Combined Elbow Release and Humeral Rotational Osteotomy in Arthrogryposis

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Purpose The purpose of this study was to determine if a simultaneous posterior elbow release and humeral osteotomy to correct both the elbow extension contracture and the humeral internal rotation contracture in children with arthrogryposis can produce similar results as a posterior elbow release alone.

Methods This study was a retrospective chart review of consecutive patients with arthrogryposis treated surgically for elbow extension contracture between 2007 and 2014. A total of 43 procedures in 36 patients had adequate available follow-up data and were included in the study. The postoperative range of motion reported was measured at the early follow-up (3–6 months), midterm follow-up (between 1 and 2 years), and the most recent long-term follow-up (after 2 years) from the date of surgery. Patients were grouped into 2 groups (simultaneous and release) based on the necessity of performing an ipsilateral humeral rotation osteotomy at the time of the release.

Results At early follow-up, patients in both groups increased their total arc of motion. There was a significant difference in extension and arc of motion at midterm follow-up (between 1 and 2 years) between the simultaneous and the release groups with the simultaneous group significantly losing both terminal extension and total arc of motion. At more than 2 years follow-up, there remained a statistically significant difference in arc of motion, with the release group having a significantly larger arc of motion. Patients who underwent dual plating had a much larger arc of motion at early follow-up than the K-wire or single-plate fixation group, despite having similar preoperative extension, flexion, and arc of motion. This difference was also significant at late follow-up.

Conclusions Patients with posterior release alone had significantly greater improvement in total arc of motion and significantly better elbow extension than patients who underwent a simultaneous humeral osteotomy. However, rigid fixation with early mobilization may yield results comparable with the release alone group. (J Hand Surg Am. 2017;■(■):1.e1-e9. Copyright © 2017 by the American Society for Surgery of the Hand. All rights reserved.)

Type of study/level of evidence Therapeutic IV.

Key words Arthrogryposis, elbow flexion contracture, humeral osteotomy, posterior elbow release.

Arthrogryposis is a descriptive term for any patient born with contracture of multiple joints. There are multiple subtypes of arthrogryposis, including syndromes with congenital contractures. The extent and severity of contracture varies between diagnoses and from patient to patient. Overall, 25% of patients may have elbow involvement and 19% shoulder involvement. The most common diagnosis is

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Amyoplasia congenita, which is a sporadic (non-heritable) condition often characterized by nearly symmetrical limb involvement. The upper extremity typically assumes a posture of shoulder adduction, shoulder internal rotation, elbow extension, and wrist flexion. This posture is due to hypoplasia or aplasia of muscles throughout the limbs. The contractures are present at birth and improve variably with time. Maximal gains without intervention are typically made in the first 2 years.  

The elbow extension contracture in arthrogryposis prevents the child from bringing the hand to the mouth for functions such as eating. Internal rotation contractures that prevent external rotation to less than 30° shy of neutral can limit bimanual tasks and force a cross-over grasp pattern (Fig. 1). Internal rotation may also contribute to preventing the hand from reaching the mouth. When nonsurgical treatments fail to allow the patient to self-feed, a combination of surgeries (humeral osteotomy to improve the external rotation arc of motion of the arm and posterior elbow release to improve elbow flexion) can be done to improve these problems.  

Posterior elbow release improves passive elbow flexion and thus ability to bring the hand to the mouth. However, for patients with elbow extension contractures who also have fixed shoulder internal rotation contractures, a release at the elbow will achieve only limited functional gains. With the shoulder internally rotated, it is impossible for the child to use elbow flexion to reach the mouth or face. Unlike the elbow, soft tissue releases have not been shown to be effective in improving shoulder range of motion. Humeral osteotomy is therefore recommended to permit the arm to be rotated into greater external rotation. Although osteotomy does not increase the actual arc of motion, it does allow that arc to be placed in a more usable space for the patient.  

Because these 2 problems (internal rotation contracture and elbow contracture) often coexist, and because they may both be addressed via a common surgical approach, it has been our practice to perform both procedures simultaneously when indicated. However, the rehabilitation protocols for these 2 procedures may conflict. Contracture releases mandate immediate and aggressive postoperative mobilization to preserve the gains that are made. However, the healing of osteotomies may require different degrees of immobilization depending on the method of fixation used.  

To determine the effect of combining these procedures, we have compared our results from recent years during which simultaneous procedures have been performed with the results from patients at our institution who had separate procedures. In particular, we were interested in whether similar gains were made in elbow range of motion. We hypothesized that patients who undergo release alone will have improved range of motion at long-term follow-up compared with patients who underwent simultaneous release and humeral osteotomy. Further, we hypothesized that a more rigid fixation construct allowing for earlier initiation of range of motion will result in similar range of motion gains compared with those in patients who underwent release alone.  

**MATERIALS AND METHODS**

This study was a retrospective chart review of patients with arthrogryposis treated surgically for elbow extension contracture between 2007 and 2014 at our institution. The study was approved by our institutional review board, and the guardians of all patients signed informed consent regarding the use of their deidentified data in the study. All surgeries were performed by 1 of the 2 senior authors (S.H.K. and D.A.Z.). We reviewed the medical records for age,
sex, preoperative and postoperative range of motion, procedures performed, hardware used for fixation, concomitant procedures performed on the same extremity, and complications. A standard hand-held goniometer was used for all measurements and measurements were recorded by either 1 of the 2 senior authors (S.H.K. and D.A.Z.) or the occupational therapist. The measurement taken at the last clinic visit prior to surgery was taken as the preoperative range of motion. The postoperative range of motion reported was measured at the early follow-up (3–6 months), midterm follow-up (between 1 and 2 years), and the most recent long-term follow-up (after 2 years) from the date of surgery.

We reviewed 44 patients. One patient was excluded because she was treated simultaneously for an olecranon fracture. Another patient was excluded because he had a different procedure to increase elbow flexion (anterior closing wedge osteotomy of the humerus). One patient was excluded because he initially had only elbow manipulation and humeral osteotomy performed. He then lost some of the range of motion of his elbow, and so a triceps lengthening was done. Five patients were excluded because they had only a humeral osteotomy performed. This left 36 patients, of whom 6 had bilateral procedures. The remaining 36 patients had sufficient data available to be included in the study.

There were no intraoperative complications. Additional procedures performed during the same surgery included flexor carpi ulnaris tenotomy, flexor carpi radialis lengthening, flexor pollicis longus lengthening, carpal wedge osteotomy,\(^6\) ulnar nerve transposition, extensor carpi ulnaris to extensor carpi radialis brevis transfer, thumb metacarpal osteotomy, thumb-index web space deepening, adductor pollicis brevis lengthening, thumb metacarpophalangeal joint arthrodesis, and extensor pollicis longus rerouting.\(^{2,4}\) None of these procedures affected the rehabilitation protocol for the elbow release or osteotomy. A summary of concomitant and subsequent surgeries is listed in Table 1.

The patients were sorted into 2 groups. In the simultaneous group, the patients had a simultaneous posterior elbow release and humeral rotational osteotomy. In the release group, the patients had only a posterior elbow release. Mann-Whitney U test was used for comparison of means between groups. Statistical significance was set at \(P\) less than .05.

### Operative technique

The patient is placed supine with the arm prepared to the axilla. A thin sterile tourniquet is occasionally used for hemostasis if the arm is of adequate size. The skin is incised directly posteriorly from the musculotendinous junction of the triceps to just past the olecranon. Full-thickness flaps are raised to the level of the triceps fascia. The ulnar nerve is identified and transposed anteriorly into a subcutaneous pocket.

The triceps tendon is incised in a distally based V-shaped incision just distal to the musculotendinous junction. The triceps tendon flap is elevated off the triceps using bipolar electrocautery. The posterior elbow joint capsule including the medial and lateral gutters is divided up to the level of the lateral and medial collateral ligaments, being sure to leave the ligaments intact.

When a humeral osteotomy is to be performed, the radial nerve is identified and protected. The medial head of the triceps is elevated subperiosteally off of the humerus from medial to lateral and distal to proximal up to the level of the spiral groove ensuring that the radial nerve is protected. If a plate is to be used, a 6-hole plate is placed on the bone and the distal holes drilled and measured for screws (Fig. 2). An osteotomy is then made with an oscillating saw while protecting the radial and ulnar nerves. The plate is then affixed distally and the arm rotated externally until elbow flexion is aimed to bring the hand toward the mouth, typically in a position of 15\(^\circ\) neutral rotation in adduction. The proximal screw holes are then drilled and the plate is fixed in compression. A second plate may be added for additional stability (Fig. 3). If K-wires are used, an osteotomy is performed at the metaphysealsupracondylar junction and crossing K-wires are used to stabilize the osteotomy (Fig. 4). The choice of rotation is made based on 2 factors: sufficient external rotation to reach the mouth without having to abduct the shoulder and to perform bimanual tasks with the hands facing each other. Parents routinely state that hand function improves, an improvement we credit to the new ability to see the palm of the hand after osteotomy.

The triceps is then repaired in a lengthened V-Y fashion using nonabsorbable suture. Range of motion is again checked to make sure that the hand can reach the mouth and that the ulnar nerve does not kink. The skin is then closed in layers and a long-arm cast or posterior orthosis with slabs is placed with the elbow in flexion.

In the release group, patients were fitted with an orthosis for 2 to 3 weeks. In the simultaneous group, long-arm cast immobilization was 4 to 6 weeks for a single plate or pins and 2 to 3 weeks for double plates. Following immobilization, patients in both
groups were then begun on a range of motion protocol. If more aggressive therapy was needed to maintain motion, patients were supplemented with alternating flexion and extension orthoses as warranted for a minimum of 2 months. The transition from a single plate to K-wires and finally to dual plating represented a change in practice at our institution over time in an effort to reduce the immobilization time and eliminate retained hardware secondary to the osteotomy. Patients were selected for simultaneous release and osteotomy if they had an internal rotation contracture sufficient to preclude hand-to-mouth and bimanual tasks from doing a release alone.

### RESULTS

Forty-three procedures in 38 patients were included. There were 18 girls and 20 boys. The simultaneous group consisted of 13 procedures and the release group of 30. The simultaneous group was older than the release group (5.7 years vs 3.3 years). Average long-term follow-up was 39.8 months for the simultaneous group and 44.5 months for the release group. Average preoperative flexion and arc of motion were significantly higher in the simultaneous group than the release group (Table 2).

At early follow-up, patients in both the release and the simultaneous groups increased their total arc of motion (release group, mean of 27° to mean of 70°; simultaneous group, mean of 40° to mean of 59°). The increase in total arc of motion was mainly through an increase in flexion. Both groups lost some passive extension after surgery (release group, 27° flexion contracture postoperative vs 0° preoperative; simultaneous group, 40° flexion contracture postoperative vs 2° preoperative). At early follow-up, the simultaneous group lost more extension and had a significantly smaller total arc of motion than the release group (Table 3; Fig. 5). There was a significant difference in postoperative flexion and postoperative arc of motion at 1- to 2-year follow-up with the simultaneous group lacking more terminal extension and with a smaller arc of motion than the release group. At more than 2 years follow-up (average of 44.5 months and 39.8 months between the release and the simultaneous groups, respectively), the release group had a smaller flexion

### TABLE 1. Concomitant Procedures Performed

<table>
<thead>
<tr>
<th>Concomitant Procedures</th>
<th>Release Group, n = 30</th>
<th>Simultaneous Group, n = 13</th>
</tr>
</thead>
<tbody>
<tr>
<td>Thumb MCP osteotomy</td>
<td>1 (3.3)</td>
<td>1 (7.7)</td>
</tr>
<tr>
<td>Syndactyly release</td>
<td>1 (3.3)</td>
<td></td>
</tr>
<tr>
<td>First adductor release</td>
<td>1 (3.3)</td>
<td></td>
</tr>
<tr>
<td>Campylodactyly reconstruction</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Finger z-plasty</td>
<td>2 (6.7)</td>
<td>1 (7.7)</td>
</tr>
<tr>
<td>FDS tenotomy</td>
<td>2 (6.7)</td>
<td></td>
</tr>
<tr>
<td>ECU to ECRB transfer</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Carpal wedge osteotomy</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Thumb contracture release</td>
<td>1 (3.3)</td>
<td></td>
</tr>
<tr>
<td>Stiletto flap</td>
<td>1 (3.3)</td>
<td></td>
</tr>
<tr>
<td>EPL transposition</td>
<td>1 (3.3)</td>
<td></td>
</tr>
</tbody>
</table>

ECRB, extensor carpi radialis brevis; ECU, extensor carpi ulnaris; EPL, extensor pollicis longus; FDS, flexor digitorum superficialis; MCP, metacarpophalangeal.

**FIGURE 2:** Single-plate fixation of humeral rotational osteotomy. (Used with permission of Shriners Hospitals for Children—Philadelphia. All rights reserved.)
contracture and maintained a larger arc of motion. The difference in the arc of motion between the release group and the simultaneous group was statistically significant.

We also looked at a subset of our data, comparing the results of crossed K-wires and single plating (given the similar immobilization times) versus dual plating, which had a shorter immobilization time (Table 4; Fig. 6). Patients who underwent dual plating (4) had a much larger arc of motion at early follow-up than the K-wire or single-plate fixation group (8), despite having similar preoperative extension, flexion, and arc of motion. This difference was also significant at late follow-up. When comparing the subgroup of patients who underwent dual plating with the release group, there were similar postoperative flexion contractures and similar arc of motion at late follow-up (Tables 3, 4; Figs. 5, 6).

Complications

One patient in the simultaneous group refractured 1 week after the pins were removed. The osteotomy had been fixed with crossed K-wires with pins removed at 6 weeks. He was treated to union in a cast and his ultimate range of motion at 17 months was from 40° to 95°. Another patient in the simultaneous group whose osteotomy was fixed with a 2.7-mm limited-contact dynamic compression plate had bending of the plate noted 1 month after surgery. He went on to union in a cast and his ultimate range of motion was 65° to 100°. There were no infections or other complications noted in either group.

DISCUSSION

For patients with amyoplasia, some authors have recommended performing all surgeries at once rather than sequentially. We hypothesized that combining an external rotation osteotomy of the humerus with a posterior elbow release would allow treatment of 2 common problems under 1 surgical approach. Across both treatment groups, the total arc of motion increased in all patients. However, patients who had only a posterior capsular release had greater gains in flexion (102° vs 88°), less loss of extension (35° vs 45°), and greater total arc of motion (67° vs 43°) at long-term follow-up than patients who also had a simultaneous humeral rotational osteotomy.
Our results confirm previous reports that a posterior elbow release improves elbow flexion and total arc of motion in these patients.\textsuperscript{1,5,8,9} The difference was achieved at the 1- to 2-year follow-up and maintained through long-term follow-up. The largest series published prior to this study included 29 elbows in 23 patients.\textsuperscript{5} Average postoperative flexion was 100\(^\circ\), loss of extension 34\(^\circ\), and arc of motion 66\(^\circ\). These results are similar to the results from our posterior elbow release only group (flexion, 102\(^\circ\); loss of extension, 35\(^\circ\); arc of motion, 67\(^\circ\)).

In patients with simultaneous humeral osteotomy and posterior elbow release, the gains in arc of motion were significantly less. This is largely due to loss of elbow extension. It is unclear if the difference in outcomes is due to the increased scarring from the additional surgery, the difference in rehabilitation protocols, or an unknown factor. Because patients who underwent dual plating achieved the same results as the release-only group, while following the same postoperative protocol, and the single plate or pinning group had a worse result after following a delayed-mobilization protocol, our conjecture is that the difference is due to the delay in mobilization. Early range of motion is a mainstay of elbow contracture release. Based on these findings, we implemented an institutional change in our clinical practice in which patients who are indicated for simultaneous osteotomy and elbow release receive dual plating to allow for early range of motion.

Our prior experience with isolated humeral rotational osteotomies was to use a single age-appropriate compression plate placed dorsally just proximal to the olecranon fossa. Owing to several peri-implant fractures in this high fall-risk and relatively osteoporotic population, we switched to a slightly undersized single compression plate but added long-arm orthosis immobilization for 4 to 6 weeks to our postoperative protocol. Our initial fixation method for the combined procedures was, therefore, a single compression plate with immobilization maintained until union, typically for 4 to 6 weeks. Seeing that our early anecdotal
TABLE 4.  Range of Motion Data for Dual Plating Versus Single Plating or K-Wires

<table>
<thead>
<tr>
<th></th>
<th>K-Wires or Single Plate</th>
<th>Dual Plating</th>
</tr>
</thead>
<tbody>
<tr>
<td>Number of patients</td>
<td>8</td>
<td>4</td>
</tr>
<tr>
<td>Preoperative flexion, ° (range)</td>
<td>42 (0 to 110)</td>
<td>53 (50 to 60)</td>
</tr>
<tr>
<td>Preoperative extension, ° (range)</td>
<td>0</td>
<td>7.5 (0 to 30)</td>
</tr>
<tr>
<td>Preoperative arc, ° (range)</td>
<td>42 (0 to 110)</td>
<td>45 (30 to 50)</td>
</tr>
<tr>
<td>Early postoperative flexion, ° (range)</td>
<td>96 (90 to 100)</td>
<td>105 (100 to 110)</td>
</tr>
<tr>
<td>Early postoperative extension, ° (range)</td>
<td>45 (10 to 60)</td>
<td>32 (15 to 45)</td>
</tr>
<tr>
<td>Early postoperative arc, ° (range)</td>
<td>51 (30 to 90)</td>
<td>73 (65 to 85)</td>
</tr>
<tr>
<td>Mid postoperative flexion, ° (range)</td>
<td>91 (45 to 110)</td>
<td>95</td>
</tr>
<tr>
<td>Mid postoperative extension, ° (range)</td>
<td>44 (0 to 60)</td>
<td>45</td>
</tr>
<tr>
<td>Mid postoperative arc, ° (range)</td>
<td>47 (15 to 90)</td>
<td>50</td>
</tr>
<tr>
<td>Late postoperative flexion, ° (range)</td>
<td>85 (50 to 120)</td>
<td>93 (80 to 110)</td>
</tr>
<tr>
<td>Late postoperative extension, ° (range)</td>
<td>53 (0 to 75)</td>
<td>27 (15 to 30)</td>
</tr>
<tr>
<td>Late postoperative arc, ° (range)</td>
<td>32 (15 to 50)*</td>
<td>67 (50 to 95)*</td>
</tr>
</tbody>
</table>

*Statistically significant at P < .05.

FIGURE 5: Preoperative, early postoperative, mid postoperative, and late postoperative follow-up of arcs of motion for patients who underwent posterior elbow release alone and posterior elbow release with simultaneous humeral rotational osteotomy. The green area represents that range of motion that was achieved whereas the red areas represent the ranges of motion that are lacking in both terminal flexion and terminal extension.
results were not as good as for the release-only patients, we switched to crossed K-wires to avoid retained hardware in the arm. A similar 4 to 6 weeks of immobilization was required. Again, unsatisfied with our early results, we switched a third time to bicolumnar plate fixation, creating a sufficiently rigid construct to mobilize at 2 to 3 weeks after surgery, but staggering smaller plates to mitigate the risk of peri-implant fracture down the road. Our current protocol is to mobilize the elbow releases at 2 weeks, but we had stopped concurrent rotational osteotomies based on the preliminary results of this study. It appears on longer follow-up that a humeral rotational osteotomy may be able to be performed concurrently with an elbow release if sufficiently rigid fixation is achieved to allow early range of motion. Further research will be needed to determine how simultaneous procedures with early mobilization fare compared with staged procedures.

Among the limitations of this study, it is possible that differences between the groups may have affected our results. The patients in the simultaneous group were older. Younger patients may do better after this type of contracture release. There may also be significant differences in the severity of arthrogryposis between the 2 groups. This study was retrospective and not randomized. Most patients in the release group did not need a humeral osteotomy, and this may suggest that the release group patients had less severe arthrogryposis. However, compared with the simultaneous group, the patients in the release group had less elbow flexion and total arc of motion before surgery, suggesting no difference in the severity of arthrogryposis with regard to the elbow. In addition, little information on patient function was available. Future research would include data on functional outcome, such as the ability to self-feed.

Patients with posterior release alone had significantly greater improvement in total arc of motion and significantly less loss of elbow extension than patients who underwent a simultaneous humeral
osteotomy. However, rigid fixation with early mobilization may yield results comparable with those of the release alone group.

REFERENCES