American Journal of Case Reports

e-ISSN 1941-5923
© Am J Case Rep, 2018; 19: 426-430
DOI: 10.12659/AJCR.908482

Received: 2017.12.11
Accepted: 2018.02.15
Published: 2018.04.11

Anterolateral Bowing of Congenital Pseudoarthrosis of Tibia Treated by Percutaneous Osteotomy and Gradual Correction Using Taylor Spatial Frame, then Late Insertion of a Fussier-Duval Nail: A Case Report

Authors' Contribution:
Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
Funds Collection G

ABEF 1 Rami Jahmani
E 2 Mohammed Alorjani

1 Department of Special Surgery, Faculty of Medicine, Jordan University of Science and Technology, Irbid, Jordan
2 Department of Pathology, Faculty of Medicine, Jordan University of Science and Technology, Irbid, Jordan

Corresponding Author: Rami Jahmani, e-mail: dr.Jahmany@yahoo.com
Conflict of interest: None declared

Patient: Male, 12
Final Diagnosis: Congenital pseudoarthrosis of tibia
Symptoms: Limp • deformity
Medication: —
Clinical Procedure: Percutaneous osteotomy and gradual correction
Specialty: Orthopedics and Traumatology

Objective: Unusual or unexpected effect of treatment

Background: Congenital pseudoarthrosis of the tibia is one of the most difficult orthopedic diseases to treat. Many authors consider osteotomy of the bowed non-broken tibia a contraindication. However, leaving the patient with a deformed, bowed leg is intolerable and is associated with deformity progression and increased risk of fracture.

Case Report: A 12-year-old boy presented with a bowed leg. X-ray tibia showed partially healed congenital pseudoarthrosis of the tibia and 30 degrees of flexion deformity. Deformity was successfully treated by percutaneous osteotomy made through the site of congenital pseudoarthrosis and gradual correction using a computer-assisted external fixator, the Taylor spatial frame (TSF; Smith and Nephew, Memphis, TN). At the end of the correction, a telescopic Fassier-Duval intramedullary nail was used prophylactically to prevent re-fracture. After 2 years of follow-up, X-ray images show bone-healing without any bowing.

Conclusions: Closed percutaneous osteotomy, without excision of the pseudoarthrosis, and gradual correction then insertion of Fassier-Duval telescopic nail may serve as a treatment to correct deformed bone in congenital pseudoarthrosis of the tibia.

MeSH Keywords: Congenital Abnormalities • External Fixators • Osteotomy • Pseudarthrosis • Tibia • Bone Nails

Full-text PDF: https://www.amjcaserep.com/abstract/index/idArt/908482

Background

Congenital pseudoarthrosis of tibia (CPT) is one of the most difficult orthopedic conditions to treat. The disease is rare, with reported incidence rates of about 1: 53 000 to 1: 190 000 population [1,2]. The exact pathogenesis is not understood. Patients are not always born with established pseudoarthrosis; many are born with sclerotic or cystic bone changes with united bone, which gradually bends then breaks to form pseudoarthrosis [3–5]. Because of that, many authors prefer to call the disease "infantile dysplasia of the tibia" [6]. Bowing and pseudoarthrosis are not the only problem of CPT patients – almost most cases develop what Paley calls "secondary changes". These include: calcareous ankle joint contracture, cavus foot deformity with verticalization of the body of the os calcis, weak push-off, valgus ankle, lateral subluxation of the talus with proximal migration of the fibula, and leg length discrepancy [7].

Although there is consensus regarding the need for surgical intervention for established pseudoarthrosis, treatment of the anterolateral bowing is controversial. When it is associated with duplicated hallux varus, it is benign and spontaneous correction is expected [8]; otherwise, it should be considered as a pre-CPT. The standard treatment of bowing is bracing in clamshell orthosis or patellar tendon-bearing (PTB) orthosis, which is somewhat successful in fracture prevention but not in stopping progression of the deformity [7]. A brace should be worn during the daytime until skeletal maturity, and this requirement is intolerable to many children. Many authors try to avoid bone osteotomy of the deformed bone to avoid the development of pseudoarthrosis. However, leaving the bone deformed is associated with difficulty in wearing an orthosis and progression of the development of secondary changes, which make the functional outcome of any future surgery poor. Based on this, we believe in the need for surgical intervention to correct the deformity in CPT.

Numerous surgical treatment options based on biological and mechanical concepts have been reported, with variable success rates [9,10]. The development of orthopedic devices has improved the treatment outcome of deformities of different natures. Percutaneous osteotomy and gradual correction of deformity using a computer-assisted fixator, the Taylor spatial frame, have proved to be a very successful method [11–13]. We reviewed the literature on percutaneous osteotomy and gradual correction to be applied for the treatment of CPT anterolateral bowing. Here, we report a case of anterolateral bowing of a congenital pseudoarthrosis treated with percutaneous osteotomy and gradual correction with the Taylor spatial frame (TSF; Smith and Nephew, Memphis, TN), followed by late intramedullary insertion of a telescopic Fassier-Duval (FD) nail. After 2-year follow-up, X-ray imaging showed bony union.

Figure 1. Case on presentation.

Case Report

A 12-year-old boy presented with right leg deformity since birth and difficulty in walking. His parents mentioned that their son underwent a surgery to correct the deformity when he was 6 years of age, but they had no documents showing details of that surgery. On examination, the patient had about 30 degrees of anterolateral bowing, ankle valgus and surgical scar on the anterior leg. We reviewed the radiographs and the patient's clinical profile and made a diagnosis of congenital pseudoarthrosis of the tibia (Figure 1), as there was no clinical evidence of neurofibromatosis. Because the classification systems describe the untreated appearance of the bone, type of the pseuoarthrosis could not be identified to our case. Surgical correction was scheduled. During surgery, after taking the nail out (used in the previous surgery), a computer-assisted external fixator TSF was mounted and percutaneous osteotomy at the site of congenital pseudoarthrosis was made using modified De Bastiani technique (percutaneous small skin incision and weakening the bone by multiple drilling, then completing the osteotomy using an osteotome), and a fibular osteotomy was done as well (Figure 2). Two rings of TSF fixator were used in this case: first, the proximal reference ring was mounted and fixed with 1 reference wire and 1 pin, then the distal ring was fixed to the bone using 2 Ilizarov wires. Struts then were connected between the 2 rings and 1 pin was added to each ring. On day 10 post-surgery, gradual correction using the TSF software program was initiated to correct flexion deformity by

Figure 2. Osteotomy and application of TSF.

a rate of 1 degree per day around a center of rotation of angulation (CORA) at the middle of the medullary canal at the site of the osteotomy [11]. Once alignment had been corrected (Figure 3), on day 45 post-surgery, the patient was encouraged to bear partial weight as tolerated.

Two months later, the TSF was taken off, pin holes were curetted and cleaned, and a Fassier-Duval telescopic intramedullary nail was inserted (Figure 4). The surgical technique for insertion of the FD rod was performed as described by Pega Medical, Quebec, Canada [14]. Initially a K-wire was inserted in the medullary canal from the proximal tibia, then the canal was reamed over by the cumulated reamer. We started reaming using the smallest reamer diameter and proceeded sequentially to larger reamer diameters under fluoroscopy guidance until the reamer came in contact with the internal cortical surface. After surgery, the leg was protected by a cylindrical cast for 2 months, which was then replaced by the orthosis. After 2-year follow-up, X-ray images showed bone-healing without tibia bowing (Figure 5).

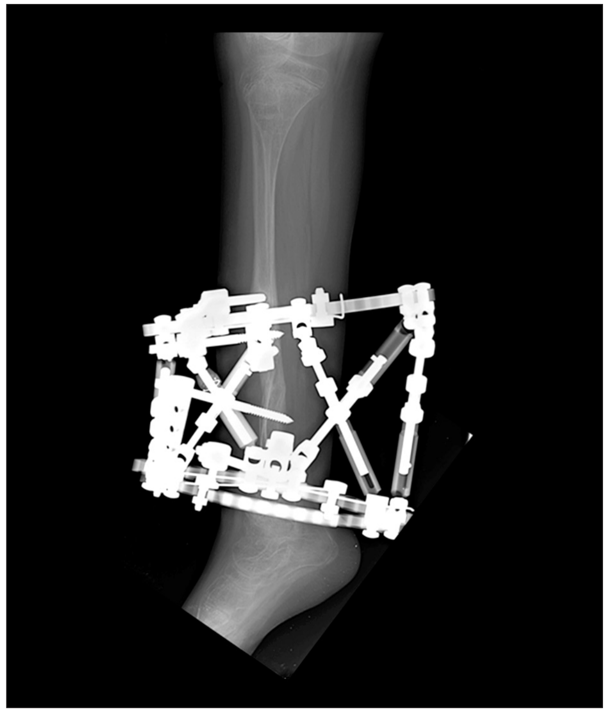

Figure 3. The bone has been aligned.

Figure 4. X-ray image after taking the fixator off, showing FD nail.

Indexed in: [PMC] [PubMed] [Emerging Sources Citation Index (ESCI)] [Web of Science by Clarivate]

Figure 5. After 2-year follow-up (lateral view and anteroposterior view).

Discussion

CPT is one of the most difficult orthopedic conditions to treat. The goal of treatment is to achieve union and maintain it, as in many cases the bone breaks after achieving of union. Two facts about the natural history of the disease are well known. First, there is no chance of spontaneous correction of the bowing. Second, once a fracture has occurred, there is very low chance for spontaneous healing and even surgical intervention is associated with a high failure rate. Therefore, all efforts should be directed to avoid bone breakage. Protecting the anterolateral CPT bowing from fracture is difficult. The literature presents 2 well-known ways to prevent fractures: orthosis and prophylactic bypass grafting [15]. They may be somewhat successful in preventing fracture but not in preventing the progression of deformity and development of secondary deformities [16]. The inconsistent results of prophylactic bypass grafting [15] make orthosis the standard treatment for anterolateral bowing of a CPT nature [7]. However, bracing the bone and leaving the deformity to progress is associated with major morbidity and makes the correction of deformity very hard in any future surgery. We believe it is important to correct the mechanical axis of the bone to stop progression of the deformity. Four decades ago, achieving of union in CPT patients was extremely difficult. Hardinge reported only 6 cases of union among 384 treated CPT cases [17]. These results made the osteotomy for correcting anterolateral bowing of CPT tibias one of the contraindications. Nowadays, with the development of new orthopedic manufacturing and technology, the outcome of CPD treatment has significantly improved. The main challenge in CPT treatment is to maintain healing, as more than half of the cases have a re-fracture [18]. Two main new inventions in orthopedic manufacturing have significantly changed the outcome of CPT treatment: the ring external fixator and a new generation of telescopic intramedullary nails. A multicenter study conducted by the European Pediatrics Orthopedic Society from 13 countries including 340 patients showed a 75.5% fusion rate using the Ilizarov ring fixator [10]. The use of an external fixator facilitates the fixation of small bone, simultaneous treatment of limb length discrepancy, deformity correction, and dynamization and compression of the healed bone. The combined use of an intramedullary nail with the Ilizarov fixator for the treatment of CPT provided lower re-fracture rates [9,19]. The FD nail is the newest generation of telescopic nails. It is an expandable intramedullary nail that follows the growth of the bone (telescopic); it has a unique mechanism of fixation through the epiphysis, which makes it more stable. Early reports of case series combining the use of a ring fixator and FD nail are promising, and 100% initial union has been reported [20]. To the best of our knowledge, there have been only a few case series that described the use of the Fassier-Duval (FD) rod in CPT patients [20–22].

With this development of orthopedic manufacturing and advancement in the treatment of CPT, we decided to revisit percutaneous osteotomy and gradual correction to correct a deformity in a CPT case. In general, percutaneous osteotomy and gradual correction show high union rates in the treatment of bone deformity. Many successful results have been published using this technique for the treatment of deformity due to various causes [12,23]. Percutaneous osteotomy decreases soft tissue damage, preserves hematoma with the contained inflammatory phase mediators within the surgical wound, and is associated with low thermal necrosis; thus, it enhances bone healing. In addition, the process of gradual correction using external fixator is well known for stimulation of bone and soft tissue blood supply. The development of a new generation of external fixators, computer-assisted fixators, allows precise correction of deformity with a less complex construct and more stability [24]. We used this fact to correct a deformity in CPT where bone is very sensitive to minimal error in alignment. In addition, the use of an FD nail with intramedullary reaming after the bone had been corrected acts as a stimulus to blood circulation and provides the osteotomy side with an intramedullary bone graft. These factors together (percutaneous osteotomy, gradual correction and intramedullary reaming) contribute to achieving union.

The advantages of using this technique is prevention soft tissue from being jeopardized by the classical open technique; thus, it increases the success of any revision future surgery whenever it is needed. The use of a reamed FD nail eliminates the need for bone grafting and preserves the ankle joint from being jeopardized by classical rodding through the ankle. The disadvantage of this technique is its inability to excise pseudoarthrosis. In the literature, "end-to-end compression" of established pseudoarthrosis using an Ilizarov fixator without excision of pseudoarthrosis has achieved initial union, but the quality of healed bone was poor and the re-fracture rate was high [19]. The situation in our reported case is different: we induced acute fracture, performed gradual correction, and inserted a nail to prevent re-fracture. Although we could achieve union by this technique, this is just a single case, and larger-scale studies on cases with different types of CPT and including patients from different age groups is required before widespread use of this approach. Long-term follow-up until skeletal maturity is also mandatory.

Conclusions

Closed percutaneous osteotomy, without excision of the pseudoarthrosis, and gradual correction then insertion of Fassier-Duval telescopic nail, can be an effective treatment to correct deformed bone in congenital pseudoarthrosis of the tibia.

Conflict of interest

None.

References:

1. Zhu GH, Mei HB, He RG et al: Effect of distraction osteogenesis in patients with tibia shortening after initial union of Congenital Pseudarthrosis of the Tibia (CPT): A preliminary study. BMC Musculoskelet Disord, 2015; 16: 216
2. Horn J, Steen H, Terjesen T: Epidemiology and treatment outcome of congenital pseudarthrosis of the tibia. J Child Orthop, 2013; 7(2): 157–66
3. Borzunov DY, Chevardin AY, Mitrofanov AI: Management of congenital pseudarthrosis of the tibia with the Ilizarov method in a pediatric population: Influence of etiological factors. Int Orthop, 2016; 40(2): 331–39
4. Shah H, Rousset M, Canavese F: Congenital pseudarthrosis of the tibia: Management and complications. Indian J Orthop, 2012; 46(6): 616–26
5. Song MH, Park MS, Yoo WJ et al: Femoral overgrowth in children with congenital pseudarthrosis of the Tibia. BMC Musculoskelet Disord, 2016; 17: 274
6. Weber M: Congenital pseudoarthrosis of the tibia redefined: congenital crural segmental dysplasia. In: Rozbruch SR, Ilizarov S (eds.), Limb lengthening and reconstruction surgery. New York: Informa Healthcare USA Inc., 2007; 495–509
7. Johari A, Luk KDK, Waddell J: Current progress in orthopedics. Mumbai, India: TreeLife Media (A Div of Kothari Medical), 2014; 318–48
8. Weaver KM, Henry GW, Reinker KA: Unilateral duplication of the great toe with anterolateral tibia bowing. J Pediatr Orthop, 1996; 16: 73–77
9. Ohnishi I, Sato W, Matsuyama J et al: Treatment of congenital pseudarthrosis of the tibia: A multicenter study in Japan. J Pediatr Orthop, 2005; 25: 219–24
10. Grill F, Bollini G, Dungl P et al: Treatment approaches for congenital pseudarthrosis of tibia: Results of the EPOS multicenter study. European Paediatric Orthopaedic Society (EPOS). J Pediatr Orthop B, 2000; 9(2): 75–89
11. Taylor JC: Correction of General Deformity with The Taylor Spatial Frame Fixator™. 2002. [available from: *http://www.jcharlestaylor.com/tsfliterature/01TSF-mainHO.pdf*]
12. Horn J, Steen H, Huhnstock S et al: Limb lengthening and deformity correction of congenital and acquired deformities in children using the Taylor Spatial Frame. Acta Orthop, 2017; 88(3): 334–40
13. Dammerer D, Kirschbichler K, Donnan L et al: Clinical value of the Taylor Spatial Frame: A comparison with the Ilizarov and Orthofix fixators. J Child Orthop, 2011; 5(5): 343–49
14. Fassier F, Paley D, Duval P: Fassier-Duval Telescopic IM System. Surgical Technique. 2013. [Available from: *http://www.pegamedical.com/medias/iw/fassier-duval_surgical_techniques_en.pdf*]
15. Khan T, Joseph B: Controversies in the management of congenital pseudarthrosis of the tibia and fibula. Bone Joint J, 2013; 95-B(8): 1027–34
16. Ofluoglu O, Davidson RS, Dormans JP: Prophylactic bypass grafting and long-term bracing in the management of anterolateral bowing of the tibia and neurofibromatosis. J Bone Joint Surg Am, 2008; 90(10): 2126–34
17. Hardinge K: Congenital anterior bowing of the tibia: the significance of different types in relation to pseudarthrosis. Ann Roy Coll Surg Engl, 1972; 51: 17–30
18. Vanderstappen J, Lammens J, Berger P, Laumen A: Ilizarov bone transport as a treatment of congenital pseudarthrosis of the tibia: A long-term follow-up study. J Child Orthop, 2015; 9(4): 319–24
19. Agashe MV, Song SH, Refai MA et al: Congenital pseudarthrosis of the tibia treated with a combination of Ilizarov's technique and intramedullary rodding. Acta Orthop, 2012; 83(5): 515–22
20. Alzahrani MM, Fassier F, Hamdy RC: Use of the Fassier-Duval telescopic rod for the management of congenital pseudarthrosis of the tibia. J Limb Lengthen Reconstr, 2016; 2: 23–28
21. Hamdy RC: Congenital pseudarthrosis of tibia and fibula: An introduction. In: Rozbruch RS, Hamdy CR (eds.), Limb Lengthening and Reconstruction Surgery Case Atlas: Pediatric Deformity. Switzerland: Springer International Publishing, 2015; 195–96
22. Wagner P, Herzenberg JE: Case 33: Congenital tibial pseudoarthrosis treated with internal and external fixation using the 4 in 1 technique. In: Rozbruch RS, Hamdy CR (eds.), Limb Lengthening and Reconstruction Surgery Case Atlas: Pediatric Deformity. Switzerland: Springer International Publishing, 2015; 223–27
23. Dabis J, Templeton-Ward O, Lacey AE et al: The history, evolution and basic science of osteotomy techniques. Strat Traum Limb Recon, 2017; 12: 169–80
24. Reitenbach E, Rödl R, Gosheger G et al: Deformity correction and extremity lengthening in the lower leg: Comparison of clinical outcomes with two external surgical procedures. Springerplus, 2016; 5(1): 2003

Indexed in: [PMC] [PubMed] [Emerging Sources Citation Index (ESCI)] [Web of Science by Clarivate]