



Surgical treatment of Sprengel's deformity : A modified Green procedure

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Twelve cases of Sprengel's deformity were treated surgically by a modification of Green's procedure. After clavicular osteotomy, all muscular attachments to the scapula were freed ; an omovertebral band if present was cut and the scapula was rotated and moved caudal to a more normal position, to be sutured into a pocket of the latissimus dorsi. The patients included 10 female and 2 male patients (age range at the time of operation : 3 to 9 years ; mean : 5.2 years). The deformity involved the left shoulder in 8 patients, the right shoulder in 4. All patients were followed for an average of 2 years (range : 6 months to 4.5 years). The range of flexion improved from an average of 95° to 105° and the range of abduction from an average of 81° to 97°. Cosmesis was improved in all 12 patients but three patients had an unsightly surgical scar. Surgical treatment by a modified Green procedure for patients with Sprengel's deformity is indicated when the patient and family want cosmetic or functional improvement.

mality occurs when the scapula does not descend to its normal position on the thoracic wall (Th2 - Th7 level) during week 9 to 12 of foetal development. The scapula is rotated medially ; the glenoid faces inferiorly (7). The inferior pole of the scapula is adducted and the superior angle is prominent in the web of the neck. In 25% to 35% of cases (in 55% of our cases) an omovertebral bone is present ; this is a rhomboidal plaque of cartilage, bone and/or fibrous tissue lying in a strong fascial sheath that extends from the superior angle of the scapula to the spinous processes, lamina, or transverse process of one or more lower cervical vertebrae (7, 9, 10, 18). The results of surgery occasionally are disappointing because the deformity is never simply elevation of the scapula ; it is always complicated by malformations and contractures of the soft tissue structures of the region (1, 6, 8, 12, 15, 16).

INTRODUCTION

Congenital high scapula, also named Sprengel's deformity or congenital undescended scapula is uncommon, but is the most frequent congenital abnormality of the shoulder (9). It is characterised by an apparent elevation of a hypoplastic and misshapen scapula, caused by failure of the scapula to descend (2).

The scapula normally differentiates at about five weeks of gestation opposite to the fourth, fifth and sixth cervical vertebrae (9). The Sprengel abnor-

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Musculature defects are most common in the trapezius, rhomboids and levator scapulae; the pectoralis major/minor, latissimus dorsi, sternocleidomastoid, serratus anterior are less commonly affected. The affected muscles undergo degeneration, necrosis, fibrosis and secondary contracture (7, 18). These factors all contribute to the limitation of scapulothoracic movement.

Other congenital anomalies may be present such as: absence/fusion of ribs, cervical ribs, Klippel-Feil Syndrome, congenital scoliosis with hemivertebrae, cervical spina bifida, syringomyelia, paraplegia, platybasia, situs inversus, mandibulofacial dysostosis, clavicular abnormalities, cardiac anomalies and kidney malformations (4, 11, 14).

Apart from the loss of function of the upper limb, the prominent superior medial angle of the scapula results in a more or less disturbing cosmetic appearance.

Congenital elevation of the scapula in Sprengel's deformity remains a complex therapeutic problem (11, 13, 16). If deformity and impairment are mild, no treatment is indicated. Sprengel's deformity does not respond to conservative treatment. Surgery may be indicated to correct cosmetic deformity, improve shoulder motion and relieve neck pain if present. Most patients with this deformity should receive surgical treatment as a child or adolescent (3).

An operation to bring the scapula inferiorly close to its normal position can be attempted when the child is about 3 years old. However, the earlier the surgery is performed after 3 years of age, the better the results, because the operation becomes more difficult as the child grows. In older children an attempt to bring the scapula inferiorly to its normal level can injure the brachial plexus (3, 7, 17, 18).

In this study, we aimed to present results of the surgical treatment of congenital high scapula by a modification of Green's procedure.

MATERIALS AND METHODS

Patients

We retrospectively reviewed 12 cases of Sprengel's deformity operated between 1998 and 2004 by a modification of Green's procedure. The patients included

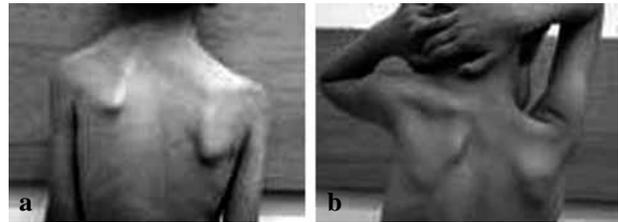


Fig. 1. — Preoperative photographs of a patient : a) extension ; b) abduction.

10 female and 2 male patients (age range : 3 to 9 years ; mean : 5.2 years) at the time of operation. The deformity involved the left shoulder in 8 patients, the right shoulder in 4. The associated anomalies included Klippel-Feil anomaly in 3 patients and congenital scoliosis with fully segmented hemivertebrae in 3 patients. Five patients (42%) demonstrated radiographic evidence of an omovertebral bone.

Before the operation, the elevation and hypoplasia of the involved scapula was a prominent feature. Flexion before operation averaged 95° (range : 80° to 110°) and abduction averaged 81° (range : 75° to 100°). The difference in height of both shoulders was 4.5 cm on average (fig 1). None of the patients exhibited a neurological abnormality.

The preoperative screening protocol included a meticulous physical examination, a radiological assessment of the cervical spine, and an intravenous pyelogram in an attempt to identify any associated anomalies that might accompany Sprengel's deformity. None of the patients displayed a renal anomaly.

Surgical procedure

The surgical technique is the modification of the Green procedure described by Leibovic *et al* (15) associated with a clavicular osteotomy (fig 2). Clavicular osteotomy was performed first, with the patient lying in supine position and under general anaesthesia. The patient was then turned to prone position, re-prepared and re-draped. A midline skin incision was made beginning at the spinous process of the fourth cervical vertebra and extending distally to the spinous process of the tenth dorsal vertebra (C4 to Th10). A plane between the subcutaneous tissue and fascia overlying the trapezius muscle was developed. The insertion of the entire trapezius muscle on the scapular spine was sectioned, and the muscle was elevated extraperiosteally, reflected medially, and separated from the lower fibers of the

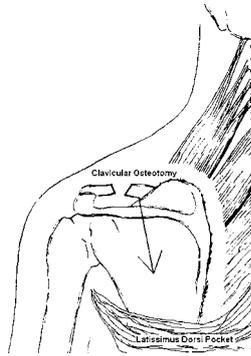


Fig. 2. — Surgical technique ; clavicular osteotomy, release of scapular muscular attachments, section of the omovertebral bone if present, suturing of the scapula into a pocket of the latissimus dorsi after the scapula has been rotated and moved caudad to a more normal position.

latissimus dorsi, exposing underlying muscles and the scapula. The supraspinatus muscle was then detached from the scapula extraperiosteally to the greater scapular notch. The omovertebral band which was present in 5 of our 12 cases was cut. When an omovertebral bone was present, it was resected (2). The insertions of the levator scapulae muscles on the superior-medial angle of the scapula and of the rhomboideus muscles on the medial border of the scapula were extraperiosteally dissected and retracted. The superior margin of the scapula was then retracted posteriorly, and starting medially, the supraspinatus portion of the subscapularis muscle was elevated extraperiosteally from the anterior surface of the scapula. Then the supraspinatus portion of the scapula along with its periosteum was excised with the help of a bone cutting forceps or with an osteotome. Afterwards, all attachments of the latissimus dorsi muscle to the scapula were extraperiosteally divided, and by blunt dissection a large pocket was created deep to the superior part of the latissimus dorsi muscle. Then by direct pressure, the scapula was gently displaced distally and rotated into a more normal position and it was sutured into a pocket of the latissimus dorsi. The inferior pole of the scapula was attached to the adjacent rib with two or three absorbable sutures to prevent winging of the scapula. The divided muscles were then re-attached.

During the postoperative care, the shoulder was immobilised in a Velpeau bandage reinforced with plaster of Paris. About four weeks postoperatively, the cast was removed and active shoulder abduction and scapular depression exercises were performed to increase muscle strength and range of motion.



Fig. 3. — Clinical presentation of the patient in figure 1 four year postoperatively : a) extension ; b) abduction. Note the improved cosmesis and function.

RESULTS

All of the 12 patients improved functionally and cosmetically with surgery, but an unsightly surgical scar was seen in three patients. The mean difference in shoulder height decreased from 4.5 cm to 1.4 cm after the operation. The average range of flexion improved from 95° (range : 80° to 110°) before operation to 105° (range : 95° to 120°). Abduction averaged 81° (range : 75° to 100°) before, and 97° (range : 90° to 125°) after surgery (fig 3).

In all of the patients, the scapular body remained somewhat smaller than its contralateral counterpart. None of the patients complained of pain in the region of the operative site. No brachial plexus palsy was seen. All patients and parents were satisfied with the final result of the treatment.

All patients were followed for an average of 2 years (range : 6 months to 4.5 years). None of the children displayed loss of cosmetic appearance or range of motion of the relevant shoulders. The position of the scapula did not change with time.

DISCUSSION

Numerous operations have been described to correct congenital high scapula. Two surgical techniques are commonly used. Green (7) described surgical release of muscles from the scapula along with excision of the supraspinatus portion of the scapula and any omovertebral bone. The scapula is then moved inferiorly to a more normal position and the muscles are reattached. Woodward (18) described transfer of the origin of the trapezius muscle to a more inferior position on the spinous processes.

Leibovic *et al* (15) modified the Green procedure by suturing the scapula into a pocket in the latissimus dorsi after rotating the scapula and moving it caudally to a more normal position. We preferred Leibovic's modification together with clavicular osteotomy in our patients as their major problem was cosmesis. By applying this approach, better cosmetic improvement was achieved in our patients. Bellemans and Lamoureux (1) also described a modification of the Green procedure in which the serratus anterior muscle was not dissected and the postoperative mobilisation began immediately. They reported 77° of abduction gain in 7 patients.

Greitemann *et al* (8) in their 23 cases operated on for congenital high scapula, reported that, for patients with impaired function, the Woodward procedure was preferable and for patients with cosmetic problems only, resection of part of the superior angle of the scapula was performed. They believed that better results were obtained with the Woodward procedure because it is followed by less scar formation that may fix the scapula in a poor position; it allows a larger mobilisation and leaves a less obvious postoperative scar. Khairouni *et al* (12) reported 79% improvement in cosmesis and function in 19 cases surgically treated by a modification of the Woodward procedure, associated with a correction of glenoid tilting. The modified Green procedure can be used instead of Woodward's procedure especially for patients in whom the cosmetic deformity is the major problem (5).

Mears *et al* (16) described partial resection of the scapula, removal of an omovertebral bone associated with a release of the long head of the triceps from the scapula. They reported improvement of flexion from 100° to 175° and abduction from 90° to 150° in 8 patients. According to these authors, contracture of the long head of the triceps represents a significant obstacle to full abduction and release of this contracture allows increased abduction.

Due to the aggressive efforts to descend the scapula during surgery, brachial plexus palsy is the most severe complication of correction of congenital high scapula. To avoid brachial plexus palsy, some authors recommended clavicular

osteotomy on the ipsilateral side as a first step in the operative treatment (1, 6, 8). We routinely followed this advice and clavicular osteotomy was performed in all our cases.

Finally we want to emphasise that functional outcome and cosmetic appearance are the main elements of a good result, rather than the radiographic position of the scapula.

In conclusion, we believe that the modified Green procedure for patients with Sprengel's deformity is indicated when either improved cosmesis or improved function is desired by the patient and/or family. It is more advantageous than the Woodward operation in terms of surgical easiness, operating time, postoperative follow-up and patient's expectancies.

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