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Use of an Intramedullary Rod for Treatment of Congenital Pseudarthrosis of the Tibia
A Long-Term Follow-up Study
BY MATTHEW B. DOBBS, MD, MARGARET M. RICH, MD, PhD, J. ERIC GORDON, MD, DEBORAH A. SZYMANSKI, RN, AND PERRY L. SCHONECKER, MD

Investigation performed at St. Louis Shriners Hospital for Children, St. Louis Children's Hospital, and Washington University School of Medicine, St. Louis, Missouri

Background: The treatment of congenital pseudarthrosis of the tibia remains difficult and controversial. The purpose of this study was to evaluate the long-term results of a technique consisting of excision of the pseudarthrosis, autologous bone-grafting, and insertion of a Williams intramedullary rod into the tibia.

Methods: Twenty-one consecutive patients with congenital pseudarthrosis of the tibia were managed with this technique between 1978 and 1999, and the results were retrospectively reviewed. The mean age of the patients at the time of the latest follow-up was 17.2 years (range, seven to twenty-five years), and the mean duration of postoperative follow-up was 14.2 years (range, three to twenty years).

Results: Initial consolidation occurred in eighteen of the twenty-one patients. Refracture occurred in twelve patients; five fractures healed with closed treatment, five healed after an additional surgical procedure, and two ultimately required amputation. Ten patients had an ankle valgus deformity after tibial union. Eleven patients had a residual limb-length discrepancy of >2 cm; six required a contralateral distal femoral and/or proximal tibial epiphyseodesis, two had a tibial lengthening, and one used a shoe-lift. Five patients had an amputation: two, because of a recalcitrant fracture; two, because of a limb-length discrepancy (6 and 9 cm); and one, because of a chronic lower-extremity deformity.

Conclusions: This technique produced a satisfactory long-term functional outcome in sixteen of twenty-one patients and should be considered for the management of congenital pseudarthrosis of the tibia.

Level of Evidence: Therapeutic study, Level IV (case series [no, or historical, control group]). See Instructions to Authors for a complete description of levels of evidence.

Congenital pseudarthrosis of the tibia is one of the most challenging problems in pediatric orthopaedics. The natural history is persistent instability and progressive deformity1-5. Treatment options have varied greatly and have included both operative and nonoperative approaches6-9. Although no single method has proven ideal, the highest rates of union have been reported after surgery10-16. Currently, three different surgical approaches—vascularized bone-grafting, intramedullary stabilization, and external fixation—have been relatively successful in obtaining initial union of the pseudarthrosis16-24. However, despite that success10,12,14-20,23, the quality and longevity of the consolidation and the future function of the involved extremity remain uncertain15,17,26. This uncertainty stems from the lack of long-term follow-up studies of the different surgical methods, particularly the lack of studies identifying the rates of refracture, limb-length discrepancy, and residual angular deformity. All of those complications can compromise the functional outcome, even though the pseudarthrosis may have healed14,17,26.

Since 1978, we have used a solid intramedullary rod combined with excision of the pseudarthrosis and autogenous iliac crest bone-grafting for the treatment of congenital pseudarthrosis of the tibia. This is a modification of the original technique described by Charnley29, and it involves use of the two-part Williams intramedullary solid rod to stabilize the pseudarthrosis site and transfix the ankle joint30. Our preliminary results were re-
ported in 1992\(^a\). The purpose of the current study was to both evaluate the long-term results of this technique and provide an update of our surgical and post-consolidation strategy when utilizing the transarticular Williams-rod stabilization technique.

**Materials and Methods**

We retrospectively reviewed the results in twenty-one consecutive patients with congenital pseudarthrosis of the tibia who had been managed with this technique at the St. Louis Shriners Hospital for Children and the St. Louis Children’s Hospital between 1978 and 1999. The mean age of the patients when the surgery was performed was 5.1 years (range, eleven months to eleven years) (Table I). Twelve patients were girls and nine were boys. All patients had an established pseudarthrosis of the tibia, which had developed after a fracture in twenty patients and after an osteotomy in one. The initial fracture occurred at a mean age of 2.5 years (range, one month to six years). Twelve children had neurofibromatosis, one had Ehlers-Danlos syndrome, and the remaining eight had no associated diagnoses. The left tibia was involved in eleven patients and the right, in ten.

Previous surgery had been performed in eleven patients prior to referral to our institution. A single compression plate and screws and autogenous bone graft had been used in five patients; bone-grafting without internal fixation had been attempted in three patients (Cases 4, 19, and 21); a vascularized fibular graft and an intramedullary rod had been used in one patient (Case 7); and an intramedullary rod with autogenous bone graft had been used in another (Case 3). A pseudarthrosis developed after an osteotomy and fixation with crossed Kirschner wires for treatment of anterolateral tibial bowing in one child. Eight of the remaining ten patients had been managed nonoperatively with casts and/or orthoses. Two patients had had no previous treatment. Electrical stimulation, either external (four patients) or with an implanted device (two patients), had been used in six patients as adjunctive treatment.

The pseudarthroses were classified radiographically with use of Boyd’s system, which includes six distinct radiographic types of anterolateral bowing deformity\(^b\). Three patients were born with anterolateral bowing and a defect in the tibia (Type I). Nine pseudarthroses developed in an hourglass constriction of the tibia (Type II) (Figs. 1-A, 1-B, and 1-C). Six pseudarthroses developed, without narrowing, at a sclerotic segment of bone (Type IV) (Figs. 2-A through 2-E), and one of these pseudarthroses developed after an osteotomy. The remaining three patients had an attenuated, dysplastic fibula at birth, and a tibial pseudarthrosis developed (Type V). All but three of the pseudarthroses were located in the distal third of the tibia.

Anteroposterior and lateral radiographs were made every three months postoperatively until osseous union was achieved and variably thereafter. Scanograms were used to measure any clinically evident limb-length discrepancy. All patients were examined by one of the senior authors at the time of the most recent follow-up. Particular attention was paid to the range of motion of the ankle, residual limb-length discrepancy, and any residual angular deformity at the ankle.

The number and type of additional surgical procedures following our initial procedure were recorded. The mean age at the latest follow-up examination was 17.2 years (range, seven to twenty-five years). The mean duration of follow-up after insertion of the intramedullary rod was 14.2 years (range, three to twenty years). Fourteen patients were skeletally mature at the time of the latest follow-up.

**Treatment Protocol**

The same operative technique was used for all patients. The patient is placed in a semilateral position on a radiolucent operating table with a support beneath the hip, and a sterile tourniquet is applied to the proximal part of the thigh. The entire lower extremity and ipsilateral iliac crest are prepared and draped. An iliac crest bone graft, consisting of adequate corticocancellous strips and cancellous bone, is obtained first, and the wound is closed. The hip support is removed, and the patient is positioned supine. An anterior longitudinal incision is made, extending several centimeters both proximal and distal to the pseudarthrosis site. The pseudarthrosis is completely excised until there is normal bleeding bone of the medullary canal of both the proximal and the distal tibial fragment. The resection shortens the tibia by 1.0 to 3.0 cm. The medullary canal of both tibial fragments isreamed with a drill and/or a curet.

The tibia is then stabilized with a Williams rod (Figs. 3-A and 3-B). This rod consists of an indwelling rod and an insertion rod that either are available commercially (Zimmer, Warsaw, Indiana) or are custom-made for each patient from a hospital stock of Kirschner, Rush, and Ender rods. Preoperative planning is essential for selecting a rod of appropriate length and diameter. The length of rod that is needed is calculated on the basis of the expected length of the tibia after the pseudarthrosis has been excised and any angular deformity is corrected. The rod typically transfixes the ankle joint, and the anticipated remaining growth determines the appropriate location of the distal end of the rod at the time of surgery. With longitudinal growth, the distal part of the tibia, the ankle, and the foot migrate distally. Our current strategy is to minimize the duration of ankle immobilization by the rod. Usually, the distal end of the rod should extend well into the body of the calcaneus in children four years of age or less, whereas it should end in the talus in children five to ten years of age. Following consolidation, at approximately one to two years after rod placement, we perform a second operation to push the rod across the ankle joint. The rod is not placed across the ankle joint when the patient has a relatively proximal pseudarthrosis or is nine years of age or older.

The entire rod assembly is inserted into the medullary canal of the distal segment and is advanced with a power source antegrade across the distal part of the tibia, the ankle joint, and the subtalar joint and out through the heel pad. During the passage of the rod across the ankle joint, it is imperative that the foot be positioned to minimize the tendency for calcaneal and valgus angulation of the distal segment and/or hindfoot. The desired ankle position is neutral dorsiflexion-plantar flexion, verified clinically and with fluoroscopy.
The tibial segments are anatomically aligned at the pseudarthrosis site, and the rod is then driven retrograde into the proximal fragment. Again, direct visualization and the use of fluoroscopy are imperative to confirm that the rod is within the medullary canal and that the tibia is aligned in the coronal and sagittal planes. Occasionally, because of extensive deformity, an additional osteotomy of the proximal fragment is necessary to ensure both intramedullary passage of the rod and a more anatomic alignment of the tibia. Unscrewing the insertion rod one turn and noting the rod junction on fluoroscopy confirms the exact location of the distal end of the indwelling rod. The insertion rod is removed when an appropriate position has been achieved.

Fixation of the fibula adds stability to the construct if the fibula is fractured or if fibular shortening was necessary to achieve adequate apposition of the tibia. Fibular fixation is often less than optimal because of the small size of the medullary canal. When stabilization is performed, we prefer intramedullary fixation with an appropriately sized Kirschner wire, which is placed into the distal fragment through an open incision after excision of the pseudarthrosis. The wire is directed antegrade, through the distal tip of the fibula, and then retrograde into the proximal fragment.

The previously harvested cancellous and corticocancellous bone graft is then placed around the tibial pseudarthrosis site and is secured with circumferentially placed absorbable heavy suture material. Syngraft (GenSci Regeneration Sciences, Irvine, California) is added in children who are four years old or younger, because earlier bone growth will lead to a more durable construct.
years of age or less. A suction drain is placed over the pseudarthrosis site. The deep fascia of the leg is not closed. The subcutaneous tissue and skin are closed in separate layers.

According to the protocol, children six years of age or less are treated with a one and one-half spica cast to ensure minimal rotatory stress at the pseudarthrosis site. The spica cast is replaced with an above-the-knee cast at six to eight weeks postoperatively. Cast immobilization is discontinued at approximately four to six months postoperatively, once clinical and radiographic examination indicates that union has occurred. Older children are treated with an above-the-knee cast with the knee flexed 30° for an average of four months. Once the cast is removed, the involved extremity is protected with a custom-fabricated clamshell knee-ankle-foot orthosis with a locked ankle joint and a free knee joint. Adequate circumferential leg contact is an essential feature of this orthosis, which is worn twenty-four hours a day. Walking with progressive weight-bearing is initiated. For children nine years of age or older, the knee-ankle-foot orthosis can be converted to a total contact ankle-foot orthosis as consolidation progresses.

With longitudinal growth of the tibia, the rod remains in place and the distal part of the tibia and the foot migrate away from its distal end until the ankle joint is free. This took approximately three years to occur in the patients treated earlier in our study. Currently, to minimize the duration of ankle joint immobilization and the potential for disruption of the articular cartilage during the gradual migration of the rod across the ankle joint, we surgically advance the rod across the

<table>
<thead>
<tr>
<th>Age When Tibial Rod Inserted (yr + mo)</th>
<th>Age at Refracture (yr + mo)</th>
<th>Treatment of Refracture</th>
<th>Fibular Surgery</th>
<th>Age at Latest Follow-up (yr + mo)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1 + 9</td>
<td>6 + 9</td>
<td>Cast</td>
<td>Fibular osteotomy; no fixation</td>
<td>14 + 2</td>
</tr>
<tr>
<td>6 + 1</td>
<td>—</td>
<td>—</td>
<td>Fibular osteotomy; no fixation</td>
<td>10 + 2</td>
</tr>
<tr>
<td>5 + 6</td>
<td>11 + 8</td>
<td>Cast; electrical stimulation</td>
<td>Pseudarthrosis excision; intramedullary rod</td>
<td>25 + 5</td>
</tr>
<tr>
<td>4 + 4</td>
<td>10 + 4</td>
<td>Cast; electrical stimulation</td>
<td>Fibular osteotomy; no fixation</td>
<td>23 + 6</td>
</tr>
<tr>
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<td>7 + 6</td>
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<tr>
<td>10 + 4</td>
<td>—</td>
<td>—</td>
<td>None</td>
<td>16 + 11</td>
</tr>
<tr>
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<td>11 + 8</td>
<td>Plate and screws × 2</td>
<td>Fibular osteotomy; no fixation</td>
<td>10 + 8</td>
</tr>
<tr>
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<td>2 + 6</td>
<td>Rod revised, bone graft</td>
<td>Fibular osteotomy; no fixation</td>
<td>7 + 0</td>
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<tr>
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<td>6 + 0</td>
<td>Rod revised, bone graft</td>
<td>Fibular osteotomy; no fixation</td>
<td>11 + 8</td>
</tr>
<tr>
<td>2 + 2</td>
<td>—</td>
<td>—</td>
<td>None</td>
<td>18 + 6</td>
</tr>
<tr>
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<td>10 + 3</td>
<td>Rod revised, bone graft</td>
<td>None</td>
<td>20 + 8</td>
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<tr>
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<td>10 + 4</td>
<td>Rod revised, bone graft</td>
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<td>20 + 7</td>
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<td>13 + 8</td>
<td>Rod revised, bone graft</td>
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<td>15 + 8</td>
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<td>Cast</td>
<td>Fibular osteotomy; no fixation</td>
<td>23 + 5</td>
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<tr>
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<td>9 + 9</td>
<td>Plate and screws</td>
<td>Pseudarthrosis excision; no fixation</td>
<td>24 + 9</td>
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<tr>
<td>3 + 11</td>
<td>—</td>
<td>—</td>
<td>Fibular osteotomy; no fixation</td>
<td>21 + 8</td>
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<tr>
<td>4 + 3</td>
<td>—</td>
<td>—</td>
<td>Pseudarthrosis excision; no fixation</td>
<td>22 + 8</td>
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<tr>
<td>7 + 0</td>
<td>—</td>
<td>—</td>
<td>Pseudarthrosis excision; intramedullary rod</td>
<td>24 + 2</td>
</tr>
<tr>
<td>3 + 8</td>
<td>—</td>
<td>—</td>
<td>Pseudarthrosis excision; intramedullary rod</td>
<td>12 + 5</td>
</tr>
</tbody>
</table>
ankle joint, with a pusher rod, when its distal end approaches the articular surface of the talus. The orthosis is unlocked at the ankle once the rod lies proximal to the ankle joint. The orthosis is worn until the patient reaches skeletal maturity and then for sports activities thereafter.

**Results**

The technique resulted in initial consolidation of the pseudarthrosis in eighteen of the twenty-one extremities, at a mean of sixteen months (range, five to forty-three months) (Figs. 2-A through 2-E). Union was achieved in the three remaining tibiae after a second bone-grafting procedure. Two patients had both a proximal and a distal tibial osteotomy in order to realign the tibia at the time of the initial insertion of the rod. Growth of the tibia resulted in the distal end of the rod being located proximal to the ankle joint in twelve patients, at a mean of thirty-four months (range, twenty to fifty-eight months) postoperatively. In three patients, treated early in the series, the rod continued to traverse the ankle joint and was removed in an attempt to regain ankle motion; all three patients sustained a refracture, which required repair of the pseudarthrosis and reinsertion of the rod (Table I). An additional three patients, in whom the distal part of the tibia failed to migrate away from the distal end of the nail, had delayed union of the pseudarthrosis and were treated with repeat bone-grafting and replacement of the rod. In those patients, the proximal portion of the replacement rod was surrounded with methylmethacrylate, through a separate bone window created in the proximal tibial metaphysis, to help anchor it to the proximal part of the tibia. With subsequent growth, the distal part of the tibia and the foot migrated away from the rod in all three patients. Eight patients in whom the rod was no longer traversing the ankle joint regained functional motion of the ankle, which we defined as a total arc of motion (maximum dorsiflexion to maximum plantar flexion) of ≥25°. Four patients had nonfunctional ankle motion even though the rod was no longer crossing the ankle joint, and in one patient the rod was still transfixing the ankle joint at the time of this review. We purposefully did not transfix the ankle joint with the Williams rod in three patients; all had a functional ankle range of motion at the time of final follow-up.

Twelve patients had a refracture after the initial consoli-
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Reduction and internal fixation with a plate and screws and autogenous iliac crest bone-grafting, and the remaining three patients were treated with revision intramedullary nailing and autogenous bone-grafting. Two patients with recurrent fractures did not have healing and underwent amputation.

Ten patients had a fibular pseudarthrosis at the time of the intramedullary nailing of the tibia. Four of these patients were treated with excision of the fibular pseudarthrosis and intramedullary fixation of the fibula, two had excision of the pseudarthrosis alone, and the remaining four patients had no fibular surgery. Two of the six patients treated with excision of the fibular pseudarthrosis with or without internal fixation had a refracture of the tibia compared with two of the four patients who had a fibular pseudarthrosis but no fibular surgery. There was a trend toward increased ankle valgus deformity postoperatively in the patients who had had a fibular pseudar-

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throsis preoperatively (six of ten patients), even when the fibular lesion had been excised and had subsequently united. Only four of the eleven patients who had not had a fibular pseudarthrosis preoperatively had a clinically relevant ankle valgus deformity postoperatively. Of the eleven patients who had an intact fibula at the time of surgery, eight had a fibular osteotomy to aid in the alignment of the tibial fragments; of these eight patients, six had a refracture of the tibia, and three of the six patients eventually had an amputation. Of the three patients in whom an intact fibula was not treated with an osteotomy, two had a refracture of the tibia but no amputations were required.

Eleven patients had a clinically relevant limb-length discrepancy, averaging 5 cm (range, 2 to 9 cm). Six of these patients were treated definitively with a contralateral distal femoral and/or proximal tibial epiphyseodesis, with a mean predicted limb-length discrepancy of 4 cm (Table II). Two patients were treated with tibial lengthening with the Ilizarov method, with the corticotomy for the lengthening performed in the proximal part of the tibia. One patient with a 3-cm discrepancy was treated with a shoe-lift. Two other patients, with 6 and 9-cm limb-length discrepancies and associated ankle valgus deformity, underwent amputation. Of the remaining ten patients, who had a limb-length inequality of <2 cm, three were still skeletally immature at the time of writing.

Anteroposterior weight-bearing radiographs of the ankle revealed an ankle valgus deformity, after consolidation of the pseudarthrosis and growth of the distal part of the tibia to free the ankle joint, in ten patients. In nine patients, the valgus deformity was successfully treated with medial distal and/or proximal physeal stapling of the tibia at a mean age of eleven years (range, eight to fourteen years). After the valgus deor-
Five patients required amputation because of chronic lower-extremity dysfunction: two, because of recurrent fracture nonunion; two, because of limb-length discrepancy; and one, because of residual angular deformity. Three of the five patients had a below-the-knee amputation, and the other two initially had a Syme amputation. However, the two Syme amputations were revised to below-the-knee amputations because of instability of the stump and difficulty with prosthetic fitting. All five patients who had an amputation were fully weight-bearing with the use of a standard below-the-knee prosthesis for walking at the time of follow-up. There were no deep infections, neurovascular complications, or broken rods.

Discussion

Currently, three surgical techniques (vascularized fibular grafting, intramedullary stabilization, and external fixation) are being used with relative success for the treatment of congenital pseudarthrosis of the tibia\(^1\)\(^{-}\)\(^{23}\). However, there is a lack of long-term studies of these methods. As Boyd suggested years ago, the true success of treatment of congenital pseudarthrosis of the tibia in a growing child can be known only by following the patient until maturity\(^1\).

The current technique of free vascularized fibular grafting has been well described\(^24\), and several authors have reported success in obtaining initial tibial consolidation\(^15\),\(^17\),\(^24\),\(^29\),\(^30\),\(^32\). However, there are many potential pitfalls. A secondary bone-grafting procedure is frequently necessary to obtain union at the pseudarthrosis site\(^24\). In addition, refracture and recurrent nonunion at one end of the graft site\(^17\),\(^24\), or in the graft itself\(^31\), are not uncommon following consolidation. Another problem is malalignment—either anterior bowing or valgus deformity—of the affected tibia\(^24\). These angular deformities do not remodel and often are progressive\(^12\),\(^23\). In addition, progressive valgus ankle deformity on the donor side with proximal migration of the distal part of the fibula is a problem following harvest of a vascularized fibular graft in children\(^31\).\(^33\). To prevent this complication, tibiofibular metaphyseal synostosis (the Langenskiöld procedure) has been recommended\(^24\),\(^33\). However, in a recent review, this procedure was found to only delay, not prevent, the development of ankle valgus in children after harvesting of a vascularized fibular graft\(^33\).

Numerous authors have reported on the use of a circular external fixator in the treatment of congenital pseudarthrosis of the tibia\(^20\),\(^26\),\(^34\)-\(^36\). Fifteen patients with a total of sixteen congenital pseudarthroses of the tibia treated with the Ilizarov apparatus were reported on by Paley et al.\(^20\). Initial union occurred in fifteen of the sixteen extremities after one treatment and in one extremity after one additional treatment. The mean age at the time of treatment was eight years compared with a mean age of five years at the time of the initial rod placement in the current series. Five refractures occurred in the series by Paley et al., and all were successfully treated with a variety of additional procedures. In a more recent study, Boero et al.\(^26\) reported failure of the Ilizarov technique in eight of twenty-one patients with congenital pseudarthrosis of the tibia. Consolidation was obtained in twelve of fourteen patients who were operated on at the age of thirteen years.
of five years or older and in only one of seven patients who were treated at the age of four years or younger.

Umber et al.\textsuperscript{10} popularized the use of the two-part Williams intramedullary solid rod in North America. We reported our initial results with the use of this technique in 1992\textsuperscript{16}. Although union was obtained in all ten patients at a mean of six years postoperatively, we observed problems in many patients, including failure of distal tibial migration, stiffness of the ankle, limb-length discrepancy, and residual angular deformities at the ankle. In the interim, we have continued to follow these patients, added others to our series, and modified the technique to improve our results.

Despite reports of ankle stiffness following long-term transfixation of the tibiotalar and subtalar joints\textsuperscript{1,37}, only a few patients in our study did not regain functional ankle motion once the rod no longer transfixed the ankle joint. In order to minimize loss of ankle motion, we now recommend surgically advancing the rod out of the hindfoot and ankle joint soon after the pseudarthrosis has healed (usually less than two years after rod insertion). The initial choice of rod length is crucial to ensure that, at the time of rod advancement, it is not pushed across the proximal tibial physis and/or into the knee joint. In general, if the ankle joint is going to be transfixed for less than two years, the rod should be initially placed no less than 2 cm from the proximal tibial physis. In cases in which the pseudarthrosis is more proximal and/or the patient is nine years of age or older, intramedullary fixation should be obtained without transfixing the ankle joint.

In an attempt to decrease the prevalence of ankle stiffness, custom interlocking intramedullary nails that do not transfix the ankle joint were developed and are now available; they may be appropriate for patients in whom the segment distal to the pseudarthrosis is large enough to accommodate adequate fixation\textsuperscript{38}. Unrestricted ankle motion, however, may allow the distal tibial segment to move sufficiently to delay or even prevent union of the pseudarthrosis. Another disadvantage of locked nails is that even custom nails are too large for use in many younger children. The solid two-part Williams intramedullary rod can be used in children of all ages and has distinct advantages in terms of strength and the ability to provide rigid immobilization and protect the consolidating pseudarthrosis. In the current series, temporary transfixation of the tibiotalar and subtalar joints did not negatively affect the long-term functional result.

Failure of the tibia to migrate distally from the rod was another problem in our patients. Early in the series, this was treated by simply removing the rod, but refracture soon occurred in all three patients. Currently, failure of the tibia to migrate distally is treated with reinsertion of a rod at the initial pseudarthrosis site. The replacement rod is secured to the proximal part of the tibia with methylmethacrylate applied to its proximal 2 cm through a cortical window. Fixing the rod proximally facilitates migration of the rod through the hindfoot with growth of the distal part of the tibia.

Refracture is common in patients with congenital pseudarthrosis of the tibia, despite apparently solid clinical and radiographic union\textsuperscript{16,39-41}. This complication occurred in twelve of our patients. Three of the refractures occurred early in the series, after removal of the rod in an attempt to regain ankle motion. We agree with others who have concluded that it is inadvisable to remove the rod after union\textsuperscript{20,21,29,42}. An attempt was made to identify risk factors for refracture in this patient population. The twelve patients who had a refracture were younger at the time of the initial fracture than those who did not have a refracture. This finding is consistent with the findings of Roach et al.\textsuperscript{43} as well as those of Kim and Weinstein\textsuperscript{44}, who stated that late-onset fracture and pseudarthrosis is a more benign form of the lesion.
TABLE II Secondary Procedures and Outcomes

<table>
<thead>
<tr>
<th>Case</th>
<th>Syndesmosis Placement</th>
<th>Additional Procedures</th>
<th>Ankle Range of Motion*</th>
<th>Final Limb-Length Discrepancy† (cm)</th>
<th>Assistive Device‡</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Yes</td>
<td>1. Proximal tibial osteotomy for valgus deformity; new rod inserted 2. Syme amputation 3. Conversion to a below-the-knee amputation</td>
<td>NA</td>
<td>NA</td>
<td>Prosthesis</td>
</tr>
<tr>
<td>2</td>
<td>No</td>
<td>1. Rod advancement 2. Med. proximal physeal stapling of tibia</td>
<td>Functional</td>
<td>2.5</td>
<td>KAFO</td>
</tr>
<tr>
<td>3</td>
<td>No</td>
<td>None</td>
<td>Functional</td>
<td>2.0</td>
<td>AFO</td>
</tr>
<tr>
<td>4</td>
<td>No</td>
<td>Med. proximal physeal stapling of tibia</td>
<td>Functional</td>
<td>3.0</td>
<td>—</td>
</tr>
<tr>
<td>5</td>
<td>No</td>
<td>Rod removal</td>
<td>Rigid</td>
<td>0.4</td>
<td>—</td>
</tr>
<tr>
<td>7</td>
<td>Yes</td>
<td>Contralateral epiphyseodesis</td>
<td>Functional</td>
<td>1.0</td>
<td>AFO</td>
</tr>
<tr>
<td>8</td>
<td>No</td>
<td>Rod advancement</td>
<td>Functional</td>
<td>None</td>
<td>KAFO</td>
</tr>
<tr>
<td>9</td>
<td>No</td>
<td>None</td>
<td>Rod still across ankle</td>
<td>1.5</td>
<td>KAFO</td>
</tr>
<tr>
<td>10</td>
<td>No</td>
<td>1. Below-the-knee amputation 2. Stump revision for overgrowth</td>
<td>NA</td>
<td>NA</td>
<td>Prosthesis</td>
</tr>
<tr>
<td>11</td>
<td>No</td>
<td>Rod advancement</td>
<td>Rigid</td>
<td>None</td>
<td>KAFO</td>
</tr>
<tr>
<td>12</td>
<td>Yes</td>
<td>1. Contralateral epiphyseodesis 2. Med. proximal physeal stapling of tibia</td>
<td>Functional</td>
<td>None</td>
<td>AFO</td>
</tr>
<tr>
<td>13</td>
<td>No</td>
<td>1. Rod revised and methylmethacrylate applied to proximal part of rod; bone graft 2. Syme amputation 3. Conversion to a below-the-knee amputation</td>
<td>NA</td>
<td>NA</td>
<td>Prosthesis</td>
</tr>
<tr>
<td>14</td>
<td>No</td>
<td>1. Rod revised and methylmethacrylate applied to proximal part of rod; bone graft</td>
<td>Rigid</td>
<td>None</td>
<td>KAFO</td>
</tr>
<tr>
<td>15</td>
<td>No</td>
<td>1. Ilizarov lengthening 2. Med. distal physeal stapling of tibia</td>
<td>Functional</td>
<td>1.0</td>
<td>KAFO</td>
</tr>
<tr>
<td>16</td>
<td>Yes</td>
<td>1. Med. distal physeal stapling of tibia 2. Staple and syndesmosis screw removal</td>
<td>Functional</td>
<td>None</td>
<td>—</td>
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<tr>
<td>17</td>
<td>No</td>
<td>1. Ilizarov lengthening 2. Contralateral epiphyseodesis</td>
<td>Rigid</td>
<td>1.0</td>
<td>AFO</td>
</tr>
<tr>
<td>18</td>
<td>Yes</td>
<td>1. Rod exchange and bone-grafting 2. Contralateral epiphyseodesis 3. Med. distal physeal stapling of tibia</td>
<td>Functional</td>
<td>1.5</td>
<td>AFO</td>
</tr>
<tr>
<td>20</td>
<td>Yes</td>
<td>1. Med. proximal physeal stapling of tibia 2. Contralateral epiphyseodesis 3. Rod advancement</td>
<td>Functional</td>
<td>1.5</td>
<td>KAFO</td>
</tr>
<tr>
<td>21</td>
<td>Yes</td>
<td>1. Med. proximal physeal stapling of tibia 2. Proximal tibial cortical window—application of methylmethacrylate to proximal rod</td>
<td>Functional</td>
<td>0.5</td>
<td>AFO</td>
</tr>
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</table>

*A functional range of motion was considered to be a total arc of ≥25°. †NA = not applicable because of amputation. ‡AFO = ankle-foot orthosis; KAFO = knee-ankle-foot orthosis.
However, with the small numbers of patients in our series, neither the presence of associated disorders (neurofibromatosis and Ehlers-Danlos syndrome) nor the Boyd type of the pseudarthrosis had significance in terms of predicting the prevalence of refracture.

The need for fibular surgery remains controversial. Johnston concluded that it is crucial to resect a fibular pseudarthrosis or, if the fibula is intact, to perform a fibular osteotomy in order to achieve optimal limb alignment and union. Similarly, in our series, the prevalence of tibial refracture was higher in the patients in whom the fibular pseudarthrosis was not resected (two of four) than in those who had a resection with or without internal fixation (two of six). However, unlike Johnston, we found no difference in outcome between the patients in whom an intact fibula had been osteotomized and those in whom an intact fibula had been left alone. The numbers of patients were too small for us to be able to draw conclusions on the efficacy of performing an osteotomy of an intact fibula.

Valgus deformity of the ankle can compromise the functional result in patients treated successfully with an intramedullary rod. We have speculated that the valgus deformity is more a natural outcome of the initial deformity (deficient fibula and lateral support) and less an iatrogenic complication of traversing the tibial physeal plate with the rod since the rod is positioned in the central portion of the physis. In our study, the majority of the ankle valgus deformities occurred in patients with a concomitant fibular pseudarthrosis, even when the fibular lesion had been treated and had subsequently united. Early in the series, we noted a tendency toward recurrent ankle valgus deformity in patients with a united fibular fragment and deficient lateral support. To prevent this complication, we now recommend placement of a syndesmosis screw once valgus correction has been achieved in patients with a deficient fibula. This technique has maintained correction in all six of our patients so treated.

Limb-length discrepancy is common after treatment of congenital pseudarthrosis of the tibia and was clinically relevant in half of our patients. It is primarily the result of chronic resorption of bone at the pseudarthrosis site prior to repair and secondarily as a result of the operative resection of the pseudarthrosis. We found that most cases can be treated successfully with an appropriately timed contralateral distal femoral and/or proximal tibial epiphyseodesis, a tibial lengthening procedure, or a shoe-lift.

Most cases of congenital pseudarthrosis of the tibia can be successfully treated with the modified Williams technique, but attention to detail is critical. On the basis of our twenty-four years of experience with this technique, we can make several recommendations. The essential surgical steps of the technique include complete resection of the tibial pseudarthrosis, intramedullary stabilization of the tibial and if possible the fibular pseudarthrosis, and iliac crest bone-grafting. In a young child with a typical distal tibial deformity, the intramedullary rod should extend into the hindfoot to temporarily immobilize the subtalar and ankle joints; the desired ankle position is neutral dorsiflexion-plantar flexion. For less common, more proximal lesions and/or in older children, the intramedullary rod should extend only to the distal tibial physis. A fibular pseudarthrosis, if present, should be resected and intramedullary fixation should be attempted with an appropriately sized Kirschner wire; an intact fibula should be osteotomized if shortening appears to be necessary to achieve adequate apposition of the tibial fragments. Although consolidation typically occurs following this treatment, refracture, failure of the rod to migrate, shortening, and ankle valgus deformity are typical problems. In many cases, these problems can be treated successfully with additional surgical procedures that spare the limb. Occasionally, despite aggressive surgical treatment, amputation is required because of chronic lower-extremity dysfunction, as occurred in five patients in our series. However, the Williams technique produced a satisfactory long-term outcome in sixteen of our twenty-one patients and should be considered as the initial operative procedure for congenital pseudarthrosis of the tibia.

References


