Reconstruction of active elbow flexion in patients with radial ray deficiency: report of two cases

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Radial ray deficiency is a complex congenital abnormality of the radial side of the upper extremity. Stiffness of the elbow in extension is frequently observed in association with a radial clubhand involving radial angulation and displacement of the carpus with respect to the distal ulna. In such cases, corrective surgery on the carpus should not be attempted in the absence of an adequate range of elbow flexion because insufficient elbow flexion precludes bringing the hand to the face and feeding oneself. Active elbow flexion has been reported to improve spontaneously after the first year or two of life, although less than 90° of flexion is finally obtained in many patients. In some patients, the elbow remained stiff in extension or gained flexion to only 40° or less. The insufficient elbow flexion might be one of the causes of gradual bowing of the ulna or recurrence of radial clubhand deformity after wrist corrective surgery. To date, there have been only a few case reports of reconstruction of active elbow flexion with a good outcome, which was achieved by triceps transfer. However, the triceps is important as an antagonist of flexion of the elbow joint and should therefore be preserved as much as possible. Here, we report two cases of radial clubhand without active elbow flexion in which transposition of the rudimentary insertion of the biceps to the ulna resulted in a good outcome.

Case reports

Case 1

A boy was born with a Bayne type 4 radial clubhand associated with a Blauth type 5 hypoplastic thumb in the right upper extremity and a Bayne type 3 radial clubhand associated with a Blauth type 5 hypoplastic thumb in the left upper extremity (Fig. 1). When the patient was 10 months old, he underwent the right wrist centralization procedure and transposition of the rudimentary insertion of the ipsilateral biceps to the ulna.

Although the wrist was splinted as straight as possible before surgery, the passive range of motion (ROM) of the right elbow joint was 0° to 110° and the active ROM was 0° to 20°. The passive pronation/supination was 90°/90°. There was severe radial deviation of the right hand.

An anterior approach was used to expose the distal part of the biceps in the elbow joint. The thin fascia-like insertion of the biceps was fused with the fascia in the proximal radial forearm. A very thin fibrous band was found between the ulnar side of the distal biceps and the fascia in the proximal ulnar forearm and considered to represent a rudimentary lacertus fibrosus.

After separation of the accompanying artery from the biceps, the thin fascia-like insertion of the biceps was detached from the radial side of the forearm and held with 2-0 Ti-Cron suture (Covidien, Dublin, Ireland). Two drill holes (0.7 mm in diameter) were made anteroposteriorly a few millimeters distal to the coronoid process of the ulna.

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Then, the Ti-Cron suture holding the distal stump of the biceps was passed through the holes and secured in the bone under tension with the elbow in 45° of flexion. Postoperatively, a posterior splint was applied for 3 weeks, after which active flexion of the elbow joint was allowed.

Ten years postoperatively, the active ROM of the right elbow joint was 0° to 135°. Active supination of the forearm was 55° and active pronation was −20° (Fig. 2). Manual muscle testing of elbow flexion resulted in a score of grade 5.

Case 2

A girl was born with radioulnar synostosis associated with a Blauth type 5 hypoplastic thumb in the left upper extremity and a Bayne type 1 radial clubhand associated with a Blauth type 2 hypoplastic thumb in the right upper extremity (Fig. 3). When the patient was 15 months old, she underwent surgical closure of a ventricular septal defect and annuloplasty for treatment of tricuspid insufficiency. She was thought to have Holt-Oram syndrome but did not undergo genetic analysis of TBX5. When the patient was 22 months old, she underwent left index finger pollicization and transposition of the rudimentary insertion of the ipsilateral biceps to the ulna. Before surgery, the passive ROM of the left elbow joint was 0° to 90°, with no active flexion. The forearm was fixed in 60° of pronation. Manual muscle testing of elbow flexion resulted in a score of grade 5.

Discussion

In radial ray deficiency, most of the elbow flexors are abnormal. The long head of the biceps is almost always absent. If it is present in conjunction with total aplasia of the radius, it inserts abnormally into the lacertus fibrosus. The short head is generally present but is usually fused with the coracobrachialis, brachialis, or forearm flexor muscles. The brachialis may be present but is often fused with the biceps at its origin and becomes continuous with the common flexor muscles without any specific insertion. The brachioradialis is usually absent in patients with total aplasia of the radius. If it is present, the brachioradialis can be rudimentary or fused with the extensor muscles and insert aberrantly on the carpus or ulna. Despite all this abnormality of the muscles around the elbow joint, Smith
Figure 2  Case 1. (A) The patient’s elbow extension 10 years postoperatively. (B) The elbow flexion. The elbow actively flexed to 135°. (C) Radiograph of the elbow joint 10 years postoperatively. The site of osteosclerosis was located distal to the coronoid process of the ulna. This is the remnant of the drill holes used for the attachment of the biceps insertion to the ulna with suture.

Figure 3  (A) The preoperative appearance of the left upper extremity in case 2. No crease was found in the antecubital fossa. (B) The preoperative radiograph. Radioulnar synostosis and absence of the thumb were observed.
reported never observing a patient fail to regain at least 90° of elbow flexion after wrist centralization, even when the elbow was stiff preoperatively. The brachialis may act as the primary flexor of the elbow joint in such patients. The brachialis is a monarticular muscle and is considered to act as an antigravity and skill-related muscle, whereas the biceps is a biarticular muscle and is considered to act as a propulsive muscle. The propulsive muscle produces gross power and accelerates reaching movements. In normal active elbow flexion, the bulging of the contracted brachialis muscle belly pushes the belly and tendon of the biceps muscle forward. This increases the biceps lever arm and thus the flexion power of the elbow. Although the biceps might be unnecessary for active flexion of the elbow joint, it would remain necessary for power flexion against resistance or maintenance of elbow flexion. In our cases, the thin fascia-like insertion of the biceps fused with the fascia in the proximal radial forearm, leading to inefficient transmission of the traction force produced by the biceps to the forearm. Direct suturing of the rudimentary insertion of the biceps to the ulnar cortex was therefore desirable to allow efficient transmission of the biceps traction force and thus improvement in the activities of daily life.

Several procedures have been reported for reconstruction of active elbow flexion in patients with congenital elbow malformations; these include triceps transfer, pectoralis major transfer, latissimus dorsi transfer, the Steindler flexorplasty, and free muscle transfer. In patients with radial ray deficiency, the short head of the biceps is usually present without a secure distal insertion. Therefore, transfer of a muscle from outside the upper arm is unnecessarily invasive. The Steindler flexorplasty would be inadequate because of the weakness of the wrist extensors. The triceps muscle should generally be preserved as much as possible because of its importance as an antagonist of flexion of the elbow joint, but triceps transfer could be used as an alternative in cases in which the insertion of the short head of the biceps is completely absent.

We secured the rudimentary insertion of the biceps to the ulna under tension with the elbow in 45° of flexion in case 1 and 90° of flexion in case 2. The optimal elbow position and degree of tension with which to secure the biceps remain unclear. In case 1, the maximum active elbow flexion was 90° after 1 year and had improved to 135° 10 years postoperatively. In case 2, because sufficient length of the rudimentary insertion could not be obtained, we secured the insertion to the ulna with 90° of flexion. The maximum active elbow extension was −40° after 2 months and 0° after 2 years postoperatively. The secure distal insertion might force the biceps itself to adapt to the length of the humerus as the patient grows, by the downward pull of gravity or by the traction force of the triceps.

In case 1, because complete absence of the radius on radiographic examination obviously indicated the absence of radial biceps insertion, we were afraid that spontaneous improvement of elbow flexion would be insufficient to bring the hand to the face. Then, transposition of the rudimentary insertion of the biceps was decided to be performed with the wrist centralization surgery and resulted in excellent active ROM and flexion power of the elbow joint. This transposition surgery should be performed on patients with radial ray deficiency who cannot gain sufficient active elbow flexion after the wrist corrective surgery. However, especially in cases of complete absence of the radius (Bayne type 4), it would be an option to combine the transposition of the rudimentary insertion of the biceps with the wrist corrective surgery.

We did not perform any other imaging examinations except radiography before the elbow surgery. Although the short head of the biceps was reported to be generally...

Figure 4  Case 2, intraoperative photographs. (A) Just after the separation of the rudimentary insertion of the biceps (white arrow) from the fascia of the common flexor muscles (asterisk). The black arrow indicates the proximal side. (B) The new insertion of the biceps onto the ulna (white arrow). The black arrow indicates the proximal side.
present, it might be better to perform a noninvasive imaging examination, such as magnetic resonance imaging or ultrasound, to define the presence of the biceps itself and its insertion. However, it would be difficult to find a thin fascia-like insertion of the biceps by magnetic resonance imaging or ultrasound.

Conclusions

Transposition of the rudimentary insertion of the biceps is an option for reconstruction of active elbow flexion in patients with radial ray deficiency without active elbow flexion.

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References


Figure 5  Case 2. (A) Elbow extension 3 years postoperatively. (B) Elbow flexion. The elbow actively flexed to 80°. (C) Radiograph showing the site of osteosclerosis distal to the coronoid process of the ulna (arrow).