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Review Article

Lower Extremity Rare Preaxial Polydactyly. A Case Report and Literature Review

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Abstract: The condition known as pediatric foot polydactyly can manifest itself in a broad range of malformations, from a single extra digit that is only connected to the rest of the foot by a thin band of connective tissue to intricate central foot duplications that involve the duplication of tarsal bones. The presentation of preaxial polydactyly of the foot is crucial to understand, even though it is quite uncommon. This is because in over half of the cases, several congenital malformations, such as syndactyly and atrial septum defects, have been described. The result of surgical reconstruction should be a foot that is stable, mobile, and pain-free, with five aesthetically pleasing toes. This should allow the patient to wear standard footwear and walk without experiencing any discomfort.

Keywords: Duplications, Polydactyly, Preaxial Polydactyly, Congenital Foot Abnormalities, Surgical Management.

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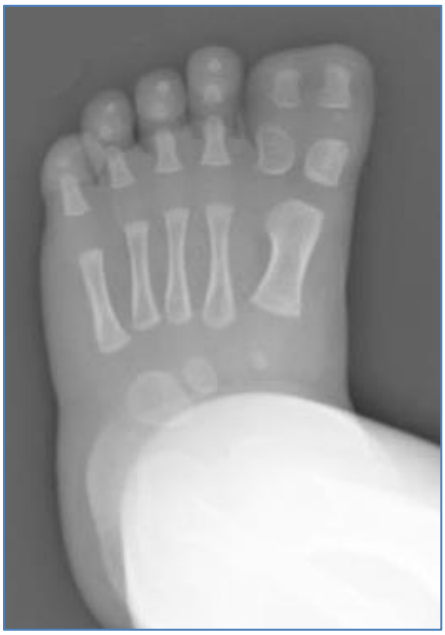
INTRODUCTION

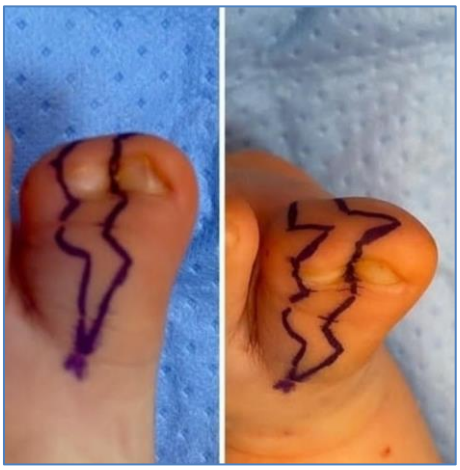
Foot polydactyly is well recognized as the prevailing congenital anomaly affecting the forefoot. The condition is characterized by the presence of six or more digits on the foot. Duplication can manifest at several anatomical levels, ranging from the midfoot to the distal phalanges. In very extreme cases, tarsal deformity may be observed. It is worth noting that around 50% of deformities exhibit bilateral occurrence. Furthermore, concomitant syndactyly is frequently observed in conjunction with these abnormalities. While there is no known sex predisposition, there is a significant variance in the incidence rates based on ethnicity. The rates of incidence are significantly elevated in patients of African heritage, ranging from 3.6 to 13.9 per 1,000 live births. In contrast, the prevalence within the Caucasian cohort is between 0.3 to 1.3 per 1,000 live births. Foot polydactyly is frequently observed as a separate disorder, however it can also be linked to other abnormalities or be a component of a congenital syndrome. Hand polydactyly is the most often observed concomitant condition. According to Venn-Watson's findings, out of a sample size of 65 individuals diagnosed with polydactyly of the foot, around 33% exhibited concurrent polydactyly of the hand. Additionally, these patients presented with many additional related disorders such as vertebral abnormalities, tibial deficits, cleft deformities of the lip and palate, and cardiac defects. When foot polydactyly occurs as an independent disorder, around 30% of individuals exhibit a positive familial association. Several syndromes have been shown to be related with foot polydactyly. These include Ellis-van Creveld syndrome, trisomy 13, tibial hemimelia, trisomy 21, and Greig cephalopolysyndactyly syndrome. The classification of polydactyly is conventionally based on the site of the duplication, which can be categorized as preaxial, central, or postaxial. Approximately 79% of duplications manifest as postaxial anomalies, specifically affecting the most lateral rays of the foot. A total of 6% of the sample population is classified as central, encompassing

the second through fourth rays. The remaining 15% are categorized as preaxial, specifically pertaining to the first ray.

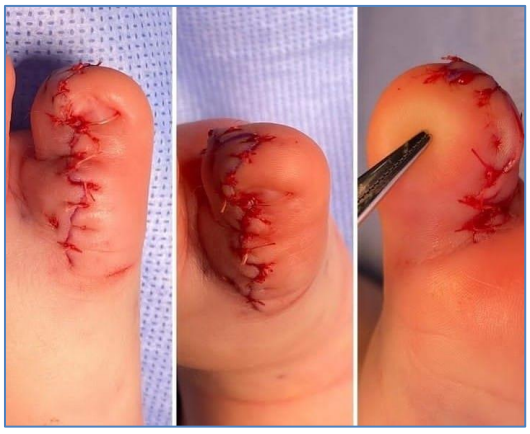
FIGURES



1: Anteroposterior radiograph showing distal phalangeal type of preaxial polydactyly



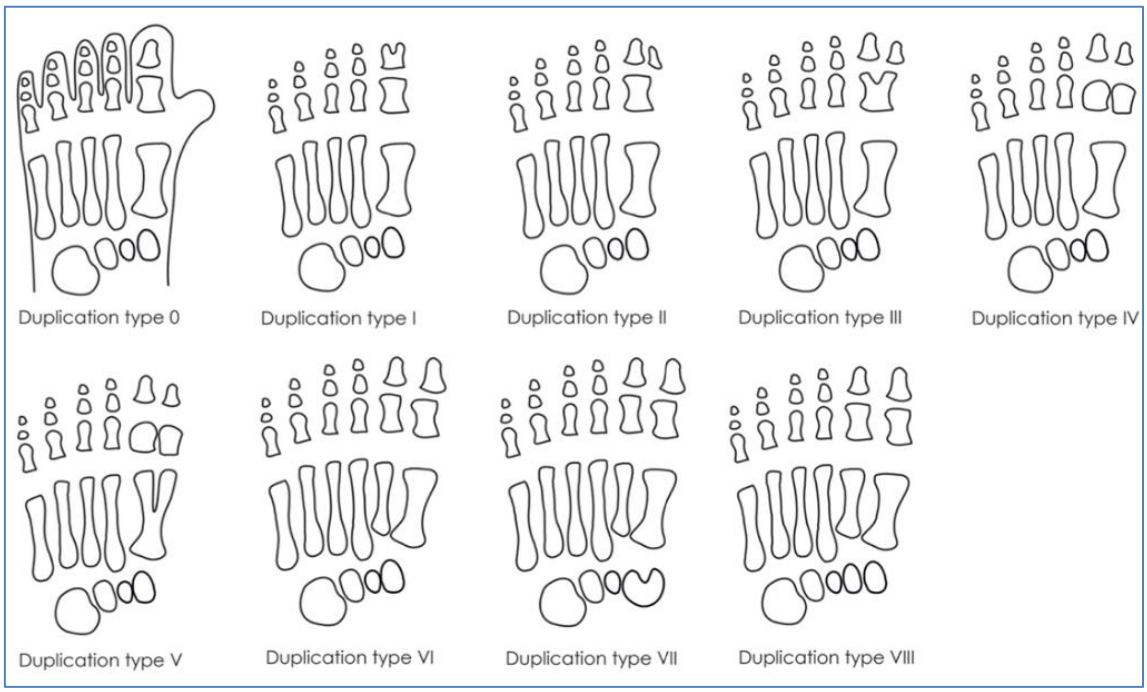
2: Prior surgical markings



3: The intermediate postoperative outcome

DISCUSSION

Polydactyly is a prevalent congenital foot malformation that includes a range of anomalies, varying from insignificant duplications of soft tissue to significant abnormalities in the bone structure. Preaxial polydactyly is characterized by the presence of an additional digit located on the medial aspect of the foot. Treating this particular group of individuals might be challenging as a result of the presence of concomitant abnormalities in the first metatarsal. The initiation of limb development begins around the conclusion of the fourth week of embryonic development, with the lower limb exhibiting a little temporal delay of around 1 to 2 days in comparison to the upper limb. During the sixth week of embryonic development, the hand and footplates begin development. The regulation of limb creation involves an intricate cascade of molecular signals, encompassing HOX genes, SHH genes, and retinoic acid. The anteroposterior patterning of the limb is determined by the zone of polarizing activity situated on the postaxial boundary of the limb bud. Preaxial polydactyly has been linked to several syndromes and malformations, such as cephalopolysyndactyly (Noack, Carpenter, Grieg), Rubinstein-Taybi, facio/digital, Holt-Oram, and Down syndromes. The congenital malformations most often linked with this condition encompass digital polydactyly, digital syndactyly, vertebral anomalies, anorectal anomalies, tibial deficits, cleft deformities of the lip and palate, and heart problems. Polydactyly can be categorized based on its anatomical position, namely as preaxial (involving the first ray), central (affecting the middle rays), or postaxial (pertaining to the fifth ray). The postaxial deformity is the prevailing kind, accounting for approximately 80% of all duplications. Approximately 15% of cases have preaxial malformation, whereas central duplication accounts for approximately 5% of cases. There is a variety of categorization schemes available for foot polydactyly. The categorization proposed by Temtamy and McKusick is solely predicated upon the anatomical site of the abnormality. The Venn-Watson (V-W) classification system is predicated upon the morphology of the corresponding metatarsal bone, encompassing a total of six distinct types: (1) a metatarsal bone that is normal but exhibits distal duplication; (2) a metatarsal bone that is block-shaped; (3) a metatarsal bone that assumes a Y-shaped configuration; (4) a metatarsal bone that assumes a T-shaped configuration; (5) a metatarsal bone that possesses a normal shaft but an expanded head; and (6) a metatarsal bone that exhibits duplication of the ray. Masada *et al.*, made additional modifications to the V-W system. The researchers classified the two preaxial kinds into four distinct categories: type 1, characterized by complete duplication of the metatarsal bone; type 2, characterized by complete duplication of the phalangeal bone; type 3, characterized by partial duplication of the metatarsal bone; and type 4, characterized by incomplete duplication of the phalangeal bone. In their study, Watanabe *et al.,* categorized foot polydactyly into four distinct types: tarsal type (including one subtype), metatarsal type (including three subtypes), proximal phalangeal type (including five subtypes), and distal phalangeal type. Regrettably, the majority of these categorization schemes are mostly reliant on morphological characteristics and offer limited assistance in informing surgical strategizing or prognosticating treatment outcomes.



1: Duplication types in the Rotterdam foot classification

Foot polydactyly results in a range of functional and aesthetic defects in affected individuals, encompassing challenges in shoe fitting and intermittent discomfort when walking. The initial step in assessing the newborn polydactylous foot is identifying the precise positioning of the additional digit or digits. In cases with intricate abnormalities, the determination of which toe is supernumerary may not be readily apparent. However, it is customary to retain the more well-formed toes during the process of surgical repair. The mechanism by which the additional digit is connected to the remainder of the foot is also a crucial consideration in the context of surgical intervention planning. Certain attachments consist of narrow soft-tissue bridges, while others encompass many tissue types such as bone, cartilage, and occasionally the nail plate. The examiner should carefully observe and document the occurrence of syndactyly, nail malformations, as well as any deformities in the metatarsals or phalanges in the axial, coronal, or sagittal planes. These deformities may require corrective osteotomy. In conclusion, while polydactyly often manifests in the forefoot, there are instances where other regions of the limb may also be impacted. It is important to observe and document the existence or absence of constriction bands, as well as conduct a thorough comparison of the legs to identify any asymmetries in terms of length and alignment. Furthermore, it is important to thoroughly evaluate the flexibility and deformity of the hindfoot and midfoot. When there is a suspicion or confirmation of bony involvement based on physical examination, it is recommended to request standard anterior-posterior (AP) and lateral radiographs of the foot. These radiographs will offer a more comprehensive assessment of the degree of metatarsal involvement and deformity. For more complex deformities, delaying the surgery until around 1 year of age is preferable. This allows better preoperative clinical and radiographic evaluation to better define the extent of involvement. Most authors recommend surgery before walking age to avoid any developmental delay related to footwear difficulties or any psycho logical issues related to cosmesis.

In the majority of cases with preaxial polydactyly, the innermost toe is often excised, unless certain circumstances arise when the outermost big toe is visibly undeveloped or elevated in relation to the other toes. In cases when a preaxial polydactyly is present, characterized by the presence of an additional digit with a soft-tissue connection, it is possible to address this condition by the use of suture ligation at the base of the supernumerary toe. To provide a more comprehensive soft-tissue connection, a racquet-type incision may be employed. The management of preaxial polydactyly has a greater level of complexity. The observed unfavorable results were attributed to the occurrence of recurrent hallux varus and insufficient removal of all supplementary osseous components. The importance of soft-tissue rebalancing in preaxial polydactyly surpasses that of postaxial polydactyly due to the potential development of hallux varus or valgus deformity resulting from inaccurate alignment of the medial components. In order to prevent the occurrence of a hallux valgus deformity, it is necessary to surgically reconnect the abductor hallucis and collateral ligaments to the remaining phalanx following the excision of the medial hallux. In cases when the lateral hallux is surgically excised, the adductor hallucis muscle is afterwards affixed to the remaining phalanx in order to mitigate the development of a progressive varus deformity. In cases when the removal of the additional preaxial digit and subsequent adjustment of soft tissues do not lead to the desired alignment of the great toe, it is advisable to contemplate the implementation of a wedge osteotomy. One of the most complex scenarios in preaxial polydactyly pertains to the existence of an epiphyseal bracket that impedes the medial longitudinal development of the toe. In such cases, the use of physiolysis with a wedge osteotomy is necessary. The concepts behind congenital hand reconstruction can also be extended to the field of foot reconstruction.

CONCLUSION

A detailed foot examination should be conducted on every infant, and if foot polydactyly is found, hand polydactyly or other abnormalities should be examined. Preoperative radiography helps determine duplication extent. The objective of surgical repair of foot polydactyly is a pain-free, flexible foot with normal curves and form. Duplication type and location need more complicated techniques. Soft-tissue balance is crucial for stabilizing the joint following duplicate removal. Block metatarsals, medial epiphyseal brackets, and central polydactylys are more likely to develop and require surgery later in adulthood.

CONFLICTS OF INTERESTS: The researchers have disclosed no conflicts of interest.

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