MEDICAL SCIENCE

A rare case report of congenital pseudoarthrosis of tibia and fibula managed with dual modality of treatment: Ilizarov external fixator and plate osteosynthesis

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ABSTRACT

Congenital pseudoarthrosis is specific kind of non-union that is either present or developing at birth. Though its cause is unknown, it commonly affects people with neurofibromatosis. The most common sites of congenital pseudoarthrosis are the distal portion of the tibia and commonly the fibula of the same leg. Approximately one in 250,000 live pregnancies results in congenital pseudarthrosis of the tibia. A 4-year-old child with a history of his right leg being shorter since, birth and being unable to use it for walking arrived at the orthopaedic outpatient clinic. The patient was managed with resection of the callus and pseudoarthrosis site and the tibia shaft was fixed with low contact dynamic compression locking device. The Ilizarov ring fixator was used and corticotomy was performed at the junction of the proximal and middle tibial shafts in order to lengthen the leg using the distraction osteogenesis principle. The goal of surgical therapy is to achieve pseudoarthrosis bone union while restoring limb alignment in order to lessen the risk of a subsequent fracture and keep bone development and function in the leg. It appears as CPT and is commonly accompanied with distinctive anterolateral bending. The Ilizarov procedure offers patients with CPT a therapeutic choice that is secure, useful and realistic. It can achieve a number of goals, including ankle stability, osteosynthesis and leg-length equality, when compared to alternative therapy methods. The fixation technique using a locking compression plate along with a bone graft improves the treatment of complicated congenital pseudarthrosis of the tibia.

Keywords: Pseudoarthrosis, Neurofibromatosis, Ilizarov, Osteosynthesis

1. INTRODUCTION

The two main types of congenital angular abnormalities of the leg are those in which the angulation's apex is anterior and those in which it is posterior. In both, the tibia frequently bows medially or laterally in addition to anteriorly or posteriorly (Pannier, 2011). Neurofibromatosis is frequently linked to anterior tibial bending. Congenital pseudoarthrosis is a particular kind of non-union that is either present or developing at birth. Its origin is uncertain, but enough people with neurofibromatosis or stigmata associated with it have it to raise the possibility that neurofibromatosis, if not the actual cause of congenital pseudarthrosis, is at least closely connected to it (Kong et al., 2018). The distal portion of the tibia and commonly the fibula in the same leg are most frequently affected by congenital pseudoarthrosis. The real reason why the bone at the site of the pseudoarthrosis has the poor healing capability is unknown; nonetheless, hamartomatous thickened fibrous tissue with restricted vascular expansion is always present there approximately one in 250,000 live births results in congenital pseudarthrosis of the tibia. A similar disease in the ipsilateral tibia frequently precedes or is present together with congenital pseudoarthrosis of the fibula (Shah et al., 2012). Depending on the patient's age and whether or not they have a fracture, congenital pseudoarthrosis of the tibia is treated differently. Pseudarthrosis requires little treatment before the kid can walk, but after they can, the limb has to be covered and immobilized in a clamshell orthosis. If there is no fracture, the kid can be managed with close monitoring while wearing a brace until skeletal maturity. When casts or braces are used as the sole treatment for a real pseudoarthrosis of the tibia, healing cannot be anticipated (Thakur et al., 2015). The first step in treating tibial pseudoarthrosis surgically is resecting the whole pseudoarthrosis and the surrounding hamartomatous tissue, restoring mechanical alignment and fixing the implant intramedullary. A combination of primary shortening, bone transport, extra bone grafting and bone morphogenetic protein is frequently used to supplement these three fundamental principles. Even when the union is achieved, it often only lasts a short while. Refracture leg-length disparity and malalignment may necessitate further surgical care and even amputation, in cases where they occur. We report a Case of a Child with a consanguineous marriage shortening of the Right leg since birth and inability to use the right leg for walking.

2. CASE REPORT

4 years older boy arrived at the orthopaedic outpatient clinic with a history of his right leg has been shorter since birth and he cannot walk on his right leg. The patient is born of consanguineous marriage. There was no family history of such deformity. There is no significant obstetric history. It was a full-term normal vaginal delivery. A bony abnormality in the right leg was noticed (Figure 1). Milestones were developed at appropriate for the age except walking. On general examination, he is a healthy and vitally stable boy, afebrile with no signs of pallor, icterus or clubbing. There is café au lait spots on the back (Figure 2). Local examination shows a bony discontinuity in the right leg, while the left leg is normal. The systemic examination is within normal limits. An X-ray of the right leg reveals tibial and fibular bony discontinuity, tapering ends for both bones and callus production where the deformity is located (Figure 3, 4). He was scheduled for surgery to repair congenital pseudoarthrosis of the tibia (CPT), Boyd's type 4, after the radiographs and clinical characteristics were examined. The intersection of the tibia's middle and distal thirds is where pseudoarthrosis was seen.

The patient was planned for surgical management. Intra-operatively Callus and Pseudoarthrosis site was resected and the tibia shaft was fixed with low contact dynamic compression locking plate. Ilizarov ring fixator was applied and corticotomy was done at the proximal and middle tibial shaft junction for limb lengthening by the principle of distraction osteogenesis (Figure 5, 6, 7). The patient was postoperatively monitored in the recovery room, given appropriate intravenous antibiotics and analgesics for three days and later shifted to oral medications. The patient recovered very well. On Post-op Day 5, the distraction of 1 mm/day started of Ilizarov ring fixator for distraction osteogenesis. The gap between the Fracture fragments was filled with a bone graft made from resected pseudoarthrosis.

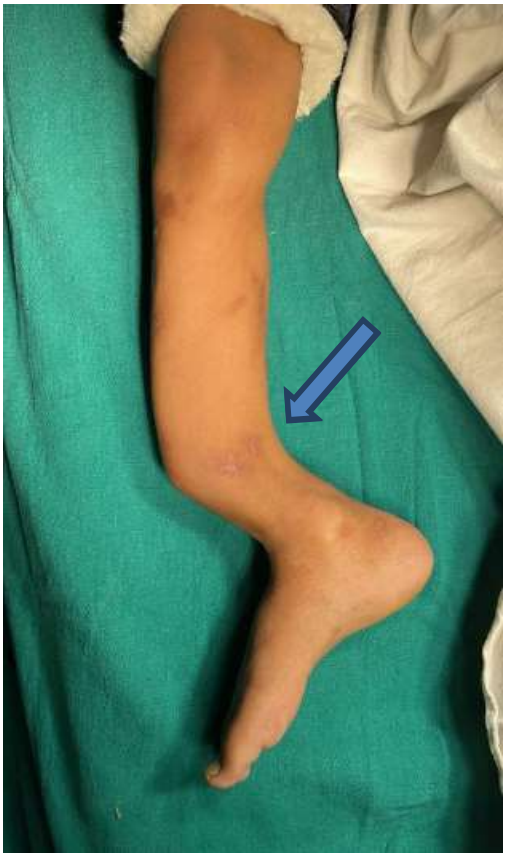

Figure 1 Shows visible deformity of right leg

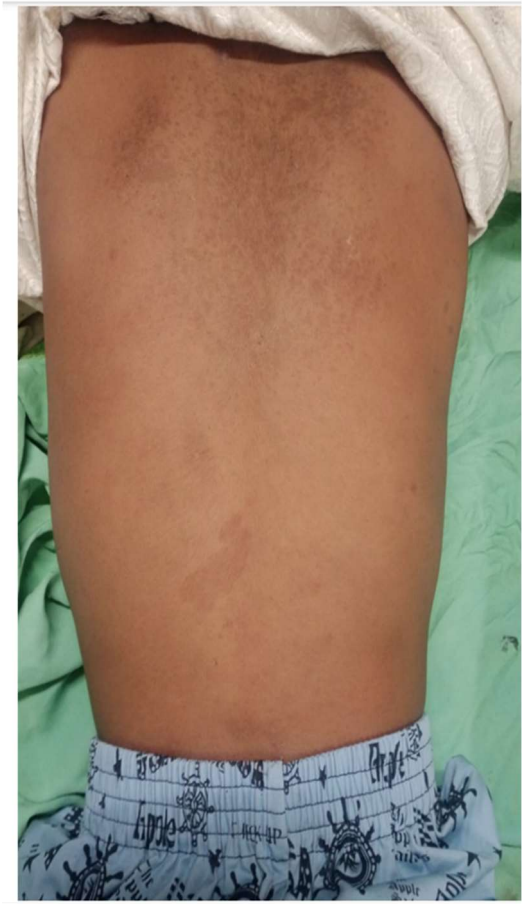

Figure 2 Shows Café' au lait spots on back

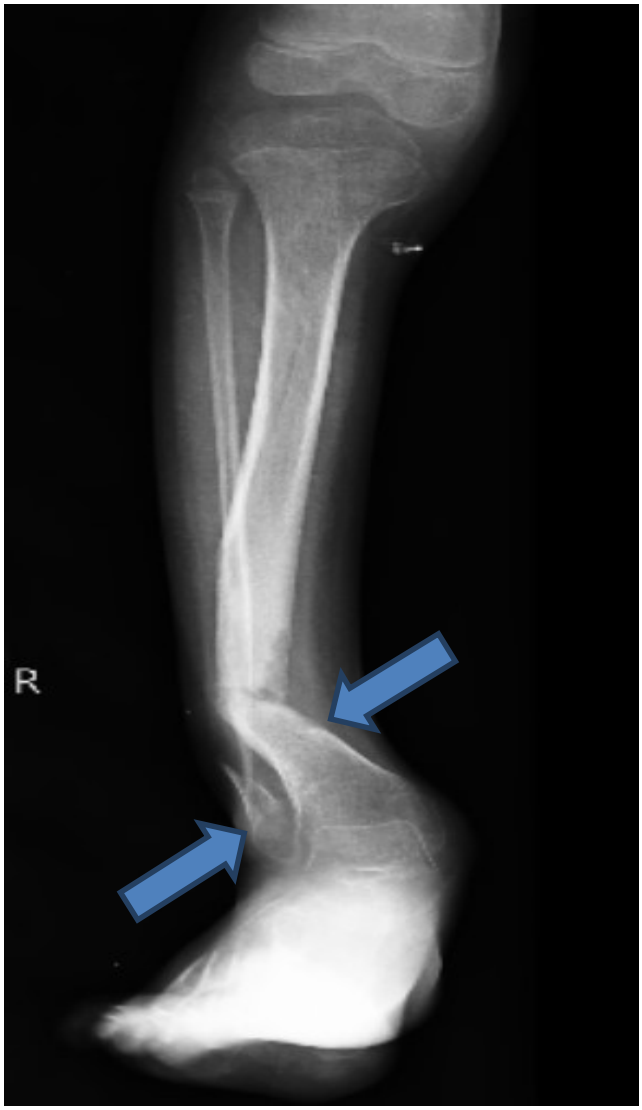

Figure 3 Shows X ray of the child Anteroposterior view shows Pseudoarthrosis at the junction of Middle and Distal third of the tibial shaft and distal fibula shaft with medial angulation

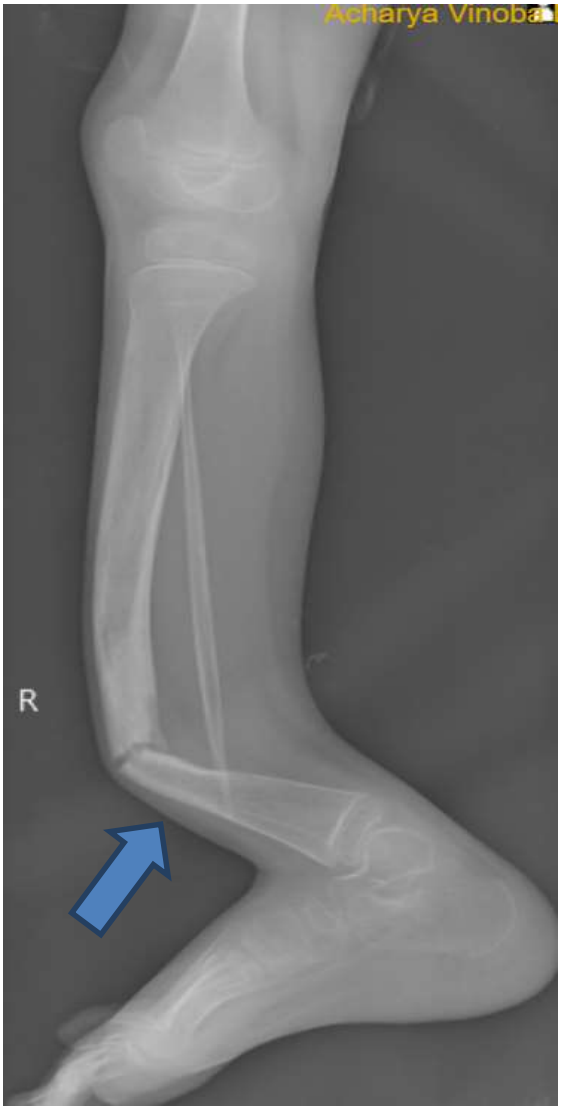

Figure 4 Shows X ray of the child lateral view shows posterior angulation of the tibia

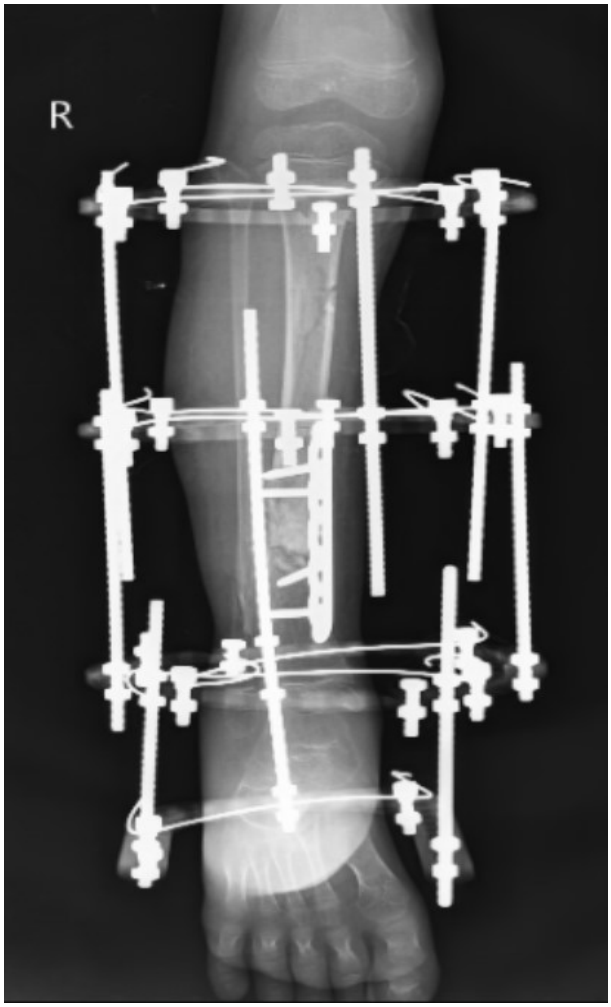

Figure 5 Shows Post-operative X-ray of the child Anteroposterior view shows plate and ilizarov ring fixator with bone graft

The patient was later managed by removing the Ilizarov fixator and above knee cast application for four weeks for protected weight bearing. Later, after four weeks of casting, the above knee cast was removed. Physiotherapy in the form of Quadriceps and Hamstrings strengthening exercises, Ankle pumps, Knee ROM exercises and Partial to full weight-bearing mobilization started. After proper recovery and physiotherapy patient was discharged and advised for regular follow-up (Figure 8 and 9).

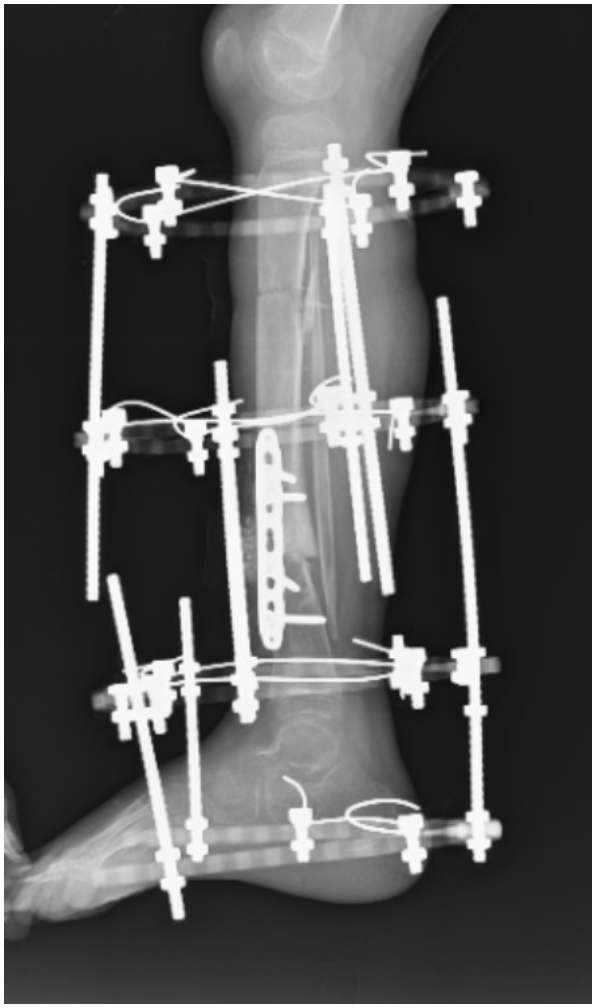

Figure 6 Shows X ray of the child lateral view shows plate and ilizarov ring fixator with bone graft and corticotomy site

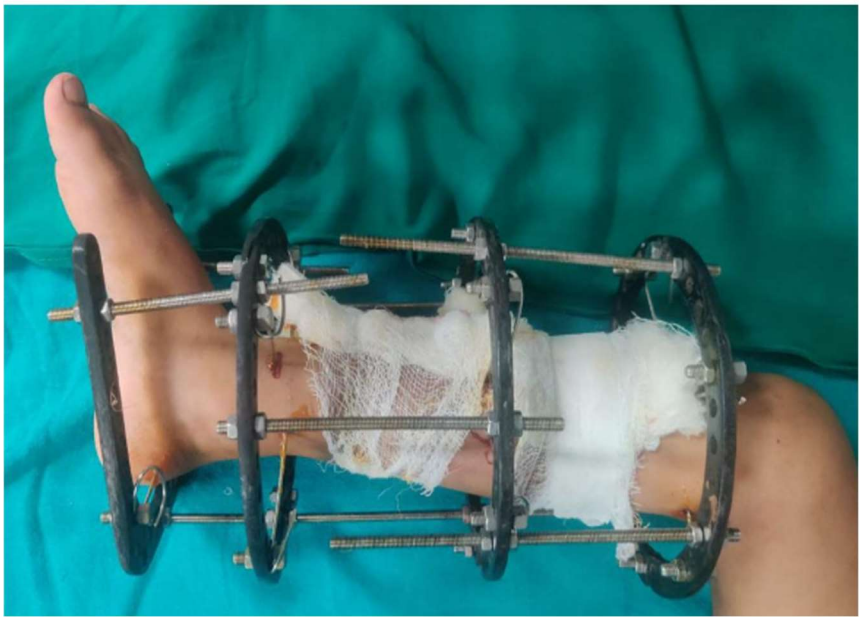

Figure 7 Shows clinical image of the leg with ilizarov ring fixator and deformity correction

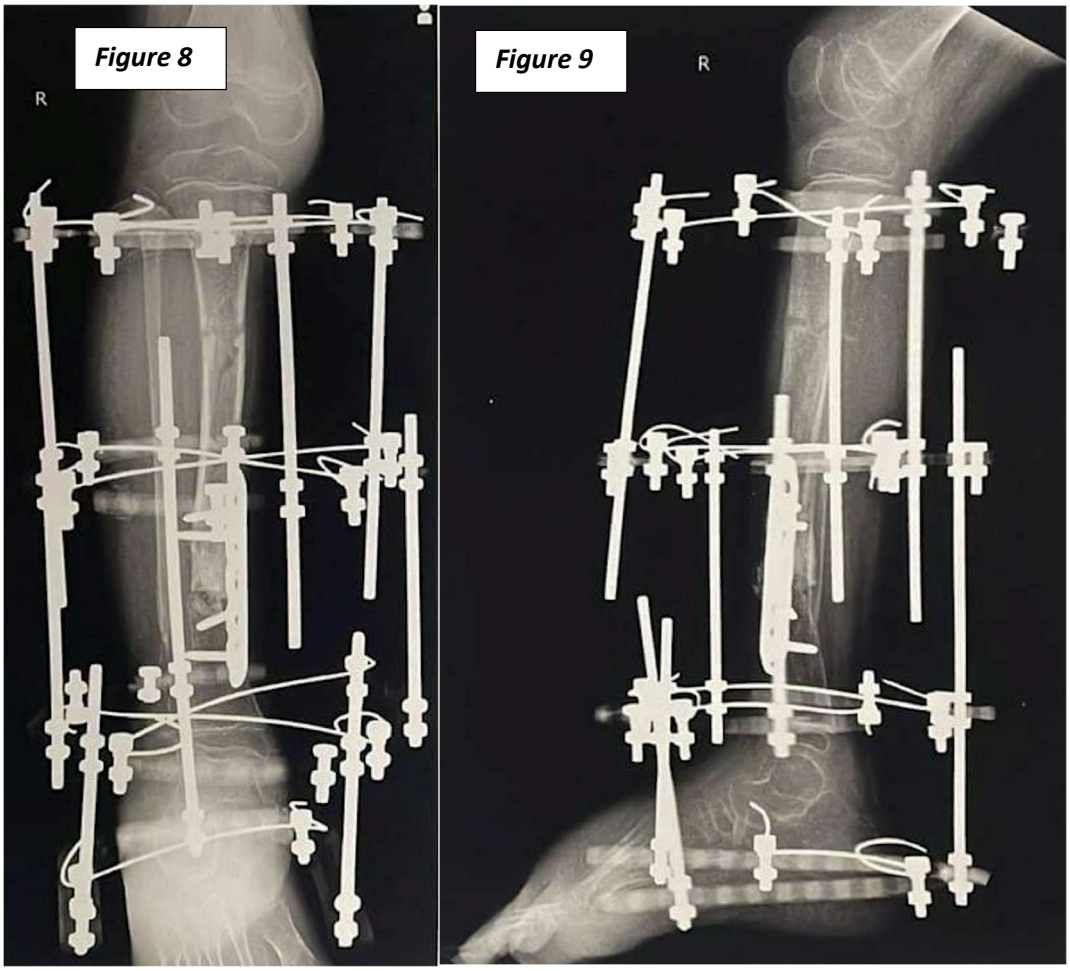

Figure 8 and 9 Shows 2 months follow up x-ray of the child showing uniting tibial shaft with plate and ilizarov fixator in situ with osteogenesis at corticotomy site in anteroposterior and lateral view respectively

3. DISCUSSION

Because it is tough to establish and keep a strong union, the CPT continues to be challenging for orthopaedic doctors. Ohnishi et al., (2005) found that vascularized fibular graft and Ilizarov technique were the most effective ways to treat CPT patients in multicenter research (Stammers and Cope, 2015). These techniques do not, however, always ensure a strong relationship. In order to reduce the danger of a recurrence fracture and maintain bone development and function in the leg, surgical treatment aims to accomplish pseudoarthrosis bone union while restoring leg alignment. In our experience, the Ilizarov treatment is very useful when other surgeries fail or refracture occurs (Boyd, 1982).

Additionally, if this method doesn't work, there are still all other traditional drugs available. In order to encourage healing, cortical bone grafts have been employed as an anabolic stimulation. It often comes on as CPT and has unique anterolateral bending. It is commonly misdiagnosed since the initial symptoms resemble fractures. Lower limbs are stretched to restore balance once the osteotomy heals (Vander-Have et al., 2008). We think that the same technique might be used to cure both shorter deformity and lower extremity angulation. It could reduce hospital stays and healthcare costs. Even though the cure rate for CPT is gradually increasing, the final outcome is often impacted by a number of factors, including leg-length discrepancy, refracture, axial deformity, progressive malalignment and ankle valgus. Refractivity is one of the main issues. All weight-bearing tension will be applied to the connection's weakest component (Grill et al., 2000). Common biological and mechanical defects of the united bone include a tiny docking site, a tibial valgus or rotation deformity and a sclerotic bone lacking a medullary cavity. This calls for the adoption of a preventative brace.

Despite disagreement on the best course of action for CPT, there is consensus in the literature on these problems. Whatever the technique, a strong internal fixation and tibial section realignment are required for the union. All kinds and presentations of CPT cannot be properly treated with surgery. The kind of pseudoarthrosis and in particular, the degree of the bone anomalies, should be taken into account while choosing the surgery (Ohnishi et al., 2005; Alzahrani, 2022). Despite disagreement on the best course of

action for CPT, there is consensus in the literature on these problems. Whatever the technique, a strong internal fixation and tibial section realignment are required for the union. All kinds and presentations of CPT cannot be properly treated with surgery. The kind of pseudoarthrosis and in particular, the degree of the bone anomalies, should be taken into account while choosing the surgery (Ohnishi et al., 2005).

4. CONCLUSION

Treatment for challenging congenital pseudarthrosis of the tibia now includes a bone transplant as well as a fixation operation employing a locking compression plate. The Ilizarov treatment offers patients with CPT a therapy alternative that is secure, useful and realistic. It can achieve a number of goals, including ankle stability, osteosynthesis and leg-length equality, as compared to alternative treatment methods. The patient's postoperative condition is also steady. If the patient has mastered tractor adjustment, they could be permitted to go home with an external fixation. This might reduce hospital stays and healthcare costs. However, the results of surgery also need continued observation. Future research should potentially provide light on the etiopathogenesis of CPT and result in the development of a fresh approach.

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Informed consent

Written & Oral informed consent was obtained from individual participant included in the study. Additional informed consent was obtained from individual participant for whom identifying information is included in this manuscript.

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Conflict of interest

The authors declare that there is no conflict of interests.

Data and materials availability

All data sets collected during this study are available upon reasonable request from the corresponding author.

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