Unique Case of Polydactyly and A New Classification System

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Abstract: Polydactyly of the hands or feet is a common birth deformity. We recently encountered a female infant with a case of a crossed type 1 polydactyly with a mixed polydactyly of the feet. A mixed and crossed polydactyly is a rare finding with only one other reported case. This is the first report of crossed and mixed polydactyly of the feet presenting with 7 complete toes on each foot without syndactyly. In addition to a discussion of the treatment, this case has lead us to propose a more complete and less confusing classification system.

Key Words: polydactyly, postaxial, preaxial, mixed polydactyly, crossed polydactyly

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Polydactyly of the foot is a common deformity occurring in approximately 1.7 in 1000 live births.[1] The incidence is even higher in the black race with estimates ranging from 3.6 to 13.9 in 1000 live births.[1] Polydactyly of either the hand or the foot, when nonsyndromic, is thought to occur as an autosomal dominant trait with variable penetrance.[1–4]

Polydactyly is classified as preaxial, central, or postaxial depending on the location of the duplication.[1] Preaxial polydactyly is defined as a supernumerary digit on the medial aspect of the foot or hand. Postaxial polydactyly involves the lateral aspect of the foot or hand. Central ray polydactyly involves duplication of the second, third, or fourth digits.[2] In all types, the metatarsal may be partially or completely duplicated. There are few published reports of the phenomenon known as crossed polydactyly, classified as either type I or type II.[5–7] Type I occurs when the child has postaxial polydactyly of the hands and preaxial polydactyly of the feet. Type II represents preaxial polydactyly of the hands combined with postaxial polydactyly of the feet. It is much less common than type I.[5] In 1975, Nathan and Keniston proposed another classification known as *mixed polydactyly*. This classification allows for the presence of preaxial and postaxial polydactyly on the same extremity. Mixed polydactyly was further subclassified as upper extremity mixed, lower extremity mixed, and crossed. *Upper* and *lower extremity mixed* indicate preaxial and postaxial polydactyly of either the hands or feet. *Crossed polydactyly* retains its definition as stated above. In addition, an individual with mixed polydactyly of one set of extremities and a preaxial or postaxial deformity of the other set was termed *mixed* and *crossed*.[5]

We recently encountered a female infant with a case of a crossed type 1 polydactyly with a mixed polydactyly of the feet. A mixed and crossed polydactyly is a rare finding with only one other reported case.[8] This is the first report of crossed and mixed polydactyly of the feet presenting with 7 complete toes on each foot without syndactyly. The case also led us to propose a more complete, less confusing classification system.

CASE REPORT

A 1-month-old girl was seen for evaluation of polydactyly of her hands and feet (Fig. 1). The postaxial polydactyly of the hands was treated with suture ligation postnatally before our involvement with the child. The family history included both a maternal grandmother and maternal great grandmother with bilateral polydactyly of the feet. The maternal grandmother was reported to have had 13 toes. The mother was born with 12 toes. Information about the exact pattern of their involvement was not available. Neither had hand involvement.

Physical examination revealed a healthy well-nourished female infant with complete preaxial and postaxial polydactyly of the feet. Small scars were present laterally on both hands from the suture ligation of the rudimentary postaxial duplications of her fingers. The remainder of the upper and lower extremity examination, including the neurological examination, was normal. There were no other orthopaedic or systemic anomalies.

Radiographs of the feet demonstrated preaxial and postaxial polydactyly including the metatarsals (Fig. 2). The right foot had complete duplication of first and fifth metatarsals and digits. The left foot had a complete duplication of the first metatarsal and digit with duplication of the fifth toe with a Y-shaped fifth metatarsal.

Reconstructive surgery was performed at 8 months of age. Both first and seventh digits were excised bilaterally using racquet-type incisions. During removal of the preaxial duplications, the abductor hallucis tendon was preserved and reattached to the remaining hallux. The most lateral arm of the Y-shaped left fifth metatarsal was resected at its branch point. The lateral most sixth metatarsal on the right foot was also excised. The child's recovery

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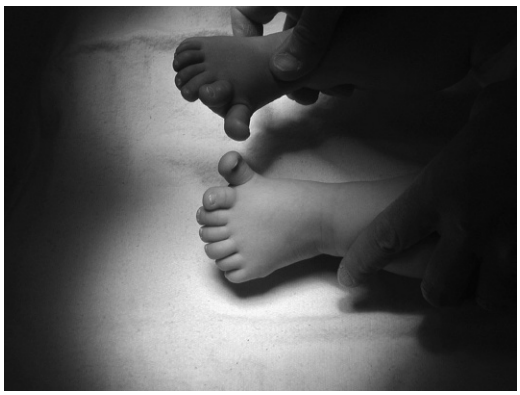

FIGURE 1. Preoperative photograph showing 7 digits bilaterally with both preaxial and postaxial duplication of digits.

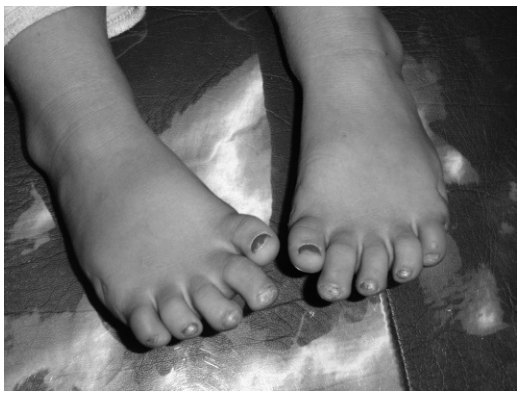

FIGURE 3. Postoperative photograph of both feet at 4 years showing normal contours with the exception of mildly hypoplastic first rays.

was uneventful. At 4 years, cosmesis was excellent, and her function was normal (Fig. 3).

DISCUSSION

This is a rare case and, to our knowledge, the first reported case of a child with crossed and mixed polydactyly with 14 toes without syndactilization. We feel that it is confusing to combine the terms *crossed* and *mixed polydactyly*. The purpose of the term *crossed polydactyly* is to imply that the deformities of the upper extremities are opposite to the lower extremities. If mixed polydactyly, existence of preaxial and postaxial polydactyly on upper or lower extremities, is present, how can there be an opposite orientation on the other set of extremities? Therefore, combining the terms *mixed* and *crossed* is confusing. We propose a simpler and more inclusive classification. This is presented in Table 1. All subtypes are described as a unilateral or bilateral. Type A polydactyly would be preaxial or postaxial and could be unilateral or bilateral in the hands or feet. Type B indicates upper and lower extremity polydactyly, either preaxial or postaxial. Type C represents upper and lower extremity involvement with opposite orientation. This type can be subdivided into C1 and C2. C1 is upper extremity postaxial with lower extremity preaxial polydactyly; C2 is the opposite. Type D has preaxial and postaxial on the same extremity, previously referred to as mixed. Type E is preaxial and postaxial polydactyly of the upper or lower extremities along with preaxial or postaxial deformities in the other set of extremities. Descriptive terms including *rudimentary*, *complete*, *upper and lower extremity* and references to the side involved can still be used within this framework. The term *central ray polydactyly* would retain its current definition.[2] Although this classification has no implications in regards to treatment, it is important for description and delineation. In addition, as we investigate inheritance, it may aid in studying pedigrees and genetic patterns.

Our case is unique with a child having preaxial and postaxial polydactyly of the feet and postaxial polydactyly of the hands. This would be classified as type E in our new system. A review of the literature revealed one similar case. The individual had complete syndactylization of the left great toe and partial syndactylization of the right great toe.[6] There were no cases with complete preaxial and postaxial duplications of the feet without syndactyly.

The genetic disposition for polydactyly is not clearly understood. Most patients relate a positive family history. In a

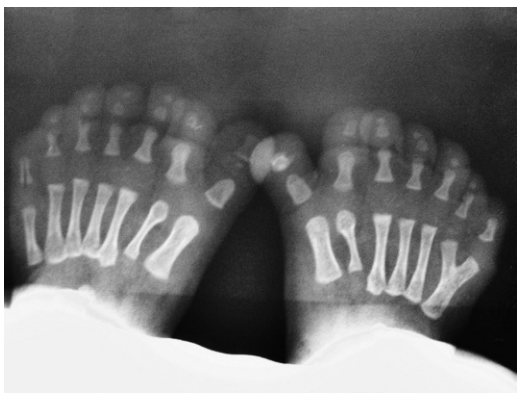

FIGURE 2. Radiograph reveals complete duplication of preaxial and postaxial metatarsals and digits of the right foot and complete duplication of the preaxial metatarsal and digits of the left foot. The postaxial metatarsal of the right foot was Y-shaped with complete duplication of the digit.

TABLE 1. Classification of Polydactyly

Type A	Preaxial or postaxial polydactyly of a hand or foot, unilateral or bilateral
Type B	Preaxial or postaxial polydactyly of the hands and feet, same orientation
Type C	Involvement of hands and feet, opposite orientation
C1:	Upper postaxial, lower preaxial
C2:	Upper preaxial, lower postaxial
Type D	Preaxial and postaxial polydactyly of the same extremity, usually bilateral, notate if unilateral
Type E	Type D polydactyly of the upper or lower extremities with type A of the other set of extremities

study by Venn-Watson of 72 patients, at least 30% had a positive family history of polydactyly.[8] In our case, the patient's mother had type A and maternal grandmother likely had type C polydactyly of the feet. Genetic studies have indicated that each type of polydactyly is a result of a different autosomal dominant gene with irregular modifiers.[3,4] Associated anomalies have been found in individuals with polydactyly, including trisomy 13, Meckel syndrome, and Down syndrome.[7] Down syndrome is strongly associated with first ray duplication, but not with postaxial polydactyly.[9] Our patient had no associated anomalies or syndromes.

It has been recommended that surgical intervention take place between 6 and 12 months of age. This allows the bones and soft tissues to become well defined and reduces the risk of anesthetic complications.[9] Performing surgery before independent ambulation will allow for proper fitting shoes in addition to improved cosmesis. In most cases, the most medial or lateral digit is removed. However, if this digit is more functional, it may be spared and the adjacent digit removed. Care must be used in regard to soft tissues to bring the digit into an appropriate position.[7,8] Syndactylization is sometimes useful.

Complications of polydactyly reconstruction of the medial or preaxial side of the foot include hallux valgus, residual bone or soft tissue, and short first metatarsal with plantar callosities.[7,8] In efforts to avoid hallux valgus during reconstruction, the abductor tendon should be reattached in addition to capsular repair.[7,8] Complications of polydactyly reconstruction of the lateral or postaxial side of the foot include superficial infection, residual bone or soft tissue, wide metatarsal head, wide foot with a gap between fourth and fifth toes, and varus toes.[7,8]

CONCLUSION

This is the first case of polydactyly with 7 complete toes bilaterally and postaxial duplications of the fingers, classified as type E in our proposed new classification system. This system would aid in study and discussion of these deformities. The involved 8-month-old child was treated surgically with an excellent result. She now wears normal shoes and has no functional limitations.

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