Radial Longitudinal Deficiencies With Hypoplastic/Absent Thumbs and Cutaneous Syndactyly of the Most Radial Digits

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Purpose To describe radial longitudinal deficiency with hypoplastic or absent thumb and cutaneous syndactyly between the most radial digits. In addition, to discuss the clinical relevance and unique treatment decisions involved in optimizing functional outcomes in these patients.

Methods A total of 163 extremities of 122 patients with radial longitudinal deficiencies were reviewed. We reviewed radiographs and clinical images that were available, with most radial hypoplastic digit and cutaneous syndactyly to the adjacent finger.

Results There were 7 hands with this type of deformity. Four cases had a hypoplastic thumb associated with cutaneous syndactyly between the thumb and index finger. Three cases had a deformity in which the thumb was absent and the hypoplastic index finger was syndactylized to the long finger. Proximal and distal radioulnar synostoses were associated with these deformities in 3 patients.

Conclusions Because the most radial digit is severely hypoplastic in this type of deformity, pollicization is usually indicated. However, the pollicization procedure must be modified due to associated syndactyly with different degrees of hypoplasia or absence of the intrinsic muscles. This type of deformity should be distinguished from hypoplastic thumb without syndactyly. *(J Hand Surg 2010;35A:1497–1501. Copyright © 2010 by the American Society for Surgery of the Hand. All rights reserved.)*

Type of study/level of evidence Therapeutic IV.

Key words Absent thumb, classification, congenital anomaly, hypoplastic thumb, radioulnar synostosis.

IN RADIAL DEFICIENCY, abnormalities can occur in the hand, radius, and elbow. Blauth[1] classified hypoplastic thumb into 5 grades according to radiographic findings. Heikel[2] reported several cases of hypoplastic thumb associated with syndactyly between the index and long fingers and hypoplasia of the radius. Sometimes we encountered cases with syndactyly between the most radial digit and the adjacent ulnar digit in the presence of hypoplastic or absent thumb. However, this type of deformity cannot be categorized on the basis of Blauth's classification. Graaff et al.[3] described 2 cases of hypoplastic thumb deformity associated with syndactyly of the index finger in a family. However, there are few reports that discuss cutaneous syndactyly between a hypoplastic thumb and the index finger. Because the most radial digit is severely hypoplastic in this type of deformity, pollicization of the next ulnar digit is usually indicated after removal of the most

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radial digit. However, when the most radial hypoplastic digit is associated with cutaneous syndactyly to the adjacent finger, the pollicization procedure must be modified.

In the present study, we investigated cases of syndactyly of the most radial digit in the presence of hypoplastic or absent thumb. These cases are referred to as *syndactylous* type of hypoplastic or absent thumb in this report, and we discuss the position of this type of deformity in the classification of radial longitudinal deficiency.

MATERIALS AND METHODS

All children with a diagnosis of radial longitudinal deficiencies in a senior author's practice, seen between 1974 and 2008, had screening of their hand photographs or radiographs to identify cases of syndactylous type of hypoplastic or absent thumb. A total of 163 extremities of 122 patients were investigated. Seven hands with syndactylous type of hypoplastic or absent thumb were identified. In these patients, the clinical features and deformities of the hand were observed, and the relationships between syndactylous type of hypoplastic or absent thumb and deformities of the forearm and/or abnormalities of other systems of the body were analyzed.

RESULTS

Clinical features and deformities of the hand

There were 7 hands with syndactylous type of hypoplastic or absent thumb in 7 patients; 2 were boys, and 5 were girls. The right hand was affected in 4 cases, and the left in 3.

The thumb or index finger was severely hypoplastic. The intermetacarpal space between the most radial digit and the next ulnar one was quite narrow in all cases. The thenar eminence was flat in appearance in all cases.

Four cases had a hypoplastic thumb associated with cutaneous syndactyly between the thumb and index finger (Figs. 1, 2). The degree of cutaneous syndactyly varied. In 2 cases, complete cutaneous syndactyly was present (Fig. 1). In the remaining 2 cases, cutaneous syndactyly existed at the distal end of the proximal phalanx of the hypoplastic digit (Fig. 2).

Three cases had a deformity in which the thumb was absent and the hypoplastic index finger was syndactylized to the long finger (Fig. 3). In 2 cases, cutaneous syndactyly existed at the proximal end of the distal phalanx of the hypoplastic digit (Fig. 3). In the remaining case, complete cutaneous syndactyly was present.

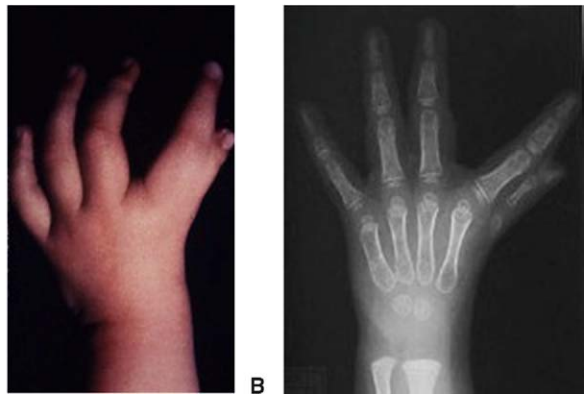

FIGURE 1: A, B The proximal 2/3 of the thumb metacarpal is absent, and remaining phalanges are severely hypoplastic on the left hand. The hypoplastic thumb is associated with tight cutaneous syndactyly between the thumb and index finger.

As for surgical treatment, of the 4 cases with hypoplastic thumb associated with cutaneous syndactyly between the thumb and index finger, separation of syndactyly and opponensplasty were performed in one case. Resection of hypoplastic thumb and pollicization of the index finger were performed in 2 cases. No surgical treatment had been performed in one case. On the other hand, of the 3 cases with absent thumb associated with cutaneous syndactyly between the hypoplastic index and long fingers, resection of the hypoplastic index finger and pollicization of the long finger were performed in 2 cases. No surgical treatment had been performed in one case.

As for abnormal findings found during pollicization, it was confirmed that no thenar muscles had developed. Flexor and extensor tendons of the most radial hypoplastic digit, together with the interosseous muscles on the radial side of the pollicized digit, were severely hypoplastic in all cases. After pollicization, one case had a weak adduction (Table 1, case 2), one case had weak abduction, adduction, and extension of the pollicized digit (Table 1, case 4), one case had a weak abduction (Table 1, case 5), and the last case had a stiff finger (Table 1, case 7). Among them, in one case with a stiff pollicized digit, the thumb was stable and pinch was possible; therefore, the parents did not want further treatment (Table 1, case 7). In one case, opponensplasty by using a flexor digitorum superficialis of the ring finger and first web plasty were performed, and a pinch was possible after the second surgery (Table 1, case 5). In 2 cases, tendon transfer was recommended to get better pinch function, but the parents did not agree because they were satisfied with the results (Table 1, cases 2 and 4).