The Journal of Foot & Ankle Surgery 58 (2019) 814–820

Contents lists available at ScienceDirect

The Journal of Foot & Ankle Surgery

journal homepage: www.jfas.org

Surgical Reconstruction Technique of Two Patients With Tarsal Type Preaxial Polydactyly: Two True Prehalluces

Daniel H. Ferman, BS[1], Laura D. Bauler, PhD[2], Karen Bovid, MD[3]

[1] *Third-Year Medical Student, Western Michigan University Homer Stryker M.D. School of Medicine, Kalamazoo, MI*
[2] *Assistant Professor, Department of Biomedical Sciences, Western Michigan University Homer Stryker M.D. School of Medicine, Kalamazoo, MI*
[3] *Assistant Professor, Department of Orthopaedic Surgery, Western Michigan University Homer Stryker M.D. School of Medicine, Kalamazoo, MI*

ARTICLE INFO

Level of Clinical Evidence: 4

Keywords:
congenital
extradactyly
foot deformities
hallux abnormalities
supernumerary toe/digit
tarsal type preaxial foot polydactyly
true prehallux

ABSTRACT

Polydactyly of the foot occurs in 1.7 cases per 1000 live births, comprising 45% of congenital abnormalities of the foot. Most reported cases of polydactyly of the foot are postaxial, and 15% are preaxial; of those, tarsal type preaxial polydactyly (a true prehallux) occurs in only 3% of cases. Because of this rarity, there is minimal literature available to guide management or surgical reconstruction. Two newborns presented with similar tarsal type preaxial polydactylies in the context of multiple congenital anomalies at a single institution. Patient 1 presented at birth with an accessory digit arising medially from the right foot at the medial malleolus. Two weeks later, genetically unrelated, patient 2 presented at birth with an accessory digit arising medially from the right foot at the talus. Both patients underwent resection of the extra digit and reconstruction including transfer of the accessory anterior tibial tendon arising from the preaxial extra digit to the remaining first ray. Two years after surgery, both patients are walking well with preserved dorsiflexion strength. Given the rarity of true prehallux cases, reported surgical treatment and outcomes are lacking. This case demonstrates the management of 2 patients to better guide future patient care. Although nonsurgical treatment with shoewear modification is an option, surgical reconstruction facilitated wearing typical shoes while preserving ambulatory ability. Both patients in this series had an accessory anterior tibial tendon. Surgical transfer of the tendon prevented loss of dorsiflexion strength and foot drop postoperatively.

Polydactyly is a common congenital abnormality involving extra digits of the feet or hands. With an overall incidence of 1.7 per 1000 live births, a significant racial disparity exists, with incidences of 0.3 to 1.3 per 1000 live Caucasian births and 3.6 to 13.9 per 1000 live African American births (1–4). There is no difference between sexes, and it is twice as likely to be unilateral than involve bilateral limbs. Polydactyly of the foot can be divided into 3 large classes depending on the location of the accessory digit: preaxial (referring to the medial foot with duplication of the first digit), central (referring to duplication of the second, third, or fourth digits), and postaxial (referring to the lateral foot with duplication of the fifth digit). Of these, the postaxial type is by the far the most prevalent, comprising ~79%; of the remaining cases, the preaxial type comprises ~15% and the central type 6% (3). Polydactyly occurs both in isolated instances and in association with established genetic syndromes and congenital anomalies. Although both environmental and genetic factors play a role in the etiology of polydactyly, currently less is known about the genetic basis for the preaxial type.

Here we highlight the unusual presentation of 2 tarsal subtype preaxial polydactyly cases, both true prehalluces. The term "true prehallux" has come to mean an entire great toe arising from the medial border of the navicular bone (5). Although multiple polydactyly classification systems exist, the Watanabe classification system—based on both ray involvement and level of duplication—is used in this report for both its direct inclusion of the true prehallux as a subtype and its usefulness for surgical planning (3,6). Because of the rarity of preaxial polydactyly cases, the existing surgical literature is limited; this case report discusses the surgical technique used for these 2 patients to guide future surgical treatment.

Case Report

Timeline

Two patients presented at birth, within 2 weeks of each other, each with multiple congenital anomalies and right foot tarsal type preaxial polydactyly. Treatment occurred between June 2014 and February

Financial Disclosure: None reported.
Conflict of Interest: None reported.
Address correspondence to: Laura D. Bauler, PhD, Department of Biomedical Sciences, Western Michigan University Homer Stryker M.D. School of Medicine, 1000 Oakland Drive, Kalamazoo, MI 49008.
E-mail address: laura.bauler@med.wmich.edu (L.D. Bauler).

1067-2516/$ - see front matter © 2018 by the American College of Foot and Ankle Surgeons. All rights reserved.
https://doi.org/10.1053/j.jfas.2018.12.011

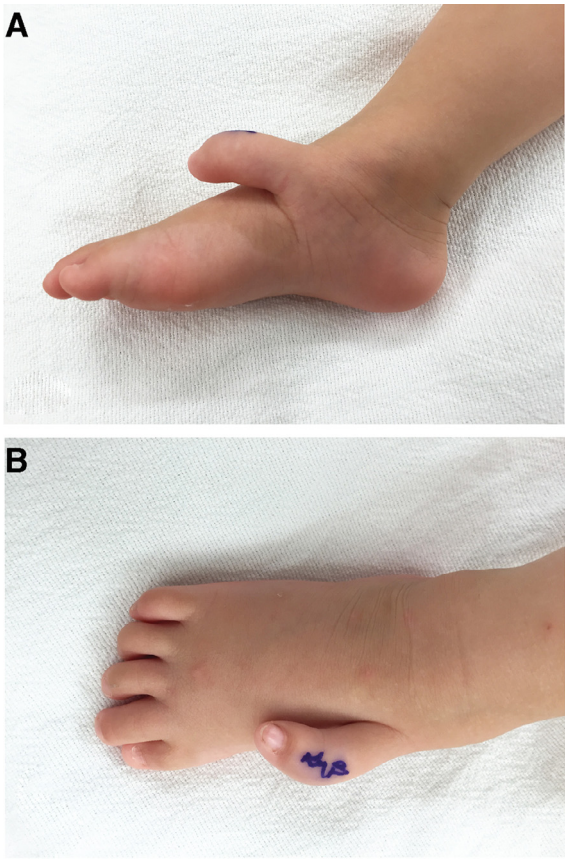

Fig. 1. Patient 1, preoperative clinical photos. Medial (*A*) and dorsal (*B*) aspects of the right foot at age 13 months.

2018. Consent was obtained from the parents of both patients for use of the images and information contained in this report.

Patient 1

A female, born at 39 weeks by cesarean delivery, was diagnosed with congenital scoliosis with rib fusions, a right scapular malformation, left thumb hypoplasia, bilateral developmental dysplasia of the hip, and right foot tarsal type preaxial polydactyly. At her 13-day-old presentation to clinic, the hallux was hypoplastic, and the extra digit was noted on the medial proximal side of the right foot, appearing to articulate with the ankle near the medial malleolus. At her 9-month visit, the digit had independent extensor tendon motion, noted to move over the dorsum of the right foot. Surgery was presented as an option to mitigate shoewear problems and cosmetic strain, even though the toe was not expected to cause difficulty with gait. At 1 year of age, radiographs demonstrated preaxial polydactyly with ossification of an apparent metatarsal and distal phalanx in the extra digit and a normal-appearing remainder of the foot (Figs. 1 and 2).

A right foot preaxial polydactyly removal with reconstruction and tendon transfer was performed at 13 months of age. Postoperative visits showed a well-healed right foot, and the patient was walking at 6 weeks postoperation with full function of the foot and ankle. She was able to walk, run, and climb without difficulty or pain at 7 months postoperation. At 3 years of age (~2 years postoperatively), she ambulated well with a heel-to-toe gait. Physical examination demonstrated normal ankle and hindfoot range of motion; stability to anterior drawer, inversion, and eversion testing; physiologic hindfoot alignment and arch; and hypoplasia of the remaining great toe, as well as mild hypoplasia of the right foot and calf (Fig. 3). Dorsiflexion strength was 5 of 5. Radiographs demonstrated appropriate alignment and development of the medial malleolus and first ray, with mild hypoplasia of the first metatarsal and phalanges (Fig. 4).

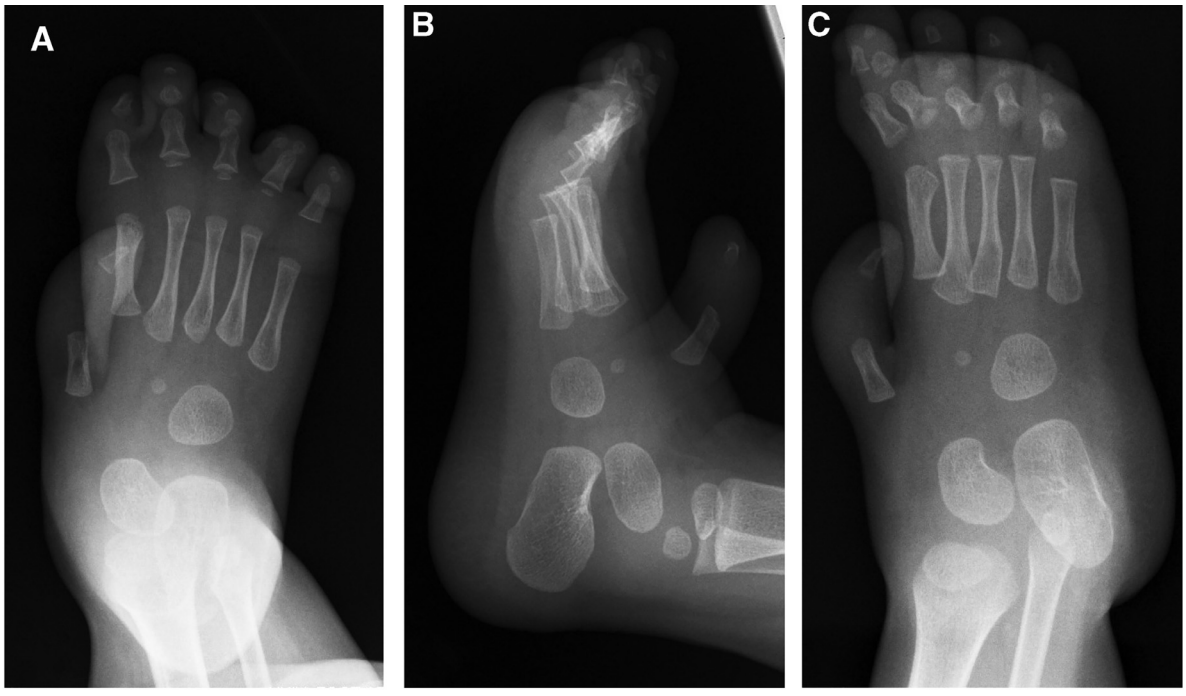

Fig. 2. Patient 1, preoperative radiographs. Anteroposterior (*A*), lateral (*B*), and oblique (*C*) radiographs of the right foot at age 12 months.

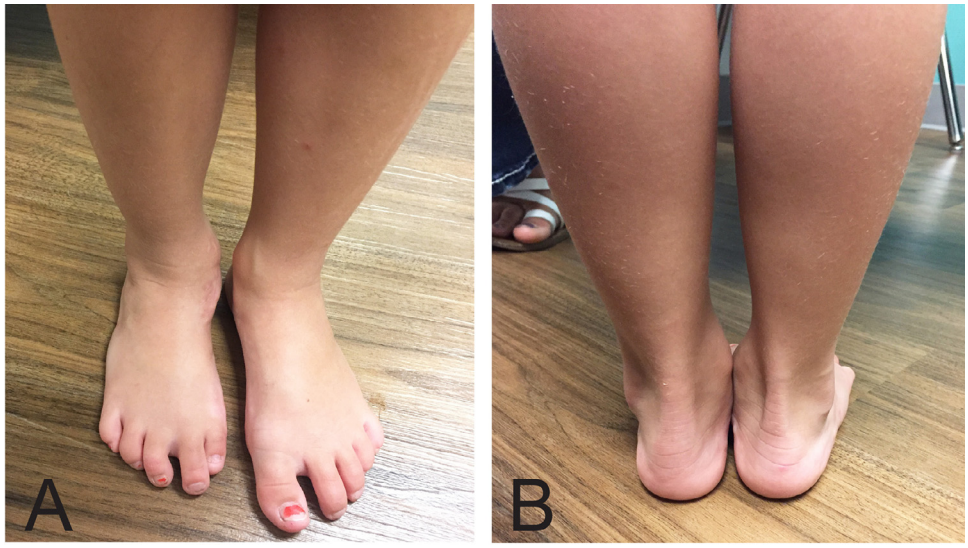

Fig. 3. Patient 1, 2-year postoperative clinical images. Dorsal feet (*A*) and hindfoot (*B*) at age 3 years.

Patient 2

A male born prematurely, at 35 weeks' gestational age, was diagnosed with VACTERL (vertebral defects, anal atresia, cardiac defects, tracheoesophageal fistula, renal anomalies, and limb abnormalities) syndrome including tracheoesophageal fistula (esophageal atresia and stenosis), multicystic kidney disease, a ventricular septal defect, inguinal and umbilical hernia, a single umbilical artery, bilateral hypoplastic thumbs, right foot tarsal type preaxial polydactyly (Fig. 5), and a Gollop-Wolfgang complex consisting of a left bifid femur with complete

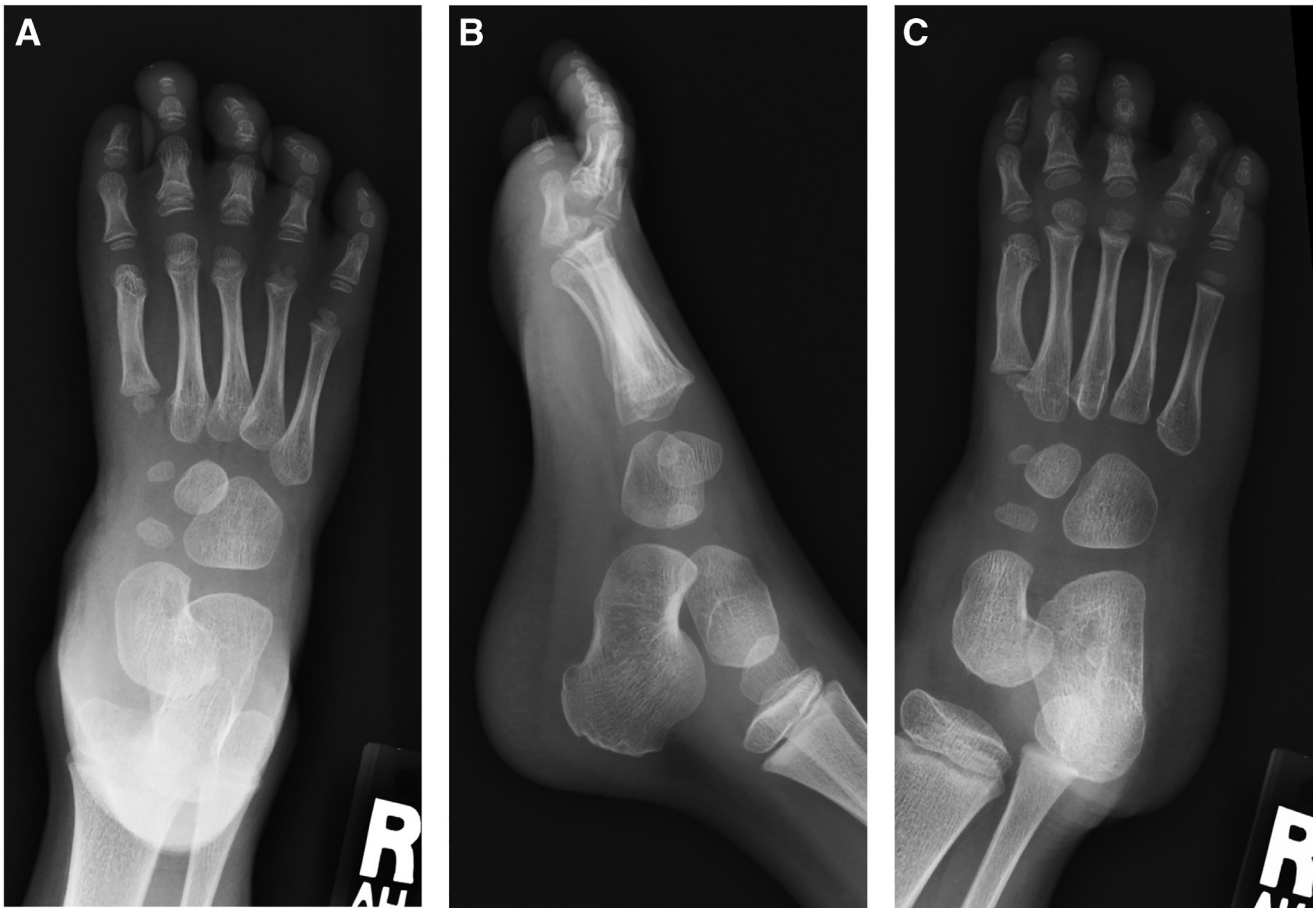

Fig. 4. Patient 1, 2-year postoperative radiographs. Anteroposterior (*A*), lateral (*B*), and oblique (*C*) radiographs of the right foot at age 3 years.

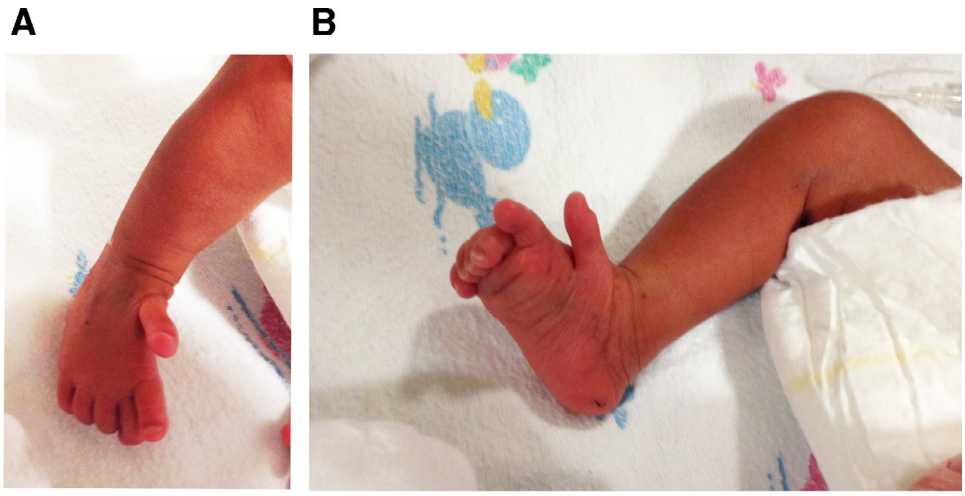

Fig. 5. Patient 2, newborn clinical images. Dorsal (*A*) and plantar (*B*) medial aspects of the right foot with the patient actively extending the prehallux.

tibial hemimelia. At 12 months of age, his right hindfoot and ankle had normal alignment and flexibility. The hallux was hypoplastic, and the extra digit was present over the level of the first metatarsal medially, appearing to articulate with the navicular or talus. The extra digit could flex and extend voluntarily. There was good capillary refill, sensation to light touch was intact over the right foot, and a dorsalis pedis pulse was palpable. Radiographs demonstrated a normal-appearing foot with the addition of the preaxial accessory digit, with only the distal phalanx ossified at that time (Fig. 6). At 16 months, the patient, despite attempts, was unable to stand secondary to his lower limb deformities.

At 19 months of age, a left through-knee amputation, femoral osteotomy, and resection of medial distal femur concurrently with the right preaxial polydactyly reconstruction were performed under the same anesthesia. The 2-week postoperative visit showed well-healed surgical incisions with active dorsiflexion of the right foot and grossly intact sensation. The patient continued to progress and was able to crawl by 4 months postoperation, with sensation and motor function grossly intact on the right foot, active dorsiflexion of the right ankle, a palpable dorsalis pedis pulse, and brisk capillary refill. By 2 years and 3 months of age, he was able to pull himself to stand and remain standing for up to 20 minutes with the help of his walker. A supramalleolar orthosis (SMO) was delivered for the right foot because slight valgus hindfoot alignment was noted while standing. Two years postoperation, the patient was ambulating well with a right SMO and left lower extremity prosthesis without need of an assistive device. There was mild hypoplasia of the hallux (Fig. 7). He stood with flexible hindfoot valgus and had full ankle and hindfoot range of motion. Dorsiflexion strength was 5 of 5. Radiographs of the right foot demonstrated mild hypoplasia of the great toe proximal and distal phalanges with normal alignment and appearance of the navicular ossification center (Fig. 8).

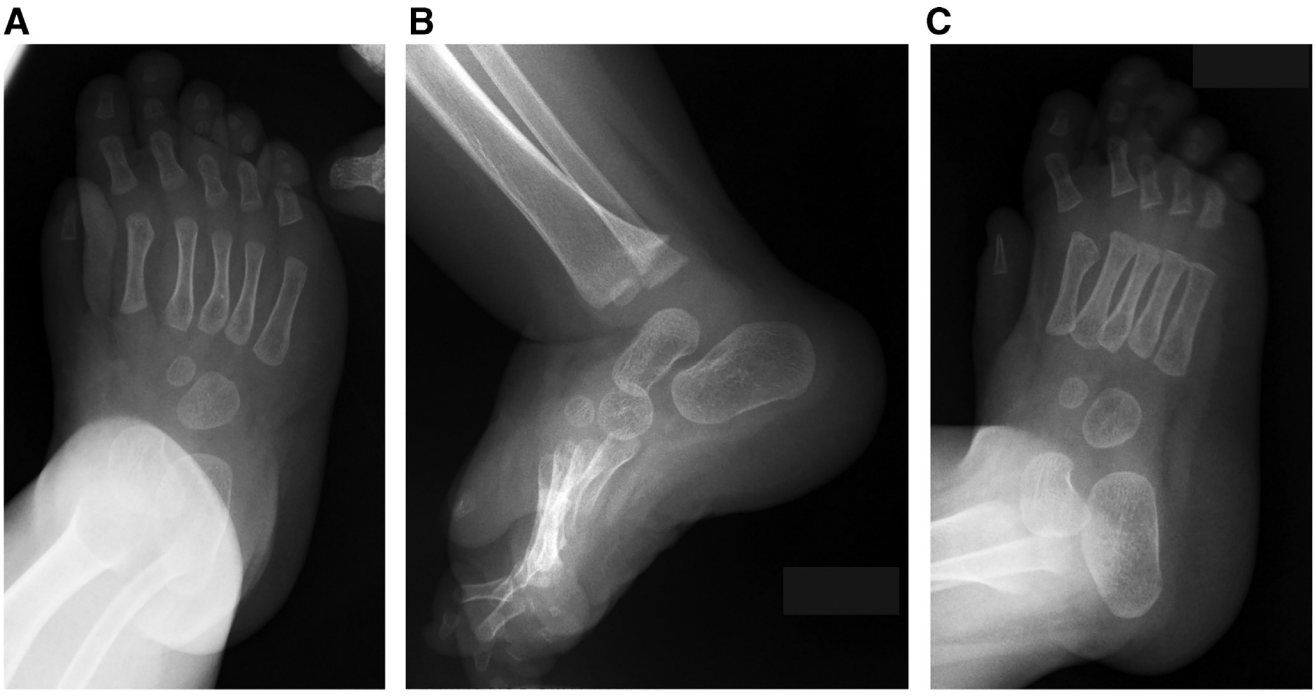

Fig. 6. Patient 2, preoperative radiographs. Anteroposterior (*A*), lateral (*B*), and oblique (*C*) radiographs of the right foot at age 12 months.

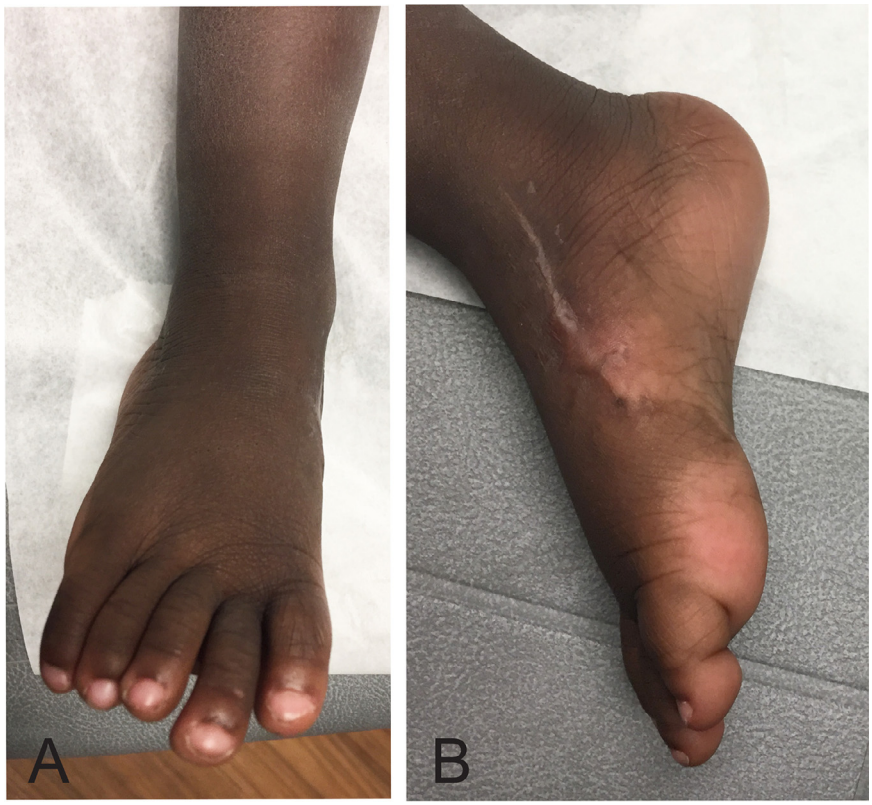

Fig. 7. Patient 2, 18-month postoperative clinical images. Dorsal (*A*) and medial (*B*) aspects of the right foot at age 3 years.

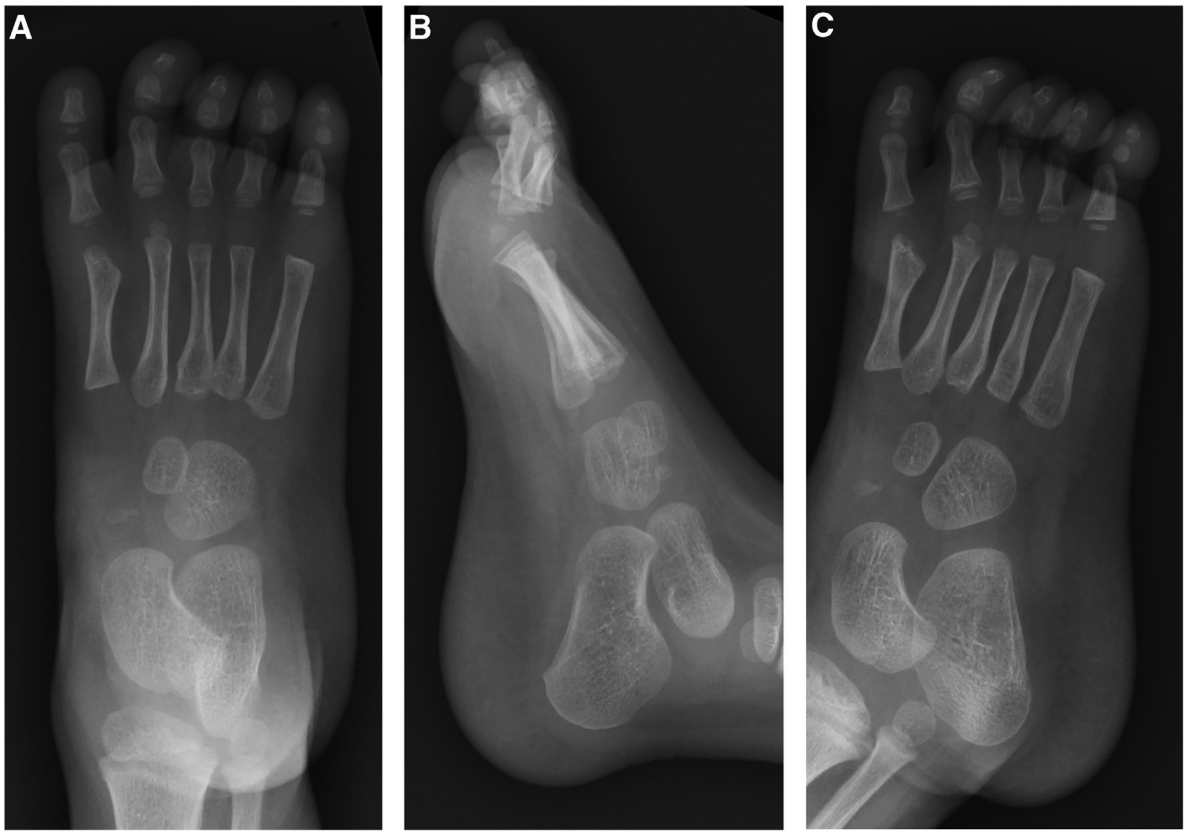

Fig. 8. Patient 2, 18-month postoperative radiographs. Anteroposterior (*A*), lateral (*B*), and oblique (*C*) radiographs of the right foot at age 3 years.

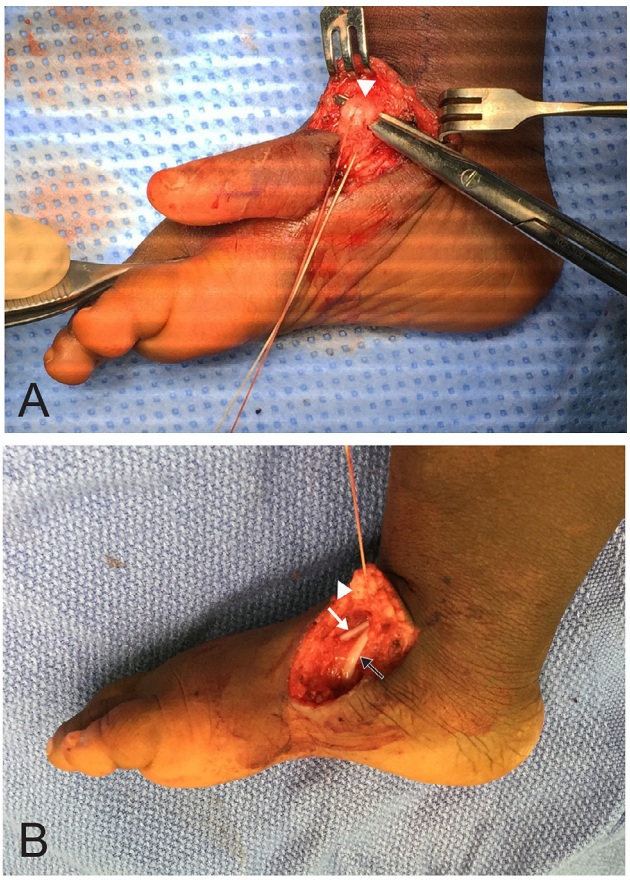

Fig. 9. Patient 2, intraoperative images highlighting anterior tibial tendons. (*A*) Accessory anterior tibial tendon over the instrument (white arrowhead) tagged with suture as it inserts on the prehallux. (*B*) Extensor hallucis longus (white arrow) and anterior tibial (black arrow) tendons. The accessory anterior tibial tendon (white arrowhead) has been tagged with suture and resected from the prehallux. Note the larger diameter of the accessory tendon compared with the anterior tibial tendon.

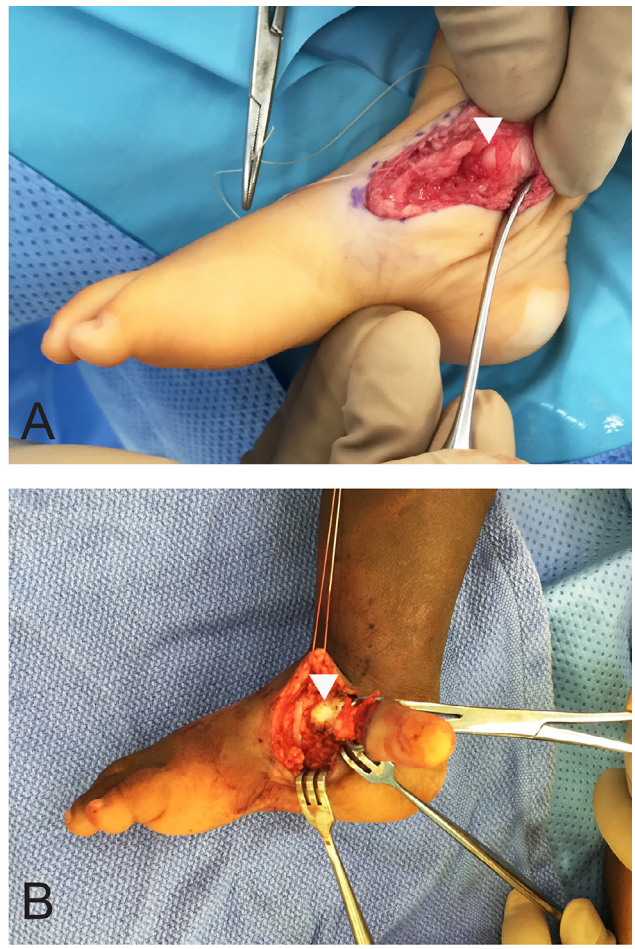

Fig. 10. Intraoperative images highlighting articular facets. Articular facets (white arrowheads) in patient 1 (*A*) on the anterior aspect of the medial malleolus where the preaxial digit articulated and in patient 2 (*B*) at the articulation of the prehallux with the talus.

Surgical Procedures

The procedures were performed with the patients under general anesthesia, positioned supine, and using a thigh tourniquet. A longitudinal incision was made over the medial foot and ankle with a racquet around the base of the extra digit in both patients. What appeared to be an accessory anterior tibial tendon was found inserting in both accessory digits in the region of the metatarsal. This anomalous tendon was significantly larger in patient 2 than the anterior tibialis tendon that coursed over the midfoot and inserted on the remaining first ray (Fig. 9). The anomalous tendons were divided, keeping as much length as possible, and later repaired to the periosteum over the medial cuneiform in both patients, adjacent to the more normal-appearing tendon.

The accessory digit of patient 1 was found to contain 4 bones: a distal phalanx, a proximal phalanx, a metatarsal, and a second more proximal long bone that appeared to articulate with the anterior portion of the medial malleolus. At this joint on the enlarged anteromedial aspect of the medial malleolus was an area of cartilage appearing to be an articular facet (Fig. 10*A*), which was sharply divided after disarticulation to provide a more normal contour. Care was taken to stay anterior to the deltoid ligaments, which were left intact. Mini C-arm fluoroscopy of the removed digit confirmed ossification of the distalmost phalanx and the metatarsal, whereas the proximal phalanx was nonossified and the proximal extra long bone remained cartilaginous. No extra bones remained attached to the foot via C-arm fluoroscopy or via direct inspection and palpation.

The accessory digit of patient 2 was found to contain 5 bones: a distal phalanx, a proximal phalanx, a long bone consistent with a metatarsal, a second more proximal long bone, and a proximal-most very small round bone that appeared to articulate with the head of the talus (Fig. 10*B*). This joint was disarticulated, and cartilage surfaces were visualized. After transferring and repairing the accessory anterior tibial tendon to the remaining first ray, the wound was irrigated and the incision closed in standard layered fashion. A sterile bandage and short leg cast were applied. Weightbearing was allowed as tolerated in the cast for 6 weeks. The cast was then removed and ambulation begun as tolerated in standard shoewear.

Discussion

To date, only a few cases of tarsal type preaxial polydactyly have been published in the literature. In a review of all preaxial polydactyly cases encountered at a single institution over 30 years, of the 22 patients (36 feet) that had medial ray polydactyly, only 3% (1 foot) were found to be of the tarsal type (3). Since the original 1966 Cobey and Cobey (5) case of the true prehallux, at least 4 additional tarsal type preaxial polydactyly cases have been reported: a 1979 case in California of a patient with a single accessory phalanx and metatarsal articulating with the

medial cuneiform, a 2014 case in Iran of an adult patient with an accessory digit deviating from the tarsometatarsal joint, a 2015 case in Japan of a patient with 2 accessory phalanges articulating with the medial border of the navicular bone, and a 2016 case in Maryland of a rare combined tarsal and metatarsal type preaxial polydactyly with 2 accessory digits (5,7–10). This highlights the rarity of cases that present with a true prehallux and thus a limited amount of surgical guidance.

Whereas most cases of true prehalluces in which tendons were found attached to the prehallux identified accessory tibialis posterior tendons, with the exception of the 2015 Okumoto et al (9) case, both patients we present in this case report had accessory tibialis anterior tendons. Preservation and transfer of the accessory extensor tendon, rather than simple surgical resection of the extra digit with release of the accessory tendon, was elected with the hope of maximizing both patients' dorsiflexion strength and preventing foot drop, especially in the instance that the extra tendon appeared to be the dominant dorsiflexor of the ankle. Tendons were divided during surgery, keeping as much length as possible, and later repaired to the periosteum over the medial cuneiform in both patients, adjacent to the more lateral, nonanomalous anterior tibial tendon. In patient 2, we believe the transferred tendon to be the dominant dorsiflexor, although on neither patient did the dissection extend far enough to reveal either the body of the muscle attached to the tendons or the location of the full insertion of the nonaccessory tendon. For patient 2, the accessory tendon was much larger than the anterior tibial tendon attaching to the nonanomalous first ray and was therefore presumed to have larger muscle belly associated with it.

General surgical guidelines involved resection of the projecting toe with preservation of the digit that best aligned axially with the foot, rebalancing of soft tissues, and reduction of medial tarsal prominences. Indications were poor shoe fit and cosmetic concerns. Surgery was delayed in both patients until further skeletal ossification could occur within the affected ray to allow accurate assessment of the anatomy during the procedure and to limit risks of anesthesia by performing foot reconstruction under the same anesthesia as other planned procedures, preferably closer to 1 year of age. Postoperative casting and taping were used to prevent angular deformities and encourage normal contour.

Our patients are also unique for the bones found within the accessory digits. The inclusion of a second long bone proximal to the metatarsal in both cases, with an additional small round bone in patient 2, is inconsistent with previous reports of prehalluces. The 2 accessory digits also differ in their articulation with the rest of the foot, appearing to do so with the anterior portion of the medial malleolus in patient 1 and the head of the talus in patient 2. This upsets the traditional definition of a true prehallux as an accessory digit arising from the medial border of the navicular; it is reasonable to speculate that the 4 bones represent 2 accessory phalanges, an accessory metatarsal, and either an accessory medial cuneiform or an accessory navicular bone, whereas the fifth bone represents an accessory navicular bone.

On an interesting note, the finding of articular facets may support the well-accepted evolutionary theory of the prehallux as originating in lower vertebrates. Mammals such as the opossum and echidna retain this descendent of a sixth toe, which when seen in humans often has true articulation including hyaline cartilage and ligaments, differentiating it from a sesamoid bone (11).

In conclusion, the fact that both patients presented within 2 weeks of each other, born at the same hospital, is most likely coincidental, although it is of course possible that in combination with genetic predisposition, there were environmental factors in common. There is evidence that environmental factors such as maternal diabetes are involved in preaxial polydactyly; however, it is difficult to distinguish such factors from underlying genetic syndromes. In such cases, both obtaining a complete family history for congenital anomalies and consultation with a clinical geneticist are warranted. When planning surgical reconstruction of a tarsal type preaxial polydactyly, we recommend evaluation for an accessory anterior tibial tendon and, if present, transferring it to the remaining first ray to preserve dorsiflexion strength.

References

1. Venn-Watson EA. Problems in polydactyly of the foot. Orthop Clin North Am 1976;7:909–927.
2. Phelps DA, Grogan DP. Polydactyly of the foot. J Pediatr Orthop 1985;5:446–451.
3. Belthur MV, Linton JL, Barnes DA. The spectrum of preaxial polydactyly of the foot. J Pediatr Orthop 2011;31:435–447.
4. Woolf CM, Myranthopoulos NC. Polydactyly in American Negroes and whites. Am J Hum Genet 1973;25:397–404.
5. Cobey MC, Cobey JC. A true prehallux. The first to be described in the literature. J Bone Joint Surg Am 1966;48:953–954.
6. Watanabe H, Fujita S, Oka I. Polydactyly of the foot: an analysis of 265 cases and a morphological classification. Plast Reconstr Surg 1992;89:856–877.
7. Rao BR. Supernumerary toe arising from the medial cuneiform. A case report. J Bone Joint Surg Am 1979;61:308.
8. Jalili A. Preaxial polydactyly of foot, report of a rare case. Shafa Ortho J 2014;1:e24623.
9. Okumoto T, Aramaki-Hattori N, Taguchi Y, Inoue Y, Matsuzaki K, Kishi K. A case report of true prehallux. JPRAS Open 2015;4:26–29.
10. Granite G, Herzenberg J, Wade R. Rare case of tibial hemimelia, preaxial polydactyly, and club foot. World J Clin Cases 2016;4:401–408.
11. Kidner FC. The prehallux (accessory scaphoid) in its relation to flat-foot. J Bone Joint Surg 1929;11:831.