THORACOSCAPULAR FUSION FOR FACIOSCAPULOHUMERAL DYSTROPHY

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Eleven thoracoscopic fusion operations have been done on six patients. The indication is symptomatic winging of the scapula caused by thoracoscopic muscle paresis with intact function in the deltoid. This situation almost exclusively occurs in facioscapulohumeral dystrophy. The operation is successful in achieving stability of the scapula and in greatly improving function and cosmesis. Although the course of this type of muscular dystrophy is variable, the benefits of operation have not deteriorated with progression of the disease over a maximum follow-up period of twenty-three years.

The operation was designed by one of us (R. C. H.) in 1954 to help a patient who had severe winging of the scapula as a result of facioscapulohumeral dystrophy. The outcome of the operation was gratifying and produced greatly improved function of the arm.

This paper describes the technique of thoracoscopic fusion using tibial cortical grafts with screw fixation. It is also an independent review by one of us (S. A. C.) on the late results of the operation.

Duchenne in 1868 described the clinical course of a progressive muscular atrophy in thirteen patients, and his name has been associated with the early onset sex-linked recessive form of muscular dystrophy. In 1884 Landozzy and Déjirine reported on a small group of cases in which muscular involvement was initially limited to the face and shoulder girdle. This form of the disease has since been known as facioscapulohumeral dystrophy and it is transmitted as an autosomal dominant with a wide range of expressivity. The age of onset is usually fifteen to thirty years, commonly involving the serratus anterior, trapezius, rhomboids, and latissimus dorsi muscles and also the muscles about the eyes and mouth. This form of the disease runs a very variable course but is generally benign and a normal life span is probable (Walton 1961).

Fortunately, the deltoid muscle is nearly always spared or only partially involved. Without a stable origin the deltoid loses its mechanical advantage, and in attempted abduction the scapula rotates and active sustained abduction and flexion is severely limited. If the scapula can be stabilised, then the deltoid can exert a powerful action on the humerus (Figs. 1 and 2).

Several methods of achieving stability of the scapula have been described but unfortunately they have all been extremely small series with short periods of follow-up.

Putti (1906) described an interscapular fixation which apparently caused compression of the vessels under the clavicle. Rinaldi (1964) fixed the scapula to the thorax using a fascial band taken from the fascia lata and passed around the spinous processes of the second and third and fifth and sixth thoracic vertebrae and then through a hole in the spine of the scapula. This was a modification of the techniques used by Whitman (1932) and Dewar and Harris (1950). They considered it necessary to retain some thoracoscopic movement. Although the initial results are satisfactory the fascial sling stretch and function deteriorates. More recently one case was described by Bunch (1973) in which true

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bony fusion had been achieved between the scapula and thorax using wire and bone grafts. It was noted that after a bilateral procedure the vital capacity was reduced.

The technique to be described was first carried out using tibial cortical grafts alone but subsequently supplementary cancellous iliac crest grafts were sometimes also used. Bony union has been achieved in all patients (Fig. 3). The technique and early results of the first three cases were reported to the British Orthopaedic Association in 1960 (Howard 1961). A long-term follow-up of these and further patients is presented.

CLINICAL MATERIAL
There were six patients, five of whom had had bilateral operations; a total of eleven thoraco scapular fusions. The average age at the time of operation was thirty years (range 15 to 59 years). There were five women and one man.

All had had the diagnosis of facioscapulohumeral dystrophy made and were complaining of symptoms related to winging of the scapulae. The details of each patient are shown in Table I.

OPERATIVE TECHNIQUE
The patient lies supine and a cortical tibial bone graft, 9 centimetres by 1 centimetre, is taken. Cancellous bone from the proximal tibia is scooped out at the same time. This wound is closed, and the patient is then turned to lie prone with the arm hanging over the side of the table. This almost always brings the scapula into the ideal position for fixation to the ribs with the vertebral border lying parallel and 5 to 7 centimetres lateral to the spinous processes.

Table I. Details of the patients

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<tr>
<th>Case number</th>
<th>Sex</th>
<th>Side</th>
<th>Length of follow-up (years)</th>
<th>Age at time of operation (years)</th>
<th>Age at onset of symptoms (years)</th>
<th>Family history</th>
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The incision is made along the medial edge of the scapula. The underlying muscles, which are sometimes atrophied, are divided and the deep surface of the vertebral border of the scapula is denuded of tissue for 2 centimetres laterally, detaching part of the origin of subscapularis. The superficial medial margin of the scapula is also cleared of muscle attachment for about 2 centimetres laterally.

Three ribs lying under the most convenient part of the scapula are chosen: these are usually the fourth, fifth and sixth ribs (Fig. 4). The periosteum is incised in the line of the rib, and the periosteum and parietal pleura are separated so that a retractor can be put under the ribs to prevent damage to the pleura.

The tibial graft is divided into two or three struts, and a hole is drilled at each end. Each graft is then fixed to the scapula and the underlying rib by means of screws passing through the whole thickness of the ribs but obviously not allowed to stand proud of their deep surface. Any gaps left between the scapula and the ribs are then packed with cancellous bone chips. If more cancellous graft is needed, this can be taken from the posterior superior iliac crest. Sound fixation of the scapula to the ribs is achieved at the end of the operation (Fig. 5). The wound is closed with drainage and the patient is carefully turned over, with an assistant holding the arm. A shoulder spica is applied with the arm at 80 degrees abduction and 30 degrees forward flexion, the hand being in front of the mouth.

The patient is nurtured after the operation in a sitting position. The average length of stay in hospital was seventeen days.
Three months after operation the arm section of the spica is bivalved so that the upper section may be removed and the patient can start active abduction. Once control of abduction has been regained, the plaster of Paris is removed. A triangular pillow may be placed under the arm to allow gradual adduction of the arm over a period of one week.

Physiotherapy is then needed to help regain full glenohumeral movements and to strengthen the deltoid muscle.

On two occasions the scapular muscles were found to be hypertrophic and the scapula could not be made to lie against the ribs. Some of the muscle belly of subscapularis was excised, and the tibial grafts were used as spacers and placed between the ribs and scapula; screws were used in the same way (Fig. 6). The number of ribs involved and the number of struts used varied, depending upon the shape of the chest wall and scapula and the fixation achieved.

RESULTS

Five of the six patients were seen personally and the other was contacted by letter.

The longest follow-up was twenty-two years and the shortest six months (average 11.3 years). The time between the onset of symptoms and operation was an average 7.6 years (range 2 to 19 years) (Table 1).

The main complaints before operation were of weakness of the arm and inability to sustain the arm in the abducted or flexed position, as it would just flop down after a short time (for example, patients complained that they could not reach things out of high cupboards nor get books down from high shelves); of cosmetic deformity—the scapula would ride up in the neck and wing posteriorly, looking very ugly and causing great difficulty with clothes; and of a dragging sensation in the shoulder and aching and pain after use.

**Power and function.** The power of the deltoid muscle did not diminish over the years and in fact became stronger. All patients experienced an ache in deltoid for several months after the operation but this gradually disappeared.

Two of our patients had had a slow relentless progress to their disease and were severely affected at time of follow-up, both eventually having involvement of the lower limb. They both, however, managed to do sedentary part-time jobs—one as a book-keeper, and the other as a telephonist involving constant reaching upwards and forwards to take the plugs out of the switchboard.

All patients were independent before operation and could do most things but great effort was required in any movements involving the shoulder girdle. The operation allowed them to do sustained manoeuvres involving the shoulder, and simple tasks such as getting dressed and cooking were achieved without pauses for rest, which was not possible before operation. Three patients who could not wash their hair or backs before operation could do so afterwards.

**Cosmesis.** The majority of our patients were women and cosmesis was a big factor in their decision to accept
operative treatment. Before operation the scapulae not only wing but are elevated on attempted flexion or abduction, giving an ugly appearance of widening the base of the neck.

Although fixation of the scapula leaves a considerable scar the patients were happy to accept this because of the general improvement in appearance (Fig. 7). The scapulae are fixed in the position previously described, which gives maximum range of active movements. This position is further lateral than the natural position of the scapula, and tends to produce a slightly square-shouldered appearance (Fig. 8). This was noted by us but was never remarked upon by the patients.

**Pain.** Pain was never a major factor in that the most described was a persistent aching discomfort before operation. This was relieved in all patients.

**Range of movements.** It is interesting to note the range of movements that these patients can achieve, all of which obviously takes place at the glenohumeral joint alone and is remarkably consistent (Fig. 9). The average range of movements was: abduction 100 degrees, flexion 90 degrees, extension 35 degrees, external rotation 20 degrees, and internal rotation 90 degrees.

The ability to sustain abduction or flexion was, to the patient, the greatest improvement.

**Complications.** *Early:* Two patients complained of pleuritic pain but in both it settled within a week. This was presumably due to protruding screws on the deep surface of the ribs but could not be demonstrated radiographically due to the difficulties of radiography in the plaster spica. Two patients developed localised pain in the chest wall while in plaster, and subsequently this was thought to be due to stress fractures of the ribs that were later seen on radiography to have healed (Fig. 10).

*Late:* One patient complained of pain at the site of fusion and the graft was shown to be ununited. She had a successful secondary graft with iliac cancellous bone. Two patients had to have screws removed as they were
causing superficial irritation. No particular anaesthetic difficulties have been experienced with these patients.

DISCUSSION

Five out of the six patients returned to have a thoracoscopic fusion done on the remaining shoulder. This fact alone speaks for the success of the operation to the patient. The sixth patient had other disabilities of greater priority.

Often it appears that the disease is not symmetrical in its onset and only one shoulder may require treatment; several years may elapse before the other shoulder needs the operation. For example, one patient (Case 4) with a particularly slowly progressive form required the first shoulder to be done at age forty-five and the second at age fifty-nine.

When the disease is asymmetrical the patient complains of feeling “unbalanced” and this balance is restored by operation. This complaint may recur later when the other shoulder is affected and again is corrected by thoracoscopic fusion.

Lung function studies done on some of the earlier patients in the series showed only minimal loss of vital capacity, and none of the patients at review had any functional problem related to this as they were limited by peripheral muscle weakness rather than by any diminution in respiratory reserve.

We know of four further patients who have had bilateral operations for this disease. In two patients the technique described above was used and in the other two fusion was obtained with iliac crest grafts alone. Fusion was achieved in all four patients with similar good clinical results. These were done at other centres and are not included in this series as follow-up was limited.

Facioscapulohumeral dystrophy is a rare disease, and hence the operation is seldom required, but when indicated the results are very successful, fully justifying the major surgery involved.

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REFERENCES


