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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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