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Clinical Outcomes of Congenital Pseudoarthrosis of Tibia Treatment: Long Term Study Experience from Our Tertiary Referral Center

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Abstract

Congenital pseudoarthrosis of the tibia (CPT) is a rare orthopedic condition marked by abnormal bone development, leading to anterolateral bowing of the tibia and a propensity for pathological fractures. With an incidence of approximately 1 in 140,000 live births, CPT often presents in early childhood and poses significant challenges in achieving bone union due to its progressive nature. CPT commonly presents with anterolateral bowing of the tibia, progressing to spontaneous fractures and pseudoarthrosis. Over 50% of cases are associated with neurofibromatosis type 1. The condition is characterized by poor bone quality and abnormal vascularization, resulting in progressive deformity, recurrent fractures, limb length discrepancy, and altered limb function. Effective treatment is crucial to prevent long-term disability. The primary objectives are to achieve bone union, maintain limb function, minimize complications, reduce refracture rates and limit surgical interventions. Various treatments include intramedullary nailing, vascularized fibular grafts, external fixation (e.g., Ilizarov fixator) and plaster of Paris casts. The choice depends on the condition's stage and individual patient factors. We aim to analyze the clinical outcomes of 22 CPT patients treated with different surgical modalities, contributing insights into effective management and potential areas for improvement. A retrospective analysis was conducted on 22 patients treated at Indira Gandhi Institute of Child Health, Bangalore, from 2011 to 2015, with follow-ups ranging from one to five years. Data were collected on demographics, clinical and radiographic findings, treatment modalities, surgical details, complications, and outcomes. Patients were classified using the Boyd system to guide treatment. Of the 22 patients (14 males, 8 females), Boyd classification distribution was: Type I - 2 patients, Type II - 2 patients, Type IV - 6 patients, Type VI (associated with NF1) - 12 patients. Treatments included K-wires (4 patients), telescopic nails (10 patients), Ilizarov fixators (6 patients), and Rush nails (2 patients).Bone union was achieved in 12 patients (54.5%), with 8 requiring multiple procedures. Refracture occurred in 2 patients (9%). Limb length discrepancies were noted in 8 patients (36.3%). Complications included refracture and amputation in severe cases. Managing CPT is challenging, requiring individualized treatment plans. While bone union was achieved in over half of the cases, the study highlights the necessity of addressing complications like refracture and limb length discrepancy. Further research with larger cohorts and extended follow-ups is essential to enhance treatment strategies and patient outcomes.

Keywords
Congenital pseudoarthrosis of the tibia, CPT, Tibial pseudoarthrosis, tibial deformity, deformity of leg.

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INTRODUCTION:

Congenital pseudoarthrosis of the tibia (CPT) is a rare orthopedic condition characterized by abnormal bone development, typically presenting with anterolateral bowing of the tibia and a propensity for pathological fractures[1]. The incidence of CPT is approximately 1 in 140,000 live births. This condition often manifests in early childhood and poses significant challenges due to its progressive nature and the complexities involved in achieving bone union.

Clinical Features: CPT is commonly associated with anterolateral bowing of the tibia[2], which may be noticeable at birth or develop during infancy. The deformity progresses over time, leading to spontaneous fractures that fail to heal properly, resulting in a false joint or "pseudoarthrosis." [3] More than 50% of CPT cases are linked to neurofibromatosis type 1 (NF1)[4], a genetic disorder that affects the development of nerve tissues.

Pathophysiology and Natural History: The pathophysiology of CPT involves poor bone quality and abnormal vascularization, contributing to the high likelihood of fractures and non-union[5]. If left untreated, CPT leads to several complications:

- Progressive deformity of the tibia
- Recurrent fractures
- Limb length discrepancy
- Altered limb function

These complications necessitate prompt and effective treatment to prevent long-term disability.

Goals of Treatment: The primary objectives in managing CPT are:

- Achieving sound bone union
- Maintaining the functional capability of the limb
- Minimizing complications
- Reducing the rate of refracture
- Limiting the number of required surgical interventions

Research shows numerous treatment modalities have been explored for CPT, but no single approach is universally superior. The choice of treatment often depends on the stage of the condition and individual patient factors. Commonly used treatments include Intramedullary nailing, Vascularized, fibular grafts, External fixation (e.g., Ilizarov fixator)[6], Plaster of Paris casts, Different surgeons advocate for various methods, each reporting varying success rates[7].

LITERATURE REVIEW:

Recent journals and standard reference text books have been searched and checked data base for the literature.

The Boyd Classification system is widely used to categorize CPT:

- **Type I:** Fracture present at birth
- **Type II:** Hourglass constriction of the tibia
- **Type III:** Bone cyst
- **Type IV:** Sclerotic segment of the tibia without constriction, resulting in a stress fracture
- **Type V:** Dysplastic fibula
- **Type VI:** Presence of intraosseous neurofibroma

This classification aids in determining the appropriate treatment strategy and predicting outcomes. This study aims to present the results of 22 patients with CPT, treated with different surgical modalities at our hospital. By analyzing the clinical outcomes, the study seeks to contribute valuable insights into the management of this challenging condition, highlighting the effectiveness of various treatment options and identifying potential areas for improvement in patient care.

MATERIALS AND METHODS:

This study is a retrospective analysis of 11 patients with congenital pseudoarthrosis of the tibia (CPT) who were treated at Indira Gandhi Institute of Child Health (IGICH) in Bangalore. The study period spans from 2011 to 2015, with a minimum follow-up period of one year and a maximum of five years. Total 22 patients from 5 to 12 years, among them 14 were males and 8 females from IGICH. The inclusion criteria was patients from IGICH diagnosed to be congenital pseudoarthrosis of tibia. Patients were followed up for a minimum of 1 year.Various surgical techniques were employed based on the patient's age, the extent of deformity, marrow size, and involvement of the fibula. The selection of implants was tailored to individual patient needs.

Implants Used:

K-wire (used in 4 patients), Expandable (telescopic) nails (used in 10 patients), Ilizarov external fixator (used in 6 patients), Rush nail (used in 2 patients)

Outcome Measures:

The primary outcomes measured were rate of sound bone union, Functional capability of the limb, incidence of complications (e.g., refracture, limb length discrepancy), Number of surgical

interventions required. Descriptive statistics were used to summarize patient demographics and clinical characteristics. Outcomes were expressed as percentages for categorical variables (e.g., rate of union, complication rates). Patients were followed up for a minimum of one year post-surgery, with periodic clinical and radiographic evaluations conducted to monitor healing, limb function, and the occurrence of complications. All final evaluations were conducted by the same doctor to ensure consistency.

RESULTS AND DISCUSSION:

Total Number of Patients:

22, Range from 5 months to 12 years. Among them 14 were males and 8 were females (Male-to-female ratio: 7:4). The Boyd Classification Distribution was Type I: 2 patients (9%), Type II: 2 patients (9%), Type IV: 6 patients (27%), Type VI (associated with neurofibromatosis): 12 patients (55%).Treatment Modalities and Implant Usage were, 4 (18%) patients had k wire fixation, Telescopic expandable nails were used in 10 (45%) patients. Illizarov external fixator was used in 6 (27%) and 2 (18%) patients had rush nail. The criteria for Implant Selection were:

- **Age of Patient:** Younger patients often received K-wires or expandable nails[8] (fig 1-6).
- **Marrow Size:** Larger implants were used in patients with sufficient marrow size.
- **Extent of Deformity:** More severe deformities often required the use of the Ilizarov fixator (Fig 7- 10)
- **Involvement of Fibula:** Presence of fibular involvement influenced the choice of implant and surgical technique.
- **Limb Length Discrepancy:** Consideration was given to anticipated limb lengthening needs.

Clinical Outcomes was sound Union achieved in 12 patients (54.5%), Single Surgerywas in 4 patients (18%) and multiple procedures in 8 patients (36%), Union with Bone Grafting: Required in 4 patients (18%), Refracture Rate: 2 patient (9%) experienced refracture after discontinuing brace at 3 months. Fracture Healing in Progress: 4 patients (18%) still undergoing healing with the last surgery performed less than 5 months prior to the final follow-up. Limb Length Discrepancy was observed in 8 patients (36.3%), all discrepancies were less than 5 cm. There was a need for Multiple Definitive Procedures in more severe Boyd types (e.g., Type VI and IV).

Complications:

There was re fracture in 2 patients (9%) post-treatment. Two (18%) of the patients were recommended amputation in view of chronic debility and to mobilize early. Two (18%) patients lost follow up.

Comparative Data with with previous studies:

Sound Union Rates:

- Our Study: 54.5%
- Boerco and Catagni et al.: 66.7%
- Dobbs MB et al.: 76%

Refracture Rates:

- Our Study: 9%
- Boerco and Catagni et al.: 23.5%
- Dobbs MB et al.: 57%

Limb Length Discrepancy:

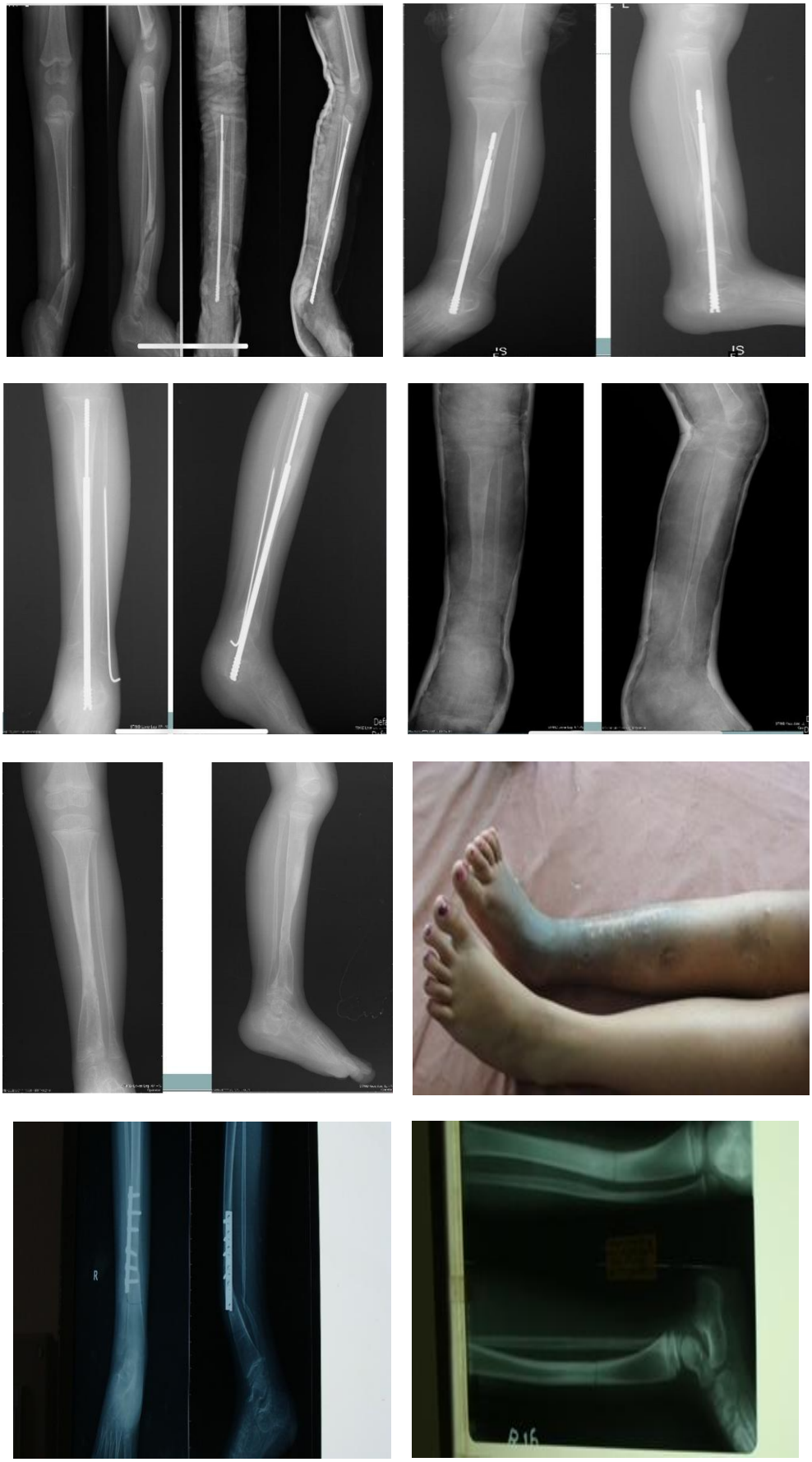
- Our Study: 36.3%
- Anderson DJ *et al*.: 63%

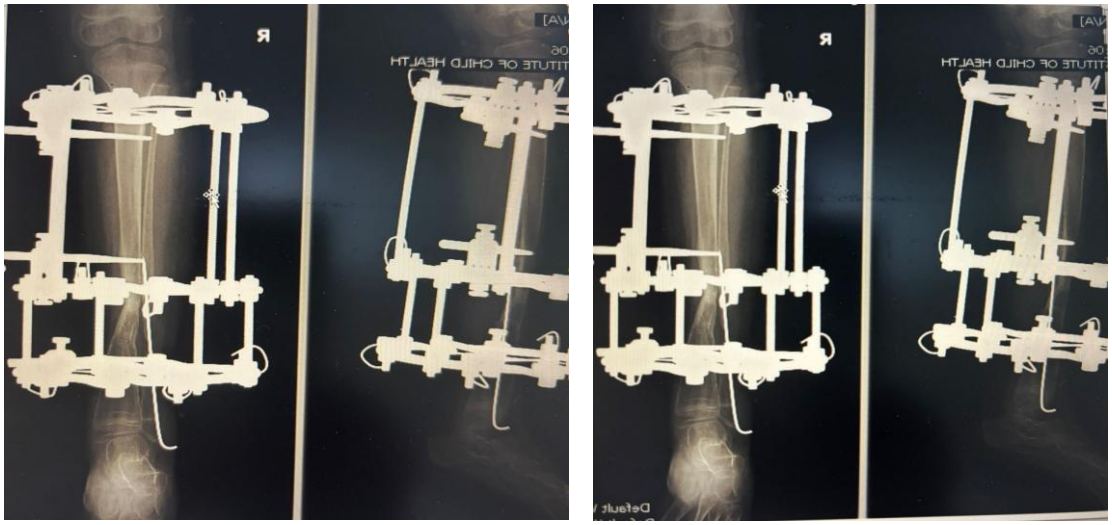
Union with Single Surgery:

- Our Study: 33.33%
- EPOS Study: 63%

Summary of Key Findings:

1. **Sound Union:** Achieved in a significant proportion of patients, particularly those undergoing multiple procedures.
2. **Complications:** Refracture and limb length discrepancy were notable complications, but refracture rates in our study were lower compared to some literature reports.
3. **Need for Multiple Surgeries:** Patients with more severe classifications (e.g., Boyd type VI) often require more than one definitive surgical procedure.
4. **Treatment Modalities:** Various surgical techniques were employed with tailored approaches based on individual patient characteristics and needs.





CONCLUSION:

The findings from this study highlight the challenges in managing congenital pseudoarthrosis of the tibia and underscore the importance of individualized treatment plans. While sound union was achieved in over half of the cases, the need for multiple surgeries and the management of complications such as limb length discrepancy and refracture remain critical aspects of patient care. Future studies with larger patient cohorts and more longer follow-up periods are essential to further refine treatment strategies and improve outcomes for patients with CPT.

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