Elbow and forearm reconstruction in patients with ulnar dimelia can improve activities of daily living

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Ulnar dimelia (mirror hand with double ulna) is a rare congenital anomaly in which both forearm bones develop as a normal ulna and polydactyly is present in the hand, the thumb and radius bone being absent.6,14 Elbow and forearm problems remain after removal of excess digits and reconstruction of a radial finger to serve as a thumb to allow prehensile movement in all cases reported in the literature.14 No reports of elbow reconstruction have been identified, although reports of hand and thumb reconstruction have been published.3,5 We present our experience of improved activities of daily living (ADLs) after elbow reconstruction for a patient with ulnar dimelia. The elbow reconstruction led to better forearm range of motion but no significant improvement in elbow flexion.

Case report

A boy born in December 1994 with right-sided ulnar dimelia and polydactyly had a surgical history of a radial finger amputation at the age of 3 months and pollicization at the age of 4 years for his 6-fingered hand. He was referred to our hospital for follow-up after these surgeries.

He had loss of elbow flexion and forearm rotation from birth and started to feel a disturbance in ADLs from the beginning of 2010 with increasing activity. He had no other obvious deformity, and there was no contributory family history.

Clinical examination demonstrated a decreased range of right elbow motion, with flexion of 90° and extension of −30°. The patient presented with no forearm mobility and a fixed position at 70° of supination. Neurovascular examination findings were normal, and the surgical scars had healed well. The Disabilities of Arm, Shoulder and Hand (DASH) scores (ranked from 0 indicating least disability to 100 indicating the most disability)7 were 12.9 (Disability/Symptom) and 18.8 (Sports/Music), and the Hand 20 score (ranked from 0 indicating least disability to 100 indicating most disability)16 was 51.8.

Standard radiographs showed 2 identical ulna bones articulating with the lower end of the humerus (Fig. 1), and 7 carpal bones grouped in a perfect arch about a central bone and a pisiform bone on the medial side (Fig. 2). Cineradiographs (Video 1, anteroposterior view; Video 2, lateral view; available online) showed that the medial forearm bone functioned in the role of the ulna and that the lateral forearm bone functioned as the radius. However, the proximal end of the lateral olecranon-like shaped bone seemed to serve as a bony block for both elbow flexion and forearm rotation.

In March 2011, arthrography and surgery were performed. The proximal forearm joint (radioulnar joint) and distal forearm joint (radioulnar joint) were identified (Figs. 3 and 4). The right upper extremity was then prepared and draped in the standard sterile fashion. The patient and the procedure were identified and confirmed by all present in the room before the onset of the procedure. The procedure began with an Esmarch tourniquet being placed around the right upper extremity. Once the tourniquet had been inflated to 250 mm Hg, a longitudinal incision was made starting at the lateral condyle of the humerus and carried down to the subcutaneous tissue. The extensor carpi ulnaris was separated from the anconeus, and the distal fibers of the anconeus were divided. Standard neurovascular anatomy surrounding the elbow was observed. Care was taken to protect the posterior interosseous nerve.

The common extensor muscle origin was reflected anteriorly by subperiosteal dissection from the lateral epicondyle. The joint...
capsule was incised longitudinally, and the lateral surface of the proximal lateral forearm bone was exposed. A part of the biceps brachii tendon was inserted to the proximal end of the lateral forearm bone (Fig. 5). We then proceeded with the “radial head”plasty, the proximal end of the lateral olecranon-like shaped bone was resected, and a radial head-shaped proximal end was produced using a bone saw and a chisel (Fig. 6).

Next, a medial incision was made under the tip of the medial epicondyle with the elbow flexed at a right angle. The ulnar nerve was isolated in its groove posterior to the epicondyle and retracted anteriorly exposing the medial collateral ligament. The posterior oblique ligament with strong fibrous tissue was identified and incised (Fig. 7). A bone spur on the anterior side of the distal humerus was also excised, and arthrolysis was performed around the insertion of the triceps brachii. Elbow extension/flexion 0°/110° and forearm supination/pronation 90°/−10° were acquired during the surgery. Once this had been completed, the tourniquet was released.
Hemostasis was achieved in all incision sites using bipolar cautery. The lateral and medial incisions were closed with 4-0 PDSII (Ethicon, Somerville, NJ, USA) in an interrupted dermal pattern and then with several interrupted 5-0 Vicryl Rapide (Ethicon) sutures on the skin. Sterile dressings and a soft bandage were applied. An elbow splint maintaining the elbow at a right angle and the forearm in neutral was also applied. The patient tolerated the procedure well and was transferred to the recovery room in stable condition.

The patient was permitted to start continuous passive elbow motion using the EL-2500 device (Medical Engineering System, Tokyo, Japan) 5 days after surgery within the limits of pain. Progressive active mobilization of the elbow and forearm were allowed during the first 2 postoperative weeks.

The range of forearm motion was pronation 0°/supination 80°, and elbow motion was flexion 0°/extension −15° at final follow up at 2.5 years after surgery in September 2013. The DASH (Disability/Symptom), DASH (Sports/Music), and the Hand 20 scores improved from 12.9 to 6.9, from 18.8 to 0.0, and from 51.8 to 30.0 (Table I), respectively. The patient was asymptomatic and fully functional for ADLs. In addition, he was satisfied with his function for sporting activities, including lacrosse and football. The patient had no complaints of pain before or after the surgery.

Standard radiographs 2.5 years after the surgical procedure showed appropriate positioning of the proximal lateral ulna functioning as the radial head, as well as proper positioning of the medial ulna, with no signs of recurrence or ectopic ossification (Fig. 8).

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<thead>
<tr>
<th>Variable</th>
<th>Preoperative</th>
<th>Postoperative</th>
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<tbody>
<tr>
<td>Forearm</td>
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<tr>
<td>Supination, °</td>
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<td>80</td>
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<td>Pronation, °</td>
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<td>Elbow</td>
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<td>Extension, °</td>
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<td>−20</td>
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<tr>
<td>Flexion, °</td>
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<td>80</td>
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<tr>
<td>DASH score</td>
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<tr>
<td>Disability/Symptom</td>
<td>12.9</td>
<td>8.6</td>
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<tr>
<td>Sports/Music</td>
<td>18.8</td>
<td>25.0</td>
</tr>
<tr>
<td>Hand 20 score</td>
<td>51.8</td>
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DASH, Disabilities of the Arm, Shoulder and Hand.
Discussion

Ulnar dimelia is classified as a type of mirror hand.\textsuperscript{2} It can be experimentally provoked by transplantation of a zone of polarizing activity of one limb bud to the cranial side of another limb bud.\textsuperscript{4} The cause remains unknown, although another type of mirror hand, Laurin-Sandrow syndrome with bilateral mirror hand and feet, absent radii and tibias, and nasal defects,\textsuperscript{11,15} is known as an autosomal-dominant syndrome.\textsuperscript{12}

Limited movements of the elbow and flexion deformity of the wrist are some of the features associated with ulnar dimelia.\textsuperscript{1} Reduction of the number of digits and thumb reconstruction are the mainstays of treatment.\textsuperscript{1} Others have noted the need for elbow and forearm surgeries for ulnar dimelia patients.\textsuperscript{9}

In typical ulnar dimelia, the forearm and hand elements show sagittal mirror symmetry. However, asymmetrical vascular anatomy was also reported on computed tomography angiography.\textsuperscript{1} In fact, we found that the posterior interosseous nerve ran onto the proximal lateral forearm bone, and the ulnar nerve was located in the groove posterior to the medial epicondyle when we operated in this case using lateral and medial approaches. The vascular and nerve anatomy was more standard than we expected after reviewing the bony anatomy. In addition, we confirmed asymmetrical motion of 2 forearm bones when the elbow was moved on fluoroscopy. The medial forearm bone served in the role of the ulna, and the lateral forearm bone functioned as a radius; therefore, the olecranon-like proximal prominence of the lateral forearm bone would limit forearm rotation.

The proximal and distal forearm joints were both present on arthrography in the present patient. We therefore acquired improved forearm rotation after resecting the prominent proximal lateral forearm bone structure. In general, obtaining joint motion in congenital ankylosis or contracture cases is more difficult than in traumatic cases and is even more difficult in more chronic cases. However, forearm rotation would be relatively improved after arthrolysis in the congenital cases.\textsuperscript{10,13} The DASH (Disability/Symptom) score was improved from 12.9 preoperatively to a score of 6.9 at 2.5 years after surgery. The mean DASH score of the general population is reported to be 10.1.\textsuperscript{8} The DASH (Sports/Music) score improved from 18.8 preoperatively to a score of 0.0 postoperatively, whereas the mean general population score for the DASH (Sports/Music) is reported to be 9.7.\textsuperscript{8} The Hand 20 was also used as a measure of ADLs function, with a preoperative score of 51.8, which improved to a postoperative score of 30.0 (Table I). We experienced a patient who showed improvement in ADLs after gaining forearm rotation, whereas elbow flexion/extension motion did not change remarkably.

Surgical release of proximal forearm and elbow joints with “radial head” plasty should be considered for obtaining forearm motion and improving ADLs function in patients with ulnar dimelia. In addition, better elbow flexion may result if surgery is performed at an earlier age. This particular patient began to feel a disturbance in ADLs with increasing activities at approximately the age of 15 years; however, he might have had a better result if surgery had been performed at a younger age. We also suggest that improvement may be enhanced with continuous passive elbow motion immediately after surgery.

Conclusion

A patient with ulnar dimelia treated with proximal forearm and elbow joint surgical release with “radial head” plasty resulted in ADLs improvement after acquiring forearm rotation. This surgery should be...
considered for improving forearm motion and ADLs for ulnar dimelia even in young adulthood.

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Disclaimer

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Supplementary data

The videos related to this article can be found online at http://dx.doi.org/10.1016/j.jse.2013.12.001.

References