

Tetrapolydactyly in a female neonate -A case report

Dillip Kumar Das¹, Nijwm Mahilary², Sourajit Routray², Suprabha Shukla³

¹Asst. Professor,² PG Pediatrics, Hi Tech medical college, Bhubaneswar.

³Senior Resident pediatrics, SCB Medical College, Cuttack.

Abstract

Polydactyly is the most common congenital anomaly of the hands and feet consisting of supernumerary fingers or toes. It can be isolated or seen in association with other congenital anomaly. Polydactyly of one limb is more common, but involvement of all the four limbs (tetrapolydactyly) is very rare. We are reporting a case of post-axial tetrapolydactyly in a female neonate without syndactyly and any other congenital anomaly.

Key words: Neonate, Tetrapolydactyly, Polydactyly24, Post-axial.

INTRODUCTION

Polydactyly is a common congenital digital variation of the hand and foot, which is characterized by super numerary digits. It can occur either as preaxial deformity (involving the thumb) or as postaxial deformity (involving the small finger). Small finger duplication (postaxial deformity) has been further subdivided into two types. Type A is a well-formed digit. Type-B is a small, often underdeveloped supernumerary digit.^[1] Previous theories suggest that polydactyly is due to defect in the programmed cell death cycle of fetal limb development. Recently, it is thought to be due to specific genetic malfunction. Hexadactyly of all the four limbs is a very rare skeletal malformation and more common seen in male children.^[2] We are reporting a female baby born to a Hindu Telugu family from India with isolated postaxial tetrapolydactyly without any other congenital anomaly.

CASE REPORT

The index case is a term (38 week and 5 days), female neonate, was delivered through caesarean section from 23 years old Telugu mother. Birth weight was 2834 grams and head circumference was 34 cm and body length was 47 cm. Mother of the baby had similar postaxial tetrapolydactyly without any other congenital anomaly. Mother underwent operation for polydactyly of both the hands for cosmetic reason while both the feet has intact 6 digits. No other family member has history of isolated polydactyly or tetrapolydactyly in previous 2 decades. No history of any drug intake other than the routine drugs prescribed to her during antenatal care.

On examination, the baby had 6 digits in all 4 limbs. The extra fingers were post-axial type and on the ulnar and the fibular side in the upper and the lower limb respectively. All the fingers were well formed. On palpation, each extra digit had bone with mobile joints. Spine and external genitalia were normal. All other systems were clinically normal. No other congenital



Figure 1: Mother's feet showing 6 digits in both feet.



Figure 2: Baby's feet showing 6 digits in both feet

malformation noted. Ultrasonography of the abdomen and the pelvis revealed normal findings. Neurosonogram also revealed no abnormality. Clinico-radiological examination revealed no other congenital anomaly apart from tetrapolydactyly.

DISCUSSION

Polydactyly is the most common congenital anomaly of the hand and foot. The reported incidence of polydactyly varies from about 1 in 300 in black people to about 1 in 3000 in whites^[3]

Address for correspondence*

Dr Dillip Kumar Das

C/O- Bijay Mohan Biswal,

House no- 14, Malhasahi, Mangalabag,
Cuttack, Odisha.

Mob: 07504901123

Email- dr.dillipdas@gmail.com



Figure 3: Baby with 6 digits in bilateral hands and feet.

The incidence of tetrapolydactyly is not known as only case reports are there.^[4] Pre-axial polydactyly occurs more in white children and is often unilateral whereas post-axial polydactyly occurs in African-Americans and may be bilateral, and inheritance pattern is autosomal dominant.^[1] The post-axial polydactyly seen in white people is usually associated with various syndromes and inheritance is autosomal recessive.^[5]

According to the location, polydactyly can be subdivided into preaxial, postaxial and central. Preaxial polydactyly refers to supernumerary digit on the medial aspect of the hand or the foot where as postaxial polydactyly involves the lateral aspect and central ray polydactyly involves duplication of the second, third or fourth digit. It can occur in isolation or in association with other congenital anomaly. Mutations in the *GLI3* gene and another locus on chromosome 13 were found to be associated with post-axial polydactyly but the majority of cases remain unexplained. Recently, another locus on chromosome 19p13.1-13.2 is found to be associated.^[6] The study on polydactyly revealed that the postaxial polydactyly has been significantly seen to be associated with cephalocele, microcephaly, anophthalmia, cleft lip, polycystic kidney disease, and cyclopia.^[7]

As per available literature, males are affected more than the females. Radulescu et al had reported a case of a female

newborn with tetrapolydactyly in a family where 3 generations of males were affected by polydactyly.^[8] Polydactyly in blacks usually occurs in isolation. The digits may be functional or non-functional.^[9] Our case has non-functioning digits in both hands but functioning digits in both feet.

CONCLUSION

Tetrapolydactyly can occur in isolation without any other congenital anomaly, but occurrence of tetrapolydactyly in female neonate still remains a rare occurrence globally, and in that context our case is worth reporting..

REFERENCES

1. Ani CC. Isolated Wassel type II preaxial polydactyly. *Jos Journal of Medicine*. 2010;5:48-9.
2. Mukherjee S, Paul R, Bandhopadhyay M, Das P. Case report on Post-axial Polydactyly with different bony configurations. *International Journal of Anatomical Variations*. 2011;4:77-79.
3. Watt AJ, Chung KC. Duplication, *Hand Clinics*. 2009; 25(2): 215-227.
4. Hosalkar HS, Shah H, Gujar P, Kulkarni AD. Crossed polydactyly. *J Postgrad Med*. 1999;45(3):90-92.
5. Castilla EE, Fonseca RL, Dutra MD, Bermejo E, Cuevas L, Martinez ML. Epidemiological analysis of rare polydactylyes. *Am J Med Genet*. 1996;65(4):295-303.
6. Zhao H, Tian Y, Breedveld G, Huang S, Zou Y, Y J, et al. Postaxial polydactyly type A/B (PAP-A/B) is linked to chromosome 19p13.1-13.2 in Chinese kindred. *Eur J Hum Genet*. 2002;10:162-6.
7. Castilla EE, Lugarinho R, Dutra MD, Salgado LJ. Associated anomalies in individuals with polydactyly. *Am J Med Genet*. 1998;80:459-65.
8. Radulescu A, David V, Puiu M. Polydactyly of the hand and foot. Case report. *Jurnalul Pediatrului*. 2006; 9: 33-34.
9. Sadler TW. *Langman's Medical Embryology*, Lippincott Williams & Wilkins, Philadelphia, Pa, USA, 10th edition, 2006.