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Ectrodactyly Fibular Aplasia

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Amongst the various skeletal hypoplasias, absence of fibula is the most frequent one, yet it does not find a place in standard pediatric books. The condition was first reported in 1936. Fibular aplasia or hypoplasia, when present in association with oligodactyly, absent metatarsals, absent tarsals and cleft hand, is termed as ectrodactyly fibular aplasia (EFA)(1). Other associated anomalies include femoral defects, absence of patella, congenital heart defects and renal anomalies(1).

Case Report

A full term boy, weighing 2800 g was born of a vaginal delivery to a third gravida mother. On examination, the baby was otherwise well but for the deformity of right leg and foot. He measured 49 cm in length and head circumference was 35.5 cm. There was no other apparent skeletal malformation. Systemic examination was unremarkable.

The right lower limb was 3.5 cm shorter than the left and the fourth and fifth toes were absent. There was an anteromedial angulation of tibia with dimpling of overlying skin. The right foot was in equinovalgus position and lateral malleolus was not palpable. The hip and knee joints on the right side were normal. *X*-ray of right foot and leg revealed absence of fibula, cuboid, talus, IV and V metatarsals and phalanges of IV and V toes (*Fig. 1*). There was an anteromedial angulation of tibia. There were no soft tissue calcification or abnormality of knee or hip joint. The *X*-rays of left foot and leg were normal. There was no history of similar anomaly in previous sibs or in the father. There was no history of irradiation, intake of drugs during pregnancy or any significant illness in the mother during first trimester.

Discussion

The fibula is partially or completely absent more often than in any other long bone. In a review of 291 patients with congenital shortening of an extremity, shortening of fibula of 10% or more was found in 44% of the

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Received for publication: September 13, 1991; Accepted: September 8, 1992

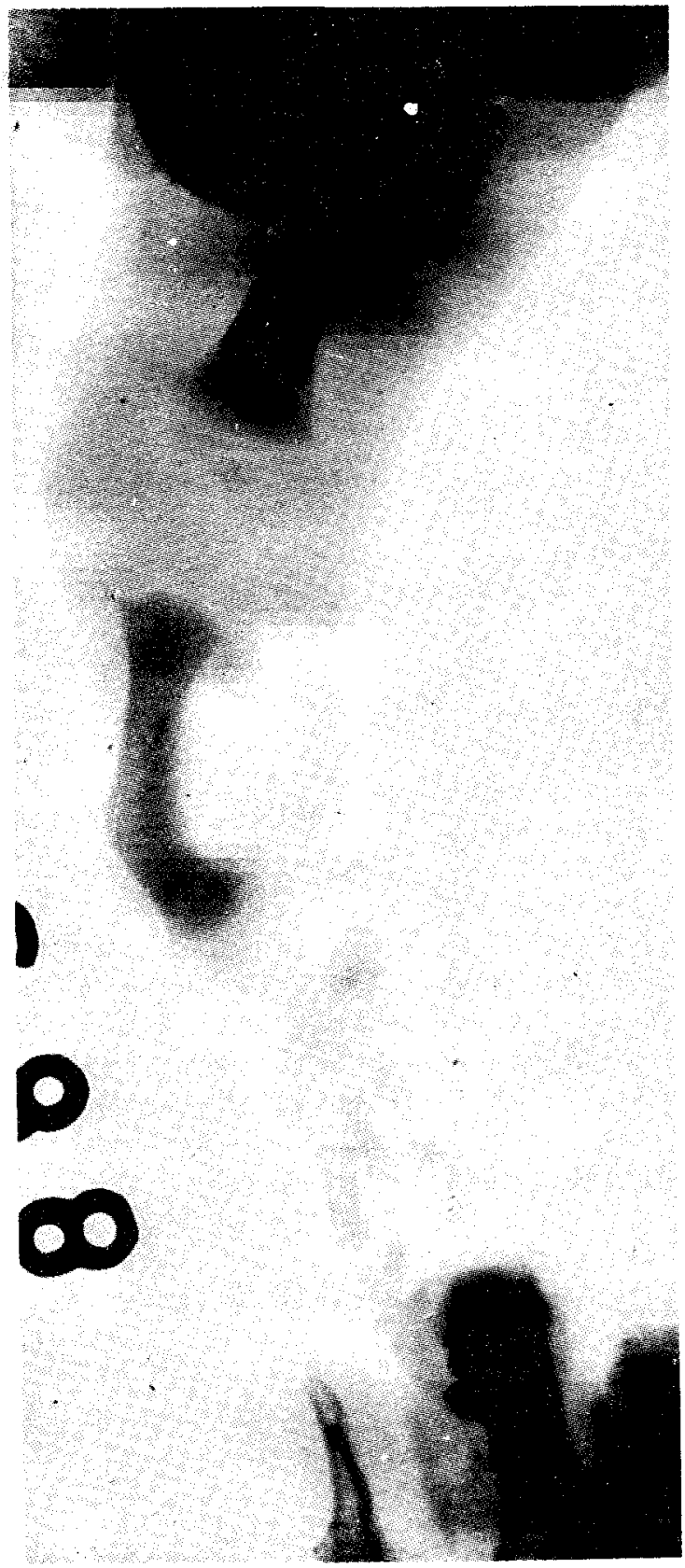

Fig. 1. Radiograph showing absent fibula, tibial shortening with anteromedial bowing and absent IV and V toes.

patients(2). This congenital fibular deficiency or ectrodactyly fibular aplasia (EFA) has been classified into 3 types(3), *Type I:* fibula is hypoplastic; *Type II:* fibula is completely absent or only a distal vestigial segment is present with tibial shortening; *Type III:* bilateral fibular aplasia with other congenital abnormalities.

The management of these cases varies with the age of the patient when first seen, the severity of the deformity, the tightness of soft structures and whether the anomaly is unilateral or bilateral(4). In Type I, epiphyseal arrest of opposite limb is carried out to equalize the leg lengths if practical or elevation of the shoe on the affected side. In Type II, Modified Syme amputation is recommended because it preserves the distal tibial epiphysis and the heel pad and an end bearing prosthesis can be worn. If patient is first seen after 5 or 6 years, any indicated amputation should be preceded by osteotomy. In Type III, the treatment depends on severity of the deformities and nature of associated anomalies.

Early recognition and timely surgical intervention may be helpful in achieving maximal functional ability especially in EFA-Type II whose prognosis is better(5).

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Autoantibodies to SS-A/RO in an Infant with Congenital Heart Block

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Neonatal lupus erythematosus is a rarely reported condition characterized by congenital heart block or transient cutaneous lupus or both(1). Mothers of these infants may have (connective tissue diseases) or may be asymptomatic, but consistently have circulating antibodies to SS-A/RO antigen.

Case Report

A new-born-girl was born to a third gravida mother by emergency cesarean section at 38 weeks gestation. During labour, the fetal heart rate varied betwen 50 and 70 beats per minute. Examination at birth showed a full term baby weighing 2.95 kilograms, with a length of 49 cm and head circumference of 33 cm. The heart rate was 62 beats minute with all peripheral pulses well felt. The blood pressure was 60/45 mm of Hg in the right arm. Auscultation of heart was normal. There was no skin rash and the rest of the systemic examination was normal. Routine hematological investigations were normal. The chest roentgenogram was normal. The electrocardiogram revealed a complete heart block with an artial rate of 140/minute and ventricular rate of 60/minute. The 2 dimensional echocardiography done on the infant showed a structurally normal heart.

The mother did not have any evidence of skin rash, joint pains or urinary complaints. Investigations of the mother revealed normal hematological, hepatic and renal parameters. Serology for connective tissue disorders showed anti-nuclear antibody (ANA), anti-Sm antibody and anti-double standard DNA (anti-ds-DNA) antibody tests to be negative. However, antibodies to SS-A/RO were detected in a titre in 1 : 64. The neonate's blood, when examined, showed antibodies to SS-A/RO in a similar titre.

The neonate was given a trial of intravenous atropine under cardioscopic monitoring with no resultant change in heart rate.

Since the child had no evidence of

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Received for publication: June 14, 1991; Accepted: April 16, 1992