

KURT STENPORT, HÄLSINGBORG:

CONGENITAL PSEUDARTHROSIS OF THE LEG

In the last 20 years congenital pseudarthrosis of the leg has been observed in altogether 7 cases under treatment in the Orthopedic Hospital of Stockholm and the Orthopedic Hospital of Hälsingborg. In 4 of these cases the pseudarthrosis involved both the tibia and the fibula, in 3 cases the tibia alone. The location of the pseudarthrosis is nearly always at the border between the middle and the lower third of the leg. These cases often involve actual defects in the tibia and fibula. In the first years of life the fragments of the bones are often seen to be in end-to-end contact, separated by a pseudarthrosis, but gradually the fragments undergo concentric subperiosteal absorption. When all contact between the fragments is gone, the typical roentgenographic finding is that both ends of the pseudarthrosis become pointed and lose the normal bone structure. The etiology has been the subject of a great deal of discussion. At present the prevailing explanation is a foetal impairment of the mesoderm due to a wanting blood supply of this particular area.

Briefly these cases may be described as follows:

CASE 1. Girl, normally developed, born 1926. At the age of 5 months a deformity of the right leg was observed. The child had not been exposed to any traumatic injury. The deformity was corrected and the leg was put in plaster cast in 1927, and the child wore a bandage until 1932.

Roentgenography showed pseudarthrosis of the tibia alone with a diastasis of 5 cm. In 1932—*i.e.*, when the child was 6 years old—she was admitted to the Orthopedic Hospital, Stock-

holm, and submitted to operation for the pseudarthrosis, with transplantation of a bridge taken from the other tibia. From the lower fragment of the tibia a small piece of tissue was removed at the same time for microscopic examination. The

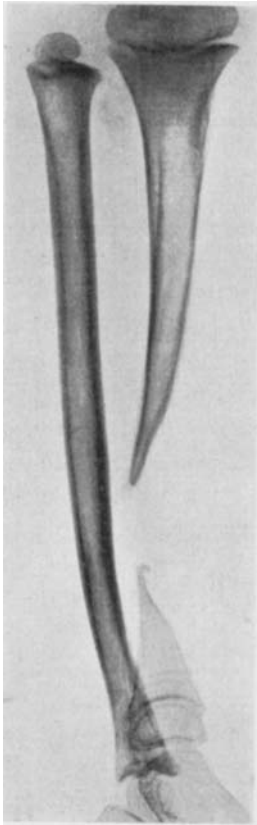


Fig. 1.



Fig. 2.

cord connecting the two tibial fragments was found to consist of collagenic connective tissue, and the little bit of bone consisted of spongiöse osseous tissue. At the border between these two tissues, the bone was undergoing destruction through halisteresis and lacunar erosion. Small islands of the so-called osteoplasts were seen here and there in the connective tissue.

The marrow spaces of the spongiöse bone contained some fibrous bone-marrow, poor in cells.

Already within a few months after the operation the lower part of the tibial bridge was seen to undergo absorption, and as this process progressed, pseudarthrosis developed anew. A little over one half of the tibial bridge healed in completely, however, after the operation the whole extremity was put in plaster; later on the girl was given an unjointed foot-casing which she is wearing yet.

In 1936 she was advised to submit to a new operation in about a couple of years. As the first operation had succeeded

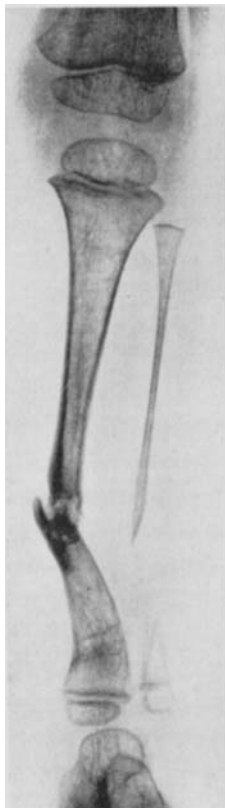


Fig. 3.

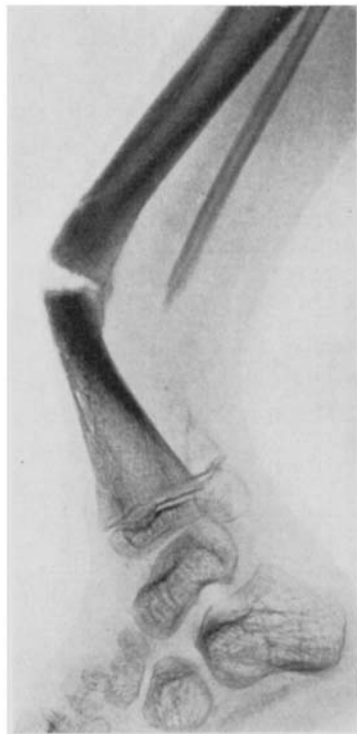


Fig. 4.

halfways, naturally a new attempt has to be made. Now the diastasis is only 2 cm.

CASE 2. Boy, born 1925. Defect of the right tibia and fibula. At the age of 1 year a deformity of the right leg was noticed, and the child was operated on in the local hospital. How the operation was performed I have not been able to find out. The leg was kept in plaster till 1928, when the child was admitted to the Orthopedic Hospital, Stockholm, where he was given a leather bandage for the foot and leg. At that time there was a distinct pseudarthrosis (Fig. 3).

In 1932 his leg was so firm and straight that he was able to walk without any bandage. On December 20, 1932, he met with a traumatic injury which gave a marked deformity of the leg, and he was unable to put any weight on it (Fig. 4). He was then readmitted to the Orthopedic Hospital, Stockholm. In January 1933 an operation was performed, with transplantation of a tibial bridge from the other leg—without favourable result (Fig. 5). In 1935 the pseudarthrosis was repaired again, this time with a bony bridge taken from the upper fragment of the tibia and moved down over the pseudarthrosis. He has been using an unjointed foot-casing since, but the pseudarthrosis has failed to heal.

CASE 3. Boy, born 1920. Pseudarthrosis of the tibia alone. Deformity of the leg since birth. No treatment in childhood. In 1934 the boy was admitted to the Orthopedic Hospital, Hälsingborg, where an amputation was advised, and carried out at once. The boy is very satisfied with his artificial leg, and he is able to walk and run without any inconvenience.

CASE 4. Boy, born 1921. Left leg crooked since birth. At the age of 2 years he was operated on in the local hospital, but the pseudarthrosis did not heal. In 1925 he was admitted to the Orthopedic Hospital Hälsingborg, where the Lane plate and screws, which had been applied at the operation 2 years before, were now removed, and the boy was given a leather bandage.

A new operation was performed in 1927, at the age of 6 years: the tibial fragments were adjusted and fixed with end-to-end sutures of catgut, and a bridge of periosteum from the upper part of the tibia was fixed over the pseudarthrosis. Plaster cast; later on, unjointed foot-casing. Reexamination in 1933 showed persistent pseudarthrosis, with shortening of the leg amounting to about 4 cm. In 1934 this lesion was complicated with pulmonary tuberculosis and tuberculosis of the knee-joint, requiring a low femoral amputation.

CASE 5. Man, born 1910. The mother noticed a deformity of his right leg immediately after birth. Still, at the age of 1 year, the child was able to walk on his crooked leg. In his second year of life he suffered a slight blow on the leg and was unable since to support himself on it. He was admitted to the local hospital, and operated on with Lane's plate and screws. The pseudarthrosis failed to heal. In 1913 he entered the Orthopedic Hospital, Hälsingborg, where Lane's plate was removed. The ends of the fragments were snipped off so that the marrow



Fig. 5.



Fig. 6.

cavity was laid open. Then the fibula was divided lengthwise and this bridge of bone and periosteum was inserted into the marrow cavities of the tibial fragments. One year later the pseudarthrosis was persisting yet. Then a new operation was performed: The pseudarthrosis tissue between the bone fragments was removed and the ends of the fragments were trimmed and fixated end-to-end by means of a silver wire. Then a band of bone and periosteum was resected by chisel from the upper tibial diaphysis and fixed over the pseudarthrosis (Fig. 6).

The next two years the patient was wearing a bandage. In 1917 it says in his case record: Clinical consolidation, but not roentgenographical. Hence he is told to keep wearing the bandage. In 1918, when the bandage became too small, and he quit using it, the leg began to bend again. Reexamination showed an angular deformity of nearly 45° , with the apex pointing forwards. He was given a new bandage. The pseudarthrosis now became firm again, but the position was poor for the foot. In 1927 a bone wedge was chiseled out of the front of the bend, and the angular position was straightened out in plaster. Since then, the patient has been wearing an unjointed foot-casing. After this, there has been no healing of the bone—on the contrary, the result is a loose pseudarthrosis with faulty position.

As late as last year I advised this patient to have his leg amputated, but he refused.

CASE 6. Boy, born 1931. When the child was a couple of months old his mother noticed a deformity (faulty position) of his right leg. The leg was put in plaster in the local hospital, and when the child was 1 year old he was given a leather bandage from the Orthopedic Hospital, Hälsingborg. The pseudarthrosis was located at an unusually high level on the tibia (between the middle and the upper thirds) and at a very low level on the fibula. In 1934—*i.e.*, when the boy was 3 years old—an operation was performed: the pseudarthrosis was loosened and the ends of the fragments were trimmed; then extension was applied in plaster in order to eliminate the shortening. The leg did not heal, and the boy has used an unjointed leg bandage

since. A new operation is to be performed when he is 10 years old.

CASE 7. Girl, born 1927. The mother noticed a faulty position of the left leg when the child began to stand on her feet.



Fig. 7.



Fig. 8.

She was limping when she began to walk, at the age of $1\frac{1}{2}$ years. In 1929 the mother brought the child to the Orthopedic Hospital Hälsingborg, where a congenital pseudarthrosis of the tibia was ascertained at the border between the middle and lower thirds of this bone, with abnormal mobility and a slight forward curvature. The fibula was not involved. The displacement was corrected under narcosis, and the leg was put in plaster. In 1930 examination showed no consolidation. Then the pseudarthrosis was treated operatively, at the age of 3 years: A bone-periosteum bridge, about 4 cm. in length, was chiseled out from the upper tibial fragment and moved down over the pseudarthrosis, after this had been trimmed; and a good fixation was established with catgut. After five months of continual

and roentgenographically satisfactory callus formation, the plaster cast was cut. Since then the girl was playing and running around with other children for about two months. Then she met with a slight injury, resulting in a new fracture. Then we tried again with a plaster cast involving the whole extremity, but the fracture at the site of the old pseudarthrosis would not heal, and again there developed a pseudarthrosis. Now the child was again allowed to move about with an unjointed foot-casing until 1936, when she returned again to the Orthopedic Hospital, Hälsingborg.

At this time the bone showed a markedly angular deformity, pointing forwards, without any stability in the pseudarthrosis, and the lower tibial fragment, which could be made out as pointed, threatened to break through the skin (Fig. 8).

At that time this girl was 9 years old—and something had to be done: amputation or operation for pseudarthrosis? The latter way was chosen.

On August 5, 1936: *Seance 1*: A bridge of pure bone, 12 cm. in length, and measuring 6 mm. at its greatest height, was introduced edgewise under the periosteum of the tibia of the healthy leg. 6 weeks later, on September 16th: *Seance 2*: Operation for pseudarthrosis with transplantation of the tibial bridge + pure bone + new bone + overgrowing periosteum. I selected this combined method because at the present I am rather skeptical about the efficiency of pure bone and new bone alone in operations for pseudarthrosis—especially as Orell has himself reported a case of congenital tibial pseudarthrosis on whom operations had been performed no less than five times within a period of 1933—35, with pure bone and new bone, and in which the pure bone is discharged in part, and in which the pseudarthrosis had not been healed as late as the summer 1936. This child was operated on at the age between 6 months and 3 years. Theoretically, however, a good deal of importance may be attached to the fact that even at the time of the operation the autoplasmic tibial bridge was covered by a good deal of new bone—that is, soft osseous connective tissue (soft callus; about 6 weeks old), promoting thus the course of healing.

Operation: Longitudinal section over the area where the new bone is to be implanted. With Albee saw removal of a splinter of the tibia, measuring about 0.5 cm. in width and 8 cm. in length, comprising both the implanted pure bone and the

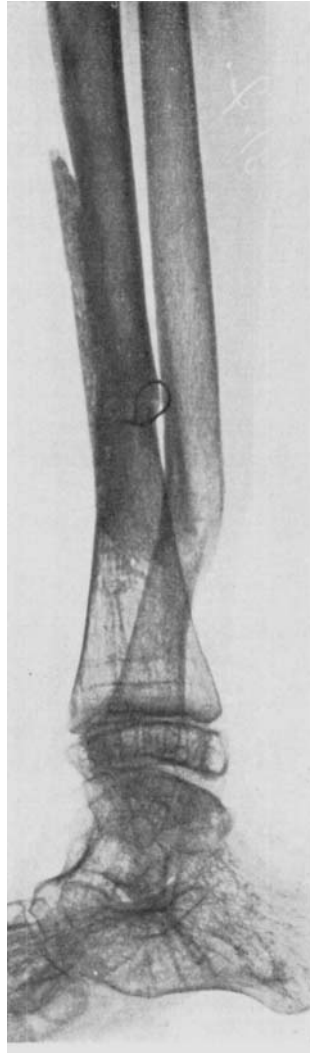


Fig. 9.

production of new bone. Suitable amounts of new bone were found in the angles between the new bone and the tibia. A curved incision, about 15 cm. in length, was laid over the site of the pseudarthrosis. The pointed fragments of the pseudarthrosis were trimmed for a distance of about 2 cm., leaving a surface of fresh spongiöse bony tissue. Then the tip of the proximal stump was resected, leaving the marrow cavity open. The combined bridge was inserted well into this cavity and its location was fixed by the means of two coarse catgut sutures. Of course, the bone bridge did no longer have any direct function in this combination bridge as new bone was formed already, but its presence meant a good deal in avoidance of disturbances by contact. After this: Hip and thigh in plaster. Walking plaster cast after 3 months. About 5 months later, when the plaster cast was cut, the leg was firmly united, but a new leg plaster bandage was prescribed, and this the patient has used until June, this year—that is, about 9 months after the operation. Now she is walking about without any cast—and without any pain. Yet she has been given an unjointed foot-casing which she is instructed to keep on using for at least one year, and perhaps longer.

Comments.

Let us look for a minute on these 7 cases in which so many operative methods have been employed with particularly unfavourable results. In all the publications I have read so far, the authors have reported similar discouraging results. The most favourable statistics give healing of the bone in about 1 out of 10 cases. The autoplasmic bridges have yielded most of the successful results.

Then the question arises: What are we to do in cases of congenital tibial pseudarthrosis? Amputation or operation?

Everything considered, I would be inclined to suggest operative treatment. It is to be hoped that the technique will advance, and that the results will be better.

Then comes the other question: When is the operation to be performed?—Not before the age of 6 years. Good results before

that age are rare. On the other hand, in suitable cases it may be advisable to wait with the operation till puberty.

In the first years of childhood the bones are to be supported by means of a suitable bandage. If, later on, an operation gives no favourable result, there is time enough left to consider an amputation. For reasons concerning the question of an artificial limb, amputation is not to be performed before school

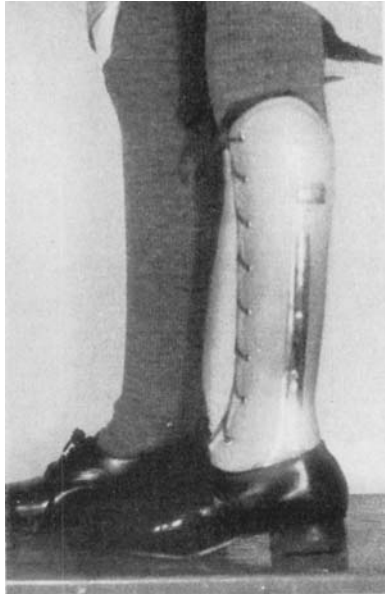


Fig. 10.

age. Whenever operative treatment it to be instituted it ought to be made so radical and thorough as possible. The pseudarthrotic ends are to be cut off to such an extent that fairly normal bone is laid bare in the operative wound. Preferably, the holes of the marrow cavities are to be opened, and the ends of the fragments are to be fixated end-to-end. If the fibula is intact, oblique osteotomy is to be performed at once. The transplantation ought to be made with an autoplasmic tibial bridge, and it is not difficult to obtain such a bridge if the child is not operated on before the age of 6—7 years.

It may be that the combined tibial bridges I have used in these cases constitute a good transplantation material. The after-treatment with plaster and unjointed foot-casing should be very long, at least 2 years.¹⁾ For, in cases of healed congenital pseudarthrosis, the risk of a new fracture is extremely great (Fig. 10).

DISCUSSION

F. Langenskiöld, Hälsingfors:

In pseudarthrosis involving only the tibia, the implantation of the upper part of the fibula into the upper fragment of the tibia as suggested by *Holm*, is to be recommended strongly.

On the other hand, in attempts to bring tibial pseudarthrosis to healing it is not essential—in contrast to *Böhler*—to make use of parts resected from the fibula.

Still, immobilization of the limb is of the greatest importance. Through this procedure the transplantation of the bone transplant ought to be firmly fixed with the fragments. For this purpose I have found it serviceable to make use of the *Parham* band which, I think, deserves more recognition than it has attained before.

Nils Silfverskiöld, Stockholm:

In the literature it has been pointed out several times that congenital pseudarthrosis of the leg is closely related to the congenital curvature of the legs. There are also transitional cases with curvature and partial bone defect, which does not involve the entire width of the bone (see Figs. 1 and 2).

In the curvature the compact substance of the bone is thickened and dense like ivory. On cutting into it with a chisel, one finds this bone to be unplastic, hard and brittle. Nor does the bone heal afterwards (poor vascularization), and pseudarthrosis develops. If the patient after such an operation con-

¹⁾ In the treatment of congenital tibial pseudarthrosis, like in all other forms of treatment of pseudarthrosis, the most important point in order to obtain a healing of the lesion may simply be a protracted fixation in the correct position.

sults another physician, the diagnosis may easily be: congenital pseudarthrosis, poorly operated on. Judging from the roentgenograms shown in the introductory lecture, at least one of those cases might fall under this category.

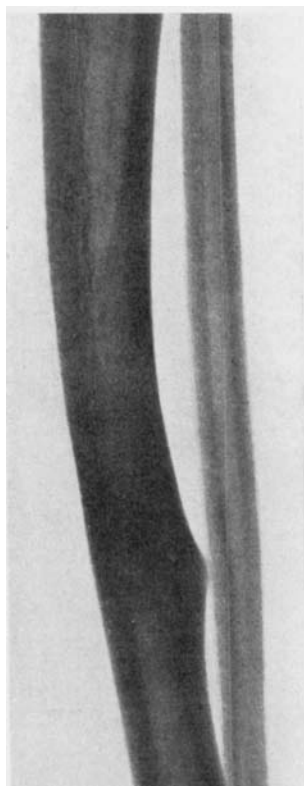


Fig. 1.

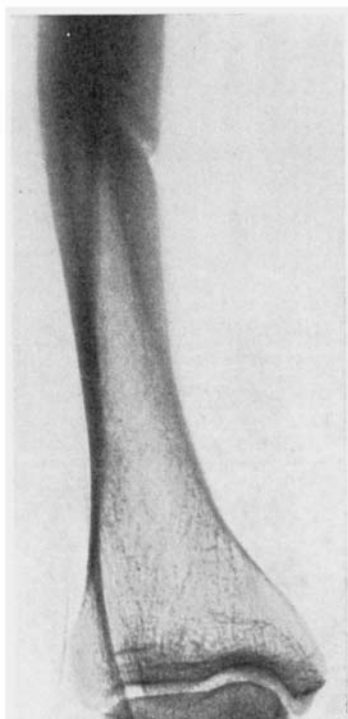


Fig. 2.

The congenital curvature of the leg, described, among others, by Professor *B. Valentin*, Hannover, should not be corrected operatively within the markedly sclerotic part of the curvature. I regret to say that my own experiences confirm this view.

As to the treatment of all this group of cases, I wish to recommend an operative correction in the spongiöse metaphysis distally to the pseudarthrosis, or the deformity. I have treated

two cases of this kind in whom healing of the congenital pseudarthrosis took place after wedge-formed metaphyseal osteotomy.

Other conditions being equal, healing of bone, as you know, proceeds always more rapidly in spongiöse bone tissue than in diaphyses. Besides, the width of the bone is greater in the metaphysis than in the diaphysis and thus the post-operative fixation



Fig. 3.

Fig. 4.

will be better here. Furthermore, spongiöse bone is more plastic for chiseling. That even the proximal pseudarthrosis subsequently might undergo bony consolidation is attributable, I think, to the fact that after the angular correction the weight is put on the pseudarthrosis perpendicularly—not obliquely as before.

I beg to be allowed to demonstrate one of these cases by means of some roentgenograms:

K. C., female, born 1922 (abstract of case record No. 6234). At her birth a swelling of the left leg was noticed. At the site of this swelling the bone was soft, and it was bent during the first year of life.

(A cousin of this patient has a congenital curvature (Figs.



Fig. 5.



Fig. 6.

1 and 2) in which, as mentioned above, pseudarthrosis developed at the site of osteotomy at the curvature). At the age of 11½ years the patient was submitted to an operation, performed by another surgeon. Since then the patient has been wearing a supporting bandage. Her state at that time is shown in Figs. 3 and 4.

On May 12, 1933: Cuneiform osteotomy in the distal part of the metaphyses of the tibia and fibula + correction + plaster cast. Cf. Figs. 5 and 6.

July 24, 1933: Plaster cast removed. Beginning consolidation of the pseudarthrosis, which keeps on healing. The patient is allowed to begin to walk.

June 14, 1937: *Reexamination*. The patient has now been walking about for 4 years without any difficulty—and without

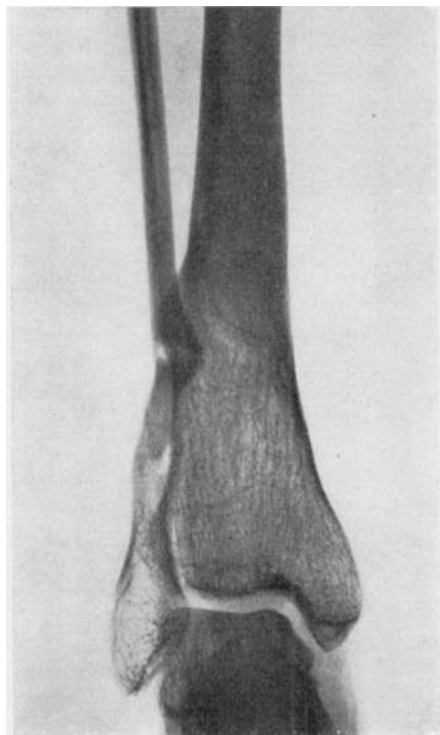


Fig. 7.

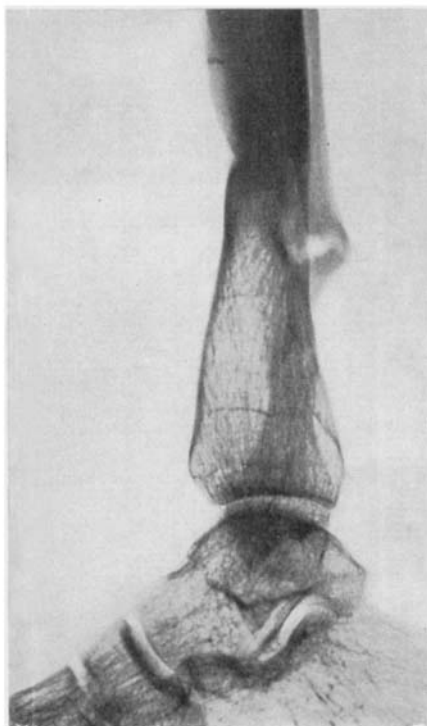


Fig. 8.

the support of any form of bandage. The bone is clinically well, firm and strong. Furthermore, even the foot has increased relatively more in length and width than the other foot, so that the two feet now look more alike than before the operation. Figs. 7 and 8 show roentgenographically good consolidation of the former pseudarthrosis; besides, the leg is straight.