Surgical Treatment of Arthrogryposis of the Elbow

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The purpose of this study was to analyze our results of surgical treatment of arthrogryposis of the elbow and to compare our tendon transfer results using range of motion (ROM) criteria versus functional use criteria. Eighteen tendon transfers for elbow flexion in 14 children with arthrogryposis with an average follow-up period of 4 years (range, 1–14 years) and 6 elbow capsulotomies with triceps lengthening in 6 children with arthrogryposis with an average follow-up period of 5 years (range, 2–9 years) were evaluated. Each child was assessed by a questionnaire regarding functional use of the upper extremity, physical examination of ROM and strength, and a videotaped activities of daily living evaluation. Tendon transfer results were classified and compared using 2 methods of evaluation: postoperative strength and ROM and effective functional use of the tendon transfer to perform activities of daily living. The 6 elbow capsulotomies improved from an average preoperative arc of 17° of motion (average extension, −2°; average flexion, 19°) to an average final follow-up arc of 67° (average extension, −25°; average flexion, 92°). The 18 tendon transfers evaluated by strength and ROM criteria showed 9 triceps to biceps transfers in 9 arms (7 good, 1 fair, and 1 poor), 5 pectoralis to biceps transfers in 4 arms (1 good, 3 fair, and 1 poor), and 4 latissimus dorsi to biceps transfers in 3 arms (2 good and 2 fair). Evaluation by functional use criteria gave the same result in 13 transfers and downgraded the result in 5; the downgraded results were due to resultant flexion contracture or limited functional use because the transfer was in the nondominant arm. Based on this review, optimal surgical candidates for tendon transfer are children older than 4 years, who have full passive ROM of the elbow in the dominant arm, and at least grade 4 strength of the muscle to be transferred. (J Hand Surg 1998;23A:1063–1070. Copyright © 1998 by the American Society for Surgery of the Hand.)

Arthrogryposis multiplex congenita is a disease involving multiple joint contractures with deficient musculature. Elbow joint involvement is frequent. In a review of 114 patients with arthrogryposis, Gibson and Urs[1] reported that 72% had upper extremity involvement; of these patients, the wrist was most commonly affected (32%), closely followed by the hand (26%), elbow (25%), and shoulder (19%). Similarly, Lloyd-Roberts and Lettin[2] examined 39 patients with arthrogryposis and reported that 59% had elbow involvement; extension contracture was the most common deformity.

Elbow dysfunction poses a significant functional disability for these children. The elbows are frequently contracted in extension. Most children with arthrogryposis have bilateral involvement, adjacent joint contractures at the wrist and shoulder, and significant muscle weakness, adding further difficulty in adapting to the elbow deformity. Surgery is indicated for improvement of passive or active elbow functions. For the child with bilateral elbow extension deformities unresponsive to splinting and stretching,

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Received for publication November 16, 1994; accepted in revised form May 1, 1998.

No benefits in any form have been received or will be received from a commercial party related directly or indirectly to the subject of this article.

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0363-5023/98/23A06-0002$3.00/0

elbow capsulotomy and triceps lengthening provide passive ability for hand-to-mouth activities. For the child with functional passive elbow range of motion (ROM) that lacks an active flexor, tendon transfer provides significant functional improvement. Because of the complexity of the deformities in these children, a complete functional evaluation of the severity of disease involvement, including an assessment of adjacent joint involvement, muscle strength testing, and determination of the functional needs of the arms as assists for impaired ambulation are necessary to appropriately plan surgical intervention.

The purpose of this study is twofold. First, this study analyzes our treatment results of 4 surgical procedures for elbow flexion dysfunction in children with arthrogryposis: elbow capsulotomy/triceps lengthening and 3 types of tendon transfers. This includes comparing the results of 3 specific transfers for active elbow flexion: pectoralis major, latissimus dorsi, and triceps. Second, this study compares tendon transfer results using traditional ROM and strength criteria versus using functional use outcome criteria.

Table 1. Upper Extremity Assessment

	Affected	*Surgery*
Shoulders	Yes (18)	Humeral derotation osteotomy (1)
Elbows	Yes (18)	Triceps lengthening and capsulotomy (6)
		Triceps-to-biceps transfer (9)
		Pectoralis-to-biceps transfer (5)
		Latissimus-to-biceps transfer (4)
Wrist	Yes (18)	Radius extension osteotomy (3)
		Radius extension osteotomy with FCU/FCR to ECRB transfer (2)
		Carpal extension osteotomy (1)
		Repeat radius osteotomy (1)
		Proximal row carpectomy (1)
		Flexor tendon lengthening (1)
Hands	Yes (18)	Syndactyly releases (8)
		Thumb extensor transfer (1)

Numbers of patients indicated in parentheses.
FCU, flexor carpi ulnaris; FCR, flexor carpi radialis; ECRB, extensor carpi radialis brevis.

Materials and Methods

Sixty-one children at Children's Hospital of Boston and Shriners Hospital for Crippled Children-Twin Cities Unit (Minneapolis, MN) diagnosed with arthrogryposis multiplex congenita consented and completed a comprehensive evaluation. By chart review and patient/parent interview, all medical problems and previous surgical and nonsurgical interventions were documented. Forty-three of the 61 children with arthrogryposis (70%) had involvement of their elbows. Eighteen of the 43 patients with elbow involvement (42%) were treated surgically and are the subjects of this study.

The 18 patients underwent 24 elbow procedures: 6 elbow capsulotomies with triceps lengthening in 6 children and 18 tendon transfers for elbow flexion in 14 children. Tendon transfers included 9 triceps to biceps transfers, 5 pectoralis to biceps transfers, and 4 latissimus to biceps transfers. Four of these patients had 2 procedures: 1 staged bilateral pectoralis to biceps transfer (patient 11), 1 staged bilateral latissimus to biceps transfer (patient 14), 1 staged right triceps to biceps and left pectoralis to biceps transfer (patient 6), and 1 concomitant elbow capsulotomy with triceps to biceps transfer. One patient (no. 9) underwent 3 staged procedures: elbow capsulotomy with triceps lengthening followed by a failed triceps to biceps transfer at an outside institution, then by a latissimus to biceps transfer at our institution. Three of the 24 procedures were performed at outside institutions. Elbow capsulotomy with triceps lengthening was evaluated only in the 4 children not treated with additional tendon transfer.

Elbow capsulotomy with triceps lengthening was indicated in children lacking 90° of passive elbow flexion after completing a program of ROM exercises and/or serial static splinting or casting. Tendon transfer was indicated in children lacking active elbow flexion with at least 90° of passive elbow motion. Tendon transfer choice was at the surgeon's discretion.

All 18 children had classic arthrogryposis multiplex congenita with disease confined to limb abnormalities and no evidence of central nervous system involvement or other syndromic features. Ten of the patients walked independently without ambulatory aids, 3 walked limited amounts with ambulatory aids, and 5 were wheelchair dependent. All 18 patients had involvement of their shoulders, elbows, wrists, and fingers; all upper extremity surgical procedures are listed in Table 1.

All patients were physically examined for active and passive ROM and strength in each of the patients' upper and lower extremity joints. Those children treated by tendon transfer had, in addition, a functional assessment, including completion of a questionnaire regarding functional use of the upper extremity and examination by observation of activi-

Table 2. Tendon Transfer Results

Result	*Strength and ROM Assessment*		*Functional Use Assessment*
	Postoperative Strength	*Postoperative Active Flexion*	*Use in ADL*
Good	4/5	>90°	Actively incorporates transfer; rare adaptive mechanisms* used
Fair	3/5	<90°	Transfer augments elbow flexion with occasional adaptive mechanisms*
Poor	0–2/5	None	Uses adaptive mechanisms* for elbow flexion or significant loss of other ADL function due to tendon transfer

ROM, range of motion; ADL, activities of daily living.
* Adaptive mechanisms for elbow flexion: table push, trunk thrust, or cervical bending (see text for descriptions).

ties of daily living (ADL). The ADL evaluation assessed each child's ability to self-feed, brush their own hair, drink from a cup, write, use the toilet, and use ambulatory aids as necessary. The ADL evaluation was performed in the office and was also reviewed later by video tape.

Those children who had elbow flexion tendon transfers were evaluated by 2 methods of evaluation (Table 2). The first method correlated results according to postoperative strength and ROM. Criteria included grade 4 strength with over 90° of active flexion for a good result, grade 3 strength with less than 90° of active flexion for a fair result, and grade 0 to 2 strength for a poor result.

The second method of evaluation rated the tendon transfer result on how effectively the patient incorporated the elbow tendon transfer in carrying out the ADL. Three "adaptive mechanisms" to compensate for loss of active elbow flexion were seen among the children: table push, trunk thrust, and cervical bending. With table push, the children bent their elbows by pushing their forearms against the table at which they were seated to bring their hands to their mouths. With trunk thrust, the patients would swing their arms into an elevated position by swaying their trunk and using gravity to bend their arms. With cervical bending, the patients would bend their necks and use their mouths to manipulate objects or to eat food directly off the table. Those patients who had a *good* functional result rarely need to use any adaptive mechanisms, patients with a *fair* functional result frequently have to augment their elbow flexion through occasional adaptive mechanism use, and patients with a *poor* functional result were able to use the transfer to help stabilize their hand in space, but required continued use of adaptive mechanisms for elbow flexion. Additionally, any significant deleterious effects of the tendon transfer on loss of other ADL functions (writing, toiletry, ambulation) demerited the tendon transfer result to *poor.*

Operative technique included standard surgical procedures as previously described for posterior elbow capsulotomy with triceps lengthening,[3] triceps to biceps transfer,[4] pectoralis major transfer to biceps,[5,6] and bipolar latissimus dorsi to biceps transfer.[7,8] In 1 case, a myocutaneous bipolar latissimus dorsi transfer was used. The bipolar transfers were secured to the coracoid proximally; if the coracoid was hypoplastic, the acromion was used for proximal fixation. In all transfers, the tendon insertion was performed through a separate anterior Henry incision in the antecubital fossa. If the biceps tendon was present, it was used for the insertion; otherwise, a drill hole was placed into the radial neck at the level of the normal biceps insertion. The tendon or muscle-fascial extension was passed through the radial neck to a posterior incision where it was secured to the periosteum or looped back onto itself. In 1 case, the radial head was dislocated preoperatively and the insertion was secured through a drill hole in the ulna. Moderate tension of the transfer was set with the elbow between 70° and 90° of flexion.

Postoperative immobilization was in approximately 90° of flexion for 4 weeks using a long arm cast with a sling for the triceps transfer and a shoulder spica for the latissimus dorsi and pectoralis transfers. Active use of the transfer was begun at 4 weeks and resistive exercises were begun at 8 weeks after surgery. The elbow was protected in a sling until the transfer could be actively controlled.

Results

Elbow Capsulotomy and Triceps Lengthening. Six children underwent this procedure at an average age of 4 years (age range, 1–10 years). Four children

Table 3. Tendon Transfer Patient Data

Patient No.	Type of Transfer	Age at Surgery (yr)	Age at Follow-up (yr)	Operated Side	Preoperative PROM (E/F)	Follow-up PROM (E/F)	Follow-up AROM (E/F)	Follow-up Strength	Strength/ROM Outcome	Functional Outcome
1	Triceps	6	8	R (D)	0/105	−55/105	−55/105	4	Good	Good
2	Triceps	5	6	R (ND)	0/110	−35/110	−35/110	4	Good	Fair
3	Triceps	5	9	R (D)	0/95	−35/95	−35/95	4	Good	Good
4	Triceps	11	25	R (D)	0/100	0/90	0/90	4	Good	Good
5	Triceps	7	11	R (D)	0/110	−30/90	−30/90	4	Good	Good
6	Triceps	5	7	R (D)	−5/105	−15/140	−15/140	4	Good	Good
7	Triceps	8	9	R (D)	−10/105	−40/130	−40/95	4	Good	Good
8	Triceps	2	11	L (A)	+10/20*	−95/150	−95/120	3	Fair	Poor
9	Triceps†	2	10	R (D)	NA†	−35/110	−35/35	2	Poor	Poor
10	Pectoralis	4	11	R (ND)	−10/95	−30/100	−30/100	4	Good	Fair
11	Pectoralis	4	13	L (ND)	NA	−20/90	−20/70	3	Fair	Fair
11	Pectoralis	5	13	R (D)	NA	−40/90	−40/80	3	Fair	Fair
12	Pectoralis	6	13	L (ND)	0/95	−15/70	−15/70	4	Fair	Fair
13	Pectoralis†	6	7	L (ND)	NA†	0/90	0/0	1	Poor	Poor
14	Latissimus	13	9	R (ND)	0/100	0/100	0/100	4	Good	Fair
1	Latissimus	14	15	L (ND)	+10/100	+10/100	+10/85	4	Good	Fair
1	Latissimus	14	15	R (D)	0/95	0/95	0/70	3	Fair	Fair
9	Latissimus	9	11	R (D)	−35/110	−35/110	−35/105	3	Fair	Fair

PROM, passive range of motion; E, extension (in degrees); F, flexion (in degrees); AROM, active anti-gravity range of motion (in degrees); D, dominant arm; ND, nondominant arm; A, ambidextrous; NA, not available.
* Concomitant capsulotomy.
† Surgery at outside institution.

underwent elbow capsulotomy and triceps lengthening alone and 2 children had concomitant or subsequent tendon transfer. Before surgery, the average arc of passive motion was 17° (average extension, −2°; average flexion, 19°). All children achieved at least 90° of passive flexion intraoperatively. After an average follow-up period of 5 years (range, 2–9 years), the average arc of passive motion was 67° (average extension, −25°; average flexion, 92°). All children achieved and maintained increased elbow ROM. Four of 6 children maintained over 90° of elbow flexion. Intra-articular incongruity limiting further flexion was specifically noted in 2 children.

Triceps to Biceps Transfer. Nine children underwent a triceps to biceps transfer at an average age of 6 years (age range, 2–11 years) with an average follow-up period of 5 years (range, 2–13 years) (Table 3). All children had at least a 0° to 95° arc of passive motion before surgery, with the exception of the 1 child who underwent a capsulotomy and triceps tendon transfer concomitantly. The preoperative triceps strength was at least 4+/5 in all patients. Seven transfers were in the dominant extremity.

At the final follow-up examination, 1 patient had grade 5 strength, 6 patients had grade 4 strength, one child had grade 3 strength, and one child had grade 2 strength. Excluding the child undergoing concomitant capsulotomy, the average preoperative arc of passive motion was 103° (average extension, −1°; average flexion, 104°). In all patients, the final postoperative arc of passive motion was 76° (average extension, −38°; average flexion, 114°). Eight of the 9 patients had loss of passive extension.

The preoperative arc of active motion was 0°. The postoperative arc of antigravity active motion was 61° (range, 0° to 125°). The average active flexion was 87°, with 8 of 9 patients achieving greater than 90° of active flexion.

Using ROM and strength as a measure of outcome, 7 patients were graded as good, 1 was graded fair, and 1 was graded poor. On the basis of functional use as a measure of outcome, 6 patients were rated as good, 1 was rated fair, and 2 were rated poor. The 2 children with poor strength underwent the tendon transfers at the age of 2 years and both also had elbow capsulotomy. The functional outcome result downgraded in 2 children: 1 due to transfer into the nondominant extremity and 1 due to significant loss of ADL function from the resultant flexion deformity. Loss of ADL function due to a flexion contracture of 95° which, combined with extension contracture in the nonoperated elbow, disabled the patient in bimanual activities; specifically, toiletry was hampered because of inability for either hand to help with urination.

Pectoralis to Biceps Transfer Results. Five pectoralis to biceps transfers were performed in 4 children at an average age of 4 years (age range, 4–6 years) with an average follow-up of 5.8 years (range, 3–9 years) (Table 3). All children had at least a 95° arc of passive flexion before surgery. The preoperative pectoralis strength was at least 4/5 in all patients. Three were performed on the left arm, 2 on the right arm. Four were performed on the nondominant extremity and 1 on the dominant extremity. One patient had bilateral procedures.

At the final follow-up examination, strength was grade 4 in 2 children, grade 3 in 1 child bilaterally, and grade 1 in 1 child with surgery at an outside institution. Loss of preoperative muscle strength was noted in 3 of 5 children.

The average preoperative arc of passive motion was 90° (average extension, −5°; average flexion, 95°). The average postoperative arc of passive motion was 75° (average extension, −21°; average flexion, 95°). On average, 16° of passive extension was lost and no passive flexion was gained.

The preoperative active motion was 0°. The postoperative arc of active antigravity motion was 41° (range, 0° to 70°). The average active flexion was 64°, with 1 of 5 patients achieving greater than 90° of active flexion.

On the basis of strength and ROM, 1 transfer was categorized as good, 3 as fair, and 1 as poor. On the basis of functional use, 4 transfers had fair results and 1 had a poor result. The functional use scale downgraded the result in 1 child; although he had grade 4 strength with 90° of flexion in the transfer, he continued to use his nonoperated dominant hand with adaptive mechanisms for ADL because this was more efficient.

Latissimus Dorsi to Biceps Transfer. Four latissimus dorsi to biceps transfers were performed in 3 children at an average age of 11 years (age range, 6–14 years) with an average follow-up period of 1.5 years (range, 1–3 years) (Table 3). All children had a preoperative passive flexion of at least 95°. The preoperative latissimus strength was assessed as at least 4/5 in all cases. At postoperative follow-up, strength was grade 4 in 2 transfers and grade 3 in 2 transfers. Loss of preoperative muscle strength was noted in 2 of 4 children.

The average preoperative arc of passive motion was 92° (average extension, −6°; average flexion, 96°). The average postoperative arc of passive motion was 93° (average extension, −6°; average flexion, 96°). No change in passive motion was noted.

The preoperative active motion was 0°. The postoperative arc of active antigravity motion was 84° (range, 70° to 100°). The average active flexion was 90°, with 2 of 4 patients achieving greater than 90° of active flexion.

On the basis of strength and ROM, 2 transfers were graded as good and 2 were graded as fair. On the basis of functional use, 2 good results were downgraded to fair. The fair results were due to lack of use in the nondominant hand and to inadequate muscle strength requiring occasional use of adaptive mechanisms to aid elbow flexion.

Complications. No intraoperative or perioperative complications were noted. Two patients underwent subsequent surgery. One patient had undergone a triceps to biceps transfer at an outside institution, with results of 0/5 strength and no active arc of motion. She subsequently was augmented with a latissimus dorsi to biceps at our institution, with improvement of muscle strength to grade 3/5 and an increased active arc of motion to 70°. One patient had an imbrication of the pectoralis to biceps transfer because of weakness and limited functional use, but unfortunately did not have particular improvement of the transfer strength.

Discussion

The first purpose of this study is to analyze our treatment results for surgical intervention for elbow flexion dysfunction due to arthrogryposis. This review, similar to others in the literature (Table 4), is a retrospective study with a relatively small number of patients. For elbow capsular release, our results are similar to those of other studies in that all children improved their passive elbow motion. Other reports have not discussed our operative findings of intra-articular incongruity, which in this study limited passive flexion in 2 of 6 children. For tendon transfers overall, using ROM and strength criteria as a measure of surgical outcome, good, fair, and poor results were obtained in 10, 6, and 2 children, respectively, similar to findings in other studies (Table 4).

Choices for donors available for transfer to the biceps include the triceps,[3,4,9,10] pectoralis,[2,5,9–11] latissimus dorsi,[7,12] Steindler flexorplasty,[13] and sternocleidomastoid.[14] We have no experience with Steindler flexorplasty or sternocleidomastoid transfer

Table 4. Literature Review of Surgical Treatment of the With Arthrogryposis Elbow

Source	*No. of Surgical Procedures*	*Results and Criteria for Results*	*Success*	*Partial*	*Failure*
Elbow capsule release and triceps lengthening					
Lloyd-Roberts and Lettin[2]	7	Permanent, temporary, or no improvement	7	0	0
Williams[3]	4	Improved ROM	4	0	0
Bennett et al.[9]	3	PROM to 90°	3	0	0
Total			14	0	0
Triceps to biceps transfer					
Carroll and Hill[4]	8	Ability to bring hand to mouth	5	1	2
Williams[3]	20	4 strength success 2–4 strength partial; 0 fail	12	7	1
Doyle et al.[10]	7	Improved ROM and single-hand feeding	4	0	3
Bennett et al.[9]	2	Active function	1	0	1
Total			22	8	7
Pectoralis major transfer to biceps					
Lloyd-Roberts and Lettin[2]	8	Permanent, temporary, or no improvement	6	0	2
Carroll and Kleinman[5]	2	>60° arc with active flexion to 120°	1	0	1
Doyle et al.[10]	7	Improved ROM and single-hand feeding	6	1	0
Atkins et al.[11]	6	Grade 4 strength, ROM, subjective improvement	2	3	1
Bennett et al.[9]	3	Improved function	2	0	1
Total			17	4	5
Steindler flexorplasty					
Lloyd-Roberts and Lettin[2]	1	Permanent, temporary, or no improvement	1	0	0
Doyle et al.[10]	2	Improved ROM and single-hand feeding	2	0	0
Bennett et al.[9]	1	Self-feeding	1	0	0
Total			4	0	0

ROM, range of motion; PROM, passive range of motion.

in elbows with arthrogryposis. Because these children most commonly have severe wrist palmar flexion/pronation deformities that could be exacerbated by proximal relocation of the flexor pronator origin as in a Steindler flexorplasty, we have not used this procedure. The sternocleidomastoid also has not been used due to its cosmetic disfigurement of the neck.

Based on the comparative results of this report, as shown in Table 5, the triceps transfer has shown to offer several distinct advantages. First, the procedure is the least extensive dissection of the options available. The triceps requires only a transfer of the insertion, rather than a bipolar transfer with more extensive dissection necessary for the pectoralis major or latissimus dorsi. Second, most children with arthrogryposis have a strong triceps and its strength can be reliably tested preoperatively. Finally, in our experience, the results show the best maintenance of preoperative strength of the muscle; thus, it has been most predictable of the transfers in our experience.

The 7 patients not requiring elbow capsulotomy all had good results in strength, ROM, and functional outcome. The 2 triceps transfers with poor results had concomitant elbow capsulotomy or previous elbow capsulotomy with triceps lengthening. Whether the results were poor due to their young age, to the short length of time between release and transfer, to a poor triceps muscle associated with poor passive ROM, or to the previous lengthening of the triceps cannot be concluded based on 2 cases. We recommend, however, elbow capsular release with triceps lengthening be staged with subsequent triceps transfer, noting that the results of transferring a previously lengthened triceps are unknown.

The disadvantages of triceps transfer are loss of extension power and elbow flexion contracture. Some claim that loss of extension power is unacceptable for patients with lower extremity involvement who require active elbow extension for transferring or ambulation aids. However, in the present series, its donor morbidity was minimal, as gravity contin-

Table 5. Comparative Results for Tendon Transfers

	Triceps	*Pectoralis*	*Latissimus Dorsi*
No. of transfers	9	5	4
Average age at surgery (yr)	6	4	11
Length of follow-up (yr)	5	5.8	1.5
Average postoperative PROM arc	76°	75°	93°
Average loss of extension	37°	16°	0°
Average postoperative active flexion	87°	64°	90°
No. with >90° active flexion	8/9	1/5	2/4
Average strength	3.8	3.0	3.5
Strength/ROM result			
Good	7	1	2
Fair	1	3	2
Poor	1	1	—
Functional use result			
Good	6	—	—
Fair	1	4	4
Poor	2	1	—

PROM, passive range of motion; ROM, range of motion.

ues to extend the elbow; however, each patient must be evaluated individually. Two of our patients had significant lower extremity involvement and were not compromised by triceps transfer. For example, 1 patient was confined to a wheelchair at a young age and was dependent on help for transfers due to severe lower extremity involvement. Transfer of the triceps was performed; loss of extension strength did not change the patient's inability to independently transfer from the wheelchair, but did improve her ability to independently perform hand-to-mouth functions. In this series, however, loss of passive extension after triceps transfer was noted to be 37°, significantly more than either the pectoralis major or the latissimus dorsi transfer. Eight of 9 patients undergoing triceps to biceps transfer developed flexion contractures. We advocate including the high incidence of elbow flexion contracture as part of the preoperative discussion. Thus, previous elbow capsulotomy with triceps lengthening and possible unacceptable loss of elbow passive extension and/or strength are reasons for consideration for other tendon donors.

The pectoralis transfer can be used as a unipolar,[15] partial bipolar,[8] or complete bipolar[7] transfer. In the elbow with arthrogryposis, we have used the complete bipolar transfer with the entire available muscle mass to maximize potential for strength. The advantage of this type of transfer is that additional muscle mass is added to the hypoplastic limb. There is a theoretical loss of muscle strength in the chest wall and shoulder internal rotation; however, the shoulder with arthrogryposis is most frequently internally rotated without active external rotators so the functional loss is minimal in our experience. The disadvantage is the extensive dissection necessary, and the distal attachment can be technically difficult due to the muscular bulk of the pectoralis. Although other investigators have discouraged its use in females, 1 of the 4 patients in this series is female and was treated with a modified procedure using 2 incisions (a horizontal clavicular incision and a smaller vertical incision parallel to the sternum) with careful undermining. At 7-year follow-up at the age of 13 years, she had normal breast development with minimal chest wall asymmetry. As reported in this series, the greatest problem encountered with this transfer was its lack of predictability of strength.

The third available option for transfer is the latissimus dorsi. Similar to the pectoralis transfer, muscle mass is added from the chest wall to the hypoplastic limb without significant loss of function in our experience. Its disadvantage is also similar in that the surgical dissection necessary is extensive and the resultant strength is somewhat less predictable. Although the preoperative muscle mass can be palpated and its strength estimated, accurate preoperative assessment of muscle strength can be difficult. As with the pectoralis major muscle, the muscle may be atrophied and infiltrated with fat, which can be a source of suboptimal strength. Additionally, due to its shape as a long muscle, its tension can be difficult to set.

The second purpose of this study is to compare tendon transfer results based on the traditional assessment of ROM and strength versus a functional assessment of use in ADL. Using traditional ROM and strength criteria as a measure of surgical outcome, 10 good, 6 fair, and 2 poor outcomes were obtained. Using a questionnaire regarding evaluation of functional use of the upper extremity, along with active demonstration and videotaping of 6 ADLs, 6 good, 9 fair, and 3 poor outcomes were obtained. The results that were downgraded by the functional assessment analysis were due to transfer in the nondominant extremity and to loss of hand-to-hand function due to 95° flexion deformity of 1 elbow in a child with an extension deformity of the other elbow. The functional use analysis showed more use of the transfer if done in the dominant extremity and also showed the affect of multijoint involvement. Improvement of upper extremity function cannot be assumed to be due solely to improved elbow function. As shown in Table 1, several patients had surgical procedures of the shoulder, wrist, and hand, which may in part account for the improved upper extremity function. The functional data are also limited by their retrospective nature, with no comparative preoperative functional data collected. Yet, patients and parent substantiated functional improvement.

Finally, review of upper extremity dysfunction in 61 children with arthrogryposis has led to observations that have allowed formulation of our treatment recommendations for elbow dysfunction. These recommendations are primarily based on clinical observation, which have proven useful in our clinical practice. The first goal of caring for the upper extremities of a child with arthrogryposis is to provide passive mobility of at least 1 elbow to allow hand-to-face activities. To achieve passive elbow mobility, physical therapy is the initial type of care. Repetitive, gentle, passive manipulation of the joint by the therapist and parents may progressively lessen the contracture. Casts and splints also are used with varying clinical success. The goal of therapy is to achieve at least 90° of passive flexion. In this series, 43 patients had elbow involvement and passive ROM was obtained without surgical intervention in 25 patients (58%). The indication for surgical release of the elbow is less than 90° of passive elbow flexion after at least 6 months of supervised elbow stretching. In this study, passive elbow flexion was obtained and maintained using an elbow capsulotomy and lengthening of the triceps in the subset of patients who failed physical therapy.

The second goal of upper extremity treatment is to provide active elbow flexion so that the hand can effectively be positioned in space. Evaluation for a tendon transfer requires integration of multiple factors, including patient age, intelligence, extremity dominance, ipsilateral shoulder/wrist/hand function, contralateral upper limb function, need for ambulatory aids, and available motors for transfer. Tendon transfer is indicated in children with arthrogryposis lacking active elbow flexion who have at least 90° of passive elbow motion and an available donor. Choice of transfer donor includes consideration of donor strength, morbidity associated with donor function loss, and predictability of tendon transfer results as discussed above.

Based on this review of surgical indications and outcomes assessment, we make the following treatment recommendations. Exercises to obtain and maintain passive ROM of the elbow are initiated at birth. If at least 90° of passive elbow flexion has not been achieved by 18 to 24 months of age after at least 6 months of supervised elbow stretching, an elbow capsulotomy with triceps lengthening is recommended. After the age of 4 years, tendon transfers for elbow flexion on the dominant arm are recommended, with triceps to biceps giving the most predictable results. The optimal surgical candidate for tendon transfer is a child older than 4 years who has full passive ROM of the elbow, arthrogryposis in the dominant arm, and at least grade 4 strength of the muscle to be transferred.

The authors acknowledge Drs William Cooney, Michael Millis, and John Hall for their case contributions.

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