

Dept. of Orthopaedic Surgery T2, Gentofte Hospital,
Copenhagen, Denmark.

CONGENITAL ANGULATION OF THE LOWER LEG AND CONGENITAL PSEUDARTHROSIS OF THE TIBIA IN DENMARK

K. SKOU ANDERSEN

Accepted 19. vi. 72

So-called congenital pseudarthrosis is rarely congenital, but has developed during early childhood, in consequence of a pathological fracture. Congenital pseudarthroses have been reported in the tibia, clavicle, femur, and ulna (Moore 1957). The most common site is the tibia, and such a case was reported in 1708 by Hatzoecher (Camurati 1930).

In order to collect a sizable number of cases having congenital pseudarthrosis of the tibia, I have tried to determine the number of patients in Denmark having congenital angulation of the lower leg (*crus curvatum congenitum*) and congenital pseudarthrosis of the tibia.

M A T E R I A L

Congenital angulation of the lower leg (*crus curvatum congenitum*) is taken to mean all congenital angulations of the lower leg insofar as systemic skeletal diseases can be ruled out as their cause (Lindemann 1961). Congenital pseudarthrosis of the tibia is taken to mean all congenital fractures of the tibia as well as all pseudarthroses of the tibia in children following upon a pathological fracture, insofar as such fractures are not due to systemic skeletal diseases.

All cases of congenital angulation of the lower leg and of congenital pseudarthroses of the tibia in patients born in Denmark during the period 1940 to 1965 were recorded after perusing the files of all Danish orthopaedic departments as well as departments of orthopaedic surgery. Moreover, questionnaires were sent to and completed by all Danish departments of surgery.

A total of 46 patients with congenital angulation of the lower leg had been admitted. Twelve had posterior and all the others anterior bowing of the lower leg. Among the 34 patients of the latter group, 15 showed complete and one partial aplasia of the homolateral fibula, and 3 exhibited pronounced hypoplasia of the homolateral foot. The distribution by sex and side affected is shown in Figures 1 and 2.

Figure 1. Distribution of congenital angulation of the lower leg by sex.

	Male	Female	Total
Congenital angulation of the lower leg with anterior bowing and aplasia of the homolateral fibula	13	3	16
Congenital angulation of the lower leg with anterior bowing and hypoplasia of the homolateral foot	2	1	3
Congenital angulation of the lower leg with anterior bowing (pre-pseudarthroses)	8	7	15
Congenital angulation of the lower leg with posterior bowing	6	6	12
Total	29	17	46

Figure 2. Distribution of congenital angulation of the lower leg by side affected.

	Right	Left	Bilateral
Congenital angulation of the lower leg with anterior bowing and aplasia of the homolateral fibula	9	5	2
Congenital angulation of the lower leg with anterior bowing and hypoplasia of the homolateral foot	1	2	—
Congenital angulation of the lower leg with anterior bowing (pre-pseudarthroses)	4	11	—
Congenital angulation of the lower leg with posterior bowing	7	5	—
Total	21	23	2

Figure 3 gives the frequency of tibial fractures and pseudarthroses among the 46 patients with congenital angulation of the lower leg. Pseudarthrosis of the tibia due to pathological fractures occurred only in children with congenital angulation of the lower leg and never in children with a normal lower leg. Out of the 12 tibial fractures, which occurred among 15 patients with congenital angulation of the lower leg without attendant aplasia of the fibula or hypoplasia of the foot, two had been present at birth, whereas the other 10 occurred at an age of from 12 to 39 months. The healing tendency following fracture or osteotomy was normal in only 2 patients of this group, whereas 13 patients developed pseud-

Figure 3. Fractures, osteotomies and pseudarthrosis in congenital angulation of the lower leg.

	Fracture	Osteotomy	Pseudarthrosis
Congenital angulation of the lower leg with anterior bowing and aplasia of the homolateral fibula	—	2	—
Congenital angulation of the lower leg with anterior bowing and hypoplasia of the homolateral foot	1	1	—
Congenital angulation of the lower leg with anterior bowing (pre-pseudarthrosis)	12	3	13
Congenital angulation of the lower leg with posterior bowing	—	—	—
Total	13	6	13

arthrosis of the tibia. Three patients had associated congenital deformities, i.e. constriction band on the homolateral leg, contralateral pes equino-varus, and syndactyly on the hands. In two of these patients the tibia united normally—in one of them after a congenital fracture and in the other one following osteotomy at the age of 6 months (Figure 4).

For 14 patients X-rays of the fracture prior to treatment were available. In 11 these films of the tibia could be naturally assigned to 3 groups of uniform radiological appearance. Two patients exhibited cystic lesions (Figure 5), 5 showed narrowing of the middle part of the tibial shaft giving an impression of segmental dysplasia (Figure 6), and 4 had a pronounced anterolateral convex curvature with severe sclerosing and thickening of the cortex (Figure 7). This type was found in the two patients with contralateral club foot.

In one patient whose fracture did not occur until the age of 39 months, there was only moderate anterior bowing with localized sclerosis at the junction of the middle and lower third of the tibia (Figure 8).

When the analysis was concluded in 1970, union had occurred in 6 patients. However, only 3 of these patients have reached skeletal maturity. In 3 cases amputation had been done because of non-union. In 4 cases with union the primary X-ray films had shown a congenital angulation of the lower leg of the "club-foot type" (Figure 7).

The incidence of congenital pseudarthrosis of the tibia in Denmark is apparent from Figure 9.

DISCUSSION

Little has been published about the incidence, aetiology, and pathogenesis of congenital pseudarthrosis of the tibia. Its treatment, on the

Figure 4. Results of treatment.

No.	Age at fracture (months)	Age at osteotomy	Associated congenital deformities	Pseudarthrosis	Number of operative procedures for pseudarthrosis	Age at operation	Result at follow-up (1970)	Skeletal maturity
1	--	6 mo	yes	none	—	—	union	no
2	birth		yes	yes	1	2 years	union	yes
3	birth		yes	none	—	—	union	no
4	28 mo		no	yes	3	2-4 years	amputation	yes
5	3 mo		no	yes	5	4 months-4 years	non-union	no
6	9 mo		no	yes	5	3-4½ years	amputation	no
7	13 mo		no	yes	5	2-14 years	union	yes
8	39 mo		no	yes	4	4-9 years	non-union	no
9	3 mo		no	yes	1	3½ years	union	yes
10	--	14 mo	no	yes	3	3-5½ years	amputation	yes
11	3 mo		no	yes	1	3 months	amputation	no
12	15 mo		no	yes	—	—	amputation	no
13	--	18 mo	no	yes	4	2-3 years	union	no
14	2 mo		no	yes	1	4 years	non-union	no
15	5 mo		no	yes	—	—	amputation	yes

mo = months



Figure 5. Congenital pseudarthrosis of the tibia with cystic lesions in the bones.



Figure 6. Congenital pseudarthrosis of the tibia with hour-glass constriction ("dysplastic type").



Figure 7. Congenital pseudarthrosis of the tibia as seen in the cases with contra-lateral club-foot ("club-foot" type).

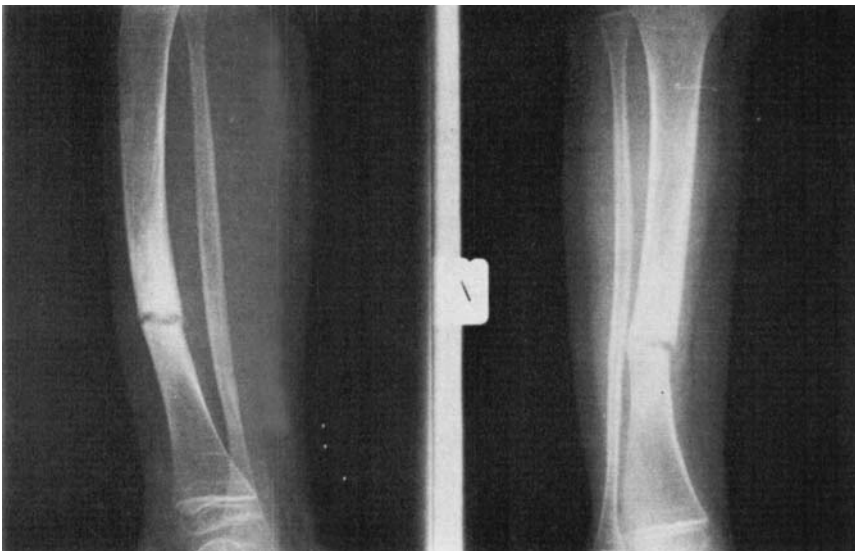


Figure 8. Congenital pseudarthrosis of the tibia recognized as late as at the age of 39 months.

Figure 9. Incidence of congenital pseudarthrosis of the tibia in Denmark.

Period	No. of live-born	Cases with pre-pseudarthrosis
1940-1944	395,932	1
1945-1949	447,744	2
1950-1954	387,686	3
1955-1959	377,443	4
1960-1964	396,093	4
1965	85,796	1
Total	2,090,694	15

Incidence: 1:140,000

other hand, has been the subject of much discussion, but although numerous therapeutic methods have been described, the chance of union is still not even 20 per cent (Stelling 1971). Only a few of the so-called congenital pseudarthroses of the tibia are in fact present at birth, the greater part developing in the course of the first years of life in consequence of a pathological fracture in an anteriorly bowed congenital angulation of the lower leg (Froelich 1910, Henderson 1925, Kite 1941, Lindemann 1943, Büttner & Eysholdt 1950, Moore 1957, Van Nes 1966). A few authors (Henderson 1925, Dillehunt & Le Concq 1928, Compère 1936, Moore 1957, Evans & Eggers 1962, Eichenholtz 1970) have included cases which have certainly not been congenital pseudarthrosis of the tibia. In other publications (Camurati 1930, Bischofberger 1949, Charnley 1956, Boyd & Sage 1958, Van Nes 1966, Eyre-Brook et al. 1969) it is not quite clear whether all the cases have been congenital tibial pseudarthroses. This lack of certainty and uniformity in classifying the various materials, combined with the rare occurrence of congenital pseudarthrosis of the tibia and the ignorance concerning its spontaneous course, in particular during the pre-pseudarthrosis stage (Lloyd-Roberts & Shaw 1969), has led to quite some doubt in assessing the therapeutic results and has increased the number of confusing and conflicting results reported in the literature (Nicoll 1969).

Congenital pseudarthrosis of the tibia never seems to develop in cases of congenital angulation of the lower leg with posterior bowing (Debrunner 1933, Heyman & Herndon 1949, Badgley et al. 1952). Pseudarthrosis in congenital angulation of the lower leg with anterior

bowing and homolateral aplasia of the fibula has been reported in only 4 cases since the advent of X-rays (Froelich 1910, Camurati 1930, Madsen 1956, Aitken 1959), but in general the prognosis with respect to spontaneous fracture and pseudarthrosis is considered favourable. All other cases of congenital angulation of the lower leg with anterior bowing without aplasia of the fibula or foot may give rise to spontaneous fracture and pseudarthrosis, and this entire group should thus be considered as non-manifest congenital pseudarthrosis of the tibia or pre-pseudarthrosis (Lloyd-Roberts & Shaw 1969). As previously demonstrated (Andersen et al. 1968), cases with other associated congenital deformities, however, seem to have a somewhat better prognosis than those without. A so-called congenital pseudarthrosis of the tibia hardly occurs in a previously normal-looking tibia. In the present material, which also comprised all Danish departments of surgery, there was no such case in which a pathological fracture could be suspected as the cause of tibial pseudarthrosis.

The distribution by sex and side affected by fracture and pseudarthrosis shows, as in other investigations (Moore 1957, Sofield 1971), a slight preponderance of left-sided cases and of males. The reported time of occurrence of the fracture has differed somewhat, but the most common age interval is 0-18 months (Lloyd-Roberts & Shaw 1969), and only a minority of the fractures are truly congenital. Therefore, the term congenital pseudarthrosis of the tibia is a misnomer, but it will be maintained, as it is generally used in the literature.

Several authors (Gasne 1907, Henderson 1925, Compère 1936) have tried to classify the congenital tibial pseudarthroses on the basis of the radiological appearances, including the degree of atrophy and the size of the distal fragment in the assessment. Camurati (1930) classified the pseudarthroses of his material by the degree of instability at the pseudarthrosis site. Others (Badgley et al. 1952, Boyd & Sage 1958, Lloyd-Roberts & Shaw 1969) have mentioned the cystic and dysplastic changes that may at times be found on the X-ray films. However, there has not been any analysis on the number of the various types and their prognosis, although Nicoll (1969) believed that there must presumably be 2 or 3 different clinical types of congenital tibial pseudarthrosis.

On the basis of the materials published so far, which have rarely included the primary X-ray films, it is not possible to decide whether the classification used in the present study is adequate. Presumably there is a fourth type, as the present material did not include cases of

late congenital tibial pseudarthroses as described by several previous authors (Büttner & Eysholdt 1950, Moore 1957, Sofield 1971).

The literature is scarce on the incidence of congenital tibial pseudarthrosis. In 1950 Büttner & Eysholdt were able to collect only 276 cases from the world's literature, whereas McFarland (1951) did not find a single case among 48,000 new borns. Among 105,119 newborns, Madsen (1956) found only 60 congenital fractures of the long bones, and less than 10 per cent were truly congenital. In the Shriners Hospitals, U. S. A., congenital pseudarthrosis of the tibia was found in less than 0.1 per cent of the patients treated (Sofield 1971), and in the Instituta Rizzoli, Bologna, only 50 cases had occurred in 50 years (Païs 1953). In the present study the incidence was 1 in 140,000 newborns. Whether the incidence is increasing is difficult to ascertain, as the study is retrospective and as such carries some uncertainty.

In the cases where pseudarthrosis developed the treatment was operative with a few exceptions. The 13 patients had 15 different operations, so that it is not possible to assess the value of each individual procedure.

SUMMARY

This is a retrospective study of congenital angulation of the lower leg (crus curvatum congenitum) and/or congenital pseudarthrosis of the tibia in children born in Denmark during the period 1940–1965. Congenital pseudarthrosis of the tibia was found only in patients with congenital angulation of the lower leg with anterior bowing without homolateral aplasia of the fibula or hypoplasia of the foot. This group is designated pre-pseudarthroses (non-manifest congenital pseudarthroses of the tibia).

The incidence of congenital pseudarthrosis of the tibia was 1 in 140,000 newborns. On the basis of the primary X-ray films the congenital tibial pseudarthroses could be divided into 3 radiological types: (1) cystic, (2) dysplastic, and (3) "club-foot type". Presumably a fourth type exists. In the present material union of the pseudarthrosis occurred mainly in the "club-foot type".

REFERENCES

- Aitken, G. T. (1959) Amputation as a treatment for certain lower-extremity congenital abnormalities. *J. Bone Jt Surg.* **41-A**, 1267–1285.
Andersen, K. S., Bohr, H. & Sneppen, O. (1968) Congenital angulation of the lower leg. *Acta orthop. scand.* **39**, 387–397.

- Badgley, C. E., O'Connor, S. J. & Kudner, D. F. (1952) Congenital kyphoscoliotic tibia. *J. Bone Jt Surg.* **34-A**, 349-369.
- Bischofberger, C. (1949) Erfahrungen in der operativen Behandlung der kongenitalen Unterschenkelpseudarthrose. *Z. Orthop.* **78**, 432-461.
- Boyd, H. B. & Sage, F. P. (1958) Congenital pseudarthrosis of the tibia. *J. Bone Jt Surg.* **40-A**, 1245-1270.
- Büttner, A. & Eysholdt, K.-G. (1950) Die angeborenen Verbiegungen und Pseudarthrosen des Unterschenkels. *Ergebn. Chir. Orthop.* **36**, 165-222.
- Camurati, M. (1930) Le pseudartrosi congenite della tibia. *Chir. Organi Mov.* **15**, 1-162.
- Charnley, J. (1956) Congenital pseudarthrosis of the tibia treated by the intramedullary nail. *J. Bone Jt Surg.* **38-A**, 283-290.
- Compère, E. L. (1936) Localized osteitis fibrosa in the newborn and congenital pseudarthrosis. *J. Bone Jt Surg.* **18**, 513-523.
- Debrunner, H. (1933) Zur natürlichen Aufrichtung verbogener Knochen. *Arch. orthop. Unfall-Chir.* **32**, 512-514.
- Dillehunt, R. B. & LeCocq, J. F. (1928) Pseudarthrosis of the tibia in young children. *J. Amer. med. Ass.* **90**, 1615-1617.
- Eichenholtz, S. N. (1970) Congenital pseudarthrosis of tibia. *N. Y. med. J.* **70**, 1049-1058.
- Evans, E. B. & Eggers, W. N. (1962) Nonrigid fixation in the treatment of childhood pseudarthrosis of the tibia. *Amer. Surg.* **28**, 510-517.
- Eyre-Brook, A. L., Baily, R. A. J. & Price, C. H. G. (1969) Infantile pseudarthrosis of the tibia. *J. Bone Jt Surg.* **51-B**, 604-613.
- Froelich, M. (1910) Traitement des pseudarthroses congénitales. *Rev. Chir. orthop.* **21**, 1-19.
- Gasne, E. (1907) Coudures et pseudarthroses congénitales de la jambe. *Rev. Chir. orthop.* **8**, 267-286.
- Henderson, M. S. (1925) Pseudarthrosis of the tibia in children. *J. Bone Jt Surg.* **7**, 340-354.
- Heyman, C. H. & Herndon, C. H. (1949) Congenital posterior angulation of the tibia. *J. Bone Jt Surg.* **31-A**, 571-580.
- Kite, J. H. (1941) Congenital pseudarthrosis of tibia and fibula. *Sth. med. J. (Bgham., Ala.)* **34**, 1021-1032.
- Lindemann, K. (1943) Die Pathogenese der angeborenen Unterschenkelpseudarthrose. *Z. Orthop.* **74**, 256-272.
- Lindemann, K. (1961) Die angeborenen Deformitäten des Unterschenkels. *Handbuch der Orthopädie*, Band IV, Teil II, pp. 741-780. Stuttgart.
- Lloyd-Roberts, G. C. & Shaw, N. E. (1969) The prevention of pseudarthrosis in congenital kyphosis of the tibia. *J. Bone Jt Surg.* **51-B**, 100-105.
- McFarland, B. (1951) Pseudarthrosis of the tibia in Childhood. *J. Bone Jt Surg.* **33-B**, 36-46.
- Madsen, E. T. (1956) Congenital angulations and fractures of the extremities. *Acta orthop. scand.* **25**, 242-280.
- Moore, J. R. (1957) Congenital pseudarthrosis of the tibia. *Instructional Course Lectures*, vol. XIV, pp. 222-237. (American Academy of Orthopaedic Surgeons.) J. W. Edwards, Ann Arbor, Michigan.

- Nicoll, E. A. (1969) Infantile pseudarthrosis of the tibia. *J. Bone Jt Surg.* **51-B**, 589–592.
- Païs, C. (1953) La pseudarthrose congénitale du tibia. *Rev. Chir. orthop.* **39**, 701–707.
- Sofield, H. A. (1971) Congenital pseudarthrosis of the tibia. *Clin. Orthop.* **76**, 33–42.
- Stelling, F. (1971) Personal communication.
- Van Nes, C. P. (1966) Congenital pseudarthrosis of the leg. *J. Bone Jt Surg.* **48-A**, 1467–1483.

Correspondence to:

K. Skou Andersen
Dept. of Orthopaedic Surgery T2
Gentofte Hospital
2900 Hellerup
Denmark