SPRENGEL’S DEFORMITY: SURGICAL CORRECTION BY A MODIFIED GREEN PROCEDURE

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ABSTRACT

Objective: To evaluate the cosmetic and functional results of patients submitted to surgical correction of Congenital High Scapula (Sprengel’s Deformity) using modified Green’s Procedure, as well as patients’ satisfaction and complications.

Methods: Nine patients submitted to surgical treatment from September 1993 to April 2008 have been assessed. The modification from original technique was: subperiosteal muscle detachment, resection of superomedial scapular portion and fixation of medial portion of scapular spine to contralateral posterior iliac crest instead of skeletal traction, with subcutaneous wire. The mean age was 7 years and 3 months. The mean follow-up time was 3 years and 7 months. Results: The mean improvement in forward elevation was 39° (range 0 to 80°). According to the Cavendish Classification, cosmetic improvement of two degrees was achieved in eight cases, and three degrees in one. All patients were satisfied with results.

Conclusions: Surgical correction of Sprengel’s Deformity by a modified Green’s procedure with contralateral posterior iliac crest fixation instead of skeletal traction, showed both cosmetic and functional improvements; all patients and/or family members were satisfied with the results, and the complications associated to the surgical technique did not interfere on end results.

Keywords – Sprengel’s deformity; Scapula

INTRODUCTION

Congenital elevation of the scapula is a rare condition of unknown etiology that results from the abnormal termination of the caudal migration of the scapula in the embryonic period (1-4). Eulenberg first described this condition in 1863, and after the publication of Kolliker’s study in 1891, which credited Sprengel for this finding, it was referred to as the Sprengel deformity (1,2,5,6). The main clinical changes are the hypoplasia and abnormal positioning of the scapula, which may cause aesthetic problems and limit the movements of the shoulder girdle (1,2,7,8) (Figure 1). Chinn reports the possibility of the diagnosis being made in the prenatal period by ultrasound (9).

The presence of omovertebral bone, which joins the scapula to the spine, is a characteristic change cited by several authors (1,7,10-12). Mooney et al. reported a case in which a bone structure was identified extending from the medial border of the scapula toward the clavicle and the occipital region, unlike the usual pattern of omovertebral bone (13).

Surgical treatment of deformity seeks to improve aesthetics and, in some cases, function, and may consist of osteotomies, bone resections, muscle releases with repositioning of the scapula, or a combination of these (4,6,8,10,11,14-16). The surgical technique described by Green in 1957 is one of the most used and involves bone resection of the supraclavicular portion of the scapula, and lowering of the same, which is maintained by means of skeletal traction (17). Leibovic et al. (4) describe a modification of this technique in which the scapula is kept lowered by means of a suction with a pocket made in the latissimus dorsi muscle.
All patients underwent surgical treatment. Besides the aesthetics, the indication for surgical treatment also aimed to improve shoulder function, mainly through a gain in elevation.

The preoperative clinical evaluation consisted of measuring the elevation of the affected shoulder, which ranged from $80^\circ$ to $160^\circ$ (average $110^\circ$); the aesthetic deformity was assessed using the Cavendish criteria\(^6\) (Table 1), five patients were classified as grade III and four grade IV.

The surgical technique was a modification of that described by Green, which originally consisted of a detachment of the extraperiosteal upper and medial muscles of the scapula, omovertebral bone resection (if present), supraclavicular excision of the scapula, lowering of the scapula and reinsertion of the muscle in the new position, maintaining this position with skeletal traction wire\(^{17}\). As a modification of the original technique, subperiosteal muscle detachment and resection of only the superior medial portion of the scapula were performed. Instead of using skeletal traction, we opted for subcutaneous steel wire fixation of the medial scapular spine to the contralateral posterior iliac crest\(^{20}\) (Figures 3 and 4).

Postoperatively, patients remained immobilized in a Velpeau cast for about four weeks. The fixation material (steel wire) was removed, on average, 69 days (ranging from 30 to 110 days) after the initial surgical procedure.

**Table 1 – Cavendish scale used to aesthetically classify patients with a Sprengel deformity\(^6\).**

<table>
<thead>
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<th>Grade</th>
<th>Description</th>
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<tr>
<td>I (very mild)</td>
<td>Leveled shoulders and practically invisible deformity when the patient is clothed.</td>
</tr>
<tr>
<td>II (mild)</td>
<td>Shoulders practically leveled, but the deformity is visible when the patient is clothed.</td>
</tr>
<tr>
<td>III (moderate)</td>
<td>Shoulder elevated from 2 to 5 cm and the deformity is easily visible.</td>
</tr>
<tr>
<td>IV (severe)</td>
<td>Shoulder significantly elevated, with the superior angle of the scapula close to the occipital.</td>
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**Figure 1** – Clinical changes in a Sprengel deformity. Hypoplastic and elevated left scapula (a) and limitation of elevation (b) (case 9).

**Figure 2** – Omovertebral (OV) bone. Radiographic (a) and intraoperative (b) appearance (case 6).
The same aesthetic and functional criteria used preoperatively were used for postoperative evaluation, with the addition of the degree of patient and/or family satisfaction (satisfied/dissatisfied), the appearance of the scar (normotrophic/hypertrophic), and the presence of complications related to the surgical technique. Lowering of the scapula as described by Carson et al.\(^{11}\) was measured in the radiographic evaluation, that is, the difference between the preoperative and postoperative scapular unevenness.

The mean postoperative follow-up was three years and seven months, ranging from eight months to seven years and 11 months.

**RESULTS**

There was an increase in the average elevation of about 39° (ranging from 0° to 80°), the variation of elevation in the postoperative period was from 130° to 160°, with an average of 149°. According to the Cavendish classification, eight cases showed an improvement of two grades and one case showed an improvement of three grades. Six patients were raised to grade I and three to grade II.

The average radiographic lowering was 2.7 cm (ranging from 1 to 5.8 cm).

All scars were considered normotrophic.

All nine patients and/or their families were satisfied with the results.

The postoperative complications included breaches...
of the fixation material in three cases and a superficial infection in one case. These complications did not interfere directly with the final results.

Table 2 contains a summary of the sample and results.

**DISCUSSION**

Sprengel deformity is the most common congenital deformity of the shoulder(3,10). It is the result of a scapula blocked from its normal descent, probably between the ninth and 12th weeks of pregnancy, causing a bone to be in an abnormal position and usually hypoplastic, and the periescapular muscles present varying degrees of hypoplasia and degenerative changes(2,3,6,7,11).

Skeletal deformities are frequently associated with Sprengel deformity, ranging from 67% to 100%(1,3,4,16,21). Seven of our cases (78%) had associated deformities, the most frequent of which were in the spine (scoliosis and Klippel-Feil syndrome) and changes in the ribs.

A change considered by many authors to be characteristic is the presence of an omovertebral connection joining the superior medial angle of the scapula to the spinous process, lamina, or the transverse process of the lower cervical spine. This connection can be of a fibrous, cartilaginous, or osseous nature or a combination of these(1,10,11). Sartoris(22) considers the anteroposterior radiograph and the computed tomography reconstruction in three dimensions to be the best tests for the detection of omovertebral bone. Reports of the presence of omovertebral bone in the literature indicate an incidence of 20% to 46%(6,8,11,12,16), but only one patient in our series (11%) had this connection.

Surgical treatment is not indicated for mild deformities; it is reserved for patients with more pronoun-
cised deformities\(^{(1,2,6,14)}\). According to Cavendish\(^{(6)}\), the primary goal of treatment is cosmetic improvement, since functional improvement is not consistent. Carson et al.\(^{(11)}\) suggest that improved function can also be a goal in patients with important functional limitations. Farsetti et al.\(^{(23)}\) observed that after 25 years of follow-up in 14 patients treated medically, there were no aesthetical and functional changes in relation to the initial clinical evaluation, and concluded that whenever there is an expectation of improvement from the family and patient, surgical treatment is indicated, preferably using a technique that includes resection of the upper part of the scapula.

Boon et al.\(^{(24)}\) reported a case treated surgically where dissection was performed in 16 cadavers prior to the procedure to improve knowledge of the local anatomy. They observe that the dorsal scapular nerve located in the upper medial border of the scapula is at the greatest risk of injury; the accessory and suprascapular nerves are rarely at risk.

Dendane et al.\(^{(25)}\) emphasize that the ideal candidates for surgery to reposition the scapula are those with less than four years of age, while Greitemann et al.\(^{(26)}\) consider the ideal to be an operation before six years of age. Khairouni et al.\(^{(27)}\) report that the patient’s age and the presence of omovertebral bone do not influence the results. Doita et al.\(^{(28)}\) achieved good aesthetic results in two patients operated in adulthood, but used only bone resection techniques without lowering the scapula. Ross and Cruess\(^{(29)}\) analyzed 77 operated cases and found the best results with respect to elevation gain to have occurred with the Woodward\(^{(8)}\) and Green\(^{(17)}\) techniques, both of which include the lowering of the scapula. Zhang et al.\(^{(30)}\) do not consider lowering the scapula to improve aesthetics to be the main objective and affirm that only resection of the superior medial portion of the scapula and the omovertebral bone, when present, is a safe and effective method of treatment.

In only two of our patients (cases 4 and 7) were the indications for surgery purely for aesthetic purposes. All were operated on during the first decade of life. By understanding the repositioning of the scapula to be essential to achieving functional improvement, besides aesthetic improvement, we chose to use the Green technique with some modifications that were introduced to our department by Dr. Ruy Rocha de Macedo, of which there were no reports in the literature. The main modification was to fix the scapula in the contralateral posterior iliac crest with subcutaneous steel wire instead of performing skeletal traction. The possibility of wire breakage, which occurred in three patients, and the need for a second procedure to remove this wire can be considered disadvantages of the surgical technique. But these factors did not alter the final results.

The morcellation (osteotomy) of the clavicle associated with the procedure for lowering the scapula is regarded as a first surgical procedure for many authors, and can be performed in severe deformities to avoid compression of the brachial plexus\(^{(1,2,3,16)}\). In our study, this procedure was not performed, since there were no such problems. McMurtry et al.\(^{(31)}\) consider avoiding excessive correction of the position of the scapula to be the best way to prevent neurological complications.

Carson et al.\(^{(11)}\) obtained 29° of elevation gain with an average lowering of 1.6 cm, while Grogan et al.\(^{(32)}\) gained 37° with 2 cm of lowering. Both used the Woodward technique. In our cases, we obtained mean gain of 39° of elevation and 2.7 cm of lowering.

Wilkinson and Campbell\(^{(14)}\) and Cavendish\(^{(6)}\) state that the association of Sprengel deformity and severe skeletal changes would be a contraindication to surgery because it would not offer significant aesthetic benefits. Despite a gain of 40° of elevation, the patient (case 1) who had associated severe scoliosis was not satisfied in the early postoperative period. However, in the later follow-up period, both the patient and his family said they were pleased with the outcome of surgery.

Regarding aesthetics, there was an improvement of at least two grades according to Cavendish’s classification, which contributed to the high rate of patient satisfaction.

**CONCLUSIONS**

The results of nine patients with Sprengel deformity operated by a modified Green technique, lowering and fixing the scapula to the contralateral posterior iliac crest with steel wire, showed both aesthetic and functional improvement. All patients and/or their families were satisfied with the treatment and the complications related to the technique did not affect the final results.
REFERENCES