

Management of Congenital Pseudarthrosis of the Tibia in Infants

the Effect of Early Surgery on Achieving Union in
Pseudarthrosis and on Growth of the Limb—

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Abstract The study was undertaken to determine whether the results of surgical treatment for pseudarthrosis of the tibia in young children is so poor as to recommend that surgery be deferred till after three years of age. Radiographs and records were reviewed of twenty six consecutive surgical patients with Crawford Type IV congenital pseudarthrosis of the tibia. Growth abnormalities in the tibia, fibula and in the femur of the affected limb at the time of presentation were identified. The outcome from treatment, in terms of achieved union in pseudarthrosis, refracture rate and limb length, in thirteen children treated before the age of three years was compared with those treated later.

Union in pseudarthrosis was achieved in 12 (92%) of the 13 children treated before three years of age by excision of the pseudarthrosis, intramedullary rod insertion and dual onlay cortical bone grafting. Union was achieved in only 5 of 7 children who underwent the same procedure between 3 and 12 years of age. The extent of shortening in the limb at the time of surgery was least in those below three years of age. The limb length discrepancy remained unchanged in 11 children who had transarticular Rush rod insertion before the age of three years. Growth abnormalities in the fibula, tibia and in the femur were less pronounced and the rate of a refracture were not frequent in those treated early.

The study suggests that there is no need to defer surgery for pseudarthrosis of the tibia till the child is older than three years of age.

Introduction

Contrary to earlier reports, the problem of achieving union in congenital pseudarthrosis of the tibia does not appear to be insurmountable.

A recent review of 340 patients conducted by the EPOS from has found that union could be achieved in over 75% of instances⁽⁵⁾⁽³⁰⁾. The results of this multicenter study also suggested that the chances of achieving union in pseudar-

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throsis were lowest in children under the age of three years. Consequently, there has been consensus among the participants of that study that surgery should be avoided until after three years of age¹⁵⁾. Sharrard²⁴⁾ and Hardinge¹⁶⁾ have recommended deferring surgery till four years of age.

However, over the last 13 years we have treated 26 patients with established pseudarthrosis of the tibia and performed surgery on these children as soon as they presented to us. In 13 cases, we undertook surgery in children less than three years of age since we assumed that the sooner union was achieved, the less pronounced would be any growth abnormalities in the limb. In the light of the reservations for such early surgery expressed by the members of the EPOS study group, we felt we ought to evaluate the results of surgery in our children who were under three years of age, more closely.

The specific aims of the present study were to :

1. Determine the union rate in congenital pseudarthrosis in children treated under the age of three years.
2. Identify various growth abnormalities in the affected limb in children with congenital pseudarthrosis of the tibia, and determine whether these growth problems, including shortening, were less in children who were treated before the age of three years.

Material and methods

Case records and radiographs were reviewed of 30 consecutive patients with congenital anterolateral bowing of the tibia treated at this center, during the last 13 years. The patients

included 21 males and 9 females. The right side was involved in 13 patients, and the left was involved in 17. Cutaneous features of neurofibromatosis were evident in 26 patients. Two patients had radiological changes of fibrous dysplasia that were confirmed subsequently by biopsy.

Two patients were treated by bracing and did not develop pseudarthrosis. Of the remaining 28 patients who developed established pseudarthrosis of the tibia, two were lost to follow-up. The outcome of the remaining 26 patients with established Crawford Type IV pseudarthrosis⁹⁾ was analyzed in detail.

Twenty patients were skeletally immature when they presented for treatment, and thirteen of these children were less than three years old at the time of surgery.

The skeletally immature children underwent excision of the pseudarthrosis, internal fixation with an intramedullary rod and onlay cortical bone grafting. Bone graft was harvested from the subcutaneous surface of the opposite tibia in 19 children. At least two struts of cortical bone were placed alongside the pseudarthrosis site and no attempt was made to anchor the grafts to the tibia. Sheffield telescoping rods were implanted with the T pieces in the proximal and distal tibial epiphyses in two patients. All the other skeletally immature children had an intramedullary Rush rod passed from the calcaneum across the subtalar and ankle joints into the tibia. In one child simultaneous limb lengthening was also undertaken.

The six skeletally mature patients were treated by compression at the pseudarthrosis site. Proximal metaphyseal lengthening of the tibia with the help of a monolateral external fixator was performed in four of these six

Table 1. Frequency of Primary Union in Pseudarthrosis of the Tibia after the Index Operation in 26 Patients treated at Different Ages

Age at surgery	n	Mean age at surgery (years)	Union after index operation	
			n	%
<3	13	1.93±0.65	12	92.3
3-12	7	6.82±2.14	5	71.4
>12	6	18.50±3.21	4	66.6

patients.

The children who were under three years at the time of surgery have been followed up for a mean period of 6.27 years (range 2 to 11.4 years). Children who at the time of surgery were between 3 and 12 years have been followed up for a mean period of 5.44 years (range 2 to 8.1 years). The sequential radiographs of each patient were studied to identify changes in length of the femur, tibia and fibula. Tibial, femoral and fibular length measurements were made with the help of grid films and expressed as percentage difference as compared to the normal side. The initial tibial length measurement was made from the immediate postoperative film and not from the pre-operative film in view of the bowing and angulation at the pseudarthrosis site before surgery.

Growth abnormalities were noted in the tibia such as abnormal inclination in the proximal tibial physis, bowing in the shaft of the tibia, alterations in the shape, and alignment in the distal tibial epiphysis. The length of the fibula, the position of the lateral malleolus based on Malhotra's grading¹⁸⁾ and the presence of a fibular pseudarthrosis were noted.

The frequencies of these growth abnormalities were compared among the three groups of patients based on the timing of surgery *viz.* children under the age of three years, children between three and twelve years, and patients who were over twelve years of age, at the time of surgery.

The outcome of treatment in terms of achieving union in the pseudarthrosis, correction of deformities and equalization of limb length was compared among the groups. Eleven children who had a follow-up of more than five years were studied in greater detail to identify the alterations in growth, the frequency of any re-fracture, and any need for any secondary surgical procedure.

Results

Union rates

When the pseudarthrosis was united after the index operation, without the help of any supplementary procedure, it was referred to as "primary" union. If union occurred after some additional procedure then it was referred to as "secondary" union. The frequency of primary union was highest among children who were less than three years of age at the time of surgery (Table 1). The average time for union was 3.91 ± 2.31 (SD) months in children under 3 years, and 3.90 ± 1.24 (SD) months in children between 3 and 12 years. One supplementary bone grafting procedure was sufficient to obtain secondary union in the one child under three and in the two children between three and twelve years of age in whom primary union was not achieved. The pseudarthrosis remained ununited in two skeletally mature patients.

Re-fracture

A re-fracture at the site of the original pseudarthrosis occurred in two patients who were treated before the age of three years. One fracture occurred at three years after the primary surgery. In the other child, the re-fracture occurred at two years after the first Rush rod had been replaced with a longer rod. In both these instances of re-fracture, the implants also

Table 2. Extent of Shortening in the tibia in the immediate Postoperative Period and at Final Follow up

Age at surgery	n	Mean initial shortening (%)	Range(%)	Mean final shortening (%)	Range(%)
<3 years	11*	5.26	1.2 18.5	5.71	0 11.5
3 12 years	7	17.58	0.9 35.8	18.34**	5.3 37
>12 years	6	43.53	32 57.5	19.05***	4.2 25.2

* : Initial shortening could not be measured in two cases since the radiographs of the normal tibia were not available

** : Limb lengthening performed on one limb

*** : Limb lengthening performed on four limbs

Table 3. Frequency of Growth Abnormalities in the Tibia, Fibula and Femur in Skeletally Immature Children with Congenital Pseudarthrosis of the Tibia

Growth abnormality	<3 years (n = 13)		3 12 years (n=7)	
	At presentation	At follow up	At presentation	At follow up
Proximal tibial physeal tilt	0/13	0/13	2/7	2/7
Posterior bowing in the proximal third of the tibia	1/13	1/13	3/7	3/7
Ankle valgus	4/13	8/13	6/7	7/7
Proximal migration in the distal fibular physis	7/13	11/13	6/7	7/7
Fibular pseudarthrosis	7/13		6/7	
Fibular hypoplasia	6/13		1/7	
Femoral lengthening	5/13		6/7	
Femoral shortening	3/13		0/7	

failed. A repeat Rush rod implant with cortical bone grafting lead to union within four months in both these patients.

Both the children who had telescoping rods implanted, developed new pseudarthrosis at a more distal site. One of these children required three further operations to achieve union while in the other patient, a single bone grafting operation with the original rod in situ was sufficient to facilitate union.

Re-operation for rod exchange

The rush rod recedes into the distal half of the tibia as the child grows. At this stage the

short rod was removed and a longer rod was inserted in eight children who were under three at the time of initial surgery and in two children initially treated between three and twelve years of age. In two instances, rod exchange was possible without having to osteotomise the tibia. However, in eight patients the tibia had begun to bow just proximal to the rod, and an osteotomy was required to facilitate rod exchange. Autogenous cortical bone graft re-harvested from the contralateral tibia was placed around the osteotomy site in these cases. Union of the fresh osteotomy occurred within a

mean period of 3.8 months. The mean number of operations (including the index operation) performed per patient was 2.15 in children who were under three years and 1.7 in those who were between three and twelve years at the time of the index operation.

Limb length inequality

The degree of shortening in the affected tibia was least in children who were treated before the age of three years, and the shortening remained virtually unchanged till the final follow-up (Table 2). In 4 of the 13 children who were less than three years of age, the extent of tibial shortening gradually became reduced over time. In one child, the shortening remained virtually unchanged for more than three years. Tibial shortening increased in the remaining patients. Among children who were between 3 and 12 years of age at the time of the index operation, a spontaneous reduction in tibial shortening was noted in only one patient. In five children the shortening increased modestly. In one child, limb lengthening was done at the time of initial grafting and rod implantation. Though almost complete equalization of limb lengths had been achieved at the time, the residual shortening in the tibia at skeletal maturity was 26%.

Since the follow-up periods varied from patient to patient, the alteration in limb length was calculated as the alteration in length per year. The mean alteration in length of the tibia, per year was a 0.4% reduction in limb length discrepancy per annum in children treated under three years of age, and a 0.69% increase in limb length discrepancy per annum was noted in children treated between three and twelve years of age.

Growth abnormalities of the limb :

Growth abnormalities in the tibia, fibula and femur were more frequently seen in children treated later (Table 3).

Discussion

The results of the present study challenge the view expressed by the members of the EPoS study group that it is difficult to achieve union of pseudarthrosis in children under the age of three years. Not only was union achieved in most children under three years of age, but the primary union rate of 92% was one of the highest reported in the literature⁽¹⁾⁻⁽⁴⁾⁽¹⁰⁾⁻⁽¹⁷⁾⁽¹⁹⁾⁻⁽³⁰⁾. What was even more interesting was that in the present study, the union rate in children under the age of three years was higher than that obtained in older children. On the other hand, Boero et al.⁷⁾ had abysmally poor results in children less than five years of age, with a union rate of 14%, while they had 86% success in older children. Ghanem et al. also reported very poor results in young children¹³⁾. What could be the reason for such conflicting results? The method of treatment adopted by those authors was the Ilizarov technique. In contrast, our operative approach entailed excision of the pseudarthrosis, intramedullary nailing and autogenous cortical bone grafting. Grill et al.¹⁵⁾ highlighted the problems of applying the Ilizarov apparatus to young children and suggested that these factors may have contributed to their poor results. With the technique we employed, no such problems were encountered. The rationale of the use of cortical graft rather than cancellous graft¹⁷⁾, and the benefit of the insertion of an intramedullary rod, has been noted in previous studies⁽²⁾⁽³⁾⁽⁵⁾⁽⁶⁾⁽¹²⁾⁽¹⁷⁾. The high rate of union achieved in the present study and in other

earlier reports²⁾³⁾⁶⁾¹²⁾¹⁷⁾ do not support the view expressed by the EPoS study group that bone grafting and internal fixation methods were of little value for this condition¹⁵⁾.

The present study shows that union can be achieved in young children by bone grafting and intramedullary rod fixation. However, as Weintraub and Grill²⁰⁾ emphasized, "viewing the healing of the pseudarthrosis alone as a surgical issue underestimates many other problems that affect the outcome". We endorse this view and feel that it is imperative that other problems such as growth abnormalities in the tibia and fibula, which are associated with the condition, are also addressed.

Limb length inequality encountered in pseudarthrosis of the tibia may be due to inherent growth abnormalities seen in this condition or due to the ill effects of treatment.

Comparison of the length of the limb at commencement of treatment, shows that the longer the pseudarthrosis had untreated remained un-united, the greater the extent of shortening in the tibia. This observation justifies our approach to early intervention. However, does early surgery in any way hamper the subsequent growth of the limb? The treatment adopted for managing skeletally immature children in this study entailed transfixing the distal growth plate of the tibia with an intramedullary rod. Damage to the growth plate either while passing the reamer or the rod at the time of surgery, and the retention of the rod for several years post-operatively might adversely affect the growth plate resulting in progressive growth retardation. However, the present study showed that in children who were followed up for over five years, a smooth Rush rod inserted from the calcaneum across the distal tibial

growth plate did not result in any progressive shortening in the limb. The percentage shortening in the limb remained virtually unchanged in some of these patients, while in others there was an actual reduction in the limb length inequality. On the other hand, the two children who underwent Sheffield telescoping rod implantation did show progressive shortening indicating that the growth plate was damaged. Both these patients had the telescoping rod inserted from the ankle joint.

Yet another important problem in the management of congenital pseudarthrosis in the tibia is the tendency for a refracture after union. The frequency of a refracture following rod implantation was not unduly high in those children who were treated before the age of three years.

The need for a re-operation again was not appreciably greater in children treated before three years of age. The main reason to re-operate was for rod exchange. We believe that it is prudent to retain an intramedullary rod till skeletal maturity as an internal splint to minimize the risk to a re-fracture. The fact that a sizeable proportion of children showed a tendency for bowing in the tibia as they outgrew the initial intramedullary rod indicated that the risk to a refracture was very real. If rod exchange could be performed before bowing in the tibia occurred, then the need for osteotomy of the bone might be avoided. This would reduce the morbidity of the re-operation. We opted to perform autogenous bone grafting at the time of rod exchange whenever an osteotomy was needed as we felt that union might not be achieved without the bone graft.

The higher frequency of other growth abnormalities such as abnormal physeal inclination,

posterior bowing in the proximal tibia, and abnormalities in the distal fibular physis noted in older children suggested that all these abnormalities developed over a period of time in children with pseudarthrosis. It would follow that the sooner the union was achieved, then the less would be the chance of these growth abnormalities developing.

In conclusion, the frequency of union was higher, the shortening in the limb was less and associated growth abnormalities were less frequent in children with congenital pseudarthrosis of the tibia who were treated before the age of three years. In the light of these results we see no justification in deferring surgery for pseudarthrosis of the tibia till four years of age. However, in these young children we recommend that the procedure to adopt is excision of the pseudarthrosis, cortical bone grafting and intramedullary rod implantation in preference to the Ilizarov technique since the latter procedure has been reported to have poor results in young children⁷⁾⁽³⁾⁽⁵⁾⁽⁹⁾.

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