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CASE REPORT
Hand/Peripheral Nerve

Reconstruction of a Monodactylous Hand with Microsurgical Free Foot-to-Hand Transfer in Split-Hand/Split-Foot Malformation with Tibial Aplasia

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Summary: Split-hand/split-foot malformation with long bone deficiency (SHLFD syndrome) is a rare congenital disorder, which may be sporadic or autosomal dominant with incomplete penetrance. When complete tibial aplasia is seen, the mainstay of treatment is amputation and lower limb prosthesis. This rare constellation of congenital differences presents an opportunity for microsurgical free tissue transfer using the principle of "spare parts" to improve the functionality of the hand. We present a rare case of split-hand/split-foot malformation with a monodactylous right hand and complete tibial aplasia, treated with microsurgical free foot-to-hand transfer at the time of lower limb amputation, reconstructing key pinch. At the latest 8 months follow-up, the patient had no pain, active key pinch, and ambulated independently with prostheses. He was able to use his right hand independently for a number of daily activities, such as stacking blocks, drinking from a cup, and playing with toys.*(Plast Reconstr Surg Glob Open 2020;8:e2356; doi: 10.1097/GOX.0000000000002614; Published online 28 February 2020.)*

Split-hand/split-foot malformation with long bone deficiency (SHLFD syndrome) is a rare congenital disorder.[1] The complete expression of this disorder consists of deficiency of the central rays and complete tibial aplasia.[2,3]

Adactylous or monodactylous hands do not allow for grasping or pinching, and have limited function.[4] Complete tibial aplasia is typically treated with amputation and lower limb prosthesis.[5] This rare constellation of congenital differences presents an opportunity for microsurgical free tissue transfer using the principle of "spare parts" to improve functionality of the hand. We present such a case treated with microsurgical free foot-to-hand transfer at the time of lower limb amputation, reconstructing key pinch.

CASE REPORT

History and Examination

A 6-year-old boy presented to our clinic for evaluation of split-hand/split-foot malformations with longitudinal deficiencies of all four limbs. He was the second-born child of parents of a non-consanguineous marriage. The antenatal period was uneventful with no maternal illness or drug use. Limb malformations were diagnosed by fetal scan at 8 months of gestation. The patient was born via normal vaginal delivery.

On examination, the right hand had a single radial-sided digit with a stable basilar joint and restricted interphalangeal motion (Fig. 1). A rudimentary digit was attached to the distal forearm by soft tissue. No sensory deficits were noted. Radial artery pulsations were palpable.

The patient ambulated using bilateral prostheses with his knees hyperflexed in the sockets. Both legs were hypoplastic with nonfunctional knee and ankle joints (See **figure, Supplemental Digital Content 1**, which displays clinical photograph showing congenital anomalies of all four limbs in patient with SHLFD syndrome, http://links.lww.com/PRSGO/B284). Each foot had two toes. No craniofacial, chest, spine, genitourinary, or anal abnormalities were seen.

Imaging Studies

Plain radiograph of the right-hand shows a single well-formed thumb (Fig. 2). Plain radiograph of the right lower extremity shows aplasia of the patella and tibia and hypoplasia of the foot (See figure, **Supplemental Digital Content 2**, which displays plain radiograph of the

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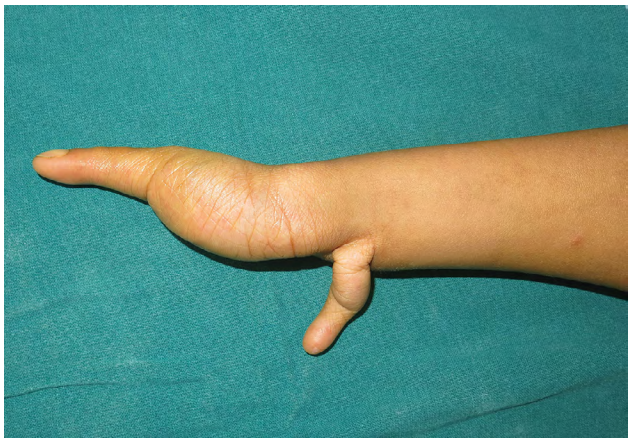

Fig. 1. Clinical photograph of the right-hand show one well-formed radial-sided digit with a rudimentary digit proximal to the wrist crease.

right lower extremity shows aplasia of the patella and tibia and hypoplasia of the foot, http://links.lww.com/PRSGO/B285).

Surgical Treatment

After discussion with the pediatric orthopaedic team and the patient's parents, a decision was made to pursue bilateral above-knee amputations to allow for prosthetic fitting and early mobilization. Right foot transfer from the amputated limb to the right hand was planned to create an ulnar post for key pinch (**see Video** [online], which displays (1) preoperative planning for positioning of the transferred foot to ensure that the thumb can meet the new ulnar post, (2) postoperative key pinch, and (3) improved functional use of the right hand, as seen by the ability to stack multiple blocks, following free foot-to-hand transfer).

Surgery was performed under general anesthesia and tourniquet control. Structures were identified in the recipient wrist. Volarly, the radial artery, ulnar artery, median nerve, and flexor tendons were identified. Dorsally, two subcutaneous veins, one digital nerve, and an extensor tendon were identified. The floating digit was explored and found to have one digital nerve, which was preserved. An area was prepared at the wrist for attachment of the foot.

Since the distance from the wrist to the finger-tip was 10 cm, we planned to transfer the entire foot (Video). The posterior tibial artery and nerve, plantarflexor tendon, dorsiflexor tendon, digital nerve, and three dorsal veins were identified. The tendons were found fused as one bunch (See **figure, Supplemental Digital Content 3,** which displays intraoperative photographs showing dissection of the posterior tibial artery and nerve, http://links.lww.com/PRSGO/B286). The foot was detached and transferred to the hand, and fixation was achieved with one 1.25 mm K-wire. The new cleft created became the webspace (See **figure, Supplemental Digital Content 4**, which displays intraoperative photographs showing transfer of the foot to the hand, http://links.lww.com/PRSGO/B287) The two veins were anastomosed. Respective tendons were repaired. The posterior tibial artery was anastomosed to the radial artery in end-to-end fashion, and on clamp release, the reconstructed hand pinked up immediately (See **figure, Supplemental Digital Content 5**, which displays intraoperative photographs showing pink, perfused digits after vascular anastomoses, http://links.lww.com/PRSGO/B288). Ischemia time was 3 hours. The nerve of the foot was coapted to the rudimentary ulnar nerve and the nerve to the floating digit.

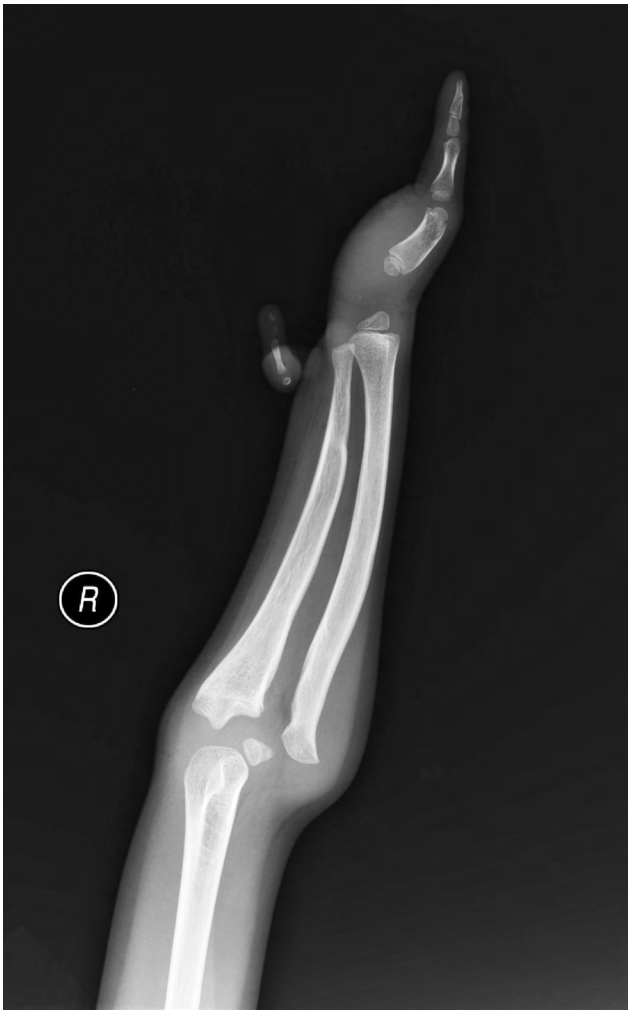

Fig. 2. Plain radiograph of the right upper extremity shows a single well-formed thumb, two bones of the forearm, and congenital radial head dislocation.

Above-knee amputation was completed proximal to the distal femoral physis. The floating digit was excised. The skin flaps of the right hand were closed with the help of split-thickness skin grafts. Plaster immobilization was used. Total procedural time was 10 hours.

Postoperative Course

Postoperative monitoring was uneventful. The patient underwent staged left above-knee amputation during the same hospitalization. At one month, The K-wire was removed in clinic, and he was transferred into a

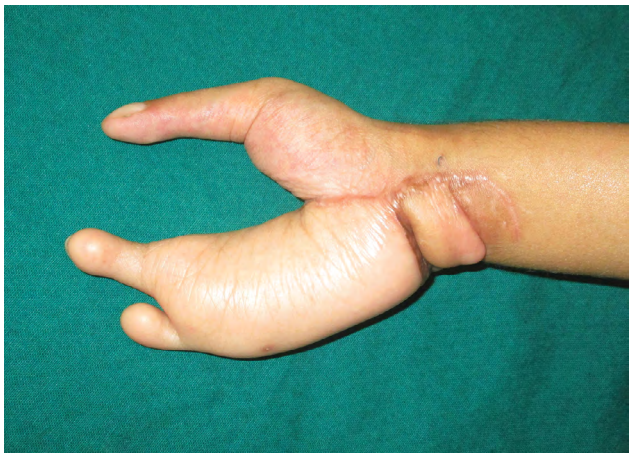

Fig. 3. Postoperative photograph of the right hand.

thermoplastic splint. He was able to touch his thumb to the transferred toes, and physiotherapy was begun for mobilization. At 5 months, new above-knee prostheses were fitted and gait training was initiated.

At the latest 8 months follow-up, the patient had no pain and active key pinch. He was able to use his right hand independently for a number of daily activities such as stacking blocks, drinking from a cup, and playing with toys (Fig. 3 and Video) (See **figure, Supplemental Digital Content 6**, which displays postoperative plain radiograph of the right hand showing a foot-to-hand transfer at the level of the tarsus, http://links.lww.com/PRSGO/B289). Key pinch strength was 0.5 kg compared with 3.2 kg on the contralateral hand. Both static and moving two-point discrimination were 9 mm on the transferred foot compared with 3 mm on the thumb. He walked independently with bilateral above-knee prostheses.

DISCUSSION

The use of multiple toe transfers to reconstruct hand deficiencies has been reported.[6,7] Consideration of transfer of spare parts is especially salient in the setting of concurrent lower extremity amputation.[8] The free, microsurgical transfer of an entire foot to the hand is exceedingly rare. Somerville et al. reported a foot-to-hand transfer in a 2-year-old patient who sustained a severe lawn-mower foot injury with independent use at 6-year follow-up.[9] Hashem and Al-Qattan reported a foot-to-hand transfer in an 8-month-old patient with congenital differences of all four limbs with a good functional outcome but guarded aesthetic outcome at 18-month follow-up; a subsequent commentary by Ogino raises the question of optimal timing of complex microsurgical reconstruction.

In our case, good key pinch function was achieved by foot-to-hand transfer. Since the transferred part was hypoplastic and expected to have little movement, care was taken in preoperative planning to place the ulnar post in a position such that the native thumb could meet it in lateral pinch. The age of the patient allowed us to best position the transferred foot and allowed him to participate in the postoperative rehabilitation. Care should be taken to harvest adequate lengths of vessels, nerves, and tendons. Finally, skeletal stability and nerve repair are important to final functional outcome.

Congenital limb differences may afford opportunities to improve function by free tissue transfer of a spare part to a more functional role. We have reported a successful case of foot-to-hand transfer in split-hand/split-foot malformation with tibial aplasia.

SUMMARY

SHLFD syndrome is a rare congenital disorder, which may be sporadic or autosomal dominant with incomplete penetrance. When complete tibial aplasia is seen, the mainstay of treatment is amputation and lower limb prosthesis. This rare constellation of congenital differences presents an opportunity for microsurgical free tissue transfer using the principle of "spare parts" to improve the functionality of the hand. We present a rare case of split-hand/split-foot malformation with a monodactylous right hand and complete tibial aplasia, treated with microsurgical free foot-to-hand transfer at the time of lower limb amputation, reconstructing key pinch. At the latest 8 months follow-up, the patient had no pain, active key pinch, and ambulated independently with prostheses. He was able to use his right hand independently for a number of daily activities, such as stacking blocks, drinking from a cup, and playing with toys.

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