Abstract

Introduction: With a less than ideal health infrastructure and a vast underprivileged population in our country, many cases of Sprengel's deformity are initially missed and are subsequently untreated. Although for best surgical results, the patient must be under the age of five, many patients present late at our tertiary care center. Thus, we evaluated the clinical results of the modified Woodward's Procedure in such neglected cases.

Methods: A retrospective study, of 16 patients aged 8 year or above, with Sprengel's deformity who underwent the modified Woodward's procedure between 2006 and 2011. Clavicular osteotomy/morselization was avoided. The Cavendish scoring system was used to grade cosmesis and shoulder abduction was used to assess the functional outcome. The patients were prospectively followed up at two years and after skeletal maturity.

Results: There were ten females and six males and the average age was 9.4 years. The mean follow up was 4.2 years. Omo- vertebral bar excision was done in 13 (81.25%) patients. The mean increase in postoperative shoulder abduction and Cavendish grades at a 2 years follow-up were, 19.1° and 1.32 grades, respectively. None of our patients needed Clavicular osteotomy/morselisation and none developed neurological abnormalities. The was no change in the values at skeletal maturity.

Conclusion: Satisfactory outcomes can be acquired by the Modified Woodward’s procedure in neglected Sprengel’s deformities. Besides a definitive cosmetic correction, the procedure also results in improvement of shoulder abduction. Clavicular Ostetomy/Morselization is not always required in cases presenting after the age of 8 years.

Keywords: Woodward’s procedure, Sprengel’s shoulder, Cavendish grading, Modified Woodward’s procedure, Neglected Sprengel deformity.

Introduction

Sprengel's deformity (SD) or congenital elevation of the scapula is a rare congenital abnormality of the shoulder girdle. It is characterized by a high riding, mal-rotated scapula and is usually associated with Spino-cervical anomalies. The disorder ranges from mild restriction in shoulder abduction to severe cosmetic deformity and shoulder dysfunction. Due to uncoordinated growth between the fibrous bands and the hypoplastic bone, SD can be progressive requiring early surgical intervention. Several methods have been described with the techniques described by Woodward [1] and Green[2] being most commonly performed. The condition is usually obvious at birth and gets more evident as the child ages. If surgery is performed below the age of five, there is significant improvement in cosmesis and shoulder abduction [3, 4]. Due to a vast underprivileged population in
India, many cases of SD are initially missed and subsequently neglected. Such untreated cases, where the primary presentation is after 8 years, are not uncommon at our tertiary care center. Thus, the purpose of our study was to evaluate the functional results of the Modified Woodward's Procedure in such neglected cases.

Materials and Methods
This is a retrospective study of patients between the age of 8 and 12 years with SD operated via the modified Woodward procedure. The cases were operated by a single surgeon between 2006 to 2011, at a tertiary care center. Patients were followed up at 2-year post-operatively and after skeletal maturity.

Evaluation for cosmesis was according to the Cavendish classification [5] and functional outcome was measured by shoulder abduction in comparison to the other side. The indications for surgery were severe limitation in shoulder abduction and/or an unacceptable cosmetic appearance. Sixteen patients who underwent the below described modification of the Woodward procedure, were included in the study.

Surgical Technique
1) Under general anaesthesia and in the prone position, a midline skin incision was made extending from mid-cervical spine to D-10 vertebral level. All muscles between the scapula and spine were released sequentially from their spinal insertion.
2) The omovertebral bar was excised if present; partial excision of the supraspinous portion of scapula was also performed.
3) The scapula was brought down with gentle traction and the muscles were reattached to the spine at their new insertion using No.5 Ethibond sutures.
4) Clavicular osteotomy was not required in any of the patients. For the initial 3 weeks, the patients were immobilized with an arm-chest bandage. Mobilization was started in the third week with active range-of-motion exercises for the shoulder. The Cavendish grading and range of shoulder abduction were measured at 1 year follow up and at skeletal maturity.

Results
The average age of the cohort was 9.4 years (Range: 8-12 years) with 10 females and 6 males. The mean follow-up was 4.6 years. Three patients were Cavendish grade 2, six patients were Cavendish grade 3 and seven patients were with Cavendish grade 4. Average pre-operative shoulder abduction was 82.5° which improved post-operatively to 101.6°. Omo-vertebral bar excision was performed in 13 (81.25%) patients. The mean increase in postoperative shoulder abduction and Cavendish grades was 19.1° and 1.32 grades, respectively. There was a cosmetic improvement of at least 1 Cavendish-grade in 14 shoulders (87%) at 2-year post-operatively (Table 1). The was no change in the values at skeletal maturity.

One patient had a superficial skin infection, and two patients suffered inadvertent skin damage over the scapula. Dural puncture during omovertebral bar excision was noted in one patient. There were no brachial plexus or neurological injuries.

Discussion
First described by Eulenberg in 1863, as congenital elevation of the scapula [6], it later came to be associated with the name Sprengel after his description of a series of four cases in 1891 [7]. The two major problems encountered with SD are cosmesis and reduced shoulder abduction. Conservative management has been largely unsuccessful in addressing these problems [3]. Thus, several surgical options were introduced to encounter these issues. The surgical options mainly consist of a combination of lowering of the scapula, muscle detachment and scapular osteotomy. The Green's [2] and Woodward's [1] procedures, along with their modifications are among the commonly practiced surgeries. Due to the relative rarity of this deformity, it has been difficult to generate large sample sizes and conduct prospective trials to establish the superiority of any single procedure over the other. Associated anomalies vary on a case-to-case basis and impact the functional outcome. However, several case series have noted good outcomes following the Woodward procedure in children below 8-years [8–13]. None of the studies report the outcomes of neglected SD, its associated challenges and complications.

Ever since its first description in 1961, the Woodward procedure [1] has become the most widely used technique for the management of Sprengel shoulder. The correction is obtained by moving the spinous insertions of the trapezius and rhomboids downwards after resection of the omovertebral bone. Several modifications to the Woodward procedure have been proposed [8–13]. These include, an additional excision of the medial scapular border, thereby reducing its horizontal width and preventing apparent winging of the scapula; anterior release and subsequent rotation of the scapula, thus correcting the varus angulation to the glenoid, or a combination of the above.

In our study, all muscles between the scapula and spine were released sequentially from their spinal insertions. If present the omovertebral bar was excised, partial excision of supraspinous portion of scapula was also performed. The scapula was gently pulled down and the muscles were reattached to the spine at

| Table 1: Post-operative changes at follow-up |
|-----------------------------|-----------------------------|
| Variables                  | Pre-operative (Mean)         | 2-yr follow-up (Mean)   |
| Cavendish Grading          | 3.25                        | 1.93                     |
| Shoulder Abduction         | 101.6                       | 82.5                     |
| Change                     | 1.93                        | 101.6                    |
| Change in %                | 40.62                       | <0.001                   |
| p’ Value                   | 1.32                        | 3.25                     |

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their new insertion. We did not feel the need for a clavicular osteotomy in any patient.

Patients with SD are painless and have a reduction in shoulder abduction, affecting activities of daily living. They are usually associated with other anomalies. For these reasons, we used the Cavendish system to objectively evaluate the outcome of surgical treatment. In previous studies, the Cavendish scale improved by 1 or 2 grades, implying the overall cosmetic appearance of the patients improved substantially [8–13]. Majid et al [14] had a mean Cavendish grade change of 1.8 in their cohort. There was an average change of 1.32 grade in our study and fourteen cases had at least one Cavendish grade improvement in cosmesis. Clearly, neglected cases can have substantial improvement following the procedure, and enthusiastic traction to place the scapula more caudally does not appear to be necessary.

The studies on Woodward’s procedure have shown a mean increase in abduction of 32° to 59° [6, 7, 10–14]. Although, not the sole criteria to measure the functional outcome, the principal reason for functional disability is reduced shoulder abduction. The mean increase in shoulder range of abduction in our series was 19.1°. However, overhead activities was possible in all patients and there was no loss of movement during longer term follow-up. In our experience, the improvement in abduction after the modified Woodward’s procedure was substantial. Our results suggest that surgeries performed on cases presenting after 8 years of age restores functional abduction of the shoulder, but the range of abduction achieved is less as compared to cases operated at a younger age.

Opinion regarding the optimal age for surgical correction remains divided. Surgical correction before the age of 3 years is more challenging compared to an older child [15]. If surgery is performed below the age of five, there is significant improvement in cosmesis and shoulder abduction [3, 4]. However, with increasing age, no substantial results may be expected due to loss of biological plasticity of soft tissues, fibrosis and rapid growth. Loss of function due to fixed structures and psychological maladjustment may be additional factors. However, there is no evidence to state that older children have a worse prognosis after surgery. Our study focusses on neglected cases with the mean age at surgery being 9.4 years. None of our patients experienced problems, during surgery or at follow-up.

Complications of this procedure have been frequently reported in the literature [10–14]. Post-operative infections are rare but the scar is often hypertrophic or results in a keloid. Winging of the scapula, brachial plexus palsy and scar dehiscence have been known to occur. Our patients did experience complications in the form of superficial skin infection, inadvertent skin damage over the scapula and dural puncture during omovertebral bar excision. There was no recurrence of deformity in any of the patients. Brachial plexus palsy, is the most dreaded complications of the procedure. Children above the age of 8 years are at a higher risk for this [15], probably due to the distorted anatomy and often fibrotic structures with limited elasticity. The role of clavicular osteotomy in the prevention of brachial plexus palsy remains

<table>
<thead>
<tr>
<th>Study authors</th>
<th>Average Age (years)</th>
<th>No. of cases</th>
<th>Procedure performed</th>
<th>Clavicular osteotomy</th>
<th>Increase in function (degree)</th>
<th>Improvement in cosmesis (Cavendish scale)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Woodward(1)</td>
<td>8.1</td>
<td>9</td>
<td>Woodward</td>
<td>No</td>
<td>32</td>
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<tr>
<td>Crha and Ga’l(8)</td>
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<td>18</td>
<td>Woodward</td>
<td>Not routinely performed</td>
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<td>0.67</td>
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<tr>
<td>Grogan et al.(9)</td>
<td>6.5</td>
<td>20</td>
<td>Woodward</td>
<td>Not routinely performed</td>
<td>37</td>
<td>-</td>
</tr>
<tr>
<td>Wu et al.(10)</td>
<td>4.7</td>
<td>9</td>
<td>Woodward</td>
<td></td>
<td>78</td>
<td>2.23</td>
</tr>
<tr>
<td>Carson et al.(15)</td>
<td>9</td>
<td>11</td>
<td>Woodward</td>
<td></td>
<td>28</td>
<td>-</td>
</tr>
<tr>
<td>Ross and Cruess(13)</td>
<td>-</td>
<td>17</td>
<td>Woodward</td>
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<tr>
<td>Cho et al (11)</td>
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<td>10</td>
<td>Modified Woodward</td>
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<td>58</td>
<td>-</td>
</tr>
<tr>
<td>Jindal et al(12)</td>
<td>5.6</td>
<td>12</td>
<td>Woodward</td>
<td>Not routinely performed</td>
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<td>1.92</td>
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<td>Borges et al(16)</td>
<td>-</td>
<td>14</td>
<td>Modified Woodward</td>
<td>Not routinely performed</td>
<td>34</td>
<td>-</td>
</tr>
<tr>
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<td>8.4</td>
<td>16</td>
<td>Modified Woodward</td>
<td>No</td>
<td>19.1</td>
<td>1</td>
</tr>
</tbody>
</table>

Table 2: Demographics and results of patients of Sprengel's deformity treated by the Woodward procedure by different authors
It is usually performed prophylactically to prevent the palsy. The osteotomy is also presumed to better mobilise the scapula in older children where the joints are not supple enough for surgical descent. However, there is no direct evidence to suggest that clavicular osteotomy prevents neurological complications. We did not perform clavicular osteotomy in any of our patients, and found acceptable cosmetic and functional results. We suggest that a clavicular osteotomy is not always necessary even in the older child with Sprengel's deformity.

**Limitations of the studies**

A retrospective study design with a small study group are the major limitations of our study. However, a child presenting with SD after the age of eight years is rare, and the cohort among them requiring surgery is even fewer. We consider 16 such cases present an adequate sample size to be able to draw meaningful inferences.

**Conclusion**

Modified Woodward's procedure gives good results in neglected SD. In addition to providing definitive cosmetic correction, the procedure also results in improvement of shoulder abduction. Clavicular osteotomy is not always required in most neglected cases.

**References**


**Declaration of patient consent**: The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given the consent for his/her images and other clinical information to be reported in the journal. The patient understands that his/her names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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