

Split Hand and Foot Malformation

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Case Report

A 15 months old female child was seen with deformities of both hands and feet present since birth. The child was first in birth order and product of non- consanguineous marriage with no significant antenatal or family history. In both the hands (Figures 1, 2), there was a wide central cleft with presence of only four digits. The right hand in addition had syndactyly of thumb and index finger. Both the feet (Figure 4) had a cleft with absence of second and third toes. There were no dysmorphic features nor were any other anomalies detected on systemic examination. Developmental milestones had been achieved normally. Plain radiographs showed the absence of central ray in both the hands (Figure 3) and syndactyly (soft tissue) of thumb and index finger in right hand.

The plain films of feet (Figure 5) showed cleft on both sides with five metatarsals and three toes. On right side, the second and third metatarsals were rudimentary whereas the middle toe had two proximal phalanges, each articulating with a metatarsal (3rd & 4th) and thereby representing syndactylous 3rd and 4th toe. Rest of the skeletal survey was normal. Chromosomal analysis did not reveal any defect. Functions of both feet were satisfactory; however the functions of hands were impaired. Besides the deformity was a cause of psychological distress and social embarrassment for the parents. The parents were counseled and offered option of staged correction of deformities.



Figure 1. Bilateral cleft hands (palmar view) with absence of central ray; the right hand also shows syndactyly of thumb and index finger.



Figure 2. Bilateral cleft hands (dorsal view)

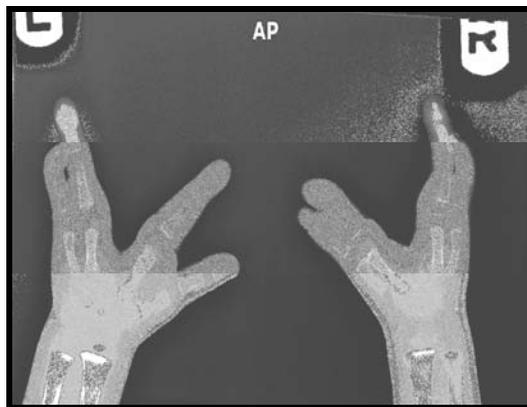


Figure 3. Plain AP film of bilateral cleft hands as shown in Figure 1, 2.



Figure 4. Bilateral cleft feet with absence of 2nd and 3rd digit

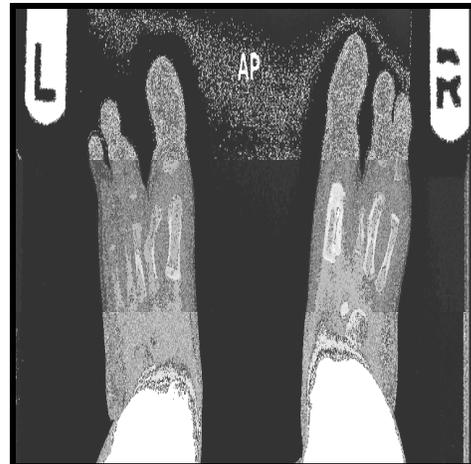


Figure 5. Plain AP film of bilateral cleft feet as shown in Fig 4

Discussion

Split-hand/split-foot malformation (SHFM) is a limb malformation involving the central rays of the auto pod (the distal division of the limb such as the foot or hand) and presents with syndactyly, median clefts of the hands and feet, and aplasia and/or hypoplasia of the phalanges, metacarpals and metatarsals ¹. Ectrodactyly, lobster foot and hand or split foot and hand are used as synonyms. Genetically split hand/split foot malformation (SHFM) has been found to be heterogeneous, with mutations identified at five loci (SHFM1 at 7q21.3, SHFM2 at Xq26, SHFM3 at 10q24, SHFM4 at 3q27 and SHFM5 at 2q31) ².

Barsky has described two types of cleft hand ³. One is typical cleft hand with a deep palmar cleft which separates the two central metacarpals. It is often bilateral and may involve the feet. Our case belonged to this type. Usually the middle or index ray is deficient in a V-shaped deformity. Its incidence has been reported to be about 1 in 90,000 new births with no sex predilection ^{2,4}.

In the atypical cleft hand, the central rays are absent and only short radial and ulnar digits remain with a shallow cleft giving a U-shaped deformity. The thumb and little finger usually are the best-developed digits on the hand. This deficiency is usually unilateral and a family history seldom is present. Other associated abnormalities include cleft lip and palate, reduction in number and size of the phalanges, syndactyly, polydactyly, triphalangeal thumb, scalp defect, genitourinary anomalies with atresia of the nasolacrimal duct and buphthalmos⁵.

Treatment is surgical and is directed at attainment of the features of a basic hand and feet, functionally and cosmetically ⁶. Counseling of parents is important with respect to management of deformed offspring and the possibility of recurrence of the disease in the future siblings and antenatal diagnosis by ultrasonography should be offered⁷.

References

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