The outcome of 56 children (61 shoulders) treated surgically at the Rizzoli Institute between April 1975 and June 2010 for congenital elevation of the scapula is reported. There were 31 girls and 25 boys with a mean age at surgery of 6.4 years (2 to 15). The deformity involved the right shoulder in 20 cases, the left in 31 and was bilateral in five. The degree of the deformity was graded clinically and radiologically according to the classifications of Cavendish and Rigault, respectively. All patients underwent a modified Green procedure combined, in selected cases, with resection of the superomedial portion of the scapula and excision of any omovertebral connection. After a mean follow-up of 10.9 years (1 to 29.3), there was cosmetic improvement by at least one Cavendish grade in 54 shoulders (88.5%). The mean abduction of the shoulder improved from 92° (50° to 155°) to 112° (90° to 170°) and the mean flexion improved from 121° (80° to 160°) to 155° (120° to 175°). The unsatisfactory cosmetic result in seven shoulders was due to coexistent scoliosis in two cases and insufficient reduction of the scapular elevation in the other five. An incomplete upper brachial plexus palsy occurred post-operatively in three patients but resolved within seven months.

We suggest that a modified Green procedure combined with resection of the superomedial portion of the scapula provides good cosmetic and functional results in patients with Sprengel’s shoulder.

Sprengel’s shoulder is a rare congenital deformity resulting from failure of the scapula to descend properly.1-3 Affected children have two major problems: cosmetic and functional. The clinical appearance, a result of elevation and malrotation of the scapula, may be worsened by frequently associated anomalies such as scoliosis, Klippel-Feil syndrome and asymmetry of the chest-wall.

The condition has been widely reported and since Putti1 in 1908 described transplantation of the scapula to a lower level by excision of the protruding upper part and division of muscle attachments, various forms of treatments have been described.2-7

Cavendish8 reviewed 110 patients from different orthopaedic centres, and Gonen et al9 and Ross and Cruess10 described 28 and 77 cases, respectively – besides these, publications on the surgical treatment of Sprengel’s shoulder are based on small studies, ranging from five to 22 patients.11,12

The aim of this study was to present the largest series hitherto of patients with Sprengel’s shoulder treated operatively at a single centre.
All the patients were classified clinically and radiologically. Their clinical appearance was assessed according to the Cavendish scale \(^8\) and evaluated on plain radiographs in the standing position using Rigault’s grading system \(^13\) (Table I, Figs 1 and 2). Active and passive movement of the affected shoulder was tested in abduction and flexion. A total of 15 patients (27%) also underwent three-dimensional CT reconstruction (3D-CT).

Surgery was indicated for marked limitation of movement, moderate or severe clinical deformity (Cavendish grades III or IV) and a Rigault grade of 2 or 3 (Fig. 2a).

All the patients underwent a modified Green procedure (Fig. 2b).\(^{4,14}\) Trapezius was elevated extraperiosteally from the scapula and reflected medially. The insertions of levator scapulae on the superior angle and rhomboids on the medial border of the scapula were dissected. The whole superomedial region was always explored and any scapulovertbral or scapulothoracic connection was detached. The omovertebral bone, when present, was excised. Supraspinatus was then detached from the scapula along with its periosteum. The superomedial angle of the scapula was excised in some cases. The scapular attachments of latissimus dorsi and serratus anterior were detached and the scapula was displaced distally to the level of the healthy side and fixed to a rib using an absorbable suture or a pouch of latissimus dorsi. After checking the orientation of the glenoid, the muscles were reattached to the scapula.

Post-operatively, a Velpeau bandage was used for two weeks when active-assisted range of movement exercises were initiated.

The patients were examined at one, three, six, nine and 12 months, and annually thereafter. The clinical appearance (Fig. 2c) at the last available follow-up including the Cavendish grade and the appearance of the scar were evaluated from photographs shown to an independent examiner. The appearance of the scar was considered normal if it looked similar to the surrounding tissue, hypertrophic if it was thicker than the surrounding tissue and keloid if it was abnormal, thicker and different in colour and texture from the surrounding tissue. The range of movement of the shoulder was compared with the pre-operative value. All intra- and post-operative complications were recorded.

**Results**

There were 36 patients with Cavendish grade III deformity and 15 with grade IV. Five others had bilateral grade IV involvement. According to Rigault’s classification, 41 shoulders were grade 2 and 20 were grade 3.

In all unilateral cases, the incision was along the medial border of the scapula. In three bilateral cases a midline paraspinous vertebral incision was used. At operation an embryonic remnant was seen in 30 shoulders (49%). This was a bony connection in 22 shoulders (36.1%), cartilaginous in two (3.3%) and fibrous in six (9.8%). Excision of the superomedial angle of the scapula was performed in

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**Table I. Distribution of the 61 shoulders pre- and post-operatively according to the classifications of Cavendish\(^8\) and Rigault et al\(^13\)**

<table>
<thead>
<tr>
<th>Classification</th>
<th>Description</th>
<th>Pre-operative (n, %)</th>
<th>Post-operative (n, %)</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Cavendish</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Grade I</td>
<td>Very mild, shoulder level and deformity invisible when dressed</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Grade II</td>
<td>Mild, shoulders almost level, lump visible in web of the neck when dressed</td>
<td>0</td>
<td>22 (36.1)</td>
</tr>
<tr>
<td>Grade III</td>
<td>Moderate, shoulder elevated 2 cm to 5 cm. Deformity easily visible</td>
<td>41 (67.2)</td>
<td>32 (52.5)</td>
</tr>
<tr>
<td>Grade VI</td>
<td>Severe, superior angle of the scapula near occiput, with or without neck webbing</td>
<td>20 (32.8)</td>
<td>7 (11.5)</td>
</tr>
<tr>
<td><strong>Rigault</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Grade 1</td>
<td>Superomedial angle lower than T2 but above T4 transverse process</td>
<td>0</td>
<td>15 (24.6)</td>
</tr>
<tr>
<td>Grade 2</td>
<td>Superomedial angle located between C5 and T2 transverse process</td>
<td>41 (67.2)</td>
<td>34 (55.7)</td>
</tr>
<tr>
<td>Grade 3</td>
<td>Superomedial angle above C5 transverse process</td>
<td>20 (32.8)</td>
<td>12 (19.7)</td>
</tr>
</tbody>
</table>

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Diagram showing Rigault’s radiological classification \(^13\) for Sprengel’s deformity of the shoulder. Grade 1, superomedial angle lower than T2 but above T4 transverse process; Grade 2, superomedial angle between C5 and T2 transverse process; Grade 3, superomedial angle above C5 transverse process.
42 shoulders (69%). Following distal displacement to the level of the healthy side the scapula was fixed to a rib with an absorbable suture in 50 shoulders (82%), whereas a pouch of latissimus dorsi was used in 11 shoulders (18%).

Three patients developed an incomplete palsy affecting the upper brachial plexus a few days post-operatively; they received electro-stimulation therapy and the symptoms resolved within seven months.

At a mean follow-up of 10.9 years (1 to 29.3) the mean abduction of the shoulder improved from 92° pre-operatively (50° to 155°) to 112° (90° to 170°). The mean forward flexion improved from 121° pre-operatively (80° to 160°) to 155° (120° to 175°).

There was cosmetic improvement by at least one Cavendish grade in 54 shoulders (89%, in 50 patients) (Table I). The seven unsatisfactory cosmetic results were due to coexistent progressive scoliosis in two cases and insufficient reduction of the scapular elevation in the other five.

The surgical scar was acceptable in 34 shoulders (55.7%), hypertrophic in eight (13.1%) and keloid in 16 (26.2%). A paraspinous incision was used in the remaining three patients with bilateral deformity (Table II). The mean post-operative difference between the superior-internal angles of both scapulae was 1.8 cm (0 to 5).

The patients generally showed a radiological improvement except for 12 shoulders (19.7%) with persistent grade 3 deformity (Table I). The only late complication was recurrence of a bony omovertebral connection, which required a further procedure in one patient.

Discussion

Following operative correction by a modified Green technique an improvement was achieved both functionally and cosmetically in 80% (49 of 61) and 88.5% of shoulders (54 of 61), respectively. Over the years, the operation has remained the same, but the percentage of patients where the lowering of the scapula was combined with a resection of its superomedial angle has increased. There has also been the tendency to operate at an earlier age, thereby resulting in better functional outcomes. The cosmetic results were tarnished by a keloid scar in 27% of shoulders (16 of 58).

In our opinion surgical correction of Sprengel's shoulder is indicated in patients with a Cavendish grade III or IV and Rigault grade 2 or 3 deformity. The Cavendish scale, based on cosmetic criteria, is popular but subjective, and should be used in combination with Rigault's classification. The main goals of surgery in this condition are to improve the cosmetic appearance and function.
techniques available include inferior relocation of the scapula \(^4,17,19\) and/or scapular osteotomy with or without resection of the scapular body.\(^7,20,21\)

There are few studies reporting the results of Green’s procedure \(^8\) or its modification.\(^9,14,22,23\) The cosmetic results are satisfactory, with a mean lowering of the scapula by 1.5 vertebrae\(^14\) and 1.9 vertebrae\(^22\) in two series, respectively. In our series there was cosmetic improvement by at least one Cavendish grade in 54 shoulders (88.5%) (Table I).

We agree with other authors that the final appearance is dictated by the pre-operative state and the presence of associated spinal deformities has a marked negative impact,\(^12,14,15,20\) as shown by the low profile outcomes in the present series with 25 children with marked brevicollis in Klippel-Feil syndrome.

It has been argued that operations based on lowering of the scapula overlook the functional aspect.\(^16\) In the present series there was a minor improvement in function compared with previous series.\(^14,22\) However, this might be due to the mean age, which was significantly higher in our patients (mean 6.4 years compared with means of 3\(^14\) and 3.7 years\(^22\)) and the high number of patients with Klippel-Feil syndrome (40% in the present series compared with 0%\(^14\) and 13%\(^22\)).

In order to improve function in selected patients, such as those with Klippel-Feil syndrome with homolateral thoracic scoliosis, the Mears technique \(^7\) might be a viable option, because the spinal deformity impedes the lowering of the scapula and an oblique osteotomy through the body of the scapula with subsequent resection allows better orientation of the glenoid. In the report by Masquijo et al\(^16\) 14 patients with and without associated Klippel-Feil syndrome were treated by this method with significant improvement of shoulder movement.

In the surgical treatment of Sprengel’s shoulder, 3D-CT can help in pre-operative planning and plays an important role in the detection of an bony omovertebral bar.\(^24\) Even if the connection is cartilaginous or fibrous, the surgical approach for Green’s procedure allows exploration of the whole superomedical region and removal of all scapular connections to the spine.

In our patients it was not possible, as in the 3D-CT study of Cho et al,\(^24\) to evaluate the true dimensions of the costoclavicular space that contains the brachial plexus. Therefore the risk of brachial palsy, which occurred in only three shoulders in the present series, might be avoided by performing a clavicular osteotomy, whatever the age of the patient.

Green’s scapuloplasty for the correction of Sprengel’s shoulder is less commonly undertaken than Woodward’s technique.\(^6\) However, both procedures show similar outcomes based on a lowering of the scapula and better orientation of the glenoid. Resection of the superomedical portion of the scapula and its fixation to the rib cage also contribute to better cosmetic results.

No benefits in any form have been received or will be received from a commercial party related directly or indirectly to the subject of this article.

References