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OPERATIVE TREATMENT OF CONGENITAL PSEUDARTHROSIS OF THE TIBIA

Factors Influencing the Primary Result

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Congenital pseudarthrosis of the tibia is a rare condition of unknown aetiology. A large number of operative methods have been suggested for its treatment, "establishing the fact that there is some doubt concerning the best form of treatment and emphasizing the need for systematic treatment continued until the completion of bone growth" (Van Nes 1966).

Interest in the phenomenon has increased. During the past 25 years more than 50 papers have appeared in English, German and French. However, the therapeutic problems still remain unelucidated. It therefore seemed of interest to publish a material collected from several countries in an attempt to elucidate whether some operative methods were better than others or whether the therapeutic results were influenced by some special factors, operative or non-operative.

CLINICAL MATERIAL

Congenital pseudarthrosis of the tibia is taken to mean all congenital fractures of the tibia as well as pseudarthroses of the tibia after a pathological fracture or osteotomy on a lower leg with congenital anterior angulation. Cases of generalized skeletal diseases were excluded, if these diseases could be assumed to have contributed to the occurrence of fracture or pseudarthrosis.

From 18 orthopaedic departments and hospitals in Denmark, Norway, Sweden and the USA, all cases diagnosed in the individual departments as congenital pseudarthrosis of the tibia were reviewed by the author. In 60 cases, all fulfilling the above-mentioned definition, complete records and x-ray films dating from the time of the first treatment until treatment was completed or until 1970 were available. In 40 cases the patients were born between 1930 and 1960. In four of these cases the pseudarthrosis arose after an osteotomy. To obtain a more accurate definition

of the disease concept these four cases were not included and the present material thus consists of 36 patients.

It was demonstrated in a previous paper (Andersen 1973) that on the basis of the primary x-ray films a distinction may be made between four different radiological types of congenital tibial pseudarthrosis. In the present material primary x-ray films were available in 18 cases. In the remaining cases x-ray films from before the first treatment, but after the fracture were available.

Union of the pseudarthrosis is taken to mean all cases where the affected leg had been stable with radiological union for at least 24 months and where no re-fracture later occurred in the observation period. Union followed by re-fracture is taken to mean all cases where the affected leg had been stable with radiological union for at least 12 months but where a new fracture later followed. The re-fractures occurred from 1½ to 8 years after the last operation on the pseudarthrosis. For patients who at the time of the follow-up had union of the pseudarthrosis and who had reached skeletal maturity the mean observation time for the union was 5 years (2–9 years): For patients with union who had not reached skeletal maturity the mean observation time was 7 years (3–9 years).

Table 1. The result in 1970 of the treatment in relation to radiological type of pseudarthrosis.

Radiological type	Total no.	No. with union	No. with union who have reached skeletal maturity	Average no. of operations in cases with union
Dysplastic	14	6	4	6
Cystic	11	5	3	5
Sclerotic	8	5	3	4
Clubfoot	3	3	2	1
Total	36	19	12	–

Table 2. The number of operative methods used in the treatment of 34 cases of congenital tibial pseudarthrosis.

Type of operation	No.
Diaphyseal fragmental reversal and intramedullary fixation	23
Dual onlay bone grafting	10
By-pass graft	1
Translocation of the fibula	1
Telescoping	4
Intramedullary rodding and bone grafting	22
Simple bone grafting	29
Osteosynthesis and bone grafting	12
Supplementary operations	34
Others	7
	143

Table 3. The results of certain operative methods in the treatment of congenital tibial pseudarthrosis.

Type of operation	Primary operations			Later operations			
	No.	Mean age at operation	No. with union	No.	Mean age at operation	No. with union	No. with union after suppl. op.
Diaphyseal fragmental reversal and intramedullary fixation	5	40 months	0	18	60 months	2	5
Dual onlay bone grafting	0			10	74 months	1	0
Intramedullary rodding and bone grafting	4	28 months	0	16	69 months	2	0
Simple bone grafting	13	36 months	0	15	69 months	0	0
Osteosynthesis and bone grafting	2	19 months	0	8	80 months	1	1

The results of the treatment at the time of the follow-up in 1970 are listed in Table 1. As the prognosis for the clubfoot type is very favourable and quite different from the prognosis of the other types of pseudarthroses, these cases have been omitted in the following analyses.

The operative methods are listed in Table 2. Simple bone grafting means the insertion of a bone graft over the pseudarthrosis without any fixation of the graft with metal. No distinction is made between the various ways of placing the graft. Supplementary operations means the application of additional bone grafts to the pseudarthrosis site (30 cases) or replacement of an intramedullary rod (4 cases), without any other operative intervention.

Union of the pseudarthrosis followed after two operations with diaphyseal fragmental reversal and intramedullary fixation, after two operations with intramedullary rodding and bone grafting, after one operation with dual onlay bone grafting, after one operation with telescoping and after one operation with a single bone graft fixated with screws. The results of the individual operative procedures are given in Table 3. In no case was the primary operation followed by union. The distribution of the different types of pseudarthroses submitted to the various operative methods varied but could not be related to the results. The possible influence of the previous operations on the result of a single operation in relation to type of operative method is seen from Table 4. In order better to evaluate the

Table 4. The results of certain operative methods in relation to the previous operation in the same case of pseudarthrosis.

Type of operation	Type of preceding operation: Diaphyseal fragmental reversal	Dual onlay bone graft	Telescoping	Intramedullary rodding	Osteosynth. and bone graft	Simple bone graft
Diaphyseal fragmental reversal	9 (5)	0	0	5 (1)	0	3 (1)
Dual onlay bone graft	0	2 (2)	0	0	2 (2)	4 (1)
Telescoping	0	0	0	1 (1)	1 (1)	0
Intramedullary rodding and bone grafting	5 (1)	2	0	8 (2)	0	1
Osteosynth. and bone grafting	1 (1)	3 (1)	0	0	3	1 (1)
Simple bone grafting	0	0	0	0	2	11 (4)

Figures indicate no. of operations.
Figures in brackets indicate no. of operations followed by union, or by union and refracture, or by union after a supplementary operation.

Table 5. The influence of resection of the pseudarthrosis on the result of the operative treatment.

Radiological type of pseudarthrosis	Operation with resection of pseudarthrosis		Operation without resection of pseudarthrosis		
	No.	Union	No.	Union	
Dysplastic	28	3	14	0	($P < 0.005$)
Cystic	22	0	7	0	($P > 0.05$)
Sclerotic	13	2	8	1	($P > 0.05$)

P-value is calculated from the Four Fold Table Test.

Table 6. The number of operations and the results in relation to the age of the patient at the time of operation.

Age (years)	No. of operations	No. followed by union
0–3	45	0
3–6	53	4
6–9	20	5
9–12	10	3
12–15	8	3
15–18	2	0

tendency to union, the number of operations followed by union as well as the number of operations followed by union after supplementary operations and operations followed by union and refracture are given.

An accurate survey of the complications following the operative interventions is difficult to present because of the retrospective nature of the study. In two cases serious infection followed operation, one case after a diaphyseal fragmental reversal and intramedullary fixation and one case after simple bone grafting. Following the diaphyseal fragmental reversal, non-union at the proximal osteotomy site followed the operation in six cases, five of these being of the cystic type.

No case operated upon without bone grafting was followed by union of the pseudarthrosis. In 46 operations not including supplementary operations autologous bone was used, in three operations followed by union and in six operations by union and refracture. In 21 operations homologous bone was used, in three operations followed by union and refracture.

Intramedullary fixation was not used in the treatment of pseudarthroses of the sclerotic type except for a few cases. Among cases of the dysplastic and cystic types 11 were treated with intramedullary rodding and fixation of the tarsus. One operation was followed by union and five operations by migration or breakage of the intramedullary pin. In 26 operations intramedullary rodding without fixation of the tarsus was used, in two operations followed by union and in 12 by migration of the intramedullary pin.

Table 7. Results of operative treatment in relation to the total number of operations.

Total no. of operative interventions	No. of pseudarthroses	Pseudarthroses with union	Amputations
1–3	12	3	8
4–6	12	8	4
7–9	7	5	2

Table 8. Number of operations and their results in cases treated in Scandinavia and in USA.

	No. of hospitals	No. of pseudarthr.	No. of ord. op.	No. of suppl. op.	Pseudarthr. with union
Scandinavia	7	11	33	2	1
USA	3	22	72	32	15

The effect of resection of the pseudarthrosis in operations other than supplementary ones is seen in Table 5. The results of all operations including supplementary operations in relation to age is shown in Table 6. Two cases of the dysplastic type had no operative treatment but were immobilised in plaster and brace and finally amputated at the age of 7 and 9 years, respectively. The remaining 31 cases were operated on from one to nine times each. The rate of union in relation to number of operative interventions is shown in Table 7. The distribution of the cases in relation to nationality of treating department and the result hereof is shown in Table 8.

DISCUSSION

Analysing the results of operative treatment of congenital pseudarthrosis of the tibia one comes up against several difficulties. Owing to the rare occurrence of the condition even large clinics can hardly reach a double figure within a reasonable period. This being the case it seems acceptable to evaluate this material a little closer and involve all operative interventions in the analysis.

Several authors have classified congenital tibial pseudarthrosis by type (Camurati 1930, Févre 1954, Guilleminet & Ricard 1958, Hardinge 1972, Sage 1971) but no one has attempted an actual analysis of incidence and prognosis of the individual types, although the question has been raised (Févre 1954, Nicoll 1969). The radiological classification used in the present analysis (Andersen 1973) is based upon the

radiological findings in the pre-pseudarthrosis stage. Although x-ray films from prior to or immediately after the fracture were available in only 18 cases all cases could be classified on the basis of the x-ray films.

From Table 1 it is apparent that the prognosis of congenital pseudarthrosis of the tibia depends upon its radiological type. By far the best results were found after treatment of pseudarthroses of the clubfoot type in which union was obtained after fewer operations and even by plaster cast alone. This was not previously considered possible (Boyd & Sage 1958). However, in a large material Hardinge (1972) found union in three per cent after immobilization in plaster as the only treatment. As the prognosis for the clubfoot type differed essentially from the other types of pseudarthroses these three cases were not considered in the following analysis.

To-day it is not possible to decide with certainty which type of operation should be selected for each individual case of pseudarthrosis of the tibia. However, it seems beyond doubt that simple bone grafting affords a poorer result than other methods (Hardinge 1972, McBryde & Stelling 1972, Sofield 1971). Operation by dual onlay bone grafting and by intramedullary rodding affords good results according to several authors (Apoil 1970, Sage 1971, Sofield 1971, Van Nes 1966). In the present material the results of the various operative methods showed no significant differences when all primary and supplementary operations were excluded. Considering all operations the results after the diaphyseal fragmental reversal and intramedullary fixation might be better than after any other operation but the difference is not statistically significant ($P > 0.20$). Lastly the diaphyseal fragmental reversal operation carries an appreciable number of complications, the most important being non-union at the proximal osteotomy site. However, new operative methods (d'Aubigné et al. 1970, Witt & Refior 1970) as well as prophylactic operations (Lloyd-Roberts & Shaw 1969) could not be evaluated due to the retrospective nature of the analysis.

Considering the single elements in the operative procedures bone grafting is a *sine qua non* in any operation for congenital pseudarthrosis of the tibia. In most cases grafting was done by autologous bone but there was no difference between the results after autologous and after homologous bone grafting ($P > 0.20$) considering all other factors equal.

Regarding the fixation of the pseudarthrosis there was no statistically significant difference between the results after plating and after intra-

medullary rodding. Probably there was a more pronounced tendency towards refracturing after plating. Of four pseudarthroses of the sclerotic type operated upon with intramedullary fixation there were no refractures, whereas there was one refracture after three sclerotic pseudarthroses operated upon with dual onlay bone grafting. The refractures after plating always occurred at the distal end of the plate. Among all operations there were 11 refractures, four of which occurred after dual onlay bone grafting and one after plating and grafting, whereas only one occurred after intramedullary rodding. Whether or not intramedullary rodding with fixation of the tarsus gave a better fixation could not be shown ($P > 0.05$).

Some authors have recommended lengthening of the Achilles tendon or amputation of the forefoot in order to improve the chance of union in the pseudarthrosis. In the present analysis only two amputations of the forefoot were done and only one lengthening of the Achilles tendon. As resection of the pseudarthrosis may mimic the same effect the results after operations with and without resection of the pseudarthrosis were considered. For pseudarthroses of the dysplastic type resection of the pseudarthrosis gave a significantly better result ($P < 0.005$), whereas there were no differences for the cystic and sclerotic types of pseudarthrosis.

Previously it has been claimed that operative treatment of congenital tibial pseudarthrosis should not be started until after the age of 4 to 6 years. Now several authors emphasize the advantages of an early operation, and also of a rapid re-operation if the primary operation does not lead to union (McBryde & Stelling 1972, Sage 1971, Sofield 1971, Sulamaa & Vilkki 1963). According to Hardinge (1972), however, "long term union" is not obtained by any operation before the age of 33 months. As seen from Table 6 one might get the impression that age is a factor of importance. This is probably not correct and as seen from Table 7 the total number of operative interventions presumably is of much more importance.

The influence of a previous operation is difficult to evaluate. From Table 4 one might get the impression that the continuous use of the same operative procedure may better the chance of union. Obviously this impression is not statistically valid. From the analysis one might conclude that so far there seems to be no operative method of choice, as long as one uses a method which gives a solid fixation of the pseudarthrosis and includes transplantation of bone.

The present material is selected in that not all cases treated in the

different hospitals are included. From Table 8 it is apparent that the treatment in American and Scandinavian departments differs in respect to the use of supplementary bone grafting and it is obvious that the latter procedure favours a good result.

On the basis of the analysis, one might conclude that there so far seems to be no operative method of choice in the treatment of congenital tibial pseudarthrosis. The prognosis is dependent on the type of pseudarthrosis. The result of the operation is favoured by solid fixation, resection of the pseudarthrosis and abundant bone grafting. Experience in treating cases of this type seems to be of equal importance.

SUMMARY

A total of 36 cases of congenital tibial pseudarthrosis born 1930–1960 and treated in American and Scandinavian orthopaedic hospitals have been analysed. There seems to be no operative method of choice. The prognosis depends on the radiological type. Factors favouring a good result from the operation are solid fixation, resection of the pseudarthrosis and abundant bone grafting. Experience in treating cases of this type seems to be of equal importance.

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