Thoracoscapular Fusion for Winging of the Scapula with Screw Fixation for Fascioscapulohumeral Dystrophy (Modified Copeland-Howard Procedure)

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Introduction

This article describes our technique of thoracoscapular fusion with screw fixation for treatment of winging of the scapula in patients with fascioscapulohumeral dystrophy. Winging of the scapula may be caused by dysfunction of the long thoracic nerve with a resultant anterior serratus muscle palsy and/or trapezius palsy due to spinal accessory nerve injury. This can be caused by a single traumatic episode; repetitive trauma; neuralgic amyotrophy; poliomyelitis; and the muscular dystrophies, particularly fascioscapulohumeral dystrophy. Thoracoscapular fusion is indicated when deltoid and the other glenohumeral motor units are intact but the scapula-stabilizing muscles are permanently nonfunctioning. The most common indication is fascioscapulohumeral dystrophy.

Fascioscapulohumeral dystrophy is one of the most common types of muscular dystrophy and was first described by Landouzy and Dejerine in 1884¹. Fascioscapulohumeral dystrophy is a genetic disorder and characterized by weakness in facial and shoulder girdle musculature². It is an autosomal dominant myopathy, although 10% to 30% of the cases arise from de novo mutation^{3,4}. Symptoms usually begin in childhood, and >90% of patients have some evidence of disease on examination by the age of twenty years. The course of the disease is slowly progressive, although many patients have long periods of relatively stable function³. Life expectancy is normal, but up to 15% of affected individuals become severely disabled and eventually must use a wheelchair. Fascioscapulohumeral dystrophy is currently untreatable⁵⁻¹⁰.

There are currently more than 10,000 children with all types of muscular dystrophy living in the United States, and approximately 350 per million are born with the disease in the United States¹¹.

Shoulder girdle weakness is the most constant feature of fascioscapulohumeral dystrophy and leads to scapular winging. The severity of the effect of the disease on the muscles of the shoulder varies among the different types of disease. The muscles commonly involved are the serratus anterior, trapezius, rhomboids, latissimus dorsi, and muscles around the eyes and mouth. The deltoid invariably is spared. Fascioscapulohumeral dystrophy affects tasks of repetitive use requiring abduction and flexion of the shoulder. Even during such simple daily activities as overhead dressing, the muscles fatigue quickly and are unable to hold the arm in sustained abduction and elevation. Gross winging of the scapula can be a substantial cosmetic deformity (Fig. 1, Video 1). Female patients in particular regard this as their main reason for surgery. In addition, winging may pose problems with routine garment wear. Shoulder straps on clothes tend to slip off the shoulder and down the arm.

When shoulder elevation is attempted through use of the deltoid, the scapula rotates and lifts off the chest wall, losing strength at the glenohumeral joint (Fig. 2). This prevents the person from sustaining shoulder abduction or flexion¹². Patients are unable to hold heavy objects or perform overhead activities such as combing hair or shaving. Patients may also dislike the cosmetic deformity, and the shoulder girdle can be painful¹².

Mechanical fixation of the scapula to the thoracic wall provides a stable fulcrum on which the deltoid can exert its powerful action on the humerus and abduct the arm without rotation of the scapula¹. The procedure is described in the following steps.

Step 1: Preoperative Evaluation

Perform the Horwitz test.

- To determine whether the patient might benefit from a thoracoscaphular fusion, perform the scapula stabilizing test (Horwitz test)^{1,13,14}, which is a relatively simple clinical examination. Ask the patient to raise both arms forward and to maintain the arms in this horizontal position until fatigue causes them to drop to the side.
- Have the patient repeat the test while you manually hold the scapula downward and against the chest wall to prevent winging. If this greatly improves the patient's ability to maintain flexion or abduction and the fatigue test is substantially prolonged, then thoracoscaphular fusion might be indicated (Figs. 3-A and 3-B, Video 2).

Step 2: Position the Patient and Mark the Skin for the Operation

With the patient on a Montreal mattress, position the arms in 90° to 110° of elevation in the scapular plane and approximately 90° of external rotation and mark the skin.

- Perform the operation with the patient under general anesthesia and prone on a Montreal mattress mounted on the operating table. A Montreal mattress is a rectangular mattress with a central hollow that helps to prevent compression of the abdomen during respiration and avoids interference with the venous return. It has a gutter at the head end to ensure mild flexion of the neck, which assists airway maintenance (Figs. 4-A and 4-B).
- Position the arms in 90° to 110° of elevation in the scapular plane and approximately 90° of external rotation on a board (Fig. 5). This position brings the scapula into the ideal position for fixation to the ribs with the ventral border of the scapula lying parallel to the ribs. In this position, the fixed scapula will allow the best range of movement of the shoulder.
- Drape the upper extremity free to allow easy intraoperative handling. Also drape the ipsilateral posterior iliac crest free if bone-graft harvesting is planned.
- Mark the line of the spinous processes of the thoracic vertebrae, the medial border of the scapula and the scapular spine with a marker pen. Mark the incision line 5 to 7 cm lateral to the spinal processes (Fig. 5).

Step 3: Surgical Approach

Make an incision along the medial scapular edge, incise the trapezius muscle, and detach the levator scapula, rhomboid major, and rhomboid minor muscles.

- Make a straight incision along the medial edge of the scapula, 5 to 7 cm lateral to the spinal processes (Fig. 5). The underlying muscles often are atrophied, but occasionally they are hypertrophic but nonfunctioning.
- Incise the trapezius muscle parallel to the skin incision.
- Detach the levator scapula, rhomboid major, and rhomboid minor muscles from the medial border of the scapula and retract them medially.

Step 4: Prepare the Scapula

Do not decorticate the scapula to avoid weakening it.

- Detach the supraspinatus, infraspinatus, and teres minor muscles subperiosteally from the medial border of the scapula to expose a 3-cm-wide strip on the medial side of the scapula (Fig. 6).
- Detach the subscapularis muscle subperiosteally from the medial 5 to 6 cm of the scapula and resect the impinging part of the muscle to allow good direct contact between the denuded scapula and the ribs. Do not decorticate the scapula as the scapular bone is very thin and weakening of the scapular bone should be avoided.

Step 5: Select and Prepare the Ribs

Expose three or four ribs subperiosteally, from their superior border to avoid the neurovascular bundle.

- Choose three or four ribs lying under the most convenient part of the scapula; these are usually the fourth, fifth, and sixth ribs. (The third or the seventh may be fixed if needed.) The first chosen rib (usually the fourth rib) is the rib nearest to the scapular spine as this is the location of the best and strongest bone on the medial part of the scapula (Fig. 7).
- Incise the periosteum in the line of the rib and then separate subperiosteally the periosteum and the parietal pleura on the deep surface of the rib (from the superior part of the rib to avoid the neurovascular bundle) with a small periosteal elevator (Figs. 7 and 8). Introduce and place a thin retractor (McDonald dissector) under the rib to prevent damage to the pleura.
- Remove any excessive intercostal muscles and soft tissue that may prevent direct contact be-

tween the ribs and the scapula. Gently roughen the bone and partly decorticate the superficial surface of the ribs to create “fish scale” with a ferret gouge or an osteotome. Do not fully decorticate the ribs to avoid weakening them. Roughen the superficial surface to promote osseous healing.

Step 6: Drill the Scapula and Ribs

Drill the ribs with a McDonald dissector underneath them to prevent damage to the pleura.

- Place the scapula in the correct position over the denuded ribs, drill the scapula with a 3.2-mm drill bit, and make a mark on the cortex of the underlying rib. Drill the underlying rib with a 3.2-mm drill bit, with a protective McDonald dissector in place underneath the rib to prevent damage to the pleura.
- Measure the rib and the scapular drill holes to determine the appropriate screw length. Over-drill the scapular drill holes with a 4.5-mm drill bit. Use a self-tapping cortical or cancellous screw with a washer, tightened loosely to fix the scapula.
- Insert the second and third screws, and occasionally the fourth, in the same manner as the first. It is best to mark the drilling point on the rib for the second hole with the first screw in place, drilling through the scapula and then partially drilling into the external cortex of the rib. Then unscrew the first screw and complete the drilling of the rib with the McDonald dissector in place to prevent damage to the pleura (Fig. 8). Repeat this for the third hole with the previous two screws in place so that the scapula is fixed during this procedure and has the best placement of the screws in the ribs (Fig. 9). Then unscrew the first two screws and complete the drilling of the rib with the McDonald dissector in place to prevent damage to the pleura (Fig. 8). We use cortical screws with washers. If the screw grip in the rib is not optimal, we use a cancellous screw. We tend to avoid using a second screw in the same rib as this increases the risk of rib fracture.

Step 7: Harvest and Introduce Bone Graft, or Use Bone-Graft Substitute, and Close

Pack bone chips or bone-graft substitute between the ribs and the deep surface of the scapula.

- Harvest cortical and cancellous bone grafts from the ipsilateral posterior iliac crest or use tricalcium phosphate (TCP) granules or paste

as a bone-graft substitute to eliminate donor site morbidity. In the past few years, we have preferred to use bone-graft substitute (Actifuse ABX; Baxter, Deerfield, Illinois).

- Loosen the screws between the ribs and the scapula (Fig. 10) and pack bone chips or bone-graft substitute between the ribs and the deep surface of the scapula (Fig. 11). Then tighten the screws until good fixation of the scapula to the ribs is achieved (Fig. 12).
- Close the wound in layers, consisting of trapezius to infraspinatus and supraspinatus, subcutis, and skin. You may inject local anesthetics for intercostal nerve blocks, which are safe when performed appropriately. Take care to avoid pneumothorax or hemothorax.

Step 8: Postoperative Immobilization and Rehabilitation

An adjustable brace with the arm in 60° of abduction and 30° of forward flexion is worn for three months.

- Take special care when turning the patient over from the operating table and adjusting the brace. Ensure that no stress is placed on the fusion site as the patient is turned over. As many people as possible should assist at this stage.
- Immobilize the shoulder in an adjustable brace with the arm in about 60° of abduction and about 30° of forward flexion so that the patient can reach his or her mouth by flexing the elbow (Fig. 13). Several companies manufacture similar braces.
- Postoperatively, the arm is held in this position with the adjustable shoulder brace for three months (Fig. 14). Encourage the patient to perform elbow and hand movements in the brace during this time.
- At three months after surgery, remove the brace. Advise the patient to use a pillow under the arm for about a week in order to bring the arm gradually down from the abducted position. The pillow is strapped to the chest with a body belt.
- The patient commences physiotherapy with passive motion followed by active exercises in elevation and rotation. Physiotherapy continues to restore full glenohumeral movement and strengthen the deltoid muscle.

Results

Between July 1997 and July 2010, a thoracoscapular fusion was performed in thirty-five shoulders of twenty-four patients with fascioscapulohumeral dystrophy¹⁵. Eleven patients had a bilateral procedure. In bilateral cases, the

dominant side was operated on first when the involvement was symmetric whereas the side with more severe involvement was given priority in asymmetric cases. The mean interval between the two operations was sixteen months (range, six to thirty-six months). The mean age at the time of thoracoscaphular arthrodesis in the twenty-four patients was twenty-six years (range, sixteen to sixty-two years).

Thirty-two shoulders in twenty-one patients (fourteen males and seven females) were available for long-term evaluation. These patients were followed for a minimum of twenty-four months (average, eighty-eight months; range, twenty-four to 174 months).

Active elevation range increased, on average, from 65° preoperatively to 119° postoperatively.

The Constant score¹⁶ is the most acceptable functional score for the shoulder, having been adopted by international, American, and European shoulder and elbow societies. It consists of subjective and objective measurements, and the total score is the sum of pain, activities of daily living, range of motion, and strength scores. In our study¹⁵, the total Constant score increased, on average, from 30 points (range, 17 to 41 points) preoperatively to 61 points (range, 30 to 90 points) postoperatively, whereas the Constant subscores increased from 9.8 points (range, 3 to 15 points) of 15 points preoperatively to 13.2 points (range, 8 to 15 points) postoperatively for pain, from 5.4 (range, 2 to 13) of 20 points to 14.6 points (range, 6 to 20 points) for activities of daily living, from 14.6 (range, 8 to 22) of 40 points to 25.6 points (range, 12 to 36 points) for range of motion, and from 0 of 25 points to 8 points (range, 0 to 20 points) for strength. There was a clear improvement in the ability to carry out daily and leisure activities as most of the patients went from having severe limitation to having moderate or less limitation. Our results are comparable with those in our previous study in 1999¹, in which the mean Constant score was 58 points (range, 13 to 90 points) postoperatively. This confirms that these results are reproducible.

The mean patient satisfaction score (Subjective Shoulder Value [SSV])¹⁷ improved from 1 (range, 0 to 4) of 10 points preoperatively to 8.4 points (range, 4 to 10 points) postoperatively (Fig. 15, Video 3).

We found no difference in the functional outcomes for the eight shoulders with the longest follow-up (more than ten years) and those for the twelve shoulders with shorter follow-up (between two and five years). The Constant score averaged 62.5 points (an improvement of 34.1 points from a preoperative score of 28.4 points) in the former group compared 59 points (an improvement of 28 points from a preoperative score of 31 points) in the latter. Thus, there was no obvious deterioration of function over time in these patients.

Radiographic evaluation of the thirty-two shoulders that underwent long-term evaluation showed clear callus formation and no signs of loosening of the screws

in all but two patients. Good union with callus formation was observed both after use of autologous bone graft and after use of bone-graft substitute (Figs. 16-A and 16-B).

Two patients showed early signs of nonunion with loosening of the screws. Both underwent revision surgery, resulting in solid fusion at the time of the latest follow-up.

Before the operation the scapulae not only winged but also were elevated on attempted flexion or abduction, giving an unsightly appearance of widening of the base of the neck. Although fixation of the scapula leaves a considerable scar, the patients were willing to accept this because of the general improvement in appearance. The position in which the scapulae are fixed allows a maximum range of active movements. This position is farther lateral than the natural position of the scapula and tends to produce a slightly “squared shoulder” appearance. However, the male patients liked this “Superman” appearance, and the female patients liked the fact that it kept their clothes’ straps and handbag shoulder straps from falling (Fig. 17).

One patient (one shoulder) had a pneumothorax, which was treated successfully with conservative means and resolved spontaneously.

What to Watch For

Indications

- Weakness of the arm and an inability to sustain the arm in the abducted or flexed position causing limitations such as an inability to reach items in high cupboards or obtaining books from high shelves.
- The scapula riding up in the neck and winging posteriorly, appearing unsightly and causing great difficulty with clothes.
- Shoulder straps of bags and clothes slipping off the shoulder and down the arm.
- A dragging sensation in the shoulder.
- Aching and pain after use of the arm. Pain is not usually a primary indication but there can be an important pain component in fascioscapulo-humeral dystrophy¹⁵.

Contraindications

- Weakness severely affecting the deltoid muscle as well.
- Inability or unwillingness to tolerate immobilization in a brace for three months.

Pitfalls & Challenges

- Prepare and measure the brace on the patient preoperatively. It is advisable to ask the patient to bring a T-shirt to be worn underneath the brace after surgery.

- Patient positioning and scapular position in the operating theater prior to incision and surgery are crucial to the success of thoracoscopic fusion.
- Expose the ribs from their superior border to avoid injury to the intercostal neurovascular bundle. Subperiosteal exposure is important to avoid pneumothorax and lung damage. Remaining subperiosteal, with the McDonald dissector very close to the rib bone, minimizes the risk of pneumothorax and pleural and lung damage. Place the McDonald dissector underneath the ribs to provide protection when drilling the ribs to avoid pleural or lung injury.
- Use one screw through the scapular spine, where the scapular bone is thickest and strongest.
- Using the sequence described in Step 6 allows the best placement of the screws in the ribs. Mark the drilling point on the second rib with the first screw fixed in situ in the first rib, unscrew the first screw, and complete the drilling with McDonald dissector providing protection under the rib. Mark the drill hole in the third rib with the two previous screws fixed in situ in the ribs, unscrew the screws, and complete the drilling with McDonald dissector under the rib for protection.
- At the end of the procedure, it is crucial to protect the fusion by taking special care when turning the patient from the operating table and applying and adjusting the brace. As many people as possible should assist at this stage.
- The whole procedure, including patient positioning, surgery, turning the patient from the operating table, and fixing the brace, takes approximately three hours. The procedure should be performed with care and should not be rushed.
- The difficulties presented by brace immobilization for three months should be clearly and repeatedly explained to the patients preoperatively, as their compliance is crucial for a successful outcome. Most of the complications in our patients occurred as a result of noncompliance.
- Complications include pneumothorax due to perforation of the pleura during drilling of the ribs or by a screw. Hence it is mandatory to obtain a chest radiograph immediately postoperatively. Other complications include hemothorax from injury to the intercostal vessels, wound infection, and rib stress fracture. The screws can pull out from the ribs or through the scapula, with loss of fixation, or a scapular fracture can occur if the patient fails to keep the brace on for three months or as a result of trauma. Nonunion or failure to fuse between the scapula and the ribs is another possible complication. In our series¹⁵

and in the one reported by Kiss¹⁸, patients with nonunion were commonly older men (in their thirties) who were obese.

Clinical Comments

- Mechanical fixation of the scapula to the ribs provides the deltoid muscle with a stable post from which to function, prevents scapular winging, and improves the function of the deltoid muscle in patients with fascioscapulohumeral dystrophy.
- Thoracoscopic fusion results in significant improvement in active shoulder elevation as well as the endurance when holding the arm elevated⁴. The active elevation in our series seems to be slightly better than that in some other series^{2,4,12,19-31}. We believe that this improved range of motion was due to our positioning of the scapula when it was fixed.
- Using our screw fixation technique reduces substantially the risk of lung injury and pneumothorax, as no wires need to be passed under the ribs. Furthermore, with the use of screws there is only a small drill hole in the rib; this decreases the risk of rib fracture compared with that associated with wires around the ribs, which may strangulate the blood supply to the rib and increase risk of fracture.
- Thoracoscopic arthrodesis can be achieved with use of screws, wires, or plates, with or without bone graft. We used screws with washers and posterior iliac bone graft or bone-graft substitute. In four cases, fixation failed in the early stage of the healing process. We attribute two cases to noncompliance with the postoperative regimen. There is no clear cause for the early failure in the other patients. Patients with failure have been found to be older men (in their thirties) with relative obesity^{15,18}, an important observation.
- Respiratory function is not substantially affected by thoracoscopic fusion. Only a minimal loss of vital capacity, no change in forced expiratory volume after unilateral fusion, and a minimal decrease of 0.12 L after bilateral fusion were shown previously^{14,20,23}.

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References

1. Copeland SA, Levy O, Warner GC, Dodenhoff RM. The shoulder in patients with muscular dystrophy. *Clin Orthop Relat Res*. 1999 Nov;368:80-91.
2. Demirhan M, Uysal O, Atalar AC, Kilicoglu O, Serdaroglu P. Scapulothoracic arthrodesis in facioscapulohumeral dystrophy with multifilament cable. *Clin Orthop Relat Res*. 2009 Aug;467(8):2090-7. Epub 2009 Mar 31.
3. Kissel JT. Facioscapulohumeral dystrophy. *Semin Neurol*. 1999;19(1):35-43.
4. Giannini S, Faldini C, Pagkrati S, Grandi G, Digennaro V, Luciani D, Merlini L. Fixation of winged scapula in facioscapulohumeral muscular dystrophy. *Clin Med Res*. 2007 Oct;5(3):155-62.
5. Carpenter, Karpati G. Pathology of skeletal muscle. New York: Churchill Livingstone; 1984. p. 507-10.
6. Chyatte SB, Vignos PJ Jr, Watkins M. Early muscular dystrophy: differential patterns of weakness in duchenne, limb-girdle and facioscapulohumeral types. *Arch Phys Med Rehabil*. 1966 Aug;47(8):499-503.
7. Johnson EW, Braddom R. Over-work weakness in facioscapulohumeral muscular dystrophy. *Arch Phys Med Rehabil*. 1971 Jul;52(7):333-6.
8. Vinken PJ, Bruyn GW. Fascio-scapulo-humeral dystrophy. In: *Handbook of clinical neurology*. Amsterdam: North-Holland Publishing Company; 1979. p 415-31.
9. Walton JN. Muscular dystrophy and its relation to the other myopathies. In: Adams RD, Eaton LE, Shy GM, editors. *Research Publications*. Vol 38. Association for Research in Nervous and Mental Disease. Baltimore: Williams and Wilkins; 1960.
10. Walton J, Gardner-Medwin D. Progressive muscular dystrophy and the myotomic disorders. In: Walton J, editor. *Disorders of voluntary muscle*. Edinburgh: Churchill Livingstone; 1981.
11. Siegel IM. Diagnoses, management, and orthopaedic treatment of muscular dystrophy. Chicago: Yearbook Medical; 1986.
12. Orrell RW, Copeland S, Rose MR. Scapular fixation in muscular dystrophy. *Cochrane Database Syst Rev*. 2010;(1):CD003278. Epub 2010 Jan 20.
13. Horwitz MT, Tocanitus LM. Isolated paralysis of the serratus anterior (magnus) muscle. *J Bone Joint Surg Am*. 1938;20:720-5.
14. Copeland SA, Howard RC. Thoracoscapular fusion for facioscapulohumeral dystrophy. *J Bone Joint Surg Br*. 1978 Nov;60(4):547-51.
15. Van Tongel A, Atoun E, Narvani A, Sforza G, Copeland S, Levy O. Medium to long-term outcome of thoracoscapular arthrodesis with screw fixation for facioscapulohumeral muscular dystrophy. *J Bone Joint Surg Am*. 2013 Aug 7;95(15):1404-8.
16. Constant CR, Murley AH. A clinical method of functional assessment of the shoulder. *Clin Orthop Relat Res*. 1987 Jan;(214):160-4.
17. Gilbert MK, Gerber C. Comparison of the subjective shoulder value and the Constant score. *J Shoulder Elbow Surg*. 2007 Nov-Dec;16(6):717-21.
18. Kiss J. Scapulothoracic fusion in case of facioscapulohumeral muscular dystrophy (FSHD)-Results and problems. In: 24 Congress of the European Society for Surgery of the Shoulder and the Elbow (SECEC); 2012 Sep 19-22. Dubrovnik, Croatia; 2012. p 149. OP – 40.
19. Bunch WH. Scapulo-thoracic fusion. *Minn Med*. 1973 May;56(5):391-4.
20. Bunch WH, Siegel IM. Scapulothoracic arthrodesis in facioscapulohumeral muscular dystrophy. Review of seventeen procedures with three to twenty-one-year follow-up. *J Bone Joint Surg Am*. 1993 Mar;75(3):372-6.
21. Ketenjian AY. Scapulocostal stabilization for scapular winging in facioscapulohumeral muscular dystrophy. *J Bone Joint Surg Am*. 1978 Jun;60(4):476-80.
22. Letournel E, Fardeau M, Lytle JO, Serrault M, Gosselin RA. Scapulothoracic arthrodesis for patients who have fascioscapulo-humeral muscular dystrophy. *J Bone Joint Surg Am*. 1990 Jan;72(1):78-84.
23. Jakab E, Gledhill RB. Simplified technique for scapulocostal fusion in facioscapulohumeral dystrophy. *J Pediatr Orthop*. 1993 Nov-Dec;13(6):749-51.

24. Twyman RS, Harper GD, Edgar MA. Thoracoscapular fusion in facioscapulohumeral dystrophy: clinical review of a new surgical method. *J Shoulder Elbow Surg.* 1996 May-Jun;5(3):201-5.
25. Andrews CT, Taylor TC, Patterson VH. Scapulothoracic arthrodesis for patients with facioscapulohumeral muscular dystrophy. *Neuromuscul Disord.* 1998 Dec;8(8):580-4.
26. Berne D, Laude F, Laporte C, Fardeau M, Saillant G. Scapulothoracic arthrodesis in facioscapulohumeral muscular dystrophy. *Clin Orthop Relat Res.* 2003 Apr;(409):106-13.
27. Diab M, Darras BT, Shapiro F. Scapulothoracic fusion for facioscapulohumeral muscular dystrophy. *J Bone Joint Surg Am.* 2005 Oct;87(10):2267-75.
28. Mummery CJ, Copeland SA, Rose MR. Scapular fixation in muscular dystrophy. *Cochrane Database Syst Rev.* 2003;(3):CD003278.
29. Krishnan SG, Hawkins RJ, Michelotti JD, Litchfield R, Willis RB, Kim YK. Scapulothoracic arthrodesis: indications, technique, and results. *Clin Orthop Relat Res.* 2005 Jun;(435):126-33.
30. Rhee YG, Ha JH. Long-term results of scapulothoracic arthrodesis of facioscapulohumeral muscular dystrophy. *J Shoulder Elbow Surg.* 2006 Jul-Aug;15(4):445-50.
31. DeFranco MJ, Nho S, Romeo AA. Scapulothoracic fusion. *J Am Acad Orthop Surg.* 2010 Apr;18(4):236-42.

Figures

Fig. 1
Scapular winging resulting in cosmetic deformity in a patient with facioscapulohumeral dystrophy.

Fig. 2
Fig. 2-A When thoracoscapular muscles are paralyzed, the deltoid loses its stable origin. As the deltoid contracts, the scapula rotates and the arm remains by the side. **Fig. 2-B** Fixation of the scapula to the ribs provides a stable fulcrum for the action of the deltoid (Reproduced, with permission, from: Copeland SA, Levy O, Warner GC, Dodenhoff RM. The shoulder in patients with muscular dystrophy. *Clin Orthop Relat Res.* 1999 Nov;368:80-91.)

Fig. 3-A

The scapula stabilizing test (Horwitz test) is begun by the patient raising both arms forward and maintaining this position with the arm horizontal until fatigue causes it to drop to the side (right side).

Fig. 3-B

The Horwitz test is then repeated with the examiner manually holding the scapula downward to the chest wall to prevent winging. If the patient finds it easier to maintain flexion or abduction and the fatigue test is prolonged, a thoracoscaphular fusion may be indicated.

Fig. 4-A
The Montreal mattress.

Fig. 4-B
A patient positioned prone on the Montreal mattress.

Fig. 5
Patient positioning for surgery with the arms in 90° to 110° of elevation in the scapular plane and approximately 90° of external rotation on a board. With the patient in this position, the line of the spinous processes of the thoracic vertebrae, the medial border of the scapula, and the scapular spine are marked with a marker pen. A straight incision is made along the medial edge of the scapula, parallel to the line of the spinous processes.

Fig. 6
Exposure of the medial border of the scapula.

Fig. 7
Exposure of four ribs.

Fig. 8
Positioning of the drill for the third hole with the previous two screws in place. Note the thin retractor (McDonald dissector) under the rib to prevent damage to the pleura. Exposure of the ribs from their superior border avoids injury to the intercostal neurovascular bundle.

Fig. 9
Fixation with the third screw.

Fig. 10
Loosening of the screws to enable insertion of bone graft or bone-graft substitute.

Fig. 11
Packing of corticocancellous autograft or bone-graft substitute after loosening of the screws.

Fig. 12
The completed screw fixation.

Fig. 13
The shoulder is immobilized in an adjustable brace with the arm at 60° of abduction and 30° of forward flexion so that the patient can reach the mouth by flexing the elbow.

Fig. 14

Postoperatively, the arm is held in the position described in Figure 13 with the adjustable shoulder brace for three months.

Fig. 15

Clinical outcome at 3.5 years after one procedure and three years after the other in a patient who underwent bilateral thoracoscaphular fusion.

Fig. 16-A
Anteroposterior radiograph showing union at the site of a thoracoscaphular arthrodesis with screw fixation.

Fig. 16-B
Transscapular radiograph showing union at the site of a thoracoscaphular arthrodesis with screw fixation.

Fig. 17
The “squared shoulders” appearance, which the male patients liked as they perceived it to be a “Superman” appearance and the female patients liked as it kept their clothes’ straps and handbag shoulder straps from falling.