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CONGENITAL ABSENCE OF THE FIBULA

By

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Next to absence of the radius, the commonest defect of the long bones of the extremities is congenital absence of the fibula. It is one of those malformations which have become very much more frequent during the past years, chiefly as the result of the Thalidomide accident (13).

We have earlier discussed the embryological, aethiological and patho-anatomical problems connected with defects of the long bones of the extremities in general, as well as the general principles of treatment (10). The present paper presents some experience gained in the treatment of six cases of congenital absence of the fibula.

The classical picture of fibular defect includes complete absence of the fibula, a bent tibia which is shorter than normal, a dimple in the skin on the anterior surface of the leg, talipes equinovalgus position of the foot, absence of one or more fibular rays of the foot, absence or fusion of one or more tarsal bones and a hypoplastic femur. The defect may also be partial, bilateral and/or combined with other defects (3, 4, 5, 6, 11).

Opinion regarding treatment is divided. Conservative measures (manipulation, casts) are generally resorted to first. The recent trend seems to be towards surgical intervention (2, 3, 4, 5, 7, 8, 11). Besides the deformity of the foot, the shortness of the extremity constitutes a difficult problem. Amputation is often said to be inevitable. *Perkins* follows a conservative line, carrying out amputation at the final stage of growth. *Coventry & Johnson* perform palliative operations (soft tissue correction and osteotomy of the tibia) from the second or third year onwards. *Thompson et al.* attribute the foot deformity and the bowing of the tibia to the presence of a taut fibrous band replacing the fibula which they believe should be cut in infancy. According to them, considerable correction of the tibial bowing and the valgus position is achieved in this way. *Thompson* (12) is of the opinion that the best

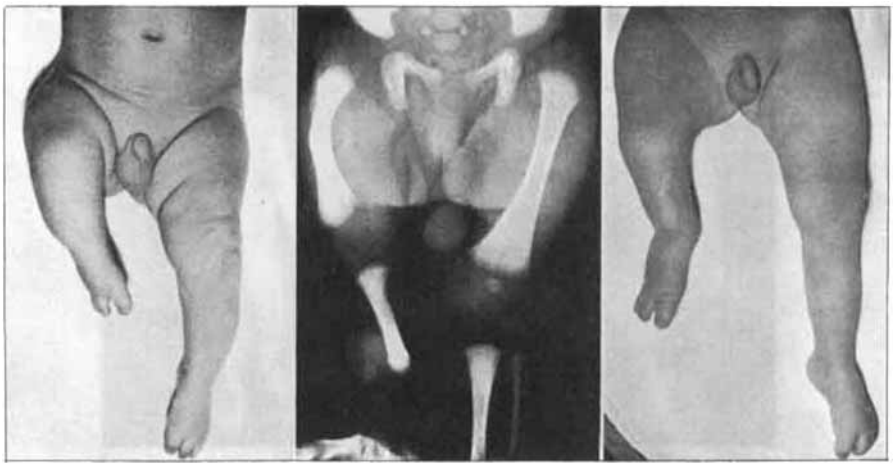

Fig. 1 A. *Fig. 1 B.* *Fig. 1 C.*

Case 1. Figures 1 A–B: before treatment. Fig. 1 C: at the age of 2 years.

result is obtained by combining this treatment with Syme's amputation carried out at the completion of growth. He stresses the fact that below-knee amputation during the growth period often leads to unsatisfactory development of the stump during the course of growth. *Aitken* recommends amputation during the early growth period, particularly if considerable shortening must be expected. *Farmer & Laurin* likewise carry out amputation in severe cases at an early age.

CASE REPORTS

Case 1. (5737/58), A.S. Male. Total absence of the right fibula (Figs. 1 A–C). He was brought for treatment at the age of 3 months. The right lower extremity was shorter than the left, the shortening comprising both the thigh and the leg. The knee was in pronounced flexion contracture, the foot being fixed in the talipes equino-valgus position. There was a dimple in the skin on the anterior surface of the leg. There were only four toes on each foot. The left lower extremity was otherwise normal. The radiogram showed a short and bowed right femur. The tibia was bent, the convexity being antero-medial.

At the age of 3 months osteotomy of the tibia was carried out, the proximal epiphysis of the left fibula with its epiphyseal cartilage being transferred to the site of the osteotomy. In a short time this transplant fused, however, showing no trace of the expected growth in length. At the age of 1½ years osteotomy was repeated and achillotenotomy carried out, since the malposition had increased. After this operation the position was satisfactory. Because of contraction of the knee joint a cuneiform resection of the femur was performed at the age of 3 years. At the age of 3½ years the motility of the knee joint is 140 to 110 degrees. The motility of the foot

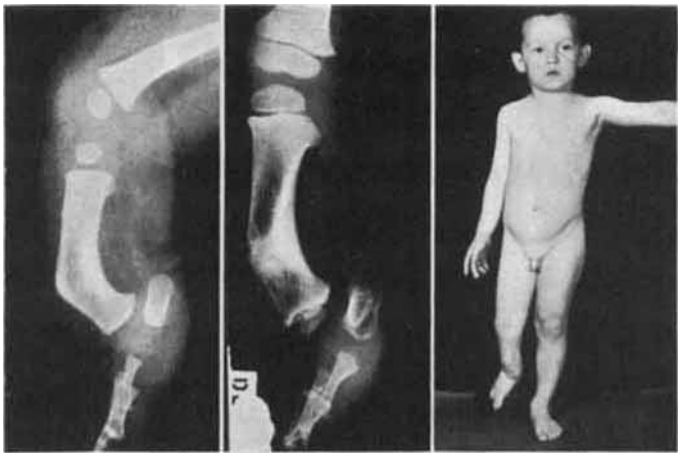

Fig. 2 A. *Fig. 2 B.* *Fig. 2 C.*

Case 2. Fig. 2 A: before treatment. Fig. 2 B–C: at the age of 4 years.

in the ankle joint is almost normal. The extremity is 12 cm. shorter than the left lower extremity. The patient walks well with the aid of an orthopaedic boot.

Case 2. (609/59), V.K. Male. Ttotal absence of the right fibula (Figs. 2 A–C). He was brought for treatment at the age of 6 months. The femurs were of equal length. The right tibia was short, thick, and convex in the antero-medial plane. The foot was in moderate valgus and slight equinus position. The patient was first treated with redressement and a brace but without result. At the age of 2 years and 9 months osteotomy of the tibia and achillotenotomy were carried out, with simultaneous excision of the fibular fibrous bands. At the age of 4 years and 3 months the right tibia is 9 cm. shorter than the left and moderate bowing remains in the tibia. The motility in the knee joint is normal. The patient walks well with an orthopaedic boot.

Case 3. (494/59), J.J. Male. Total absence of the right fibula (Figs. 3 A–D). He was brought for treatment at the age of 3½ months. The right femur was slightly shorter than the left. The tibia was short and convex in the antero-medial plane. There was moderate flexion contracture in the knee and the foot was in extreme equino-valgus position. Two fibular toes and metatarsal bones were missing. Osteotomy of the tibia was carried out. At the same time the third metatarsal bone was transferred to the site of the fibular malleolus. Further, achillotenotomy and excision of the taut fibular bands were carried out. Because of impairment of the position, osteotomy and achillotenotomy were repeated 1½ years later. The position was then satisfactory. Two years after the last operation the tibia is now quite straight, but it is barely two-thirds the length of the left tibia. The position of the foot is comparatively good. The boy, who is now almost 4 years old, walks well with the aid of an orthopaedic boot.

Case 4. (4677/59), S.H. Female. Total absence of the left fibula (Figs. 4 A–C). She was brought for treatment at the age of 4 days. The right lower extremity was entirely missing. The upper end of the left femur was dysplastic and in pronounced

Fig. 3 A. *Fig. 3 B.* *Fig. 3 C.* *Fig. 3 D.*

Case 3. Fig. 3 A–B: before treatment. Fig. 3 C–D: at the age of 2 years.

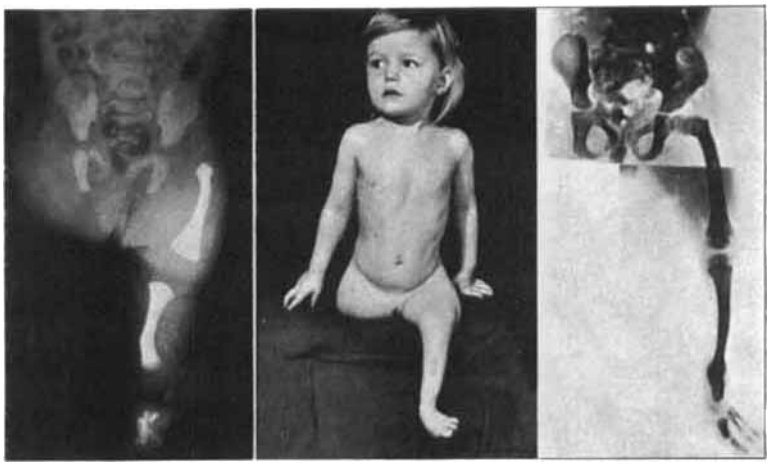

Fig. 4 A. *Fig. 4 B.* *Fig. 4 C.*

Case 4. Fig. 4 A: before treatment. Fig. 4 B–C: at the age of 3 years.

coxa-vara position. The tibia was convex in the antero-medial plane and the foot was in marked equino-valgus position. Three fibular metatarsal bones and toes were missing.

At the age of 2 weeks osteotomy of the tibia, achillotenotomy and excision of taut fibular bands were carried out. Follow-up at the age of 3 years and 3 months showed good function of the hip and knee. The tibia is straight. The foot is in slight equino-valgus position. Since the function of the left lower extremity is good, the patient will probably be able to walk with the aid of a prosthesis.

Fig. 5 A. *Fig. 5 B.* *Fig. 5 C.* *Fig. 5 D.*

Case 5. Fig. 5 A: before treatment. Fig. 5 B–D: at the age of 3 years.

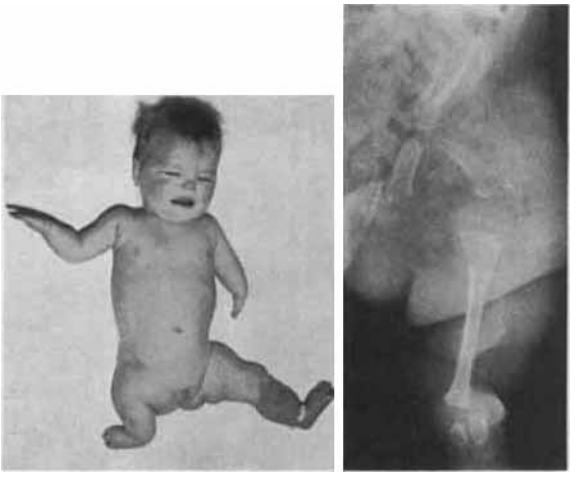

Fig. 6 A. *Fig. 6 B.*

Case 6. Fig. 6 A–B: before treatment.

Case 5. (5585/60), J.M. Male. Total absence of the right fibula. He was brought for treatment at the age of 10 months. The right femur was normal. The tibia was short and curved. The foot was in moderate valgus position. The left foot lacked the fifth toe and a metatarsal bone.

Osteotomy of the tibia was carried out at the age of 10 months. Two months later the operation was repeated, the position being corrected. Achillotenotomy and excision of a fibrous fibular band were also made. At the age of 3 years there is still moderate bowing of the tibia. The foot is of a slightly rocking chair-shape with

the calcaneus turned in the proximal direction. The patient walks very well with the aid of an orthopaedic boot.

Case 6. (1179/62), R.N. Female. Total absence of the right fibula.

She was brought for treatment at the age of ½ year. The right knee was almost at the level of the hip in rather pronounced flexion contracture with the foot in marked valgus position. Otherwise the foot was almost normal. The radiograph showed only the distal part of the femur. The tibia was straight. In addition, the patient had a subtotal radius defect on the left side and hypoplasia of the right radius. There were two fingers only on either hand. Since treatment was primarily directed at the femoral defect, treatment of the fibula has so far been conservative.

Clinical Picture.

All the cases described above are unilateral total defects of the fibula. 4 of the patients were boys and 2 girls. With one exception all defects were right-sided. In all but one of our cases the tibia was thicker and shorter than normal, and bowed with antero-medial convexity. At the the site of the tibial curvature there was a dimple in the skin on the anterior surface of the leg. The foot was in most cases in more or less marked equino-valgus position. The Achilles tendon, the fibular tendons and the fibrous bands were taut and prevented redressement of the ankle joint. In the fibular portion of the leg and foot there were often defects of the muscles and other soft tissues as well. In some cases the talus seemed to be missing, either entirely or partly. The fibular metatarsal bones and toes were mostly absent. The knee was often in flexion contracture which, however, was slighter and was less resistant than the contracture of the ankle. The femur was normal in only two out of the six cases. In two cases it was shorter than normal, in one there was marked coxa vara and in one case there was a subtotal defect of the femur.

Treatment.

In one case in which the fibular defect was combined with subtotal defect of the femur in the same extremity, the treatment has so far been conservative. In the remaining five cases operation, as shown in Fig. 7, has been carried out. The tibial surface was exposed subperiosteally through a longitudinal incision and a cuneiform osteotomy of the tibia performed. Achillotenotomy was further carried out in all cases, and in four cases the taut bands were severed at the site of the missing malleolus. We did not aim at complete correction of the equinus position. The tibia was fixed in position with a Kirschner nail introduced from under the foot into the tibia. Immobilization was achieved

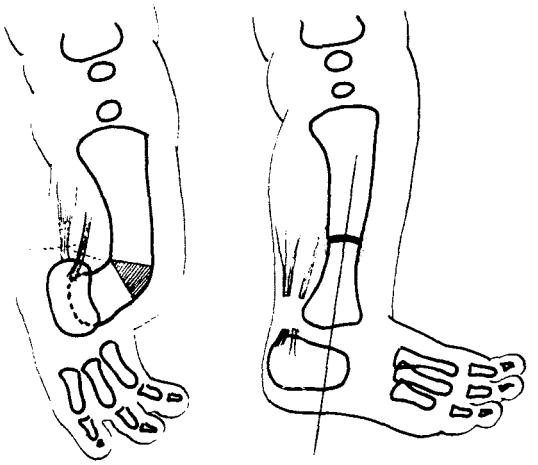

Fig. 7.

with a plaster cast extending from the base of the thigh to the toes. The nail was removed two weeks after the operation. The tibial fusion was normal in all cases. In a couple of cases satisfactory correction was only obtained after reoperation.

RESULTS OF TREATMENT

The age of the patients at the time of the operation ranged from 2 weeks to 2 years and 9 months. The postoperative period of observation varies between 1½ years and 3 years and 3 months. This period of observation is too short to allow conclusions as to the final results of treatment but during this period the tibial angulation and the position of the foot have not changed. Four of the five patients treated operatively are able to walk well with the aid of an orthopaedic boot. In the fifth patient (Case 4) it is the fact that one extremity is entirely missing that prevents her from walking. From the point of view of function the knee joint was good in all cases. The tibia is either quite straight or there remains a slight anterior bowing.

There is often a rocking chair-like deformity in the foot. The shortness of the tibia necessitates the use of an orthopaedic boot, which compensates the shortening of the affected leg.

DISCUSSION

In addition to the severity of the defect, the result of the treatment largely depends on how early operative measures can be undertaken.

The fibular defect is probably the result of a disturbance at the stage of the formation of the fibular anlage in the fifth week of pregnancy. The experiences gained in the Thalidomid cases in particular indicate the truth of this assumption (10, 13). Heredity has also been established (6). The disturbance is not confined to the skeletal parts alone but other tissues in the same area are also affected. The deformation in the extremity does not automatically become corrected during the course of growth, as often happens, for instance, in a sound extremity in children with fractures healed in a poor position. During the period of the foetal development a severe deformity may often have developed from secondary causes.

By conservative measures, redressement and bandages, satisfactory results may possibly be achieved in mild cases seen at an early age. If the tibia is badly curved, however, and if there is a taut fibrous band at the site of the fibula, the value of conservative treatment is doubtful.

It is our opinion that osteotomy of the tibia, in conjunction with tenotomies and excision of the fibular bands, is the best method of treatment. The operation should preferably be carried out in early infancy. With the method adopted by us, a comparatively straight tibia and satisfactory correction of the valgus position have been achieved. The marked shortness of the leg constitutes the most difficult problem. It makes the use of an orthopaedic boot inevitable. The degree of the final shortening of the leg and the necessity of amputation at a later stage cannot yet be evaluated in these cases. In our opinion, amputation during childhood is not to be recommended.

SUMMARY

On the basis of a series of 6 cases of total congenital absence of the fibula, the clinical picture and treatment of this deformity is discussed. The authors stress the importance of early operative treatment. Surgical correction comprising a cuneiform osteotomy of the tibia, achillotenotomy and, when necessary, excision of taut fibrous fibular bands, was carried out. By this procedure it was possible to achieve satisfactory straightening of the tibia and correction of the valgus position in the majority of cases. The patients were then able to walk with the aid of an orthopaedic boot. The authors do not recommend amputation during childhood.

RESUME

Basés sur l'étude d'une série comprenant 6 cas d'absence totale congénitale du péroné, le cadre clinique et le traitement de cette anomalie sont discutés. Les auteurs soulignent l'importance d'un traitement précoce. La correction chirurgicale comprend l'ostéotomie (résection en coin) du tibia, l'achilloténotomie et, lorsqu'elles sont présentes, l'excision des bandes fibreuses péronéales. En utilisant cette technique, il a été possible d'obtenir dans majorité des cas un redressement satisfaisant du tibia et de corriger la position en valgus. Les malades ont été capables de marcher avec l'aide d'une botte orthopédique. Les auteurs ne sont pas partisans de l'amputation pendant l'enfance.

ZUSAMMENFASSUNG

Das klinische Bild und die Behandlung von sechs Fällen totales, kongenitales Defekt der Fibula wird diskutiert. Die Wichtigkeit der frühzeitigen operativen Behandlung wird betont. Keil-osteotomie der Tibia, Achillotenotomie und Resektion der fibrösen Strängen wurden ausgeführt. In meisten Fällen ist es möglich gewesen eine genügende Verbesserung der Deformität zu erreichen. Die Patienten konnten mit Hilfe einer orthopädischen Schuhe gehen. Die Amputation im Kindesalter ist nicht zu empfehlen.

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