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.

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).

FSHD is inherited in an autosomal dominant pattern, and the clinical severity and progression rate is highly variable. There are two well-defined subtypes, FSHD1 and FSHD2. 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 FSHD1 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 FSHD2, 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 FSHD 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 FSHD, 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 dyskinesis (37). Recently, Basem el Hassan redefined the spectrum of scapular dysfunction, introducing the term scapulothoracic abnormal motion (38). The scapular winging pattern in FSHD 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 scapula, 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.
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 forestated, 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.
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. Spongious 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             | Detoid 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 deltoide, no winging | Contraindicated. No functional benefits |

Figure 3
(A) Stabilization of the scapula mimics postoperative range of motion; (B) without stabilization.
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

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

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

Anaesthesia is maintained with total i.v. anaesthesia with propofol and remifentanil 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. Remifentanil 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° ± 15.8° to 98.8° ± 20.3°; P < 0.001), elevation (60.6° ± 17.2° to 123.7° ± 26.7°; P < 0.001) and with quick disabilities of the arm, shoulder and hand (qDASH) scores (from 34.7 ± 11.4 to 13.3 ± 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
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$_2$, 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$_2$ is observed, a chest tube is indicated.

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

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

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

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 et al. (20) | 4 (44%) | 1 | 1 | 1 | 5 | 1 | 1 | 1 | 1 | 2 | 4 | 1 | 2 | 3 |
| Andrews et al. (21)   | 4 (40%) | 1 | 1 | 1 | 2 | 1 | 1 | 1 | 1 | 1 | 1 | 1 | 1 | 1 |
| Berne et al. (22)     | 19 (38%) | 4 | 1 | 5 | 2 | 1 | 1 | 1 | 1 | 4 | 2 | 1 | 1 | 2 |
| Boileau et al. (23)   | 4 (40%) | 1 | 1 | 1 | 1 | 1 | 1 | 1 | 1 | 1 | 1 | 1 | 1 | 1 |
| Bunch et al. (24)     | 1 (6%)  | 1 | 1 | 1 | 1 | 1 | 1 | 1 | 1 | 1 | 1 | 1 | 1 | 1 |
| Cooney et al. (25)    | 10 (71%) | 1 | 1 | 1 | 1 | 1 | 1 | 1 | 1 | 1 | 1 | 1 | 1 | 1 |
| Copeland et al. (26)  | 9 (64%)  | 1 | 1 | 1 | 1 | 1 | 1 | 1 | 1 | 1 | 1 | 1 | 1 | 1 |
| Diab et al. (27)      | 2 (18%)  | 1 | 1 | 1 | 1 | 1 | 1 | 1 | 1 | 1 | 1 | 1 | 1 | 1 |
| Eren et al. (19)      | 16 (23%) | 3 | 1 | 4 | 1 | 3 | 4 | 2 | 1 | 5 | 0 | 0 | 0 | 0 |
| Jakab et al. (28)     | 0                   | 0 | 0 | 0 | 0 | 0 | 0 | 0 | 0 | 0 | 0 | 0 | 0 | 0 |
| Le Hanneur et al. (29)| 7 (87.5%) | 4 | 2 | 1 | 1 | 1 | 1 | 1 | 1 | 1 | 1 | 1 | 1 | 1 |
| Letournel et al. (30) | 9 (56.3%) | 2 | 3 | 3 | 3 | 3 | 3 | 3 | 3 | 3 | 3 | 3 | 3 | 3 |
| Rhee et al. (31)      | 1 (11%)  | 1 | 1 | 1 | 1 | 1 | 1 | 1 | 1 | 1 | 1 | 1 | 1 | 1 |
| Twyman et al. (32)    | 7 (58%)  | 1 | 3 | 1 | 1 | 1 | 1 | 1 | 1 | 1 | 1 | 1 | 1 | 1 |
| Van Tongel et al. (33)| 8 (25%)  | 1 | 2 | 1 | 1 | 1 | 1 | 1 | 1 | 1 | 1 | 1 | 1 | 1 |
| Wolfe et al. (64)     | 2                   | 2 | 2 | 2 | 2 | 2 | 2 | 2 | 2 | 2 | 2 | 2 | 2 | 2 |

*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.

ICMJE Conflict of Interest Statement
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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References
1. Statland JM & Tawil R. Facioscapulohumeral muscular dystrophy. Continuum (Minneapolis, Minn) 2016 22 1916–1931. (https://doi.org/10.1212/CON.0000000000002399)
2. Tawil R & Van Der Maarel SM. Facioscapulohumeral muscular dystrophy. Muscle and Nerve 2006 34 1–15. (https://doi.org/10.1002/mus.20522)
3. Gerevini S, Scarlato M, Maggi L, Cava M, Caliendo G, Pasanisi B, Falini A, Previtali SC & Morandi L. Muscle MRI findings in facioscapulohumeral muscular dystrophy and their relationship with the clinical course in patients affected by facioscapulohumeral muscular dystrophy. J Neurol Neurosurg Psychiatry 2012 83 556–561. (https://doi.org/10.1136/jnnp.2011.237910)
dystrophy. European Radiology 2016 26 693–705. (https://doi.org/10.1007/s00330-015-3890-1)

4. Goseling RJM, Voermans NC, Okkersen K, Brouwer OF, Padberg GW, Nikolic A, Tugler R, Dorobek M, Mah JK, van Engelen BGM et al. Early onset facioscapulohumeral dystrophy - a systematic review using individual patient data. Neuro muscular Disorders 2017 27 1077–1083. (https://doi.org/10.1016/j.nmd.2017.09.007)

5. Padberg GW, Frants RR, Brouwer OF, Wijmenga C, Bakker E & Sandkuijl LA. Facioscapulohumeral muscular dystrophy in the Dutch population. Muscle and Nerve. Supplement 1995 2 S81–S84. (https://doi.org/10.1002/mus.880181315)

6. Loonen TGI, Horlings CGC, Vincenten SCC, Beurskens CHG, Knuijt S, Padberg GWAM, Statland JM, Voermans NC, Maal TJJ, van Engelen BGM et al. Characterizing the face in facioscapulohumeral muscular dystrophy. Journal of Neurology 2021 268 1342–1350. (https://doi.org/10.1007/s00415-020-10281-z)

7. Wang LH & Tawil R. Facioscapulohumeral dystrophy. Current Neurology and Neuroscience Reports 2016 16 66. (https://doi.org/10.1007/s11910-016-0667-0)

8. Deenen JC, Arnts H, van der Maarel SM, Padberg GW, Verschuuren JJ, Bakker E, Weineich SS, Verbeek AL & van Engelen BG. Population-based incidence and prevalence of facioscapulohumeral dystrophy. Neurology 2014 83 1056–1059. (https://doi.org/10.1212/00003086-199805010-00011)

9. Mostacciolo ML, Pastorell E, Vazza G, Miorin M, Angelini C, Tomelleri G, Galluzzi G & Trevisan CP. Facioscapulohumeral muscular dystrophy: epidemiological and molecular study in a north-east Italian population sample. Clinical Genetics 2009 75 550–555. (https://doi.org/10.1111/j.1399-0004.2009.01158.x)

10. Zatz M, Marie SK, Cerqueira A, Vainzof M, Pavanello RC & Passos Bueno MR. The facioscapulohumeral muscular dystrophy (FSHD1) gene affects males more severely and more frequently than females. American Journal of Medical Genetics 1998 77 155–161. (https://doi.org/10.1002/(SICI)1096-8628(19980501)77:2<155::AID-AJMG9>3.0.CO;2-R)

11. van der Maarel SM, deidda G, Lemmers RJ, van Overveld PG, van der Wielen M, Hewitt JE, Sandkuijl L, Bakker B, van Ommen GJ, Padberg GW et al. De novo facioscapulohumeral muscular dystrophy: frequent somatic mosaicism, sex-dependent phenotype, and the role of mitotic transchromosomal repeat interaction between chromosomes 4 and 10. American Journal of Human Genetics 2000 66 26–35. (https://doi.org/10.1086/302730)

12. Ehrlich M, Jackson K, Tsumagari K, Camaño P & Lemmers R. Hybridization analysis of O424 repeat arrays linked to FSHD. Chromosoma 2007 116 107–116. (https://doi.org/10.1007/s00412-006-0080-6)

13. Ricci G, Sciotti I, Sera F, Govi M, D’Amico R, Frambooli I, Mele F, Filosto M, Vercelli L, Ruggiero L et al. Large scale genotype-phenotype analyses indicate that novel prognostic tools are required for families with facioscapulohumeral muscular dystrophy. Brain 2013 136 3408–3417. (https://doi.org/10.1093/brain/aws226)

14. Nikolic A, Ricci G, Sera F, Bucci E, Govi M, Mele F, Rossi M, Ruggiero L, Vercelli L, Rovaglia S et al. Clinical expression of facioscapulohumeral muscular dystrophy in carriers of 1–3 D4Z4 reduced alleles: experience of the FSHD Italian National Registry. BMJ Open 2016 6 e007798. (https://doi.org/10.1136/bmjopen-2015-007798)

15. Brooke MH. A Clinician’s View of Neuromuscular Diseases. Baltimore, MD: Williams & Wilkins 1977.

16. Brouwer OF, Padberg GW, Wijmenga C & Frants RR. Facioscapulohumeral muscular dystrophy in early childhood. Archives of Neurology 1994 51 387–394. (https://doi.org/10.1001/archneur.1994.00531601080111)

17. Whitman A. Congenital elevation of scapula and paralysis of serratus Magnus muscle. JAMA 1933 99 1332–1334. (https://doi.org/10.1001/jama.1933.02740680028007)

18. Howard RC. Thoraco-scapular arthrodesis. Journal of Bone and Joint Surgery. 1961 43-B 175.

19. Eren I, Erjen A, Birsel O, Atalar AC, Oflazer P & Demirhan M. Functional outcomes and complications following scapulothoracic arthrodesis in patients with facioscapulohumeral dystrophy. Journal of Bone and Joint Surgery. American Volume 2020 102 237–244. (https://doi.org/10.2106/JBJS.19.00571)

20. Alshameeri ZA, Garg S & Wallace WA. Scapulothoracic fusion for facio-scapulo-thoracic dystrophy. Shoulder and Elbow 2011 3 56–61. (https://doi.org/10.1111/j.1758-5740.2010.00103.x)

21. Andrews CT, Taylor TC & Patterson VH. Scapulothoracic arthrodesis for patients with facioscapulohumeral muscular dystrophy. Neuromuscular Disorders 1998 8 580–584. (https://doi.org/10.1096/0960-8966(98)00081-9)

22. Berne D, Laude F, Laporte C, Fardeau M & Saillant G. Scapulothoracic arthrodesis in facioscapulohumeral muscular dystrophy. Clinical Orthopaedics and Related Research 2003 409 106–113. (https://doi.org/10.1097/01. blo.0000057970.10364.35)

23. Boileau P, Pison A, Wilson A, van der Meijden O, Sacconi S, Trojani C & Gauco MD. Bilateral scapulothoracic arthrodesis for facioscapulohumeral muscular dystrophy: function, fusion, and respiratory consequences. Journal of Shoulder and Elbow Surgery 2020 29 931–940. (https://doi.org/10.1016/j.jse.2019.10.006)

24. Bunch WH & Siegel IM. Scapulothoracic arthrodesis in facioscapulohumeral muscular dystrophy. Review of seventeen procedures with three to twenty-one-year follow-up. Journal of Bone and Joint Surgery. American Volume 1993 75 372–376. (https://doi.org/10.1099/0004623-199303000-00008)

25. Cooney AD, Gill I & Stuart PR. The outcome of scapulothoracic arthrodesis using cerclage wires, plates, and allograft for facioscapulohumeral dystrophy. Journal of Shoulder and Elbow Surgery 2014 23 e8–e13. (https://doi.org/10.1016/j.jse.2013.04.012)

26. Copeland SA, Levy O, Warner GC & Dohenhoff RM. The shoulder in patients with muscular dystrophy. Clinical Orthopaedics and Related Research 1999 368 80–91. (https://doi.org/10.1097/00004623-199911000-00010)

27. Diab M, Darras BT & Shapiro F. Scapulothoracic fusion for facioscapulohumeral muscular dystrophy. Journal of Bone and Joint Surgery. American Volume 2005 87 2267–2275. (https://doi.org/10.2106/JBJS.D.02952)

28. Jakab E & Gledhill RB. Simplified technique for scapulothoracic fusion in facioscapulohumeral dystrophy. Journal of Pediatric Orthopaedics 1993 13 749–751. (https://doi.org/10.1097/00124139-199311000-00011)

29. Le Hanneur M & Saint-Cast Y. Long-term results of Letournel scapulothoracic fusion in facioscapulohumeral muscular dystrophy: a retrospective study of eight cases. Orthopaedics and Traumatology, Surgery and Research 2017 103 421–425. (https://doi.org/10.1016/j.otsr.2016.12.012)

30. Letournel E, Fardeau M, Lytte JO, Serrault M & Gesselin RA. Scapulothoracic arthrodesis for patients who have facioscapulohumeral muscular dystrophy. Journal of Bone and Joint Surgery. American Volume 1990 72 78–84. (https://doi.org/10.2106/00004623-199007201-00013)
31. Rhee YG & Ha JH Long-term results of scapulothoracic arthrodesis of facioscapulohumeral muscular dystrophy. Journal of Shoulder and Elbow Surgery 2006 15 445–450. (https://doi.org/10.1016/j.jse.2005.10.015)

32. Twyman RS, Harper GD & Edgar MA Thoracoscopic fusion in facioscapulohumeral dystrophy: clinical review of a new surgical method. Journal of Shoulder and Elbow Surgery 1996 5 201–205. (https://doi.org/10.1016/s1058-2746(05)80006-o)

33. Van Tongel A, Atoun E, Narvani A, Sforza G, Copeland S & Levy O. Medium to long-term outcome of thoracoscopic arthrodesis with screw fixation for facioscapulohumeral muscular dystrophy. Journal of Bone and Joint Surgery. American Volume 2013 95 1404–1408. (https://doi.org/10.2106/JBJS.L.01097)

34. Ludewig PM & Cook TM Alterations in shoulder kinematics and associated muscle activity in people with symptoms of shoulder impingement. Physical Therapy 2000 80 276–291. (https://doi.org/10.1093/ptj/80.3.276)

35. De Baets L, Van Deun S, Monari D & Jaspers E. Three-dimensional kinematics of the scapula and trunk, and associated scapular muscle timing in individuals with stroke. Human Movement Science 2016 48 82–90. (https://doi.org/10.1016/j.humov.2016.04.009)

36. Ludewig PM, Hoff MS, Osowski EE, Meschke SA & Rundquist PJ. Relative balance of serratus anterior and upper trapezius muscle activity during push-up exercises. American Journal of Sports Medicine 2004 32 484–493. (https://doi.org/10.1177/0363546503258911)

37. Martin RM & Fish DE. Scapular winging: anatomical review, diagnosis, and treatments. Current Reviews in Musculoskeletal Medicine 2008 1 1–11. (https://doi.org/10.1007/s12178-007-9000-5)

38. Elhassan BT, Dang KH, Huynh TM, Harstadt C & Best MJ. Outcome of arthroscopic pectoralis minor release and scapuloplasty for the management of scapulothoracic abnormal motion. Journal of Shoulder and Elbow Surgery 2022 31 1208–1214. (https://doi.org/10.1016/j.jse.2021.10.046)

39. Flanigan K. Facioscapulohumeral muscular dystrophy and scapulonneral disorders. Myology 2004 2 1123–1133.

40. Lamperti C, Fabbri G, Vercelli L, D’Amico R, Frusciante R, Bonifazi E, Fiorillo C, Bosarto C, Cao M, Servida M et al. A standardized clinical evaluation of patients affected by facioscapulohumeral muscular dystrophy: the FSHD clinical score. Muscle and Nerve 2010 42 213–217. (https://doi.org/10.1002/mus.21671)

41. Tawil R. Facioscapulohumeral muscular dystrophy. Neurotherapeutics 2008 5 601–605. (https://doi.org/10.1016/j.nert.2008.07.005)

42. Ricci G, Ruggiero L, Vercelli L, Sera F, Nikolic A, Govi M, Mele F, Daolio J, Angelini C, Antonini G et al. A novel clinical tool to classify facioscapulohumeral muscular dystrophy phenotypes. Journal of Neurology 2016 263 1204–1214. (https://doi.org/10.1007/s00415-016-8123-2)

43. Zampatti S, Colantoni L, Straffella C, Galota RM, Caputo V, Campoli G, Pagliaroli G, Carboni S, Mela J, Peconi C et al. Facioscapulohumeral muscular dystrophy (FSHD) molecular diagnosis: from traditional technology to the NGS era. Neurogenetics 2019 20 57–64. (https://doi.org/10.1007/s10048-019-00575-4)

44. Schätzl T, Kaiser L & Deigner HP. Facioscapulohumeral muscular dystrophy: genetics, gene activation and downstream signalling with regard to recent therapeutic approaches: an update. Orphanet Journal of Rare Diseases 2021 16 129. (https://doi.org/10.1186/s13023-021-01760-1)

45. Tawil R, Figlewicz DA, Griggs RC & Weiffenbach B. Facioscapulohumeral dystrophy: a distinct regional myopathy with a novel molecular pathogenesis. FSH Consortium. Annals of Neurology 1998 43 279–282. (https://doi.org/10.1002/ana.410430303)

46. Personius KE, Pandya S, King WM, Tawil R, McDermott MP & Group TFD. Facioscapulohumeral dystrophy natural history study: standardization of testing procedures and reliability of measurements. The FSH DY Group. Physical Therapy 1994 74 253–263. (https://doi.org/10.1093/ptj/74.3.253)

47. Mackenzie WG, Riddle EC, Earley JI & Sawatzky BJ. A neuromuscular complication after scapulothoracic arthrodesis. Clinical Orthopaedics and Related Research 2003 408 157–161. (https://doi.org/10.1097/00003086-200303000-00019)

48. Bhatia S, Hsu AR, Harwood D, Toleikis JR, Mather RC, 3rd & Romeo AA. The value of somatosensory evoked potential monitoring during scapulothoracic arthrodesis: case report and review of literature. Journal of Shoulder and Elbow Surgery 2012 21 e14–e18. (https://doi.org/10.1016/j.jse.2011.12.003)

49. Hamel J & Tawil R. Facioscapulohumeral muscular dystrophy: update on pathogenesis and future treatments. Neurotherapeutics 2018 15 863–871. (https://doi.org/10.1007/s13311-018-00675-3)

50. Awerbuch GI, Ngo MR & Wishnow R. Browner’s sign and facioscapulohumeral dystrophy. Archives of Neurology 1990 47 1208–1209. (https://doi.org/10.1001/archneur.1990.005310066018)

51. Olsen DB, Gideon P, Jeppesen TD & Vissing J. Leg muscle involvement in facioscapulohumeral muscular dystrophy assessed by MRI. Journal of Neurology 2006 253 1437–1441. (https://doi.org/10.1007/s00415-006-0230-z)

52. Horwitz MT & Tocantins LM. Isolated paralysis of the serratus anterior (Magnus) muscle. Bone and Joint Surgery 1938 20 720–725.

53. Eren İ, Birsal Ö, Öztop Çakmak O, Aslanger A, Gürsöy Özdemir Y, Eraslan S, Kayserili H, Ofazer P & Demirhan M. A novel shoulder disability staging system for scapulothoracic arthrodesis in patients with facioscapulohumeral dystrophy. Orthopaedics and Traumatology, Surgery and Research 2020 106 701–707. (https://doi.org/10.1016/j.otsr.2020.03.002)

54. Struyf F, Nijs J, Mottram S, Roussel NA, Cools AM & Meeusen R. Clinical assessment of the scapula: a review of the literature. British Journal of Sports Medicine 2014 48 883–890. (https://doi.org/10.1136/bjsports-2012-091059)

55. Struyf F, Nijs J, Baeyens JP, Mottram S & Meeusen R. Scapular positioning and movement in unimpaired shoulders, shoulder impingement syndrome, and glenohumeral instability. Scandinavian Journal of Medicine and Science in Sports 2011 21 352–358. (https://doi.org/10.1111/j.1600-0838.2010.01274.x)

56. Sobus DC, Simonneau GG, Dietz KE, Levene JA, Grossman RE & Smith WB. The lennie test for measuring scapular position in healthy young adult females: a reliability and validity study. Journal of Orthopaedic and Sports Physical Therapy 1996 23 39–50. (https://doi.org/10.2519/jospt.1996.23.1.39)

57. Copeland SA & Howard RC. Thoracoscopic fusion for facioscapulohumeral dystrophy. Journal of Bone and Joint Surgery (British Volume) 1978 60-B 547–551. (https://doi.org/10.1302/0301-620X.60B4.7118075)

58. Scollan JP, Chuhtai M, Evans PJ & Styron JF. Scapulothoracic fusion using nonabsorbable suture fixation: surgical technique and review of the literature. JSES Reviews, Reports, and Techniques 2021 1 118–126. (https://doi.org/10.1016/j.jsr.2021.01.003)

59. Forero M, Adhikary SD, Lopez H, Tsui C & Chin KJ. The ector spinae plane block: a novel analgesic technique in thoracic neuropathic pain. Regional Anesthesia and Pain Medicine 2016 41 621–627. (https://doi.org/10.1097/AAP.0000000000000451)
60. Attarian S, Salort-Campana E, Nguyen K, Behin A & Andoni Urtizberea J. Recommendations for the management of facioscapulohumeral muscular dystrophy in 2011. Revue Neurologique 2012 168 910–918. (https://doi.org/10.1016/j.neurol.2011.11.008)

61. Trevisan CP, Pastorello E, Armani M, Angelini C, Nante G, Tomelleri G, Tonin P, Mongini T, Palmucci L, Galluzzi G et al. Facioscapulohumeral muscular dystrophy and occurrence of heart arrhythmia. European Neurology 2006 56 1–5. (https://doi.org/10.1159/000094248)

62. Trevisan CP, Accorsi A, Morandi LO, Mongini T, Savoia G, Gravino E, Angelini C & Tegazzin V. Undiagnosed myopathy before surgery and safe anaesthesia table. Acta Myologica 2013 32 100–105.

63. Bunch WH. Scapulo-thoracic fusion. Minnesota Medicine 1973 56 391–394.

64. Wolfe GI, Young PK, Nations SP, Burkhead WZ, McVey AL & Barohn RJ. Brachial plexopathy following thoracoscopic fusion in facioscapulohumeral muscular dystrophy. Neurology 2005 64 572–573. (https://doi.org/10.1212/01.WNL.0000150907.82191.05)