e-ISSN: 0976-822X, p-ISSN:2961-6042

Available online on http://www.ijcpr.com/

International Journal of Current Pharmaceutical Review and Research 2025; 17(9); 1260-1261

Original Research Article

Bridging the Gap: Advances in the Treatment of Congenital Pseudoarthrosis of the Tibia

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Received: 01-06-2025 / Revised: 15-07-2025 / Accepted: 21-08-2025
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Conflict of interest: Nil

Abstract

Congenital pseudoarthrosis of the tibia (CPT) is a rare but challenging pediatric orthopedic condition characterized by spontaneous fracture, non-union, and progressive deformity. Despite advances in surgical techniques and adjuvant therapies, achieving durable union with optimal limb function remains difficult. This paper provides an extensive review of current treatment modalities, including intramedullary fixation, vascularized fibular grafting, Ilizarov external fixation, and combined approaches. The role of pharmacological adjuvants such as bisphosphonates and bone morphogenetic proteins (BMPs), alongside emerging biological therapies, is also discussed. Outcomes, complications, and future directions in management are analyzed to provide a comprehensive understanding of CPT treatment.

Keywords: Congenital Pseudoarthrosis, Tibia, Pediatric Orthopedics, Vascularized Fibular Graft, Ilizarov Fixation, Bone Morphogenetic Proteins.

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Introduction

Congenital pseudoarthrosis of the tibia (CPT) is a rare entity, occurring in approximately 1 in 140,000 to 250,000 live births. It is frequently associated with neurofibromatosis type 1 (NF-1) and less commonly with fibrous dysplasia. The pathophysiology involves dysplastic periosteum, poor vascularization, and fibrous hamartomatous tissue, leading to bone fragility and spontaneous fracture, most often in early childhood. Management of CPT remains one of the most debated areas in pediatric orthopedics. The primary treatment goals include achieving bone union, preventing refracture, correcting deformity, preserving limb length, and ensuring long-term function. Despite decades of research, no single treatment modality has proven universally successful[5].

Review of Literature and Treatment Modalities

Non-Operative Management: Historically included splinting and bracing. These approaches were rarely successful in achieving union and are currently reserved for temporary stabilization until definitive surgical intervention is feasible.

Intramedullary Fixation: The aim is to provide stable fixation across the pseudoarthrosis site using Rush rods, Williams rods, or telescopic rods. While relatively simple, the technique carries a high refracture rate, risk of rod migration, and potential angular deformities.

Vascularized Fibular Grafting (VFG): For many years considered the gold standard, VFG provides structural support and biological activity.

Reported union rates range from 60–80%. Complications include stress fractures, ankle instability, and donor site morbidity[2,3].

Ilizarov External Fixation: Based on principles of resection of pathological tissue, compression at the pseudoarthrosis site, and distraction osteogenesis for limb lengthening. It allows simultaneous correction of deformity and limb length discrepancy, with union rates reported at 70–90%. Limitations include pin tract infections, patient compliance issues, and prolonged treatment duration[1,7].

Combined and Modified Approaches: Increasingly, surgeons are combining intramedullary fixation with Ilizarov external fixators to improve stability and reduce refracture risk. The cross-union technique, where tibia and fibula are united with external fixation and bone grafting, has demonstrated promising union rates[9].

Adjuvant Biological Therapies: Bone morphogenetic proteins (BMP-2, BMP-7) stimulate osteogenesis and are often used alongside grafting. Bisphosphonates decrease osteoclastic activity, reducing bone resorption. Stem cell and tissue engineering approaches are still in the experimental phase but hold promise for the future[8].

Discussion

The treatment of CPT requires an individualized, multi-modal strategy. Vascularized fibular grafting and Ilizarov fixation remain the most widely accepted techniques, yet refracture and progressive deformity are persistent challenges. Recent literature emphasizes the benefits of combined approaches that integrate stable fixation, bone grafting, and biological augmentation. The success of treatment hinges on meticulous resection of dysplastic tissue, stable fixation, and prolonged postoperative bracing to prevent refracture. Importantly, long-term follow-up is crucial, as children are at risk for refracture, especially during growth spurts[6,9].

Conclusion

Congenital pseudoarthrosis of the tibia remains one of the most difficult conditions to treat in pediatric orthopedics. No single technique guarantees long-term success. Current evidence supports combined approaches utilizing stable fixation, vascularized grafting, and biologic augmentation.

Future management will likely focus on regenerative medicine and genetic therapies, particularly in cases associated with NF-1. A multidisciplinary approach involving pediatric orthopedicians, geneticists, and rehabilitation specialists is key to optimizing outcomes.

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