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Unilateral lobster claw syndrome with pulmonary hypertension: A Rare entity

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Abstract

Ectrodactyly, is a congenital malformation of limb, presenting with deficiency or absence of one or more central digits of hand or foot along with/without syndactyly, presented as median clefts of the hands or feet with aplasia/or hypoplasia of the phalanges, metacarpals or metatarsals. Presence of median cleft in the hand or feet gives the appearance as lobster hand/feet, hence also referred as various names Lobster- claw syndrome, pincer split hand, claw hand and split hand complex or split hand /feet anomalies (SHFM), resulting from insult in intra-embryonic life either genetic or environmental. Lange's typical Cleft hand is often presented with syndactyly or polydactyly, also associated with other anomalies such as Anencephaly, mental retardation, tibial aplasia, ectodermal dysplasia, craniofacial findings, orofacial clefting, imperforate anus, onychia, cataracts and deafness. While atypical cleft hand is unilateral in upto 50% of cases and feet are usually not involved. The condition does not show any inheritance pattern and is not associated with any other associated anomaly. Another mode of expression exist for cleft hand (SHFM) are Syndromic and Nonsyndromic forms, an isolated Nonsyndromic form limited just to the limbs and Syndromic expression involving other anomalies as like Lange's typical cleft hand. There are five types of SHFM syndrome based on the chromosomal associations and genes thought to be responsible for SHFM. Congenital heart diseases are mostly found in SHFM type 1 and in SHFM type 5. Here we report a case of unilateral Lobster- claw left hand associated with congenital heart diseases (ventricular septal defect), with pulmonary hypertension reported.

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INTRODUCTION