CONGENITAL PSEUDARTHROSIS OF THE TIBIA

RAYMOND T. MORRISSY, EDWARD J. RISEBROUGH, JOHN E. HALL

From the Children’s Hospital Medical Center, Boston, and the Hospital for Sick Children, Toronto

Forty cases of congenital pseudarthrosis of the tibia were reviewed. The results were assessed so as to emphasise function rather than simply the presence or absence of union. At the time of review, 14 patients had undergone amputation; eight others had non-union or tenuous union. No surgical procedure except the Farmer operation (a composite skin and bone pedicle graft from the other leg) showed any clear superiority. Among the factors associated with a poor result were considerable shortening, older children, and rapid resorption of the bone graft. It is felt that congenital pseudarthrosis of the tibia is a biological problem and not merely a mechanical one; consequently biological approaches to its treatment are needed.

Two questions are of immediate importance in treating a patient with congenital pseudarthrosis of the tibia: what is the best surgical procedure to use?; and, when is amputation indicated? Unfortunately, few articles in the English literature contain more than 20 cases (Van Nes 1966; Sofield 1971; Masserman, Peterson and Bianco 1974; Andersen 1976b), and of these, only two make it clear how many patients have reached, or nearly reached, maturity (Sofield 1971; Andersen 1976b). Several reports claim that one particular surgical procedure is better (Wilson 1941; McFarland 1951; Farmer 1952; Charnley 1956) and some suggest that there is no indication for amputation (Van Nes 1966; Sofield 1971). No previous series has compared a wide variety of procedures in a large number of patients followed for a long period of time.

The purpose of this present study is to assess the eventual outcome in 40 patients who were treated over 25 years by several different surgeons. We have tried to identify which factors might help the surgeon in treating any individual patient.

MATERIAL AND METHODS

The records and radiographs of all patients with congenital pseudarthrosis of the tibia treated at the Hospital for Sick Children, Toronto, and the Children’s Hospital Medical Center, Boston, since 1940 were reviewed. Patients were considered for inclusion if they had reached an apparent end-point—for example, amputation, skeletal maturity, or near skeletal maturity with no hope of a better result even if union was achieved. Forty-nine patients met these criteria. Nine of these had to be excluded: five who were near skeletal maturity, but whose final result was nevertheless still in doubt; one whose lesion included absence of the distal half of the tibia; two whose tibiae, though bowed anteriorly, never fractured; and one who could not be traced for follow-up. Those with fracture of the fibula alone were, of course, excluded from the outset.

The critical question we sought to answer was this: which of the patients in whom the limb had been retained would have been better off, both functionally and cosmetically, with an amputation? Each patient who had not been amputated was examined by at least one of the authors other than his own surgeon; if, however, the result was not really in question, then an evaluation (including a photograph, radiographs, and clinical measurements) from another doctor was accepted instead. Apart from assessing the tibia for union, the following also were examined and recorded: the range of movement at the knee and ankle; deformity of the foot, ankle, and leg-length discrepancy; the gait; the use of braces or supports; the patient’s job and physical activities; and the presence or absence of pain. In addition, an attempt was made to find out what the patient and his parents really felt about the success or otherwise of his treatment.

A result was rated as good if the patient had solid union of the tibia, used no support or splint, was fully active, had no pain, and was not troubled by his leg; in short, his pseudarthrosis had been part of his childhood but did not affect his adult activities. A result was considered fair if union had been achieved, supports or splints were minimal, deformity, though noticeable, was neither severe nor incapacitating, and the patient’s daily activities were not affected; though not ideal, such a result is better than an amputation. A poor result was one with non-union or tenuous union requiring a support, with a severe functional and cosmetic handicap, and in which it was judged that amputation would have been preferable. Amputations, since they represented a definitive act to end the disease before healing, were considered separately; though clearly failures of treatment, the final functional and cosmetic results were usually superior to those in the “poor” group.

In an effort to identify those factors in the disorder or its treatment which would allow prediction of the result in any given patient, a number of factors were recorded; these included the sex of the patient, which side was involved, the presence or absence of neurofibromatosis, the age at which the first fracture occurred, the type of pseudarthrosis and the treatment. A modification of the method of Andersen (1973, 1976a) was used to classify the pseudarthroses: the terms “cystic” and “dysplastic” were used as he used them, but “late” was used only if the first fracture occurred after the age of 48 months. Factors noted during treatment were the number of bone grafts used in attempting to obtain union, the time required to unite, the age at the last grafting operation, the skeletal response to grafting, the amount of

R. T. Morri ssy, MD, University of Arkansas for Medical Sciences Department of Orthopaedic Surgery (and Pediatrics), Arkansas Children’s Hospital, 804 Wolfe Street, Little Rock, Arkansas 72201, USA.
E. J. Riseborough, MD Harvard Medical School, Department of Orthopaedic Surgery.
J. E. Hall, MD The Children’s Hospital Medical Center, 300 Longwood Avenue, Boston, Massachusetts 02115, USA.

Requests for reprints should be sent to Dr R. T. Morri ssy.

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VOL. 63, No. 3, 1981

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<th>Age at follow-up (years)</th>
<th>Type of pseudarthrosis</th>
<th>Age at first graft (months)</th>
<th>Age at last graft (months)</th>
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<th>Result</th>
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* Includes prophylactic grafts
† Second number indicates refracture after spontaneous union
#, First number is at time of initial review in 1972; second number is at follow up in 1977
shortening, the type of bone graft and the method of fixation. It was very difficult to determine from the charts and radiographs exactly when the pseudarthrosis united; the age at the last grafting operation was therefore used as the index of when union occurred, since this operation represented the final attempt to achieve or improve union.

RESULTS
Twenty-one patients were male, 19 female. Fourteen pseudarthroses involved the right tibia, 26 the left. Neurofibromatosis was present in 20 patients. Nine patients had a good result; nine, a fair result; eight, a poor result; 14 had amputations. The average age at follow-up, excluding those with amputation, was 16 years, ranging from 12 to 27 years. The results are summarised in Table I.

**Good results.** Nine patients, six boys and three girls, achieved good results. Four pseudarthroses were of the right tibia, five of the left. Four of the nine patients had neurofibromatosis. The average age at which fracture occurred was 19.6 months with a range of 3 to 47 months. Four of the pseudarthroses were of the dysplastic type, three were cystic, and in two the first radiographs were not available so that the type could not be determined. The average number of bone grafts for this group (excluding one prophylactic graft) was 2.2: two patients needed one graft each, three needed two grafts, and four patients needed three. The average time to achieve union (the time between first and last bone graft) was 13.4 months, ranging from nil (where the first graft succeeded) to 32 months. The average age at which the last grafting operation was performed was 40.2 months with a range of 27 to 70 months. It follows that no patient in this review achieved a good result if he required more than three bone grafts, took more than 32 months to achieve union, or failed to show union by the age of six. The operation resulting in union was a Farmer procedure in five cases, a bypass in one, homogenous bone without fixation in two, and autogenous bone with transverse pins in one. The average amount of shortening in this group was 1.4 centimetres, ranging from nil to four centimetres. One patient in this group had an epiphysiodesis resulting in two centimetres of shortening. In four patients, the fibula did not bow or fracture during the course of treatment.

**Fair results.** Nine patients, four boys and five girls, achieved fair results. The right tibia was involved in one and the left in eight. Four of the nine patients had neurofibromatosis. The average age at which fracture occurred was 21.3 months, the earliest occurring at one month and the latest at 62 months. Five of the pseudarthroses were dysplastic, two were cystic, one was late, and in one the type could not be determined. The average number of bone grafts for this group was 5.6; the least number was three, and the most 10. The average time to achieve union was 63 months, ranging from 15 to 127 months. The average age at which the last grafting operation was performed was 92.2 months, the latest union occurring at 150 months. The operation preceding union was a Farmer operation in two patients, a dual onlay graft in three, a sliding graft in one, the addition of homogenous bone without fixation in one, and the addition of autogenous bone without fixation in two. The average amount of shortening was 3.4 centimetres, the greatest amount being 6.0 centimetres. Epiphysiodesis was used in two patients, each with 3.0 centimetres of residual shortening at maturity. An additional patient had a femoral shortening operation to correct a deficit of 8.1 centimetres.

**Poor results.** Eight patients, two boys and six girls, had poor results. The right tibia was involved twice, the left six times. Neurofibromatosis was present in five of the eight patients. The average age at which fracture occurred was 44.3 months; but if late pseudarthroses were excluded, the average age was only 24.7 months. Two of the pseudarthroses were of the dysplastic type, two were cystic, two were late, and in two the type could not be determined. The average number of bone grafts in this group (excluding two prophylactic grafts) was 3.8, ranging from three to six. The average time to achieve union was 67 months, ranging from 25 to 132 months. The earliest union was achieved at 72 months and latest at 278 months, with an average of 124.1 months. If the two late cases are excluded, the average is 108 months. The type of graft resulting in union was a Farmer procedure in one, a dual onlay in one, a Sofield in one, homogenous bone without fixation in one, autogenous bone with an intramedullary rod in two, and autogenous bone without fixation in two. The average shortening in this group was 5.5 centimetres, ranging from two to eight centimetres; all but one patient was at least 4.0 centimetres short. Two patients with non-union died from neoplasms of the central nervous system, aged respectively 12 and 19 years. Five patients had tenuous union, as evidenced by their radiographs, and needed to wear a brace or to use crutches. One patient's fracture was solidly united but with 7.0 centimetres of shortening despite an epiphysiodesis; he still needed a special shoe, and had gross deformities of the leg, ankle and foot.

**Amputation.** Fourteen patients, nine boys and five girls, had undergone amputation. There were equal numbers of right and left tibiae involved and exactly half the patients had neurofibromatosis. The average age at which fracture occurred was 16.9 months, ranging from birth to 62 months.

Eight of the pseudarthroses were classified as dysplastic, three as cystic, and one as late; in two the type could not be determined. The average number of bone grafts in this group (excluding four prophylactic grafts) was 4.6, not significantly different from the average in the fair or poor groups; the number ranged from 2 to 11. The average age at amputation was 9.7 years (not significantly different from the time to achieve union in the poor group) ranging from 1 to 28 years. Two of the patients with union requested amputation because of poor function.
Acceptable versus unacceptable
In order to discover which factors determined whether the result was acceptable or unacceptable, the good and fair results were combined and labelled "acceptable"; similarly the poor results were added to the amputations and labelled "unacceptable". Ten boys and eight girls had acceptable results; in 11 boys and 11 girls the result was unacceptable. The left side was involved in 13 of the 18 acceptable cases and in 13 of the 22 unacceptable cases. Neurofibromatosis was present in eight of the acceptable cases (44.4 per cent) compared with 12 of the unacceptable (54.5 per cent). The average age at fracture was virtually the same for the two groups: 20.5 months for the acceptable group and 26.9 months for the unacceptable. However, the two with fractures present at birth and two of the three with fractures occurring during the first month were all in the unacceptable group. On the other hand, three of the four patients with fractures occurring after the age of 48 months were also in the unacceptable group. On the whole, there was little at the beginning of treatment that distinguished one group from the other. The types of pseudarthrosis in the acceptable and unacceptable groups were virtually identical (dysplastic in nine and 10 respectively, cystic in five and five, late in one and three, and unknown in three and four). The average number of grafts was 3.9 for the acceptable group and 4.5 for the unacceptable group.

We did find a statistical difference, however, in the age of the grafting operation: 72 per cent of those with an acceptable result were aged 72 months or less at the time of their last graft (P<0.05). Moreover, no patient who was still being grafted at 13 years of age achieved an acceptable result. There was likewise an indication that the shorter time between the first and the last graft in the acceptable group was significant (P<0.075). Shortening also (though seldom recorded before amputation) was significantly different (P<0.0017) in the two groups; it was less for the acceptable group. Five of the six patients with shortening of more than six centimetres were in the unacceptable group.

ANALYSIS OF RESULTS

Neurofibromatosis
As stated earlier, half the patients in this series had neurofibromatosis, and, when the results were analysed, approximately half the number in each group were so affected. In itself, therefore, neurofibromatosis does not make union less likely or the result less favourable.

Of considerable significance, however, is the fact that, of the 20 patients with neurofibromatosis, no less than five developed gliomata of the central nervous system. Three had gliomata of the brain: two of these are dead, each with tenuous union and a poor result at the time of death; one remains alive with a good result as far as the leg is concerned, but with a poor prognosis for life. Another patient, with an extensive glioma of the cervical cord, is in a nursing home; he has extensive paralysis and had an amputation for non-union. The fifth patient had an optic glioma and also had a partial glossectomy; the result in her leg was rated as poor.

We also looked at the relationship between the presence of neurofibromatosis and the type of pseudarthrosis. According to Andersen (1976a), neurofibromatosis is not associated with the late or the cystic type of pseudarthrosis, but is always present in the dysplastic type. But in our series, of the 19 pseudarthroses classified as dysplastic, 11 had neurofibromatosis and eight did not; of the 10 cystic cases, three had neurofibromatosis and seven did not; none of the "late" cases had neurofibromatosis. We also found (as stated earlier) that the type of pseudarthrosis bore no relationship to the result.

Bone grafting
In all, 169 bone grafting procedures had been performed on the 40 patients; seven operations were prophylactic, leaving 162 grafts performed in the hope of achieving union. In order to establish which type of graft most often led to union, each case was analysed and the type of graft thought most responsible for union was recorded. It was not always the last graft. If, for example, an onlay graft united proximally and the major portion of the graft survived, but the addition of homogenous bone at the distal end was necessary for union, then the onlay graft was credited with producing the union; but if a major portion of the graft had resorbed and additional procedures were necessary, the graft was not so credited. The graft was called successful if the final result was good or fair.

Nine of the 17 Farmer procedures (53 per cent) resulted in union: five were good results and two fair, an acceptability rate of 41 per cent. Four onlay grafts resulted in union (12.5 per cent); there were no good results but three fair results, a success rate of nine per cent. Only one of the bypass procedures resulted in union (seven per cent); the result in this case was good, so that the success rate was seven per cent. Of four Sofield procedures only one resulted in union (25 per cent); although united, the result here was poor. A sliding graft was used in three cases, with union in one (33 per cent); the result here was fair.

Homogenous bone grafting was used on 22 occasions and by itself produced union in four (17 per cent); two of these were good results and one fair, a success rate of 13 per cent. Autogenous bone was used on 68 occasions and produced union in seven (10 per cent); one was a good result and two fair, a success rate of four per cent. No particular method of fixation of the fracture or of the graft could be shown to be related in any significant way to success despite suggestions in the literature that one particular method might yield better results than others. Electrical stimulation was used once but failed.

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Response to grafting. A striking feature in many patients was that large amounts of the grafted bone resorbed within a few months; in other patients no resorption occurred. Since varying amounts of bone usually remain after grafting, only the response to the initial bone graft can be studied and then only if adequate radiographs (just before the first graft, immediately after, and three months later) are available—which was not always the case. However, there were 12 patients with adequate radiographs in whom almost complete resorption of the graft had occurred within three months; 11 of these had poor results or came to amputation. In the twelfth patient the leg was salvaged by a Farmer procedure and the result was fair. In no patient with rapid resorption of the graft was there a good result. In patients with fair results, when resorption did occur, it was neither so extensive, nor so rapid as in those with poor results. In addition to resorption of the graft, some patients also showed increased destruction and resorption of the tibia itself following grafting (Figs 1, 2 and 3); this guaranteed a poor result.

Prophylactic grafting. Bone grafting before fracture occurred was performed in seven cases. The type of pseudarthrosis was dysplastic in four, cystic in one, and unknown in two. The results were good in one and poor in two; four came to amputation. The average age at which prophylactic grafting was done was 13.4 months, ranging from 4 to 55 months. The average age at which fracture occurred was 27.4 months. There were two additional patients who had prophylactic grafting who did not sustain fractures: one had a cystic lesion which, nine years after curettage and grafting, is still satisfactory; the other, with a dysplastic tibia, had a bypass graft, but was lost to follow-up after five years.

Neither prophylactic bracing nor casting was used routinely; those in whom either method was used did not seem to do any better. We were unable to identify any patient who avoided operation because of cast or brace treatment alone.

Shortening
Shortening is difficult to evaluate because it cannot be measured at the end of treatment; the difficulty is that the other leg may have been operated upon in an attempt to achieve equality. Despite such attempts there was still a significantly greater amount of shortening in the patients with tenuous union or with non-union. On cursory examination, the radiographs of many of these patients seemed to show the pseudarthrosis migrating distally; closer inspection, however, showed that this was secondary to failure of growth in the distal physis while growth of the proximal physis continued (Figs 4 and 5).

Spontaneous healing
In two patients a pseudarthrosis healed spontaneously. The first (Figs 6, 7 and 8) was a girl without
neurofibromatosis who walked at 13 months, when mild anterior bowing of the tibia was noted. At 18 months she had a minor fall and the tibia fractured. After three months of immobilisation in a cast she resumed walking without support. Although she walked with a mild limp, no deformity was noticed until she was five and a half, when a lump appeared over the fibula. Radiographs were reported to show fracture of the fibula through a cyst. Over the next year, progressive anterior bowing of the tibia occurred, ending in a fracture. After three bone grafts in three years, she underwent amputation.

The second patient at six and a half years of age had a minor injury to her leg which resulted in a fracture of her tibia. After four months of immobilisation in plaster she resumed full unsupported weight-bearing for an additional 18 months; then the tibia again fractured, with minimal trauma. This patient was subsequently classified as having a poor result.

Osteotomy
Osteotomy of the affected tibia was performed in four patients. In one this, the first operation, was performed at the age of 11 months; the result was a pseudarthrosis but, after two bone grafting operations, healing eventually occurred. In the other three patients, the osteotomy was performed to correct deformity after union had occurred. Two were done within a year of achieving union, both through the site of union of the distal end of the graft; both, after further bone grafting, eventually healed. In the remaining patient the osteotomy was through the distal tibia, below the affected area, after nine years of solid union; this osteotomy healed promptly with a good result.

DISCUSSION
The first problem facing the surgeon who treats congenital pseudarthrosis of the tibia is to decide which procedure he should use, for it is generally supposed that the type of bone grafting and the method of fixation play a large part in determining the success or failure of treatment. In our series 162 bone grafting operations were performed but no method emerged as superior to any other, with the exception of the Farmer procedure.

After no other technique was so much viable bone incorporated by the tibia; this remained true whether the end-result was success or failure (Figs 9 to 13). The chief difficulty was in achieving union between the graft and the short distal segment of the tibia. The Farmer procedure possesses significant biological differences from other procedures in that it brings its own blood supply. This fact not only invites speculation regarding the aetiology of congenital pseudarthrosis, it may also have clinical relevance. Using modern techniques it may be possible to achieve the same effect without such an extensive operation on the opposite limb; thus the opposite fibula, or a rib on a microvascular pedicle, might be anastomosed to a pedicle on the affected limb (Taylor et al. 1978).

The second problem in treating congenital pseudarthrosis is to decide when to abandon further attempts at achieving union and to amputate the leg. Although some authors feel there may be no indications for amputation (Van Nes 1966; Sofield 1971), most accept that sometimes it is necessary and is indeed in the patient’s best interests (Boyd and Sage 1958).

When our results were subjected to rigorous analysis, only two factors emerged as statistically significant in separating acceptable from poor results. The first was age: the older the patient at the time of his last graft, the less likely he was to achieve union, and no patient who was still being grafted at 13 achieved an acceptable result. This contrasts with the view of some surgeons (Henderson 1928; Lloyd-Roberts and Shaw 1969) that the older the child at the time of grafting, the better the chance of success, and the later the fracture occurs the better the result. This view has been challenged (Sofield 1971; Masserman et al. 1974) and our data support that challenge. The age of the first fracture did not emerge as statistically significant, but two curious factors were noted: of the four patients who first fractured after 48 months only one achieved a fair result; and of the five patients with fractures at birth or
within the first month of life four were amputated and in
the fifth the result was only fair.

The second statistically significant factor was the
greater amount of shortening in the unacceptable
results. In all but one patient with shortening of more
than six centimetres the result was unacceptable. This
shortening is associated with abnormal growth of the
distal tibial epiphysis. Bagdley, O'Connor and Kudner
(1952) noted that the appearance of this tibial epiphysis
was often delayed, but stated that it eventually appeared
and developed normally. We often observed angulation
at the ankle, combined with a thin angulated physis and a
small epiphysis. The fact that this thin, angulated physis
may not be growing leads to an apparent distal migration
of the pseudarthrosis and to shortening. This distal area
is where grafts usually fail to unite, and the cause of
failure may well be more biological than mechanical.
This ties up with the observation that, in some cases we
have studied since this present review, there was
decreased uptake of $^{99m}$Tc-polyphosphate in the distal
tibial epiphysis. We therefore suggest that there is a
continuing physiological disturbance in the affected
region of the bone; this disturbance may be related to
the aetiology of the pseudarthrosis, or it may be a
complication of treatment. Further experience with
bone imaging may help to answer this question and
provide a method of establishing an earlier prognosis.

Unfortunately, the response to bone grafting, which
should be one of the best indicators of how successful
treatment is likely to be, is also the most difficult to
assess objectively. Rapid resorption of a bone graft and,
sometimes, spreading destruction at the site of
the pseudarthrosis, are easy to observe and, in our
opinion, should play a large role in the decision for
amputation.

The presence of neurofibromatosis, which has
usually been thought to imply a worse prognosis, in our
series made no significant difference. Nor, in contrast
with Andersen (1976a) could we relate the type of
pseudarthrosis to the presence or absence of
neurofibromatosis. Incidentally, although we agree that
there are different radiographic types of pseudarthrosis,
we found that they merged: in time, or after operation,
the late sclerotic type often became the dysplastic type;
and the cystic type could, after operation, evolve into the
dysplastic type.

The most remarkable aspect of neurofibromatosis
in our series, however, was the finding that 25 per cent of
those so affected developed a glioma of the central
nervous system. We have no explanation for this
association, any more than we have for the association of
neurofibromatosis with congenital pseudarthrosis of the
tibia. However, the high incidence of gliomata in
patients with neurofibromatosis may temper enthusiasm
for continued grafting as the child grows older.

There are no absolute criteria for amputation. This
was illustrated by the contrast between two patients who
achieved union but who subsequently (because of the
functional impairment imposed by their weak, short,
deformed limbs) requested amputation, and a group of
patients who, though dissatisfied with the appearance
and function of their legs, would not consent to
amputation. To the patient, success is not always
equivalent to union, nor is amputation necessarily
preferable to functional and cosmetic failure. Some patients had invested an enormous amount of emotional energy into their poorly functioning, deformed limbs and had developed an unreasoning attachment to them nourished by many operations, many months in casts, and many years of social deprivation. This psychological response imposes a twofold burden on the surgeon: first, since many cases will end in amputation, he should early in the course of treatment, prepare the parents and patient for such an eventuality; secondly, he should not continue treatment beyond the point where there is a reasonable hope of success.

The factors we have been discussing should, when considered in combination, help to guide the surgeon. Thus, a 10-year-old child who fractured either at birth or after the age of six, whose grafts are rapidly resorbed, whose distal epiphysis is abnormal, whose leg is six centimetres short, and who has already had five bone grafts without union, is not a good candidate for further grafting. On the other hand, the four-year-old patient who fractured at 10 months and now, after two grafts, is only two centimetres short and has good incorporation of his graft (except for a pseudarthrosis at the distal end) is quite likely to achieve an acceptable result.

One of the problems in studying congenital pseudarthrosis of the tibia is its tremendous variability. This is illustrated by the contrast between one patient who remained healed after only three months in plaster, and another who never healed despite 11 bone grafts in six and a half years. Between such extremes were many who appeared to achieve clinical and radiographic union, only to refracture within 3 to 25 months. Clearly it is almost impossible to determine how much influence any one particular surgical procedure exerts on the natural history of the condition (Nicoll 1969). Moreover, the achievement of union is only half the problem; the other half is maintaining it (Birkett 1951). We need a long-term follow-up of a large number of patients who have undergone a large number of different procedures. Even when the patient with union reaches skeletal maturity there is no assurance that union will be permanent. Thus the patient shown in Figures 14, 15 and 16 when seen in 1972, was, despite numerous operations, gross shortening (equalised by operating on the other leg) and the long time it took to achieve union, classified as a good result because he had excellent function. Then, at the age of 27, a spontaneous fracture led to amputation.

From a study of the multiple factors in our cases, we are led to the conclusion that the conventional surgical procedures may, in any particular patient, be less important than the severity of the disorder. Consequently, new biological approaches to treatment, for example electrical stimulation (Brighton et al. 1975; Bassett, Caulo and Kort 1981) and microvascular bone grafting, (Taylor et al. 1978), are worthy of clinical investigation in those patients who do not respond to conventional methods.
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REFERENCES


