gauzes.

Post-operative management:
On return from the operation theatre, patients were allowed with partial weight bearing walking on next day and with full weight bearing in a week time as tolerated. Parenteral antibiotics were given for upto 3-4 days and with parenteral and oral analgesia. Check x-rays were done on next day and any adjustments were made on 2nd or 3rd day if required. Patients were trained for daily wash of fixator, pins, pin care and mobility of joints and quadriceps strengthening exercises.

Follow-up in OPD:
In the follow up in OPD, fixators were checked thoroughly, each and every nut and bolt tightened, wires tensionized if needed, pin sites were cleaned with pyodine solution and hydrogen per oxide and washed thoroughly with normal saline. If there was any deep pin tract infection, or pin loosening, the patients were readmitted for surgical toilets and treated according to culture and sensitivity report isolated organisms and readjustments of fixators respectively. Follow up were made on every 14th day in first month and then once a month until solid union has been achieved at the pseudoarthrosis site. Check radiograph were performed in two planes every time.

Before removal, fixators were dismantled first without anesthesia and fracture site was examined for movement and tenderness. If there was any tenderness and mobility at fracture site, fixators remained in situ for more 3-4 weeks. All the fixators had been removed without anesthesia as an out-patient procedure. First, the distal rings were removed and the wires were taken out with help of chuck and then similarly proximal construct removed. All the pin site wounds were washed with saline and pyodine soaked gauzes.

Results:
Out of the 10 patients, six were female (60%) and four (40%) were male (Figure 1). Mean age was 8.5 years (3 to 14 years). The right tibia was affected in 7 patients (70%) and left one in 3 patients (30%) (Figure 2). The mean fixation duration was 8 months. All the patients were treated initially by thorough resection of the pseudoarthrosis site and compression and fixation with ilizarov ring fixator alone. Good radiologic and clinical result was achieved in 8 patients (80%). In the remaining two patients, a second surgery was performed with resection of the sclerotic bone ends and packing the non-union site with autologous iliac crest bone grafting with good radiologic union achieved subsequently. Four patients developed complications, two patient with pin site infection alone. Out the remaining two patients one patient had refracture alone and one had refracture at the union site with pin track infection as well. Out of the two patient with refracture, union was achieved in one patient with intramedullary rod fixation and ilizarov ring fixator with iliac crest bone grafting. One patient ultimately end with ampu-
tation of the affected leg. The average leg length discrepancy was 2.5 cm which was compensated with shoe raise. In our study, good results were achieved mainly in patients with age more than 5-6 years with overall success rate of 90%.

**Discussion:**

Currently, vascularized fibular grafting, intramedullary stabilization and external fixation are being used with relative success for the treatment of CPT.5,10,19,23 The superiority of any specific procedure is difficult to determine. The different types of pseudo-arthrosis, the effects of previous treatment, the need for follow-up until skeletal maturity, the timing of the procedure and the definition of what is a successful result all make it difficult to compare the outcome of treatment in different series.28

Numerous authors have reported on the use of the Ilizarov circular external fixator in the treatment of CPT.5,15,16,23 The Ilizarov technique is a comprehensive approach to all aspects of CPT. It simultaneously attends to various aspects of the condition, including resection of the pseudoarthrosis, deformity correction, shortening defect, infections, articular function, weight bearing and the valgus ankle. The disadvantages of this technique are the duration of treatment, its complexity, pin tract infections, ankle valgus and refractures.28

Our results (80% with Ilizarov fixator alone and 90% with combination treatment) are comparable to result of study of Boero et al and Paley et al whose union rate were 81% and 94% with one treatment and 100% with two treatment in contrast to ours of 80% with one treatment and 90% with two treatments respectively.23

Grill et al, in a multicentre study, analyzed the different therapeutic methods used by the European Paediatric Orthopaedic Society and had the results as nearby to ours.27 There treatment data of 340 patients who underwent 1287 procedures for CPT were analyzed. The therapeutic modalities which were reviewed included the McFarland bypass graft, plating, rodding and grafting, the Ilizarov fixator as well as conservative measures. The findings of that study demonstrated that plating and rodding seemed to afford inadequate stability to allow the pseudoarthrosis to heal, and that surgeons who used that kind of fixation resected too little of the pseudoarthrotic bone in an attempt to avoid shortening. The results of that study also showed that the Ilizarov technique was the method of choice in the treatment of CPT. In addition to success in correction of the other deformities, this method achieved the highest rate of union (75.5%).27,28

In another study by Ghanem et al,7 the mean fixation duration was 7.8 months and union was achieved in 10 out of 14 patients (71.4%) with compression and use of Ilizarov fixator in 7 patients and in 3 out of remaining 6 patients with use bone graft as well but all with out excision of pseudoarthrosis site. The mean fixation duration in our study was 8 months.

In our study, two patient failed to achieve union and two patients had refracture similar to others studies despite apparently solid clinical and radiological union.5,6,10,13,22,23 There is consensus that surgery should be avoided before the third year of life, and if possible, it should be postponed until the age of five years10,27 and we had also good result with those patients aging 5-6 years.

The Ilizarov technique is useful in many cases of CPT in which union failed to occur inspite of many previous surgeries. The use of this method does not preclude the use of other procedures. The Ilizarov method takes considerable time and effort to obtain good results. The surgeon must know when to abandon this procedure and

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**Table 1: Outcome**

<table>
<thead>
<tr>
<th>Treatment</th>
<th>Number of patients</th>
<th>Union</th>
<th>Non-Union</th>
<th>Amputation</th>
</tr>
</thead>
<tbody>
<tr>
<td>Resection + Compression + Ilizarov fixator</td>
<td>10</td>
<td>08(80%)</td>
<td>02(20%)</td>
<td>_</td>
</tr>
<tr>
<td>Resection + Compression + ICBG + Ilizarov fixator</td>
<td>02</td>
<td>02(100%)</td>
<td>_</td>
<td>_</td>
</tr>
<tr>
<td>Resection + Compression + IMR + Ilizarov fixator for Refracture</td>
<td>02</td>
<td>01(50%)</td>
<td>01(50%)</td>
<td>01</td>
</tr>
</tbody>
</table>

IMR= Intramedullary rod fixation, ICBG= Iliac crest bone graft.
perform an amputation which will make the patient more functional. 26

Amputation, although undesirable, is often an effective means of treating congenital pseudarthrosis of the tibia. 29 Indications for amputation vary from case to case. Some indications as stated by McCarthy 30 are (a) failure to achieve bony union after three surgical attempts; (b) significant leg-length discrepancy (> 5 cm); (c) interference with growth distal to the pseudarthrosis causing a non-functional foot; and (d) significant suffering by the patient from repeated operations and hospitalizations. In our study, one patient had amputation after the failure of three surgeries.

Conclusion:
Ilizarov fixator is an excellent treatment modality for the treatment of congenital pseudoarthrosis of the tibia regarding bone union, deformity correction, limb lengthening procedures and especially in situations when healing has failed to occur despite many previous surgeries.

References: