Congenital Cleft Foot (Lobster Claw): An Unusual Association with Ano-Rectal Malformation

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Although the vertebral and radial anomalies are commonly seen in infants with ano-rectal malformations, the major abnormalities of lower limbs in the form of hypoplastic leg, absent tibia or fibula, ilial hypoplasia, arthrogryphosis have been infrequently reported(1). We report a case of high ano-rectal malformation having multiple anomalies of the para-axial skeleton. The purpose of this report is to highlight the association of ano-rectal malformation with a rarely encountered skeletal anomaly–unilateral cleft foot.

Case Report

A full term baby weighing 2 kilograms having high ano-rectal malformation was treated in the Department of Pediatric Surgery (*Fig. 1*). Antenatal history of mother did not reveal any exposure to drugs or radiation. Clinical examination of the baby revealed multiple anomalies of the para-axial skeleton. These included hypoplasia of tibia and fibula of the right side along with cleft foot of the same side, and brachydactyly involving the metacarpal of the right thumb (*Fig. 2*). The maldevelopment of the foot involved absence of the middle digital ray and fusion of the bases of the fourth and fifth metatarsals. The medial ray consisted of normal first and second metatarsals. No other major abnormalities involving gastro-intestinal tract, genito-urinary system and heart were present.

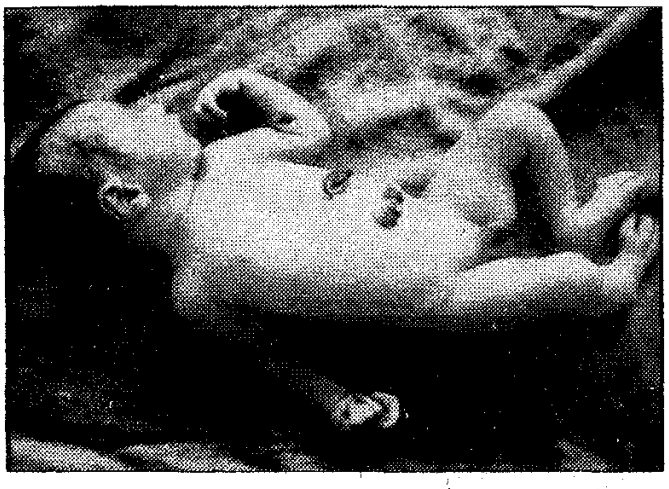

Fig. 1 Clinical photograph showing hypoplastic right leg with cleft foot and loop colostomy for ano-rectal malformation.

A left transverse loop colostomy was performed. The post-operative period was uneventful. The child is under followup and definitive procedures for high ano-rectal malformations and cleft foot will be performed at a later date.

Discussion

Congenital cleft or split foot is a form of ectrodactyly characterised by the absence of two or three central digital rays of the foot. The cone-shaped cleft in the forefoot tapers proximally. The first metatarsal may be of normal size or it may be broad and connected with the intermediate cuneiform at its base representing fusion of the first and second metatarsals. Valgus deformity of great toe is common. The lateral

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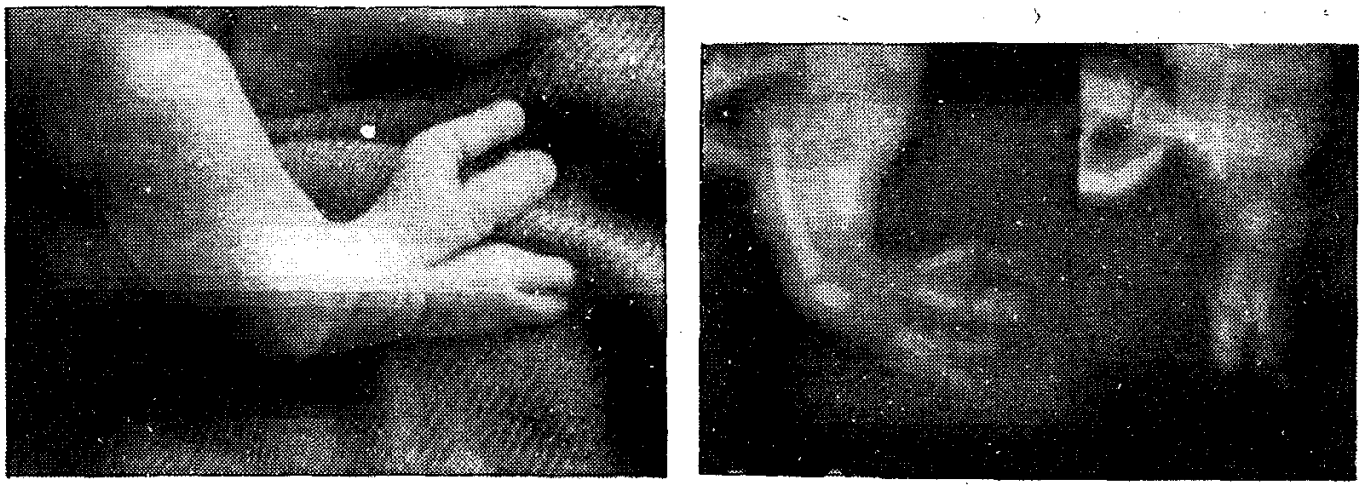

A *B*

Fig. 2. Clinical appearance and roentgenogram of congenital cleft foot.

digits may consist of only the fifth metatarsal or the fifth and fourth metatarsals. The phalanges of the lateral ray usually deviate towards the midline. The hindfoot is normal.

Cleft foot is a very rare malformation; it exists in two forms. In the typical form, the deformity is always bilateral. It is inherited usually as an autosomal dominant trait with incomplete penetrance(2), although an uncommon autosomal recessive transmission has also been reported(3). In the less common atypical form, the deformity is unilateral (as seen in our case) and there is no evidence of familial inheritance. In view of the existence of sporadic, dominant and recessive forms of the cleft foot defect, the propensity for skipped generations and the fact that there is probably considerable heterogenecity, genetic counselling can be very difficult(4). It is interesting to note a similar heterogenecity in ano-rectal malformations also.

Although bilateral cleft foot may be an isolated deformity, it usually occurs in conjunction with clawing of the hand(2). Other associated abnormalities are cleft lip and palate, reduction in number and size of phalanges, syndactyly and polydactyly, triphalangeal thumb, scalp defects and genito-urinary anomalies(4,6). Two distinct syndromes, EEC syndrome and REEDS syndrome have been described(7,8). We have not come across association of cleft foot with severe hypoplasia of tibia, fibula and high anorectal malformation before this report.

Surgical correction of cleft foot is indicated to facilitate the fitting of shoes and to improve the objectionable appearance. Surgery is performed between one and two years. The divergent metatarsals are approximated by osteotomy at their bases, the deformed toes are normally aligned, and the cleft forefoot and toes are surgically syndactylized to maintain correction.

REFERENCES

1. Gruchalski J, Irving IM, Lister J. Skeletal anomalies associated with esophageal, duodenal and ano-rectal atresias. Lancet 1976, 2: 17.

2. Brasky AJ. Cleft hand: Classification, incidence and treatment. J Bone Joint Surg 1964, 46: 1707-1720.

3. Mosavy SH, Vakushuri P. Split hands and feet. S Afr Med J 1975, 49: 1842-1844.

4. Preus M, Fraser FC. The lobster-claw defect with ectodermal defects, cleft lip-palate, tear duct anomaly and renal anomalies. Clin Genet 1973, 4: 369-375.

5. Phillips RS. Congenital split foot (Lobster claw) and triphalangeal thumb. J Bone Joint Surg 1971, 53: 247-257.

6. Adams FH, Oliver CP. Hereditary deformities in man due to arrested development. J Hered 1945, 36: 3-8.

7. Rudiger RA, Hasse W, Passarge E. Association of ectrodactyly, ectodermal dysplasia and cleft lip-palate. Amer J Dis Child 1970, 120: 160-163.

8. Reed WB, Brown AC, Sugarman GI, Schlesinger L. The REEDS syndrome. Birth defects 1975, 10: 61-73.

NOTES AND NEWS

WORKSHOP ON GROWTH MONITORING AND NUTRITION SURVEILLANCE

A Workshop on 'Growth Monitoring and Nutrition Surveillance Problem' will be organized by the Growth and Development Chapter and Nutrition Chapter of IAP Subspeciality on *17th January 1993* during the XXX National Conference of IAP, Calcutta. The Registration Fee is Rs. 100/- which is to be paid by Demand Draft drawn on 'XXX National Conference of IAP, Calcutta' payable at Calcutta and to be sent to the Organizing Secretary at 9/1 Ramanath Pal Road, Calcutta 700 023.

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