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Lobster Claw Hand Foot Syndrome: Rare Congenital Orthopedic Disorder

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Case

A term male infant was born to 28 year old primigravida mother, non-consanguineous marriage, with birth weight of 2.8 kg and with normal Apgar score 8/8/9. At birth baby was noted to have deformed left foot with syndactyly of the lateral two toes with median cleft of the left foot with associated hypoplasia of nails (Figures 1 and 2). There were no other facial and limb abnormalities noted in infant. The infant was diagnosed as case of diagnosed with a case of cleft foot (lobster foot). The lower limb x-ray showed presence of tibia as these patients are known to have tibial agenesis. The infant was evaluated with echocardiography and brain ultrasound which was normal. There was no orthopedic abnormality in the parents or their relatives. The baby was discharged and now in regular orthopedician follow up.

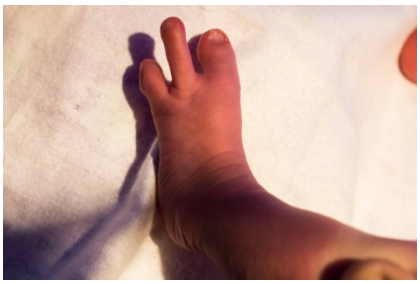

Figure 1: Lobster foot with syndactyly of the lateral two toes with median cleft of the left foot with associated hypoplasia of nails

Figure 2: Lobster foot with syndactyly of the lateral two toes with median cleft of the left foot with associated hypoplasia of nails (Note: the normal contralateral foot and fused nails of the lateral two great toes)

Discussion

Ectrodactyly–ectodermal dysplasia–cleft syndrome is an infrequently seenorthopedics malformation with reported incidence being1: 90 000 live births. It is known by varied names including split hand–split foot–ectodermal dysplasia–cleft syndrome or split hand, cleft hand or lobster claw hand/foot. It is called lobster claw hand/ foot because of the presence of median cleft in upper and lower limbs due to is absence of central digital rays, giving the affected limbs the appearance of lobster claws [1]. Embryologically it is thought to arise due to wedge shaped defect of the apical ectoderm of the limb buds [2]. The most commonly seen inheritance pattern is autosomal dominance though rarely X linked and autosomal recessive are also seen [3]. The associated malformations includes tibial aplasia, learning disabilities, ectodermal and cranio-facial findings, orofacial clefting, renal abnormalities such as VUR, recurrent UTI, enamel hypoplasia and conductive hearing loss [4]. Manske and Halikis surgical classification is most commonly used even though there are many classifications available [5].

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