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CONGENITAL ANGULATION OF THE LOWER LEG

Crus Curvatum Congenitum

By

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Congenital angulation of the lower limbs makes up a rare and ill-defined syndrome which may be the precursor of so-called "congenital" pseudarthrosis of the tibia.

Since *Camurati*, in 1930, published his report on these conditions, there has been an increasing number of reports, but the designations have differed, *e.g.* intrauterine fracture, crus varum congenitum, congenital bent tibia, congenital angulation of tibia, the kyphoscoliotic tibia, and congenital pseudarthrosis of the tibia.

Since the aetiology of these congenital angulations is unknown, the syndrome is only a clinical concept maintained chiefly because of the risk of pseudarthrosis.

Froelich (1910) stated that this pseudarthrosis was not always congenital, but often resulted from fracture of the lower leg which had been bowed from birth. According to *Kosic* (1941) pseudarthrosis developed only in anterolaterally convex legs; his term referring to the entire condition was crus varum congenitum. Other directions of angulation are more uncommon, but may also give rise to pseudarthrosis (*Camurati* 1930).

Büttner & Eysholdt (1950) do not feel that it is possible to decide with certainty whether all congenital angulations of the lower leg make up a pathogenetic entity. According to *Lindemann* (1961) there is not, however, much doubt that intrauterine fracture, crus varum congenitum, and the congenital pseudarthroses of the tibia are pathogenetically closely related.

At our present stage of knowledge it seems reasonable to collect all

cases of congenital angulations of the lower leg under one common designation.

Lindemann, in 1961, suggested the term crus curvatum congenitum (c. c. c.), meaning all congenital angular deformities of the lower leg in so far as the more systemic diseases of bone could be ruled out. This term will be used here.

Interpreted in this way, c. c. c. makes up a characteristic syndrome. It is most often unilateral, although bilateral cases have been reported, *e.g.* by *Camurati* (1930) and *Ducroquet & Cottard* (1939). The angulation is present at birth or appears within the first days of life. It nearly always affects the distal half of the lower leg, the most characteristic site being at the junction of the middle and distal third. Radiography often shows thickening of the cortex on the concave aspect of the bone on a level with the apex of the angulation. At times, there are cystic changes or fracture of the bone at the site of the angulation. While these changes are invariably present in the tibia, the fibula may be radiologically normal. As a rule, however, it shows the same changes as the tibia. In certain cases the fibula may be absent, and these cases have sometimes been classified in a separate group (*Farmer* 1960). Since they are accompanied, in the majority of cases, by angulation of the tibia, and since pseudarthrosis has been described in this group too (*Aitken* 1959), it seems reasonable to assign them to c. c.c.

Previously, most interest was devoted to the aetiology and treatment of the congenital pseudarthroses of the tibia. We felt that it must be of interest to elucidate the prognosis of c. c. c. with a view to spontaneous correction of the angulation and to the risk of pseudarthrosis.

MATERIAL

During the period 1934–1966 a total of 40 cases of c.c.c. were treated in the Orthopaedic Hospital, Copenhagen.

Twenty patients were seen in the hospital for the first time within the first 8 weeks of life, and only 11 patients were over one year of age when first seen. In all cases the angulation had been present at birth.

The follow-up period ranges from 6 months to 32 years, average 13.5 years. When the analysis was completed about new year 1966–1967 6 patients were under 5 years of age, while 8 were from 5 to 10 years, 7 from 11 to 15 years, and 19 were over 15 years of age.

The sex distribution is given in Table 1. There was only one bilateral case, and of the remaining 39 cases 20 were right-sided and 19 left-sided.

To divide the cases of c.c.c. into sub-groups as uniform as possible from the

Table 1. Distribution of Crus curvatum congenitum by sex and type of angulation

Sex	Type of angulation		
	Anterior *with* associated congenital deformities	Anterior *without* associated congenital deformities	Posterior
Female	5	7	6
Male	16	3	3
Total	21	10	9

prognostic point of view, we felt that it would be expedient, as done by *Heymann* (1949), to distinguish between cases with anterior angulation without associated congenital deformities, cases with anterior angulation with associated congenital deformities, and cases with posterior angulation.

The angulation of the antecurved legs was most often antero-lateral convex or purely anteroconvex. In the group without other associated deformities all 10 cases were antero-laterally convex. In the group with associated deformities the angulation was in 4 cases antero-laterally convex, in 10 cases purely anteroconvex. In 4 cases the angulation was antero-medially convex.

Of the cases with posterior angulation 8 were postero-medially convex and one posteroconvex.

Patients with c.c.c. often have other congenital deformities. Among the 40 cases of c.c.c. there were 29 with hypoplasia of the ipsilateral lower limb. 19 had congenital deformity of the ipsilateral foot, 6 had dimpling of the skin, and 8 had a congenital constricture at the site of the angulation. In 12 cases there was aplasia of the ipsilateral fibula and in 3 syndactyly of fingers.

Out of the 31 cases with anterior angulation 6 had a congenital constricture and 5 dimples in the skin. In 12 there was aplasia of the fibula. In 8 of these latter cases there was also absence of either the 5th or the 4th ray in the foot. 6 patients had congenital equino-valgus foot, one congenital equino-varus. In 2 cases there was such pronounced hypoplasia of the foot that it presented merely as a lump without definite ossification.

Out of the 9 cases with posterior angulation 2 had associated congenital constrictures, 1 had dimpling of the skin, 1 had calcaneo-valgus, and 1 pronounced hypoplasia of the foot without ossification.

Two factors are of essential importance to the functional ability obtainable in c.c.c. One is the degree of hypoplasia and the attendant shortening and the other is the occurrence of fractures and their tendency to unite.

Hypoplasia is present in varying degree in c.c.c. Among 17 cases seen at birth or shortly after 7 had mild hypoplasia with up to 1 cm shortening of the lower leg. 6 had 2–3 cm shortening and 4 more than 4 cm shortening.

Hypoplasia occurred in legs with anterior as well as posterior angulation. Out of 31 patients with anterior angulation 20 exhibited considerable hypoplasia and shortening. And among these 20 cases the hypoplasia was so marked in 8 that in combination with a deformed foot it made up an indication for amputation. In 3 cases epiphysiodesis of the contralateral leg had to be performed.

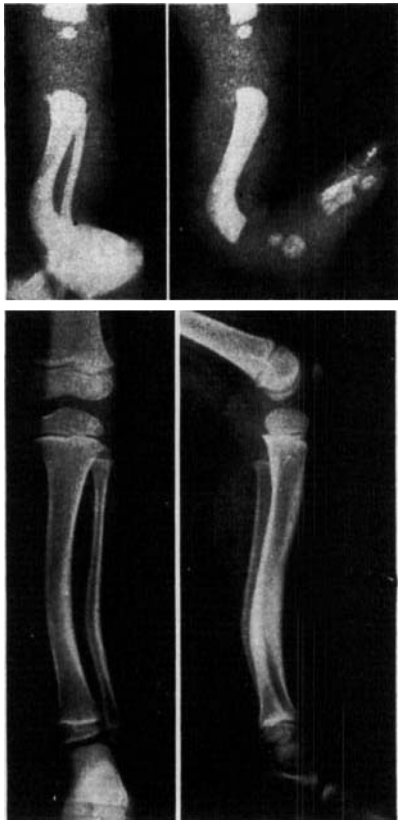

Figure 1 a + b. OH 983/62. The spontaneous correction in a Crus curvatum congenitum with posterior angulation, a) at the beginning of follow-up, b) 4 years later.

Out of 9 patients with posterior angulation 8 had hypoplasia of major degree. Five required epiphysiodesis and one patient was fitted with a Syme prosthesis.

The tendency for the angulation to become spontaneously corrected was assessed by measuring the angulation in X-ray films from the beginning and from the end of the follow-up period. In this connection, it should be mentioned that the material is derived from a period of more than 30 years, and therefore the therapeutic principles in respect to correcting the angulation have not been uniform throughout. A number of cases had been treated by corrective casts, while others had had osteotomy or therapeutic fracturing.

In the 9 patients with posterior angulation primary examination showed angulation in the sagittal plane ranging from 105° to 170°, average 150°. In 2 cases the bone was fractured at an early stage of the follow-up period in an attempt to correct the angulation. The others have been followed for 4–19 years. At the end of the follow-up period 6 of these patients showed satisfactory appearances (less than 10° angulation). In one case the angulation exceeded 10°, but this patient had been followed for only 4 years during which spontaneous correction from 105° to 160° had occurred (Figure 1).

In the 31 cases with anterior angulation the angulation was 90°-170° at the primary examination. After a short follow-up period osteotomy was done in 3 cases and fracturing of the lower leg in one in an attempt of correction. In 2 cases the follow-up period had been only 6 months, so that it is not possible to assess the tendency to correction. In the remaining 8 cases the follow-up period ranged from 4–22 years. Four obtained a satisfactory appearance with an angulation of less than 10°. In 4 cases the angulation exceeded 10° at the end of the follow-up period.

The most serious complication of c.c.c. is fracture and pseudarthrosis. The fracture may be present at birth or it may occur within the first years of life. Our material includes 3 cases in which fracture was diagnosed immediately after birth. In 3 fracture occurred within the first year of life and in 6 within the 2nd–5th year of life. In another 3 cases therapeutic fracturing was done a few weeks after birth.

This gives a total of 15 fractures among the 40 cases of c.c.c. Out of these fractures 11 developed into pseudarthroses.

Out of the 3 fractures present at birth one developed into pseudarthrosis. Out of the 9 fractures occurring after birth all ended as pseudarthroses. One of the 3 therapeutic fractures developed into pseudorthrosis (Table 2).

Table 2. Cases with fracture and pseudarthrosis by type of curvation.

	Type of angulation		
	Anterior *with* associated congenital deformities	Anterior *without* associated congenital deformities	Posterior
Total number of cases	21	10	9
Number of cases with fracture	3	10	2
Number of cases with pseuarthrosis	1	10	0

Corrective osteotomy was carried out in 4 cases—three with anterior angulation with associated deformities and one with posterior angulation. In all 4 cases the osteotomy healed within the normal time.

Some authors, *e.g. Moore* (1949) and *Badgley* (1952), feel that on the basis of the X-ray films they can pick out the prognostically unfavourable types, claiming that thickening of the cortex on the concave aspect of the angulation, with consequent partial or complete obliteration of the medullary canal, predisposes to pseudarthrosis. However, *Exner* (1952) and *Matzen* (1955) are sceptical of the pathogenetic significance of this X-ray finding.

Thickening of the cortex on the concave aspect of the bone was present in 26 of our cases. Partial obliteration of the medullary canal was found in 13, 8 of whom had complete obliteration. Only 3 of these 8 patients developed pseudarthrosis.

Cystic changes at the site of the angulation have often been reported. *Compere* (1936) published a case in which he believed the pseudarthrosis to be due to localized osteitis fibrosa. *Green & Rudo* (1943), who also found cystic changes,

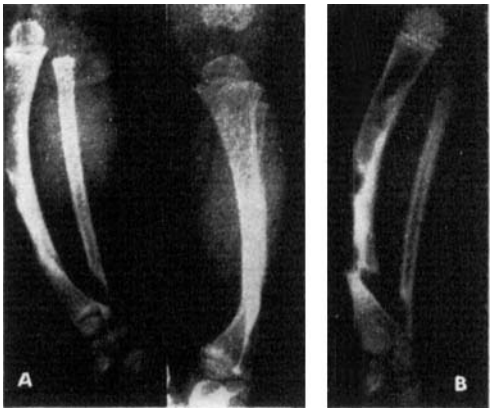

Figure 2. Case of Crus curvatum congenitum with anterior angulation and development of fracture in a cystic tibia within 4 months.

claimed that these were intraosseous neurofibromas. However, this claim has not been confirmed (*Boyd* 1958). In 4 of our cases the first X-rays showed cysts in the tibia. All later developed pseudarthrosis (Figure 2).

The angulation of the lower leg in the sagittal plane ranged from 90° to 170°. The magnitude of the angulation, measured as the numerical degree, could not be correlated to the risk of developing pseudarthrosis. Among 9 cases with an angulation below 140° no pseudarthroses occurred. Among 21 cases with an angulation of between 140° and 180° there were 4 cases of pseudarthrosis.

Pathological studies of congenital tibial pseudarthroses have been described by *Camurati* (1930). His studies as well as numerous subsequent ones have shown that histologically the congenital pseudarthroses do not differ from other pseudarthroses. *Green & Rudo* (1943) were the first to study a biopsy from a c.c.c. before fracture and pseudarthrosis occurred. This examination also failed to demonstrate specific histological features. In our material biopsies from the site of pseudarthrosis were removed for microscopic examination in 4 cases subjected to operation for the pseudarthrosis, but no notable findings were made. In 4 cases microradiographic study of the biopsies was done, and in 2 cases double labelling with tetracycline was done before removing the biopsies. These investigations did not reveal any special features of the bony structure, but in one case it seemed as if the rate of deposition of bone occurred faster than might be expected according to the patient's age (Figure 3).

DISCUSSION

The aetiology of c. c. c. is unknown. However, there is presumably a relationship between congenital pseudarthrosis of the tibia and von Recklinghausen's neurofibromatosis as demonstrated by int. al. *Barber* (1939) and *Ducroquet & Cottard* (1939). One case of neurofibromatosis occurred in our series, and this patient developed pseudarthrosis.

On the basis of the numerous theories advanced Guilleminet (1953) suggested a working hypothesis presupposing that the congenital

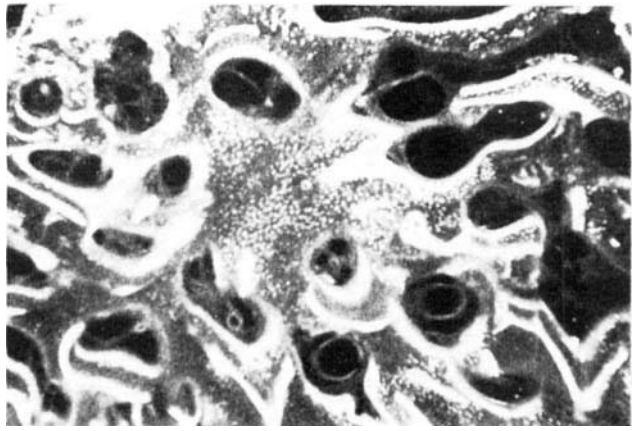

Figure 3. Section through the bone near the pseudarthrosis labelled with Tetracycline. OH 4276/60.

pseudarthroses of the tibia have invariably been preceded by segmental dysplasia or dystrophy. This dysplasia may be due to systemic causes, such as mesenchymal damage, neurofibromatosis, endocrine disturbances, or to local causes, such as a reduced blood flow through the part or intrauterine mechanical factors. This hypothesis comprises all known aetiological factors and seems to be at present the most acceptable one. Whether it can also embrace all cases of c. c. c. is not clear.

Clinically a distinction may be made between several types of c. c. c. Like *Badgley* (1952), we found cases with anterior angulation to be most common. In *Badgley's* series as well as in ours they constituted about 75 per cent. Cases with posterior angulation made up about 25 per cent. Both types are equally often associated with other congenital deformities, viz. in about one-third.

From the prognostic point of view the three groups: anterior angulation without associated congenital deformities, anterior angulation with associated congenital deformities, and posterior angulation are fairly sharply separated. Fractures ending in pseudarthrosis were by far most common in the group with anterior angulation without associated deformities. In Badgley's series this occurred in 1 out of 4 cases, and in our series in all 10 cases.

In the group with anterior angulation with other congenital deformities *Badgley* found 1 pseudarthrosis among 10, and we had 1 among 21.

In the group with posterior angulation *Badgley* had no pseudar-

throsis among 5 cases, and in our series too there was none among 9 cases.

Several workers have published cases in which the posterior angulation has become spontaneously corrected. *Heymann* (1949) as well as *Lindemann* (1961) stated that cases with posterior angulation differ clearly from the cases with anterior angulation, partly in respect to spontaneous correction and partly in respect to the risk of pseudarthrosis. In our material some spontaneous correction occurred, not only in the group with posterior, but also in that with anterior angulation. Probably the correction was more marked among the patients with posterior angulation.

Like several others before us, we have found no reason to presume that radiological findings such as thickening of the cortex and partial obliteration of the medullary canal, are of any independent prognostic importance. On the other hand, we have found cysts to be a serious prognostic sign, predisposing to pseudarthrosis. This observation too is in accordance with those of previous authors.

In further accordance with previous findings (int. al. *Heymann* 1949, *Thyge Madsen* 1956) we found dimpling of the skin and a congenital constricture at the site of the angulation to occur in anterior as well as posterior angulations. We found no evidence for assuming that c. c. c. with a congenital constricture or c. c. c. with dimpling of the skin make up separate groups in respect to the risk of developing pseudarthrosis.

Cases showing aplasia of the fibula have been set part by some workers as a separate group differing from c. c. c., presumably wrongly. In our series no case of pseudarthrosis occurred in the group having c. c. c. as well as aplasia of the fibula.

CONCLUSION

Cases of c. c. c. with anterior angulation without associated congenital deformities very often develop pseudarthrosis. The therapeutic principle must be to avoid fracture. No weight-bearing should be allowed, and any kind of osteotomy is contra-indicated. Repeated radiographic examinations should be done to detect cyst formation, if any, as this must be interpreted as a serious prognostic sign.

Cases of c. c. c. with anterior angulation and other congenital deformities seldom end in pseudarthrosis. In these cases deformities of the feet and hypoplasia call for treatment. Presumably the leg should

not be braced, as in this group the hypoplasia is frequently a problem. Correcting osteotomy should only be performed on strict indications.

Cases of c.c.c. with posterior angulation very seldom develop pseudarthrosis. In these cases treatment is often required for foot deformities and hypoplasia. Presumably, bracing of the leg is unnecessary. Osteotomy is rarely indicated. The prognosis is on the whole favourable.

SUMMARY

From 1934 to 1966 a total of 40 cases of congenital angulation of the lower leg, here called crus curvatum congenitum (c. c. c.), were treated in the Orthopaedic Hospital, Copenhagen.

The follow-up period ranges from 6 months to 32 years.

C. c. c. is sometimes associated with other congenital deformities. About three-quarters of all the cases were anterior and one-quarter posterior angulations. Associated congenital deformities were equally common in these two groups. Hypoplasia was present in varying degrees.

Fractures of the leg occurred in 15 cases. Out of 10 cases of c. c. c. with anterior angulation without associated deformities all had fractures with development of pseudarthrosis.

Assessment of the X-ray films appears to show cysts to be a serious prognostic sign in respect to pseudarthrosis, all our 4 cases with cysts developing pseudarthrosis.

An attempt to assess the tendency to spontaneous correction of the angulation showed that possibly this is most marked among the cases of posterior angulation.

The principle of treatment must be to avoid fracture as far as at all possible. In cases with associated congenital deformities treatment of foot deformities and hypoplasia is often required.

RESUME

De 1934 à 1966, on a traité à l'Hôpital Orthopédique de Copenhague 40 cas d'incurvation congénitale de la jambe (Crus curvatum congenitum = C. c. c).

La durée des observations varient entre six mois et 32 ans.

La C. c. c. est souvent accompagnée d'autres déformités congénitales. 3/4 des malades étaient des cas d'antéincurvation et 1/4 de rétroincurvation. La fréquence d'autres déformités congénitales est la même pour les deux groupes. De l'hypoplasie a été observée à des degrés variables.

Il y eut en tout 15 cas de fracture de la jambe. Dans les 10 cas d'antéincurvation sans autre déformité, il y eut dans tous les cas fracture et formation de pseudarthrose.

Lorsqu'il s'agit de porter une appréciation sur les radiographies, le kyste semble être un signe pronostique grave par rapport à la pseudarthrose.

On a essayé de juger de la tendance à une correction spontanée de l'incurvation. Elle est peut-être surtout marquée dans les cas de rétroincurvation. Le principe du traitement doit être d'éviter les fractures dans toute la mesure du possible dans les cas présentant d'autres déformités, le plus souvent des déformités du pied et de l'hypoplasie qui exigent un traitement.

ZUSAMMENFASSUNG

Von 1934 bis 1966 sind am Ortopädischen Krankenhaus in Kopenhagen 40 Fälle von Crus curvatum congenitum (C. c. c.) behandelt worden.

Die Beobachtungszeit wechselt von 1/2 bis 32 Jahren. C. c. c. is zeitweise von anderen angeborenen Verbildungen begleitet. Ungefähr 3/4 aller Fälle war antekurviert und 1/4 retrokurviert. Begleitende Deformiteten fanden sich gleich häufig in beiden Gruppen. Hypoplasie kam in wechselndem Grade vor.

Im ganzen wurden 15 Fälle von Unterschenkelbruch gesehen. Bei 10 Fällen von C. c. c. mit Antekurvation ohne begleitenden Verbildungen kam es in allen Fällen zum Bruch und zur Pseudarthrosebildung.

Bei der Betrachtung der Röntgenbilder scheinen Cysten hinsichtlich Pseudarthrosebildung ein ernshaftes prognostisches Zeichen darzustellen.

Die Neigung zur Spontankorrektur der Krümmung hat man abzuschätzen versucht und sie war möglicherweise am meisten bei den Fällen mit Retrokurvation ausgesprochen.

Das Prinzip der Behandlung muss es sein, soweit als möglich Brüche zu vermeiden. In Fällen von begleitenden Deformiteten, sind es zumeist Fussdeformiteten und Hypoplasie, die Behandlung erfordern.

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