Congenital Pseudarthrosis of the Tibia

RESULTS OF TECHNICAL VARIATIONS IN THE CHARNLEY-WILLIAMS PROCEDURE

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Background: Results of the Charnley-Williams method of intramedullary fixation for treatment of congenital pseudarthrosis of the tibia have varied, in part because of variations in surgical technique. The outcomes of three variations of this procedure were compared to determine which technique was the most likely to result in union.

Methods: The results in twenty-three consecutive patients with congenital pseudarthrosis of the tibia were reviewed at four to fourteen years following initial surgical treatment with an intramedullary rod. Three types of procedures were performed: type A, which consisted of resection of the tibial pseudarthrosis with shortening, insertion of an intramedullary rod into the tibia, and tibial bone-grafting combined with fibular resection or osteotomy and insertion of an intramedullary rod into the fibula; type B, which was identical to type A except that it did not include fibular fixation; and type C, which consisted of insertion of a tibial rod and bone-grafting but no fibular surgery. The outcome was classified as grade 1 when there was unequivocal union with full weight-bearing function and maintenance of alignment requiring no additional surgical treatment; grade 2 when there was equivocal union with useful function, with the limb protected by a brace, and/or valgus or sagittal bowing for which additional surgery was required or anticipated; and grade 3 when there was persistent nonunion or re-fracture, requiring full-time external support for pain and/or instability.

Results: Eleven patients (48%) ultimately had a grade-1 outcome; nine, a grade-2 outcome; and three, a grade-3 outcome. The final outcome was not associated with either the initial radiographic appearance of the lesion or the age of the patient at the time of the initial surgery. The results following type-A and B operations were better than those after type-C procedures. Surgery on an intact fibula resulted in a lower prevalence of grade-3 outcomes than was found when an intact fibula was not operated on (p = 0.05). Transfixation of the ankle joint by the intramedullary rod did not decrease the prevalence of grade-3 outcomes.

Conclusions: There is little justification for a type-C operation, as it either resulted in a persistent nonunion or failed to improve an equivocal outcome in every case. Leaving an intact fibula undisturbed to maintain stability or length also was not successful in this series. In addition, the presence of fibular insufficiency (fracture or a pre-pseudarthrotic lesion) was highly prognostic for subsequent valgus deformity (occurring in ten of twelve cases), whether or not the fibula eventually healed.

A common initial surgical approach to the treatment of congenital pseudarthrosis of the tibia is to combine the procedures described by Charnley and Williams and popularized by Coleman— that is, resection and shortening of the tibial diaphysis through the pseudarthrosis site, internal fixation with an intramedullary rod, and autogenous bone-grafting. Achievement of union with this procedure has varied considerably, with prevalences ranging from 28% to >80%, presumably because of substantial variations in surgical technique among the different reports. Specific technical details of the procedure—such as whether it is preferable to resect the pseudarthrosis and shorten the tibia or to simply add bone graft at the defect; whether it is necessary to perform fibular resection, shortening, and intramedullary fixation when there is a fibular pseudarthrosis; whether a fibular osteotomy should be done if the fibula is intact; whether the outcome is affected by an ununited fibula; and whether the intramedullary rod should be placed across the ankle joint—are not well documented. This review was undertaken to evaluate the effect of such variations in the Charnley-Williams technique on the rate of union in patients with congenital pseudarthrosis of the tibia.

Materials and Methods

The results of surgical treatment of twenty-three consecutive patients with congenital pseudarthrosis of the tibia, operated on between 1978 and 1992 by seven different pediatric orthopaedic surgeons, were reviewed retrospectively (Table I). Criteria for inclusion in the study were initial surgical treatment by insertion of an intramedullary rod into the tibia and suffi-
### TABLE 1 Outcomes According to Type of Procedure

| Case | Crawford Type* | Age at Op. (yr + mo) | Type of Procedure | Bone Graft as Separate Op. | Transfixation of Ankle | Duration of Orthotic Use (yr) | Grade of Outcome | Duration of Follow-up (yr) |
|------|----------------|----------------------|-------------------|---------------------------|------------------------|-------------------------------|-----------------|---------------------------|
| 1    | I (IV)         | 5 + 8                | A                 | +                         | +                      | 9                            | 1               | 12                        |
| 2A   | I (IV)         | 3 + 7                | C                 | –                         | –                      | 5                            | 2               | 5                         |
| 2B   |                | 8 + 11               | B                 | +                         | –                      | 3                            | 3               | 3                         |
| 3    | I (IV)         | 5 + 6                | B                 | –                         | –                      | 6                            | 1               | 6                         |
| 4    | IV             | 2 + 1                | A                 | (×3)                      | +                      | 5.5                          | 1               | 14                        |
| 5A   | II             | 3 + 4                | C                 | –                         | –                      | 1                            | 3               | 1                         |
| 5B   |                | 4 + 4                | A                 | +                         | +                      | 7                            | 1               | 8                         |
| 6    | II (IV)        | 9 + 5                | A                 | –                         | –                      | 8                            | 1               | 8                         |
| 7A   | II             | 6 + 11               | Hemiepiphyseodesis| A                         | +                      | 4.5                          | 2               | 5                         |
| 7B   |                | 12 + 0               | Hemiepiphyseodesis| –                         | –                      | –                            | 1               | 1                         |
| 8A   | IV             | 1 + 8                | A                 | +                         | +                      | 7                            | 2               | 6.5                       |
| 8B   |                | 8 + 2                | Hemiepiphyseodesis| –                         | –                      | 2                            | 1.5             | 8                         |
| 9A   | II             | 5 + 1                | A                 | –                         | +                      | 1                            | 3               | 1                         |
| 9B   |                | 6 + 3                | A                 | –                         | +                      | 6                            | 1               | 8                         |
| 10A  | II             | 3 + 2                | A                 | –                         | +                      | 1                            | 2               | 1                         |
| 10B  |                | 4 + 1                | A                 | –                         | +                      | 1                            | 2               | 5                         |
| 10C  |                | 9 + 0                | Hemiepiphyseodesis| –                         | –                      | 0                            | 2               | 1                         |
| 11A  | IV             | 2 + 6                | A                 | –                         | –                      | 0.5                          | 3               | 0.5                       |
| 11B  |                | 3 + 0                | A                 | –                         | –                      | 1                            | 2               | 2                         |
| 11C  |                | 5 + 0                | A                 | –                         | –                      | 0.5                          | 1               | 2.5                       |
| 12A  | III            | 1 + 0                | C                 | +                         | +                      | 4                            | 3               | 4                         |
| 12B  |                | 5 + 3                | A                 | –                         | +                      | 3                            | 1               | 3                         |
| 13   | III            | 1 + 9                | B                 | –                         | +                      | 1                            | 2               | 10                        |
| 14A  | III            | 1 + 7                | B                 | –                         | –                      | 2                            | 3               | 2                         |
| 14B  |                | 3 + 6                | B                 | –                         | +                      | 2                            | 2               | 2                         |
| 15   | I (IV)         | 5 + 9                | B                 | –                         | –                      | 0.5                          | 1               | 7                         |
| 16A  | IV             | 1 + 6                | A                 | –                         | +                      | 0.5                          | 2               | 2                         |
| 16B  |                | 3 + 4                | A                 | –                         | +                      | 0.5                          | 2               | 6                         |
| 17A  | I (IV)         | 7 + 4                | C                 | –                         | –                      | 0.3                          | 3               | 0                         |
| 17B  |                | 7 + 8                | C                 | +                         | –                      | 0.5                          | 3               | 6                         |
| 18   | IV             | 1 + 2                | C                 | +                         | +                      | 5                            | 3               | 5                         |
| 19A  | II             | 2 + 1                | A                 | –                         | +                      | 6                            | 2               | 6                         |
| 19B  |                | 8 + 3                | C                 | +                         | –                      | 5                            | 2               | 5                         |
Caretful follow-up to ascertain an outcome, as described below. The duration of follow-up ranged from four to fourteen years (mean, nine years) after the index procedure. Fourteen patients were twelve years of age or older at the time of the review. All patients had been treated with only splinting or bracing of the limb with the pseudarthrosis prior to the index surgical procedure. All had unilateral involvement, and twelve had neurofibromatosis. The age at the time of the initial diagnosis ranged from birth to nine years (mean, two years and four months), and the age at the index procedure ranged from eleven months to nine years and five months (mean, three years and eight months). According to Crawford's classification\textsuperscript{11,12} (Fig. 1), five tibiae had type-I morphology; seven, type-II; three, type-III; and eight, type-IV. Seven patients presented with an incomplete or stress fracture superimposed on a type-I or II lesion; they were not considered to have a true type-IV lesion because of the lack of displacement or erosive or atrophic morphology, which suggests a different prognosis.

Three types of procedures were performed. In a type-A operation, the tibial pseudarthrosis was resected extraperiosteally proximally and distally to what the surgeon considered to be more normal bleeding bone, the medullary canal was opened with a curet or drill, and the tibia was shortened and internally fixed with an intramedullary rod, which frequently transfixed the ankle and subtalar joints. Simultaneously, fibular resection, shortening, and intramedullary fixation was performed extraperiosteally when there was a fibular pseudarthrosis. When the fibula was intact, it was osteot-

| Case | Crawford Fracture Type* | Age at Op. (yr + mo) | Type of Procedure | Bone Graft as Separate Op. | Transfixation of Ankle | Duration of Orthotic Use (yr) | Grade of Outcome | Duration of Follow-up (yr) |
|------|--------------------------|----------------------|-------------------|--------------------------|------------------------|----------------------------|-----------------|--------------------------|
| 20A  | IV                       | 1 + 11               | A                 | –                        | +                      | 11                         | 2               | 11                       |
| 20B  | IV                       | 13 + 0               | Hemiepiphyseodesis, distal tibia-fibula synostosis | –                        | _                      | 1                           | 1               | 3                        | 14 |
| 21A  | II (IV)                  | 8 + 6                | B                 | –                        | –                      | 1                           | 2               | 5                        | 11 |
| 21B  | II                       | 14 + 2               | C                 | _                        | _                      | 0.5                         | 2               | 6                        | 11 |
| 22A  | IV                       | 1 + 10               | A                 | +                        | +                      | 4.5                         | 2               | 4.5                      |
| 22B  | IV                       | 5 + 4                | C                 | _                        | +                      | 6.5                         | 2               | 6.5                      | 11 |
| 23A  | IV                       | 0 + 11               | B                 | –                        | +                      | 6                           | 2               | 8                        |
| 23B  | IV                       | 9 + 4                | Hemiepiphyseodesis | –                        | _                      | _                           | 2               | 2                        | 10 |

*(IV) indicates that there was a superimposed incomplete or stress fracture.
omized subperiosteally, and any overlapped segment was resected. In either case, the fibula was shortened enough to allow impaction at the tibial resection site and then was fixed with an intramedullary rod. Autogenous iliac crest bone graft was applied to one or both resection or osteotomy sites.

A type-B procedure was identical to a type-A operation except that the fibula was not internally fixed. In some patients who had an intact fibula, it was not bone-grafted but simply osteotomized subperiosteally, and the ends were then allowed to overlap.

A type-C procedure involved surgery on the tibia only, including resection, insertion of a rod, and bone-grafting of the tibial defect without shortening. The fibula was either intact and not disturbed in order to preserve its length and stability or it had a pseudarthrosis that was not addressed.

Postoperative management varied according to the surgeon’s preference. A hip spica or non-weight-bearing long leg cast was worn for up to three months postoperatively, after which a weight-bearing long or short leg cast was applied, depending on the degree of consolidation, and was worn for an additional two to three months. A protective orthosis was then prescribed for a period ranging from three months to indefinitely. Prolonged orthotic protection was required when ankle transfixation had been performed (Figs. 2-A through 2-E), and it was discontinued only when the rod had migrated completely into the distal tibial epiphysis.

Repeat autogenous bone-grafting, including posterolateral placement in an attempt to gain crossunion between the tibia and fibula, was performed, at the discretion of the surgeon, as a separate additional procedure in thirteen instances, three to twelve months following the intramedullary rod procedure.

The outcome was graded according to both clinical and radiographic criteria at the time of the most recent follow-up. Unequivocal union was defined clinically by a full weight-bearing status, with or without orthotic support; absence of pain or an antalgic limp; and either a history or documentation of some physical activity approaching normal play, sports, or mobility. Radiographically, union was considered unequivocal when there were no transverse or longitudinal cortical defects or atrophy and there was remodeling and thickening of the tibial and fibular cortices circumferentially (Fig. 2-E). Any union that did not fulfill these criteria was considered to be equivocal, although this designation did not necessarily mean that addi-
tional treatment was needed or that the outcome was poorer.

A grade-1 outcome was an unequivocal union with maintenance of alignment that required no additional surgical treatment. Mild malalignment (≤10° in the coronal or sagittal plane) or limb-length discrepancy (≤3 cm) expected to require either no treatment or only contralateral epiphyseodesis did not preclude a grade-1 outcome. The outcome was considered grade 2 when there was equivocal union (residual longitudinal or transverse cortical deficiency) and/or deformity (usually >15° of valgus, procurvatum, or recurvatum) for which additional surgery was required or anticipated (Figs. 3-A through 3-E). Patients with a grade-2 outcome wore a brace, primarily to prevent refracture, but had clinically useful function. Grade-3 outcomes consisted of a persistent nonunion or refracture (Figs. 4-A through 4-E) that was painful or unstable and required full-time orthotic support. When a grade-2 or 3 outcome necessitated additional operative treatment (Table I), the grading was performed again once the outcome after the additional surgery could be determined. There was no formal evaluation of functional outcome—that is, there was no objective determination of muscle strength, ankle kinematics or kinetics, or exercise tolerance. Transfixation of the ankle was evaluated as a separate variable affecting the eventual outcome.

Statistical comparison of outcomes was performed with use of the Fisher exact test.

Results

At the most recent follow-up evaluation, eleven patients (48%) had a grade-1 outcome. Five patients had this outcome after a single operation, five required two operations, and one patient required three. Nine patients (39%) had a grade-2 outcome; one of these outcomes was achieved after just one operation, seven were achieved after two, and one was achieved after three. Three patients (13%) had a grade-3 outcome; two had had two operations and one, only one. There was no association between the initial radiographic appearance (the Crawford classification) and the most recent outcome (Table II). Of note was the fact that no patient who had had a primary type-C procedure had union without a revision, and no patient who had a type-C procedure in order to revise a previous operation with a grade-2 or 3 outcome had improvement in the quality of the union.

Fifteen patients underwent the initial operation when they were less than four years of age. Five of them had a grade-1 final outcome; eight, a grade-2; and two, a grade-3. Eight patients had the initial surgery performed after the age of four. Six of them had a grade-1 final outcome; one, a grade-2; and one, a grade-3.

| Crawford Fracture Type | Outcome (no. of patients) |
|------------------------|---------------------------|
|                        | Grade 1 | Grade 2 | Grade 3 |
| I                      | 3       | 0       | 2       |
| II                     | 4       | 3       | 0       |
| III                    | 1       | 2       | 0       |
| IV                     | 3       | 4       | 1       |

*The association between the outcome and the Crawford classification was not significant (p = 0.78).
one, a grade-3. The age at the time of the initial surgery did not affect the chance of avoiding a grade-3 outcome (p = 0.73).

The procedures that addressed the fibula (the type-A and B procedures) yielded better results. A total of thirty-six rod procedures were performed; twenty-three were primary operations, and thirteen were revisions. The twenty-six type-A or B operations produced nine grade-1, thirteen grade-2, and four grade-3 outcomes, whereas the ten type-C procedures produced no grade-1, five grade-2, and five grade-3 outcomes. Type-A or B procedures resulted in significantly fewer grade-3 outcomes (p = 0.046). When just the twenty-three primary procedures were evaluated, it was found that the eighteen type-A or B procedures produced five grade-1, ten grade-2, and three grade-3 outcomes, whereas the five type-C

Fig. 3-A through 3-E Case 10, a boy with neurofibromatosis who presented with a Crawford type-II lesion when he was two years old. Fig. 3-A At the age of three years and two months, the patient sustained the first fracture. He underwent a type-A procedure with ankle transfixation. Fig. 3-A One year later, a type-A revision was performed to replace the fibular rod, which had migrated distally, and to push the tibial rod higher into the proximal segment to hasten release of the ankle. Additional graft was placed to complete the synostosis.

Fig. 3-C Two years following revision, there was solid union and a mobile ankle. The patient refused further orthotic protection, in spite of the lesion at the proximal tip of the tibial rod. Fig. 3-D By the age of nine years, five years following revision, substantial ankle valgus had developed in spite of a healed and crossunited fibula. Medial hemiepiphyseodesis of the distal part of the tibia was performed. Fig. 3-E One year later, the ankle valgus has been corrected, but the diaphyseal valgus and recurvatum as well as the proximal lesion remained (a grade-2 outcome). The patient was fully active and asymptomatic except for a decreased range of motion of the ankle.
Case 2, a boy who presented with the insidious onset of varus deformity following trivial trauma when he was three and one-half years old. **Fig. 4-A** The lesion was classified as Crawford type I with an incomplete stress fracture. The fibula appeared to be uninvolved. **Fig. 4-B** A type-C procedure without ankle transfixation was performed following failure of healing in a cast. There was an incomplete fracture line four and one-half years postoperatively (a grade-2 outcome) despite continuous orthotic protection. The distal tip of the Williams rod had gradually protruded through the cortex, and there was a progressive anterior bow and valgus deformity. A type-B revision was performed. **Fig. 4-C** Four months following revision, the fibular osteotomy site had not united and the tibial pseudarthrosis appeared to be more atrophic. The fibula was more sclerotic and narrowed distal to the level of the tibial pseudarthrosis. Additional bone-grafting of the tibia only was performed at this point.

**Fig. 4-D** Fifteen months following bone-grafting, the tibia appeared to be united but the fibular osteotomy site was ununited. Also, a second, more distal fibular fracture had occurred, with increasing valgus. No treatment other than continued brace protection was provided. **Fig. 4-E** At the age of eleven years and six months, three years after grafting, the tibia had also broken and gone on to frank pseudarthrosis. A series of Ilizarov procedures to correct angulation and obtain union by bone transport failed. A below-the-knee amputation was eventually performed at the age of sixteen years.
operations produced no grade-1, one grade-2, and four grade-3 outcomes. Primary type-A or B procedures resulted in a much lower prevalence of grade-3 outcomes than did primary type-C procedures (p = 0.017).

Eleven patients had only one rodding procedure, although five had a separate bone-grafting operation at a later date and four had a distal tibial medial hemiepiphyseodesis and/or creation of a tibia-fibula synostosis to treat valgus deformity. Ten of the eleven patients had functional union (seven grade-1 and three grade-2 outcomes) at six to fourteen years following a type-A or B procedure. The eleventh patient, who had had a type-C operation as well as a later bone-grafting procedure, did not have union (a grade-3 outcome) at five years postoperatively.

The status of the fibula was crucial to the outcome. In ten of the twelve instances in which the fibula was not intact (had a pre-pseudarthrosis or had actually fractured) at the time of surgery, there was clinically relevant valgus deformity postoperatively, even when the fibular lesion had been treated and had subsequently united; all ten outcomes were grade 2. Cross-union between the tibia and fibula, achieved in four instances, did not prevent this valgus malalignment (Figs. 3-A through 3-E). Of the nine procedures in which an intact fibula was not osteotomized (a type-C procedure), six resulted in a nonunion (a grade-3 outcome) and three produced a grade-2 outcome. Of the eight procedures in which an intact fibula was osteotomized but not treated with a rod (a type-B procedure), three produced a grade-3 outcome and five resulted in healing without valgus, with three of the five producing a grade-1 outcome. Of the eighteen procedures in which the fibula was treated with a rod following osteotomy or resection (a type-A procedure), seventeen eventually resulted in a grade-1 or 2 outcome (Figs. 5-A through 5-E). Surgery on an intact fibula (osteotomy with or without insertion of an intramedullary rod) was therefore crucial to avoiding a grade-3 outcome (p = 0.05). Of the fourteen intact fibulae that were osteotomized, eleven subsequently healed. There was no difference in healing between the fibulae

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Figs. 5-A through 5-E Case 11, a boy with neurofibromatosis who was two and one-half years old at the time of presentation. Fig. 5-A Initial radiographs showing a Crawford type-IV lesion with an intact fibula. The patient underwent a type-A procedure with a custom interlocking rod and a single screw in the distal segment at the age of two years and six months. This was revised within three months because the interlocking screw backed out. Fig. 5-B Radiographs made three months following revision with the distal tibial segment fixed with an antegrade technique and two interlocking screws, one of which was placed in the epiphysis itself. Hypertrophic callus and crossunion can be seen to be developing. The epiphyseal screw was removed to untether the physis at this time. Postoperative protection with an articulated ankle-foot orthosis was discontinued, three months later.

Fig. 5-C Two years later, lateral bowing had recurred, with loss of fixation in the distal segment. The patient had mild pain and required restoration of alignment.
treated with intramedullary fixation (six of seven) and those treated without it (five of seven) \( (p = 0.51) \).

Of the twenty procedures that included placement of an intramedullary rod across the ankle joint \(^{14} \), five produced a grade-1 outcome; twelve, a grade-2; and three, a grade-3. Of the sixteen procedures in which the ankle joint was not transfixed by a rod, four led to a grade-1 outcome; six, a grade-2; and six, a grade-3. With the numbers available, the prevalence of grade-3 outcomes was not significantly affected by the use or avoidance of ankle transfixation \( (p = 0.123) \).

Nine of the thirteen separate bone-grafting procedures appeared to enhance union (led to a grade-1 or 2 outcome), whereas four were followed by persistent nonunion. As a result of the various types of rod procedures done prior to these procedures, the variations in postoperative management, more detailed analysis of the effect of bone-grafting alone could not be done.

An attempt was made to analyze the effects of postoperative orthotic protection following the thirty-six rod and thirteen bone-grafting procedures. The duration of brace use ranged from four months to twelve years following these procedures. Postoperative orthotic management was considered to have had no discernible effect on the outcome when union was not achieved (Fig. 4-B) or when the orthosis had been worn for less than six months, as this made any long-term effect on union doubtful. The orthosis appeared to have had no discernible effect in twenty-three instances. In fifteen instances, the effect of the orthosis was considered uncertain, such as when union was achieved following surgery but there was subsequent refracture while the orthosis was being used. Orthotic use was considered to have been beneficial in eleven instances, in which refracture was prevented for the entire follow-up period, especially in proximal or distal areas of the tibia that were no longer spanned by the intramedullary rod because of growth or when the rod had never migrated out of the ankle.

**Discussion**

Historically, congenital pseudarthrosis of the tibia has been one of the most difficult and frustrating conditions to treat. Repeated ineffective surgical attempts to achieve union have been commonplace, and refracture following what appeared to be union has occurred with enough frequency for any union to be considered only transient. Additionally, emphasis placed solely on the achievement of union obscures the many functional disabilities that result from multiple operations and the often endless bracing \(^{13,15} \). Because it is intuitive that the functional outcome will likely be poor in the absence of union, the goal of the initial treatment must be to gain union or at least to stabilize the pseudarthrosis so that the limb can function. As a result of this review, I concluded that two basic principles should direct treatment of this condition: first, the alignment of the limb should be aggressively maintained to avoid fracture due to bone deformity compounding fragility and, second, permanent intramedullary fixation is desirable in order to internally splint the tibia to achieve union, prevent refracture, and maintain alignment potentially with no external support.

These two principles are incorporated into the commonly used initial procedure described by Charnley \(^1 \), in which the pseudarthrosis is resected, bone graft is applied to the site, and the tibia is internally fixed with an intramedullary rod.

**Fig. 5-D** A type-A revision was performed at the age of five years. A tibial osteotomy performed proximal to the pseudarthrosis with an antegrade technique allowed realignment. Fibular osteotomy and fixation encouraged union of the hypertrophic callus. Bone graft was added only at the proximal tibial osteotomy site. **Fig. 5-E** Two and one-half years after revision, the patient had normal alignment and ankle motion (a grade-1 outcome).
that transfixes the ankle and subtalar joints. Use of the threaded coupled rods developed by Williams, originally for intramedullary fixation in osteogenesis imperfecta, has simplified the technique of rod placement and length determination. The tibial rod can extend as far proximally as desired (for example, to the proximal tibial physeal) and can be accurately left in the calcaneus, talus, or even the distal tibial epiphysis if adequate purchase can be obtained. Coleman is generally credited with popularization of this method, which also includes intramedullary fixation of the fibula.

Results of the Charnley-Williams technique have varied considerably. Recent proponents have reported union in >80% of cases, although additional bone-grafting, application of a new rod following fracture or angulation proximal or distal to the end of the original rod, and other procedures, as well as indefinite orthotic protection, are often necessary. Follow-up of some patients has been relatively short (two years or less), so that the prevalence of late refracture probably has been underestimated. The results have been achieved with a variety of techniques, including dual rod fixation or use of a tibial rod only with no fibular surgery. Other variations include the use of extendable rods not crossing the ankle and the use of extendable rods with or without both fibular surgery and ankle transfixation. On the other hand, in a recent multicenter review by the European Paediatric Orthopaedic Society of the results of 240 operations, only 28% to 50% achieved union.

Again, there was substantial variation in surgical technique, including, for example, use of a tibial rod without bone-grafting. Because of an impression that the outcome of Charnley’s method is strongly determined by the technical details of the procedure, I undertook a review to delineate which specific aspects of the procedure might be important.

Because of the retrospective nature of the present study, which extended back to 1978, in some cases the outcome could be determined only from notations in the medical record regarding weight-bearing status, use or discontinuation of use of protective orthoses, and radiographic evaluation. Grade-1 and grade-3 outcomes, representing the extremes of union with an aligned stable limb on the one hand and a frank nonunion or refracture with pain and/or instability requiring full-time external support on the other, were unambiguously delineated. A grade-2 outcome was assigned when useful weight-bearing function (usually with support) had been achieved even though the union was precarious as a result of radiographic defects and even though there was usually angulation exceeding 15° and/or shortening predicted to exceed 3 cm.

There appears to be little justification for performing a type-C procedure, as it resulted in either a persistent nonunion (a grade-3 outcome) or failed to improve an equivocal (grade-2) union in every instance. No patient had an improved outcome after a procedure in which the fibula was not addressed. Leaving an intact fibula undisturbed might be contemplated to provide stability following resection, bone-grafting, and rod fixation of a tibial defect. However, it can be argued more persuasively that the pseudarthrosis cannot consolidate when the defect is held distracted by an intact fibula. Experience with fibular osteotomy alone to allow impaction and consolidation of posttraumatic hypertrophic tibial nonunions supports this concept. In the present series, type-A and B procedures, in which the fibula was either resected and shortened (when a pseudarthrosis was present) or osteotomized (when it was intact), resulted in a lower prevalence of grade-3 outcomes than did type-C procedures, in which fibular surgery was not performed. Type-A and B procedures were particularly more effective than type-C procedures in preventing grade-3 outcomes when they were performed as the primary operation. Since eleven of the fourteen intact fibulae that were osteotomized healed in this series, concern over additional deformity or morbidity secondary to nonunion of the fibula may be overstated in light of the poorer outcomes when the fibula is not operated on.

The need for fibular surgery is not universally accepted. Although Baker et al. reported that union was achieved in ten of thirteen patients treated with intramedullary fixation of the tibia without fibular surgery, they could not determine if the presence of a fibular pseudarthrosis affected union. They also reported that nine of the patients with union had a valgus deformity of ≤30°, but they did not discuss such issues as refracture, gait disturbance, or poor ankle function. Similarly, Fern et al. and Joseph and Mathew reported union rates of 100% and 83%, respectively, without fibular surgery, but they did not report the prevalence of valgus deformity. Fern et al. reported a 40% refracture rate after 4.5 years of follow-up, and Joseph and Mathew reported a 25% refracture rate after three years of follow-up. The relationship between the omission of fibular surgery and valgus deformity, refracture, and the need for additional treatment cannot be directly determined from these reports, but experience in the current series suggests that progressive valgus with fibular insufficiency should be treated (Figs. 4-A through 4-E). Maintenance of alignment to minimize the possibility of refracture cannot be overemphasized.

A bowed or hypoplastic fibula, suggesting impending pseudarthrosis, or frank fracture was prognostic of subsequent valgus malalignment, which was not avoided in ten of twelve instances in which the fibular lesion was treated surgically (Figs. 3-A through 3-E). The valgus deformity occurred in spite of treatment designed to restore the integrity of the fibula, suggesting that such malalignment is a measure of the severity of the condition and is not a reflection of the treatment. Early appreciation of such valgus and corrective treatment to maintain limb alignment may avoid refracture and a subsequent resistant nonunion (Figs. 4-A through 4-E). For this reason, osteotomies at sites other than the original pseudarthrosis may be required to regain alignment and reestablish intramedullary fixation. Other methods to correct valgus, such as hemiepiphyseodesis or distal tibia-fibula synostosis (to prevent proximal migration of the lateral malleolus), can be utilized when acute realignment is not necessary.

Transfixion of the tibiotaral and subtalar joints was originally justified as a necessary evil to obtain adequate pur-
Chase in the often atrophic and short distal tibial segment. Although the rod can migrate into the tibial diaphysis with growth, objectively measured functional outcomes of ankle transfixation have been dismal and likened to the function of an amputee. Considering that ankle transfixation did not decrease the prevalence of grade-3 outcomes (nonunions), efforts to achieve stabilization without ankle transfixation, especially when the segment distal to the pseudarthrosis is large, are appropriate.

Although postoperative orthotic management was universally employed in this series, ascertaining its role in achieving or maintaining union was difficult. Indeed, in thirty-eight of forty-nine instances, postoperative bracing was considered to have had either an uncertain effect or no effect on the surgical outcome. As ankle transfixation does not seem to decrease the prevalence of nonunion, the use of intramedullary devices, such as custom interlocking or flexible rods (Figs. 5-A through 5-E), that do not cross the ankle joint would eliminate to a great extent the need for ankle bracing. Another traditional role for orthotic management has been protection against refracture. This indication may be reconsidered since only two patients (Cases 2 and 17) had a refracture that was resistant to healing and all other refractures healed after cast immobilization or revision surgery. Thus, the role of traditional, indefinite orthotic protection in maintaining union may be small, since refracture is best treated with a cast and, if that is unsuccessful, with revision surgery. For patients in whom a valgus and/or sagittal plane deformity develops proximal or distal to the tip of the rod, and who are at increased risk for refracture, the functional cost of long-term orthotic protection must be weighed against the risk of revision with osteotomy and insertion of a new, longer rod. Such decisions must be made on an individual basis.

While skeletal maturity has been considered to be a milestone that may impart some degree of protection against refracture, this has never been documented by the follow-up of adult patients. Furthermore, follow-up of patients at skeletal maturity has often merely documented the poor function of the extremity, reflecting results of treatment in infancy and childhood. Union was achieved prior to maturity and then maintained in six patients in this series (Cases 1, 4, 6, 10, 20, and 21), with a grade-1 outcome in four and a grade-2 outcome in two. All patients stopped using a brace and, although objective assessment of functional outcome was beyond the scope of this study, subjectively all had a very useful extremity, presumably as a result of early achievement and then maintenance of union without long-term external immobilization. As refracture is a potential complication of this disease at any time, and as long-term prophylactic bracing is functionally undesirable and has not been proven to prevent this complication, emphasis should be directed toward surgical procedures that have the best chance of achieving union early and maintaining it by permanent intramedullary fixation.

Operations that do not address the fibula appear to be ineffective in achieving union and limb alignment. The Charnley-Williams procedure should therefore include (1) tibial intramedullary fixation, preferably avoiding ankle transfixation, following extraperiosteal resection of the pseudarthrosis site and diaphyseal shortening; (2) extraperiosteal resection or shortening osteotomy of the fibula, depending on the preoperative status of the fibula, with or without intramedullary fixation (a type-A or B procedure); and (3) autogenous iliac crest bone-grafting of the tibia and the fibula if a pseudarthrosis is present. If the fibula is not intact preoperatively, valgus malalignment is likely to occur and treatment should be planned to control or correct it, to avoid refracture and the need for debilitating long-term orthotic protection.

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