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Management of a Case of Split Hand/Foot Malformation with Functional Triumph

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ABSTRACT

Introduction: Split-hand/split-foot malformation (SHFM). It is defined as a malformation of the limb involving the central rays of the autopod and presenting with a deep median cleft of the hand and/or foot, aplasia/hypoplasia of the phalanges, metacarpals, and metatarsals. We report a case of management of ectrodactyly involving both hands and feet.
Case Report: A 4 yr old male child who was born out of non-consanguineous marriage presented with the gap at the middle of both hands and foot since birth, with associated history of falling of small object through the gaps of the hands. Snow-Littler technique was planned first on Right hand and then after 6 months on the left hand. Syndactyly was released between 3rd and 4th digit using Cronin technique & the raw area was covered by Split Thickness skin graft. Removal of polydactylysed digit was performed from right 1st web-space and the transposition of index finger to 3rd metacarpal was done using K- wire.
Conclusion: Surgical management at the right time and proper counseling of the parents are required in order to have a functional triumph in the affected child with avoidance of recurrence of the condition in the future pregnancies.

Keywords: Split, Hand, Feet, Ectrodactyly, Malformation

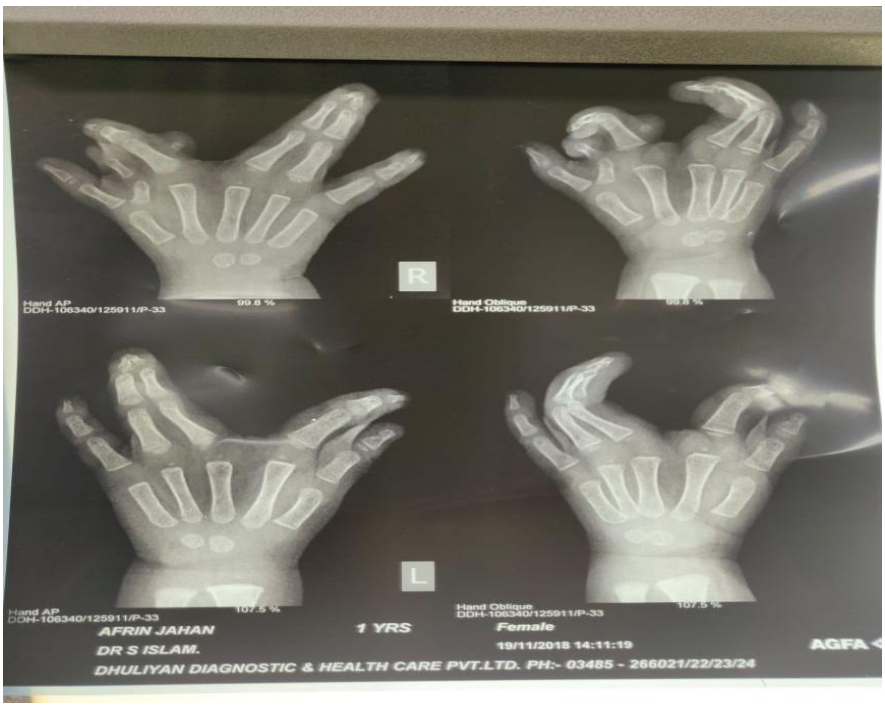
INTRODUCTION

Ectrodactyly and "Lobster claw hand" are the other terms used for the Split-hand/split-foot malformation (SHFM) .It is defined as a malformation of the limb involving the central rays of the autopod and presenting with a deep median cleft of the hand and/or foot, aplasia/hypoplasia of the phalanges, metacarpals, and metatarsals. The incidence of SHFM is around 1 in 90,000 live births. [1] It expresses in two forms, one is nonsyndromic, where the isolated involvement of limbs occurs and other is associated anomalies known as syndromic form. [4] The absence of the central digital rays secondary to the median cleft in the hand and feet which results in the appearance of a lobster. [2]
We report a case of management of ectrodactyly involving both hands and feet.

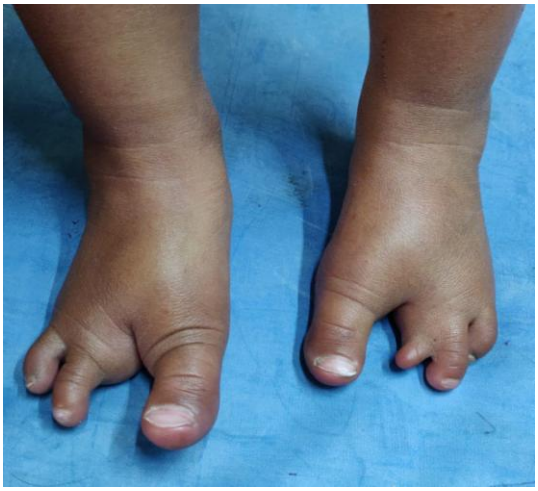
CASE REPORT

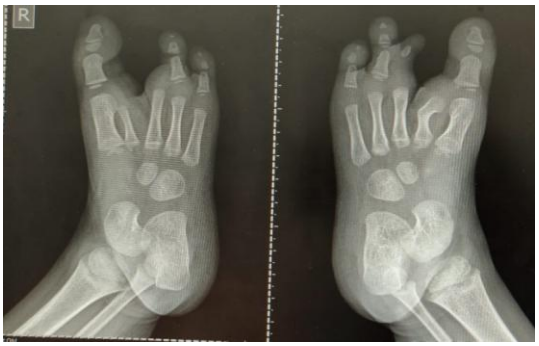
A 4 yr old male child who was born out of non-consanguineous marriage presented with the gap at the middle of both hands and foot since birth, with associated history of falling of small object through the gaps of the hands. Although the child had been using both hands for normal activities and used to grasp larger objects without difficulty, with left hand as being the dominant hand. He used to walk normally. He had born with normal vaginal delivery and was having normal body weight as per age. He had no other associated anomaly. He had undergone detailed Clinical and radiological examinations (FIGURE 1A&1B) for involved extremities. His 2D Echo was normal. A diagnosis of Split hand/foot malformation (Non Syndromic)

was made with Manske type 3 (in Right hand) Type 2 (in Left hand) classification.



1A





1B

FIGURE 1A & 1B – X RAY SHOWING SPLIT HAND/SPLIT FEET

After PAC fitness, Snow-Littler technique was planned first on Right hand and then after 6 months on the left hand. Separation of first webspace & resurface were done with palmar based full thickness flap from the cleft avoiding injury to the neuro-vascular structure, and too much thinning of the cutaneous flap.(FIGURE 2) The length of flap was not kept too long as it was a random flap, and after the elevation of flap bleeding from the margin was checked for viability.(FIGURE 3 &4)

Syndactyly was released between 3rd and 4th digit using Cronin technique & the raw area was covered by Split Thickness skin graft. Removal of polydactylysed digit was performed from right 1st web-space and the transposition of index finger to 3rd metacarpal was done using K- wire. Immobilizations of upper limbs were done using an above elbow splint for 3 weeks.

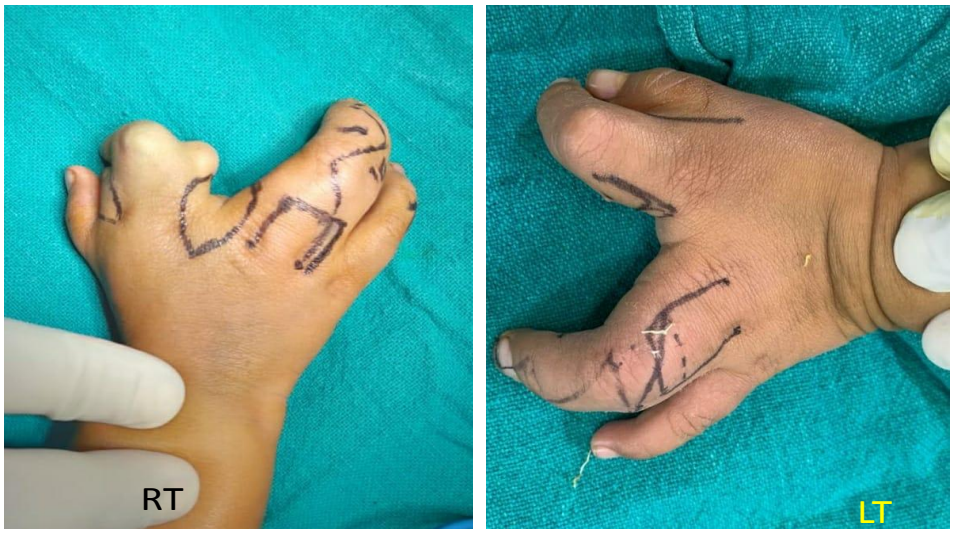


FIGURE 2 - MARKINGS FOR CORRECTION OF DEFORMITY IN RIGHT(RT) AND LEFT(LT) HANDS

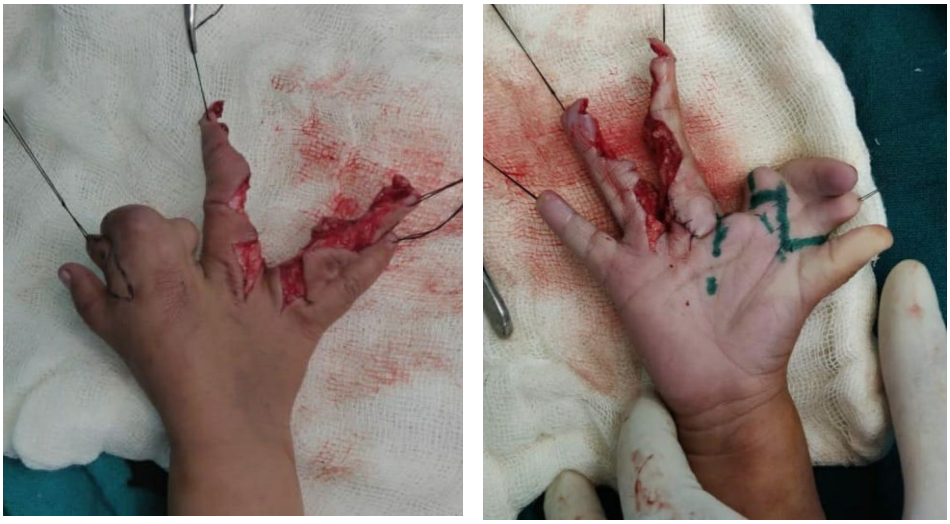


FIGURE 3- INTRAOPERATIVE VIEWS OF RIGHT HAND

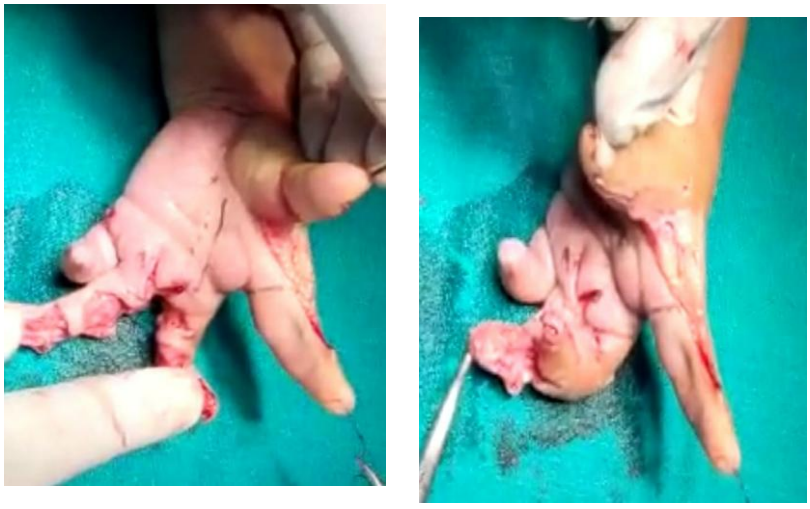


FIGURE 4 - INTRAOPERATIVE VIEWS OF LEFT HAND

Post operatively patchy graft loss was observed that healed over time with regular dressing. There was mild contracture of right Index finger was noted during follow up for

which the child was put on extension finger splint. After 12 months of follow-up, the child was able to hold various objects without fall with better grasp & control.(FIGURE 5)

FIGURE 5- FOLLOW UP PICTURES AFTER 1 YEAR

DISCUSSION

There are five different genetic mutations that are known to be associated with SHFM with Type I being the most frequent variety and it occurs as a result of mutation in chromosome 7 in a region containing two homeobox genes, i.e. DLX5 and DLX6. [2]

The non-syndromal SHFM limited to the hands and feet occurs usually with the pattern of inheritance involving the autosomal dominant gene having high penetrance. [5] However, The syndromic form has a variable degree of expression.

Verma *et al*. had described split-hand and split-foot in two sibships born out of consanguineous marriage and indicated that split-hand and -foot deformity can be inherited as an autosomal recessive trait. [6] Similarly Ray and Freire-Maia also reported autosomal recessive ectrodactyly. [6,8] Interestingly, Klein also reported ectrodactyly in two siblings born out of mating between a man and the daughter of his half-brother. [8] Zlotogora and Nubani described a family is in which four subjects in two sibships had typical SHFM. [9] However, a two-locus model has also been suggested as an alternative possibility. Ectrodactyly results in a social disaster bearing a psychosocial impact over the child which is difficult in regard of management. Surgical management is the mainstay treatment in order to improve function and appearance. Prosthetics are also used. [3] Counseling of the parents should be done regarding the possibility of recurrence of the disease in their future siblings and antenatal diagnosis by ultrasonography should be offered. [4,10-11] We have operated the child using Snow- Littler technique and Cronin method two correct the deformities of both the hands plus proper counseling was offered to the parents for the likelihood of the condition in the future pregnancies. The child was recovered satisfactorily during the post operative period.

CONCLUSION

Surgical management at the right time and proper counseling of the parents are required in order to have a functional triumph in the affected child with avoidance of recurrence of the condition in the future pregnancies.

REFERENCES

1. Barsky AJ. Cleft hand: Classification, incidence, and treatment. Review of the literature and report of nineteen cases. J Bone Joint Surg Am. 1964;46:1707–20. [PubMed] [Google Scholar]
2. %183600 split-hand/foot malformation 1; SHFM1. Available from:http://www.ncbi.nlm.nih.gov/entrez/di

spomim.cgi?cmd=entryandid=183600 [accessed on 2007 Jun 13]
3. Vadoma. Available from: http://en.wikipedia.org/wiki/Vadoma#column-one#column-one[accessed on 2007 Jun 13]
4. Pinette M, Garcia L, Wax JR, Cartin A, Blackstone J. Familial ectrodactyly. J Ultrasound Med. 2006;25:1465–7. [PubMed] [Google Scholar]
5. Zlotogora J. On the inheritance of the split hand/split foot malformation. Am J Med Genet. 1994;53:29–32. [PubMed] [Google Scholar]
6. Verma IC, Joseph R, Bhargava S, Mehta S. Split-hand and split-foot deformity inherited as an autosomal recessive trait. Clin Genet. 1976;9:8–14.
7. Ray AK. Another case of split foot mutation in sibs. J Hered. 1960;61:169–70.
8. Freire-Maia A. A recessive form of ectrodactyly and its implications in genetic counselling. J Hered. 1971;62:53. Klein IJ. Hereditary ectrodactylism in siblings. Am J Dis Child. 1932;43:136–42. [Google Scholar]
9. Zlotogora J, Nubani N. Is there an autosomal recessive form of the split hand and split foot malformation? J Med Genet. 1989;26:138–40. [PMC free article] [PubMed] [Google Scholar]
10. Arbués J, Galindo A, Puente JM, Vega MG, Hernández M, de la Fuente P. Typical isolated ectrodactyly of hands and feet: Early antenatal diagnosis. J Matern Fetal Neonatal Med. 2005;17:299–301. [PubMed] [Google Scholar]
11. Gane BD, Natarajan P. Split-hand/feet malformation: A rare syndrome. *J Family Med Prim Care*. 2016;5(1):168-169. doi:10.4103/2249-4863.184656

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