

## Strategic two-stage approach to radial club hand

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**Abstract** In patients with radial club hand (RCH), there is absence of the radius and severe hypoplasia of the thumb, so both wrist stabilization and thumb reconstruction are essential. When wrist stabilization is performed, recurrence of angulation is a problem. When thumb reconstruction is done, preservation of at least one major dorsal vein is important to prevent necrosis of the reconstructed thumb. We executed a strategic approach to RCH, which aimed to prevent both recurrence of angulation and necrosis of the reconstructed thumb.

**Keywords** Radial club hand · Centralization · Pollicization · Skin incision · Thumb reconstruction

### Introduction

Radial club hand is a notable manifestation of congenital radial dysplasia [1]. Patients with radial club hand have three main problems, which are a short forearm, radial deviation of the wrist, and an absent or hypoplastic thumb [2]. Various techniques have been reported for correction of this functional and aesthetic deformity [3, 4].

To correct radial deviation at the wrist, centralization has been the most common wrist stabilization procedure. When centralization is done, removal or breakage of the Kirschner wires stabilizing the wrist can lead to recurrence of angulation [3]. Indications for pollicization in patients with congenital abnormalities continue to be a matter of controversy, but the indications for pollicization are generally accepted as type IV (floating thumb) and type V (aplasia) of Blouth [5]. To prevent necrosis of the pollicized digit, vascular damage must be avoided. Frequently, the only vein runs along the radial aspect of the metacarpophalangeal (MP) joint of the index finger, and this must be protected while raising the flaps [6]. Therefore, skin incisions on the dorsum of the hand, which might cause poor blood supply to the flap and reconstructed thumb, should be avoided.

This report describes a strategic two-stage approach to radial club hand, which prevents recurrence of angulation and avoids necrosis of the reconstructed thumb in detail.

### Case report

A one-week-old male patient had right radial club hand deformity with complete absence of the radius and a floating thumb (Fig. 1A, B). There were also flexion deformities at the proximal interphalangeal (PIP) joints of the index finger and middle finger. The index finger had  $-30^\circ$  of active and passive extension with  $90^\circ$  of active and passive flexion at the PIP joint, while the middle finger had  $-20^\circ$  of active and passive extension with  $100^\circ$  of active and passive flexion at the PIP joint. Movement of the elbow was only slightly limited ( $-10^\circ$  of active and passive

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extension with 110° of active and passive flexion). Therapy with a dynamic wrist splint was started from 3 months after birth. However, surgery for his radial club hand was delayed due to the treatment of life-threatening heart disease associated with Holt-Oram syndrome. A two-stage operation was planned, consisting of wrist stabilization by centralization and subsequent thumb reconstruction by index finger pollicization.

### Stage one (centralization)

At the age of 22 months, centralization was performed. The goal of centralization is to stabilize the carpal bones and to correct angulation [1]. A dorsal S-shaped incision was extended from the dorsum of the hand to the distal third of the forearm. This allowed access to the radial musculature, the shaft of the ulna, and the carpus itself [3]. Except for the skin incision, the actual centralization technique was based on the descriptions of Bayne and Klug [7].

The fibrotic muscle mass attached to the carpus, including extensor carpi radialis and flexor carpi radialis, was dissected free. Limited shaving of the carpus was done while avoiding injury to the growth plate in order to sta-

bilize the carpus on the ulna. Then extensor carpi ulnaris (ECU) was advanced distally, and flexor carpi ulnaris was sutured to ECU distally and dorsally to help prevent radiovolar deformity. The wrist was fixed with a Kirschner wire that was 1.8 mm in diameter (Fig. 2). Skin closure was achieved easily by rotating the skin flap. A long arm cast was applied for 4 weeks. Then a short arm splint was applied to protect the wire and prevent the recurrence of deformity.

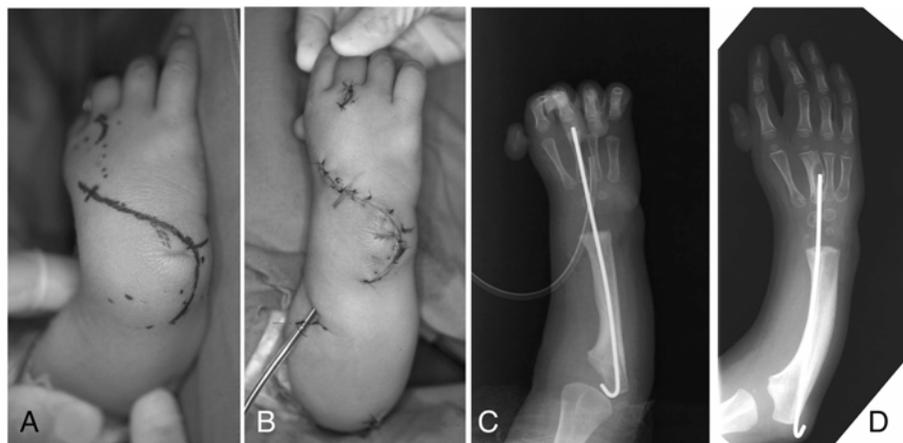
Angiography of the upper extremity was performed at the same time as the angiographic procedure that was done to examine cardiac anomalies associated with Holt-Oram syndrome. Angiography via the brachial artery indicated that the radial artery was absent and a slender vein ran along the radial aspect of the MP joint of the index finger (Fig. 3).

### Stage two (index finger pollicization)

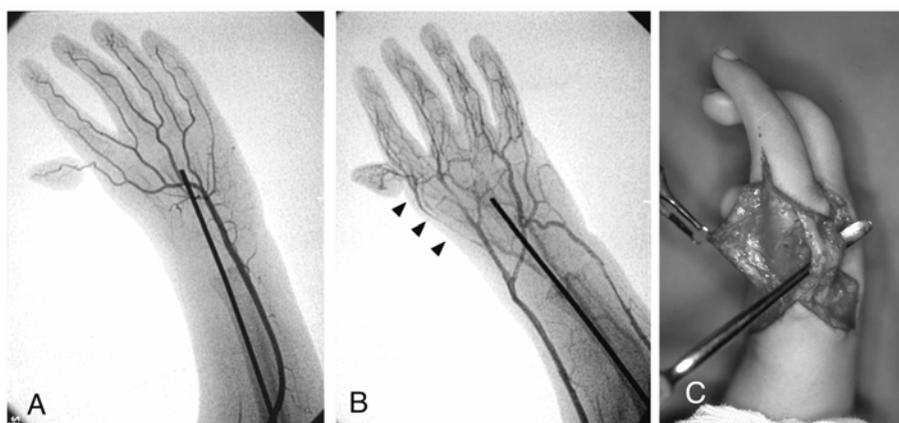
At the age of 31 months, pollicization was performed, i.e. the transposition of a digit for construction of a new thumb. In this case, the actual technique of pollicization was based on the description of Carroll [8]. The skin incision for pollicization was designed to extend from that used for centralization (Fig. 4). The procedure involved transfer of the digit with a neurovascular pedicle, adjustment of the bones (shortening, rotation, and abduction), and muscular stabilization [9]. Adjustment of the extensor tendons and the interosseous muscles played an important part in stabilization of the new thumb. The extensor indicis proprius tendon was shortened and then resutured to become the new extensor pollicis longus. The extensor digitorum communis tendon was severed at the metacarpophalangeal level. After metacarpal resection, the proximal end of this tendon was sutured to the base of the former proximal phalanx (now acting as the first metacarpal), so that extensor digitorum became the new abductor pollicis longus. The tendons of the first palmar and first dorsal interossei muscles were detached from the base of the proximal phalanx of the index finger, and then were sutured to the lateral bands of the dorsal aponeurosis after osteotomy and resection of the second metacarpal. As a result, the first palmar and first dorsal interossei became the adductor and abductor of the new thumb, respectively. Pollicization was performed without removing the Kirschner wire that had been inserted during the first operation (Fig. 5). Instead, this wire was removed when it finally broke at 23 months after centralization (14 months after pollicization). When the boy fell, the Kirschner wire broke at the ulnocarpal joint. Significant cosmetic improvement without radial deviation and a stable wrist with prehensile function were achieved at 27 months after pollicization (36 months after centralization) (Fig. 6).



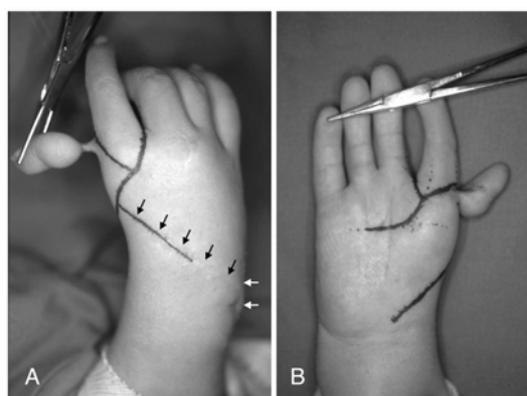
**Fig. 1** A, B. Preoperative appearance at one week of life. The hand is radially deviated by 90°. C, Preoperative X-ray film of the radial club hand at one year.



**Fig. 2** Centralization and postoperative X-ray film. **A, B, C,** Centralization at the age of 22 months. **A,** Dorsal S-shaped skin incision. **B,** Appearance immediately after operation. **C,** X-ray film immediately after operation. **D,** X-ray film shows cyst formation, possibly due to mechanical stress caused by the Kirschner wire, at 9 months after centralization (at 31-months-old).



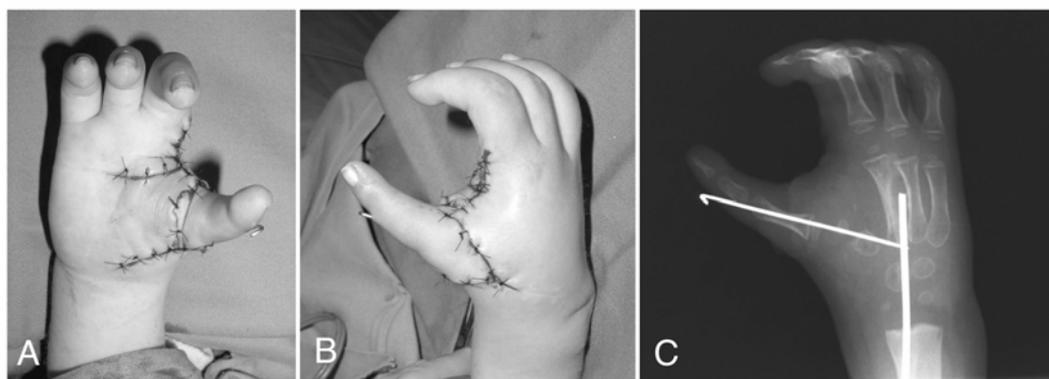
**Fig. 3** **A, B,** Angiograms obtained at 31-months-old. **A,** Angiogram (arterial phase). **B,** Angiogram (venous phase) reveals a slender vein that runs along the radial aspect of the metacarpophalangeal joint of the index finger (arrowheads). **C,** Pollicization at 31-months-old. The critical dorsal veins surrounded by connective tissue.



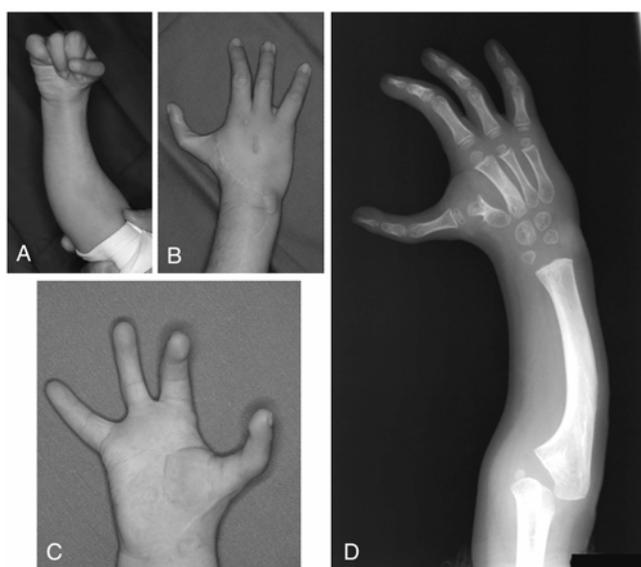
**Fig. 4** **A, B,** Pollicization at 31-months-old. **A,** The skin incision for pollicization (arrows) was designed to extend from that for centralization. **B,** Skin incision line on the palmar side.

### Discussion

Conventional wrist stabilization procedures include centralization or radicalization [10]. Recently, wrist stabilization by gradual distraction with an external fixator has also been recommended [11, 12]. The Kirschner wire for stabilizing the wrist is inserted into the ulna through the second metacarpal in the case of radialization, while it is inserted into the ulna through the third metacarpal for centralization. Therefore, the wire must be removed when index pollicization is performed after radialization, while it can be left in situ after centralization. To retain strong fixation for a longer period, centralization is preferable to radialization. Although the wire is usually removed between six and 12 weeks after centralization [1, 2, 7], the Kirschner wire was left in place for 23 months after



**Fig 5** A–C, Pollicization at 31-months-old. **A, B**, Appearance immediately after pollicization. **C**, X-ray film immediately after the operation.



**Fig 6** A–D, The patient at 58-months-old. At 36 months after centralization (27 months after pollicization), an X-ray film demonstrates a change in the distal contour of the ulna, with widening and cupping to support the carpus, so that it resembles a normal radius. Good functional and cosmetic results were obtained.

centralization until it broke in our patient. We do not recommend waiting until the intramedullary wire breaks prior to removal. However, we believe that the Kirschner wire should not be removed as long as its distal tip lies in the third metacarpal. It took approximately 6 months for the wrist to become stable because X-ray films showed cystic enlargement of the third metacarpal (possibly due to mechanical stress caused by the Kirschner wire) until 6 months after centralization. Such long-term fixation helped to prevent postoperative recurrence of angulation.

In surgery for radial club hand, the ideal incision provides additional skin on the radial side of the wrist and takes up the slack on the ulnar side, as well as giving

good access to the wrist joint and surrounding soft tissues [13]. For centralization, the reported skin incisions include a dorsal S-shaped incision, an ulnar incision with radial Z-plasty, a transverse ulnar incision, and a dorsal bilobed flap [1, 2, 7, 10, 13, 14]. Among them, the dorsal S-shaped incision can be extended to the incision line for pollicization smoothly. Therefore, our incision for pollicization was designed to extend along the line used for centralization, thus preventing damage to the dorsal veins. In addition, it gave good access to the wrist joint and surrounding soft tissue structures and could be used to correct cutaneous disproportion, i.e. a lack of skin on the radial side and excess skin on the ulnar side, by rotating the skin flap. However, flaps for correcting cutaneous disproportion might become unnecessary if preoperative soft tissue stretching was done by gradual distraction.

To treat a floating thumb, attempts at microsurgical joint transfer to restore the carpometacarpal joint have been reported, but the results appear to be relatively poor compared with index finger pollicization [15]. When index finger pollicization is done, the critical veins that run along the radial aspect of the MP joint of the index finger must be preserved on the dorsum of the hand. Although venae comitantes exist around the digital arteries, these are small and unreliable [6]. In addition to careful dissection of digital arteries, preservation of at least one major dorsal vein is very important to ensure adequate venous drainage from the newly constructed thumb supplied by two arteries [4]. In the present case, angiography revealed that the radial artery was absent, while there was a slender vein running along the radial aspect of the MP joint of the index finger (Fig. 3). A well-planned skin incision can make the pollicization procedure safer and more reliable.

In the present case, recurrence of deformity was not seen during 4 years of follow-up after centralization. However, long-term follow-up studies have revealed that some recurrence of radial club hand is universal. The wrist tends to either become flexible and deviated or stiff

and straight [11]. Concerning the outcome at skeletal maturity, the affected forearm is usually only half as long as that on the unaffected side. This length deficit can be addressed by ulnar lengthening with distraction osteogenesis [11]. Longer follow-up is required to assess the incidence of recurrence and ulnar growth retardation. To treat recurrent wrist deformity and lengthen the ulna, distraction lengthening combined with corrective osteotomy should be considered in the future [16].

In conclusion, careful preoperative planning is required for treatment of radial club hand and we believe that our strategic approach with a two-stage procedure may be useful.

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