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Case Reports and Series

Minimally Invasive Hallux Interphalangeal Joint Arthrodesis for Hallux Varus in Pfeiffer Syndrome: A Case Report

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ABSTRACT

Pfeiffer syndrome is a rare hereditary condition with an autosomal dominant transmission caused by a mutation that affects fibroblast growth factor receptors. It is one of the acrocephalosyndactyly diseases causing cranial malformations owing to early suture fusion. In the foot, it is typically associated with hallux varus, first ray hyperplasia, and partial lesser digit syndactyly. We report a clinical case of a 10-year-old patient with Pfeiffer type I syndrome with bilateral severe hallux varus due to a hypoplastic trapezoidal shaped proximal phalanx, a distal, medial-facing articular surface, and interphalangeal instability. This deformity was addressed by minimally invasive hallux interphalangeal joint arthrodesis with internal and external fixation. We report the results at the 2-year follow-up point.

First described in 1964, Pfeiffer syndrome is a rare autosomal dominant disease that occurs in approximately 1 of every 100,000 live births. It is characterized, in the classic Pfeiffer type 1 described in our patient, by centrofacial hypoplasia, hallux varus, broad thumbs, large toes, and partial lesser digit syndactyly (1–5). In the more severe Pfeiffer type 2 and 3 syndromes, the disease is less benign and can cause cloverleaf skull, severe exophthalmos, hydrocephalus, neurologic and intellectual impairment, a variable spectrum of visceral abnormalities, and, even, premature death.

To the best of our knowledge, the present study is first reported surgical case describing a minimal incision interphalangeal arthrodesis for hallux varus in a patient with Pfeiffer syndrome. We believe this technique is a useful option in the management of this pathology.

Case Report

We present the case of a 10-year-old female patient with Pfeiffer type 1 syndrome with a history of previous cranioplasty to correct craniosynostosis when she was 6 months old and hand surgery for hyperplastic thumbs at 5 years of age.

Her present complaint was the inability to wear enclosed shoes owing to bilateral severe hallux varus. She had also syndactyly between the second and third toes (Fig. 1), subtle centrofacial hypoplasia, and slight hypoacusis in the left ear. The physical examination was otherwise normal, and she was not nor had been taking any medications for this condition. She had no history of allergies or hypersensitivity.

Radiographic imaging showed that her hallux varus was secondary to severe interphalangeal deformity with a normal metatarsophalangeal joint and angle but an increased intermetatarsal (IM) angle with the first metatarsal's distal edge extending past the second metatarsal (Figs. 2 and 3). The first phalanx was hypoplastic and trapezoidal in shape. The second phalanx was hyperplastic. This combined deformity was responsible for the severe incongruency and instability.

The patient underwent surgery in August 2014 to both feet. A minimal incision hallux interphalangeal joint arthrodesis with an internal cannulated screw and percutaneous Kirschner wire fixation was performed bilaterally. Because of the first ray hyperplasia, growth arrest of the second phalanx of the hallux was not an issue. In addition, the physis of the second and third phalanxes of the lesser toes had already closed. The remaining physis of the proximal phalanx of the hallux and lesser digits were symmetrical (Fig. 2) and nearly closed (Fig. 6).

The patient was placed supine on the operating table after induction of general anesthesia. No tourniquet was used. Under fluoroscopy a 1.5-mm Kirschner wire was passed on the medial aspect of the second phalanx of the hallux, just distal to the physis and parallel to the desired joint arthrodesis orientation (Fig. 4A). A 0.3-cm minimal incision was

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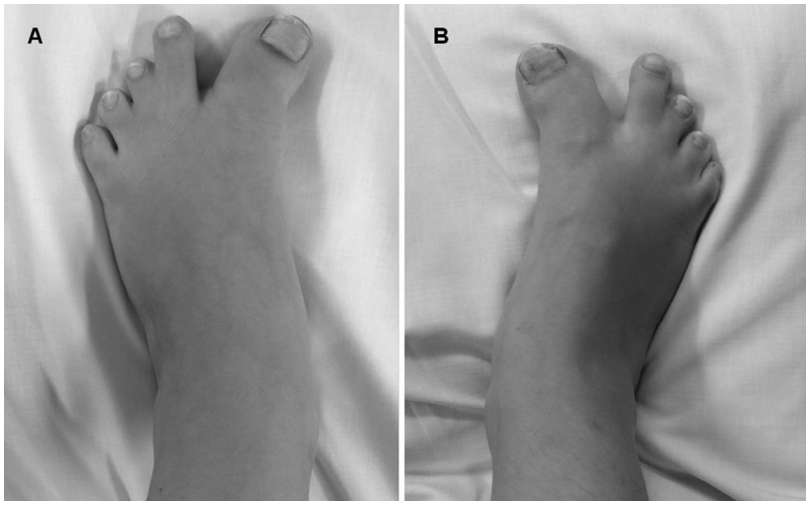

Fig. 1. Preoperative aspect of *(A)* left and *(B)* right foot showing bilateral hallux varus.

then placed proximally using a Beaver no. 64 blade. Next, using a high-torque, low-speed micromotor, a Shannon 2 × 12-mm burr was introduced proximally and parallel to the Kirschner wire, removing the physis and creating the distal flat surface for arthrodesis (Fig. 4B). A small percutaneous DPR rasp was used to extricate the cartilaginous fragments.

A similar procedure (Kirschner wire-guided percutaneous burring) was performed in the distal portion of the first phalanx (Fig. 4C), creating the proximal articular surface for arthrodesis, with both surfaces (proximal and distal) aligned with the first ray axis. Fixation was achieved using a double-compression 4-mm cannulated screw and a 1.5-mm Kirschner wire (Fig. 4D,E). A 3-0 nylon suture was used for skin closure, and the wounds were dressed with sterile compression dressings and forefoot offloading shoes.

The patient was discharged the next day. The sutures were removed 2.5 weeks after the surgery. The Kirschner wires were removed 8 weeks later. At that point, the patient started using regular shoes. She was allowed to run 4 months after surgery when clinical and radiologic evidence of arthrodesis was seen. At 14 months after the arthrodesis procedures, the cannulated screws were removed, and the second and third digit syndactyly deformities were corrected by plastic surgery (Fig. 5).

This patient had bilateral, simple, and incomplete syndactyly between the second and third toes. The goals were the creation of a web space with appropriate skin coverage without any secondary contractures. Surgery consisted of a dorsal "omega" skin flap for web space reconstruction and 2 triangular flaps for plantar and lateral skin coverage.

The final result, 24 months after the hallux surgery and 8 months after the syndactyly correction, was very satisfactory. The patient was fully satisfied without any walking or sports limitations. The alignment was adequate, bone healing had occurred between the proximal and distal phalanges, and the reduction of the hallux varus deformities was satisfactory (Figs. 6 and 7).

Discussion

Pfeiffer syndrome is a rare hereditary condition with an autosomal dominant transmission caused by a mutation in the fibroblast growth factor receptors (FGFR1 and FGFR2) (2). It is one of the acrocephalosyndactyly diseases, causing cranial malformations owing to early suture fusion, and, in the foot, it is typically associated with hallux varus, first ray hyperplasia, and partial lesser digit syndactyly

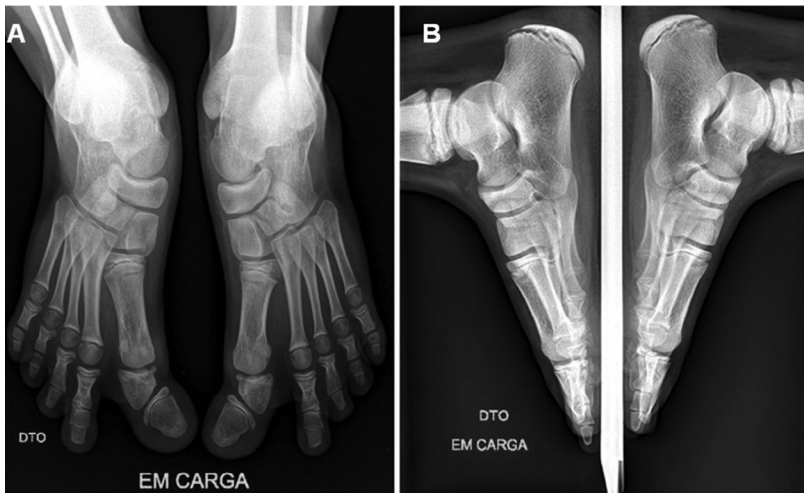

Fig. 2. Preoperative *(A)* anteroposterior and *(B)* lateral radiographs showing physis of the proximal phalanx of the hallux and lesser digits are symmetrical.