Original Article

Short Term Results of a New Comprehensive Protocol for the Management of Congenital Pseudarthrosis of the Tibia

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Abstract

Background: Treatment of Congenital Psuedarthrosis of Tibia (CPT) often poses significant challenges due to difficulty in achieving union and subsequent complications like refractures, implant failures, etc. Our new comprehensive protocol is aimed at achieving crossunion between the tibia and fibula. **Aims and Objectives:** The aim of the present study is to evaluate the short-term results of our new protocol and to compare the results with our previously used techniques. **Materials and Methods:** 10 patients with mean age 2.35 years (1 to 6.5 years) who were treated by our new comprehensive protocol were included in Group A, and 11 patients with mean age 2 years (1 to 5.5 years) who primarily underwent intramedullary rodding with bone graft were included in Group B. The new comprehensive protocol consisted of pre-operative Zolendronate infusion, surgery consisting of intramedullary fixation of tibia supplemented with Ilizarov ring fixator and bone grafting aimed at achieving tibia-fibula cross-union. Retrospective evaluation of serial radiographs was performed and outcomes with respect to union and subsequent complications were analysed. **Results:** 10/10 (100%) patients in Group A united, whereas union was achieved in only 8/11 (72%) patients in Group B. The index surgery was successful in achieving union in all 10 patients in Group A, whereas in Group B 2.25 (1 to 4) surgeries were needed to achieve union. The time to union was significantly shorter in Group A (4.68 months) as compared to Group B (30.88 months). The cross sectional area of union was significantly greater in Group A (3.82 cm2) as compared to Group B (1.18 cm2). One patient in Group A needed a subsequent corrective osteotomy for tibial valgus, and one patient underwent tibia lengthening; whereas in Group B, two patients needed corrective osteotomes for residual malaligments. **Conclusion:** Our study demonstrates that the new comprehensive protocol is extremely effective for achieving sound union in Congenital Pseudarthrosis of Tibia.

Keywords: *Congenital psuedarthrosis of tibia, ilizarov ring fixator, intramedullary rodding, neurofibromatosis, tibia fibula cross union*

Introduction

Congenital pseudarthrosis of the tibia (CPT) is a rare disorder affecting 1 in 140,000 children.[1,2] About 55% of children with CPT have underlying Neurofibromatosis Type 1.[1,3] CPT is one of the most difficult orthopedic disorders to treat due to significant difficulty in achieving union. Often multiple surgeries are needed for achieving union and even after union, complications including refractures, tibial and ankle malalignments, implant failures, etc., are fairly common.[4-9] Surgical modalities described for the management of CPT include fixation with intramedullary (IM) rods, external fixators, onlay grafts, cancellous grafts, periosteal grafts, vascularized fibula grafts and induced membrane techniques, in various permutations and combinations. The primary union rates (union after first index surgery) of various techniques range from 22% to 100% [Table 1].[9-22]

In 2011, Choi *et al*. published the results of a new surgical technique called four-in-one osteosynthesis and reported a primary union rate of 100% in 8 patients, with no refractures at a mean followup of 7.4 years.[19] Similar results were reported by Paley with his cross-union protocol.[20] Both Choi and Paley's protocols aimed at achieving cross union between the tibia and fibula, thereby increasing the cross-sectional surface area of union. We combined components of Choi's and Paley's protocols and devised a protocol suited to our patient population, which we called the new comprehensive protocol.

The aims of the present study are to evaluate the short term outcomes of the

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Table 1: Comparison of outcome parameters with previously published studies using different methods

	Treatment method	Number of patients	Primary union (%)	Final union (%)
Joseph and Mathew[10]	Rodding	26	21 (80)	24 (92)
Johnston[11]	Rodding	21	18 (86)	19 (90)
Dobbs *et al*.[12]	Rodding	23	5 (22)	20 (86)
Grill *et al*., EPOS[13]	Ilizarov	28	28 (100)	26 (93)
Borzunov *et al*.[14]	Ilizarov	108	61 (56)	82 (76)
Paley *et al*.[15]	Ilizarov	16	15 (94)	16 (100)
Agashe *et al*.[16]	Ilizarov + rodding	15	6 (40)	14 (93)
Yan *et al*.[17]	Ilizarov + rodding	51	46 (91)	47 (92)
Kalra and Agarwal[18]	Vascularised fibula graft	26	20 (79)	24 (93)
Choi *et al*.[19]	Four-in-one	8	8 (100)	8 (100)
Paley[20]	X-union protocol	17	17 (100)	17 (100)
Our series	New comprehensive protocol	10	10 (100)	10 (100)

EPOS=European paediatric orthopaedic society

new protocol mainly with respect to primary union rate and to validate its efficacy by comparing with the results of our previously used techniques.

Materials and Methods

Patients who were diagnosed and treated for CPT at our institute with minimum of 1 year followup after the index surgery were included. Index surgery was defined as the first surgical intervention performed at our institute. This was a retrospective analysis of data collected by a standardized proforma from our hospital records. Hospital review board and ethics committee approval was obtained before the commencement of the study. Patients with incomplete data sheets and radiographs and those with <1-year postsurgical followup were excluded from the study.

We studied 21 patients of operated CPT at our hospital from 2003 to 2016. Of these, 11 patients who were operated from 2003 to 2014, in whom the index surgery consisted of IM rodding with onlay grafting were included in Group B. Remaining 10 patients who were operated from 2014 to 2017, and who were treated by the new comprehensive protocol were included in Group A.

The new comprehensive protocol

Preoperative

Zolendronate infusion is administered 4 weeks before the surgery at a dose of 0.02 mg/kg intravenously slowly. Calcium gluconate at 60 mg/kg is given IV slowly 1 hour after the zolendronate infusion. Calcium at 2 g/day and Vitamin D at 400 IU/day are thereafter supplemented orally for 2 weeks.

Surgical technique Group A

The affected tibia is exposed through an anterior curvilinear incision [Figure 1a]. The hamartomatous periosteum is radically excised circumferentially [Figure 1b]. The posterior tibial neurovascular bundle is protected by careful dissection between the pathological periosteum and the attached musculature. The sclerotic bone edges at the pseudarthrosis site are excised and freshened, and the medullary canal is opened proximally as well as distally. Fibula is exposed through the same incision after careful elevation and lateral retraction of musculature, anterior tibial vessels, and deep peroneal nerve off the anterolateral surface of the tibia and interosseous membrane. The intervening interosseous membrane is excised. Hamartoma if present at the fibular site is excised. If the fibula is not affected, fibula osteotomy is done at the same level. The tibia is then fixed with an IM rod with transfixation of the ankle and subtalar joints. The fibula if dysplastic is fixed with an IM k-wire.

Bone graft is harvested from three sources as follows: (1) Onlay cortical graft from the contralateral tibia (2) a 3 cm × 3 cm rectangular cortical graft from inner table of iliac bone, and (3) cancellous graft obtained from between the two tables of ilium and the supra-acetabular area above the tri-radiate cartilage [Figure 1c]. The iliac crest cortical graft is then placed along the posterior surface of the tibia and fibula after careful retraction of the posterior tibial neurovascular bundle [Figure 1d]. The cancellous graft is placed in the interosseous space [Figure 1e] and tibial cortical graft is placed along the anterior surface after the careful elevation of the anterior tibial vessels and deep peroneal nerve laterally. The bony barrel thus formed is tied around with two Ethibond No 2 cerclage sutures [Figure 1f and g]. Surgical wounds are then closed.

An Ilizarov ring fixator is then applied to provide compression and rotational stability at the pseudarthrosis site [Figure 2a-c]. A foot extension frame is added if the distal fragment is very short.

Postoperative protocol

Postoperatively, the Ilizarov ring fixator is maintained till fracture union and one dose of injection Zolendronate is repeated at the time of fixator removal (3–5 months post-op). Union is considered to be achieved when there is continuity between the bony trabeculae of the proximal and distal segments in at least three cortices

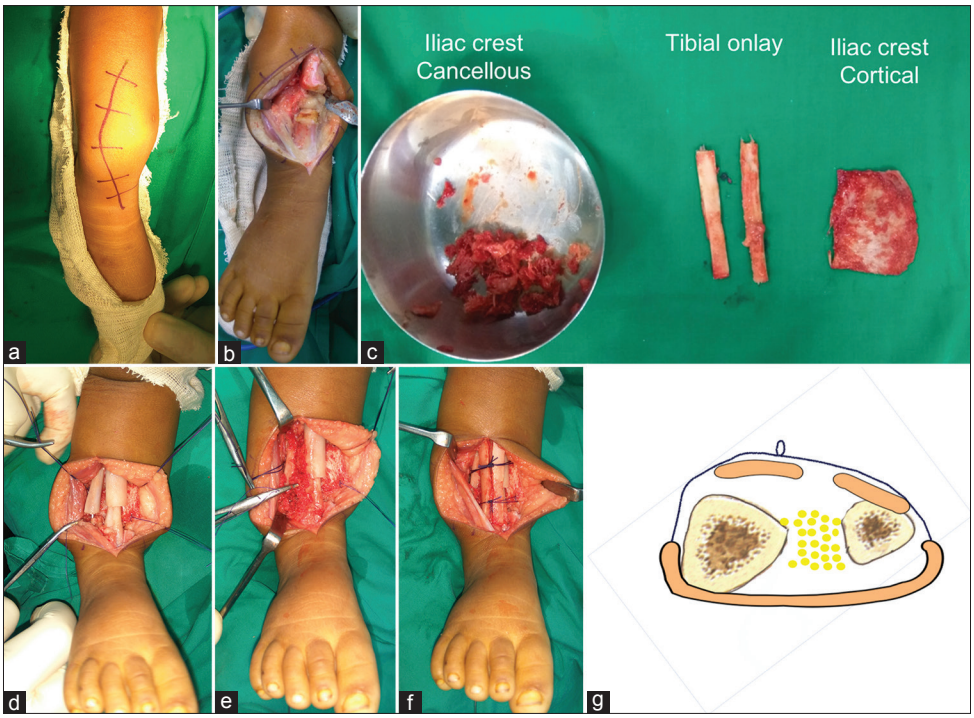

Figure 1: (a) Operative steps: Anterior curvilinear incision to expose both tibia and fibula. (b) Ciricumferential excision of hamartoma at pseudarthrosis site. (c) Harvested bone graft, cancellous bone from iliac crest, onlay cortical graft from contralateral tibia, rectangular cortico-cancellous graft from lilac crest. (d) Rectangular cortico-cancellous graft placed along posterior surface of tibia and fibula after intra-medullary fixation. (e) Cancellous graft placed in tibia-fibula interosseous space. (f) Onlay graft placed along anterior surface of tibia, bony barrel thus formed tied with Vicryl suture. (g) Diagrammatic representation of circumferential placement of bone graft

in anteroposterior and lateral views [Figure 2d and e]. Following fixator removal, an above knee cast is applied for 1 month. After cast removal, mobilization is permitted after fitting the limb with a clam-shell type Knee-Ankle-Foot-Orthosis. The intramedullary rod is retained for biomechanical stability till skeletal maturity but may be exchanged for longer rod later when the proximal nail tip recedes to the middiaphysis of the tibia with growth.

Outcome analysis

The outcome measures evaluated included tibia pseudarthrosis union rate, primary union rate (defined as union after index surgery), interval between index surgery and union, and number of surgeries needed to achieve union. The status of tibia-fibula cross-union and fibula union was also assessed. Four-in-one union was said to be achieved when all four fragments, i.e., proximal and distal tibia, and proximal and distal fibula were united in one fusion mass. Three-in-one union was said to be achieved when proximal and distal tibia and either proximal or distal fibula were united in one fusion mass, Two-in-one union denoted tibial pseudarthrosis union with no cross-union between the tibia and fibula. The cross-sectional area of union was calculated as a product of the minimum diameter of the fusion mass on antero-posterior and lateral views. Details of surgical interventions performed in both groups were noted. Late outcomes including incidence of refractures; knee, tibia, and ankle alignment in coronal and sagittal planes; and limb length discrepancy at final followup were also assessed.

Complications were categorized as minor or major. Minor complications were defined as complications or adverse effects that did not require any additional surgery or those that required only minor procedures needing daycare admissions. Major complications were defined as those requiring surgeries necessitating indoor admissions and those that adversely affected the final surgical outcome.

Statistical methods

Normalcy of data was determined based on the Shapiro–Wilk test and graphical plots.

To assess demographic data, Fisher's exact test was used. For co-relation of outcome parameters between the two groups, an unpaired *t*-test with the level of significance kept at 95% confidence intervals ($P = 0.05$) was applied. The Mann–Whitney U-test was used to assess the nonparametric data.

A paired *t*-test was used to assess the significant difference between the two groups in the preoperative and postoperative outcome variables. The statistical significance was kept at 95% confidence intervals ($P = 0.005$). Kendall rank correlation coefficient was used to measure the association.

The software used for the analysis was SPSS version 20 (IBM Corp, Armonk, NY).

Results

Demographic data

Of the 21 patients, there were 10 patients in Group A and 11 patients in Group B. The two groups were well matched with respect to age at index surgery, sex, the incidence of underlying neurofibromatosis type 2, type of CPT, incidence and type of associated fibula dysplasia (Choi's classification),[19] and number of previous surgeries (before the index surgery). Followup period was significantly longer in Group B as compared to that of Group A [Table 2].

Details of surgical interventions

The index surgery in all patients in Group A was as per the new comprehensive protocol. Rush rod was used for IM fixation in 6 patients and elongating Fassier-Duval nail was used in 4 patients. Subsequent to the index surgery, no patient in Group A needed further surgical interventions to achieve union. In one patient in Group A, in whom a nonelongating Rush rod was used for IM fixation, at 2 years followup, with growth, the tibial rod fell short and had receded below the midpoint of the tibial diaphysis with bowing proximal to the nail tip. This patient needed a surgical intervention consisting of corrective osteotomy proximal to the nail tip, bone grafting, and plating. The osteotomy healed uneventfully in 3 months [Figure 3]. Another patient in Group A underwent tibia lengthening 22 months after the index surgery.

In all 11 patients in Group B, the index surgery consisted of IM rodding with onlay grafting. Three patients remained ununited till the last followup. The index surgery succeeded in achieving union in two patients. In 3 patients, 2 surgical interventions were needed; in 2 patients, 3 surgeries were needed; and in one patient, four surgeries were needed to achieve union. In addition, five surgeries were performed for secondary complications after union: One tibial osteotomy for tibial malalignment, one supramalleolar osteotomy for ankle valgus deformity, one limb lengthening with Ilizarov, and two exchange nailings for bent or broken implants [Table 3].

Tibia and fibula union

In Group A, tibia union was achieved in 10/10 (100%) patients. The time to union and fixator removal was 4.68 ± 0.38 months (3.0–5.0 months). In 6 of these, 4-in-1 union was achieved. In 1 patient, the union achieved was 3-in-1, with persistent fibula nonunion distal to the fusion mass. In 3 patients, the tibia pseudarthrosis united, but there was no cross-union with the fibula and fibula nonunion persisted. Fibula united in all 4 patients with Grade A1/A2 fibula dysplasia, whereas in patients with Grade B1/B2/B3 fibula dysplasia, fibula union was achieved in 2 out of 6 patients.

In Group B, tibia union was achieved in 8/11 (72%) patients. Out of these fibula union was achieved in only one patient with Grade A1 fibula dysplasia. Fibula failed to unite in the remaining 10 patients (7 Grade A2, 2 Grade B1, and 1 Grade B3 fibula dysplasia).

Patients in Group A fared significantly better than patients in Group B with respect to the rate of primary union, the

Table 2: Demographic data

	Group A (new protocol)	Group B (old technique)	*P*
Age at index surgery (years)	2.35±1.67 (1.0-6.5)	2.0±1.39 (1.0-5.5)	0.52 (NS)
Classification (Crawford types)			
III	2	2	0.45 (NS)
IV	8	9	
Association with NF-1	4 out of 10	6 out of 11	0.505 (NS)
Associated fibula dysplasia (Choi's classification)	8 out of 10 A1=2 A2=2 B1=1 B2=4 B3=1	10 out of 11 A1=1 A2=7 B1=0 B2=1 B3=2	0.604 (NS)
Sex distribution			
Male	4	7	0.395 (NS)
Female	6	4	
Number of previous surgeries (before index surgery)			
0	6	10	0.07 (NS)
1	3	1	
2	0	0	
3	1	0	
Total followup (months)	21.60±6.83 (12-31)	77.73±47.60 (29-176)	0.002 (S)

S=Significant, NS=Not significant, NF=Neurofibromatosis

Table 3: Details of surgical procedures

	Group A (new protocol)	Group B (old group)
Index surgeries	New protocol=10	IM rod+OBG=11
Tibia fixation	Rush rod=6	Rush rod=9
	Fassier Duval nail=4	Fassier Duval nail=2
Fibula fixation	5/10	2/11
Revision surgeries for union	Nil	IM rod+BG=7
		BG=3
		IL+BG+lengthening=3
Surgeries to union	1	2.25+1.04 (1-4) *P*=0.000001
Revision surgeries after union for secondary problems	Osteotomy + BG + plating=1	Tibia osteotomy=1
		Supramalleolar osteotomy=1
	Tibia lengthening=1	Tibia lengthening=1
		Exchange nailings=2

IM rod=Intramedullary rod, OBG=Onlay bone graft, BG=Bone graft, IL=Ilizarov ring fixator, Ost=Osteotomy

success rate of index surgery, time to union, and number of surgeries needed to achieve union. The cross-sectional surface area of union was significantly better than in Group B [Table 4].

Limb length discrepancy

At final followup, there was no statistical difference in the Limb Length Discrepancy of the two groups. The tibial shortening averaged 3.22 ± 2.44 cm (range 0–8) in Group A, and 3.61 ± 4.47 cm (range 0.50–12) in Group B. However, the tibial shortening was partially offset by overlengthening of the ipsilateral femur in both groups: 0.83 ± 0.71 cm (range 0–2) in Group A and 1.39 ± 1.32 cm (range 0–4) in Group B [Table 4].

Alignment

In Group A, one patient needed correction for a tibial valgus deformity of 15° that developed proximal to the nail tip as it receded to the level of middiaphysis with

Table 4: Outcome parameters

	Group A	Group B	*P*
Rate of union	10/10	8/11	0.21 (NS)
Rate of primary union	10/10	2/11	0.00015 (S)
Type of union			
4 in 1	6/10	0/11	0.00021 (S)
3 in 1	1/10	0/11	
2 in 1	3/10	8/11	
Fibula union	6/10	1/11	0.0003 (S)
Time taken to union/fixator removal (months)	4.68±0.58 (3.0-5.0)	30.88±18.75 (24.0-36.0)	0.000404 (S)
Number of surgeries to union	1	2.25±1.04 (1-4)	0.000001 (S)
Cross-sectional area of union (cm^2)	3.82±0.58 (2.25-4.18)	1.18±0.19 (0.88-1.44)	0.0005 (S)
Knee, tibia and ankle malalignment at final followup			
MPTA	91.1±3.2	89.1±4.7	0.30 (NS)
PPTA	91.00±7.07	85.73±4.24	0.07 (NS)
LDTA	89.9±6.08	88.27±6.83	0.57 (NS)
Tibial alignment AP view	−2.85±5.5	−1.64±16.04	0.82 (NS)
Tibial alignment lateral view	5.3±7.9	8±19.10	0.68 (NS)
Limb length discrepancy (cm)			
Preoperative	2.05±0.36	2.14±0.45	0.27 (NS)
Postoperative	2.85±2.43	2.1±3.57	

MPTA=Medial proximal tibial angle, PPTA=Posterior proximal tibial angle, LDTA=Lateral distal tibial angle, S=Singnificant, NS=Not significant, AP=Anteroposterior

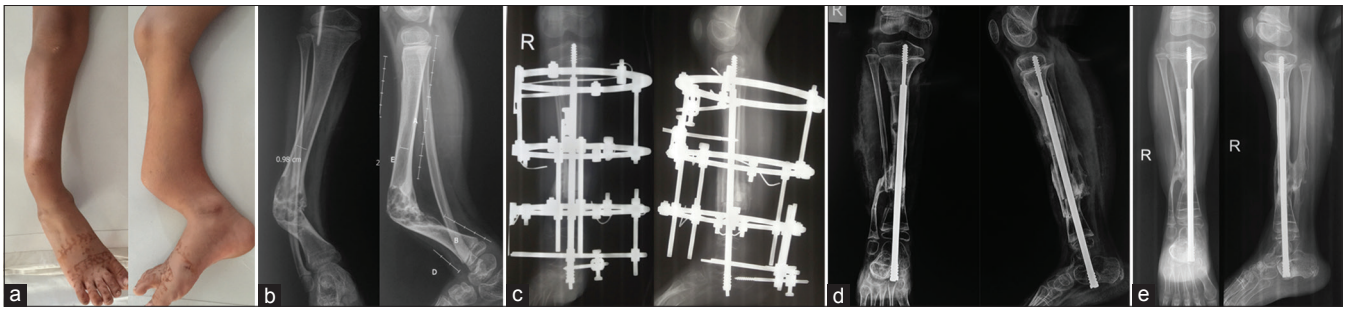

Figure 2: (a) 4-year-old boy, preoperative clinical picture. (b) Cystic type of congenital pseudarthrosis of the tibia. (c) Immediate postoperative. (d) At 6 months postoperative, tibia-fibula cross-union achieved. (e) At 1.5 years postoperative, solid consolidation of union

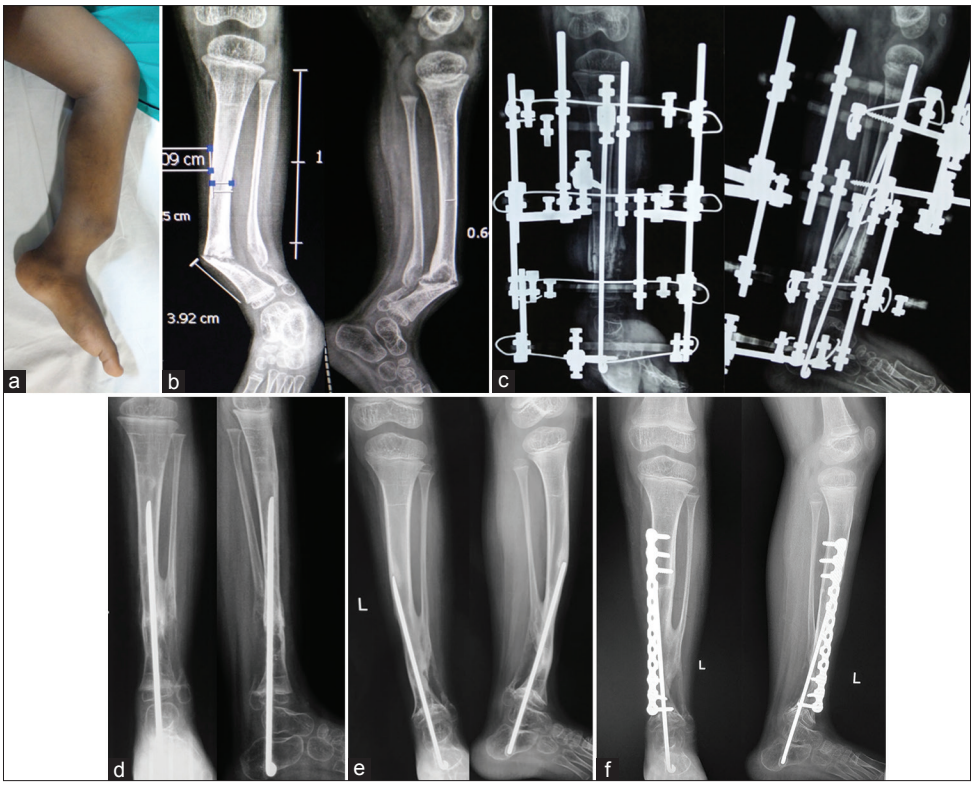

Figure 3: (a) 3-year-old girl with neurofibromatosis and congenital pseudarthrosis of tibia. (b) Atrophic type congenital pseudarthrosis of the tibia. (c) Immediate postoperative, surgery as per new comprehensive protocol. (d) At fixator removal, cross-union between tibia and fibula achieved. (e) At 1.5 years postoperative, tibial diaphysial valgus proximal to nail tip. (f) After tibia osteotomy, bone graft and plating

growth 24 months after the index surgery. In this group, at final followup, medial proximal tibial angle was mildly increased in 5 patients but did not exceed 95° proximal tibial valgus in any. The posterior slope of tibial plateau was decreased in 8 patients with the posterior proximal tibial angle (PPTA) exceeding 84°. At final followup, one patient had residual tibial valgus of 15°; and 3 patients had recurvatum deformity in the range of 10–20°. 4 patients had ankle valgus with lateral distal tibial angle (LDTA) in the range of 79–85°, and 1 patient had ankle varus with LDTA of 99°.

In Group B, during the followup period, one patient underwent supramalleolar osteotomy for ankle valgus deformity, whereas one patient underwent corrective osteotomy for tibial valgus deformity. In this group, at final followup, 7 patients had decreased posterior sloping of tibial plateau with PPTA in the range of 84–94°. Three patients had tibial diaphyseal varus, 2 had diaphyseal valgus, 1 had procurvatum and 1 had recurvatum, 3 patients had ankle valgus with LDTA measuring 75 and 84°, and 1 patient had ankle varus with LDTA measuring 100° [Table 4].

Complications

There were two cases of pin site infection in Group A and one surgical site infection in Group B, both of which settled with IV antibiotics. In Group B, there was one broken rush rod and one bent FD nail, for which exchange nailing was done. There was no incidence of refractures in either group.

Discussion

Achieving union in congenital pseudarthrosis of the tibia poses a major surgical challenge and often multiple surgeries are needed for union. The key pathology in CPT is the fibrous hamartoma at the pseudarthrosis site which does not undergo osteoblastic regeneration in response to BMP. The low osteoblastic activity and high osteoclastogenesis is the main reason for recurrence and failure of conventional treatment techniques.[4,23]

The basis of successful surgical treatment relies on three principles: Excision of the hamatomatous periosteum, stable fixation, and bone grafting. The various options for grafting have been dual onlay bone grafts of Boyd and Sage,[5] bypass graft by McFarland,[24] cortical onlay grafting,[25] vascularized bone graft,[21] wrapping bone graft,[26] and induced membrane technique.[27,28] Various options for fixation include internal fixation with plate and screws, internal fixation with IM nail (fixed or telescopic), and external fixation. Hardinge[29] found high rate of failure in fixation with plate and screws. Risk factors for poor results include a younger age at surgery, a low cross-sectional area of the healed segment, persistent fibular pseudarthrosis, residual ankle valgus, removal of an IM rod, and noncompliance with bracing. Union rates after first surgery

with various techniques described have conventioanlly ranged from 22% to 100% [Table 1].[9-13,15-18]

In their series of 26 patients treated by IM rodding and contralateral tibial onlay graft, Joseph and Mathew achieved union in 24 (92%).[10] However primary union, i.e., union after the first index surgery was attained in only 21 patients (80%). One additional bone grafting procedure was sufficient to achieve union in 3 patients. The European paediatric orthopaedic society multicenter study reported the results of various surgical techniques in 340 patients with CPT, and reported union rates of 27% with plating, 31.3% with plating and bone grafting, 28% with rodding, 50.5% with rodding-bone grafting, and 75.5% with Ilizarov ring fixator.[13] Gilbert and Brockman[12] treated 29 patients with vascularized fibula transplantation and achieved a primary union rate of 58.6% with an average of 2.9 surgeries (maximum 10). Our initial results with the conventional technique of pseudarthrosis excision-IM rodding and onlay cortical bone graft were disappointing with primary union seen in only 2 out of 11 patients (18%). Although the eventual union rate was 72%, the average number of surgeries needed to achieve union was 2.25. The average time from index surgery to union was 30 months resulting in significant morbidity.

Choi *et al*. described the four-in-one technique so called because it aimed to amalgamate four fragments, namely the proximal and distal tibia, and proximal and distal fibula into a single fusion mass.[19] In other words, the technique aimed to achieve cross union between the tibia and fibula by placing a rectangular corticocancellous graft along the under surface of the tibia and fibula at the level of the pseudarthrosis and filling the interosseous space with cancellous bone graft. In addition to IM rod fixation, they applied an Ilizarov ring fixator to stabilize the entire construct. The primary union rate in 8 patients treated by this technique was 100% and refracture rate at a followup of 3.5 years was 0%. These results were significantly superior to the results of techniques previously described in literature. On the other hand, in Choi's series, in 5 patients treated by techniques aimed at achieving end-to-end fibula osteosynthesis or distal tibio-fibular fusion, the authors reported a refracture rate of 80% at 2.7 years followup. The authors attributed the superior results of the four in one technique to the increased cross sectional area of bone contact available for union, thereby improving the primary union rates and also minimizing late complications such as refracture and proximal migration of the fibula with resultant ankle valgus.

Similar results were also reported by Dror Paley with the cross-union protocol. In Paley's protocol, instead of the cortico-cancellous graft, the author wrapped the tibia-fibula at the pseudarthrosis site with a meshed periosteal graft harvested from the inner iliac table.[20] Paley also emphasized the importance of augmenting the surgical treatment with biological treatment by administering intravenous bisphosphonate infusion in the pre and postoperative period and intraoperatively injecting the interosseous cancellous graft with Bone morphogenic protein.

At our institute, we combined components of both Choi's and Paley's protocols to make our own protocol which we called the new comprehensive protocol. This protocol was designed keeping in consideration the socioeconomic profile of the patient population visiting our institute.

The rationale for biological treatment with bisphosphonates is based on the results of previous studies which have established the role of the hamartomatous periosteum in preventing union at the pseudarthrosis sites. Cho *et al*. showed that periosteal cells in the hamartoma of CPT have decreased osteoblastic responses to BMP-2.[23] In contrast the osteoclastic activity of the periosteum was significantly higher than that of controls. They concluded that failure of healing as well as resorption of bone graft is related to increased osteoclastic activity and decreased osteoblastic activity of the CPT periosteum compared to normal periosteum. Schindeler *et al*. showed that NF1(+/-) (NF positive) mouse cells had less osteogenic potential than NF1(+/+) (NF negative) cells (controls). There was much less bone formation in response to BMP in the NF positive cells compared to the NF negative cells.[30] This suggests that inhibition of osteoclastogenesis with bisphosphonates may have a synergistic effect when combined with BMP for the stimulation of osteoblastic activity. Cost constraints rendered the use of BMP unfeasible in our patient population. In the new comprehensive protocol, biological treatment consisted of bisphosphonate (injection zolendronate) intravenous infusions, 4 weeks before the surgical intervention and repeated at fixator removal.

The choice of implant for IM fixation of the tibia needs due consideration. In the initial 6 cases treated by the new comprehensive protocol, we used nonelongating Rush rods to fix the tibia. However, with tibial growth these nonelongating rods soon fall short, necessitating rod exchange. In our first case treated by the new comprehensive protocol, at 2-year followup where the Rush rod had receded below the level of middiaphysis the patient developed tibial bowing proximal to the nail tip which required corrective osteotomy, bone grafting and plating. To prolong the interval between successive rod revisions, we now use telescopic Fassier-Duval nails for tibia fixation.

It would also be desirable if the nail used for tibial IM fixation could achieve a stable fixation of the short distal fragment. This would then permit the nail to stop short of the ankle joint thereby sparing ankle mobility. In Choi's as well as Paley's series, the authors have achieved this objective by using the latest generation Fassier-Duval nails which have the provision of interlocking by inserting a K wire through an aperture at the tip of the male rod in the distal tibial epiphysis. Seo *et al*. demonstrated satisfactory

preservation of foot and ankle function at skeletal maturity if ankle mobility is preserved and fibula stabilization is achieved after treatment.[31] Unfortunately, we did not have access to the latest generation nails and had to use the indigenously available Fassier Duval nails which do not have interlocking provision. Hence to achieve a stable fixation of the distal fragment, we had to perform a transcalcaneal fixation across the ankle and subtalar joints thereby sacrificing ankle mobility. Transfixation of the tibiotalar and subtalar joints is necessary to obtain adequate purchase in the often atrophic and short distal tibial segment. Ease of introduction and easier revision also make transfixation a feasible option.[25] A smooth rush rod that crosses the distal tibial physis need not cause growth arrest.[10]

Need and nature of surgical intervention of the fibula in CPT is a topic of considerable debate. Johnston and Birch reported a direct association between failure to operate on the fibula and poor outcomes, in terms of persistent tibial pseudarthrosis and ankle valgus.[32,33] Cho *et al*. also reported higher incidence of CPT re-fractures if the fibula remains ununited.[23] In contrast, Shah *et al*. reported 100% CPT union in all 11 patients at skeletal maturity despite not fixing the fibula in any patient.[25] Though the fibula was ununited in 10 patients, they reported ankle valgus in only 5 patients. In our new comprehensive protocol, we fixed the fibula with IM K wire in 5 patients in whom the fibula was dysplastic. However, despite the fixation the fibula remained ununited in 4 patients thus demonstrating a positive correlation between fibula dysplasia and persistent fibula nonunion. In 1 of these patients, the proximal fibula fragment cross-united with the tibia resulting in 3-in-1 union, whereas in 3 patients, there was failure of cross-union with the tibia. These patients merit close observation for refracture due to decreased cross-sectional surface area of the fusion mass, and ankle valgus due to persistent fibula nonunion. In the other 5 patients in which the fibula was not dysplastic, we divided the fibula at the level of tibia pseudarthrosis and allowed the ends to overlap to facilitate end-to-end apposition of the tibia. In all these 5 patients, the fibula united and was incorporated in the cross-union mass. However, with our earlier technique of IM rodding-onlay grafting, fibula union was achieved in only 1 out of 11 patients. In this group, even in the absence of underlying fibula dysplasia, fibula nonunion persisted in 7 indicating that interosseous bone graft placed in new technique has a role to play in fibula union.

Overlengthening of the femur was noted on the ipsilateral side in both groups. This phenomenon has been reported previously. Song *et al*. postulated that femoral outgrowth on the ipsilateral side is a compensatory response to frank pseudarthrosis or it may be a manifestation of underlying neurofibromatosis.[34]

There are a few limitations to our study. Although the new protocol fares significantly better than our previously used techniques in terms of primary union outcome measures, the followup period is too short to draw conclusions with respect to long term outcomes. However, certain observations can be made. Shah *et al*. reported that there is a direct correlation between the short term outcomes of cross-sectional surface area of union and stability of ankle mortise (status of fibula union) with the occurrence of late complications such as refractures, and tibial and ankle malalignments.[23] Since both cross-sectional area of union and fibula union are favorable with the new comprehensive protocol, we hypothesize that the prognosis for long term outcomes is also better. However, this hypothesis needs to be proven on long term followup.

Conclusion

The study demonstrates that the new comprehensive protocol is extremely effective for achieving sound union in congenital pseudarthrosis of the tibia. Longer followup would be needed to comment on the occurrence of late complications.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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