The Steindler Flexorplasty for the Arthrogrypotic Elbow

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Purpose: The arthrogrypotic elbow often lacks active flexion. If active elbow flexion can be provided by muscle transfer, patient independence increases and the patient can function in a less conspicuous manner by avoiding adaptive mechanisms. The purpose of this article is to review the outcome of patients with arthrogryposis treated with the Steindler flexorplasty to obtain active elbow flexion.

Methods: Seventeen elbows in 10 patients with an average age of 7 years were treated surgically with the Steindler flexorplasty procedure. Before surgery none of the patients was able to flex actively the elbow against gravity. All of the patients had at least 70° of passive elbow flexion. Upper-extremity active and passive range of motion, strength of flexion, functional outcome, and patient satisfaction were assessed at an average of 5 years after surgery (range, 2–9 years).

Results: After surgery all patients obtained active elbow flexion against gravity averaging 85° (range, 30°–120°); patients were able to lift an average of 1 kg through their entire arc of elbow flexion. At last follow-up evaluation patients lost an average of 27° of elbow extension. Patients lost forearm rotation but did not lose wrist or finger range of motion. Subjectively, 9 of the 10 patients were satisfied with the outcome of the surgery and would recommend the surgery to others.

Conclusions: The Steindler flexorplasty provides improved elbow flexion strength and patient function and should be considered for children with arthrogryposis. (J Hand Surg 2004;29A: 462–469. Copyright © 2004 by the American Society for Surgery of the Hand.)

Key words: Steindler, flexorplasty, arthrogryposis, amyoplasia, elbow.

Arthrogryposis multiplex congenita is a nonspecific diagnosis used to describe a mixed group of congenital disorders characterized by limited joint motion. Amyoplasia, as delineated by Hall in 1983,[1] is the most common manifestation of arthrogryposis and as indicated by its name is associated with abnormal muscle development and its replacement by fibrous and fatty tissue. Muscle imbalance contributes to the characteristic joint contractures of arthrogryposis. Limb girth is decreased because of the smaller muscle mass. It is rarely an inherited condition.[1]

The classic patient with arthrogryposis with upper-extremity involvement has an abnormality at each joint. The shoulders are internally rotated and adducted, the elbows are extended, the forearms are pronated, the wrists are flexed, and the thumbs are adducted and flexed; however, the number of joints involved and the position of contracture varies. The elbow is involved frequently and shows various presentations. The elbow may be contracted in an extended position, may be held in extension but maintain full passive flexion, or may be contracted in a position of flexion. A large number of patients have weak or completely absent active elbow flexion.[2]

If the patient lacks passive elbow flexion the first

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interventional goal is to provide functional passive elbow motion. This may be accomplished with passive stretching, casting, or surgical release. Patients with full passive motion learn adaptive mechanisms that allow the completion of activities of daily living (ADLs). For example, when eating patients bring the hand to the mouth by stabilizing the forearm against the table, leaning forward, and passively flexing the elbow. They also use shoulder musculature to flex the arm forward and allow gravity to passively swing the elbow into flexion. Activities that are typically performed with 1 hand often are performed with 2 hands by the arthrogrypotic patient. Although these adaptive mechanisms together with assistive devices make the performance of critical activities easier the lack of active elbow flexion remains disabling and socially awkward.

Active elbow flexion is the ultimate goal of intervention. Multiple procedures have been used to provide active elbow flexion in the arthrogrypotic patient including pectoralis major transfer,[3-6] latissimus dorsi transfer,[4,7] and triceps transfer.[2,8] The Steindler flexorplasty,[9] described initially to treat paralytic conditions, has been used subsequently to treat several other conditions[10-12] but has been reported in only 4 cases of arthrogryposis.[5,6,13] This procedure transfers the proximal osseous origin of the wrist and finger flexors from the medial epicondyle to a position more proximal and slightly lateral[12] on the distal humerus. The purpose of this investigation is to review the results of arthrogrypotic patients treated with the Steindler flexorplasty to provide active elbow flexion.

Materials and Methods

Patients with arthrogryposis are treated through a multidisciplinary approach at our orthopedic children's hospital. All patients are evaluated by both hand surgeons and hand physiotherapists to identify functional deficits that may be improved with therapy or surgical intervention.

The Steindler flexorplasty was performed at our hospital to treat a lack of active elbow flexion in patients with arthrogryposis. For this study we reviewed the medical records of all arthrogrypotic patients who had a Steindler flexorplasty, a minimum 2-year follow-up period, and adequate clinical records; 14 patients were identified. Four of the 14 patients did not return for final follow-up evaluation, leaving 10 patients (17 elbows) for this investigation. Seven of the patients were treated with staged, bilateral flexorplasty procedures and 3 patients were treated with unilateral surgery only. There were 6 girls and 4 boys. The average age at the time of surgery was 7 years (range, 4–12 years). The average follow-up period was 5 years (range, 2–9 years). Four of the patients had additional upper-extremity surgery including rotational osteoclasis for fixed forearm pronation, wrist fusion, and tendon transfers for wrist extension.

There were 4 entrance criteria for the surgical procedure. First, on plain radiographs there was a normal elbow articulation. Second, all patients had a minimum of 70° of passive elbow flexion (range, 70°–135° of flexion; average, 103°). Six elbows initially had less than 70° of passive elbow flexion. One improved sufficiently with serial casting; 5 required posterior elbow release and triceps lengthening at an average age of 6 years to provide adequate passive elbow flexion before consideration of flexorplasty. Third, patients had to be sufficiently cooperative to participate in postsurgical therapy, an important component for a successful outcome. Fourth, all patients had to have active finger flexion with good strength against resistance. The status of the wrist and shoulder was evaluated but no patient had treatment for these conditions near the time of flexorplasty. None of the patients were dependent on crutches for ambulation.

Surgical Technique

The surgical technique is patterned after that described by Mayer and Green.[12] A 10- to 13-cm curvilinear incision is centered over the medial epicondyle extending anterior in a proximal and distal direction (Fig. 1) The intramuscular septum proximal to the elbow and the forearm fascia are exposed; there is frequently a substantial amount of subcutaneous fat covering these structures. The ulnar nerve is identified and mobilized proximal and distal to the medial epicondyle and its entrance into the substance of the flexor carpi ulnaris muscle. The median nerve and brachial artery are also identified, mobilized, and protected. The flexor pronator mass origin is isolated from the adjacent soft tissue (Fig. 2). The medial collateral ligament complex must be preserved. A small wafer of the bony/cartilaginous medial epicondyle is separated in continuity with the muscle mass using a no. 15 blade or small osteotome and mobilized distally into the forearm (Fig. 3). The elbow is then flexed to allow fixation of the flexor/pronator origin to the anterior humerus. The amount of elbow flexion and the distance the flexor/pronator mass is mobilized proximally varies depending on the quality

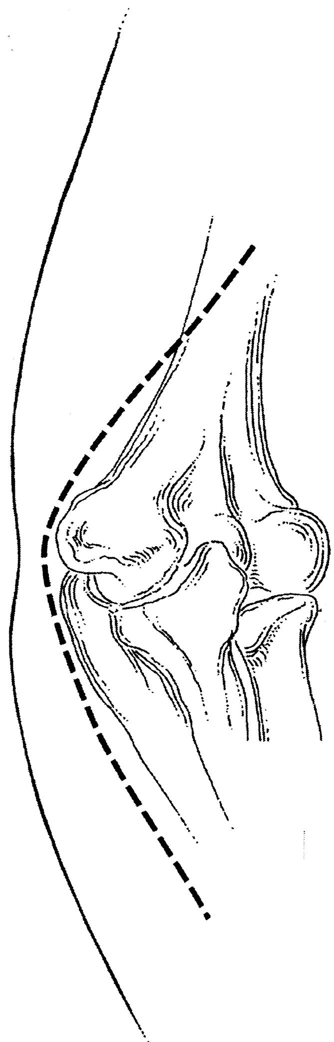

Figure 1. Incision for Steindler procedure.

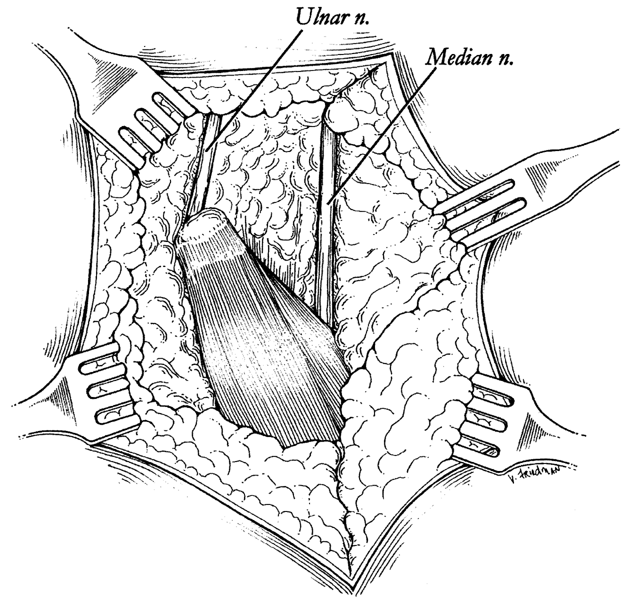

Figure 2. Exposed flexor/ pronator mass. Note that the median and ulnar nerves have been isolated.

of the muscle present and the mobility of the elbow; however, the fixation point on the anterior humerus is approximately 2 to 3 cm proximal to the medial epicondyle with the elbow flexed about 80° (Fig. 4). The brachialis is frequently hypoplastic or fibrotic; the lateral portion of the muscle is incised longitudinally and elevated at the planned insertion site on the anterior humerus. The humerus is prepared with a burr to provide a cancellous bone site for healing of the flexor pronator muscle transfer. Although several techniques have been used to secure the transfer we currently use a 3.5-mm cortical screw[14] with a soft tissue washer to secure the transfer to the humerus. Eight elbows were treated with screw fixation; 4 with heavy, nonabsorbable sutures; 3 with metal sutures; and 2 with K-wire fixation. The tourniquet is deflated before closure to attain hemostasis. Surgery time averaged 100 minutes (range, 51–126 minutes).

The elbow is splinted in a position of approximately 70° of elbow flexion and 30° of forearm supination. Four weeks after surgery the elbow is placed in a posterior splint that blocks elbow extension and allows passive flexion exercises. At 6 weeks patients are readmitted to the hospital for initiation of active range of motion exercises with flexion against

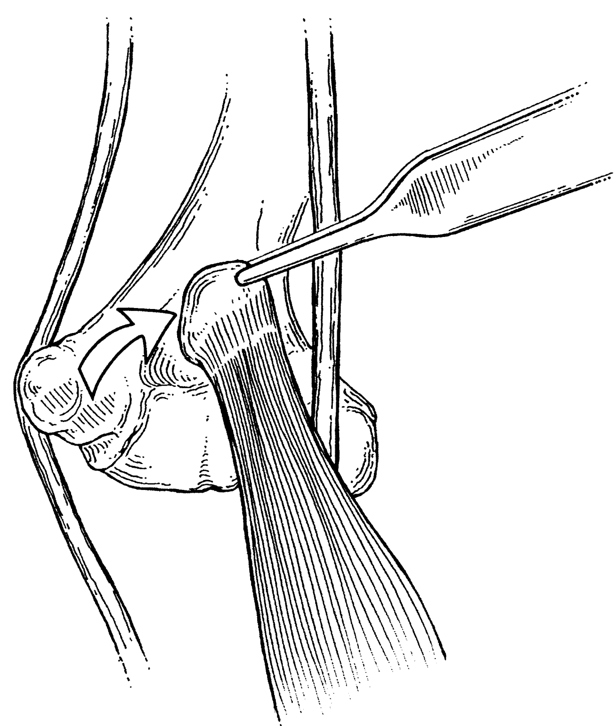

Figure 3. Transferred flexor/pronator mass.

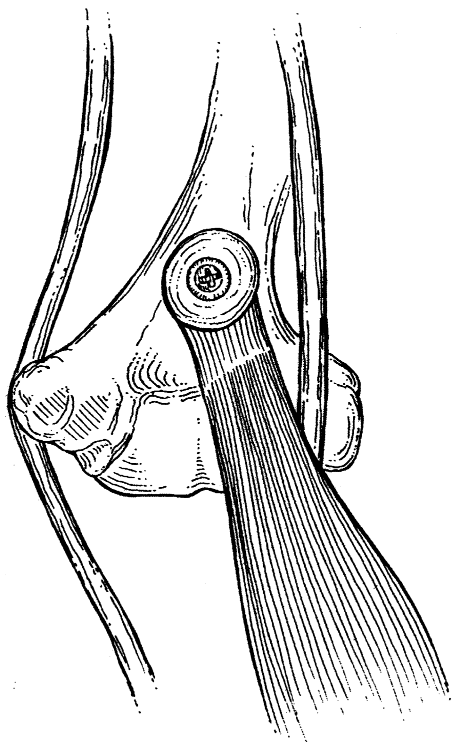

Figure 4. Flexor/pronator mass after fixation.

gravity. Resistive strengthening exercises are begun 12 weeks after surgery.

The presurgical range of motion measurements were obtained from the medical record as documented by the hand therapists. Patients returned for follow-up physical examination. A pediatric hand therapist who was not involved in the original care of the patients performed all postsurgical measurements. Passive elbow flexion and extension and active elbow flexion against gravity were measured using a goniometer with the shoulder adducted in a standardized fashion. The strength of flexion against gravity was determined by documenting the greatest amount of weight the standing patient could lift while completing a full arc of elbow flexion.

Active and passive forearm pronation and supination were measured with the shoulder adducted and the elbow flexed 90°. Active and passive wrist flexion and extension were measured; finger range of motion was also assessed.

Subjective outcome was based on questioning each patient and/or family about the level of satisfaction with the procedure. Patients were asked which ADLs were made easier by the transfer and which ADLs continued to require adaptive mechanisms. We queried the patients with bilateral involvement who did not have bilateral procedures as to what factors led them to decide not to have surgery on the opposite side. Finally, patients were asked if they would recommend the procedure to another patient with arthrogryposis.

Statistical Analysis

The 2-tailed, paired Student's *t*-test was used to evaluate the differences between presurgical and postsurgical data. Significance was considered to be $p < .05$.

Results

Before surgery no patient was able to actively flex the elbow against gravity. After surgery all patients had active elbow flexion against gravity averaging 85° (range, 30°–120°). Flexion strength through the patients' entire arc of flexion averaged 1 kg. All patients had a flexion strength of at least ½ kg with the exception of 1 patient who could lift only approximately ¼ kg through his entire arc of flexion; maximum flexion strength was 4 kg (Table 1).

The presurgical passive elbow extension averaged 1° and the presurgical passive elbow flexion averaged 103°. After surgery passive elbow extension averaged 28° and passive elbow flexion averaged 105°. The average passive arc of motion decreased significantly from before surgery (102°) to after surgery (77°) ($p < .001$). The loss of passive elbow extension was also statistically significant ($p < .001$) (Table 1).

Active pronation decreased significantly from an average of 62° before surgery to 30° after surgery ($p < .001$). Active supination also decreased from 26° before surgery to 11° after surgery ($p = .19$). The total active arc of forearm rotation decreased significantly from 88° to 41° after surgery ($p = .002$). The postsurgical passive arc of forearm rotation was 95° (Table 1).

Wrist extension decreased minimally from an average of 3° before surgery to –3° (ie, 3° of flexion) after surgery ($p = .41$). Active wrist flexion remained unchanged at 59°. Wrist arc of motion was not changed significantly from 62° before surgery to 56° after surgery ($p > .05$). The change in wrist motion was not statistically significant ($p > .05$). Postsurgical passive range of wrist motion was 78° (Table 1). No patient had a significant change in finger motion after surgery.

Subjectively, 9 of the 10 patients were satisfied with the outcome of the surgery and would recommend the surgery to others (Table 2). The specific reasons cited for satisfaction with the procedure varied; however, all 9 families believed that the procedure gave the patient more independence and im-

Table 1. Presurgical and Postsurgical Assessment

		Active Elbow Motion Before Surgery	Active Elbow Motion After Surgery			Passive Elbow Motion Before Surgery			Passive Elbow Motion After Surgery			Active Forearm Rotation Before Surgery			Active Forearm Rotation After Surgery			Active Wrist Motion Before Surgery			Active Wrist Motion After Surgery			Maximum Strength After Surgery (kg)
Patient	Side	Flex	Ext	Flex	Arc	Ext	Flex	Arc	Ext	Flex	Arc	Pro	Sup	Arc	Pro	Sup	Arc	Flex	Ext	Arc	Flex	Ext	Arc	
1	R	0	15	85	70	0	135	135	15	105	90	45	–15	30	45	–20	25	55	45	100	50	25	75	4
	L	0	0	105	105	0	120	120	0	130	130	75	–50	25	75	25	100	55	45	100	45	50	95	2
2	R	0	70	75	5	15	115	100	65	120	55	85	75	160	20	–10	10	30	0	30	40	–25	15	1/2
	L	0	65	75	10	0	70	70	60	120	60	85	60	145	35	0	35	0	0	0	45	–35	10	1/2
3	R	0	50	85	35	0	110	110	50	105	55	35	70	105	–10	75	65	80	–20	60	75	–60	15	1
	L	0	35	85	50 0	0	110	110	35	115	80	60	70	130	–30	75	45	80	–20	60	0	0	0	1/2
4	R	0	20	100	80	0	100	100	20	90	70	65	15	80	25	15	40	80	–25	55	40	–10	30	
	L	0	45	120	75	0	100	100	45	120	75	55	50	105	30	15	45	80	–35	45	60	–10	50	
5	R	0	20	60	40	0	90	90	20	80	60	80	–10	70	70	–60	10	65	30	95	65	15	80	1/2
	L	0	5	30	25	0	75	75	5	40	35	80	–80	0	0	0	0	50	15	65	35	30	65	1/2
6	R	0	15	85	70	0	110	110	15	105	90	90	55	145	35	–15	20	45	–20	25	80	55	135	1
	L	0	40	105	65	0	120	120	35	120	85	70	5	75	35	20	55	35	–15	20	80	–35	45	1
7	R	0	0	80	80	0	85	85	0	95	95	60	55	115	35	20	55	75	35	110	90	20	110	1/2
	L	0	5	90	85	0	105	105	5	110	105	50	60	110	35	35	70	75	35	110	95	20	115	1/2
8	L	0	85	120	35	0	100	100	75	130	55	45	40	85	40	15	55	75	30	105	45	20	65	1/2
9	L	0	35	85	50	0	105	105	35	100	65	50	20	70	45	15	60	70	–25	45	75	–55	20	1/4
10	L	0	15	60	35	0	100	100	0	105	105	30	20	50	25	–15	10	60	–25	35	75	–55	20	
Average		0	31	85	51	1	103	102	28	105	77	62	26	88	30	11	41	59	3	62	59	–3	56	1
p value*				.000					.000	.634	.000				.000	.193	.002				.910	.414	.462	

NOTE. All values are given in degrees except as noted.
*p values comparing presurgical and postsurgical data are reported below the postsurgical column of comparison.
Abbreviations: ext, extension; flex, flexion; pro, pronation; sup, supination.

Table 2. Postsurgical ADLs

Patient	Subjective Improvement	Adaptive mechanisms	Recommend	Eating	Hair/Teeth	Drinking	Writing	Toilet	Carrying Books	Dressing
1	Yes	Improved	Yes	Adaptive	Adaptive	1-handed	Normal	Normal	1-handed	Help w/makeup
2	Yes	Improved	Yes	Adaptive	Adaptive	Adaptive	Normal	Normal	Bimanual	Help w/fasteners
3	Yes	Improved	Yes	1-handed	1-handed	Adaptive	Normal	Normal	1-handed	Help w/fasteners
4	Yes	Improved	Yes	1-handed	Rare adaptive	1-handed	Normal	Normal	Bimanual	Normal
5	Yes	Improved	Yes	Adaptive	Adaptive	Adaptive	Normal	Normal	1-handed	Normal
6	Yes	Improved	Yes	1-handed	1-handed	Adaptive	Normal	Normal	1-handed	Help w/makeup
7	Yes	Improved	Yes	1-handed	1-handed	1-handed	Normal	Normal	1-handed	Normal
8	No	Improved	No	Adaptive	Opp hand	Adaptive	Opp hand	Opp hand	1-handed	Normal
9	Yes	Improved	Yes	1-handed	1-handed	1-handed	Normal	Normal	1-handed	Normal
10	Yes	Improved	Yes	1-handed	Adaptive	Adaptive	Opp hand	Normal	Bimanual	Help w/fasteners

Abbreviation: opp, opposite.

proved the performance of ADLs. All patients used fewer adaptive mechanisms to accomplish ADLs, making each child feel more “normal.” In many instances 2-handed activities became 1-handed activities. Before surgery all patients used either adaptive mechanisms or assistance for eating; after surgery all patients ate independently and were improved although 4 of the patients continued to use some adaptive maneuvers. Before surgery patients dressed only with assistance or using adaptive maneuvers; after surgery all patients dressed independently although 5 of the 10 patients needed assistance with buttons or fasteners. Nine of the 10 patients became independent with toileting and 6 became independent without adaptive maneuvers for brushing hair and teeth. The decreased elbow extension and the decreased forearm rotation after surgery did not affect adversely the performance of ADLs in any patient.

Seven of the patients were sufficiently satisfied with the procedure on 1 elbow to have the procedure on the opposite extremity. Three patients did not have the procedure on the opposite elbow: 1 patient had poor passive range of elbow motion and did not wish to have further treatment on that side, 1 patient had less severe muscle involvement with some maintained active elbow flexion, and 1 patient had a previous triceps-to-biceps brachii transfer on the opposite side.

One family was dissatisfied with the surgical outcome and would not recommend the surgery to others. The mother did not believe that the surgery provided any benefit to the patient and thought that he used the opposite extremity more after surgery. The strength in the operated arm remained weak, providing only ¼ kg flexion strength. The mother did not believe that any activities were made easier and she did not think that patient independence was increased.

Complications were minimal. In 1 elbow the screw attaching the flexor pronator mass to the anterior humerus dislodged; revision surgery with a longer immobilization period corrected the problem. Ulnar nerve paresthesias developed in 1 elbow immediately after surgery and the symptoms resolved by the first postsurgical day. When surgery was performed on the contralateral elbow more substantial paresthesias developed that did not resolve spontaneously; ulnar nerve transposition led to a resolution of the symptoms.

Discussion

Historically, the treatment of the arthrogrypotic elbow has been guided by the “1 up and 1 down” concept of positioning. This theory suggests that 1 elbow should be in a position of flexion for eating and the other elbow should be in a position of extension for hygiene purposes.[5,13] This concept has lost popularity as we have come to understand that bimanual activity is important for the arthrogrypotic patient.[15] The first goal of treatment must be to provide a passive arc of elbow motion. If therapy with splinting and casting do not significantly improve motion a posterior elbow release with triceps lengthening is very effective in increasing passive range of motion.[2,4,6] This report evaluates the Steindler flexorplasty as a means to provide active elbow flexion but there are 3 other muscle transfers also used commonly: the triceps-to-biceps transfer, the latissimus dorsi transfer, and the pectoralis major transfer. Although this report is not meant to be comparative a brief discussion of the other procedures for active motion is helpful.

The triceps-to-biceps transfer is advocated because it is technically straightforward to perform and improves reliably the strength of elbow flexion. Most arthrogrypotic patients have a satisfactory triceps muscle that can be transferred with minimal morbidity because gravity can assist with elbow extension. Reported strength was at least 4 out of 5 in 7 of 9 patients in a report by Van Heest et al[4] and strength averaged 4 out of 5 in 17 patients reported by Williams[2]; however, severe postsurgical flexion contracture secondary to the loss of elbow extension can be problematic. Van Heest et al[4] noted an average of 38° flexion contracture after surgery in 9 patients (who all had full elbow extension before surgery) and Williams[2] found an average of 67° flexion contracture after surgery. Doyle et al[5] noted that only 4 of 7 patients had improved motion and the ability to eat with 1 hand after this transfer. Although strength is certainly improved the postsurgical flexion contracture may affect function and the lack of active elbow extension power prohibits crutch walking.

The pectoralis major may also be transferred to provide elbow flexion. It may be used as a unipolar transfer[16] with the clavicular head insertion detached from the humerus and reinserted into the distal biceps (with or without tendon elongation). Alternatively it may be used as a bipolar transfer, rotating on its neurovascular pedicle, thus providing a more effective line of muscle pull. In 4 children Van Heest et al[4] reported a 21° elbow flexion contracture after surgery with an average active flexion against gravity of 41°. Atkins et al[3] reported on 6 patients with a 32° flexion contracture and a muscle strength of 3 to 4. Subjective outcome was very good in all. Doyle et al[5] reported on 7 patients and noted improved motion in all patients and single-handed feeding in 6 of the patients. Other reports are similar[6,13]; however, the pectoralis major transfer is not always aesthetically pleasing. The unipolar transfer adducts the arm substantially and creates an abnormally large anterior axillary fold. The bipolar transfer is a more complex procedure that deprives the arm of an adductor and can lead to significant scarring.[17]

The latissimus dorsi may be transferred in a bipolar fashion similar to the pectoralis transfer. It can be a very effective transfer if the muscle quality is good although the muscle may be hypoplastic in some cases of arthrogryposis.[15] Van Heest et al[4] reported on 3 children and noted no flexion contracture after surgery and an average postsurgical arc of active flexion of 90° with 84° of active flexion against gravity.

Although the Steindler flexorplasty has been reported commonly in the literature for polio, obstetrical palsy, and trauma[10,12,18] it has been reported rarely for arthrogryposis. It is important to note, however, that the results are good in the 4 reported cases. Lloyd-Roberts and Lettin[6] reported on 1 patient who had a satisfactory outcome. These researchers believed the surgery was most effective in children with adequate power of the wrist and hand and with some elbow flexion power. Bennett et al[13] reported on 1 patient with a good functional outcome, and Doyle et al[5] reported on 2 patients treated with the Steindler flexorplasty who both had improved motion and gained the ability to eat with a single hand.

Criticisms of the Steindler flexorplasty have focused on a few potential concerns. First, previous investigators have stated that transfer of the abnormal arthrogrypotic flexor/pronator mass provides insufficient strength.[6,17] Second, the nature of the transfer may limit elbow extension and supination.[10] Finally, some have noted increased wrist and finger flexion after the transfer, making activities that require wrist and finger extension more difficult.[4,15,17,19]

These criticisms, however, did not prove to be valid in this group of arthrogrypotic patients and the Steindler flexorplasty has met our goal of providing active elbow flexion with an ability to lift objects against gravity. The postsurgical active arc of motion averaged 58° compared with no active flexion before surgery. Postsurgical flexion contracture averaged 28°. These values are comparable to or are better than the arc of motion and flexion contracture reported with the other 3 procedures. Although this contracture may be minimized by tensioning the transfer at less than 80° of elbow flexion we are hesitant to do so because of the potential limitation of active flexion after surgery. Furthermore, this degree of flexion contracture did not limit patient function.

Patients became less dependent on adaptive mechanisms and gained independence in the performance of various tasks. Simple tasks such as brushing teeth, brushing hair, eating, and dressing become more straightforward and often done with 1 hand. The elbow flexion strength improved patients' performances of ADLs without significant postsurgical positional or aesthetic concerns.

Strength assessment in these children is difficult. Previous reports have documented postsurgical muscle strength using the British Medical Council grading strength of 1 to 5. We assessed strength objectively by evaluating how much weight a patient could hold while completing a full arc of elbow flexion.

Patients were able to flex the elbow against gravity with an average additional weight of 1 kg. This approximates grade 4 strength by British Medical Council standards. This is also comparable to most other reports.

In contrast to the predicted loss of wrist and finger flexion after proximal transfer of the flexor pronator origin,[4,15,19] patients in this series did not show an increase in wrist flexion posture or finger flexion posture after surgery. No patient was limited functionally by their positioning and all showed improved performance with activities.

Although this study is retrospective it represents a large series of Steindler flexorplasty procedures in arthrogrypotic patients. A prospective and postsurgical comparison of a functional outcome assessment tool would have been beneficial; nevertheless, the results of this study are comparable to other reported procedures to obtain active elbow flexion in patients with arthrogryposis. Furthermore, the Steindler flexorplasty is advantageous because the surgical dissection is not as extensive as is required for transfers of the pectoralis major, the latissimus dorsi, or the triceps.

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