ACTA ORTHOPAEDICA et TRAUMATOLOGICA TURCICA
www.aott.org.tr

Research Article

Congenital pseudarthrosis of the tibia: Results of circular external fixation treatment with intramedullary rodding and periosteal grafting technique

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ARTICLE INFO

Article history:
Submitted 1 July 2019
Received in revised form
23 September 2019
Accepted 27 December 2019

Keywords:
Congenital pseudarthrosis of the tibia
Success probability
Circular external fixator
Periosteal grafting
Intramedullary rodding

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ABSTRACT

Objective: This study investigated the clinical and functional results of treating congenital pseudarthrosis of the tibia (CPT) using the combined techniques of hamartoma resection, periosteal grafting, circular external fixator application, and intramedullary rodding.

Methods: The clinical and radiological data of 17 patients (mean age at the treatment time: 7.6 months (range: 4.6–9.7 months) with CPT, treated by a single surgeon between 1997 and 2017, were retrospectively analyzed. All data regarding surgical interventions, complications, deformity analysis parameters, limb length discrepancy (LLD), ankle joint range of motion, and residual deformities were reviewed. All the patients were followed up at least two years after the last surgical intervention. The mean follow-up time was 8.5 years (range: 2.2 to 15.7 years).

Results: Union was achieved with the index treatment in 15 of the 17 cases (88.2%). The mean age of the patients at the last follow-up visit was 14.2 years (range: 7.6 to 22.1). The mean LLD was 2.1 cm. Nine patients had radiological ankle valgus at the last follow-up. In the entire series, eight patients did not display any complications, four cases reported minor complications, and five cases were complicated by refractures.

Conclusion: Circular external fixator application combined with periosteal grafting is a superior method of CPT treatment. This method provides a healthy biological healing environment while correcting the mechanical problems. The combination of periosteal and cancellous bone grafts with intramedullary rods and an external fixator addresses issues that complicate obtaining and maintaining a union during the CPT treatment.

Level of Evidence: Level IV, Therapeutic study

Congenital pseudarthrosis of the tibia (CPT) is a childhood disease causing serious morbidity during its natural course. Treatment of CPT requires solving more than one issue in the affected extremity. The etiology is unknown in the majority of the cases, but neurofibromatosis is present in 40% of the patients (1, 2). However, the level of the affected area differs in each patient (3). Cases with pseudarthrosis before delivery are rare. In the majority of cases, the sclerotic segment of the tibia loses its integrity during the first two years of life (4-6).

Over the years, numerous options have been developed for the treatment of CPT, with nonunion being the focus of the treatment (7-10). Accordingly, grafting and stimulation techniques have been applied. More recently, biological stimulation applications have become popular (11, 12). Fixation is another crucial factor in the treatment of CPT. The use of plates and intramedullary rods has improved the success rates (13). Currently, however, circular external fixator (CEF) provides better fragment control and ensures simultaneous lengthening (14-18).

The popular technique proposed by Ilizarov was gradually augmented through the use of periosteal grafting (12, 14). This study reviews the clinical and functional results of cases treated by a single

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Cite this article as: *Kocaoğlu M, Eralp L, Bilen FE, Civan M. Congenital pseudarthrosis of the tibia: Results of circular external fixation treatment with intramedullary rodding and periosteal grafting technique. Acta Orthop Turkey. Acta Orthop Traumatol Turc 2020; DOI: 10.5152/j.aott.2020.03.26.*

DOI: 10.5152/j.aott.2020.03.26

surgeon using CEF, periosteal grafting, and intramedullary rodding in the treatment of CPT.

Materials and Methods

The medical and radiological data of 24 patients with CPT, treated by a single surgeon between 1997 and 2017, were retrospectively analyzed. Among these, five cases were treated with the bifocal compression-distraction method using the CEF, one patient was treated with nonvascularized fibular grafting, and a two-stage tibia-fibularization case was excluded from the study. The remaining 17 cases were treated with hamartoma resection, periosteal grafting, CEF application, and an intramedullary rod in the same session as the index operation. All the patients were followed up at least two years after the last surgical intervention. At the last follow-up visit, full-length standing lower extremity X-rays were taken to determine residual deformities and limb length discrapancies (LLD). Ankle range of motion (ROM) was also noted.

Of the 17 patients, 12 were males and 5 were females. Furthermore, 13 patients had concomitant neurofibromatosis. Crawford classification was used to classify the cases (6). Nine cases were classified as Crawford type III, and eight cases were classified as Crawford type IV. All the cases presented with LLD, with a mean value of 2.81 cm (range: 1 to 7 cm). In nine cases, the level of pseudarthrosis was in the distal 1/3 of the tibia (52.9%); in eight patients, the level of pseudarthrosis was in the middle 1/3 of the tibia (47.05%). Only three cases had intact fibula, while the remaining cases had an associated fibular pseudarthrosis. Ten patients had at least one previous surgery, and seven patients were followed up with a brace before being referred to the senior author (Table 1).

The mean age of the patients at the time of index operation was 5.6 years (range: 1 to 12.5 years). Bone resections were applied in all cases in the range of 1–3 cm during the surgery. For the resulting bone defects, physeal distraction and/or proximal metaphyseal lengthening was used in three and five cases, respectively. The mean amount of lengthening was 5 cm (range: 3–8 cm) (Table 2).

Surgical technique

The patient was placed in a supine position under general anesthesia on the radiolucent operating table, and the ipsilateral buttock was elevated with a sterile towel. The distal and proximal endpoints of the fibula, joint lines, and planned incisions were marked using a sterile marker under fluoroscopy. An anterior longitudinal incision was used to reach the nonunion site under a sterile tourniquet. The nonunion site, including the hamartoma, was resected (Figure 1). The intramedullary canal of the distal and proximal tibial segments were reamed to a size 0.5 mm larger than that of the inserted Steinman pin (2–3 mm). According to the pseudarthrosis morphology (Crawford type IV), the proximal tibial stump was split into two flaps by performing an incomplete osteotomy using a 10-mm osteotome (Figure 2). A cannulated drill opened the tract for intramedullary rod insertion in the reverse direction of

Table 1. Patient data

Case number	Diagnosis*	Crawford Type	LLD** (cm)	Pseudarthrosis Level	Fibula Pseudarthrosis
1	NF	3	2	1/3 distal	Yes
2	NF	3	1	1/3 distal	Yes
3	NF	4	4	1/3 middle	Yes
4	NF	3	1	1/3 middle	No
5	idiopathic	4	2	1/3 distal	Yes
6	NF	4	1	1/3 middle	No
7	idiopathic	4	5	1/3 distal	Yes
8	NF	3	4	1/3 distal	Yes
9	idiopathic	3	1	1/3 middle	No
10	NF	4	2	1/3 middle	Yes
11	NF	3	4	1/3 middle	Yes
12	NF	4	5	1/3 middle	Yes
13	NF	3	7	1/3 distal	Yes
14	NF	4	2	1/3 distal	Yes
15	NF	4	2,5	1/3 middle	Yes
16	idiopathic	3	2	1/3 distal	Yes
17	NF	3	1	1/3 distal	Yes

NF: neurofibromatosis; LLD: limb length discrepancy the proximal growth plate of the tibia. A Steinman rod inserted from the heel pad to the proximal tibia growth plate was kept proximal to the ankle joint. Alignment could be maintained through the same intramedullary rodding technique after transverse bone resection and acute docking for selected types (Crawford type III). An incision on the iliac crest of the ipsilateral side was performed after the tourniquet was released and hemostasis was completed. Following the exposure

Table 2. Operation details

Case number	Age at surgery (years)	Bone Resection (cm)	Combined Lengthening (cm)	Lengthening Method
1	6.4	1	no lengthening	-
2	4.5	1	no lengthening	-
3	5.4	1	no lengthening	-
4	12.5	1	no lengthening	-
5	3.3	3	5	physeal distraction
6	6.3	1	3	physeal distraction
7	11.2	2	4	physeal distraction
8	6.2	1	no lengthening	-
9	1.3	2	no lengthening	-
10	4.2	1	no lengthening	-
11	2.7	3	no lengthening	-
12	7.8	2	7	metaphyseal corticotomy
13	5.6	1	no lengthening	-
14	0.9	2	4	metaphyseal corticotomy
15	5.3	2	6	metaphyseal corticotomy
16	5.4	3	8	metaphyseal corticotomy
17	7.1	2	3	metaphyseal corticotomy

*im: intramedullary

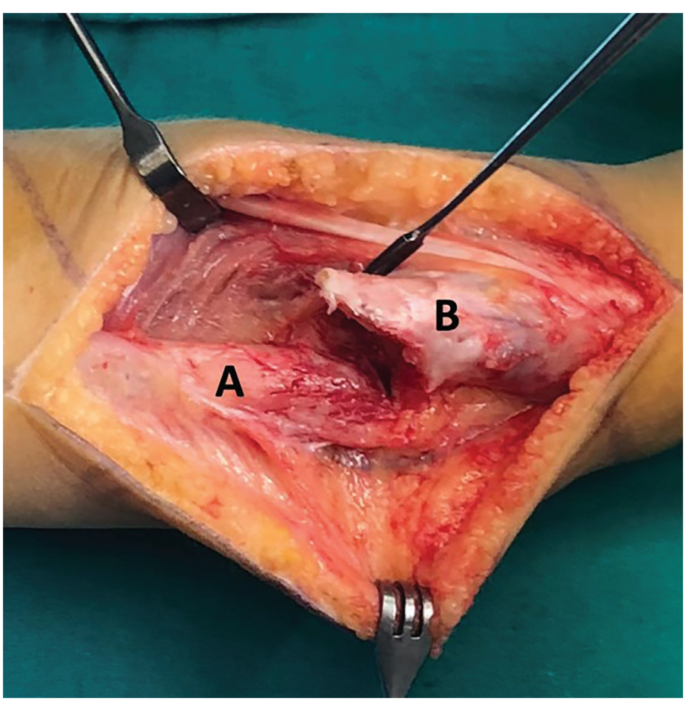

Figure 1. An intraoperative view of the atrophic pseudarthrosis site after circumferential periosteal sleeve excision, with distal (B) and proximal (A) stumps

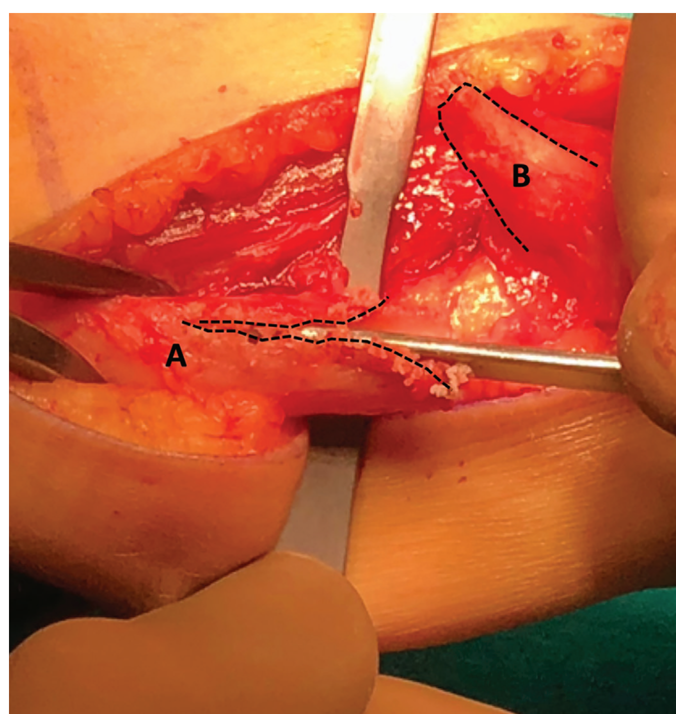

Figure 2. The proximal tibial stump is split into two flaps (A) using an osteotome in Crawford type IV cases

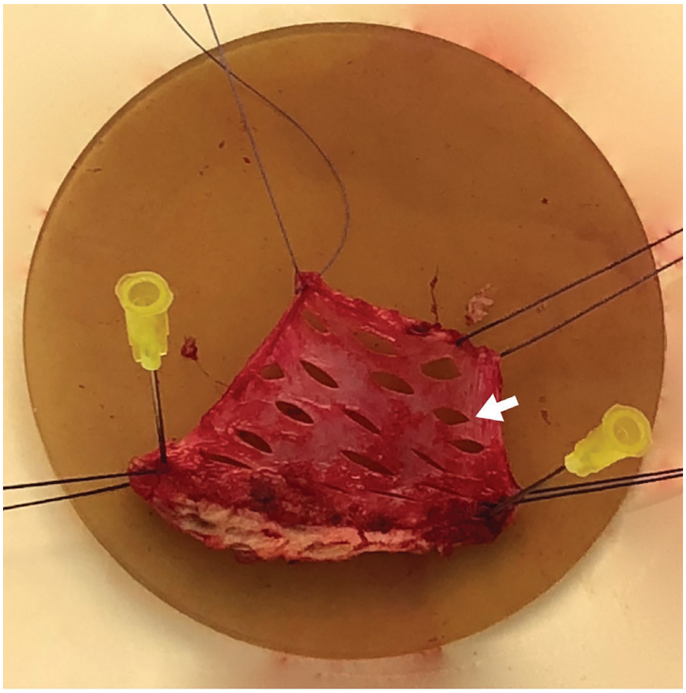

Figure 3. After harvesting a periosteal graft in a rectangular shape, meshing with small separate incisions (white arrow) increases the surface area

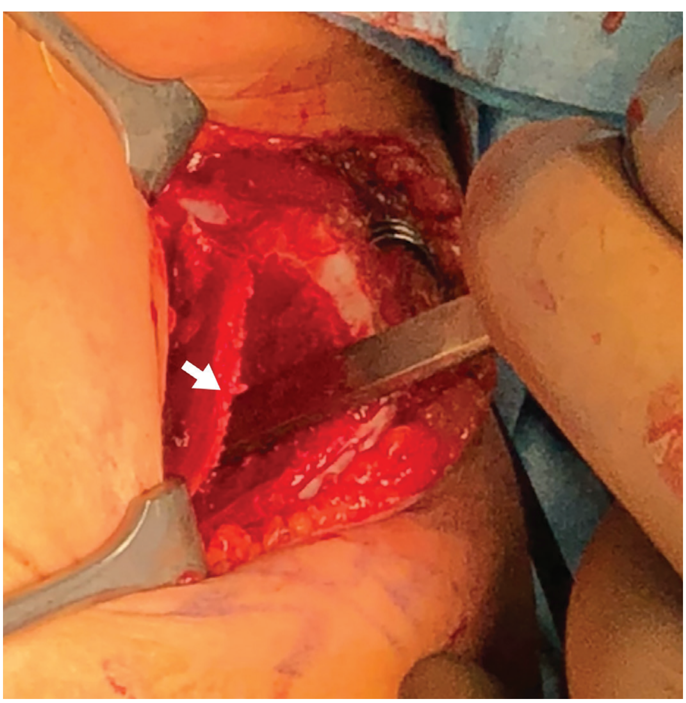

Figure 4. The retraction of the inner cortex of the ilium (white arrow) reveals the cancellous bone. An osteotome is used to harvest the spongiosa of the iliac crest

of the iliacus muscle, the fibers of the muscle were dissected and medially elevated to reveal the underlying periosteum. A scalpel was used to draw a rectangular shape in the periosteum. The periosteum was carefully dissected from the cortical bone after a split incision made to the apophysis. The periosteal graft surface was meshed onto a separate plate to increase the surface area of the graft (Figure 3). The inner cortex of the iliac bone and the spongiosa were harvested (Figure 4). The periosteal graft was wrapped around the nonunion site. Furthermore, the edges were sutured with absorbable suture material after a cancellous graft was used to fill the space between the host bone and the periosteal graft (Figure 5). The soft tissue was closed before the CEF application. The preassembled CEF was proximally and distally fixed, with 1.8 mm K-wires. Two Schanz screws of 5 mm diameter were used for additional fixation at the proximal ring level. One Schanz screw was used for each ring for distal and mid-level ring fixations. A percutaneous low-energy osteotomy was performed at the proximal metaphyseal level in cases with simultaneous lengthening. The proximal tibial epiphysis was fixed with at least two olive wires in cases with physeal distraction. The frame should be constructed, properly aligned, and positioned to allow compression at the pseudarthrosis site and distraction at the level of the proximal lengthening osteotomy or physis for bifocal compression-distraction.

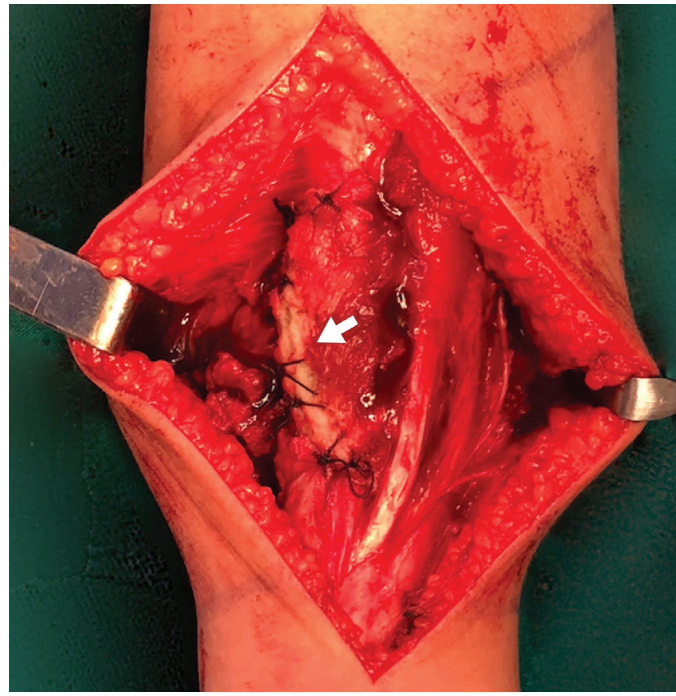

Figure 5. The periosteal graft is wrapped at the level of the interpositioned pseudarthrosis area and sutured with absorbable suture material (white arrow) circumferentially around the docking site

After the index operation, the patients were followed up monthly until fixator removal; thereafter, follow-ups were conducted every six months for two years. All patients planned to attend follow-ups until they achieved skeletal maturity. During the follow-up, pain, limping, and ROM of the ankle joint were assessed and recorded. Radiologically, long-standing X-rays as well as anteroposterior and lateral X-rays of the affected leg were utilized to detect any residual deformities and LLD.

Table 3. Results

Case Number	LLD* (cm)	External Fixator Time (months)	Healing Time (days)	Patient Total Follow Up (years)	Age at Final Follow Up (years)	Ankle ROM+	Pain	Limping	Residual Ankle Valgus	Lateral Distal Tibia Angle (aLDTA)	Refractures
1	3.0	6	251	15.7	22.2	DF=0. PF=20	moderate walking	no	no	87	no
2	4.0	7	216	12.5	17.0	DF=10. PF=20	extended walking	yes	yes	83	no
3	5.0	9	296	12.5	17.5	DF=0. PF=30	none	yes	yes	80	no
4	1.0	9	261	8.9	21.4	DF=10. PF=20	extended walking	yes	no	89	Yes (after 7y)
5	0.0	6	190	13.0	16.4	no motion	none	no	no	90	Yes (after 4y)
6	0.0	4.5	138	10.2	16.5	DF=0. PF=5	none	no	no	91	no
7	3.0	5	267	2.2	13.4	no motion	none	none	no	90	no
8	0.0	5.5	168	13.8	19.9	no motion	extended walking	yes	no	87	no Yes (after 8.6y)
9	3.0	8	245	6.4	7.7	DF=10. PF=20	none	yes	no	87	Yes (after 2y)
10	2.0	2	153	11.7	15.9	no motion	extended walking	yes	yes	79	no
11	7.5	5	no healing	8.6	11.3	no motion	none	yes	no	91	
12	0.0	8	236	2.6	11.3	no motion	none	no	no	88	no
13	8.0	4.5	175	8.5	14.1	DF=10. PF=20	extended walking	yes	no	86	no
14	2.0	4	264	6.7	7.6	DF= 5. PF=10	none	none	no	89	no
15	0.0	7	no heling	3.0	8.3	no motion	none	no	yes	72	
16	3 (o.c.)	8.5	290	3.9	9.3	DF=10. PF=20	none	no	yes	81	no
17	0.0	5.5	263	4.8	11.9	DF=10. PF=20	none	no	yes	80	Yes (after 4.2y)

*LLD: Limb length discrepancy at last follow up visit; +ROM: Range of motion; DF: Dorsiflexion; PF: Plantar flexion

Table 4. Additional surgeries after the index treatment

Case	Successive Surgeries †	Object	Technique	Interval Between Previous surgery (years)	Complications
1	1st Surg	LLD + Genu Recurvatum	Ilizarov method + intramedullary rodding	5.1	Regenerate Fracture
	2nd Surg	Regenerate Fracture	Intramedullary rod revision + casting	1.6	Delayed Union
2	1st Surg	LLD + Tibia Vara	Lengthening and Correction with CAF***	1.4	none
4	1st Surg	Profilactic TEN application	Titanium Elastic Nailing	0.9	Implant failure + Refracture
	2nd Surg	Implant Failure + Refracture	Replication of the Index Treatment with consecutive lengthening	5.9	Delayed Regenerate Maturation
	3rd Surg	Regenerate Maturation Delay + Genu Valgum	Acute Deformity Correction + Intramedullary Nailing	1.3	none
5	1st Surg	LLD + Oblique Plane Deformity	Ilizarov Method + Revision of intramedullary rodding	0.9	none
	2nd Surg	Ankle Malorientation	Supramalleolar Osteotomy	2.6	none
	3rd Surg	Refracture	Intramedullary rodding + casting	0.8	Pseudoarthrosis
	4th Surg	Pseudarthrosis	Replication of the Index Treatment with consecutive lengthening	3.1	none
6	1st Surg	Ankle Malorientation + Tibia Vara	Supramalleolar Osteotomy	2.1	none
7	1st Surg	LLD + Genu Valgum	Frame Transformation for Deformity Correction + Re-corticotomy	1.1	none
8	1st Surg	LLD + Distal tibia sagittal plane deformiy	Ilizarov Method	2.9	Delayed Union
	2nd Surg	LLD + Genu Valgum	Distal femur FAN + Proximal Tibia Correction and lengthening with CAF	3.8	none
	3rd Surg	Refracture	Osteosyntesis with Ilizarov Method	1.9	none
	4rd Surg	Contralateral Ankle Valgus (Donor Site Morbidy)	Contralateral Supramalleolar Osteotomy	2.9****	none
9	1st Surg	Refracture + LLD + Oblique Plane Deformity	Ilizarov Method	1.9	Axial Deviation, Refracture after 2 y FU
	2nd Surg	LLD + Pseudarthrosis	Replication of the Index Treatment with consecutive lengthening	3.1	none
11	1st Surg	LLD + Pseudarthrosis	Transformation to unilateral fixator + DBM grafting	0.5	No Healing
	2nd Surg	LLD + Pseudarthrosis	Vascularized Fibula Transfer	1.4	none
	3rd Surg	LLD	Lengthening with Ilizarov Method	4.9	none
14	1st Surg	LLD + Ankle Valgus	Ilizarov Method	2.3	none
15	1st Surg	Pseudarthrosis	Ilizarov Method	2.0	No Healing
	2nd Surg	Pseudarthrosis + Bone Defect	Vascularized Fibula Transfer	1.0	none
17	1st Surg	Refracture + LLD	Replication of the Index Treatment with consecutive lengthening	4.2	none

LLD: limb length discrepancy; ROM: range of motion; DF: Dorsiflexion; PF: plantar flexion; CAF: computer assisted fixator

**** Donor site morbidity related interval is 12 years 4 months

†1st Surg is considered the next surgical intervention after the index treatment

A small window in the tibial diaphysis was created to remove the Steinman rod. Forceps were used to firmly hold and remove the rod under floroscopy control from the sole of the foot or tibial tuberosity.

Results

Union was achieved with the index treatment in 15 of the 17 cases (88.2%). The mean treatment time was 7.6 months (range: 4.6–9.7 months). In all the cases, a neutral alignment was provided with intramedullary rodding during surgery; however, in case 7, axial deviation experienced by the patient during the treatment was corrected with a frame modification. Nine patients developed ankle valgus, and cases 5, 6, and 14 of these patients had supramalleolar correction osteotomy during follow-up (Table 3).

In cases 11 and 15, the index operations resulted in persistent nonunion, which was treated through free vascularized fibular transfer, using plate and screw fixation. In case 11, the preoperative LLD was 3 cm. Fibular integrity was lost during the treatment despite the preserved alignment of the fibula with intramedullary K-wire. Union was achieved after five months through free vascularized fibula transfer. This procedure was followed-up for 4.9 years without any complications until a final lengthening operation was performed using CEF (Table 4). During the last follow-up, the patient was mobile and functional. In case 15, pseudarthrosis persisted after seven months of external fixator duration. The patient was treated with free vascularized fibula transfer. Future lengthening procedures were planned for the expected LLD for this patient.

The mean follow-up period for patients in this series was 8.5 years (range: 2.2 to 15.7 years) for all surgeries. The mean age of the patients at the last follow-up visit was 14.2 years (range: 7.6 to 22.1 years). In this series, cases 9, 14, 15, 16, and 17) were skeletally immature at the last follow-up visit. In cases 4, 5, 8, 9, and 17, the index treatment technique was complicated by refracture (Table 3). The refractures were treated with a replication of the index treatment in cases 4, 5, and 17 and with the Ilizarov method (8) in cases 8 and 9 (Table 4). Separate consecutive surgeries were required for the following cases: oblique plane deformity and ankle valgus in case 5; tibia bowing and ankle valgus in case 6; an LLD of 6 cm in case 8; equinovalgus contracture of the ankle in case 14. Recorticotomy and frame modification were applied for axial deviation in cases 7 and 9 (Table 4).

The mean LLD at the last follow-up was 2.1 cm. Six patients reported radiological ankle valgus at the last follow-up. Cases 2, 4, 9, 13, 16, and 17 with ankle ROM recovered, with 10 degrees of dorsiflexion and 20 degrees of plantar flexion. Cases 5, 7, 8, 10, 11, 12, and 15 had stiff ankle joint at the last follow-up. Cases 1, 3, 6, and 14 had limited ankle joint ROM, enabling them to perform painless daily activities (Table 3) (Figure 6-8).

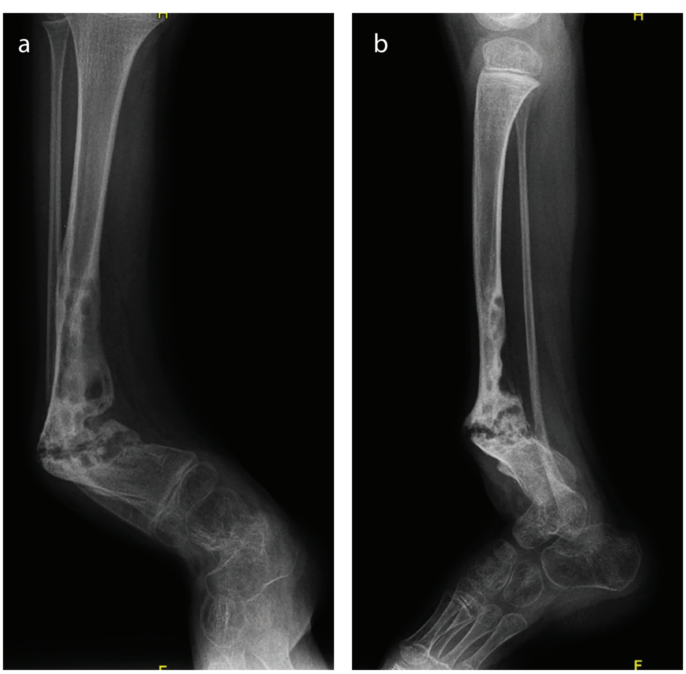

Figure 6. a, b. A case of a six-year-old boy with congenital pseudarthrosis of the tibia. A demineralized bone matrix and cancellous bone grafting was applied, with plate and screw fixation, through a previous surgery at a different center. Preoperative anteroposterior (a) and lateral (b) X-rays indicate the pseudarthrosis site with intact fibula

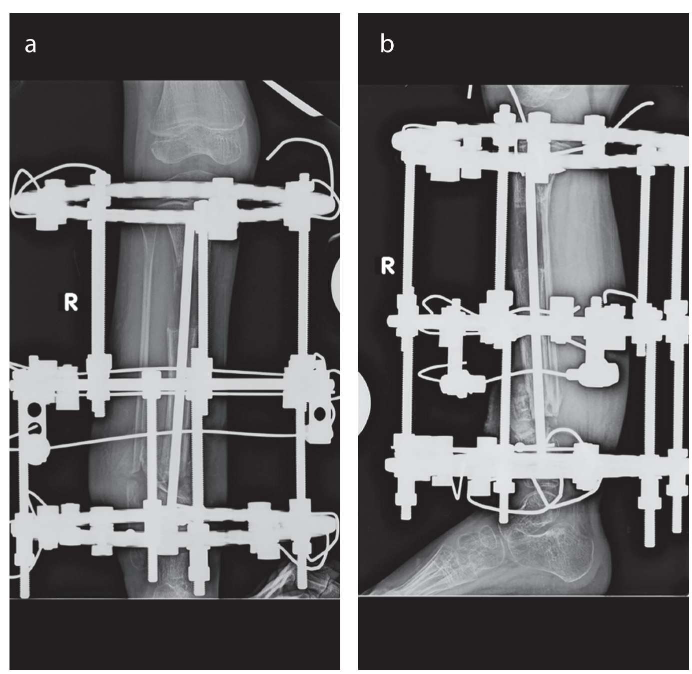

Figure 7. a, b. Postoperative anteroposterior (a) and lateral (b) radiographs of the same patient. The frame was mounted, properly aligned, and positioned with compression at the pseudarthrosis site and proximal lengthening. Proximal metaphyseal osteotomy was used for concomitant lengthening

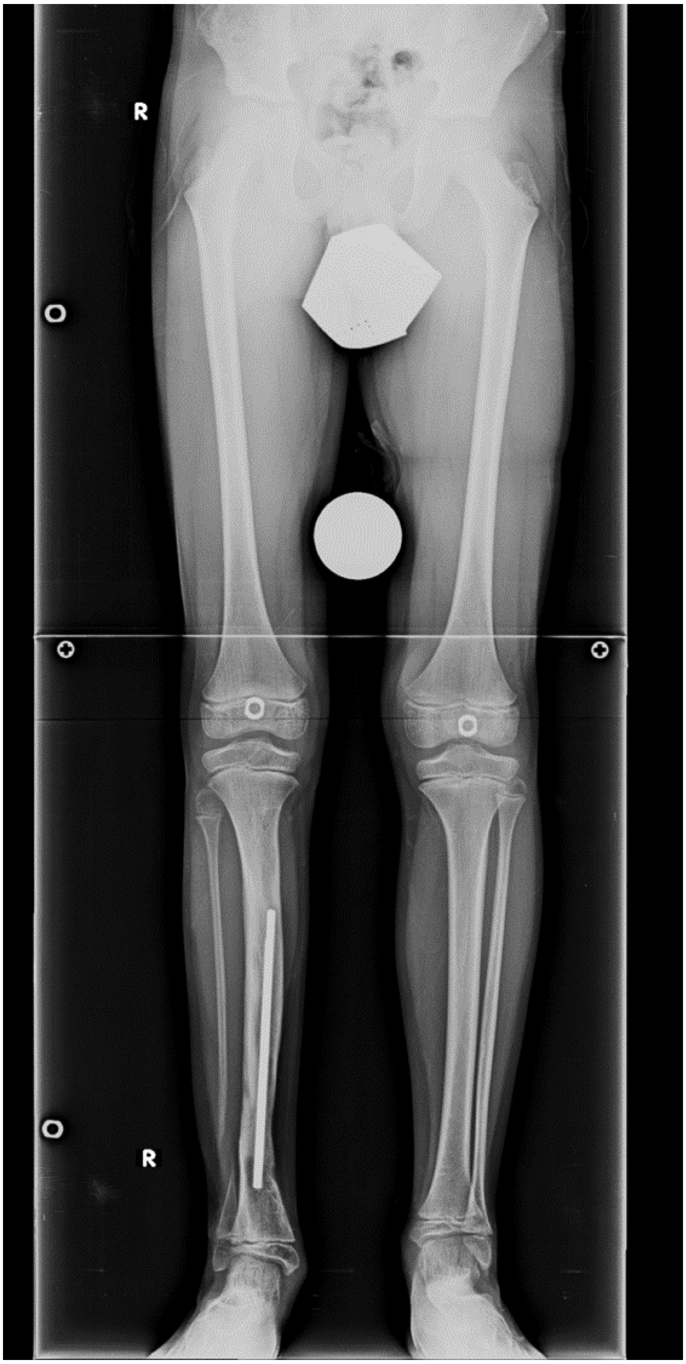

Figure 8. Solid union, good alignment, without residual ankle valgus, tibial bowing, and LLD were revealed by the long-standing lower extremity X-ray of the same patient at the three-year follow-up. The rod was maintained in the intramedullary cavity without affecting the ankle and subtalar joints

Discussion

External fixator application for the treatment of CPT has been widely used with technical modifications such as periosteal grafting, intramedullary rodding, or electrical stimulation (8, 12, 14-17). Several studies have suggested the use of bone morphogenetic proteins (BMPs) in surgical techniques for healing. However, there exists limited evidence concerning the effectiveness of BMPs (11, 19, 22, 35). Furthermore, multiagent therapy using BMP and zolendronic acid was concluded to be superior (20, 21, 34). In our series, we avoided the use of BMP or zolendronic acid.

Intramedullary rodding was the primary choice of treatment before the popularization of external fixation (23-25). Mechanical factors were indicated as the primary reasons for high complication rates during treatment. Maintaining the alignment is crucial as this creates compression forces leading to union. Coleman popularized a procedure using autogenous bone grafting (23, 26). In his series, Anderson suggested maintaining the intramedullary rod until skeletal growth. Schonecker reported union rates of 85.7%, with refracture rates of 66.6%, when reporting the long-term results of intramedullary rod treatments for CPT. Schonecker et al. suggested liberating the subtalar joint after five years and liberating the tibiotalar joint after ten years to ensure that the rod at the ankle joint was as short as possible (13, 23). We used intramedullary rodding with CEF as internal bracing as long as possible. However, consecutive surgeries were generally required to replace or remove Steinman pins due to further corrections and lengthening procedures in selected patients.

The CPT treatment aims at obtaining and maintaining union. Boero et al. reported an 81% union rate and a 23.5% refracture rate in 21 cases treated using CEF with various device assembly options. They concluded that the short-term compression of stumps after resection until biologically and mechanically viable bone growth is the primary factor for better results (3). Paley and Kocaoglu published their treatment results for 20 cases treated with a CEF combined with periosteal grafting and intramedullary rodding in a multicenter study and indicated 100% union and 40% refracture rates. More than half of the patients were aged below 4 years in this series and were skeletally immature during the last follow-up, which explains the lower refracture rates (14). Choi et al. reported eight cases of atrophic-type pseudarthrosis using a "4-in-1 osteosynthesis," with a mean age of 6.3 years. Using CEF in selected patients, they reported a 100% union rate, with no refractures during a mean follow-up period of 3.5 years (range: 2.7–10.2 years). Depending on the fibular status, a complete union between a broader cross-sectional area at the pseudarthrosis site was suggested, without losing the lateral support of the ankle caused by fibular shortening (15). In our series, broader cross-sectional areas were encountered in two patients after unintended tibio-fibular cross-union. Song reported 15 patients treated with CEF and intramedullary rod augmentation without periosteal grafting. The union rate was 40% for the initial index surgery and 93% after consecutive surgeries in patients with no primary healing. The refracture rate was 6.6% during a 4.2-year follow-up period (16). In this series, we reported 88.2% union and 33.3% refracture rates during an 8.5-year follow-up period, highlighting the importance of periosteal grafting. In the literature, success probability (union rate×[1−mean refracture rate]) is 40% in intramedullary rodding, 57% in the Ilizarov method, 58% in the Ilizarov method combined with intramedullary rodding, 58% in vascularzed fibula grafting, and 50% in all published series. According to this definition, we reported a success probability of 58.7% (34).

We excluded two series by Inan et al. and Cho et al. due to the heterogeneous external fixation treatment protocols used in these series (29, 30). Our series has the longest follow-up duration. In addition, the youngest patient was aged 11 months despite the controversy within the field regarding the appropriate age for the initial treatment. The multicenter study reported by the European Paediatric Orthopaedic Society suggested operating only on children aged over 3 years (27). In addition, Boero et al. reported better results for patients aged above 5 years when performing external fixation treatments (3). With regard to the refracture, union, and complication rates, Liu et al. reported no significant differences between patients who received operations before and after 3 years of age; however, LLDs differed between the two groups (28). In this series, we had two different groups with regard to the age of the index operation. The first group comprised patients who received initial operations elsewhere and were referred to us with complications from treatment, such as nonunion and refracture. These patients were treated using the periosteal graft method without consideration of age. The second group comprised patients first diagnosed by the senior author and followed-up by bracing. If they experienced any fractures during bracing, they were operated; otherwise, we preferred to wait until 4 years of age.

One of the main concerns during the follow-up of the treatment of CPT was residual valgus deformity of the ankle. Because of this, fibular surgeries were recommended, and longer refracture-free survival rates were expected (30, 31, 33, 34). Thabet et al. reported 35% ankle valgus in the 4.3-year follow-up period and suggested screw or plate hemiephiphysiodesis (14). Choi et al. defined a classification system according to the fibular morphology and suggested a 4-in-1 technique for patients with a specific type of fibular patology (32). In our series, nine patients developed ankle valgus deformity during follow-up. Supramalleolar osteotomy was the preferred choice of treatment for symptomatic three patients. Remaining six cases, who displayed subtalar compensation in ankle valgus, are potential candidates for future corrections; however, they have not reported any clinical symptoms such as limping or pain.

The primary difficulty in the treatment of CPT is the achievement of the union. CEF with periosteal grafting is a superior method; this method provides a healthy biologic healing milieu and achieves union by correcting the mechanical problems. The combination of periosteal and cancellous bone grafts with intramedullary rods and external fixators addresses all issues that complicate union in CPT treatment.

Ethics Committee Approval: Ethics committee approval was received for this study from the Ethics Committee of İstanbul University, İstanbul School of Medicine (23.05.2018 - 67218/14).

Informed Consent: Written informed consent was obtained from patients and/or patients' family members who participated in this study.

Author Contributions: Author Contributions: Concept - M.K., LE.; Design - M.K., L.E.; Supervision - M.K.; Resources - M.K., L.E.; Materials - M.K., L.E.; Data Collection and/or Processing - M.C., F.E.B.; Analysis and/or Interpretation - M.K, L.E.; Literature Search - M.C., F.E.B.; Writing Manuscript - M.C.; Critical Review - M.K., L.E., F.E.B.

Conflict of Interest: The authors have no conflicts of interest to declare.

Financial Disclosure: The authors declared that this study has received no financial support.

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