**Clinical Experience of the Woodward Procedure in Sprengel’s Deformity**

Shu-Jung Wu¹, Lin-Show Chin¹, Jinn-Rung Kuo²

**Objectives:** To determine the safety and outcome in the treatment of Sprengel’s deformity by the standard Woodward procedure without clavicular osteotomy.

**Methods:** This nine year retrospective study from 1998 to 2006 closely followed 9 patients, six boys and three girls, aged 3-6 years (mean, 4.7±1.1 years), with Sprengel’s deformity. The ages of patients at the time of diagnosis ranged from 0 (immediately after birth) to 5 years. Associated congenital diseases included Klippel-Feil syndrome (6 cases), congenital scoliosis (3 cases) and congenital heart disease (1 case). All patients underwent the standard Woodward surgical procedure. No clavicular osteotomy or other modified methods were performed during the operations. Changes in the shoulder abduction degrees and the appearance classified by the Cavendish grading system were recorded before and after the operations.

**Results:** In this study using the Wilcoxon signed rank test, the results indicated significant improvements in the abduction degrees of the shoulder (before vs. after: 82.22±7.12 vs. 160.56±11.84, p=0.006) and the Cavendish grades (before vs. after: 3.56±0.53 vs. 1.33±0.5, p=0.007). No surgical complications were noted during the 4.5 year-follow-up period (mean: 8 years; ranging from 3 to 10 years).

**Conclusions:** As a result of this clinical study, we conclude that the Woodward procedure is a safe and effective procedure in the treatment of Sprengel’s deformity. Besides, for patients younger than six years of age, we consider that it is not necessary to combine the clavicular osteotomy with the Woodward procedure.

**Key words:** sprengel’s deformity, woodward procedure, abduction of the shoulder, cavendish grade, omovertebrae, klippel-feil syndrome

Sprengel’s deformity is an uncommon disease, which occurs as the result of an interruption of the normal caudal migration of the scapula during development, most likely between the ninth and twelfth weeks of gestation. This disease further leads to cosmetic deformities and is often accompanied by functional impairment. Many hypotheses have been proposed to explain how this developmental failure occurs, but to date the exact cause is still unknown.

With this deformity, the scapula is elevated, hypoplastic and somewhat adducted. This deformity is usually noticed at birth and progresses with the development of the child. The left side is often more affected than the right. The main findings on physical examination include a webbed neck, a palpable scapula into the cervical region, and a limited range of motion in the shoulders. Radiographs show that the superior angle is attached to the cervical spine by an omovertebral bone or band in about 40% of the cases. Certain associated osseous anomalies, such as scoliosis, spina bifida or incomplete segmentation are often present.

Treatment goals are to correct the deformity and to...
improve the function of the affected shoulder. Conservative treatment with rehabilitation is advised in mild cases. If the deformity is especially disfiguring or the function of the affected shoulder is impaired to a significant degree, operative intervention should be considered.

Historically, various surgical procedures have been employed. For example, Putti described a procedure in which the upper protruding part of the scapula is excised and pulled downward, after division of the muscle attachment. Schrock and Green subsequently modified these procedures and described a new technique for correcting the deformity, with disappointing results. Woodward described a procedure by which the correction is obtained by moving the origins of the trapezius and rhomboids downward onto the spine. He reported 9 cases that underwent this procedure and the follow-up ranged from 9 months to 5 years. Results were satisfactory, leading to improvement in both appearance and shoulder function. Transient brachial plexus paralysis developed in only one case postoperatively. Grogan et al. reported a series of 20 patients with congenital elevation of the scapula treated using the Woodward procedure. Excellent results were obtained in 80% of the cases.

In this retrospective study, we will describe our clinical experience with this procedure and report long-term results in nine patients, who were surgically treated on account of severe functional impairment (82.22±7.12 degrees of scapulo-humeral abduction) and moderate to severe deformities in appearance (Cavendish grade: 3.56±0.53).

### Materials and Methods

From 1998 to 2006, nine patients (six boys and three girls), who had been diagnosed to have Sprengel’s deformity, were observed at the Chi-Mei Medical Center, in Tainan, Taiwan. The ages of patients ranged from 0 (immediately after birth) to 5 years. The ages at the time of surgery ranged from 3 to 6 years. The deformity was located on the right in 4 cases (44%) and on the left in 5 (56%). None of the patients had a bilateral deformity. The omovertebral bones, found in 4 cases (44%) were completely resected during each operation. All nine patients were associated with congenital abnormalities, such as Klippel-Feil syndrome, characterized by a short neck with a low hair line, due to a reduction in the number of vertebrae (six patients, 66%), congenital heart disease (one patient, 11%), or scoliosis (three patients, 33%). A detailed clinical summary is shown in Table 1.

Preoperatively, measurements of the abduction degrees of both shoulders were recorded in order to evaluate postoperative improvement. The severity of the deformities was assessed by the Cavendish four-grade system to standardize the evaluation of cosmetic improvement. Photographs are available for all patients to assess the cosmetic deformities.

All nine patients had moderate to severe deformity in appearance (Cavendish grades 3 to 4) with the arc of abduction of less than 90 degrees (severe functional loss). If the Cavendish grade 1 (very mild) and 2 (mild) patients had been diagnosed, physical therapy treatment

### Table 1. Clinical Summary of 9 Cases

<table>
<thead>
<tr>
<th>Case</th>
<th>Gender</th>
<th>Side</th>
<th>Age at operation (years)</th>
<th>Omovertebral bone</th>
<th>Associated congenital abnormalities</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>F</td>
<td>R</td>
<td>3</td>
<td>Y</td>
<td>Congenital heart disease, VSD</td>
</tr>
<tr>
<td>2</td>
<td>M</td>
<td>L</td>
<td>4</td>
<td>N</td>
<td>Congenital scoliosis</td>
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<tr>
<td>3</td>
<td>M</td>
<td>R</td>
<td>4</td>
<td>N</td>
<td>Congenital scoliosis</td>
</tr>
<tr>
<td>4</td>
<td>F</td>
<td>R</td>
<td>4</td>
<td>N</td>
<td>Klippel-Feil syndrome, Congenital scoliosis</td>
</tr>
<tr>
<td>5</td>
<td>M</td>
<td>R</td>
<td>4</td>
<td>Y</td>
<td>Klippel-Feil syndrome</td>
</tr>
<tr>
<td>6</td>
<td>M</td>
<td>L</td>
<td>6</td>
<td>N</td>
<td>Klippel-Feil syndrome</td>
</tr>
<tr>
<td>7</td>
<td>M</td>
<td>L</td>
<td>6</td>
<td>N</td>
<td>Klippel-Feil syndrome</td>
</tr>
<tr>
<td>8</td>
<td>M</td>
<td>L</td>
<td>6</td>
<td>Y</td>
<td>Klippel-Feil syndrome, Spina bifida</td>
</tr>
<tr>
<td>9</td>
<td>F</td>
<td>L</td>
<td>5</td>
<td>Y</td>
<td>Klippel-Feil syndrome</td>
</tr>
</tbody>
</table>

VSD: ventricular septal defect
would have been indicated first, and surgical intervention not advised.

Each of the nine patients underwent operative correction using the Woodward procedure. During the surgical procedure, the patient was placed prone on the operating table with his neck slightly flexed. A mid-line incision was then made extending from the spinous process of the fourth cervical vertebra, distal-ward to the ninth thoracic vertebra. The subcutaneous tissue was undermined to display the vertebral border of the scapula and the orientation of the trapezius muscle fibers. The inferior and lateral border of the trapezius muscle were identified and then separated from the deeper muscular layer of the latissimus dorsi. The origin of the trapezius was then detached from the spinous processes. After the muscle sheet had been removed from the spinous processes, the omovertebral bone or fibrous bands attached to the superior border of the scapula were exposed. The omovertebral bone, when present, was completely excised from its attachment to the cervical spine and superior angle of the scapula. Special attention was paid to neurovascular structures such as the spinal accessory nerve, which lies on the undersurface of the trapezius muscle in line with the medial border of the scapula, and they were carefully protected. Additionally, nerves to the rhomboids and the transverse cervical artery, which lie posterior to the levator scapulae muscle, and are inserted into the superior angle of the scapula, are most likely to be injured and therefore, we made it a point to protect them well during dissection or removal of the omovertebral bone.

After removal of the omovertebral bone or fibrous bands, and when the levator scapulae muscles had been released, we made sure that the scapula became free and mobile. By means of downward traction force by the muscular aponeurosis we brought the displaced scapula distally until its scapular spine line was at the same level as that of the opposite normal scapula. Then, the tough aponeurosis and fibrous origins of the trapezius and rhomboid muscles were reattached from the proximal to distal positions with heavy sutures. The subcutaneous tissue and skin were then closed with a fine synthetic suture to avoid postoperative scar formation.

After the operation, the shoulder was held in a neutral position with a Velpeau bandage for 3 weeks in order to provide more comfort for the patients and to prevent excess movement of the arm and shoulder. Following this procedure, both active and passive exercises were started.

The data of this study were analyzed using the Wilcoxon Rank Sum Test with SPSS software (Version 16.0, SPSS Inc., Chicago, II, U.S.A.). Pre and post operative changes in shoulder abduction degrees and Cavendish grades were examined and compared. P values of less than 0.05 were considered statistically significant.

Results

From 1998 to 2006, nine patients with Sprengel’s deformity (6 boys and 3 girls) underwent treatment and were retrospectively studied. They all had the same surgery (standard Woodward procedure without clavicular osteotomy or modified methods) at the ages ranging from 3 to 6 years. The duration of follow-up ranged from 3 to 10 years (average, 8 years).

The cosmetic deformity (Cavendish grade) and functional status (abduction degrees of the shoulder) before and after the Woodward procedure are presented in Table 2. Patients were classified as having a moderate to severe deformity in appearance, four were rated as Cavendish grade 3 and five were rated as grade 4 preoperatively. After their operations, all patients showed at least one-grade improvement at their follow-up appointments. The cosmetic appearance before and after the Woodward procedure were compared with photographs (Fig 1a, 1b). With regard to the radiographs of Case 8, the C3-C6 spina bifida with omovertebral bone could be identified preoperatively. Four months following surgical treatment, the undescended scapula was lowered to nearly the same level of the normal side (Fig 2a, 2b).

Regarding the functional impairment of these cases, the arc of abduction ranged from 70 to 90 degrees.

Table 2. Cavendish Grade and Range of Abduction Status Before and After Woodward Procedure

<table>
<thead>
<tr>
<th>Case</th>
<th>Cavendish grade</th>
<th>Range of abduction (degree)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Before</td>
<td>After</td>
</tr>
<tr>
<td>1</td>
<td>3</td>
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<tr>
<td>2</td>
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<tr>
<td>8</td>
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<td>1</td>
</tr>
<tr>
<td>9</td>
<td>4</td>
<td>1</td>
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</table>
preoperatively (severe functional loss). The follow-up range was 140 to 170 degrees, averaging 160 degrees. The changes in the Cavendish grade and the range of abduction before and after the Woodward procedure are listed in Table 3. Using the Wilcoxon signed rank test, the improvement in both the range of abduction (before vs. after: 82.22±7.12 vs. 160.56±11.84, p=0.006) and Cavendish grade (before vs. after: 3.56±0.53 vs. 1.33±0.5, p=0.007) was significant.

All the patients and their parents were pleased by the improvement of appearance despite the broad scars and occasional keloids, which occurred in some of the patients. None of the patients had brachial plexus palsy or vascular complications postoperatively, and additional operative procedures were not needed. No wound infection or recurrence of the deformity was noted in the long-term follow-up (mean, 8; range, 3 to 10 years).

**Discussion**

Sprengel’s deformity is the most common congenital deformity of the shoulder. Since 1908, various procedures have been described to improve the appearance and shoulder function of patients with Sprengel’s deformity. Previous surgical results are summarized in Table 4, along with the data on the patients from this study. It has been noted that the Woodward surgical technique is the most popular to use with patients with Sprengel’s deformity. Borges et al. described a modified Woodward procedure, which included excision of the medial border of the scapula to allow a better orientation of the shoulder. This method produced marked improvement in appearance, as assessed on the Cavendish scale, but did not seem to improve shoulder function significantly. The improvement of abduction ranged from 0 to 60 degrees (mean, 35; median, 30). The average age at the time of surgery was 8 years and 3 months (range, 4-17 years).

In our cases, the ages at the time of diagnosis ranged from birth to 5 years (mean±STD) and those at the time of surgery ranged from 3 to 6 (mean, 4.7±1.1) years. The average age of patients at the time of surgical intervention in our study was younger than those reported in other series in the literature (Table 4). Additionally, our patients had severe functional loss with the arc of abduction ranging from 70 to 90 degrees, and moderate to severe deformities in appearance assessed as Cavendish grade 3 to 4 (Fig 1). Compared with previous cases reported in other series (Table 4), ours had more severe functional and cosmetic impairment, but the surgical results were still satisfactory.

Cosmetic deformity is a major concern and is often accompanied by functional impairment, but not every...
patient with Sprengel’s deformity warrants surgical intervention. In the clinical application, according to the Cavendish classification, neither grade 1 nor minimal functional impairment warrants an operation. With grade 2, if the cosmetic deformity is not acceptable, surgical treatment is indicated. Grade 3 and 4 deformities often need surgical intervention because of comparatively severe functional and cosmetic impairment. Rigault’s classification is based on radiologic assessment, and seems more appropriate and objective than the Cavendish grading system. For cosmetic reasons, we prefer to use the Cavendish grade rather than Rigault’s classification in clinical assessment of patients because on the radiographs, it is difficult to measure the deformed scapula. Although the Cavendish classification is too subjective and not precise enough, it is morphologic, and proved easy to use for assessing the improvement of gross appearance in our patients.

When considering the timing of surgical intervention, the recommended age for early surgical treatment (before the age of six) by the Woodward procedure without clavicular osteotomy or modified methods, appears proper for safe and effective surgical correction of moderate to severe Sprengel’s deformity.

References

使用Woodward術式治療先天性肩胛骨高位症之臨床經驗

吳書榮¹ 秦凌霄¹ 郭進榮²

先天性肩胛骨高位症（Sprengel's deformity）是一種罕見的先天性肩胛骨異常疾病，目前確切的發生原因還不是很清楚。當病人的外觀變形或者功能受限情況嚴重的時候，常需要開刀來矯正外觀及功能上的缺陷。從1998到2006年，共有9位先天性肩胛骨高位症的病人在我們醫院進行手術治療，其中包括6個男孩及3個女孩，年紀介於3到6歲間，平均是4.7±1.1歲時接受手術矯正。這類病人出生時多半合併有其他先天性異常，包含Klippel-Feil症候群（6例）、脊柱側彎（3例）、心血管疾病（1例）等。這9位病人我們均使用標準Woodward術式來進行手術矯正。術中沒有合併鎖骨切骨術或其他術式。術前術後肩關節的外展位度、功能恢復及外觀等均詳細加以測量並分級比較。

經過臨床的統計分析，我們發現使用此術式的病人無論在外觀上或功能上均有顯著的進步：肩關節外展位度（術前／術後：82.22±7.12 / 160.56±11.84, p=0.006）；外觀分級（Cavendish grade，術前／術後：3.56±0.53 / 1.33±0.5, p=0.007）。所有病人在術後平均追蹤時間約8年（由3到10年不等），臨床上並沒有發現因手術所造成的併發症；統計來說，我們認為使用Woodward術式矯正先天性肩胛骨高位症是一種有效且安全的手術。對小於6歲的病患，術中不合併鎖骨切骨手術並不會增加術後神經血管損傷的風險，且就長期追蹤來看其預後也令人滿意。