Scapulothoracic Arthrodesis in Facioscapulohumeral Dystrophy with Multifilament Cable

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Abstract Patients with facioscapulohumeral dystrophy (FSHD) are affected mostly by impaired shoulder function. Scapulothoracic arthrodesis was introduced to improve shoulder function. We evaluated the outcomes of scapulothoracic arthrodesis using multifilament cables, performed on 13 patients with FSHD (18 shoulders). There were eight males and five females (mean age, 29 years; range, 20–50 years). Outcome criteria were active shoulder forward flexion and abduction, the Disabilities of the Arm, Shoulder, and Hand (DASH) score, respiratory function tests, and a new shoulder function score. Patients were followed for a minimum of 24 months (average, 35.5 months; range, 24–87 months). Solid fusion was obtained in all shoulders (two after revision); active abduction range increased from $47.2^\circ \pm 11.6^\circ$ to $102.2^\circ \pm 10.0^\circ$ (mean $\pm$ standard deviation) and anterior flexion range from $55.6^\circ \pm 16.1^\circ$ to $126.1^\circ \pm 20.9^\circ$. The DASH score decreased from 33.6 ± 8.9 points preoperatively to 11.6 ± 8.0 points postoperatively. Shoulder function score increased from

15.9 ± 2.4 points to 22.2 ± 1.3 points. Scapulothoracic arthrodesis provides satisfactory function in patients with FSHD. Our data suggest use of multifilament cables for fixation is a reasonable option with an acceptable complication rate.

Level of Evidence: Level IV, case series. See the Guidelines for Authors for a complete description of levels of evidence.

Introduction

FSHD is a genetic disorder with an autosomal dominant inheritance pattern and characterized by weakness in facial and shoulder girdle musculature. The incidence of FSHD is approximately one in 20,000 live births [19]. Typically, the initial symptoms are difficulty whistling and in closing the eyelids and occur within the first 30 years of life [4]. Involvement of the shoulder girdle muscles results in impairment of upper extremity functions. The most prominent functional abnormality is progressive loss of strength in muscles stabilizing the scapula to the thoracic cage, namely, the serratus anterior, trapezius, and rhomboid muscles. Inability to fix the scapula to the thoracic wall causes instability of the origin of the deltoid muscle. Subsequently, weakness in shoulder abduction and anterior flexion develop. These patients have difficulty in performing overhead activities, during which winging of the scapula is observed. Repetitive movements cause muscle fatigue.

Fixing the scapula to the thoracic cage mostly overcomes these problems, providing a stable origin for the deltoid muscle [7, 10, 17, 21]. Various techniques to achieve fixation have been published [7, 10, 17, 21]. These procedures may be classified as dynamic and static

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methods. Dynamic methods using muscle transfers are contraindicated in FSHD as the muscles to be transferred (ie, pectoralis major) either are affected or expected to be affected during the course of the disorder [4]. The first static technique was reported by Putti [17], who performed interscapular fixation in 1906. Whitman [21] introduced a scapulothoracic fixation technique without arthrodesis in 1932. Scapulothoracic fixation without arthrodesis using cables (scapulopexy) is another method that was introduced to treat winging scapula secondary to FSHD [7, 12]. Scapulothoracic arthrodesis using screws and tibial strut grafts was first described by Howard [10]. Numerous modifications have been reported to provide improvement of shoulder functions when fusion is achieved [9, 12, 17, 21]. Scapulothoracic arthrodesis using wires or other plates in patients with FSHD provided improvement in functions in previous series [1, 3, 5, 14, 15, 18]. An alternate method of arthrodesis is the use of multifilament wire cables, which likely would provide more stable and stronger fixation.

To assess this approach in patients with FSHD, we therefore determined (1) fusion rates, (2) changes in active range of motion (ROM), (3) functional outcome scores, (4) respiratory capacity, and (5) complications. We also evaluated correlation between functional scores.

Materials and Methods

We retrospectively reviewed 13 patients (18 shoulders) with FSHD. The diagnosis was established with clinical evaluations, EMG findings, and genetic tests when available. Genetic testing [6, 8] for 4q35 deletion allele was performed in 10 of 13 patients who had surgery. This standard diagnostic testing was performed with pulsed-field gel electrophoresis and included DNA double digestion with EcoRI/HindIII and EcoRI/BlnI. Diagnosis was confirmed in eight patients as they had residual 4q35 fragments ranging from 10 to 32 kb. We recommended scapulothoracic arthrodesis for patients with difficulties with overhead activities, and with active abduction and active forward flexion ranging from 30° to 90°, which increased to 100° or greater when the scapula was manually fixated to the thorax (Horwitz maneuver for abduction test) [9]. We did not perform scapulothoracic arthrodesis fusion in patients with mild involvement who were able to elevate and abduct their arm greater than 90° without manual scapula fixation. All patients were graded using the Medical Research Council grading [16]. We considered strength of 2/5 or less of the deltoid muscle a contraindication for scapulothoracic arthrodesis. The study group consisted of 18 shoulders of 13 patients (eight men, five women) (Table 1). Their mean age at the time of scapulothoracic arthrodesis was 29 years (range, 20–50 years). Four patients had surgery on the left

side, four on the right side, and five had bilateral procedures. In bilateral cases with symmetric involvement, the dominant side was operated on first, whereas priority was given to the side with more severe involvement in asymmetric cases. The mean interval between the two operations was 5.6 months (range, 4–8 months). The patients were followed for a minimum of 24 months (average, 35.5 months; range, 24–87 months). The study protocol was approved by our institutional ethics committee.

All operations were performed by the same surgeon (MD). Patients received general anesthesia and were positioned prone at the edge of the operating table. The upper extremity was draped free, to allow for easy intraoperative handling. The ipsilateral posterior iliac crest also was draped free for graft harvesting. Cortical and cancellous autografts were harvested through an incision over the posterior iliac crest. A longitudinal incision parallel to the spinous processes of the thoracic vertebrae was made over the medial border of the scapula. The trapezius muscle was incised parallel to the skin incision. The levator scapula, rhomboid major, and rhomboid minor muscles were detached from the medial border of the scapula and retracted medially. The supraspinatus, infraspinatus, and teres major muscles were detached subperiosteally from the medial border of the scapula to expose a 3-cm-wide strip on the medial side of the scapula. To provide a broader contact area on the scapulothoracic joint surface, the subscapularis muscle was detached subperiosteally from the medial 5 to 6 cm of the scapula and the impinging part of the muscle was resected. With the scapula held in 20° to 30° abduction, the adjacent ribs (ribs 3–7 in 14 shoulders and ribs 2–6 in four shoulders) and the anterior surface of the scapula were decorticated with a high-speed burr. One centimeter from the medial border of the scapula, two holes, 1 cm apart, were punched over each rib with a blunt-tipped probe. Using periosteal rib elevators passing through drainage tubes to prevent pleural injury, the multifilament cables (Dall-MilesTM Recon and Trauma Cable System; Stryker, Kalamazoo, MI) were passed subperiosteally beneath the ribs. A mixture of autogenous iliac bone and cancellous allograft bone was used in all patients, and demineralized bone matrix (DBM) was added to the mixture in all but three shoulders (Patients 2, 4, 15). Cables were passed through the holes on the medial side of the scapula. The cables were tensioned and locked with specially produced beads, starting from the relatively thicker inferior angle of the scapula and advancing superiorly. Stability of the fixation of the scapula was tested. Layers were closed over a suction drain.

We immobilized the shoulders in a 30°-abduction pillow sling for 3 months (Fig. 1). Removal of the sling was allowed during showering. Elbow and wrist ROM and

Table 1. Demographic data and functional results of the study group

Patient number	Shoulder number	Age (years)	Gender	Side	Followup (months)	Abduction preoperative (degrees)	Abduction postoperative (degrees)	Flexion preoperative (degrees)	Flexion postoperative (degrees)	DASH score preoperative	DASH score postoperative	Functional score (SFS) preoperative	Functional score (SFS) postoperative
1	1	20	M	L	87	60	100	85	150	18.10	3.4	20	24
2	2	50	M	R	46	60	120	70	160	23.21	4.5	17	22
3	3	22	F	R	38	30	80	40	90	39.28	16.5	14	19
4	4*	27	F	L	40	45	105 (90)**	45	135 (90)**	41.96	8.5	16	22 (20)**
4	5	27	F	R	35	45	120	45	150	41.96	8.5	14	24
5	6	43	M	L	35	30	90	30	100	50	22.4	14	21
6	7	25	M	L	36	60	100	75	125	22.41	5.2	20	24
6	8*	25	M	R	28	65	100	85	130	22.41	5.2	20	24
7	9	36	M	L	34	40	90	50	110	40.17	15.2	14	22
7	10	36	M	R	24	30	90	40	90	40.17	15.2	12	20
8	11	25	M	L	30	50	120	60	160	33.92	4.5	17	24
8	12	25	M	R	24	50	120	50	160	33.92	4.5	14	24
9	13	30	F	R	41	55	100	70	110	34.25	12	17	24
10	14	33	F	R	27	60	120	60	150	28.57	4.2	17	22
11	15*	20	M	L	37	45	100 (70)**	50	130 (90)**	33.33	5.6	14	22 (17)**
11	16	20	M	R	31	45	110	45	130	33.33	5.6	16	24
12	17	32	M	L	25	50	110	60	130	23.21	10.7	17	22
13	18	28	F	L	24	30	90	40	120	41.96	26.2	14	20
Mean***		29.11			35.5	47.2	103.8	55.6	129.1	33.5	9.9	15.9	22.5
SD***		8.14			14.4	11.7	13.6	16.1	24.4	8.8	6.7	2.4	1.75

* Shoulders with complications.

** Values in parentheses indicate results before revision.

*** SD = Standard deviation, excluding the values before revision.

Fig. 1 An arm sling with pillow provides approximately 30° abduction.

isometric deltoid exercises were performed during this period. At the end of 2 months, passive shoulder anterior flexion, abduction, and rotation exercises were begun. At the end of 3 months, the arm sling was discontinued and daily activities were allowed.

Patients were seen for followup every week for 1 month, then once a month until 6 months postoperative, and then every 3 months until the second postoperative year. Clinical outcome parameters were the preoperative and postoperative values of active shoulder abduction and forward flexion ranges and the DASH score [11]. The DASH score is a subjective, self-reported questionnaire designed to measure physical function and symptoms in patients with any of several musculoskeletal disorders of the upper limb. We considered the DASH score a gold-standard because it is widely used in upper extremity studies. Respiratory function tests were obtained preoperatively and postoperatively. Complications also were recorded. In addition to these standard parameters, an objective shoulder function score (SFS) was used to document the preoperative and postoperative status of the patients (Table 1). This score was developed specifically for patients with FSHD by a group of specialists including the authors. The SFS is based on five clinical parameters: active shoulder abduction, active shoulder flexion, ability to put hand above the head, shoulder flexion, and abduction strength. The SFS was evaluated independently by two examiners, an orthopaedic surgeon (OU) and a neurologist (SY). Using the Pearson correlation test, we found a high

level of interobserver agreement ($r = 0.869$ for preoperative values, 0.891 for postoperative values; both $p < 0.001$). We used manual muscle testing for grading strength according to the Medical Research Council as described previously [16]. Higher scores indicate better results and the total score was calculated by adding the scores of each parameter (Table 1).

Radiographic evaluation with direct xray was performed every 6 weeks, and CT examinations with three-dimensional (3-D) reconstruction were performed for 11 patients after the sixth month. Union was assessed using dynamic anteroposterior radiographs of all shoulders and 3-D reconstruction with CT in 11 of 18 shoulders.

We used a paired Student's *t* test to compare preoperative and postoperative results of each outcome parameter. We used Pearson's correlation test to explore any correlation between preoperative DASH and SFS scores and postoperative results of the same parameters. All statistical analyses were performed using SPSS[®] for Windows[®] standard version 13.0 software (SPSS Inc, Chicago, IL).

Results

Clinical and radiologic healing was obtained in all shoulders (after the primary surgery in 16 shoulders and after revision surgery in the remaining two shoulders). Additional CT examinations were performed in 11 of the 18 shoulders after 6 months postoperatively and bony fusion was documented in all 11 (Fig. 2).

Active abduction range increased ($p < 0.001$ mean difference, 55.0° ; 95% confidence interval [CI], 50° – 60°) from $47.2^\circ \pm 11.6^\circ$ (mean $\pm$ standard deviation) preoperatively to $102.2^\circ \pm 10.0^\circ$ postoperatively. Similarly, anterior flexion range increased ($p < 0.001$) from $55.6^\circ \pm 16.1^\circ$ preoperatively to $126.1^\circ \pm 20.9^\circ$ postoperatively (mean difference, 73° ; 95% CI, 61° – 80°) (Table 1). No deterioration was observed during followup. A decrease ($p < 0.001$) was observed in the DASH score from 33.6 ± 8.9 points preoperatively to 11.6 ± 8.0 points postoperatively (mean difference, 22.0 points; 95% CI, 19.1–25.0 points). Postoperative SFS scores (22.2 ± 1.3 points) were higher ($p < 0.001$) compared with preoperative scores (15.9 ± 2.4 points) (mean difference, 6.3 points; 95% CI, 5.4–7.1 points) (Table 1).

Postoperative forced expiratory volume ($93.0\% \pm 21.1\%$) and forced vital capacity ($83.4\% \pm 18.2\%$) values were similar to preoperative results ($92.3\% \pm 19.7\%$ and $83.5\% \pm 16.5\%$, respectively).

We observed a negative correlation between the values of DASH and FSHD shoulder function scores, preoperatively ($r = -0.841$) and postoperatively ($r = -0.751$) ($p < 0.001$ for both).

Fig. 2A–G (A) A lateral photograph of a 25-year-old man (Patient 8) shows active anterior flexion of both shoulders and the winging scapula preoperatively. (B) An intraoperative photograph shows the cables passing under the ribs. (C) The position of the cables (arrows) is shown after tightening and locking. (D) This radiograph was obtained during the early postoperative period after first (left) site surgery. (E) An anteroposterior radiograph of both shoulders taken at the last followup shows correct position of the scapula is preserved. (F) A 3-D CT scan obtained 6 months after surgery shows completed fusion. (G) A postoperative clinical photograph shows the active flexion of the left upper extremity.

No perioperative complications occurred. Two patients (two shoulders) had nonunions and cable loosening occurred in another, all three of whom had bilateral operations. In Patient 4, pain began in the superior part of the fusion area 2 months postoperatively (left side of a bilateral case). Direct radiographs showed a fracture of rib 3 and lysis in rib 6. The patient did not accept the recommended revision surgery and the contralateral side was fused 6 months after the first operation. One year later, she decided to have revision surgery, having seen the difference between her two shoulders. She was operated on using the same technique, but with larger amounts of graft material and DBM putty. At the last followup, 2 years after the right scapulothoracic arthrodesis and 8 months after the left revision, her right and left shoulder abduction were 100° and 105°, and flexion were 135° and 135°, respectively. In Patient 11, who had bilateral scapulothoracic arthrodeses, cables at ribs 3 and 7 failed 3 months after the first operation on the left side. The patient complained of pain and

recurrence of winging scapula. Cable fixation using the same ribs was preferred for revision fusion. Complete resolution of the patient's complaints was achieved after 3 months of immobilization. Maximum active abduction and flexion values increased to 100° and 130°, respectively. For the right shoulder of Patient 6, which was operated on 4 months after the contralateral side, control radiographs showed failure of the uppermost and lowermost cables (ribs 2 and 6) 7 months postoperatively (Fig. 3). No motion was present at the scapulothoracic joint; maximum active abduction was 100° and flexion was 130°. CT confirmed the presence of fusion at the 9th month, and irritating cables were removed afterward (Fig. 3).

Discussion

Facioscapulohumeral dystrophy is a disabling disease most often affecting the shoulder region. The main problem is

Fig. 3 A radiograph of both shoulders of a 25-year-old patient (Patient 6) taken 7 months postoperatively shows the failed cables on ribs 2 and 6 (arrows) in the right shoulder.

instability of the scapula during attempted shoulder elevation and abduction. Scapulothoracic arthrodesis has been suggested as an effective solution for this problem and one approach is use of multifilament fixation [1, 3, 5]. To assess this approach, we therefore determined (1) fusion rates, (2) changes in active ROM, (3) functional outcome scores, (4) respiratory capacity, and (5) complications. We also evaluated correlation between functional scores.

The major shortcoming of the study is the lack of a control group. However, establishing a control group is difficult for two reasons. First, observing a control group of

patients without treatment of such a functionally disabling disorder would be ethically unacceptable. Second, because FSHD is a rare disorder, performing another type of operation to compare the results either would take a long time or diminish the size of an already small group of patients. In addition, a minimum of 2 years followup is relatively short for a progressive neuromuscular disease. The need for validation of the new SFS is another weakness of the study. However, we observed a correlation with the DASH score. Therefore, the score may be used for evaluation of patients with FSHD after validation in larger series.

Our data suggest scapulothoracic arthrodesis using multifilament cables reduced scapular winging and improved function of the shoulder muscles in patients with FSHD. The substantial improvements in clinical parameters in our patients were in accordance with those in previous series (Table 2) [1, 3, 5, 12, 15, 18, 20]. A similar improvement in active shoulder abduction was observed in our patients, whereas the increase in mean forward flexion (73°) was relatively higher compared with values in other series.

The decrease in DASH scores (from 34 points to 12 points) indicated patients' satisfaction. In some other series [1, 18], ROM and improvement in daily activities without numeric values such as the DASH score were used. A special scoring system for patients with FSHD with scapulothoracic fusion was introduced previously [5], however that scoring system is less detailed than the SFS.

In addition to the subjective DASH score, clinical results were evaluated using an objective evaluation system. Similar to the DASH score, we observed improvements with this new SFS score. The presence of a strong correlation between DASH and SFS scores led us to assume this new score might be a useful tool for researchers. Special importance is given to shoulder flexion and abduction strengths in this scoring schema, which are affected mainly by the anterior and midportions of the deltoid muscle.

Table 2. Comparison of published series

Study	Shoulders (number)	Technique	Followup (months)	Radiologic union	Abduction (degrees)	Flexion (degrees)	Function
Berne et al. [1]	47	Wire and plates	102 (12–257)	46/47	81	81	improved (subjective)
Copeland et al. [3]	13	Bone graft, screws	192 (12–528)	NA	103	123	Constant and Murley score 59
Diab et al. [5]	11	Wire and plate/washer	75 (24–120)	All	139	134	11/13 good-excellent
Rhee and Ha [18]	9	Wire-plate	102 (56–118)	All	108	109	UCLA score improved from 18 to 28
Letournel et al. [15]	9	Wire	24–133	All	NA	NA	improved
Jakab and Gledhill [12]	3	Wire	35	All	NA	NA	improved
Twyman et al. [20]	6	Wire	49 (12–80)	5/6	NA	NA	improved
Current study	18	Multifilament cable	36 (24–87)	All (two after revision)	102	126	DASH and shoulder function scores improved

NA = not available; ranges for followup in parentheses.

Because the deltoid either is spared or affected in late stages of the disorder, severity of deltoid muscle involvement is used as a prognostic parameter. Subjective criteria such as fatigue or pain were not included in the parameters, thus increasing the reproducibility of the score.

After achieving scapulothoracic fusion, the functional capacity of the patient will be affected only by severity of the progression of FSHD. We have not observed any deterioration in the functional results and ROM after an average of 3 years followup. We will continue followup of the patients and evaluate long-term results. Because fusion was achieved in all patients, we do not expect future problems resulting from the fusion. However, FSHD is a progressive disease and if the deltoid and/or trapezius involvement worsens, shoulder functions of patients also might worsen.

Fusing the scapulothoracic articulation is a relatively difficult task compared with most other arthrodesis sites, because both sides of the fusion consist of thin and fragile bones. In some series, revisions for nonunions were reported at 3% to 6% [1, 3, 20]. The nonunion rate in our series (two of 18 shoulders) may be related to patients who started vigorous activities early during the recommended immobilization period. Both patients with nonunion had successful revision and achieved the same functional levels as the other patients. Some criteria should be considered during selection of a technique for scapulothoracic arthrodesis: (1) fixation strength should provide secure stabilization between the scapula and thorax during the healing period; (2) the complication rate should be low; and (3) routine removal of the implants should not be required. Owing to difficulties in screw fixation of thin bones and the close proximity of the pleura, wire fixation was the preferred method by some authors [12, 20]. Wires were augmented with plates on the scapular side by others [9, 15, 18]. The cables we used avoided the need for an augmenting implant such as a plate, as the beads of the cables provided a wide surface contact area, creating a buttress effect, similar to a plate or washers used by other authors [1, 18]. Loosening and stretching were minimized and stable fixation was achieved. We did not believe it was necessary to use an augmenting implant to protect the medial scapular margin. We recommend shoulder immobilization in the sling for at least 3 months.

Selecting the number of ribs involved in the fusion is controversial [9, 13, 15]. We fused five consecutive ribs, providing adequate stability in 15 of 18 shoulders. We believe five-level fusion is a practical upper limit for scapulothoracic arthrodesis, because the flat scapula cannot be approximated to the convex thorax at both ends.

Verification of a solid fusion at the arthrodesis site is difficult with clinical examination and direct radiographs. The CT examination with 3-D reconstruction was valuable

for this purpose. Therefore we evaluated most patients with 3-D CT.

The scapulothoracic region is prone to perioperative complications. Intraoperative and postoperative complications such as pneumothorax, hemothorax, rib fracture, and scapular fracture have been reported [2, 9, 12, 20]. In a series of 47 patients operated on using plate-screws and wires, fracture of the scapula and ribs during screw insertion occurred in one patient, bilateral stress fractures of the scapula occurred in one patient 2 years after the operation, and two rib fractures occurred in one patient during followup [9]. In a series of four shoulders, fused using 16-gauge Luque wires, no complications were observed [12]. In another study, the authors stabilized the scapula to six consecutive ribs (ribs 2–7) in 12 shoulders using 18-gauge wires and reported one symptomatic nonunion (8.3%) [20]. Another report revealed a brachial plexus palsy that resolved spontaneously in one patient and there were no complications related to the wire fixation technique, such as rib or scapula fracture or implant failure [2]. In our series, there were three complications, of which two (11.1%) had revision arthrodesis.

One possible consequence of scapulothoracic arthrodesis is limitation in expansion of the thoracic cage, resulting in reduced pulmonary capacity. Twyman et al. [20] reported a 21% decrease in the forced vital capacity and a 14% decrease in the forced expiratory volume postoperatively. Others reported they observed no change in pulmonary functions [1, 9, 15]. We observed no changes in the pulmonary reserve after one-sided scapulothoracic arthrodesis.

A successful scapulothoracic arthrodesis provides satisfactory clinical function in patients with FSHD. Use of multifilament cables for fixation is a reasonable option with a low complication rate.

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