Surgical Reconstruction for Upper-Extremity Paralysis Following Acute Flaccid Myelitis

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Background: Acute flaccid myelitis (AFM) is a debilitating illness that is defined by the sudden onset of flaccid paralysis in the extremities with spinal magnetic resonance imaging (MRI) demonstrating a longitudinal lesion confined to the gray matter. The purpose of this study was to report the types of upper-extremity palsy and outcomes of surgical reconstruction in patients with AFM.

Methods: Eight patients with a median age at onset of 3.8 years (range, 2.3 to 9.9 years) were identified. There was loss of shoulder abduction and external rotation in all patients, loss of elbow flexion in 5 patients, complete or partial loss of hand function in 3 patients, and spinal accessory nerve palsy in 2 patients. All patients underwent surgical reconstruction, which was categorized into 3 main groups: nerve transfer, secondary muscle transfer, and free muscle transfer.

Results: The median follow-up period was 39 months (range, 30 to 94 months). Four patients obtained ≥90° of shoulder abduction whereas the other 4 patients had shoulder abduction of ≤70°. The 5 patients who received free muscle transfer or nerve transfer to restore elbow function obtained ≥140° of elbow flexion. Two patients treated with free muscle transfer to restore finger function obtained satisfactory total active motion of the fingers (180°).

Conclusions: The patterns of paralysis and the strategy and outcomes of surgical reconstruction for patients with AFM differed from those for traumatic and obstetric brachial plexus palsy. All patients had loss of shoulder abduction, and 2 had spinal accessory nerve palsy. Restoration of shoulder function was unpredictable and depended on the quality of the donor nerves and recovery of synergistic muscles. Restoration of elbow and hand function was more consistent and satisfactory.

Level of Evidence: Therapeutic Level IV. See Instructions for Authors for a complete list of levels of evidence.

Acute flaccid myelitis (AFM) is a poliomyelitis-like illness that is characterized by an acute onset of flaccid paralysis in the extremities with spinal magnetic resonance imaging (MRI) demonstrating a longitudinal lesion confined to the gray matter. AFM was first reported in the United States in 2012, and clusters of cases were observed every 2 to 3 years since then in many countries[1-7]. It is a disease with a seasonal pattern, with most patients having the onset of AFM between August and November.

There was a high epidemic association between AFM and enterovirus D68 (EV-D68), but the definite causative agent is yet to be determined. Most of the patients experienced a few days of prodromal illness with respiratory symptoms before the onset of neurological deficits, which progressed rapidly from muscle weakness to complete paralysis of the affected limb without sensory involvement.

A nationwide survey in Japan revealed the paralysis in the acute stage to involve 1 limb in 37% of patients, 2 limbs in 39%, 3 limbs in 5%, and 4 limbs in 19%[7]. Involvement of family members of patients with AFM has not been reported to our knowledge.

While many authors reported that patients with AFM regained some strength as time passed[8-12], the majority of patients did not recover fully. About 90% of patients still have motor deficits during follow-up regardless of the mode of treatment of the disease[6,7]. Therefore, reconstructive surgery is needed to improve the function of the affected limb. The basic principle of surgical reconstruction for patients with AFM is based on the

TABLE I Case Definitions of AFM by the U.S. CDC*	
Case Designation	Criteria
Confirmed	(1) Acute onset of flaccid limb weakness AND (2) MRI showing a spinal cord lesion largely restricted to gray matter and spanning ≥1 spinal segment
Probable	(1) Acute onset of flaccid limb weakness AND (2) cerebrospinal fluid with pleocytosis (white blood-cell count >5 cells/mm^3, adjusted for presence of red blood cells by subtracting 1 white blood cell for every 500 red blood cells present)

*Source: Council of State and Territorial Epidemiologists (https://www.cste.org/resource/resmgr/2017ps/2017psfinal/17-ID-01.pdf and https://www.cdc.gov/acute-flaccid-myelitis/hcp/case-definitions.html).

treatment for traumatic or obstetric brachial plexus palsy. Nerve transfer is preferable for patients who present early after the onset of paralysis whereas free or pedicled muscle transfer is better for patients with late presentation[8-13].

The objective of this study was to report the types of upper-limb palsy and the outcomes of surgical reconstruction in patients with AFM—hence to update orthopaedic and microsurgeons about AFM and the current surgical treatment strategy for this disease.

Materials and Methods

This was a retrospective study of patients with upper-limb paralysis due to AFM who were referred to our hospital for surgical reconstruction. This study was approved by the local hospital institutional ethics committee, and written informed consent was obtained from all patients.

Diagnostic Definition of AFM
Standardized definitions of "confirmed" and "probable" cases of AFM provided by the U.S. Centers for Disease Control and Prevention (CDC)[1,2] on the basis of clinical and laboratory criteria are shown in Table I.

Patients Demographic (Table II)
Eight patients with confirmed AFM fulfilled the inclusion criteria. There were 3 girls and 5 boys with a median age at onset of 3.8 years (range, 2.3 to 9.9 years). Three patients had bronchial asthma whereas 5 patients were healthy before the onset of the AFM. Seven patients experienced prodromal illness with fever and respiratory symptoms. All patients had a sudden onset of neurological symptoms (upper-limb flaccid paralysis without sensory impairment) after 1 to 8 days (median, 3 days) of prodromal symptoms. All patients had pleocytosis in the cerebrospinal fluid, and their spinal MRIs demonstrated longitudinal abnormal signals confined to the gray matter and/or anterior horn cells and spanning ≥1 vertebral levels (Figs. 1 and 2). After the onset of paralysis, 7 patients received immunomodulation therapy consisting of high-dose intravenous immunoglobulin, pulse methylprednisolone, or a combination of the 2, but there was no substantial clinical improvement of the neurological deficits.

Seven patients underwent neurophysiological tests within 2 weeks after onset (Table III). The median nerve demonstrated no electrical responses in the 3 patients with hand palsy whereas it showed normal motor conduction velocity in the 4

TABLE II Demographic Data*						➤
Case	Age at Onset *(yr)*	Onset Date *(yr/mo/day)*	Sex	Final Involved Side	Asthma	Flu-Like Symptoms with High Fever
1	4.3	2010/9/16	F	L	Yes	No
2	2.9	2010/9/24	M	R	No	Yes
3	4.3	2015/9/9	M	R	No	Yes
4	9.9	2015/9/11	M	L	No	Yes
5	3.8	2015/9/19	F	R	Yes	Yes
6	3.6	2015/8/28	M	L	No	Yes
7	3.7	2015/9/7	F	L	Yes	Yes
8	2.3	2010/9/11	M	L	No	Yes
Median	3.8					
Range	2.3-9.9					

*NR = not reported by primary pediatric neurologist, and CSF = cerebrospinal fluid. †mPSL = high-dose methylprednisolone, PSL = prednisolone, IVIG = intravenous immunoglobulin, and acyclovir = anti-viral (herpes) drug.

Fig. 1
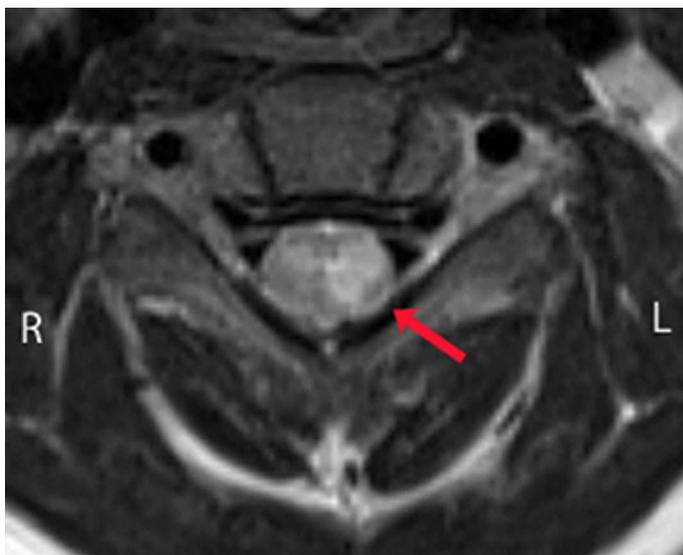
Fig. 2

Fig. 1 Sagittal view of T2-weighted cervical spine MRI demonstrating a diffuse longitudinal abnormal signal of hyperintensity over the central portion of the spinal cord (arrow). **Fig. 2** Representative axial view of T2-weighted MRI at the C4 level demonstrating abnormal hyperintensity affecting the left gray matter (arrow).

patients without involvement of the hand. The median nerve showed decreased F-wave persistence (0% to 56%) in all 7 cases, which suggested that the anterior horn cells were affected from C8 to T1.

Preoperative Evaluations

Preoperatively, all patients except Case 8 (who presented late after the onset of paralysis) were followed at 2 to 3-month intervals until the plateau of recovery was achieved.

TABLE II (*continued*)						
AFP After Symptom Onset (*days*)	Initial Paralysis	EV-D68 Test	T2-Wt. MRI High-Intensity Signal Indicating Lesion	CSF Cell Count (*cells /mm^3*)	AFM Designation	Initial Treatment†
7	Monoplegia	NR	Yes	12	Confirmed	mPSL
1	Monoplegia	NR	Yes	192	Confirmed	No treatment
3	Monoplegia	−	Yes	10	Confirmed	PSL, IVIG
3	Monoplegia	−	Yes	131	Confirmed	mPSL, IVIG, acyclovir
8	Diplegia	−	Yes	54	Confirmed	mPSL
3	Monoplegia	+	Yes	33	Confirmed	mPSL, IVIG
3	Monoplegia	−	Yes	153	Confirmed	mPSL
7	Monoplegia	+	Yes	67	Confirmed	mPSL
3						
1-8						

TABLE III Results of Preoperative Electrophysiological Tests*

| Case | Exam. Time After Onset (days) | Nerve Conduction Velocity of Median Nerve on Involved Side | | | |
		M-Wave (MCV) (m/s)	Amplitude (CMAP) (mV)	F-Wave† (%)	SCV (m/s)
1	7	0	0	0	54.1
2	1	0	0	0	NR
3	3	52.1	7.3	37	46.3
4	3	62.1	10.99	56	NR
5	8	57.7	2.2	0	57.7
6	4	0	0	0	NR
7	3	60.5	1.57	35	60.0
8	NR	NR	NR	NR	NR
Median	3				
Range	1-8				

*MCV = motor conduction velocity, SCV = sensory nerve conduction velocity, and NR = not reported by a primary pediatric neurologist. †F-wave persistence of the median nerve.

TABLE IV Preliminary Classification of AFM* ➤

| | AFM: Neck | | | AFM: Shoulder | | | |
	Sternocleidomastoid	Trapezius	Diaphragm	Deltoid	Supraspinatus/ Infraspinatus	Pect. Maj.: Clavicular	Pect. Maj.: Sternal
Spinal root segment	C2-C3, XI	C3-C4, XI	C4	C5-C6	C5-C6	C5-C6	C6-T1
Brachial plexus palsy type							
C2-C4	+	+	+	−	−	−	−
C5-T1	−	−	−	+	+	+	+
C5-C6	−	−	−	+	+	+	−
C5-C7	−	−	−	+	+	+	+
C5-C8	−	−	−	+	+	+	+
T1	−	−	−	−	−	−	−

*ECRB = extensor carpi radialis brevis, FCR = flexor carpi radialis, EDC = extensor digitorum communis, FDP = flexor digitorum profundus, C = cervical nerve root, XI = 11th cranial nerve, T1 = 1st thoracic nerve root, + = paralyzed, and − = not paralyzed.

TABLE V MRC Grades of Preoperative Muscle Power ➤

| | MRC Grade | | | | | | |
| | | | | | | Pectoralis Major | |
Case	Sternocleid omastoid	Trapezius	Phrenic N. Palsy	Shoulder Abduction	Shoulder Ext. Rotation	Clavicular	Sternal
1	5	5	No	0†	0†	0	0
2	5	5	No	0†	0†	0	0
3	4	4	No	0†	0†	0	4
4	2	0†	Yes	0†	0†	0†	0
5	3	0†	Yes	0†	0†	0†	0
6	5	5	No	0†	0†	0	0
7	4	3	No	0†	0†	0	0
8#	4	4	No	0†	1	0	3

*S = shoulder, E = elbow, H = hand, and N = neck. †Confirmed by needle electromyography or intraoperative nerve stimulation. ‡Biceps was completely paralyzed, but elbow could be flexed by forearm muscles. §Only the ulnar nerve-innervated intrinsic muscles. #Case 8, who presented late after the onset of paralysis, was the only patient not followed at 2 to 3-month intervals until the plateau of recovery was achieved.

Preoperative neurological evaluations were performed at a median of 3 months (range, 1 to 8 months) after the AFM onset. It was difficult to classify the types of paralysis according to the spinal segments that were involved—as is done in traumatic brachial plexus palsy—because there was a great variety of paralysis combinations, with certain types appearing more frequently than others[1,7]. Furthermore, the involvement of neck muscles is not uncommon in patients with AFM; this differs from brachial plexus palsy, in which the injuries are classified according to the nerve roots involved (such as C5-C6 or C5-C6-C7 palsy).

In view of the small number of AFM cases in our study, we categorized them according to the region with completely paralyzed muscles (no motor action potential on surface electromyography [EMG]) (Table IV). There were 3 patients with shoulder paralysis (loss of shoulder abduction and external rotation) without elbow paralysis, 3 patients with shoulder and elbow paralysis (loss of elbow flexion in addition to shoulder involvement) without hand paralysis,

and 2 patients with shoulder, elbow, and hand paralysis (loss of all upper-limb muscle function). Each of the patients with shoulder or shoulder-elbow palsy had additional paralysis of neck muscles, with involvement of the sternocleidomastoid, trapezius, and diaphragm as well. One patient with shoulder palsy had paralysis of the hand (wrist extensors and flexors and intrinsic muscles) due to incomplete recovery. In general, there was loss of shoulder abduction and external rotation in all patients, loss of elbow flexion in 5 patients, and complete or partial loss of hand function in 3 patients (Table V).

Surgical Reconstruction (Table VI)

Reconstruction to restore shoulder joint function: Nerve transfer surgery was preferred in cases with early presentation (<12 months after the onset of paralysis). The spinal accessory nerve was preferred as the donor nerve to address suprascapular nerve palsy. However, the contralateral C7 nerve root was used if the spinal accessory nerve was not available (due to

TABLE IV (*continued*)						
	AFM: Elbow		AFM: Hand				
	Biceps	Triceps	Wrist Ext.: ECRB	Wrist Flex.: FCR	Finger Ext: EDC	Finger Flex.: FDP	Intrinsic Muscles
	C5-C6	C6-C8	C7-C8	C6-C7	C7-C8	C8-T1	C8-T1
	−	−	−	−	−	−	−
	+	+	+	+	+	+	+
	+	−	−	−	−	−	−
	+	+	−	+	−	−	−
	+	+	+	+	+	−	−
	−	−	−	−	−	−	+

TABLE V (*continued*)							
	MRC Grade							
	Elbow Flexion	Elbow Extension	Wrist Extension	Wrist Flexion	Finger Extension	Finger Flexion	Intrinsic muscles	Type of Palsy*
	0†	0†	0	0	0	0	0	S-E-H
	2‡	0	2	0	0	0	0	S-E-H
	0†	3	4	4	4	4	4	S-E
	2†‡	2	5	5	5	5	5	N-S-E
	2	3	4	4	4	4	4	N-S
	3	2	0	0	4	2, 0§	2, 0§	S-H
	0†	2	4	4	4	4	4	S-E
	3	4	5	5	5	5	5	S

TABLE VI Types of Surgical Reconstruction

Case	Type of Palsy*	Time Between Palsy and Op. (mo)	Age at Op. (yr)	Primary Surgical Reconstruction Shoulder	Elbow	Wrist & Fingers	Secondary Reconstruction
1	S-E-H	6	5	Contralat. C7 n. root to long thoracic n. + suprascapular n.	Double free muscle transfer†	Double free muscle transfer	Zancolli metacarpophalangeal joint capsulodesis‡
2	S-E-H	13	4	Spinal accessory n. to suprascapular n.	Single muscle transfer§	Single muscle transfer	Biceps rerouting, extensor digitorum communis tenodesis, Zancolli metacarpophalangeal joint capsulodesis‡
3	S-E	5	5	Spinal accessory n. to suprascapular n.	Partial ulnar n. to musculocutan. n.		
4	N-S-E	8	10	Contralat. C7 n. root to spinal accessory n. + suprascapular n.; intercostal n. to axillary n.	Partial ulnar n. to musculocutan. nerve		
5	N-S	6	4	Contralat. C7 n. root to spinal accessory n. + suprascapular n.; thoracodorsal n. to axillary n.			
6	S-H	10	5	Spinal accessory n. to suprascapular n.		Ant. interosseous n. to ulnar n.	Tendon transfer
7	S-E	10	4	Spinal accessory n. to suprascapular n.	Partial ulnar n. to musculocutan. n.		Long head of triceps to acromion#
8	S	83	10	Multiple muscle transfer			
Median		9	5				
Range		5~83	4~10				

*S = shoulder, E = elbow, H = hand, and N = neck. †Double free muscle transfer was innervated from the spinal accessory and intercostal nerves. ‡The Zancolli metacarpophalangeal joint capsulodesis was useful to control claw-finger deformity. §Single muscle transfer was innervated from the intercostal nerves. #The transfer of the long head of the triceps to the acromion did not increase the active range of shoulder abduction.

paralysis or reserving it for free muscle transfer), and the contralateral C7 nerve root was routed to the suprascapular nerve and spinal accessory nerve. For patients with neck muscle paralysis, the function of the deltoid was reconstructed in addition to the suprascapular muscle, with the intercostal nerves or thoracodorsal nerve transferred to the axillary nerve. For the patient with late presentation (>12 months after the onset of paralysis), multiple muscle transfer was inevitable. This patient (Case 8) underwent transfer of the trapezius, pectoralis major, latissimus dorsi, and short head of the biceps for the left shoulder.

Reconstruction to restore elbow, wrist, and finger function: Five patients required reconstruction to restore elbow function, with 3 of them receiving partial ulnar nerve-to-musculocutaneous

TABLE VII Early Results: Time of Muscle Reinnervation Demonstrated by EMG

Case	Reinnervation* After Surgery (After Palsy Onset) (mo) Trapezius	Deltoid	Infraspinatus†	Biceps	Grafted Muscle	Abductor Digiti Minimi†
1			5 (11.5)		3‡, 4‡	
2			<13 (<24)		3	
3			3 (8.5)	3 (8.5)		
4	6.5 (14.5)	4 (12)	6.5 (14.5)	2.5 (10.5)		
5	6 (12)	7 (13)	8 (14)			
6			<6 (<16)			<6 (<16)
7			4 (13.5)	4 (13.5)		
8						

*The reinnervation of all muscles except 2 in Case 1 was confirmed by needle EMG. †< = far less than the estimated periods because the motor unit potentials were already mature at the time of examination. ‡Reinnervation was defined by visible contraction of the muscle.

TABLE VIII Long-Term Functional Outcomes

Case	Type of Palsy*	Follow-up (mo)	MRC Grade of Muscle Power			Range of Active Joint Motion* (°)				Power of Elbow Flexion	
			Sternocleido mastoid	Trapezius	Pectoralis Maj.	S Abd.	S Ext. Rot.	E Flex.	F Total Active Motion	MRC Grade	Hand-Held Dynamometer (% of uninvolved side)
1	S-E-H	94	5	5	0	90	60	140	180	3	26
2	S-E-H	75	5	5	2	40	80	140	180	3	Not tested
3	S-E	44	4	4	3	180†	90	150	Full	4	47
4	N-S-E	39	3	2	0	50	80	140	Full	4	34
5	N-S	39	3	3	4	160†	90	140‡	Full	4	59
6	S-H	38	5	5	4	130	70	150‡	150	3	22
7	S-E	38	4	4	0	30	90	140	Full	3	14
8	S	30	4	4	3	70	10	140	Full	3	Not tested
Median		39				80	80	140	180	3	30
Range		30-94				30-180	10-90	140-150	150-180	3-4	14-59

*S = shoulder, E = elbow, H = hand, and N = neck. †Supplementary action using shoulder flexion after 90° of shoulder abduction. ‡Spontaneous recovery of biceps.

nerve transfer and 2 patients receiving free muscle transfer (single in one and double in the other) to restore wrist and finger movement at the same time.

　　Secondary procedures: Four patients underwent secondary reconstruction to correct remaining disabilities. Procedures performed included Zancolli capsulodesis of the metacarpophalangeal joints for claw-finger correction, tenodesis to restore wrist extension, rerouting of the biceps tendon to restore forearm pronation, and transfer of the long head of the triceps to the acromion in an attempt to restore shoulder abduction.

Postoperative Management and Assessment

The shoulder and elbow were immobilized with an air cushion splint following shoulder and elbow joint reconstruction, and an additional long-arm cast was applied after hand reconstruction, for 4 weeks postoperatively. EMG was used to monitor the recovery process following nerve transfer surgery, starting at approximately 3 months postoperatively and continuing until full recovery of the motor unit potentials; EMG was needed to distinguish nerve recovery from recovery of synergic muscle function. Following electromyographic documentation of reinnervation, muscle reeducation training was

Fig. 3-A　　　　　　　　　Fig. 3-B　　　　　　　　　Fig. 3-C

Figs. 3-A, 3-B, and 3-C Case 1. A 5-year-old girl sustained complete paralysis of the left upper limb (C5~T1). She underwent transfer of the contralateral C7 nerve root to the left suprascapular nerve and the long thoracic nerve for restoration of function of the left shoulder at the first operation. Double free muscle transfers for restoration of finger and elbow function were performed at the second and third operations. The final surgery was performed to correct secondary claw-finger deformities with Zancolli metacarpophalangeal joint capsulodesis. **Fig. 3-A** Preoperative appearance showing complete paralysis of the left upper extremity. **Figs. 3-B and 3-C** Five years after the final operation. **Fig. 3-B** Photograph showing the patient's ability to abduct the left shoulder 90°. **Fig. 3-C** Photograph showing that full flexion of the left elbow was possible. Her finger motion is shown in Video 1.

Fig. 4-A

Fig. 4-B

Fig. 4-C

Fig. 4-D

Figs. 4-A through 4-D Case 5. A 4-year-old girl sustained right C2~C6 palsy with complete paralysis of the right trapezius, deltoid, supraspinatus, and biceps muscles, but flexion of the right elbow was possible with supplementary action of the forearm muscles. She underwent transfer of the contralateral C7 nerve root to the right spinal accessory nerve and suprascapular nerve at the first operation and transfer of the thoracodorsal nerve to the axillary nerve at the second operation. **Fig. 4-A** Preoperative appearance of the back, showing paralysis of the right trapezius with the left scapula higher than the right. **Fig. 4-B** Preoperative photograph showing an inability to abduct the right shoulder. **Fig. 4-C** Postoperative appearance of the back showing contraction of the right trapezius muscle with improvement of scapular elevation, although winging of the right scapula was still present. **Fig. 4-D** Photograph made 2 years after the final operation showing the patient's ability to abduct the right shoulder fully.

started using an EMG feedback technique with a myotrainer and surface electrodes.

Upper-limb function was assessed by measuring the active range of shoulder abduction and external rotation and elbow flexion and extension as well as the total active motion of the fingers. The Mallet functional scoring system was used to assess shoulder abduction and external rotation deficits. The strength of elbow flexion was measured qualitatively with the British Medical Research Council (MRC) grading scale and quantitatively with a handheld dynamometer (microFET2; Hoggan Scientific).

| | Mallet Score | | | CHQ-PF28 | | | | | |
| | | | | Physical | | | Psychosocial | | |
Case	Preop.	Postop.	Diff.	Preop.	Postop.	Diff.	Preop.	Postop.	Diff.
1	5	20	15	19	54	35	62	53	−9
2	5	17	12	25	31	6	60	61	1
3	5	25	20	8	33	25	34	47	13
4	10	12	2	19	36	17	29	46	17
5	11	24	13	15	34	19	25	46	21
6	15	21	6	38	40	2	60	64	4
7	6	12	6	24	29	5	32	45	13
8	8	16	8	21	26	5	43	50	7
Median	7	19	12	20	34	12	39	49	10
Range	5-15	12-25	2-20	8-38	26-54	2-35	25-62	45-64	−9-21

TABLE IX Preoperative and Postoperative Quality-of-Life Scores*

*Diff. = difference (change from preoperative to postoperative score).

Early and long-term outcome measures were analyzed, with the early results based on the timing of muscle reinnervation as determined with needle EMG and the long-term results based on the functional improvement of the affected limb. The Child Health Questionnaire-Parent Form 28 (CHQ-PF28) was used as a patient (parent)-reported outcome measure to assess overall improvement in quality of life.

Statistical Analysis
All data are presented as medians with ranges. Statistical analysis was not performed because of the small sample size.

Results

The reinnervation periods differed according to the muscles and the procedures performed (Table VII). The time for muscle reinnervation following contralateral C7 nerve root transfer was 6 months for the trapezius, 4 and 7 months for the deltoid, and 5 to 8 months for the infraspinatus. The infraspinatus muscles were reinnervated as early as 3 to 4 months following transfer of the spinal accessory nerve to the suprascapular nerve. Two patients had documented reinnervation at 6 and 13 months but mature motor unit potentials were evidenced at the time of these examinations; thus, the actual time of reinnervation was long before the documented period. As for the biceps muscle, the reinnervation periods were 2.5 to 4 months after partial ulnar nerve-to-musculocutaneous nerve transfer. The 3 transferred gracilis muscles in 2 patients survived well with no vascular complications and were reinnervated by 3 to 4 months after surgery.

The median duration of follow-up after the final surgical procedures was 39 months (range, 30 to 94 months) (Table VIII). The median ranges of shoulder abduction and external rotation

were both 80° (range, 30° to 180° and 10° to 90°, respectively). Four patients obtained at least 90° of shoulder abduction whereas the other 4 patients had shoulder abduction of ≤70°. The median Mallet score increased from 7 (range, 5 to 15) preoperatively to 19 (range, 12 to 25) postoperatively, with a median difference of 12 (range, 2 to 20) (Table IX). Elbow flexion in the 5 patients who received reconstruction (free muscle transfer or partial ulnar nerve-musculocutaneous nerve transfer) to restore elbow function was ≥140°, with the quantitative measurement of elbow flexion strength showing a median value of 30% (range, 14% to 59%) of the uninvolved side. Total active motion of the fingers for the 2 patients who underwent free muscle transfer to restore finger function was satisfactory (180°) (Table VIII, Figs. 3-A through 4-D, and Video 1).

In terms of quality-of-life improvement, the median physical and psychosocial summary scores of the CHQ-PF28 increased from 20 (range, 8 to 38) preoperatively to 34 (range, 26 to 54) postoperatively and from 39 (range, 25 to 62) to 49 (range, 45 to 64), respectively (Table IX).

Discussion

The patterns of motor paralysis in AFM are different from those in traumatic and obstetric brachial plexus palsy because of the involvement of the spinal cord anterior horn cells with widespread muscle dysfunction in AFM. Thus, AFM does not always follow the typical myotome distribution of a spinal segment[8-12].

All of the patients initially presented with monoplegia except for Case 5, who had diplegia; that patient's left upper limb recovered fully without surgical treatment. Our preoperative assessments (Table V) showed involvement of the shoulder and elbow muscles in the majority of patients. There was complete paralysis of the deltoid, supraspinatus,

and pectoralis major (clavicular head) in all patients and of the biceps in 5 patients. This pattern is similar to the spinal type of paralytic poliomyelitis, in which the proximal part of the limb is more commonly affected than the distal part[13]. However, the reason for the predominant involvement of the shoulder and elbow muscles in AFM is still unclear.

Although, to our knowledge, complete paralysis of the trapezius has not been reported previously in patients with poliomyelitis-like paralysis[8-12], 2 of our patients had complete paralysis of the trapezius and ipsilateral diaphragm and a third patient had partial recovery of the involved trapezius. Furthermore, Ruggieri et al. reported on 4 patients with diaphragmatic paralysis and respiratory failure requiring mechanical ventilation in a cluster of 11 patients with AFM[14]. Therefore, high cervical cord involvement is relatively common in AFM.

The prognosis of a paralytic muscle may depend on the status of associated muscles that are innervated by the same spinal segments, as is the case in poliomyelitis[15]. However, there is no evidence that this association with spinal segments occurs in the same way in AFM, the pathomechanism of which is still not well understood.

The outcomes of shoulder joint reconstruction in AFM have been inconsistent, just as they have been in brachial plexus palsy and poliomyelitis[8-12]. Transfer of the spinal accessory nerve to the suprascapular nerve was the most reliable surgical method for restoration of shoulder abduction in patients with brachial plexus palsy[16]. For our patients, the integrity of the donor nerve (spinal accessory nerve) was confirmed intraoperatively by a strong contraction of the trapezius seen on electrical stimulation. Nevertheless, half of the patients did not regain the expected shoulder abduction. The trapezius and/or spinal accessory nerve could be affected by the disease process as well, which might have prevented full recovery or allowed relapse even after a technically successful reinnervation.

Intraoperative electrical stimulation of the spinal accessory nerve is probably not reliable enough to prove a healthy donor nerve, as it is not a quantitative measurement. Also, restoration of the supraspinatus muscle alone might not be as effective as it is in patients with brachial plexus palsy, in whom concurrent partial palsy of the spinal accessory nerve is rare. Thus, we propose that, before performing shoulder reconstruction in AFM, the trapezius muscle strength be assessed with manual muscle testing[17] together with testing of compound muscle action potentials (CMAPs); neurotization of the axillary nerve would be necessary to restore deltoid function with a donor nerve not involved by the lesion, such as the triceps branch of the radial nerve.

Two of our patients had spinal accessory nerve palsy, and they were treated with transfer of the contralateral C7 nerve root to the affected spinal accessory nerve and suprascapular nerve. This technique has been proven to be useful in children with brachial plexus palsy[18,19]. Nerve transfer to the axillary nerve was also performed in the same patients to restore deltoid function. However, only 1 patient achieved a full range of shoulder abduction (Case 5) while the other patient's outcome (Case 4) was unsatisfactory because of relative weakness of synergistic muscles.

In our series, the outcomes in terms of elbow flexion were consistent for patients with either partial ulnar nerve transfer or free muscle transfer. They obtained elbow flexion of 140°, a median MRC grade of 3, and a median of 30% of the strength of the unaffected side (Table VIII). This was compatible with the reported surgical outcomes in several studies of nerve transfer for poliomyelitis-like paralysis, although half of the cases in those studies had spontaneous recovery[8-12]. Spontaneous recovery of biceps function in AFM was not unusual although the presence of elbow flexion could have been due to the action of the brachialis or forearm muscles. Two patients (Cases 5 and 6) with complete biceps paralysis initially had spontaneous reinnervation of the biceps with good elbow flexion power. Therefore, it might not be necessary to perform a reconstruction to restore elbow flexion in the early stage for patients with shoulder and elbow paralysis as long as the elbow flexion can be elicited clinically via supplementary action. Furthermore, it is always possible to restore elbow flexion at a late stage by free muscle transfer if the recovery is unsatisfactory[8].

To date, there are no standardized selection criteria for the donor nerves. Nationwide surveillance of a cluster of patients with AFM in Japan had identified normal F-wave persistence as the only significant independent factor for a good prognosis[7]. Preserved F-waves indicate an unaffected area of anterior horn cells for specific spinal segments. Thus, persistence of F-waves for median and/or ulnar nerves, although reduced or abnormal, could be a good indicator of the suitability of a donor nerve such as for a partial ulnar nerve transfer.

There is no method other than free muscle transfer that can provide satisfactory finger movement for patients with brachial plexus palsy. A similar technique can be employed effectively to restore grasping motion to patients with AFM[10,18]. Satisfactory total active motion was achieved for the 2 patients who underwent free muscle transfer (Cases 1 and 2).

The median Mallet score for the patients with AFM increased by 12 (range, 2 to 20), which was comparable with findings for patients with obstetric brachial plexus palsy[18]. Both the physical and psychological scores on the health-related quality-of-life assessment measured with the CHQ-PF28 improved postoperatively. However, the scores (Table IX) were still lower than those in children without any chronic condition, who demonstrated mean physical and psychosocial scores of 58.53 ± 4.28 and 53.86 ± 5.87, respectively, in a large population-based study by Bai et al.[20,21]. Thus, the postoperative improvements of the patients with AFM were still inadequate to meet parental expectations.

Although the small sample size of our study limits us from establishing any solid conclusions, there are a few important points that we would like to emphasize. We found a

great variety of paralytic patterns in AFM, with involvement of high cervical segments and the spinal accessory nerve not being uncommon. All patients had loss of shoulder abduction, and restoration of shoulder function was less predictable and depended on the quality of the donor nerves and recovery of the synergistic muscles. In contrast, the outcomes of the reconstructions to restore elbow and hand function were more consistent and satisfactory. ■

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