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Rare Occasion of Congenital Anomalies: Bilateral Syndactyly with Multiple Polydactyly of the Hands and Feet

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ABSTRACT

Introduction: Upper and lower limb malformations are essential things that occur related to congenital disabilities, including syndactyly and polydactyly. It may appear in association with other birth defects as a part of a syndrome and may be present in the upper extremity or lower extremity. Cases of bilateral syndactyly with multiple polydactyly of the hands and feet are the first time in our center, and they must be promptly diagnosed and treated to avoid functional and cosmetic problems.

Case presentation: A 3-month-old male toddler presented with webbed index, middle, and ring fingers with synonichia, accompanied by additional fingers on hands and feet. An x-ray examination revealed preaxial polydactyly at both hands with bifid phalanx (Wassel classification) and postaxial polydactyly type B. Duplicated postaxial metatarsal (Venn-Watson classification) and type B postaxial polydactyly also occur on both feet with webbed index and middle toe on the left foot. Direct closure was achieved with a dorsal trapezoidal and zig-zag flap to release the second and third fingers of the right hand, followed by resection of rudimentary fingers to help the child with early stages of motoric functions.

Discussion: Syndactyly can cause a length difference that results in growth and functional difficulties. Surgical release of the index to middle and middle to ring fingers should be done early between 3-6 months of age and is expected to help the child grasp, write, and subsequently manipulate objects. We prioritize operating the right hand as around 90% of people prefer to use the right hand. Simple ligation was also performed to recess the rudimentary fingers on both hands and feet. It has no functional benefit and can limit function as it may get caught or make it difficult to wear shoes and gloves. We have to remind the parents that multiple surgeries may be necessary to achieve a satisfactory result and to complete the reconstruction before 24 months of age when the patterns of function of the digits are established.

Conclusion: Bilateral complex syndactyly with preaxial and postaxial polydactyly on both hands and feet are distinctly uncommon entities that need multidisciplinary treatment. In this instance, the syndactyly releasing of the second and third fingers on the right hand is prioritized based on the function to be achieved, with excision of the rudimentary fingers.

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INTRODUCTION

Syndactyly and polydactyly are one of the rarest forms of congenital disabilities on the upper and lower limb malformations. It may appear in isolation or be associated with other birth defects as a part of a syndrome. It presents itself with upper extremity or lower extremity and either medially or laterally. Cases of bilateral syndactyly with multiple polydactyly of the hands and feet are rare, and they must be promptly diagnosed and treated to avoid functional and cosmetic problems with associated morbidity. Most cases found the incidence of syndactyly at 1 in 2,000–3,000 live births and polydactyly at a frequency of 1 in approximately 700–1,000 live births.[1] However, this is the first time in our center a patient presents himself with both.

CASE PRESENTATION

A 3-month-old male toddler was referred to the plastic surgery department from the pediatric department presented with webbed fingers and extra fingers on both hands and feet. Webbed index, middle finger, and ring finger (second, third, and fourth digits) extended past the distal interphalangeal joint with the incidence of synonychia at both hands. This is

accompanied by additional fingers on the hands and feet. X-ray examination revealed preaxial polydactyly at both hands with a bifid proximal phalanx at the right hand and bifid distal phalanx at the left hand (Wassel classification) and postaxial polydactyly type B (figure 1). Duplicated postaxial metatarsal (Venn-Watson classification) and type B postaxial polydactyly also occur on both feet with webbed index and middle toe on the left foot (figure 2). In further examinations, no significant shortening of netted digits was seen, and the range of motion was unaffected. No specific syndromic features were found, nor did a familial history of congenital malformations. Based on these findings, the whole proximodistal length of the adjacent second, third and fourth metacarpals and phalanges were attached; appeared alike and in the same region on both hands, with unions between adjacent digits in accordance with x-ray findings leading to our diagnosis of syndactyly on the hands as complex bilateral syndactyl. Preaxial polydactyly appeared bilateral on hands with type B postaxial polydactyly with six fingers in each (figure 1). Postaxial duplicated metacarpal and type B polydactyly resulting in seven toes on both feet with syndactyly of the index and middle finger on the left foot (figure 2).

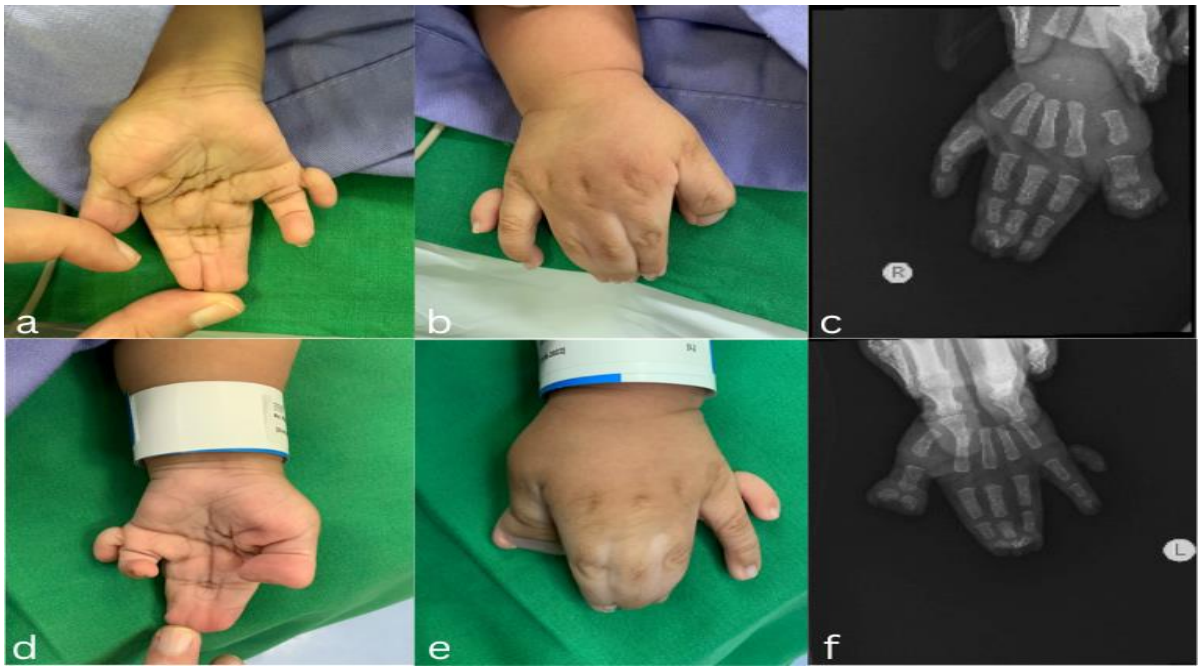

Figure 1 a. Volar view of the right hand; b. Dorsal view of the right hand; c. anteroposterior imaging of the right hand; d. Volar view of the left hand; e. Dorsal view of the left hand; f. Anteroposterior imaging of the left hand

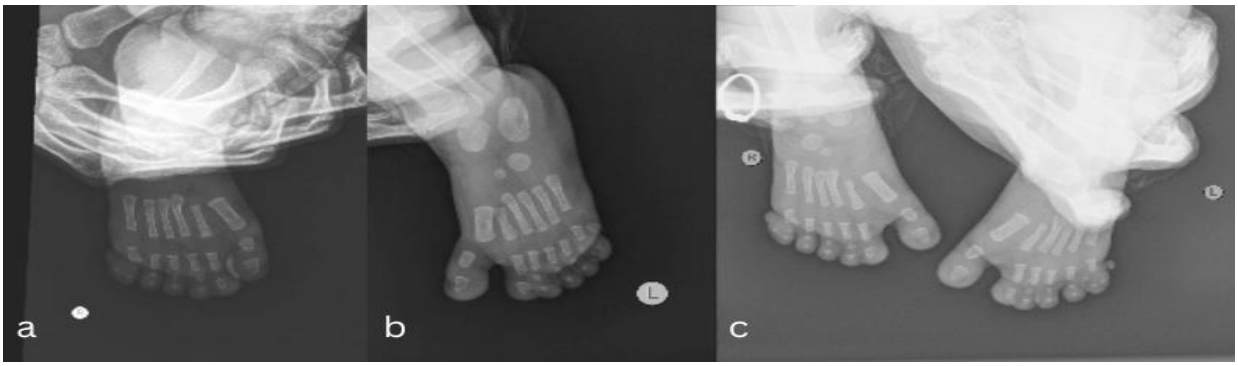

Figure 2 a. Right foot; b. Left foot; c. anteroposterior radiographic imaging of the feet

We attempted direct closure with a trapezoidal flap to release the fingers. This action takes precedence over the right hand as the dominant hand. A dorsal trapezoidal flap was then designed to two-thirds the distance between the metacarpal heads and the proximal interphalangeal joints to reconstruct the webspace of the index and middle finger on the right hand. Zig-zag dorsal and volar flaps are utilized to separate the fingers (figure 3). This step was followed by suturing of the flaps (figure 3). After releasing the webbed fingers, we followed the performance by removing the rudimentary fingers on both hands and feet. The patient was discharged from the hospital after ensuring good postoperative vascularity, motion, and function.

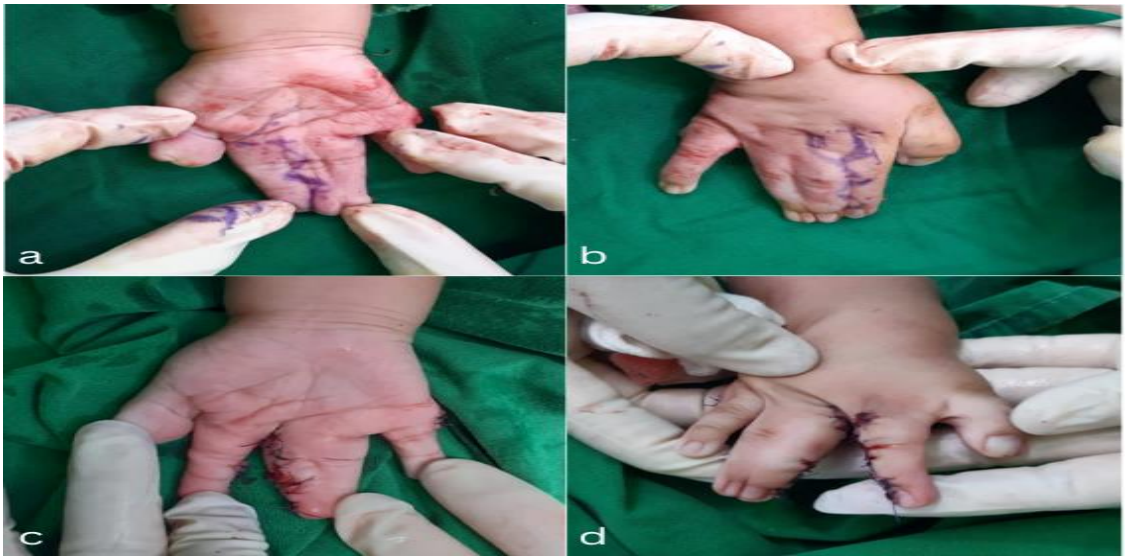

Figure 3 a. Zig-zag volar flap; b. Zig-zag dorsal flap; c., d. suturing of the flap

Four weeks following the patient returned for a follow-up checkup. The distance between the proximal interphalangeal joints of adjacent fingers and the web slope of the reconstructed web space was acceptable. On the right hand, no protruding bones, neuropathies, or other abnormalities were identified (figure 4). The webbed fingers on the middle and ring fingers remained unhampered since the index and middle fingers are utilized mostly for gripping. We suggest the parents undergo further sequential surgery to release the webbed finger of the adjacent hand and polydactyly removal after a three-month follow-up.

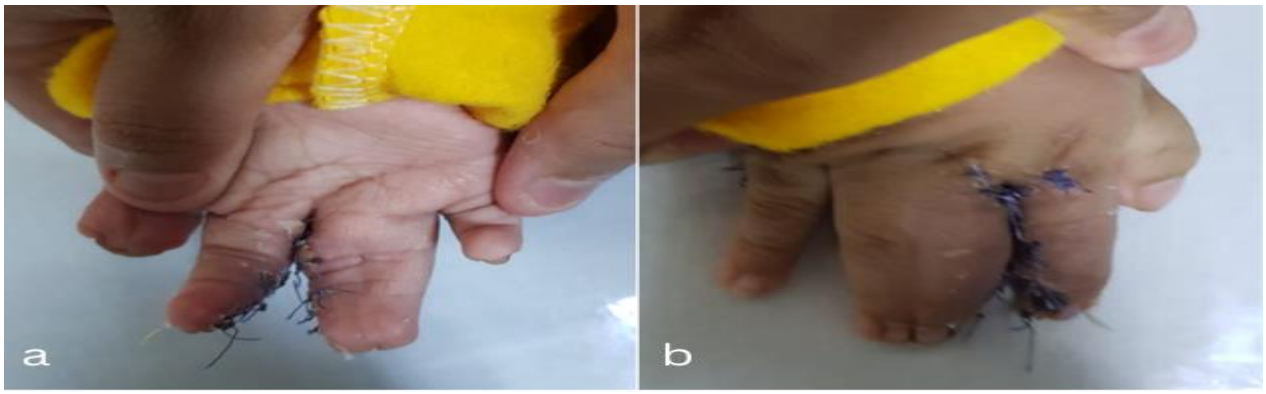

Figure 4 Four weeks after surgery

DISCUSSION

According to the Global Burden of Disease Study in 2017, birth defects are the tenth leading cause of disability-adjusted life years (DALYs) among women and ninth among men. Xu W et al.[2] stated in 2010-2018 the top 10 most frequently-occurring birth defects in China included congenital heart diseases (CHDs), polydactyly, cleft lip with or without palate, club foot, syndactyly, hydrocephalus, hypospadias, limb reduction defects, anotia/microtia, and anorectal atresia/stenosis.

Syndactyly is a congenital malformation that occurs in around two or three out of every 10,000 births. The second chromosome is related to genetic abnormalities that cause syndactyly. The fingers are webbed during normal development until apoptosis and skin recession permit the formation of the digital interspaces. Typically, complete interdigital spaces exist by the end of the sixth week of gestation. Syndactyly may be the only abnormality present or accompanied by Poland's, Cleft hand, or Apert's syndromes. The physical examinations play a key role in differentiating syndactyly and provide a general review of the affected fingers. It will be feasible to distinguish between simple, complex, and complicated syndactyly. For instance, complex synostoses, through plain X-rays of the affected hand are done to detect any synostoses, concealed polydactylies, or other bone deformities.[3] Polydactyly is nearly as typical as syndactyly, but it is ten times more prevalent among Black races than Whites.[4] They are generally separate phenomena however could be coupled with other congenital malformations and may be the only external manifestation of a syndromic condition. Most syndromic cases are preaxial and inherited in an autosomal recessive fashion with a recurrence risk of 25%.[5] Exceptions include Greig cephalopolysyndactyly syndrome and Pallister-Hall syndrome, which have an autosomal dominant inheritance. The classifying of polydactyly of the hand can be divided into

radial, central, and ulnar polydactyly, referring to the anatomical location of the extra digit or part of a digit. The Wassel classification describes the anatomical presentation of thumb duplication.[6] It is a radiographic characterization based on the level of skeletal duplication. From distal to proximal, the phalanges and metacarpal of the thumb are identified as bifid (a cleft in the bone distally with a shared base proximally), duplicated (extension of the cleft from distal to proximal, resulting in entirely independent osseous structures), or unaffected. Wassel Types I through VI are assigned according to this algorithm, but Type VII designates a triphalangeal thumb (figure 5).[7]

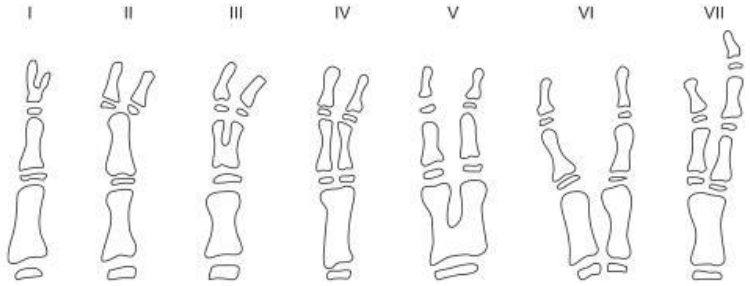

Figure 5 Wassel classification; Type I Bifid distal phalanx, Type II Duplicated distal phalanx, Type III Bifid proximal phalanx, Type IV Duplicated proximal phalanx, Type V Bifid metacarpal, Type VI Duplicated metacarpal, Type VII Triphalangeal

Venn-Watson categorized the lateral duplication of the lateral foot based on the anatomic configuration of the metatarsal and the duplicated bony parts, while The Temtamy and McKusick classification, categorizes duplications by grouping genetically related presentations developed by geneticists. It gives the most general postaxial category, with type A representing a fully formed digit and type B representing a rudimentary and pedunculated digit, commonly denoted as a *nubbin*.[8] As the quality of life declines, it is necessary to discover and evaluate irregularities as soon as possible. In the vast majority of instances, surgery is advised to improve the appearance and function of the hand, preferably during early childhood (i.e., before entering school). [7, 8]

Syndactyly of the border digits causes a length difference that results in growth and functional difficulties due to asymmetric growth, flexion contractures, and rotation deformities. In these circumstances, surgical release should be considered early, between 3 and 6 months of age. Similar to complex syndactyly, early intervention may be advantageous. In nonambulatory infants younger than 12 to 14 months with bilateral involvement, both hands should be addressed simultaneously.[9] Flaps from the dorsum of the affected fingers and dorsal and palmar interdigitating flaps have been employed in traditional surgical techniques to correct syndactyly. Nangineedi N et al.[10] conducted on 26 patients with 37 web repairs and reported the use of dorsal rectangular flap. Jose et al.[11] stated that 221 web spaces were surgically removed from 102 patients; their findings are based on a mix of flap surgeries (palmar and dorsal flaps with zig-zag incisions). The reconstruction of the nail folds requires appropriate skin and pulp. Buck-description Gramcko's of the transposition of pulp flaps is amplified to form the paronychial folds. In this instance, however, we do not need to apply the technique to achieve positive results.[12] Overcompensating the webspace by situating it proximally may reduce postoperative web creep abnormalities. In an effort to minimize or avoid the need for a skin graft, we recruit the excess dorsal tissue with the design of a dorsal pentagonal flap. This entails additional proximal dorsal hand incisions, resulting in a more visible scar. Defatting the digits before flap closure can reduce skin grafts, and must be done carefully to avoid neurovascular damage, venous congestion, and a withered appearance. Younger patients (3-6 months) have the most digital fat amenable to defatting). The goal of syndactyly release is to generate anatomically identical webspace, tension-free soft tissue closure, and restore finger function.

As for preaxial polydactyly of the hands, the reconstruction of the thumb duplication must take precedence over function and motility which is largely determined by the integrity of the carpometacarpal (CMC) joint. The CMC joint plays a larger role in thumb mobility than the metacarpophalangeal (MCP) and interphalangeal (IP) joints. It is typically expected in more distal duplications but may be underdeveloped in proximal duplications, as the thenar musculature can vary from normal to hypoplastic. Children have more difficulty with tip pinch with loss of flexion at the IP joint.[13]

Before pinch grasp or increasing thumb deviation, preaxial polydactyly surgery is usually done between 9 and 15 months of age. Options for reconstruction include complete ablation of one thumb, sharing of equal halves (Bilhaut-Cloquet technique), and reconstruction with composite parts from each of the duplicated digits.[14] According to Dijkman et al.,[15] 85% of thumb polydactyly are treated with resection and reconstruction, 5% with ablation alone, 8% with the Bilhaut-Cloquet procedure, and 1% with pollicization and on-top plasty, respectively. Similar to the reconstruction of a duplicate thumb, type A postaxial abnormalities require surgical reconstruction with consideration for retaining a stable, functional small finger with preservation of the ulnar and adductor digiti minimi, while Type B malformations require simple ligation of the skin bridge, which can be achieved using surgical resection or suture ligation.[13]

Surgery for duplicated metatarsal of the feet is indicated to cosmetically enhance and improve shoe fit and is usually performed when the patient is aged approximately 1 year, so the effect on development and walking is minimal.[16]

In this case, we prioritize operating the right hand, with a worldwide proportion of around 90% of people preferring to use the right hand for many tasks, and 10% the left hand.[17] Releasing the index finger and middle finger with a dorsal trapezoidal flap with a zig-zag incision is expected to help the child use them to practice grasping, writing, and subsequently manipulating objects. Simple ligation was also performed to recess the rudimentary fingers on both hands and feet because it has no functional benefit and can limit function as it may get caught or make it difficult to wear shoes and gloves. We then suggest the parents do sequential surgery within three months of each other. It is important to remind the parents that multiple surgeries may be necessary to achieve a satisfactory result and to complete the reconstruction before 24 months of age when the patterns of function of the digits are established.

Extra fingers and webbed fingers may come as a difficulty when a patient enters the adulthood phase and can be burdensome psychologically. Cases like this can become taboo in society. Apart from being cosmetically unsettling, in some cases, functional disturbances can occur. It is important to provide counseling and education regarding the outcome of the patient's condition and what to expect with or without the following surgery.

CONCLUSION

Bilateral complex syndactyly with preaxial and postaxial polydactyly on both hands and feet are distinctly uncommon entities that need multidisciplinary treatment. In this instance, the syndactyly releasing of the second and third fingers on the right hand is prioritized based on the function to be achieved and excision of the rudimentary fingers.

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