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ORIGINAL ARTICLE

Outcome of Congenital Pseudoarthrosis Tibia Treated with Illizarov Fixator in Children

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ABSTRACT

OBJECTIVES: To determine the outcome of congenital pseudoarthrosis tibia (CPT) managed with illizarov fixator.

METHODS: This prospective cross-sectional study was done from 20th March 2008 to 19th March 2018. All patients presenting to the Orthopaedic out patient department (OPD) with pseudoarthrosis tibia belonging to either gender aged up to 12 years were included. All patients underwent excision of tapering ends and illizarov fixator application. Outcome was assessed in terms of complications. Data regarding age, gender, side, type of CPT and any complication was noted and analyzed using SPSS version 23.

RESULTS: 16 patients were included in the study. 10 (62.5%) were males and 6 (37.5%) females. Mean age was 4 years. 10 (62.5%) patients were type-1 and 6 (37.5%) were type-2. Evidence of neurofibromatosis was seen in 9 (56.2%) patients. Most common complications were residual leg length discrepancy (LLD), mal-alignment and delayed consolidation which was seen in 8 (50%) patients.

CONCLUSION: CPT is a periosteal disease with variable results. The study concludes that good results are achieved in 50% patients.

Key Words: Congenital pseudoarthrosis tibia; Illizarov fixator; Leg length discrepancy; Neurofibromatosis

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INTRODUCTION

Congenital pseudoarthrosis tibia (CPT) that may be obvious at birth or incipient, most commonly involves the distal half of tibia and often fibula in the same limb. Incidence is 1 in 250,000 live births[1]. CPT is closely associated with neurofibromatosis that may not be the cause of pseudoarthrosis. Variable forms of fibrous dysplasia are also reported with CPT[2]. True cause of poor healing at pseudoarthrosis is unknown, however few of the cases mentioned in literature are hamartomatous thickening of fibrous tissue, limited vascular potential, thickening of adherent periosteum and osteolytic fibromatosis[3].

Natural history of CPT is neither predictable nor alterable with surgical and medical treatment. The treatment aims are to achieve

bony union at pseudoarthrosis, restore leg length discrepancy (LLD), prevent mal-alignment and to address ankle and foot problems[4]. Different methods have been used to achieve the goal like intramedullary fixation, free fibular graft and illizarov technique and at times a combination of the above surgeries[5]. Type of surgery to achieve union and prevent refracture remains controversial. Initial management is resection of entire pseudoarthrosis and surrounding hamartomatous tissue[6]. Atrophic type of CPT is more challenging. Excision of the fibrous hamartoma, removing sclerotic tapering bone ends to sufficient cross sectional tubular bone, axial mal-alignment and ankle stabilization. Osseous union is difficult to achieve having a success rate of 31 to 56 percent[7]. Union obtained may be temporary with 50% incidence of refracture[8]. Secondary surgery for knee, ankle, foot, mal-alignment and LLD increases the frustration of surgeon and agony of patient to end up with amputation of limb[9]. Involved limb is already short, removal of pathological tissue and tapering ends of bone further increases the limb-length discrepancy[10].

Illizarov fixator is used for bone lengthening, compression at fracture site after generous excision of pathological bone and soft tissue[1]. Valgus foot and ankle deformity is addressed with the same apparatus. Illizarov became available to us in early 1990's. Author experience with apparatus was boosted by 2006 earthquake. Illizarov can address pseudoarthrosis, LLD and foot component problems.[1] This technique is also useful when other modalities are exhausted. As there is scarcity of literature on CPT management and outcome, so this study was undertaken to determine the outcome of congenital pseudoarthrosis tibia (CPT) managed with illizarov fixator.

MATERIAL AND METHODS

This prospective cross-sectional study was done from 20th March 2008 to 19th March 2018 in the Department of Orthopaedics Benazir Bhutto Hospital and Holy Family Hospital, Rawalpindi, Pakistan. All patients presenting to the Orthopaedic out patient department (OPD) with pseudoarthrosis tibia belonging to either gender aged up to 18 years were included. Pre-existing infected CPT patients were excluded. Patients were assessed between 2008 to 2018 in Benazir Bhutto Hospital and Holy Family Hospital, which are attached hospitals of Rawalpindi Medical University. CPT was classified based on Boyd classification. All patients underwent excision of tapering ends and illizarov fixator application by the same consultant orthopaedic surgeon.

Illizarov procedure: Patients below the age of 5, only 3 rings were accommodatable; we used 1.5mm K-wire in these patients. Specially made sling for foot support was used made by the orthotic and prosthetic department, Rawalpindi Medical University, Rawalpindi. Hamartomatous tissue and tapering ends of bone were excised, till good bleeding bones at least 70% of the shaft diameter was gained to increase contact surface area. Attempt was made to do compression at the fracture site, to avoid kinking of the vessel, a gap more than 4cm between ends per operation compression was avoided. Later this gap was reduced to 3cm as one patient of 2 years age got kinking of the vessel; which relieved after reopening the gap. Osteotomy of the proximal tibia with predrilling of osteotome was used. Distraction in younger children started at the 8th day and in older, 10th to 12th day. 1mm/day distraction in 0.25mm/6 hourly. For large bone defect, speed was reduced to 0.75mm/day. We kept the frame till good evidence of union at distraction and non-union site. For accurate approximation of middle and distal fragment, we used wire from heel to the middle fragment to rail road the middle fragment. Data was recorded on pre-formed proformas. Data analysis done using SPSS version 23.

Table 1 Complications.

S.No.	Complication	No. of patients	Percentage
1	Pin track infection	5	31.25%
2	Loosening, breakage and refracture of pin insertion site	4	25%
3	Neurovascular injury	1	6.25%
4	Temporary kinking of vessel	1	6.25%
5	Valgus deformity	3	18.75%
6	Residual LLD	8	50%
7	Refracture	4	25%
8	Non-compliance with bracing	10	62.50%
9	Diaphyseal malalignment	8	50%
10	Delayed consolidation	8	50%

RESULTS

A total of 16 patients were included in this study, 10 (62.5%) were males and 6 (37.5%) females. Age ranged from 2 to 17 years, with a mean age of 4 years. According to Boyd classification, 10 (62.5%) patients were type 1 and 6 (37.5%) were type 2. Evidence of neurofibromatosis was seen in 9 (56.2%) patients. LLD before surgery was seen in all patients and varied from 2 to 10 cm. 10 (62.5%) patients underwent primary surgery while 6 (37.5%) patients had already undergone more than two surgeries.

Most common complications were residual LLD, mal-alignment and delayed consolidation which was seen in 8 (50%) patients (Table 1).

Bone grafting was required in 3 (18.75%) patients. 1 patient aged 7 years had gained 1.5cm more limb length than the normal limb (Figure 1).

1 patient had unexplained resorption of the proximal bone at the site of osteotomy and distraction resulting in proximal pseudoarthrosis as well Figure 2).

Another patient had a grossly deformed tibia at the age of 17 years. This patient was operated in early childhood with free non-vascularized fibular graft (Figure 3).

DISCUSSION

No standard surgical method for CPT has been defined. Basic treatment goal of these patients is to achieve union, prevent refracture, to correct LLD and correction of deformities particularly ankle joint[1]. Different modes of treatment are clinically practiced like intramedullary fixation illizarov alone, illizarov with intramedullary fixation and free vascularized fibular graft[12]. Age at which CPT should have surgery is quite controversial[13]. Joseph et al believes surgery is beneficial at the age of 3 to 4 years while others has recommended the age of 5 to 6 years as having better bone stock. Children below the age of 5, bones are not only difficult to unite as low cross section area of the bone and most of them have early fractures[14].

Illizarov technique has achieved union as well as addresses LLD, angular deformities and ankle deformities. This procedure has gained acceptance in many centers. Grill et al considered this procedure as gold standard for the treatment of CPT[15]. Cho et al reported 20/23 patients with refracture. Late apical mal-alignment may develop even after union[16]. One case of 11 year old female apparently normal looking proximal tibial bone after osteotomy and distraction had osteolysis instead of achieving bone length. Different explanation for pseudoarthrosis of distal tibia has been explained like thickening of periosteum, thickened and coherent periosteum which consists of

Figure 1 7 year old CPT case.

Figure 2 Unexplained resorption of the proximal bone at the site of osteotomy and distraction resulting in proximal pseudoarthrosis.

Figure 3 Grossly deformed tibia at the age of 17 years after early childhood surgery for CPT.

bone causing bone atrophy, fracture and pseudoarthrosis. Excision of hamartomatous tissue is considered as a part of treatment, which itself may not be surety of union and refracture after union. Pathology do not seems to be only localized to distal third of tibia as has been explained in osteolytic fibromatosis[1]. Chao *et al* believes the periosteal cell has higher osteolytic activity than osteoblastic activity which may cause failure of bone healing and cause resorption of bone[16].

In children below 5 years, we were able to apply only three rings, middle ring for distraction, one ring in lower fragment of tibia may cause anterior angulation of distal-most fragment as foot goes into planter flexion. We use sling for foot made by school of orthotics and prosthetics, Rawalpindi Medical University to prevent planter flexion. A K-wire from distal tibia to middle fragment from heel as a guide so that distraction fragment hit the distal fragment at an accurate position. Resection of sclerotic ends of pseudoarthrosis is usually done in treatment of CPT. Removing more tapered bone ends assume better cross sectional bone, better mechanical stress, chances of union and less chances of refracture[17]. Choe et al observed narrow tibial shaft unite with wide proximal metaphysis has increased refracture regardless of age[16]. Excision of more bone will increase bone gap more distraction will be required which itself become risk of fracture at distraction site. Excision of hamartomatous tissue and atrophic bone, compression of both fragments, causes early union and avoid problem of distal and middle fragment anatomical reduction. One case we have kinking of the vessel causing pallorness of foot. It was luckily diagnosed on the table. So the distraction was given and vascularity was restored but edema of the foot remained for many weeks. The high incidence of refracture, low cross sectional area of the united segment with valgus deformity at ankle causing a stress riser and compliance issue with discarding brace earlier[18]. Usually children are young with narrow medullary cavity, intramedullary device alone may not be sufficient. We have to relay on external bracing. In our series, we have not supplemented patients with intramedullary device. Once residual LLD has been restored, to achieve union of the distal fragment, we had to use a bone graft in two cases. Repeated traction is not indicated. Proximal migration of the distal fibula with valgus mal-alignment of ankle is seen 7/15 by Agashe et al. They suggest correction of the ankle valgus if greater than 5 degree is suggested by Inon et al.[12] BMP-2, BMP-7 and rh (recombinant human) BMP-2 has been used has not shown the promised clinical response, possible explanation is lack of osteoblastic differentiation in response to BMP. The fibrous hamartoma excision is considered as essential part of treatment[19]. After meticulous excision of hamartomatous tissue and atrophic bone ends, achieving union at fracture site, correction of LLD by distraction histogenesis, union and consolidation of distraction fragment may not be predictable due to ill understood disease process. Refracture of the distraction fragment

or the fracture site make it more complicated. Good results with these patients are only 50%.

CONCLUSION

CPT is still a poorly understood disease perhaps primarily a periosteal disease with decreased osteoblastic activity and increased osteoclastic activity hence making the results unpredictable.

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