

# Management of scapular dysfunction in facioscapulohumeral muscular dystrophy: the biomechanics of winging, arthrodesis indications, techniques and outcomes

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- Facioscapulohumeral muscular dystrophy (FSHD) is a common hereditary disorder which typically results in scapular winging due to wasting of the periscapular muscles affected by this condition.
- Scapulothoracic arthrodesis (STA) is the current surgical treatment for FSHD patients with severe winging and preserved deltoid muscle.
- There are several different techniques in the literature such as multifilament cables alone and cable or cerclage wires combined with single or multiple plates. We prefer cables without plates as it provides independent strong fixation points and strongly recommend utilization of autograft.
- The functional results of studies report that regardless of the technique used, shoulder elevation and thus quality of life is improved, as shown with outcome scores.
- There are several complications associated with STA. Pulmonary complications are common and usually resolve spontaneously. Meticulous surgical technique and effective postoperative analgesia may reduce the incidence. Scapular complications which are associated with the fixation may be encountered in the early or late period, which are related to the learning curve of the surgeon.
- In conclusion, STA is a reliable solution to a major problem in FSHD patients that helps them maintain their activities of daily living until a cure for the disease is found. A successful result is strongly dependent on patient selection, and a multidisciplinary team of neurologists, geneticists and orthopaedic surgeons is required to achieve good results.

## Keywords

- ▶ scapulothoracic arthrodesis
- ▶ facioscapulohumeral dystrophy
- ▶ scapular winging

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## Introduction

Facioscapulohumeral muscular dystrophy (FSHD) is the third most common hereditary muscular dystrophy, which particularly affects facial and periscapular muscles, hence the name facio-scapulo-humeral dystrophy. The disease also affects abdominal, pelvic and lower extremity muscles and rarely respiratory muscles (1). In most cases, the course of the disease is not fatal and the patients' life span is usually not shortened; however, it can cause a significant negative impact on the quality of life (QoL), by intensely interfering with the activities of daily living (2, 3). The patients typically become symptomatic in their late adolescent period. Early-onset FSHD has also been

defined, where initial symptoms are documented in the early childhood period (4). Commonly, the initial symptom is facial muscle involvement, and it is often overlooked. The expressionless face pattern is often attributed to family traits, and this is thought to be one of the reasons why patients remain undiagnosed in earlier phases (5, 6). When the shoulder girdle and periscapular muscles are involved, the scapular winging becomes apparent with patients unable to elevate their arms overhead.

The reported prevalence in the literature is variable depending on the region; however, more recent reports suggest a prevalence range of 1/8000 to 1/20 000 (7, 8, 9). This relatively wide range can be explained by an important number of undiagnosed cases due to the

paucity of apparent signs and symptoms. Moreover, a patient may present with extensive lower and upper body involvement, whereas a first-degree relative may have subtle symptoms. There is no predilection for any gender; however, in case of mosaicism, curiously, male patients become symptomatic earlier than female patients (10, 11).

FSDH is inherited in an autosomal dominant pattern, and the clinical severity and progression rate is highly variable. There are two well-defined subtypes, FSDH1 and FSDH2. In the common form, the genetic abnormality is related to a repetitive element called D4Z4 on chromosome 4q, which is seen as 11–150 repeat units in the healthy population (12). This area is contracted in FSDH1 with less repeats. In the genetically extreme cases in which the repetition count is decreased to 1–3, the symptoms are seen much earlier (13). Half of these patients are reported to become symptomatic in the first decade of life (14). The term infantile form was first described by Brooke (15). Main diagnostic criteria are mainly the same; however, the facial symptoms must be seen earlier than 5 years of age and shoulder signs must be seen earlier than 10 years of age per the diagnostic criteria that were defined by Brouwer *et al.* (16). However, the ‘infantile form’ is not necessarily related to the repetition count (14), but it is just a term based on the timing of the symptoms. In FSDH2, the condition is more likely related to heterozygous dominant mutations in the SMCHD1 gene, which is responsible for silencing this area. In both, myotoxic DUX4 protein is synthesized. The D4Z4 repeats are important for the pathologic condition to occur; however, it is not sufficient by itself and the exact genetic mechanism is yet to be determined (1, 7).

Scapulothoracic arthrodesis (STA) is a century-old solution to shoulder dysfunction due to scapular winging. It was first described as scapulopexy (17), not a fusion but a permanent fixation, which frequently failed due to material insufficiency. This technique later evolved to arthrodesis which strives for fusion of the scapula to the adjacent ribs, by means of a variety of implants. STA using screws and tibia strut grafts was first described by Howard *et al.* in 1961 (18), to stabilize the scapula to provide a stable point of rotation for the humerus. Since then, various modifications have been proposed, utilizing different types of implants, ranging from relatively non-rigid methods such as tapes to rigid plate-screw structures (19, 20, 21, 22, 23, 24, 25, 26, 27, 28, 29, 30, 31, 32, 33).

The purpose of this review is to summarize our current understanding of the scapulothoracic motion in FSDH and to provide a detailed summary of the techniques used in STA and results, as well as to describe our novel technique using non-absorbable multifilament sutures and cables.

## Normal scapula function and biomechanics

Scapula acts as a concrete base without compromising mobility. This complex function is achieved by many muscles acting in perfect coordination: trapezius, rhomboid major, rhomboid minor, levator scapulae, teres major, latissimus dorsi, serratus anterior and the rotator cuff muscles. The synergistic movement flow of the anterior muscles (pectoralis minor and major) and the posterior muscles (trapezius and latissimus dorsi) determines whether the scapula protracts, retracts or remains in neutral position during the scapular motion. Coordinated scapular movements such as upward and downward tilt or internal and external rotation require voluntary changes in the balance between those muscles in favour of the intended direction. Scapular protraction is produced by pectoralis minor, serratus anterior and pectoralis major muscles (34).

A functional shoulder can be simplified in three functions acting in perfect synergy: (i) deltoid, the main power generator of the shoulder elevation, (ii) rotator cuff muscles, stabilizers of humeral head over the glenohumeral centre of rotation and (iii) periscapular muscles stabilizing the scapula on the thoracic wall so that the force output can be transferred to the arm. Scapular stabilization is not an absolute fixation at a certain position, but instead, it is providing a limited scapular repositioning to facilitate the glenohumeral motion. It was shown that during the functional shoulder elevation, the scapula rotates upwards and outwards and tilts posteriorly (35). This sequence of motion is provided by the upper trapezius, levator scapulae and the rhomboid muscles. To establish the scapular upward rotation, the middle trapezius muscle fires in concordance with the serratus anterior, while the lower trapezius stabilizes the scapula by counterbalancing the scapular elevation and protraction produced by its upper fibres along with serratus anterior (36).

In case of FSDH, as the periscapular muscles fail to function adequately due to the decrease in their strength, scapular stabilization cannot be established during shoulder motion. This failure results in scapular winging: an excessive protraction and medial tilting of the scapula while attempting shoulder elevation. There are several scapular winging patterns: lateral winging due to trapezius dysfunction (spinal accessory nerve), medial winging due to serratus anterior dysfunction (long thoracic nerve), as well as other ill-defined patterns related to the scapular dyskinesia (37). Recently, Basem el Hassan redefined the spectrum of scapular dysfunction, introducing the term scapulothoracic abnormal motion (38). The scapular winging pattern in FSDH has not been clearly defined due to the variation in the muscle involvement and presentation. Considering that trapezius,

serratus anterior, pectoral muscles, and rhomboids are all affected to a variable degree, it can be classified as a mixed type of scapular winging.

### Patient evaluation and surgical indication

An FSHD diagnosis can be established clinically with high accuracy, but a solid diagnosis should be supported by molecular methods (39, 40, 41, 42). A clinical diagnosis alone is always prone to error due to similar dystrophies affecting shoulder girdle. A reliable FSHD diagnosis should rely on both an appropriate clinical presentation and molecular testing (7, 43). Considering that the diagnostic part of the disease poses a difficult challenge not only for clinicians but geneticists as well, and surgery is only indicated in a portion of patients, the importance of an established centre with a multidisciplinary approach becomes apparent. We strongly recommend that diagnosis, surgical treatment, rehabilitation and therapeutic trials on these patients are managed in specialized referral centres.

The evaluation of a patient starts with sorting out the traits of the disease, bearing in mind its highly variable presentation. Other muscle dystrophies affecting shoulder girdle must be excluded. Family history has a substantial role in the identification of patients with FSHD; however, it is reported that up to one-third of patients might be the first case in the family (44). Classically, FSHD has a descending progression of weakness, starting from the facial muscles and subsequently involving the periscapular, humeral and core muscles and lower extremity muscles (2). The involvement is usually bilateral, whereas the degree of weakness is not symmetrical, which may result in uneven winging of the scapula (45). Manual and quantitative muscle testing as well as functional testing have also been suggested to help with the diagnosis (46). The 'FSHD Evaluation Scale', which was developed to grade the overall severity, utilizes six different domains: facial,

scapular girdle, upper limbs, legs, pelvic girdle and abdominal muscles (40).

Shoulder examination begins with inspecting the resting position of the scapulae, which may reveal an asymmetry. The scapulae of FSHD patients are usually protracted, medially rotated and anteriorly tilted in resting position due to the weakness of trapezius, rhomboid and serratus anterior (2). The abnormal position of the scapula results in a decrease in the activation of the anterior deltoid, which exacerbates the weakness in forward flexion and manifests with an atrophic deltoid. This phenomenon conveys the importance of the thorough evaluation of the anterior deltoid when planning for surgery. It is crucial to assess both shoulders' elevation simultaneously to eliminate the truncal compensation. The assessment of active and passive ranges of motion can be performed while seated in patients with significant truncal involvement. In an FSHD patient, any attempt of shoulder elevation classically results in notable scapular winging to a variable degree depending on the remaining stabilizer muscles (Fig. 1). Complete motor function of the upper extremity should be documented before surgery. Considering that the brachial plexus neuropathy is a reported complication of arthrodesis, preoperative neurological assessment would be highly valuable to identify such an injury (19, 47, 48).

FSHD involves truncal and hip muscles as the condition progresses. The patients typically have hip extensor and abdominal muscle weakness which manifest as lumbar hyperlordosis (2, 49). The Beevor's sign, which was originally described in association with upper motor neuron lesions of the thoracic spinal cord affecting the rectus abdominis muscle, can also be present and has been found to have 90% sensitivity and specificity for FSHD (50). Another exception was thought to be the involvement of tibialis anterior muscle as lower extremity involvement often starts with a drop foot; however, this assumption has been challenged by Olsen *et al.*, as they found that the hamstrings were more severely affected (51).

As mentioned above, a deltoid power output which is enough to elevate the arm is essential for a patient to benefit from STA. Assessment of deltoid is the hallmark of operative indication (Fig. 2). A patient's potential improvement in postoperative elevation is determined by using the Horwitz manoeuvre (scapular stabilization test) (52) which simply mimics the external scapular stabilization and exhibits whether the patient can achieve further elevation if the scapula is stabilized. While assessing the active shoulder elevation, the examiner first positions the scapula in a retracted, posteriorly tilted and laterally rotated position and stabilizes the scapula manually to better document the strength of the deltoid (Fig. 3). This step is important to document the patient's benefit from surgery, as it imitates the postsurgical mechanics.



**Figure 1**  
(A) Resting position; (B) scapular winging.

The 'FSHD Evaluation Scale' (40) mentioned previously assesses overall severity and has very limited benefit to identify patients suitable for STA. We have previously described a comprehensive staging system (53) focusing particularly on scapular dysfunction and deltoid strength (Table 1). This descriptive system consists of six different stages of the disease and is based on the elevation of the arm, contraction and function of the deltoid muscle and the extent of the scapular winging.

Particular attention is required for early-onset patients (infantile cases). It is known that these patients not only become symptomatic earlier, but they also tend to deteriorate faster (4). This deterioration is accompanied by the hazard of losing the functional gains earlier, which might have been provided with surgical treatment. Moreover, early-onset patients may have more severe muscle wasting in their deltoids, which may also cause a worse outcome. A long-term follow-up of these patients is advised before attempting surgical treatment.

In patients with a shoulder elevation of 120° (stage 0 and stage 1), surgery is avoided as it may have detrimental effects on shoulder function due to missing scapulothoracic contribution. Surgery in patients with shoulder elevation from 90° to 120° with mild scapular winging (stage 2) would be rather cosmetic than functional. Patients with shoulder elevation less than 90°, full deltoid function and severe scapular winging are considered as stage 3, comprising the best candidates for surgical treatment. In cases where deltoid strength is also affected, functional benefits of surgery may be limited (stage 4). Expected outcome of STA for this patient group should be carefully explained to the patients, considering

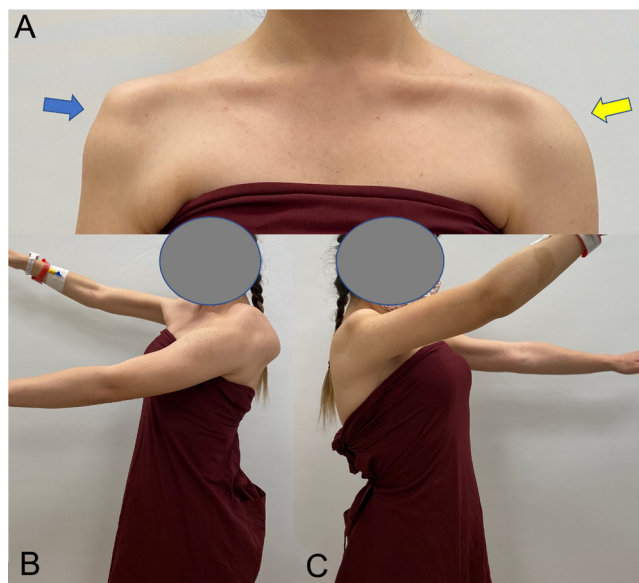
their lifestyle and expectations. Additionally, the age of the patient is important, and the surgical treatment outcome is less predictable for patients over the age of 40.

#### *The aim of surgery*

As foretold, FSHD selectively causes weakness in periscapular and pectoral muscles as well as biceps and triceps muscles while sparing the deltoid, rotator cuff and forearm muscles. This specific distribution results in a shoulder that could be fully functional but challenged with the loss of physiological motions. Since the deltoid is still functional in these patients, when a stable scapula is established by means of surgical intervention, they can regain their elevation. It should be kept in mind that the patients who have a slower progression rate are better candidates for surgical treatment.

The main objective of STA is to eliminate scapular instability and restore the contribution of the glenohumeral joint to shoulder motion. The expected outcome of the surgery is variable (19, 20, 21, 22, 23, 24, 25, 26, 27, 28, 29, 30, 31, 32, 33); however, the current literature reports that 120° of elevation may be achieved (19). The functional outcome is dependent on the preoperative strength, scapula position and glenohumeral motion. The weakness of the anterior deltoid and presence of contractures, particularly posterior tightness, have a great impact on achieved postoperative range of motion (ROM). The increase in elevation does not come without a cost: the FSHD patients have usually accustomed to their increased internal rotation, and STA may impede this motion. Surgeons should carefully manage the expectations of the patient, and the possible benefits and sacrifices of the surgery should be prudently explained.

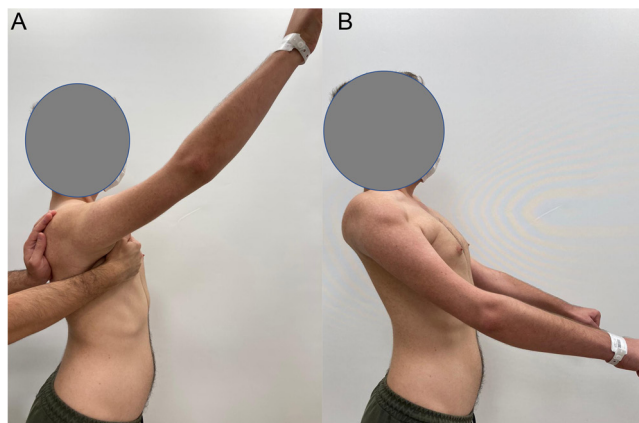
Presently, it is not clear how the arthrodesis position affects postoperative function. In a healthy individual, the scapular spine has a horizontal position that is within +5° and -5° of the scapular vertical rotation (54). The healthy scapula is in 40° of internal rotation in the coronal plane and has a 10° of anterior tilt (55) and the medial border is positioned parallel to the thoracic midline (56). It should be noted that there may be a difference in the position of the scapula in the dominant and non-dominant side (56). In the first surgery of a patient, a physiological resting position of the scapula is aimed with small variations to achieve an optimal contact surface with adjacent ribs. Contralateral surgery directly aims for the identical position of the previous side, as even minor changes in arthrodesis position would generate apparent shoulder asymmetry. Further studies are required to identify the optimal position of the scapula from a functional point of view. Typically, bilateral surgery is avoided, not only because of the long immobilization period but also due to possible pulmonary complications.



**Figure 2**

(A) Deltoid atrophy; (B, C) examination of shoulder elevation.





**Figure 3**

(A) Stabilization of the scapula mimics postoperative range of motion; (B) without stabilization.

A ‘square-shaped’ upper body was reported by several authors as an expected result of STA (22, 57). Not only the position, but prominent acromia due to an atrophic deltoid (particularly anterior) also contribute to squaring. This change in body image, although being a rare complaint, should be discussed with patients before surgery (Fig. 4).

Techniques that were previously reported in the literature are summarized in Table 2.

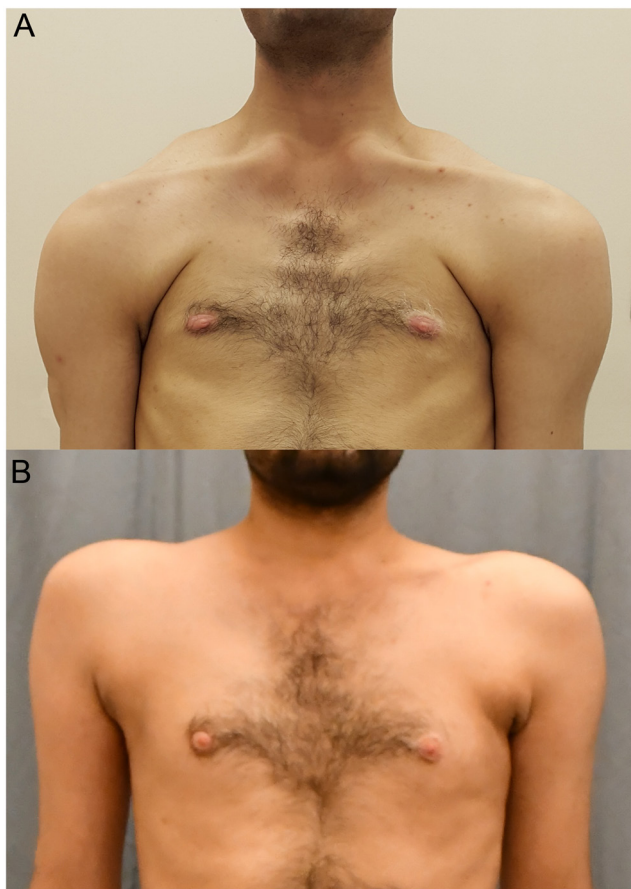
### Authors’ preferred technique

The patient is prepared in a prone position, with an arm rest placed adjacent to the table to support the abducted position. The arm is draped free and included in the surgical site along with the iliac crest and whole spine. Surgeon is positioned on the contralateral side of the affected arm to increase the visualization of the undersurface of scapula, while the assistant and surgical nurse are on the affected side. An oblique incision is made over the posterior iliac spine. Spongy and cortical grafts are harvested in a standard fashion. An oblique incision

(from medial cranial to lateral caudal) is made over the medial border of the scapula from the level of the second rib to the seventh rib. Deep dissection is carried out to expose the medial border of scapula. Fatty transformation and severe atrophy are noted on periscapular musculature depending on their involvement. The arm is placed in 90° of abduction and full external rotation, with a bulky support beneath the shoulder. This position brings the medial border of the scapula into the surgical field to allow easier visualization. The transition between subscapularis, medial border and infraspinatus is usually easy to identify. The medial border of the scapula is exposed and medial 3 cm part of infraspinatus and supraspinatus muscles are subperiosteally elevated from scapula. Now the arm is positioned in adduction and internal rotation, and scapula is elevated from the chest wall using two towel clamps. Tilting the table also helps visualizing underneath of the scapula and subscapularis. A 5 cm part of subscapularis is detached from the ventral surface of the scapula and a 3 cm wide strip is excised which usually corresponds to medial one-third to one-half of the muscle belly. Serratus anterior is detached from the inferior angle to help mobilization. The next step is to prepare the ribs that will be used for fixation. At this step, a preliminary simulation of the arthrodesis position helps in selecting the most distal rib that would be included in fusion. Depending on the size of the scapula, the sixth or seventh rib would commonly be the lowest fixation point, with the second rib being the highest one always. All the ribs are prepared in a standard fashion. The erector spinae muscle is retracted medially to expose the prominent medial angle of the ribs. This is the most medial part that requires decortication and marks the contact point with the undersurface of scapula and the ideal application point for the multifilament cable. Superficial part of intercostal muscles and periosteum is removed. It is very easy to penetrate the outer layer of the pleura with a sharp tipped cable which may cause pneumothorax. Therefore, we initially pass a 6 cm long soft feeding tube as a safety

**Table 1** A summary of the proposed staging system. Key points and surgical considerations of each stage are listed on the right side.

Stage	Elevation	Deltoid function	Scapular winging	Key points	Surgical consideration
0	Full	Full	None	Normal shoulder function with FSHD diagnosis	Contraindicated. Absolute loss of function without any benefit
1	Above 120	Full	Mild	Near normal function with slight functional limitation	Has only cosmetic benefit. May cause loss of elevation. Preferably STA is avoided
2	90–120	Full	Mild/severe	Mild functional limitation	Provides slight functional benefit but mostly cosmetic. Can be considered as a contralateral side surgery for a symmetrical body image
3	Below 90	Full	Severe	Marked functional limitation and winging. Deltoid is functional	Surgery provides clear functional as well as cosmetic benefits. Best candidates for STA
4	Below 90	Partial	Mild	Deltoid function is affected. Less winging than stage 3	Surgery provides limited functional benefit. Patient expectations and lifestyle should be considered before surgery. Can be considered as a contralateral side surgery
5	Below 30	None	None	Loss of deltoid, no winging	Contraindicated. No functional benefits



**Figure 4**  
(A) Preoperative appearance; (B) postoperative ‘squared’ shoulder.

guide, after tunnelling underneath the rib using curved dissectors and a right-angle clamp (Fig. 5A). Note that the neurovascular bundle lays in a sulcus inferior to the rib covered with a thin fascia. The cable is passed gently through the feeding tube and the tube is reused to bind together both ends of the cable until fixation. After passing all the cables, a test is crucial to check a possible pleural penetration. The surgical area is filled with saline, and the thoracic wall is entirely submerged (Fig. 5B). Patient is ventilated manually with a higher pressure than normal. If there is a penetration, bubbles are observed at the injury level, which indicates a thorax tube insertion at the end of surgery. The next step is preparation of bony surfaces with slight decortication to facilitate union. A ball tip high-speed burr is used to prepare the ventral surface of scapula and dorsal surfaces of selected adjacent ribs. Note that both scapula and ribs are fragile with thin cortices. Therefore, creating a rough surface instead of a complete decortication is recommended. The final step is determining the cable passage points for the scapula. The arm is abducted to the final fixation position. The aim is to achieve at least 15° of external rotation of the medial border to the vertebral axis, but the final position is dictated by the best fit contact between ribs and scapula. Consider that the scapula bends slightly to accommodate the thoracic curvature following tensioning. If the patient had a previous arthrodesis on the contralateral side, care must be taken to avoid asymmetry. Another important tip is to select passing points close to the medial border of scapula. A cable inserted more than 10 mm from the medial border, on the central part of the scapula, would cause a fracture since the structural support is weaker. After marking the passage points, we utilize a sharp tipped high-speed burr to create holes, but a drill can also

**Table 2** A summary of the techniques used in the literature, including fused ribs and utilization of graft.

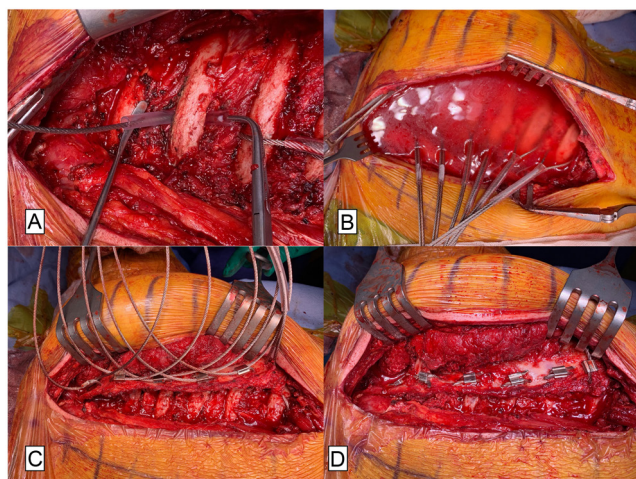
Study	Surgical method	Ribs used in fixation	Usage of grafts
Eren <i>et al.</i> (19)	Multifilament cables	5–6 (2–7 or 2–6)	Posterior iliac crest cancellous and allograft
Alshameeri <i>et al.</i> (20)	Plate and cerclage wires	5	Femoral head allograft
Andrews <i>et al.</i> (21)	Cable (as described by Bunch and Siegel)	3 (4, 5 and 6)	Iliac crest
Berne <i>et al.</i> (22)	Plate and cerclage wires: lower two ribs are fixed with a two-hole plate and steel wire. Most superior rib is osteotomized and passed through a tunnel	3 (4, 5 and 6)	None
Boileau <i>et al.</i> (23)	Plate+cerclage wire (1, 5 or 1,8 mm)	4 (3, 4, 5 and 6)	Iliac crest
Bunch <i>et al.</i> (24)	Cerclage wires	3–5 (4, 5, and 6)	Iliac crest cortical and cancellous
Cooney <i>et al.</i> (25)	Plate and cerclage wires	4–5 (3–8)	Fresh-frozen femoral head
Copeland <i>et al.</i> (26)	Screw fixation	3 (4, 5 and 6)	Iliac crest cancellous
Diab <i>et al.</i> (27)	16G cable, plate or screw with washer	5 (3–7 or 2–6)	Iliac crest cancellous
Jakab <i>et al.</i> (28)	Cerclage wire	5 (3–7)	Iliac crest
Le Hanneur <i>et al.</i> (29)	Same as Letournel’s technique	3	None (except for a revision case, iliac crest)
Letournel <i>et al.</i> (30)	The fourth rib is osteotomized, passed through a tunnel on the scapula and then plated. Two most inferior ribs are fixated with cerclage wires	3 (4–6 or 5–7)	None
Rhee <i>et al.</i> (31)	18G cerclage wire (two cases with Letournel’s technique)	3	Iliac crest (opposite)
Twyman <i>et al.</i> (32)	18G cerclage wire	5 (2–6)	Iliac crest
Van Tongel <i>et al.</i> (33)	Screw with washer	3 (4, 5 and 6)	Iliac crest or β-tricalcium phosphate

be used. The second cable from the top (third rib) usually falls to the spine of the scapula. After passing cables and crimps, the prepared graft is placed in the arthrodesis site, and cables are tightened sequentially starting from the most inferior one (Fig. 5C and D). Roughly 30–60 mL of cancellous bone graft is required. If the amount of harvested autograft is not adequate, allograft can be included (Fig. 6).

Multifilament metal cables are reliable implants, but prominent crimps that are used to secure loops may cause skin irritation and may require removal. Recently, a high-strength suture fixation method has also been proposed (58). Currently, there is no study on the efficacy of this fixation method, and it is a matter of debate if they preserve the initial compression or not. In our current practice, we utilize high-strength tape sutures (Suture Tape Cerclage System, Arthrex, FL, USA) in the most superior (second rib) and most inferior (sixth or seventh rib) fixation points to avoid irritation as these were the most frequent cause of discomfort (Fig. 6). Future studies would provide an insight if metal-free fixation would be a reliable option.

#### Anaesthetic considerations and postoperative analgesia

In our institution, we use the erector spinae plane block (ESPB) with continuous plane infusion, a recent technique described by Forero *et al.* for providing postoperative analgesia (59). An ESPB catheter (or an epidural catheter) is placed by the surgeon, deep to the erector spinae muscle, which is used with a patient-controlled analgesia device. This catheter provides analgesia over a broad region and diminishes the negative effects of pain in the postoperative period, which is not only required for comfort but also important to avoid pulmonary complications.



**Figure 5**

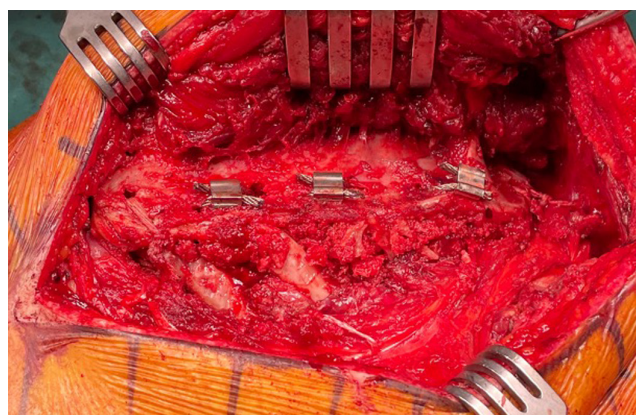
(A) Passing of the protective feeding tubes; (B) saline submersion test; (C and D) application of the cables.

Anaesthesia is maintained with total i.v. anaesthesia with propofol and remifentanyl infusions at suitable lowest doses. Although there is no known increased risk for malignant hyperthermia (60), it is prudent to have dantrolene in the armamentarium. Remifentanyl is the preferred opioid as it has a shorter half-life. In addition to the routine close monitoring process, patients are additionally monitored throughout the procedure using Bispectral Index Monitoring System (Medtronic, Dublin, Ireland). Given the increased prevalence of arrhythmias in this patient population (61), it is prudent to not use any arrhythmogenic agents such as desflurane. A list of anaesthetic drugs that are rather safe in muscular dystrophy patients has been published (62).

The impact of surgery on respiratory function has not been clearly described. In our previous series, we have not observed a negative impact on the respiratory function of patients (19). Minor pulmonary complications such as effusion or atelectasis are often associated with painful chest motion and are often managed with respiratory physiotherapy and rarely require intervention (i.e. chest tube) (19). Therefore, an effective analgesia strategy in STA is necessary. Respiratory problems may also be exacerbated by the use of opioid analgesics in the postoperative period. This block allows us to provide effective analgesia with the minimal usage of opioids. Patients are given incentive spirometers immediately after the operation, and effective analgesia allows a better rehabilitation period for respiratory function.

#### Postoperative period and rehabilitation

Although there is a consensus on the requirement of an immobilization period, the method and the duration vary in the literature. Historically, a cast or similar braces were utilized to provide an absolute protection for



**Figure 6**

Application of high-strength tape sutures in the most superior and most inferior fixation points.

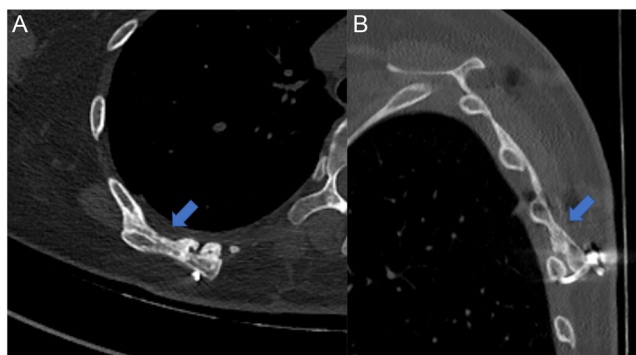


scapula (21, 22, 63). More recent publications support the use of an arm sling, from 6 weeks (20, 23, 29, 32) to 3 months (21, 22, 25, 26). We prefer to immobilize the shoulder in a sling, in 30° of abduction for 10 weeks. The patients are allowed to perform tabletop activities immediately after the surgery. It should be kept in mind that these patients have already lost their overall strength at a critical level due to their muscular dystrophies, and a prolonged immobilization might cause further detrimental functional loss.

After the tenth week, a CT scan is performed to confirm callus formation in all fixation levels (Fig. 7). Passive, active assisted and active ROM and isometric strengthening exercises are introduced gradually. At 6 months postoperatively, isokinetic strengthening and further stretching exercises are allowed. Particular attention is necessary for anterior deltoid strengthening and posterior capsular stretching. As mentioned before, due to the preoperative resting position of the scapula, the posterior capsule is tight, and the anterior deltoid is less utilized during arm movement. After STA, posterior capsular tightness becomes a limiting factor of adduction, particularly in a flexed glenohumeral joint. In addition to posterior tightness, anterior deltoid weakness also contributes to limitation of forward flexion in adduction. Patients often complain that they are unable to reach the contralateral shoulder and arm moving to the side when they try to reach the front. Therefore, posterior capsular stretching and strengthening of the relatively weaker anterior deltoid are key components of rehabilitation.

### Clinical and functional results

In our series which was published recently, bony fusion was achieved in 98% (63 out of 64 shoulders) of the cases in a mean follow-up of 71.2 months (range, 12–185 months). They observed major improvement in abduction (from  $52.7^\circ \pm 15.8^\circ$  to  $98.8^\circ \pm 20.3^\circ$ ;  $P < 0.001$ ), elevation



**Figure 7**  
Postoperative tenth week CT scan showing callus formation.

( $60.6^\circ \pm 17.2^\circ$  to  $123.7^\circ \pm 26.7^\circ$ ;  $P < 0.001$ ) and with quick disabilities of the arm, shoulder and hand (qDASH) scores (from  $34.7 \pm 11.4$  to  $13.3 \pm 13.1$ ;  $P < 0.001$ ). Only two patients reported a lower postoperative qDASH score. There were seven major pulmonary complications, five of which were treated with chest tubes. Scapular complications were seen in 10 patients. Failure of fixation was seen in three patients due to rib fractures, and two of them needed revision surgery. Brachial plexus palsy was observed in one patient. Additionally, one patient required reoperation due to scapular fracture, two due to non-union and one due to delayed union. Only two needed implant removal due to implant irritation (19).

Published functional results are summarized in Table 3. Reportedly, a preoperative elevation of 56.5–75°, improved to 81–120° after STA (Fig. 8). There is a lack of standardization in reporting the amount of thoracohumeral motion in the literature (terms elevation, flexion and abduction were used interchangeably), but it is possible to interpret these results as a combined elevation. Several authors also reported improvement in clinical scores as well. DASH and the Constant scores tend to improve significantly following STA (23, 25, 33, 53). Boileau *et al.* (23) reported a mean increase of 37 points in Constant scores as well as a mean 17 point of improvement in University of California Los Angeles (UCLA) scores. Cooney *et al.* (25) also described a mean improvement of 14 points in DASH scores. We also documented a mean improvement of 21 points in DASH scores. Visual analog scale (VAS) pain scores also tend to decrease (23, 31), following STA; however, Van Tongel *et al.* (33) reported an increase in the pain scores after the surgery. Despite all efforts in quantifying the change in the QoL following STA, none of these results are completely reliable since the implemented scores were constructed for different upper extremity problems. Constant and UCLA are shoulder-specific outcome scores; nevertheless, strength test in the objective part of Constant score would be affected directly from shoulder position and muscular dystrophy itself. On the other hand, neuromuscular QoL scores do not focus on the shoulder function and take the whole-body impairment into account. We believe DASH score would reflect the upper extremity disability better than others, but it still has similar drawbacks. There is clearly a need for an STA-specific QoL outcome score.

### Complications

The problems associated with STA can be classified into two main anatomic (pulmonary and scapular) complication groups and an additional minor problem group (Table 4). Atelectasis and pulmonary effusion are



**Table 3** A summary of clinical improvement in terms of range of motion and outcome scores.

Reference	Follow-up (months)	Preoperative elevation	Preop clinical score	Postoperative elevation	Postoperative clinical score
Eren <i>et al.</i> (19)	78	60 (50–90)	DASH: 34.7 ± 11.4	120 (100–150)	DASH: 13.3 ± 13.1
Alshameeri <i>et al.</i> (20)	17.4	59.3 ± 6.8 (45–70)		97.6 ± 9.6 (90–120)	
Andrews <i>et al.</i> (21)	73.2	56.5 ± 17.5 (45–85)		111 ± 11.7 (85–120)	
Berne <i>et al.</i> (22)	102	70 (40–90)		81 (0–130)	
Boileau <i>et al.</i> (23)	141	62 ± 20 (20–80)	Constant: 26 ± 6 UCLA: 10 ± 3 SSV: 25 ± 8 VAS: 6 ± 3	102 ± 4 (100–110)	Constant: 63 ± 3 UCLA: 27 ± 2 SSV: 62 ± 18 VAS: 1 ± 1
Bunch <i>et al.</i> (24)		63.6 ± 18.5 (10–90)		114.6 ± 34.3 (25–160)	
Cooney <i>et al.</i> (25)	29	72 (30–90)	DASH: 48 (27–74)	117 (90–130)	DASH: 34 (0.8–70)
Copeland <i>et al.</i> (26)	170.8			110.5 ± 27.7 (85–150)	Constant: 57.9 ± 26.3 (18–90)
Diab <i>et al.</i> (27)	75	75 (70–90)		105.9 (20–150)	
Jakab <i>et al.</i> (28)	35	85 ± 5.8 (80–90)		116.3 ± 11.1 (100–125)	
Le Hanneur <i>et al.</i> (29)	168	68.6 ± 19.5 (30–90)		104.3 ± 45.8 (40–160)	Constant: 57 ± 25
Letournel <i>et al.</i> (30)	68.7	76.6 ± 7.7 (60–90)		107.8 ± 10.8 (90–130)	
Rhee <i>et al.</i> (31)	102	76 (60–90)	Pain: 2.5 (0–4) UCLA: 18.4 (0–3)	109 (65–135)	Pain: 0.5 UCLA: 27.9
Twyman <i>et al.</i> (32)	48.5	63 ± 18.4 (30–80)		96 ± 8.1 (60–110)	
Van Tongel <i>et al.</i> (33)	88	65	Constant: 30 (17–41) Pain: 9.8 (3–15)	119	Constant: 61 (30–90) Pain: 13.2 (8–15)

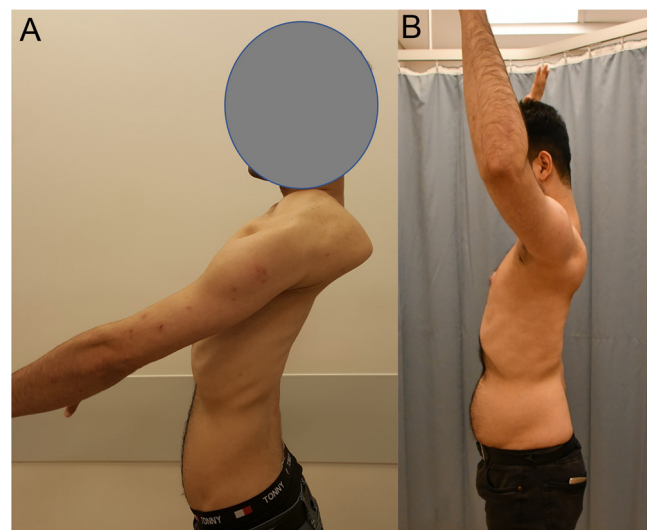
common following surgery. Effusion causes chest wall pain, which results in shallow breathing, thus impeding respiration. Additionally, the hardware may also irritate the pleura which also amplifies the negative effects on respiratory function. For asymptomatic patients with normal SpO<sub>2</sub>, close clinical follow-up, pulmonary rehabilitation and active usage of incentive spirometers are usually sufficient for spontaneous recovery. However, if the patient becomes symptomatic and a progressive decrease in SpO<sub>2</sub> is observed, a chest tube is indicated. Pneumothorax and haemothorax usually indicate pleural penetration during surgery and can be diagnosed early with the ‘saline submerge test’ intraoperatively. When diagnosed intraoperatively or postoperatively, chest tube is usually utilized. All these pulmonary complications fully recover with careful follow-up and appropriate management. We suggest daily chest x-rays in the first several days and before discharge. We have not had late pulmonary complications after discharge, and it has not been reported in the literature as well.

Scapular complications are associated with the fixation and may be encountered in the early or late postoperative period. Early complications include rib and scapula fractures, which are usually related to overtensioning of the implant. The incidence of these two complications decreases as the performing surgeon becomes more proficient in the procedure. Perforating the scapular holes close to the medial border, avoiding overtensioning and avoiding overdecortication with high-speed burr are important tips to prevent intraoperative undesirable events.

A scapula fracture can occur in the late period as a mode of overuse injury. Implant irritation is relatively common, particularly on the second rib, and may require hardware removal after bony fusion is achieved.

Non-union is another possible complication following STA. Lack of compliance with the immobilization period, strenuous activities in the early period, tobacco consumption and advanced age are possible causes of this problem. Interestingly, non-union or scapula fracture does not always affect functional outcomes. In our series, we have observed patients with only minor discomfort without apparent functional loss or completely asymptomatic patients despite these complications. A possible reason was thought to be the fibrous soft tissue connection preventing complete dissociation.

Brachial plexus palsy is a rare but most devastating complication of STA, and the exact mechanism is yet to be



**Figure 8** (A) Preoperative elevation; (B) postoperative elevation.

**Table 4** Reported complications in the literature.

Reference	Total*	Scapular complications						Pulmonary complications					ReOp	
		W	Irr	IF	SFx	RFx	BPP	NU	Ptx	Pnm	Htx	Eff		Atc
Alshameeri <i>et al.</i> (20)	4 (44%)				1						2			3
Andrews <i>et al.</i> (21)	4 (40%)					1			1	1		1	1	1
Berne <i>et al.</i> (22)	19 (38%)		1		4	5	2		5			4	1	2
Boileau <i>et al.</i> (23)	4 (40%)		1			1		1	1			1		0
Bunch <i>et al.</i> (24)	1 (6%)							1						0
Cooney <i>et al.</i> (25)	10 (71%)		1					1	1		1	1		6
Copeland <i>et al.</i> (26)	9 (64%)		2		1			1			1	2		3
Diab <i>et al.</i> (27)	2 (18%)													2
Eren <i>et al.</i> (19)	16 (23%)		3		1	4	1	3	4			2	1	5
Jakab <i>et al.</i> (28)	0													0
Le Hanneur <i>et al.</i> (29)	7 (87.5%)			4	2			1						3
Letourmel <i>et al.</i> (30)	9 (56.3%)		2			3			3			1	1	0
Rhee <i>et al.</i> (31)	1 (11%)											1		0
Twyman <i>et al.</i> (32)	7 (58%)		1	3		1	1	1	1					1
Van Tongel <i>et al.</i> (33)	8 (25%)	1		2		1		3	1					6
Wolfe <i>et al.</i> (64)	2							2						2
Total														

\*Number of complications reported and percentage of number of scapula.

Atc, atelectasis; BPP, brachial plexus palsy; Eff, pleural effusion; Htx, haemothorax; IF, implant failure; Irr, implant irritation; NU, non-union; Pnm, pneumonia; Ptx, pneumothorax; ReOp, reoperation; SFx, scapular fracture; W, wound dehiscence or superficial infection.

determined. There are no documented risk factors, which makes it more difficult to avoid this complication. Single-bundle and even pan-plexus injuries have been reported to occur (19, 47, 48). The mode of injury might be prone position, traction or compression by intraoperative manoeuvres or due to the correction itself. The rarity of the complication and heterogeneity of the reported cases prevent us to understand the exact causality and the location of this injury presently. Several authors proposed prophylactic or therapeutic mid-clavicle osteotomy, which was adapted from the correction method addressing Sprengel’s deformity. However, there are major differences between pathomechanics of these interventions, and an osteotomy cannot be generalized to FSHD patients. It may be prudent to stay on the conservative treatment side, except when a possible mechanical injury exists. In our series, we had one plexopathy which resolved with partial ulnar nerve motor paralysis after 18 months of follow-up. Cooney *et al.* also reported a case which recovered with intrinsic muscle weakness; additionally, Twyman *et al.* described a patient with upper trunk lesion who had a good functional outcome at 6 years postoperatively.

### Conclusion

FSHD results in marked disability due to the stabilization failure of the scapula as an outcome of the periscapular muscle wasting. The slow progressing nature of the disease and selective muscle involvement renders STA an effective solution to provide a stable scapula to allow better shoulder motion. Various surgical techniques were proposed which appear to be successful according to the

literature. However, the key component of success is not the technique but the patient selection. At this point, a multidisciplinary approach including a team specialized in neuromuscular diseases and geneticists with an access to a well-equipped lab will help selecting the right patient for the surgery. In the near future, a cure addressing the muscle wasting mechanisms can be expected, but until then, STA will help patients with disabilities due to scapular dysfunction.

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The authors declare that there is no conflict of interest that could be perceived as prejudicing the impartiality of the research reported.

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