The Effects of Age on the Outcomes of Elbow Release in Arthrogryposis

Christopher Richards, MD, MS,* Rey Ramirez, MD,* Scott Kozin, MD,† Dan Zlotolow, MD†

Purpose The goal of this study was to observe the effects of posterior elbow release on children with arthrogryposis at various age points: before the age of 2, between the ages of 2 and 3, and after the age of 3.

Methods This study was a retrospective chart review of patients with arthrogryposis who underwent a posterior elbow release for an elbow extension contracture between 2007 and 2014 at one institution. Eighteen procedures in 13 patients, who had a minimum follow-up of at least 2 years, were included in the study. Patients were divided into 3 groups based on their age at the time of surgery: <2 years old, 2–3 years old, and >3 years old. Comparisons of the pre- and postoperative passive arcs of motion were made.

Results The average preoperative arc of motion was 16° (0° to 30°) for the children younger than 2, 33.5° (5° to 60°) for the children 2–3, and 45° (25° to 80°) for the children older than 3. The average postoperative arc of motion was 88.2° (70° to 103°), 60° (15° to 85°), and 54.33° (23° to 70°) for the respective age groups. There was a clinically important difference in the postoperative arc of motion between the children less than 2 years old and both the children 2–3 years old and older than 3 years.

Conclusions This study demonstrates that children who underwent posterior elbow release before the age of 2 had a clinically important increase in their postoperative flexion and overall passive arc of elbow motion compared with older children at medium-term follow-up. The data suggest that earlier release may be better at restoring total passive arc of elbow motion. (J Hand Surg Am. 2018; 1.e1-e6. Copyright © 2018 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, age, posterior elbow release.

Arthrogryposis is a descriptive term for any patient born with contracture of multiple joints. The extent and severity of contractures can vary between subtypes, from patient to patient, and even from side to side. The most common diagnosis is amyoplasia congenita, which is a sporadic (nonheritable) condition often characterized by nearly symmetric limb involvement, with most patients having all 4 limbs affected. The contractures are present at birth and generally improve with time. Maximal gains without intervention are typically made in the first 2 years of life.

Because of hypoplasia or aplasia of muscles throughout the limbs, the upper extremity typically assumes a posture of shoulder adduction and internal rotation, elbow extension, wrist flexion, thumb in palm, and metacarpophalangeal joint extension. Up to 25% of patients with arthrogryposis have elbow involvement, and most of these patients have some
degree of elbow extension contractures. The elbow contractures limit hand-to-mouth motions used for activities of daily living, such as eating. Although active flexion of the elbow makes daily tasks easier and faster to perform, passive elbow flexion is necessary and sufficient to perform hand-to-mouth activities without assistance. Patients typically develop strategies on their own to get their hands to their mouths that follow 4 patterns (Fig. 1).

All of these strategies typically require at least 90° of passive elbow flexion and enough external rotation of the shoulder to clear their chest, usually between 45° and 30° less than neutral rotation. Wrist and forearm position can help or detract from hand-to-mouth function, and may need to be addressed as well. Wrist flexion and ulnar deviation may get the hand closer to the mouth, but only if the forearm can be nearly fully supinated.

When an orthosis and range of motion exercises fail to allow the patient to self-feed, surgical options include: (1) humeral osteotomy to improve the external rotation of the shoulder, (2) posterior elbow release to improve elbow flexion, (3) one-bone forearm to improve supination, and (4) tendon releases and carpal wedge osteotomy to correct ulnar deviation. Posterior elbow capsular release, triceps lengthening, and ulnar nerve transposition (posterior elbow release) have been shown to improve passive elbow flexion and provide better results when performed in isolation. Previous literature suggests good results at an average of 3 years of age. No consensus currently exists regarding the timing of

FIGURE 1: Adaptive measures that children with arthrogryposis use to self-feed. Several adaptive measures that children with arthrogryposis use to help them self-feed, including A using their knee to push their hand to their mouth, B a table, C their other arm, or D using shoulder abduction.

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surgery, although anecdotal reports have suggested that patients may do better at a younger age. The purpose of this study was to assess the effect of age at the time of surgery on the outcome of posterior elbow release. We hypothesized that surgery at an earlier age would result in better outcomes with respect to elbow total arc of motion.

**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 one of the two senior authors. We reviewed the medical records for age, sex, preoperative and postoperative range of motion, and complications. A standard hand-held goniometer was used for all measurements, which were recorded either by one of the two senior authors or the occupational therapists at our institution. The measurement taken at the last clinic visit before surgery (average of 214 days before surgery) was taken as the preoperative range of motion. The postoperative range of motion reported was measured at the most recent long-term follow-up (after a minimum of 2 years) from the date of surgery. In addition, each patient had his or her intraoperative flexion angle measured after posterior elbow release.

We reviewed 62 procedures in 44 patients during the 7-year period of this study. Of those, 18 procedures in 13 patients had a minimum follow-up of 2 years and were included in the study. Of the 6 patients who had bilateral posterior elbow releases, all surgeries were conducted within 8 months of each other and within the child’s same year of life so that they were in the same age group at the time of both surgeries. As a result, these arms were assigned to the same age group for the analysis. Patients were divided into 3 groups based on their age at the time of surgery: <2 years old (3 patients, 5 limbs, average age at surgery 1.4), 2–3 years old (7 patients, 10 limbs, average age at surgery 2.8), and >3 years old (3 patients, 3 limbs, average age at surgery 6.9). The pre-, postoperative, and changes in the passive arcs of motion were compared between groups (Table 1). There were no intraoperative complications.

**Operative technique**

The patient is placed supine with the arm prepped past the axilla. A thin sterile tourniquet (Hemaclear) is used

![Table 1. Comparison of Average Preoperative and Postoperative Elbow Range of Motion Across Age Groups](image)

<table>
<thead>
<tr>
<th>Group</th>
<th>Preoperative Flexion Angle</th>
<th>Postoperative Flexion Angle</th>
<th>Change in Arc of Motion</th>
</tr>
</thead>
<tbody>
<tr>
<td>Children &lt;2 years old</td>
<td>16° (0° to 30°)</td>
<td>102° (0° to 105°)</td>
<td>86° (90° to 120°)</td>
</tr>
<tr>
<td>Children 2-3 years old</td>
<td>33.5° (5° to 60°)</td>
<td>102° (0° to 110°)</td>
<td>68.5° (50° to 120°)</td>
</tr>
<tr>
<td>Children &gt;3 years old</td>
<td>51.7° (25° to 80°)</td>
<td>113.3° (110° to 120°)</td>
<td>61.6° (25° to 100°)</td>
</tr>
</tbody>
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for hemostasis if the arm is of adequate size. The skin is incised directly posteriorly from the musculocutaneous junction of the triceps to just past the olecranon, curving ulnarly around 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 (Fig. 2A).

The triceps tendon is incised in a distally based V-shaped incision just distal to the musculocutaneous junction (Fig. 2B). The triceps tendon flap is elevated off the triceps using bipolar electrocautery (Fig. 2D). The posterior elbow joint capsule, including the medial and lateral gutters, is divided up to the level of the medial and lateral collateral ligaments, being sure to leave the ligaments intact (Fig. 2C).

The triceps is then repaired in a lengthened V-Y fashion using a nonabsorbable suture (Fig. 1E). 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 with an absorbable suture, and a long arm cast or posterior orthosis with side struts is placed with the elbow in maximal flexion (Fig. 2F). Patients were treated with an orthosis for 2 to 3 weeks. After immobilization, patients in all groups were then begun on a range of motion protocol.

RESULTS

The average preoperative arc of motion was 16° (0° to 30°) for the children younger than 2, 33.5° (5° to 60°) for the children aged 2–3, and 45° (25° to 80°) for the children older than 3 (Table 1, Fig. 3). Children ≥3 years old also had, on average, a 6.7° flexion contracture, shifting their preoperative arc of motion into more flexion. The average postoperative arc of motion was 88° (range, 70° to 103°) for the children younger than 2, 60° (range, 15° to 85°) for the children 2–3, and 54° (range, 23° to 70°) for the children older than 3 (Table 1, Fig. 3). The average change in arc of motion was an increase in 68.2° (range, 50° to 93°) for the children younger than 2, 26.5° (range, −25° to 5°)
for the children aged 2–3, and 9.3° (range, −10° to 40°) for the children older than 3. There was a clinically important difference in the postoperative arc of motion and change in arc of motion between the children less than 2 years old and both the children 2–3 years old and older than 3 years.

**DISCUSSION**

Currently, no consensus exists regarding the timing of posterior elbow release in patients with amyoplasia. Many have hypothesized that surgery at an earlier age would result in a better arc of motion after posterior elbow release. Our data are consistent with previous reports in the literature that a posterior elbow release improves elbow flexion and total arc of motion in these patients.3,4,7–10 The data from our cohort suggest that surgical intervention at an age before 2 years results in improved arc of motion and retention of the improved arc of motion at medium-term follow-up. Elbow arc of motion improved across all groups at greater than 2-year follow-up.
Children younger than 2 had an average improvement of 68.2°, age 2–3 had an average improvement of 26.5°, and children older than 3 had an average improvement of 9° from their respective preoperative average arcs of motion. An improvement of 9° is unlikely to be clinically important, suggesting that this age group, on average, did not benefit from release.

Although it is unclear why younger children fare better, possible factors could include that (1) it is easier for parents to do passive motion exercises on younger children, (2) younger children may not scar as profoundly, and (3) selection bias leading to more informed, motivated parents seeking care at an earlier age and therefore more willing to endure the rehabilitation regimen.

Our cohort of patients also had different preoperative arcs of motion. On average, patients younger than 2 years of age had worse arc of motion (16°) compared with the children 2–3 years of age (33.5°) and older than 3 years of age (45°). Maximal gains are typically reached within the first 2 years of life. However, assuming that older children self-selected for release in this study due to inadequate nonoperative gains, these results suggest that gains continue after the first 2 years of life. Although these patients were not followed for this study before surgery, their gains before surgical intervention were achieved through therapy and activities of daily living. Despite these improvements without surgical intervention, the patients undergoing surgery at an early age achieved the largest gains in their postoperative elbow arc of motion and seemed to maintain them through follow-up (Table 1, Fig. 3). This may reflect selection bias in that patients who improved on their own were not included in this surgical cohort.

Other limitations include the following: (1) a relatively small sample size and within that sample size, some of the patients underwent a release on the opposite arm during the same year of life; (2) the study was retrospective; (3) amyoplasia has a varying severity of presentation, which could result in a selection bias for children who had surgery early rather than late; however, this would not explain the differences seen in postoperative arc of motion between groups; (4) although patients saw a gain in motion, it is unclear how long these results last and if the limb remains functional for the patients; (5) dominance was not assessed and compared in patients who underwent bilateral releases.

Patients with arthrogryposis who underwent isolated posterior elbow release at an age of less than 2 had clinically important improvements in elbow arcs of motion at long-term follow-up compared with their older counterparts. The results of this study suggest that patients with arthrogryposis who have an elbow contracture should be operated on before age 2 for maximal gains in elbow arc of motion. Further long-term studies are necessary to determine the lasting effects of posterior elbow release at an earlier age.

REFERENCES