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# Posterior Elbow Capsulotomy with Triceps Lengthening for Treatment of Elbow Extension Contracture in Children with Arthrogryposis

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*Investigation performed at the Shriners Hospital for Children-Twin Cities, Minneapolis, Minnesota, and Shriners Hospital for Children-Northern California, Sacramento, California*

**Background:** Flexion of one elbow is essential to enable children with arthrogryposis to achieve independent function such as self-feeding and self-care of the face and hair. We analyzed the outcomes of posterior elbow capsulotomy with triceps lengthening for the treatment of elbow extension contractures in a series of children with arthrogryposis multiplex congenita.

**Methods:** The medical records of all children with arthrogryposis who had been followed for a minimum of two years after treatment with elbow capsulotomy and triceps lengthening were retrospectively reviewed. The postoperative range of motion and ability to reach the mouth were compared with the preoperative status.

**Results:** Posterior capsulotomy with triceps lengthening was performed in twenty-nine elbows of twenty-three children with an average age of thirty-five months (range, seven months to thirteen years). The average duration of follow-up was 5.4 years. The arc of motion of all twenty-nine elbows improved from an average of 32° (range, 0° to 75°) preoperatively to an average of 66° (range, 10° to 125°) at the time of final follow-up. All children were able to reach the mouth using passive assistance (e.g., table-push, trunk-sway, and cross-arm techniques), and twenty-two children were able to feed themselves independently. No child underwent subsequent tendon transfer surgery.

**Conclusions:** Elbow capsulotomy with triceps lengthening successfully increases passive elbow flexion and the arc of elbow motion of children with arthrogryposis, enabling hand-to-mouth activities. In contrast to studies in which tendon transfer surgery was used to increase elbow flexion, none of the children in this series underwent subsequent tendon transfer surgery.

**Level of Evidence:** Therapeutic Level IV. See Instructions to Authors for a complete description of levels of evidence.

Arthrogryposis is characterized by congenital contractures of two or more joints<sup>1,2</sup>. The term *arthrogryposis multiplex congenita* is used to describe a large heterogeneous group of disorders, and it has been subdivided into syndromic arthrogryposis, distal arthrogryposis, and classic arthrogryposis (amyoplasia)<sup>3</sup>.

Amyoplasia is the most common form treated by orthopaedic surgeons. It is characterized by multiple congenital contractures, typical and symmetric positioning of the limbs, decreased muscle mass and strength, and sporadic occurrence with no known hereditary pattern<sup>1,4-7</sup>. Most commonly, both

upper and lower extremities are involved (quadrimelic), although involvement of the upper or lower extremities alone (bimelic) or very rarely involvement of one arm and one lower extremity (hemimelic) has been reported<sup>8</sup>. Most children with amyoplasia have normal intelligence and life expectancy<sup>9,10</sup>.

The most common pattern of deformity of the upper extremity is internal rotation of the shoulder with weak or absent shoulder girdle muscles; extension contractures of the elbow with weak or absent biceps and brachialis muscles; pronated, flexed, and ulnar deviated wrists with weak or absent wrist extension; and rigid digits with a thumb-in-palm deformity<sup>4,7-9</sup>.

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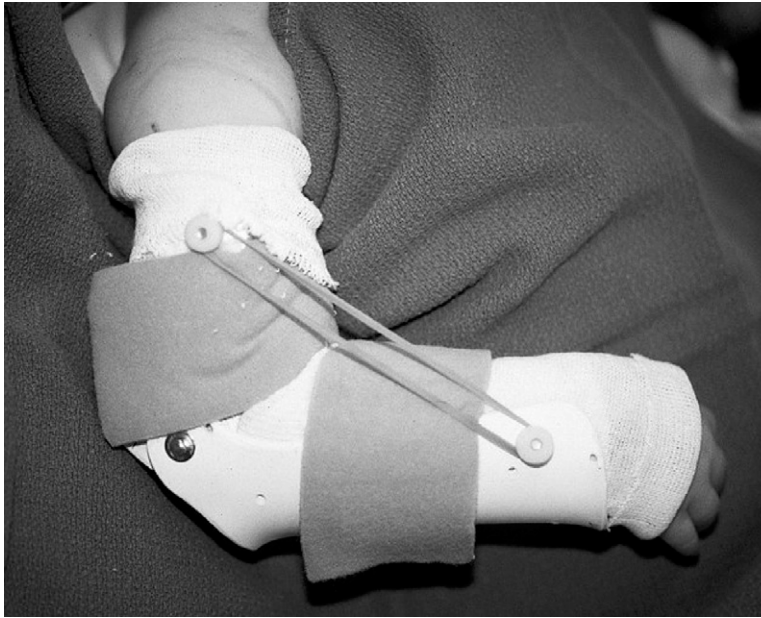


Fig. 1

A typical splint used for postoperative immobilization. The locking hinge can be opened to allow 0° to 90° of passive flexion during the first month to protect the triceps lengthening repair. After the first month of protection, the splint can be locked at night in maximum flexion for an additional two months. If the patient begins to lose elbow extension, the night splint can alternate between maximum flexion and maximum extension.

The degree of stiffness and weakness ranges from mild to severe and is not progressive. Elbow dysfunction poses a substantial functional limitation for these children<sup>2,9-11</sup>. With the elbow stiff in extension, they cannot bring the hand to the mouth to feed themselves or attend to facial hygiene.

The goal of treatment for children with arthrogryposis is to improve their quality of life by facilitating functional independence<sup>9,11,12</sup>. Treatment begins at birth and includes non-operative measures such as range-of-motion exercises, muscle and joint stretching, and splinting to improve passive joint mobility<sup>12-14</sup>. Passive motion of a joint is needed before augmentation of active motion can be considered. Treatment to improve the function of the upper limb requires comprehensive planning with simultaneous assessment of shoulder, elbow, wrist, and hand function. Of these joints, the elbow is the most critical in terms of achieving passive mobility to gain greater functional independence. Flexion of one elbow is essential to enable the child to become functionally independent with regard to self-feeding and self-care of the face and hair; extension of the other elbow is essential for independent toileting as well as transferring if the lower extremities are severely involved. If, after nonoperative treatment, elbow flexion is still insufficient to allow the hand to reach the mouth, two surgical treatment goals can be pursued: improving the passive range of motion and/or improving the active range of motion.

This retrospective review was performed to analyze the outcomes of posterior elbow capsulotomy with triceps length-

ening for the treatment of elbow extension contractures in children with arthrogryposis multiplex congenita. We hypothesized that posterior elbow capsulotomy with triceps lengthening would improve the passive range of motion sufficiently to allow hand-to-mouth function.

### Materials and Methods

The two senior authors (A.V.H. and M.A.J.) performed posterior elbow capsulotomy and triceps lengthening to treat an extension contracture due to arthrogryposis in forty-one consecutive elbows in thirty-two children at two institutions between 1991 and 2004. After institutional review board approval was obtained at both institutions, the records on these patients were reviewed to determine the preoperative and postoperative active and passive ranges of motion of the elbow, obtain information about functional ability, and identify other recorded limb operations. Range-of-motion data consisted of goniometric measurements of passive and active movement that had been routinely performed by the therapist or physician. Strength was graded with manual muscle testing. Functional abilities were identified on the basis of the history and observation. Twelve elbows of nine children were excluded from the study because they had been followed for less than two years after the surgery, leaving twenty-nine elbows in twenty-three children as the subjects of this report.

Nonoperative attempts to increase passive elbow flexion to >90° had failed for all twenty-three children prior to the

surgery. The indications for surgery were  $<90^\circ$  of passive elbow flexion and an inability to reach the hand to the mouth.

### Operative Technique

Patients undergoing bilateral simultaneous release were positioned supine, and those undergoing unilateral release were placed in a lateral decubitus position. A sterile tourniquet was used initially during dissection through a curvilinear posterior incision. In patients with minimal elbow movement, the posterior aspect of the olecranon needed to be carefully identified as the entire limb's outline was so internally rotated that the medial epicondyle could be mistaken for the olecranon. In most patients, the ulnar nerve was specifically identified as it passed through the medial intermuscular septum. The cubital tunnel was released, and the ulnar nerve was traced to the flexor carpi ulnaris innervation. A vessel loop was placed around the nerve to protect it during subsequent dissection. Once the ulnar nerve had been identified, released, and protected, the sterile tourniquet was removed to allow greater proximal dissection and triceps mobilization. The triceps was isolated, mobilized medially and laterally, and lengthened with either a z-lengthening or a V-Y advancement. Most commonly, the triceps was incised in a "V" fashion so that the central tongue was based on the olecranon and the two lateral limbs included tendon over as great a length as possible, from the proximal muscular portion distally to the olecranon insertion. The posterior aspect of the capsule was then incised at the tip of the olecranon to allow identification of the joint surface. The arthrotomy was then extended medially and laterally to allow maximal elbow flexion with a gentle passive stretch. It was necessary to extend the capsular release around the medial and lateral sides to include the posterior edges of the medial and lateral collateral ligaments. The elbow was flexed as much as possible (in excess of  $90^\circ$ ) while allowing contact between the distal ends of the lateral triceps limbs and the proximal end of the central tongue of the triceps. The triceps was then repaired in an elongated position with use of a nonresorbable or reinforced suture. The skin was closed, a light dressing was applied, and the limb was placed in a pre-fabricated custom hinged elbow brace or long arm cast.

Postoperatively, the elbow was immobilized in  $90^\circ$  of flexion with a passive range of motion allowed as soon as the patient tolerated it. Therapy was advanced to include hand-to-mouth activities with passive flexion limited to  $90^\circ$  during the first month to protect the triceps lengthening and advanced thereafter to full passive flexion. Use of the splint, shown in Figure 1, was discontinued during the day after four to six weeks but was continued at night for six months.

The Student paired t test was used to compare the preoperative and postoperative elbow flexion, extension, and arc of motion.

### Results

The study group included ten girls and thirteen boys. The age at the time of surgery averaged thirty-five months (range, seven months to thirteen years). The average duration of follow-up was 5.4 years (range, two to eleven years).

All of the children had arthrogryposis, with the disease confined to limb abnormalities. All patients had normal intelligence. Two patients had multiple joint contractures involving only the upper extremities, and twenty-one patients had multiple joint contractures in all four limbs. Five patients were unable to walk. A total of twenty-nine lower-extremity procedures had been performed in these children. Other upper-extremity procedures included six dorsal carpal wedge osteotomies, two humeral derotational osteotomies, one thumb-in-palm release, and one partial syndactyly release.

Prior to the surgery, the average arc of passive motion was  $32^\circ$  (range,  $0^\circ$  to  $75^\circ$ ), with an average of  $38^\circ$  of flexion (range,  $0^\circ$  to  $80^\circ$ ) and  $1^\circ$  of extension (range,  $40^\circ$  of extension to a  $55^\circ$  flexion contracture). An arc of at least  $90^\circ$  of passive flexion was achieved in all children intraoperatively (see Appendix).

At the time of final follow-up, the average arc of motion had increased to  $66^\circ$  (range,  $10^\circ$  to  $125^\circ$ ), with an average of  $100^\circ$  of flexion (range,  $20^\circ$  to  $155^\circ$ ) and an average lack of extension of  $34^\circ$  (range,  $0^\circ$  to  $90^\circ$ ). The average increase in the arc of motion was  $33^\circ$  (range,  $-15^\circ$  to  $105^\circ$ ). The increases in flexion, extension, and the arc of motion between the preoperative and postoperative visits were significant ( $p < 0.0001$ ).

The strength data recorded in the medical record preoperatively indicated that twenty-seven elbows had grade-0/5 biceps strength and two elbows had grade-3/5 biceps strength. At the time of final follow-up, biceps strength was grade 0/5 in twenty-two elbows, grade 1/5 in two, grade 2/5 in three, grade 3/5 in one, and grade 4/5 in one.

At the time of final follow-up, twenty-two of the twenty-three children were able to feed themselves with the hand on the operatively treated side. The one child who was unable to feed himself had inadequate hand function to carry out this task. All children were able to reach the mouth with the hand, but the twenty-one children with less than grade-3 elbow-flexion strength required the use of passive assistance, as shown in Figures 2-A and 2-B. Examples of techniques that the children used for passive assistance were propping of the arm against the leg or other arm to push it to the mouth (Fig. 2-A), pushing the limb toward the mouth against the edge of a table (Fig. 2-B), bending the neck toward the hand, and swinging the arm and using lumbar lordosis and gravity to bring the hand to the mouth.

No child underwent a subsequent tendon transfer to increase elbow flexion to facilitate self-feeding.

### Discussion

We hypothesized that posterior elbow capsulotomy with triceps lengthening would improve hand-to-mouth function. The range of flexion of the twenty-nine elbows improved by an average of  $33^\circ$  after the procedure. The average arc of motion changed from an arc in extension ( $1^\circ$  of extension to  $38^\circ$  of flexion) to an arc in flexion ( $34^\circ$  of flexion to  $100^\circ$  of flexion). Twenty-two of the twenty-three children were able to feed themselves postoperatively, and the one child who could not do so did not have adequate hand function to allow independent self-feeding. Commonly, children with arthrogryposis augment elbow flexion with wrist flexion and cervical



Fig. 2-A

**Figs. 2-A and 2-B** Postoperatively, the patients in the study had improved passive elbow flexion, with no active elbow flexion. These children used adaptive techniques to reach the hand to the mouth. **Fig. 2-A** Swinging the arm and using the contralateral arm to bring the hand to the mouth for eating was the most common adaptive mechanism while standing.

flexion in order to be able to feed themselves. Only six of these children had had a dorsal carpal wedge osteotomy of the wrist, as many children used the flexion of the wrist to augment flexion of the elbow in order to reach their mouth.

Use of a posterior elbow capsulotomy and triceps lengthening to improve passive mobility of the elbow has been described previously<sup>2,6,9,11,12,15,16</sup>. In what we believe to be the largest previous series, Axt et al.<sup>12</sup> evaluated twenty-two elbows in sixteen children who had been treated at an average age of 4.4 years. After an average duration of follow-up of eight years, there was an average improvement of 19° of elbow motion, from an average preoperative arc of movement of 20° to an average postoperative arc of movement of 39°. Hand-to-mouth function was improved in seventeen elbows and no functional gain or loss was noted in five. Two of the children had active flexion of the elbow after the capsular release.

Our results are slightly better than those reported by Axt et al.<sup>12</sup>, but they had a longer follow-up, which may account

for the difference. In both series, the children did not exhibit measurable preoperative biceps strength and, in some cases, did exhibit biceps strength postoperatively. In our series, seven of the twenty-nine elbows had some biceps strength postoperatively.

The results of posterior elbow capsular release with triceps lengthening have been examined in other, smaller studies. Lloyd-Roberts and Lettin<sup>17</sup> reported success in all of seven patients who had been so treated. Williams<sup>18</sup> reported an improved range of motion in four of four patients. Bennett et al.<sup>16</sup> reported a postoperative improvement of the passive range of motion to >90° in three of four patients who had been treated with the procedure and a change in the position of elbow motion to greater flexion (without increasing the arc) in the fourth patient. One of us (A.V.H.) and colleagues<sup>15</sup> reported on a series of six elbows in which the average preoperative arc of 17° improved to an average postoperative arc of 67° at five years postoperatively; two patients had concomitant or subsequent tendon transfers.



Fig. 2-B

Pushing the arm against a table toward the mouth was the most common adaptive mechanism for eating while sitting.

Thus, general improvement was reported in each of these studies, although they included small numbers of patients.

Most authors have recommended that, once passive motion is restored, a tendon transfer be performed to achieve active flexion of the elbow, with surgical options including triceps transfer, pectoralis transfer, latissimus dorsi transfer, and Steindler flexorplasty<sup>9,11,12,15,16,19-23</sup>. There has not been a consensus in the literature regarding which tendon transfer is most beneficial. Recent articles have highlighted the long-term complication of severe flexion contracture of the elbow after tendon transfer to improve elbow flexion in patients with arthrogryposis<sup>15,21</sup>. In the series by Axt et al.<sup>12</sup>, five of the elbows treated with posterior capsular release subsequently underwent a triceps-to-biceps transfer, with achievement of active flexion of three of the five elbows.

None of our patients were treated with subsequent tendon transfer for several reasons. First, only two children had clinically measurable strength in the biceps prior to the elbow capsular release. At the time of final follow-up, biceps strength was grade 1 of 5 in two elbows, grade 2 in three elbows, grade 3 in one elbow, and grade 4 in one elbow. Allowing a passive range of motion of the elbow may have unmasked some elbow biceps function in these patients. Subsequent tendon transfer surgery is not necessary for children with adequate biceps strength. Even in our series, in which only two elbows demonstrated grade 3 or 4 biceps strength, it does not appear that tendon transfer was necessary to improve hand-to-mouth function. Second, many of these children did not have adequate muscles to transfer. Potential donor muscles for transfer include the triceps, the latissimus dorsi, the pectoralis major,

and the flexor pronator (a Steindler flexorplasty). Children with amyoplasia often have inadequate muscle mass in either the latissimus dorsi or the pectoralis major to allow transfer. Using the triceps as a donor takes away elbow extension power and provides unopposed elbow flexion. If one arm has a flexion contracture of  $>90^\circ$  and the other arm is in extension the individual loses the ability to transfer an object from one hand to the other and all bimanual skills can be lost, creating a substantial impairment. With the Steindler flexorplasty, there is a risk of tightening of the wrist and finger flexors in patients with wrist and finger flexor contractures. Finally, the children in this study, although substantially impaired physically, were creative and found ways of compensating. Pushing the forearm against the table toward the mouth was the most common adaptive mechanism for eating while sitting. Swinging the arm and using lumbar spine lordosis and gravity to bring the hand to the mouth or face was the most common adaptive mechanism while standing. In our opinion, the risks of problems from tendon transfer after posterior elbow capsular release outweigh the benefits.

The goal of treatment of children with arthrogryposis is to improve their quality of life by facilitating functional independence. Because children with this condition are often severely impaired, each proposed treatment must be carefully assessed to ensure that unintended consequences do not decrease their overall independence. Although children with other conditions may benefit from tendon transfer surgery to restore active elbow flexion, the children in this study met the goal of self-feeding following posterior elbow capsulotomy with triceps lengthening alone, and subsequent tendon transfer surgery was not warranted.


This study had several strengths. To our knowledge, it involved the largest reported series of patients with this relatively rare condition. It provides mid-term to long-term follow-up data on a rare condition, allowing assessment of the effects of several years of growth and function on the results of the surgery. The patients were treated by two surgeons at different institutions in different parts of the country, minimizing the effects of regional or institutional biases that might be present in the treatment of this disease and demonstrating the applicability of this operation to different populations with amyoplasia.

This study also had several weaknesses. It was a retrospective review, and detailed data on shoulder, wrist, and finger involvement were not consistently available. Although we re-

ported on strength, range of motion, and the patients' ability to feed themselves independently, representing all three components of the World Health Organization International Classification of Disability (impairment [strength and range of motion], activity [hand to mouth], and participation [self-feeding]), we did not assess the subjects' overall function. Finally, the retrospective study design did not allow us to compare a group of children who had subsequent tendon transfer surgery with a group of patients who did not.

In this study, we assessed a group of children with amyoplasia who had severe elbow extension contractures that had not improved adequately with splinting and stretching exercises. A posterior elbow capsular release with triceps lengthening, performed when the children were an average of thirty-five months of age, reliably improved the passive arc of motion by an average of  $33^\circ$  and changed the arc of motion to a more flexed position, allowing the hand to reach the mouth for independent feeding, for twenty-two of the twenty-three children. The functional independence with regard to self-feeding was gained without subsequent tendon transfer surgery.

### Appendix

 A table showing the results for the individual patients is available with the electronic versions of this article, on our web site at [jbjs.org](http://jbjs.org) (go to the article citation and click on "Supplementary Material") and on our quarterly CD-ROM (call our subscription department, at 781-449-9780, to order the CD-ROM). ■

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