Polybrachysyndactyly: Diagnostic approach and treatment. A case report

José Maria Martinez Rodriguez M.D.
José Luis Villarreal Salgado M.D.
Luis Armando Lopez Rico M.D.
Gerardo Salvador Rea Martínez M.D.
Carlos E. Luna Guerrero M.D.
Samanta Cristina Arce Oliva M.D.
Marcela Rojas Hurtado M.D.
Itzel Arely Murillo Moreno M.D.
Jose Roberto Barrios Lopez M.D.
Claudia Verónica Elizabeth Vargas Hernández M.D.

Monterrey, Mexico

Case Report

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Background
Congenital anomalies of the upper limb comprise a wide group of abnormalities which present different degrees of functional alteration, many of them with the possibility of surgical intervention and rehabilitation when diagnosed in a timely manner, such is the case of polydactyly, a congenital defect that occurs in hands or feet, or both simultaneously, with the presence of one or multiple supernumerary fingers with or without soft tissue and bone between them. On this occasion, the case of a 5-year-old male patient with no important medical history is presented, who presents polybrachysyndactyly in all extremities, who was operated to correct his condition. Polydactyly occurs with an incidence of 1 per 3000 births. It is typically unilateral and sporadic. The clinical picture ranges from partial duplication of the distal end, or bifid forms, to complete digital duplications. It is a complicated anomaly to correct surgically, so it is important to evaluate the degree of hypoplasia, the best position, length and function, to achieve an intervention with adequate bone and soft tissue volume.

Keywords: Syndactyly, congenital hand anomalies.

Congenital alterations occur in 2% of newborns. 10% of cases are located in higher extremities. In the hands, the most frequent are syndactyly, followed by polydactyly.[1] In the case of polydactyly, its incidence is high, around 1:3,000 live births.[2] The etiopathogenesis of congenital hand disorders is very varied. It can occur sporadically, associated with exposure to teratogenic agents such as thalidomide or anticonvulsant drugs such as valproic acid, or be the result of an inherited genetic alteration. In preaxial polydactyly it is usually of sporadic appearance, unilateral and without predominance of sex.[1] Postaxial polydactyly may be multifaceted to or autosomal dominant with incomplete penetrance. It has been described a mutation of the gene located on chromosome 7 (7q36) responsible and encoding the protein Sonic Hedge Hog (SHH) that induces cell growth and differentiation of a large number of structures, such as limbs. The highest prevalence of polydactyly is observed in populations of Negroid origin, especially postaxial polydactyl. Many studies have revealed a higher frequency of this malformation in men than women, and a predominance of the postaxial type over the preaxial.[3]

The diagnosis is clinical and should be made at birth or at the first contact during the consultation. Accompanied by radiological evaluation to be able to classify it and decide the treatment.

The treatment is surgical and should be performed early. Reconstructive options vary by type, from total resection, finger reconstruction using duplicated finger parts, thumb in case of complicated deformities.

Case report

A 5-years-old male whit no important medical history is presented, who presents polybrachysyndactyly in all extremities, who was operated to correct his condition. Osteotomy of the supernumerary fingers was performed with creation of zeta flaps and zetoplasty in the sites where syndactyly was present, with rearrangement of flaps, as well as dermal grafts in the sites with lack of coverage which were obtained from the previously amputated fingers. Posteriorly cursed whit favorable clinical evolution

Discussion

The term polydactyly or also called supernumerary finger is part of group III or duplications of the international classification (Swanson). Preaxial polydactyly, according to Frantz and O'Rahilly, is the most common form of duplication. There are variations in size, and shape, but in general we must consider that it is not an extra

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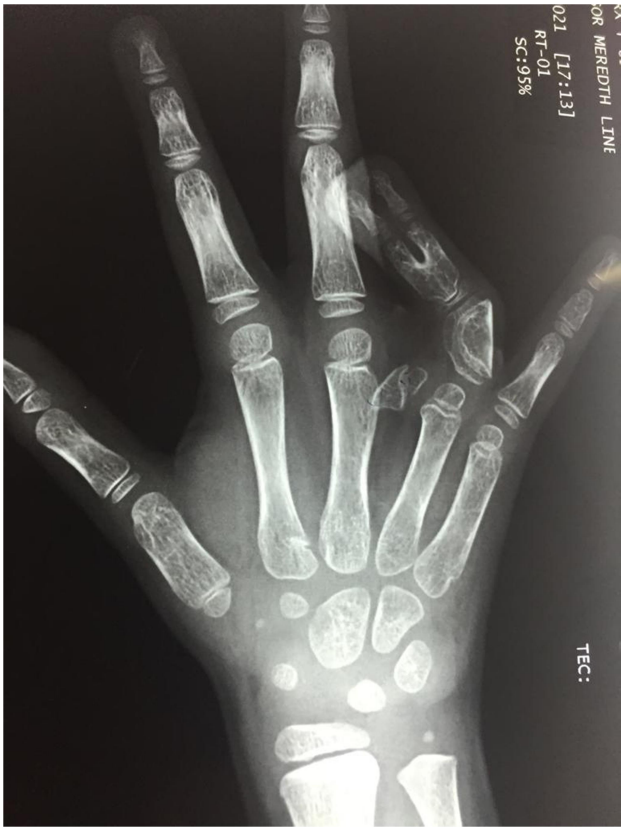

Figure 1. Anteroposterior x-ray of the right hand. The presence of duplication of the middle and distal phalanx of the 4th finger is observed.

finger, but a partition of a single finger, since the 2 most radial rays are never normal (both are hypoplastic and poorly formed), and in most cases it is not fixed with the simple resection of one of them.[4]

Classically there are 3 categories: severe hypoplasia, partial duplication and total duplication (confused with pseudo-doduplications). The most common category is partial duplication, which is further subcategorized in the well-known Wassel classification, the basis of the new modern classifications.

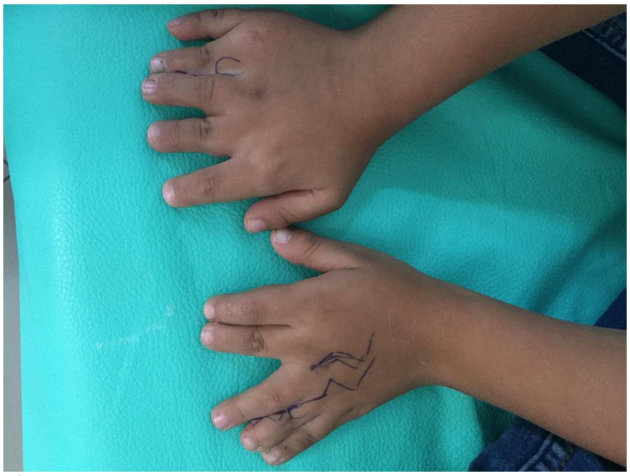

Figure 2. Marking of incision lines for resection of bone tissue and creation of flaps in thoracic extremity (hands).

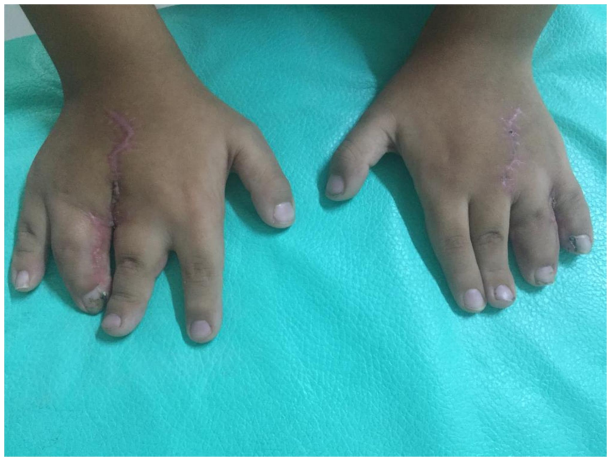

Figure 3. Postsurgical outcome of central polydactyly resection and correction of thoracic limb (hand) syndactyly

The clinical picture ranges from a partial duplication of the distal end, or bifid forms (odd Wassel numbers), to complete duplications of the digit (even) or triphalangeal and triplicate; merged or independent forms; asymmetrical or unbalanced and symmetrical or balanced; curved (clamp or pincer type) or parallel (straight). The most common form is Wassel type IV in which the phalanges are duplicated from the metacarpophalangeal joint. It is a complicated anomaly to correct surgically, so it is a priority to evaluate the degree of hypoplasia (less than 2/3 of the width of the normal thumb); Define which is in better position, has better length, width and function, and finally it is necessary to achieve with surgery an adequate bone and soft tissue volume, cosmetics and alignment of the thumb, in addition to maintaining mobility and stability and / or joint congruence of the metacarpophalangeal. It is also necessary to take into consideration the narrowness of the first space, in case it is necessary to add a z-plasty to 4 flaps.[5]

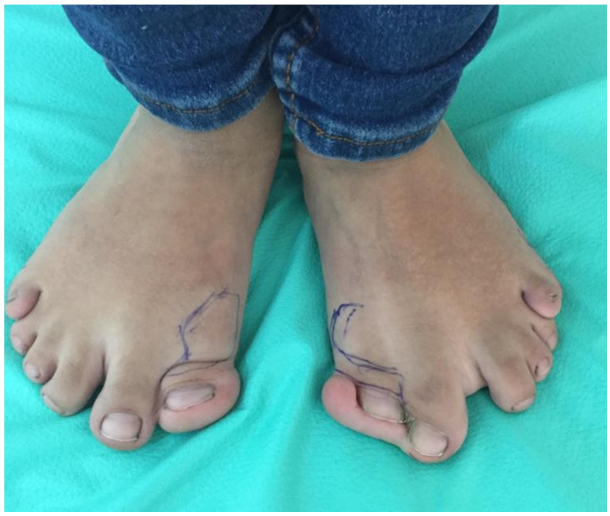

Figure 4. Marking incision lines for bone tissue resection and flap creation in pelvic extremities (feet).

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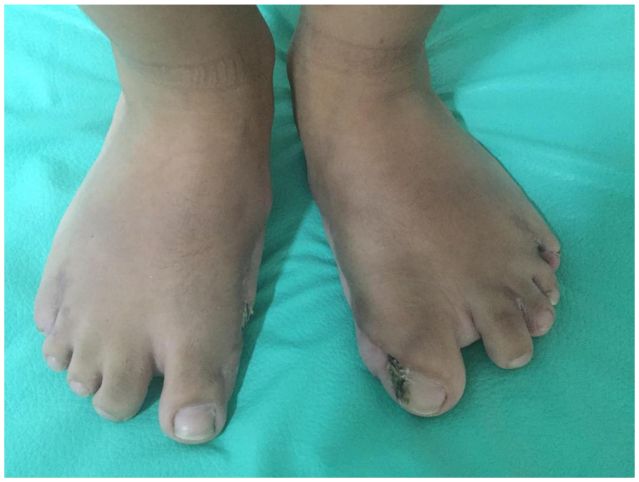

Figure 5. Postsurgical result of correction of polysyndactyly of pelvic extremities (feet).

In radial (preaxial) polydactyly, the most commonly used classification is the Wassel classification. The Wassel classification is based on radiology and is easy to remember. The levels of duplication of the different types are: type I at the distal phalanx; type II at the Interphalangeal Joint (IPJ); type III at the proximal phalanx; type IV at the Metacarpophalangeal Joint (MCPJ); type V at the metacarpal; and type VI at the Carpometacarpal Joint. Thumbs, of which at least one is triphalangeal, are type VII – this can be either at the MCPJ or at the CMCJ. In most series, the three most common Wassel types are, respectively, Wassel IV (about 50%); Wassel II (about 20%), and Wassel VII (about 12%). The percentages are variable in the different reported series.As 31% in our polydactyly series could not be well classified, we modified the classification system. The essence lies in keeping the Wassel classification but naming the differences such as deviation and triphalangism. At first consultation, following medical history and general physical examination, both upper limbs are examined. If the index to little finger is normal, with normal hand and finger creases, and a normal hypothenar region, the examination is concentrated on the radial side of the hand. In our experience, it is performing this systematically, as quite often more anomalies are present than only an extra thumb. The examination is performed from proximal to distal, starting with examination of the thenar muscles. The thenar musculature varies widely from normal to severely hypoplastic. In the Wassel I and II, the thenar musculature is mostly normal, in contrast to the Wassel V, VI, and VII. Hypermobile joints should always be related to the other joints in the hands. It is important to look for creases both dorsally and palmarly. If creases are present, then an active movement in that particular joint can be expected. The CMCJ in polydactyly can be normal, stiff or hypermobile. If abnormalities in the CMCJ are present, they are mostly encountered in the more proximal polydactylies. If polydactyly is situated at the CMCJ, the MCPJ in the best thumb can be near normal. In these cases, the movement is dependent on the presence of a syndactyly between the duplication. Depending on the location of the polydactyly, the MCPJ can be stiff, normal moving, or hypermobile and hypoplastic. For instance, in a polydactyly involving the MCPJ, quite often the duplication moves as a block. In most of these cases, the radial-sided thumb is hypoplastic and stiff, and the ulnar thumb is the better one, often moving well at the IPJ. Finally, the IPJ can present with normal movement, stiffness, or hypermobility. If the duplication is at the IPJ, both parts can move as a block. The range of motion in those cases is typically less than in a normal IPJ. In an asymmetric duplication at the IPJ, the best-developed part usually moves better. Normal examination includes extrinsic and intrinsic movement. In a newborn, it is often difficult to distinguish between these movements. However, flexion and extension can be evaluated, as well as the presence of palmar abduction. In duplicated thumbs, the flexor pollicis longus is Y-shaped in the majority of cases, with a less developed tendon to the most hypoplastic thumb. Therefore, flexion can be seen simultaneously in both thumbs. The extensor apparatus is usually less developed or absent in the more hypoplastic thumb. It can be Y-shaped as in the flexor and asymmetrically attached, therefore, deviating the distal part. The fingertips can be either normal or asymmetric. The asymmetric side is typically found on the opposing sides of the two thumbs. The nails are smaller and asymmetrical in most cases. The first web is nearly always normal in the distal duplications. In more proximal polydactylies, the first web can be narrower than the normal contralateral side.

Ulnar (postaxial) polydactyly classifications include either two or three types. In the two-stage classification according to Temtamy and McKusick, type A comprises an extra little f inger at the MCPJ, or more proximal including the CMCJ. The little finger can be hypoplastic or fully developed. Type B varies from a nubbin to an extra, non-functional little finger part on a pedicle. In the three-type classification, type I includes nubbins or floating little fingers, type II includes duplications at the MCPJ, and type III includes duplications of the entire ray. The most common presentation of ulnar polydactyly is a small appendix on a skin pedicle, in which a neurovascular bundle is present. The appendix, in most cases attached at the ulnar border of the proximal phalanx, has mostly a small nail and thus a distal phalanx, and is non-functional. Sometimes two bones are present. If the extra little finger is more developed, the more common site of duplication is at the MCPJ. In these cases, the fifth metacarpal is broad. Depending on the

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extent of development and the attachment at the MCPJ, the extra little finger flexes and extends well. Full motion of the MCPJ of the normal fifth finger can be affected. Flexor tendons and extensor tendons are usually Y-shaped and asymmetrical, as in the duplicated thumb. The extra finger is usually abducted at the MCPJ and radially deviated at the PIPJ. The hypothenar muscles are attached at the ulnar side of the extra digit and palmarly at the broad MCPJ. If the extra little finger is at the base of the metacarpal or at the CMC, the joint of the finger is usually well developed. Physical examination of ulnar polydactyly depends on the development of the extra little finger. Nevertheless, the same rules apply, as described in the examination of radial polydactyly. Central polydactyly In central polydactyly, the second, third and/or fourth digit can be involved in the duplication. Fully developed extra independent fingers are rare. In order of appearance, the most frequently affected is the ring finger, followed by the long finger, and ultimately the index finger. Frequently, the duplication of the fourth finger is partial and hidden by a syndactyly, typically to the third finger. A number of varieties are possible because the duplication often is not confined to one finger. In addition to the bony deformity and aberrant growth plates, anomalies of the flexor and extensor tendons and neurovascular structures are present. In ulnar dimelia (mirror hand), the hand looks very peculiar and is restricted in function due to the lack of a thumb and the presence of multiple digits. Seven to eight fingers are present without a thumb. Syndactyly can occur. The wrist is broad and mostly flexed. The arm is shorter with a broad elbow, an extension deficit, and restricted forearm rotation and shoulder movement.[6]

Surgery is usually performed between 6 months and 2 years of age, although there is no upper age limit for reconstruction. The basis of the problem is that both thumbs share a mechanical balance that is destroyed by the removal of one of them, leaving the flexor and extensor tendons inserted eccentrically and increasing the zig-zag deviation already existing by the lax joints. One of the surgical options is the Bilhaut procedure misnamed Bilhaut-Cloquet, in which the inner third of the most radial thumb and the outer third of the most ulnar are resected, joining the remaining parts to increase the volume of the digit. The other surgical alternative is to completely resect one digit, the most hypoplastic, and correct the deformities in the remnant by wedge osteotomies, tendon reinsertions, and ligament reconstructions, often using tissues discarded from the other finger.[7]

The most frequently affected space, in almost half of the cases, is the third interdigital space, both in simple and complex forms. It is believed that all syndactyly has surgical indication as long as functional considerations are taken into account, given mainly by the limitation of abduction and digital independence, as well as aesthetic / cosmetic and developmental. To know when to operate, it is necessary to evaluate the amount and type of syndactyly, the length of each finger, and therefore what space is affected. Faced with parental pressure, we must avoid thinking about how quickly the fingers can be separated, and think about how long the functional demand of the hand will allow delaying surgery. For fingers of similar length (second and third spaces), it is better to wait until 18 months of age, although it is always better to delay the intervention until school age, that is, 5 years, as long as there is no limitation of the extension of the shortest finger. For fingers of different lengths (first and fourth spaces), and in order to prevent the shorter finger from retracting and limiting the growth of the partner, it is preferable to intervene between 6 and 12 months of age.

As surgical principles, the corners should be covered with flaps (avoiding sutures) and in the lateral areas of the fingers there will always be a lack of skin to cover with total skin grafts. The different surgical techniques differ basically in the shape of the commissural flap and whether it is of palmar, dorsal or combined (palmo-dorsal) origin. Other techniques are differentiated by using island flaps or by the fact that they do not need skin grafts. Even today different designs continue to emerge, all with good results.[3]

Conclusion

The presence of congenital alterations of the extremities is a complex pathology due to its therapeutic complexity. You must have extensive knowledge of both the embryology and the anatomy of the extremities, as well as their alterations and their classification in order to give the optimal surgical resolution according to each case. The various surgical techniques that can be applied must be taken into account, and the one that best suits each case and that is handled efficiently by the surgeon who will perform it must be chosen. You must be extremely meticulous in each step of the treatment to be able to take the patient in question to obtain the best anatomical, functional, and aesthetic result, since the need will depend on this depending on whether or not to take a new surgical intervention.

Conflicts of interests

There was no conflict of interest during the study, and it was not funded by any organization.

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Jose Maria Martinez Rodriguez
General Surgery Department
Unidad Medica de Alta Especialidad del Noreste Monterrey IMSS
Monterrey, México.

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