

# A good index surgery for congenital pseudarthrosis of the tibia minimizes complications following surgery

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## Aims

Congenital pseudarthrosis of the tibia (CPT) has traditionally been a difficult condition to treat, with high complication rates, including nonunion, refractures, malalignment, and leg length discrepancy. Surgical approaches to treatment of CPT include intramedullary rodding, external fixation, combined intramedullary rodding and external fixation, vascularized fibular graft, and most recently cross-union. The current study aims to compare the outcomes and complication rates of cross-union versus other surgical approaches as an index surgery for the management of CPT. Our hypothesis was that a good index surgery for CPT achieves union and minimizes complications such as refractures and limb length discrepancy.

## Methods

A multicentre study was conducted involving two institutions in Singapore and China. All patients with CPT who were surgically managed between January 2009 and December 2021 were included. The patients were divided based on their index surgery. Group 1 included patients who underwent excision of hamartoma, cross-union of the tibia and fibula, autogenic iliac bone grafting, and internal fixation for their index surgery. Group 2 included patients who underwent all other surgical procedures for their index surgery, including excision of hamartoma, intramedullary rodding, and/or external fixation, without cross-union of the tibia and fibula. Comparisons of the rates of union, refracture, limb length discrepancy, reoperations, and other complications were performed between the two groups.

## Results

A total of 36 patients were included in the study. Group 1 comprised 13 patients, while Group 2 comprised 23 patients. The mean age at index surgery was four years (1 to 13). The mean duration of follow-up was 4.85 years (1.75 to 14). All patients in Group 1 achieved bony union at a mean of three months (1.5 to 4), but ten of 23 patients in Group 2 had nonunion of the pseudarthrosis ( $p = 0.006$ ). None of the patients in Group 1 had a refracture, while seven of 13 patients who achieved bony union in Group 2 suffered a refracture ( $p = 0.005$ ). None of the patients in Group 1 had a limb length discrepancy of more than 2 cm, while ten of 23 patients in Group 2 have a limb length discrepancy of more than 2 cm ( $p = 0.006$ ). In Group 1, four of 13 patients had a complication, while 16 of 23 patients in Group 2 had a complication ( $p = 0.004$ ). Excluding removal of implants, four of 13 patients in Group 1 had to undergo additional surgery, while 18 of 23 patients in Group 2 had to undergo additional surgery following the index surgery ( $p = 0.011$ ).

## Conclusion

A good index surgery of excision of hamartoma, cross-union of the tibia and fibula, autogenic iliac bone grafting, and internal fixation for CPT achieves union and minimizes complications such as refractures, limb length discrepancy, and need for additional surgeries.

## Take home message

- A good index surgery of excision of hamartoma, cross-union of the tibia and fibula, autogenic iliac bone grafting, and internal fixation for congenital pseudoarthrosis of the tibia achieves union and minimizes complications, such as refractures, limb length discrepancy, and the need for additional surgeries.

## Introduction

Congenital pseudoarthrosis of the tibia and fibula (CPT) was first described by Paget in 1891.<sup>1</sup> It is characterized by a spontaneous fracture that heals poorly with routine treatment.<sup>1</sup> The aims of surgical management of CPT are to achieve bony union of pseudoarthrosis, prevent refracture, and restore the alignment and length of the leg.<sup>2</sup> However, despite the multiple surgical options available, complication rates remain high, and there is a general acceptance that nonunion, refractures, malalignment, and leg length discrepancy are common pathways of the condition, with nonunion rates reported to be up to 68% and refracture rates up to 50%.<sup>3</sup>

Surgical approaches to treatment of CPT include intramedullary rodding, external fixation, combined intramedullary rodding and external fixation, vascularized fibular graft, and, most recently, cross-union.<sup>4,5</sup> The cross-union technique was introduced in 2011 by Choi et al<sup>6</sup> and in 2012 by Paley.<sup>7</sup> Choi et al<sup>6</sup> performed cross-union by converging the fibula towards the tibia, bone grafting between the tibia and fibula, then fixing the tibia with an intramedullary rod across the ankle joint and with external fixation. Paley<sup>7</sup> performed cross-union by placing intramedullary rods across both the tibia and fibula without crossing the ankle joint, bone grafting between the tibia and fibula with bone morphogenetic protein 2, applying a circular fixator, and using zolendronic acid preoperatively and postoperatively. Both Choi et al<sup>6</sup> and Paley<sup>7</sup> reported a 100% success rate with cross-union, achieving union and without refractures.

Multiple studies have since been published to report the outcomes of cross-union. Cross-union has also been reported to be effective in the management of refractory CPT patients.<sup>8</sup> A systematic review was conducted in 2022 to compare the outcomes of cross-union and other surgical approaches. They reported that the cross-union technique fared better than the other surgical approaches but cautioned that there are few studies on cross-union, with no direct comparison between the approaches.<sup>5</sup>

The current study therefore aims to compare the outcomes and complication rates of cross-union with other surgical approaches as an index surgery for the management of CPT. Our hypothesis was that a good index surgery for CPT achieves union and minimizes complications such as refractures and limb length discrepancy.

## Methods

A multicentre study was conducted involving two institutions in Singapore and China. All patients with CPT who were surgically managed between January 2009 and December 2021 were included. The diagnosis of CPT was confirmed based on pathological examination of the resected specimen for all patients. All patients had a minimum of one-year follow-up.

A retrospective review of the patients' medical records was performed. Demographic data including the patient's age at index surgery and sex were recorded. The patient's grade of CPT was also recorded based on the Crawford classification (Table I).<sup>9</sup> Surgical details of the patient's index surgery, as well as the subsequent surgeries, were detailed, including those that were performed prior to referral to the institution. Radiographs of the patients were reviewed by trained orthopaedic surgeons (SHST, YP) to document the presence or absence of union, refracture, and limb length discrepancy. Union was defined as a formation of bridging calluses across the pseudoarthrosis in at least three of four cortices on anteroposterior and lateral radiographs. Refracture was defined as the presence of pain and a new fracture line on radiograph following primary bone union. The limb lengths of bilateral tibia at the same timepoint were measured from the proximal tibial physis to the distal tibial physis, and any discrepancy of 2 cm or more was considered a limb length discrepancy. All complications following the index surgery were also recorded.

All patients were divided based on their index surgery. Group 1 included patients who underwent excision of hamartoma, cross-union of the tibia and fibula, autogenic iliac bone grafting, and internal fixation for their index surgery. This was done with or without bone morphogenetic protein 2 and bisphosphonates. Group 2 included patients who underwent all other surgical procedures for their index surgery, including excision of hamartoma, intramedullary rodding, and/or external fixation, without cross-union of the tibia and fibula. Comparisons of the rates of union, refracture, limb length discrepancy, reoperations, and other complications were performed between the two groups.

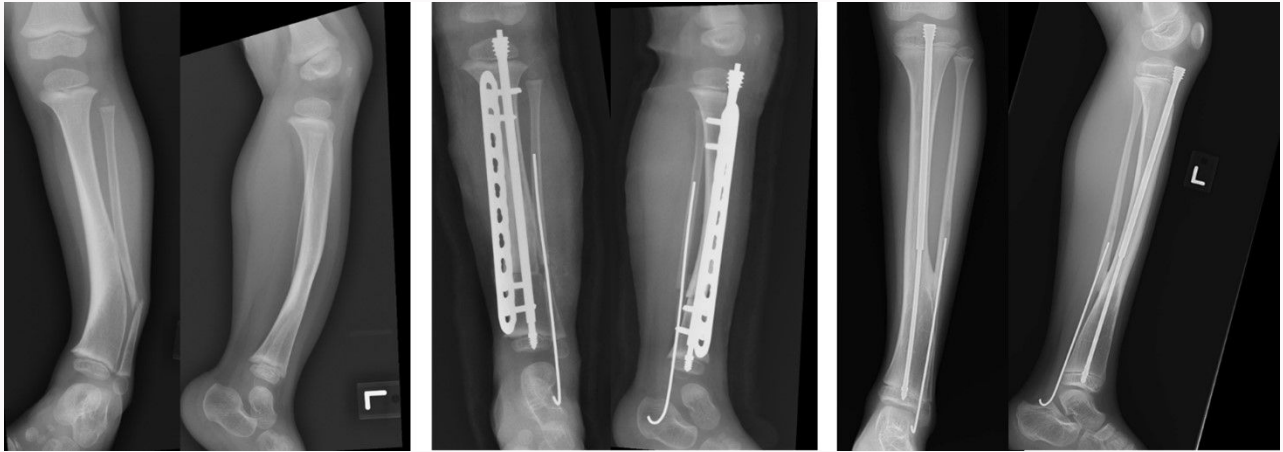
## Statistical analysis

Post hoc power analysis was performed. With the significance level fixed at 5%, the statistical power of the study was 90.3%, giving a sample size of 13 patients in Group 1 and 23 patients in Group 2, with a union rate of 100% in Group 1 and 56.5% in Group 2. SPSS v. 22.0 (IBM, USA) was used for statistical analysis. Fisher's exact test was used to perform the statistical analysis in view of the small sample size. A p-value < 0.05 was considered statistically significant.

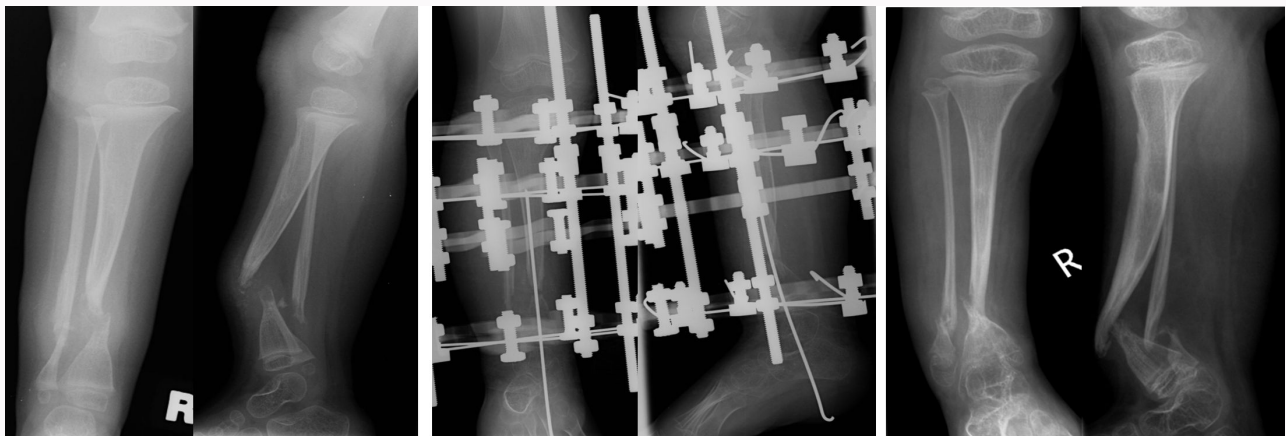
## Results

A total of 36 patients were included in the study: 13 patients underwent excision of hamartoma, cross-union of the tibia and fibula, autogenic iliac bone grafting, and internal fixation for their index surgery (Group 1; Figure 1); 23 patients underwent other surgical procedures without cross-union for their index surgery, including excision of hamartoma, intramedullary rodding, and/or external fixation (Group 2; Figure 2).

The mean age at index surgery was four years (1 to 13). There were 16 females and 14 males. In Group 1, there were four patients with Crawford 3 pseudoarthrosis and nine patients with Crawford 4 pseudoarthrosis. In Group 2, there were seven patients with Crawford 2 pseudoarthrosis, seven patients with Crawford 3 pseudoarthrosis, and nine patients with Crawford 4 pseudoarthrosis. Surgery was performed in patients with Crawford 2 pseudoarthrosis prophylactically for prevention of fracture and union of pseudoarthrosis. The mean duration of



**Fig. 1** Serial radiographs in the anteroposterior and lateral views of a two-year-old female patient in Group 1 who underwent excision of hamartoma, cross-union of the tibia and fibula, autogenic iliac bone grafting, and internal fixation. a) Initial radiographs of the patient, who was diagnosed with congenital pseudarthrosis of the tibia. b) Radiographs following surgical excision of hamartoma, cross-union of the tibia and fibula, autogenic iliac bone grafting, and internal fixation. c) Radiographs at latest follow-up of the patient at seven years old demonstrating good cross-union, with the fixation plate removed due to implant prominence.



**Fig. 2** Serial radiographs in the anteroposterior and lateral views of a two-year-old male patient in Group 2 who underwent excision of hamartoma, intramedullary rodding, and external fixation without cross-union. a) Initial radiographs of the patient, who was diagnosed with congenital pseudarthrosis of the tibia. b) Radiographs following surgical excision of hamartoma, intramedullary rodding, and external fixation without cross-union. c) Radiographs at follow-up demonstrating persistent nonunion, requiring multiple subsequent surgical procedures.

**Table 1.** Description of the Crawford classification.

Crawford classification	Description
Crawford 1	Anterolateral bow with dense medullary canal
Crawford 2	Anterolateral bow with increased medullary canal and a tubulation defect
Crawford 3	Anterolateral bow with a cystic lesion
Crawford 4	Anterolateral bow with fracture

follow-up was 4.85 years (1.75 to 14). At the conclusion of the study, three patients had reached skeletal maturity.

All patients in Group 1 achieved bony union at an average of three months (1.5 to 4); but ten of 23 patients in Group 2 had nonunion of the pseudarthrosis ( $p = 0.006$ ). None of the patients in Group 1 had a refracture, while seven of 13 patients who achieved bony union in Group 2 suffered a refracture ( $p = 0.005$ ). None of the patients in Group 1 had a limb length discrepancy of more than 2 cm, while ten of 23 patients in Group 2 have a limb length discrepancy of more than 2 cm ( $p = 0.006$ ).

Other complications in Group 1 included two patients with rod migration, two patients with osteomyelitis, and one patient with varus deformity of the ankle. Other complications in Group 2 included three patients with osteomyelitis and two patients with varus deformity of the ankle. Including all the complications, four of 13 patients in Group 1 had a complication following the index surgery, while 16 of 23 patients in Group 2 had a complication following the index surgery ( $p$

= 0.004). Excluding removal of implants, four of 13 patients in Group 1 had to undergo additional surgery following the index surgery, while 18 of 23 patients in Group 2 had to undergo additional surgery following the index surgery ( $p = 0.011$ ).

## Discussion

The principal finding of the study was that a good index surgery of excision of hamartoma, cross-union of the tibia and fibula, autogenic iliac bone grafting, and internal fixation for CPT achieves union and minimizes complications such as refractures, limb length discrepancy, and the need for additional surgeries.

These results echo the findings of the systematic review published by Rastogi and Agarwal,<sup>5</sup> which compared the outcomes of cross-union with other surgical approaches in the management of CPT.<sup>5</sup> In their systematic review, they identified a 100% union rate and 22.5% refracture rate for cross-union compared with 65.3% to 84.2% union rate and 33.7% to 48.1% refracture rate for other surgical approaches. However, in their review, while they concluded that outcomes of the cross-union technique fared better than other surgical approaches, they cautioned that studies on cross-union are few and longer follow-up is lacking.<sup>5</sup> To our knowledge, our current study therefore served as the first to compare directly between the surgical approaches for the outcomes of cross-union and other surgical approaches in the management of CPT.

The findings of the current study are promising as the outcomes of traditional methods of surgical management of CPT have always remained dismal, with a general acceptance of a high likelihood of nonunion, refractures, malalignment, and leg length discrepancy.<sup>3</sup> This therefore prompted a shift in the management of CPT towards cross-union of the tibia and fibula when introduced with good results in 2011 and 2012 by Choi et al<sup>6</sup> and Paley.<sup>7</sup> The recognition that cross-union of the tibia and fibula indeed yields better results than other surgical approaches further affirms the need for a paradigm shift in the surgical management of CPT, to minimize subsequent complications including nonunion, refracture, and limb length discrepancy in a growing child.

Despite this, it is important to recognize that surgical management of pseudarthrosis of the tibia with cross-union is still fraught with challenges. While successful with a lower complication rate, cross-union can still lead to complications, including rod migration, infection, and deformities of the ankle, as seen in our series.

Our study also faced several limitations. First, the sample size of the study is small. This is because CPT is a rare condition with a reported incidence of one in 60,000 to 250,000,000 live births.<sup>10</sup> The number of patients with CPT managed in each institution is therefore small, prompting the need for collaboration across institutions such as in this study. Further collaborations between multiple institutions

would allow larger-scale studies to be performed to validate the findings of the studies. Second, only objective outcomes were used in the current study, and there were no functional outcome scores recorded. This is because the patients treated were as young as one year old, and it is difficult to measure subjective functional outcome scores in this group of patients. Third, the majority of the patients were not followed up until skeletal maturity prior to the conclusion of this study, therefore later complications might have been missed. A follow-up study subsequently would be helpful for the long-term comparison and assessment of these patients. Lastly, the index surgeries for these patients were performed in multiple institutions, and many of the patients were operated in other institutions prior to their referral to our institution. This therefore introduced a confounding bias to the study, as not all the patients of all the original institutions were included. Further collaborations between multiple institutions would reduce this drawback, as more patients could be included.

In conclusion, good index surgery of excision of hamartoma, cross-union of the tibia and fibula, autogenic iliac bone grafting, and internal fixation for CPT achieves union and minimizes complications, such as refractures, limb length discrepancy, and the need for additional surgeries.

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### Data sharing

The data that support the findings for this study are available to other researchers from the corresponding author upon reasonable request.

### Ethical review statement

The study was performed in accordance with the ethical standards of both institutional Domain Specific Review Board research committees.

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