ECRONICON OPEN ACCESS

EC ORTHOPAEDICS
Research Article

Surgical Treatment of Sprengel's Deformity of the Scapula in Children

Nguyen Ngoc Hung* and Nguyen Do Ngoc Hien

National Hospital for Pediatrics, Hanoi Medical University, Vietnam

***Corresponding Author**: Nguyen Ngoc Hung, National Hospital for Pediatrics, Hanoi Medical University, Vietnam.

Received: December 05, 2018; **Published:** January 29, 2019

Abstract

Background: Sprengel deformity is a congenital failure of the root of the scapula bone. Limit shoulder abduction and cosmetic appearances are of great concern. Although the Woodward procedure has reported satisfactory adjustments, Long-term functional outcome after surgery has not been resolved.

Method: Fourteen patients (14 shoulders) have undergone the original and modified Woodward procedure. Woodward procedure for Sprengel distortion has been evaluated. Cosmetic appearance and function. The results of the roles are evaluated by the Cavendish classification system and continuous point function.

Classification cosmetic appearance, range shoulder movement, and radiography were obtained to explain.

Results: Original and/modified Woodward procedure: 1. Age at Surgery: 9+1 (SD = 46.10)/6+8 (SD = 26.62); P-valuate: 0.00000; Age at Lase Follow-up: 14+3 (SD 35.34)/11+1 (SD = 0.35); P-valuate: 0.10928; 2. Cavendish grade at Pre-operation: 3 (SD = 0)/3.13 (SD = 0.35); P-valuate: 0.00000; Cavendish grade at Last Follow-up: 1.16 (SD = 0.40)/1.38 (SD = 0.52): P-valuate: 0.560689 (not significantly different); 3. Motion (°) at Pre-operation: 97.5 (SD = 8.80)/93.13 (SD = 4.58); Pvaluate:0.005416; Motion (°) at Post-operation: 148.33 (SD = 10.32)/147.5 (SD = 6.54); P-valuate: 0.596971 (not significantly different); 4. Scapular Lowering on X-ray (cm): 2.53 (SD = 0.76)/2.21 (SD = 0.70); P-valuate: 0.496 (not significantly different).

Post-operatively, Comparing outcome of cosmetic and function between original and modified Woodward procedure were not significantly different.

Conclusion: The original and modified Woodward procedure offers dramatic improvement in shoulder function and cosmetic appearance for patients.

Keywords: *Sprengel Deformity; Woodward Procedure; Modified Woodward Procedure*

Introduction

Sprengel's deformity is a rare congenital anomaly of the shoulder girdle. First described by Eulenberg in 1863 [1], it was later attributed to Sprengel who reported four cases in 1891 [2].

The incidence and aetiology are unknown. Most cases occur sporadically, although there may be an autosomal dominant trait [3]. The deformity is due to a failure of descent of the scapula between the ninth and 12th weeks of intrauterine life. The scapula is originally located as a sagittally orientated structure in the neck. It then migrates to a coronally aligned posterior thoracic position. Failure of descent leaves it in an elevated, malrotated position. The periscapular muscles are hypoplastic; fibrous bands tether the scapula to the chest wall and further limit rotation. The bone is small, broad and rotated with the inferior angle medialised and the glenoid directed caudally. The superior angle is attached to the cervical spine by an omovertebral bone or band in about 40% of cases. Associated abnormalities of the cervical spine are commonly found.

With this deformity, the Scapula bone is raised, hypoplastic and partly added. This distortion is usually noticed at birth and progresses with the development of the child. The left side is often more affected than the right. Major findings on physical examination include a membrane neck, a palpable bones, and a limited range of motion in the shoulder.

This is an uncommon deformity that occurs due to interruption of normal jaw movement in the developmental process [4]. Several hypotheses have been proposed to explain this failure during development, but the exact cause is not known [5]. Cosmetic deformation is a major concern and sometimes distortion accompanies functional decline. Scapula related abductions are usually limited to the severity of Abductor shoulder anger, but functional defects are usually minimal.

Affected children have two important issues, cosmetic and functional. Clinical manifestations have been classified in an unrealistic way by Cavendish [6]. This can get worse due to the related Klippel-Feilsy syndrome which produces a short neck. From the functional point of view, the abduction of the shoulder is limited because of the scapulothoracic stiffness and a lower glenoid rotation. Most children cannot abduct more than 90°.

The goal of the treatment is to repair the deformity and improve the function of the affected shoulder. Cautious treatment with rehabilitation is recommended in mild cases. If the distortion is particularly distorted or the function of the affected shoulder is severely impaired, an intervention is considered.

Historically, various surgical procedures have been used. For example, Putti [7] describes a process in which the upper projection of the Scapula bone is removed and pulled down, after dividing the attachment of the muscle. Schrock [8] and Green [9] were later revised.

These procedures and describe a new technique to adjust the deformation, with disappointing results. Woodward [10] describes a process in which a correction is obtained by moving the origin of the trapezium and rhombus down the spine. He reported nine cases that went through this procedure and monitored the range from 9 months to 5 years. The results are satisfactory, leading to improved both appearance and shoulder function. Cerebral palsy temporarily develops only in one case after surgery.

In this retrospective study, we will describe our clinical experience with this procedure and report long-term results in Fourteen patients, who were surgically treated on account of severe functional impairment of scapulo-humeral abduction) and moderate to severe deformities in appearance.

Materials and Methods

Between 2002 and 2016, 14 Woodward procedures and modified were performed on 14 patients at the National Hospital for Pediatrics. There were Eight girls and Six boys. The age at the time of diagnosis ranged from birth to 4 years (average, 2 years 6 months). The average age at the time of surgery was 7 years 8 months (range, 4 - 15 years) (Table 1). Ten patients had surgical procedures on the right scapula, four on the left scapula, and no patients had bilateral deformity (Table 2).

All members have confirmed their agreement. This research has been approved by our Institute's Ethics Review Committee and is implemented in accordance with the principles of the Helsinki Declaration.

Preoperative evaluation included a combination of both shoulder tests (defined as combined glenohumeral and scapulothoracic motion). Appropriate photographs are available for all patients to assess cosmetic deformities. Spinal anomalies were evaluated by X-rays and in some cases by computerized tomography (CT). Intravenous or ultrasound examinations are also used to evaluate related kidney abnormalities.

The follow-up evaluation includes clinical shoulder tests, including the range of motion activity in all planes. In the attempt to standardize the evaluation of cosmetic improvement, Cavendish's classification system has been used [6] (Figure 1). In class I (very light), the shoulder joints are level and distortion is unclear when the patient is dressed. At level II (mild), the shoulder joints are level but distortions are visible when the patient is dressed. At level III (moderate), the shoulder joint is raised to 2 - 5 cm and the distortion is obvious. At level IV (heavy), the shoulder is associated with a lot of elevation and the upper corner of the jaw near the bud (Figure 2).

Figure 1: *A-B: Patient have Sprengel's Deformy.*

Figure 2: *Patient have Sprengel's Deformy and limiting elevating of the humerus.*

Preoperative grades were assigned based on patients' photographs and radiographs made before surgery. Anteroposterior radiographs of both scapulae were used to measure scapular lowering and were compared with the preoperative radiographs (Figure 3). We used the inferomedial angle of the scapula as the reference point for preoperative and follow-up measurements [11].

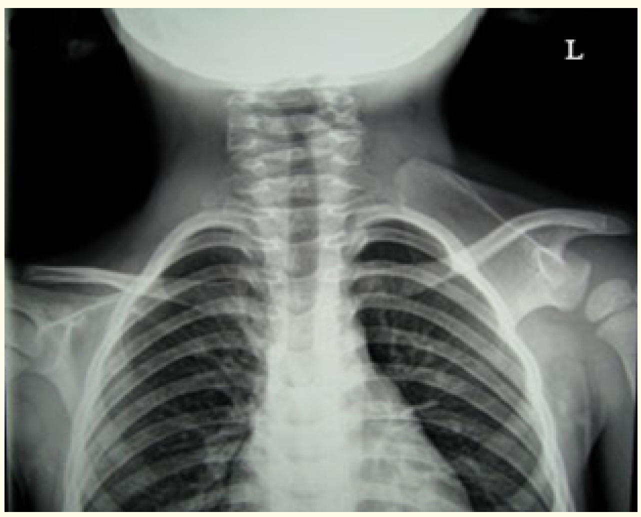

Figure 3: *Elevating Left capula in roentgenography.*

Fourteen 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 would have been indicated first, and surgical intervention not advised.

The omovertebral bones, found in 5 cases (35.7%) 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.

Classification

Numerous efforts have been made to classify Sprengel distortions to assist in guiding management and evaluating results. Cavendish classification, based on the appearance of cosmetics (for example, gravity elevation), is the most widely used classification system [3].

Rigault and his colleagues [12] developed a radioactive taxonomy of the Sprengel strain based on the superimposed projection of scapula on X-rays. Deformities are classified based on the relationship between the subliminal angle of the scapula and the relative level of vertebral. Grade 1 deformation deals with superlative angles under T1, Grade 2 deformation lies between T1 and C5, and Grade 3 deformation lies on C5.

Ross and Cruess [13] measured shoulder height based on the center level of the humeral head relative to the trunk vertical axis. Sprengel distortion has a rotational component, and Leibovic., et al. [14] measured taste displacement based on the vertical position of bone and rotational component. They assume that the center of rotation of the jaw bone is through acromioclavicular joints.

Advanced and inferior angles were developed to quantitatively and visually compare intraoperative radiographic and postoperative radiographs. However, this measurement is difficult to use and has not been widely applied.

Surgical technique

During surgery, the patient is placed easily on the operating table with a slightly curved neck. A later incision was made that lasted from the spinous process of the fourth cervical vertebra, far from the wards to the ninth thoracic vertebra. The subcutaneous tissue undermined to display the vertebral border of the scapula and the orientation of the trabecular muscle fibers. Border line and lateral vein were identified and then separated from the deeper muscular layer of latissimus dorsi.

The origin of the trapezius is then separated from the spinous processes. After the plates have been removed from the spinous processes, the omovertebral bone or strips attached to the superior border of the scapula are exposed. Omovertebral bones, when present, have been completely removed from its attachment to the cervical spine and to the upper scapula. Pay special attention to neural structures such as nerves, spine accessories, located on the undersides of Trapezius muscles are suitable for the intermediate border of scapula bone, and they are carefully protected. In addition, the nerves with rhomboids and the medial artery, located behind the levator scapulae, and inserted into the upper corners of the bones, are more likely to be injured and therefore, we have made it a point to preserve. Protect them well while dissecting or cutting off the omovertebral bone [6,15] (Figure 4). After removing the bone fibers or the omovertebral bone and as the levator muscles are freed, we ensure that the bones become free and mobile (Figure 5 and 6). By means of pulling down due to the development of skeletal muscle, we carry the knife blade away until its vertebrae is at the same level as the normal bone track opposite. Then, the hard aponeurosis and fibrous roots of the trapezius and rhomboid muscles were reattached from near to distant locations with heavy sutures. The subcutaneous tissue and skin are then closed with a good synthetic stitch to avoid scar formation after surgery [15].

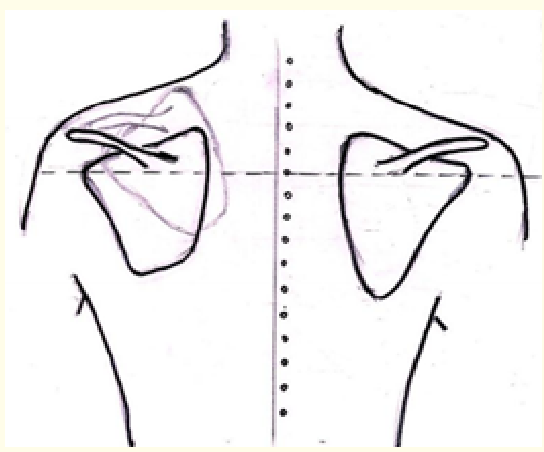

Figure 4: *Surgical technique by Woodward., et al.*

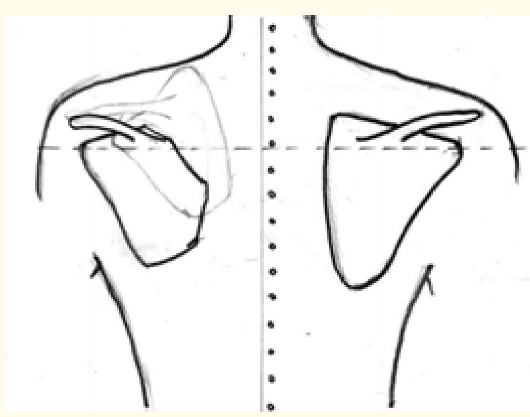

Figure 5: *Modified Woodward's surgery.*

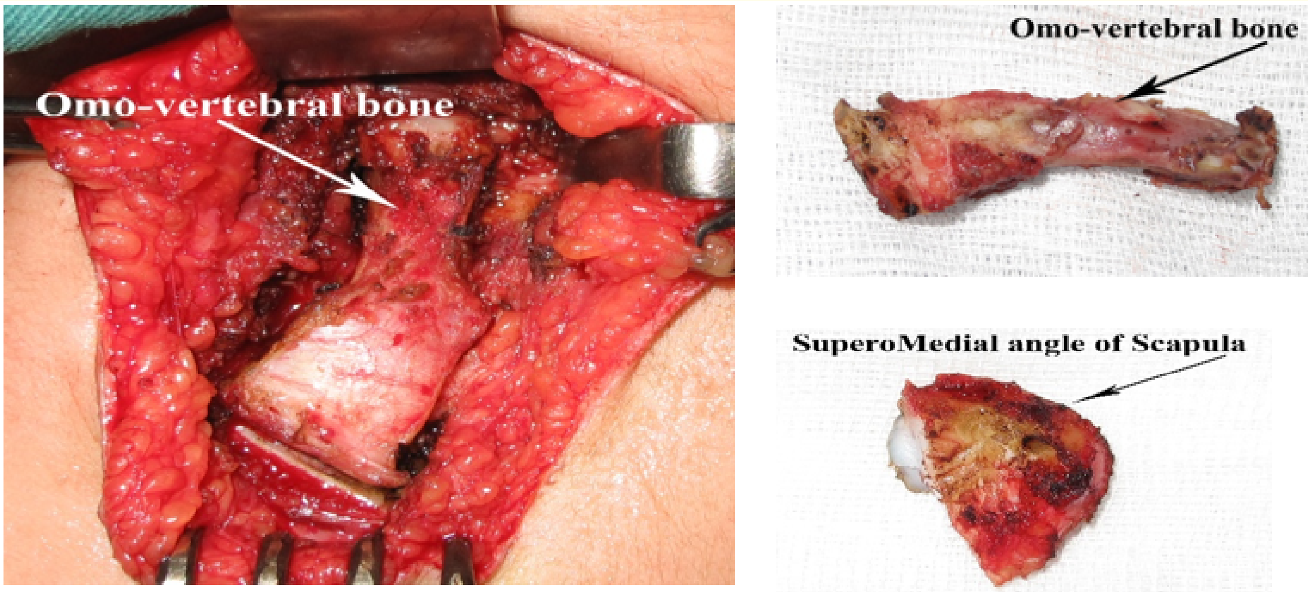

Figure 6: *Omovertebral bone and SuperoMedial angle of the Scapula were removed intra-operation.*

After the operation, the shoulders were kept neutral in Velpeau for three weeks to provide greater comfort to the patient and prevent excess movement of the arms and shoulders. In this process, both active and passive exercises are started.

The dotted line represents the reference point for the position of the scapular spine of the hypoplastic scapula at the same level as the normal contralateral scapula.

Excision of Supero-Medial portion scapula along with Omovertebral bone is removed.

Statistical analysis

Data were analyzed using the Epi Info Software Public 6.04 for epidemiology, developed by the Centers for Disease Control and Prevention (CDC) in Atlanta, Georgia, USA, http://www.ncdc.gov/epiinfo/html/prevVersion.htm. We have taken the $\chi 2$ quiz and the t-test to compare the average among the groups before and after surgery. P values below 0.05 are considered statistically significant.

Results

Between 2002 and 2016, 14 modified Woodward procedures were performed on 14 patients at the National Hospital for Pediatrics. There were Eight girls and Six boys. The age at the time of diagnosis ranged from birth to 4 years (average, 2 years 6 months). The average age at the time of surgery was 7 years 8 months (range, 4 - 15 years) (Table 1).

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, thirteen were rated as Cavendish grade 3 and one 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 (See table 3 and 4).

Regarding the functional impairment of these cases, the arc of abduction ranged from 90 to 110 degrees (average 95 degrees) preoperatively. The follow-up range was 13 to 160 degrees, averaging 147.85 degrees. The changes in the Cavendish grade and the range of abduction before and after the Woodward procedure are listed in table 3. Cavendish grade at Pre-operation: 3 (SD = 0) and Postoperation: 1.16 (SD = 0.40) P-valuate = 0.00000 was significant; Modified Woodward procedure are listed in table 4. Cavendish grade at Pre-operation: 3.13 (SD = 0.35) and Postoperation: 1.38 (SD = 0.52), P-valuate = 0.000048 was significant.

Case	Sex	Side	Age at surgery	Age at Lase F-U	Associated deformities	Type of Surgery	OVB	Cavendish grade		Motion (°)		Scapular Lowering on X-ray (cm)
								Preo	Last F-U	Preo.	Posto	
1.	F	R	10+6	15+1	Congenital Cervicothoracic Scoliosis	MW	+	III	I	90	145	1.5
2.	F	L	4+2	12+4	Congenital Cervicothoracic Scoliosis, diastematomyelia, rib deformity, kidney deform.	MW		III	I	90	160	2.6
3.	F	R	5+9	12+8	Klippel-Feil	WP	+	III	I	110	135	1.6
4.	M	R	5+2	9+4	Congenital Scoliosis	MW	+	II	I	100	150	3.2
5.	F	L	6+8	11+2	Klippel-Feil, Congenital Scoliosis	MW		IV	II	90	140	1.2
6	M	R	8+5	12+6	None	WP		III	II	105	140	2.2
7.	M	R	9+4	13+5	Congenital Scoliosis, diastematomyelia, rib deformity	MW		III	I	90	145	1.6
8.	F	R	4+6	10+6	Klippel-Feil,	WP		III	I	90	150	3.2
9.	L	L	7+5	9+8	Klippel-Feil	MW		III	II	100	140	2.4
10.	F	R	15+1	18+6	None	WP		III	I	90	160	3.4
11.	L	R	4+9	7+2	Congenital Cervicothoracic Scoliosis	MW		III	I	90	150	2.4
12	M	R	11+4	16+7	Congenital Scoliosis.	WP	+	III	I	100	160	1.8
13.	F	R	9+6	14+9	Klippel-Feil, Congenital Scoliosis, rib deformity	WP		III	II	90	145	3.0
14.	F	L	6+5	10+7	None	MW	+	III	I	95	150	2.8
Average			7+8	12+4				3.07 ± 0.26	1.28 ± 0.46	95 ± 6.78	147.85 ± 8.01	2.35 ± 0.72
P-valuate								0.00009		0.00009		

Table 1: *Details of 14 patients with modified and standard Woodward procedures for Sprengel deformity.*

MW: Modified Woodward Procedure; WP: Woodward Procedure; OVB: Omovertebral Bone; F-U: Follow-Up; L: Left; R: Right.

There are Female: 8 and Male: 6; Age at surgery: 7+8 and age at lase Follow-up: 12+4; Type of Surgery: Woodward procedure: 6 and Modified Woodward procedure: 8; Omovertebral bone: 5 (35.7%); Cavendish grade at Pre-operation: 3.07 (SD = 0.26) and last Follow-up: 1.28 (SD = 0.46) P-valuate = 000009; Motion (°) at Pre-operation: 95° (SD = 6.78) and at Postoperation: 147.85° (SD = 8.01) P-valuate = 000009; Scapular lowering on X-ray (cm): 2.35 (SD = 0.72).

Case	Age at surgery	Age at Lase F-U	Cavendish grade		Motion (°)		Scapular Lowering on X-ray (cm)
			Preop.	Last F-U	Preop.	Postop.	
3	5+9	12+8	3	2	110	135	1.6
6	8+5	12+6	3	1	105	140	2.2
8	4+6	10+6	3	1	90	150	3.2
10	15+1	18+6	3	1	90	160	3.4
12	11+4	16+7	3	1	100	160	1.8
13	9+6	14+9	3	1	90	145	3.0
Average SD	9+1 SD = 46.10	14+3 SD = 35.34	3 0	1.16 SD = 0.40	97.5 SD = 8.80	148.33 SD = 10.32	2.53 SD = 0.76
P-valuate	0.00000		0.00000		0.00000		

Table 2. *Woodward procedure.*

F-U: follow-up; L: left; R: right; Preop.: Pre-Operation; Postop.: Post-Operation.

Age at surgery: 9 years, 1 months; Age at Lase Follow-up: 14 years, 3 months; P-valute = 0.00000.

Cavendish grade at Pre-operation: 3 (SD = 0), Last Follow-up: 1.16 (SD = 0.40); P-valute = 0.00000.

Motion at Pre-operation: 97.5 (SD = 8.80), Post-Operation: 148.33 (SD = 10.32); P-valute = 0.00000.

Scapular Lowering on X-ray (cm): 2,53 (SD = 0.76).

Case	Age at surgery	Age at Lase F-U	Cavendish grade		Motion (°)		Scapular Lowering on X-ray (cm)
			Preop.	Last F-U	Preop.	Postop.	
1	10+6	15+1	3	2	90	145	1.5
2	4+2	12+4	3	2	90	160	2.6
4	5+2	9+4	3	2	100	150	3.2
5	6+8	11+2	4	1	90	140	1.2
7	9+4	13+5	3	1	90	145	1.6
9	7+5	9+8	3	1	100	140	2.4
11	4+9	7+2	3	1	90	150	2.4
14	6+5	10+7	3	1	95	150	2.8
Average	6+8	11+1	3.13	1.38	93.13	147.5	2.21
SD	SD = 26.62	SD = 0.35	SD = 0.35	SD = 0.52	SD = 4.58	SD = 6.54	SD = 0.70
P-valuate	0.001128		0.000048		0.00000		

Table 3: *Modified Woodward procedure.*

F-U: follow-up; L: left; R: right; Preop.: Pre-Operation; Postop.: Post-Operation

Age at surgery: 6 years, 8 months; Age at Lase Follow-up: 11 years, 1 months; P-valuate = 0.001128.

Cavendish grade at Pre-operation: 3.13 (SD = 035), Last Follow-up: 1.38 (SD = 0.52); P-valuate = 0.000048.

Motion at Pre-operation: 93.13 (SD = 4.58), Post-Operation: 147.5 (SD = 6.54); P-valuate = 0.00000.

Scapular Lowering on X-ray (cm): 2,21 (SD = 0.70).

Highly cited papers Comparing between original and modified woodward procedure

Original and/modified Woodward procedure: 1. Age at Surgery: 9+1 (SD = 46.10)/6+8 (SD = 26.62); P-valuate: 0.00000; Age at Lase Follow-up: 14+3 (SD 35.34)/11+1 (SD = 0.35); P-valuate: 0.10928; 2. Cavendish grade at Pre-operation: 3 (SD = 0)/3.13 (SD = 0.35); P-valuate: 0.00000; Cavendish grade at Last Follow-up: 1.16 (SD = 0.40)/1.38 (SD = 0.52): P-valuate: 0.560689 (not significantly different); 3. Motion (°) at Pre-operation: 97.5 (SD = 8.80)/93.13 (SD = 4.58); Pvaluate:0.005416; Motion (°) at Post-operation: 148.33 (SD = 10.32)/147.5 (SD = 6.54); P-valuate: 0.596971 (not significantly different); 4. Scapular Lowering on X-ray (cm): 2.53 (SD = 0.76)/2.21 (SD = 0.70); P-valuate: 0.496 (not significantly different).

Post-operatively, Comparing outcome of cosmetic and function between original and modified Woodward procedure were not significantly different.

Illustration

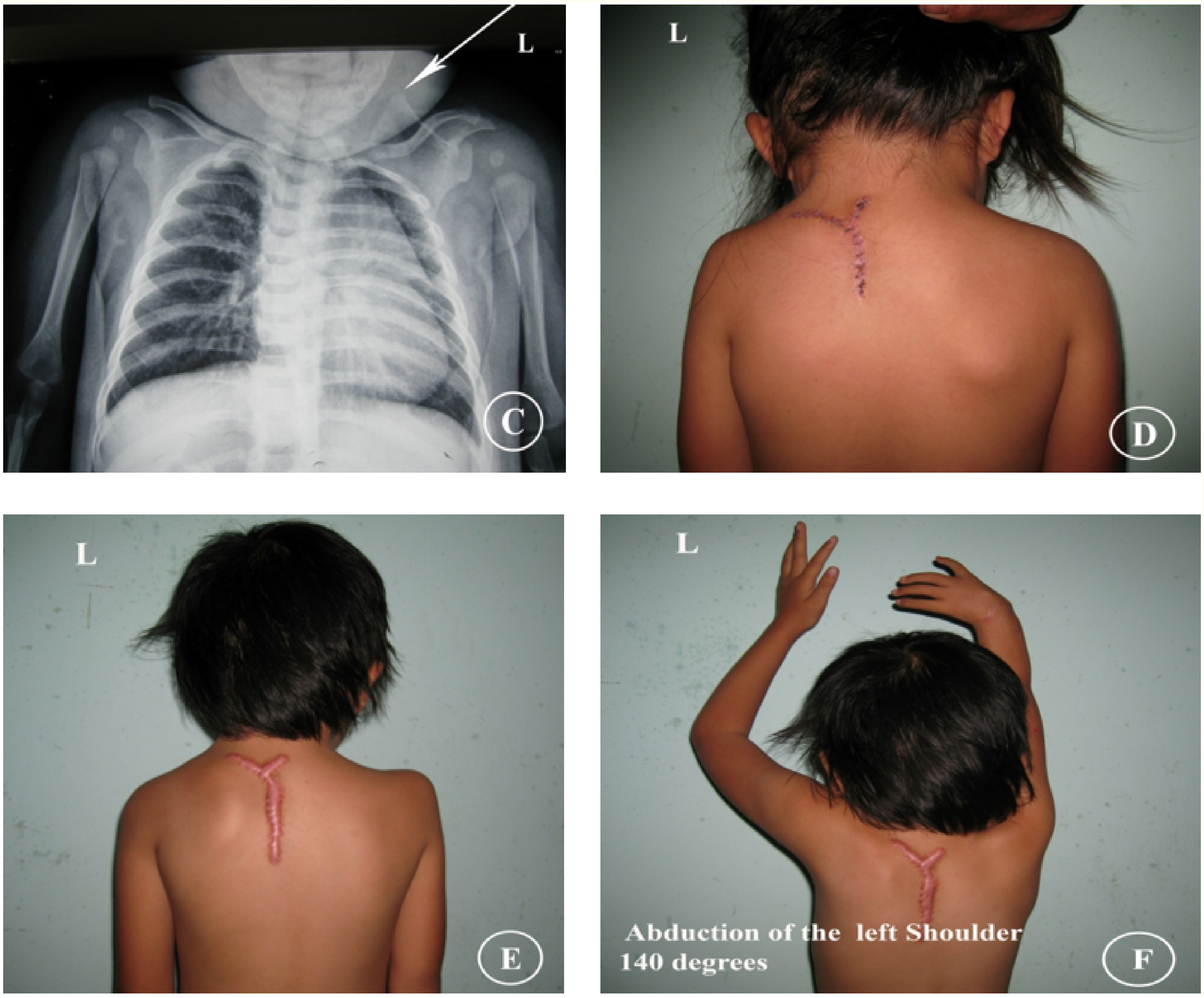

Figure 7: *Girl, Age at Operation was 4 years, 6 months. A. Preoperative with Cavendish grade 3; B. Abduction of the Shoulder was 95°; C. Roentgenography shown high left Scapula. D. Postoperative 3 weeks, Cavendish grade 1. - Postoperative 4 years 8 months, E. Cavendish grade 1; F. Abduction of the Shoulder was 140°..*

Complications

There was One patient was felt to have had an exaggeration of the scapular winging, had been present before operation; no specific information could be gleaned from the notes as to any injury during operation. This complication has also been reported by Carson., et al. [16] who recommended that scapular winging before operation should be considered a relative contra-indication to this Woodward procedure. There were no infections.

Discussion

Several theories have been put forward to explain this arrest in the development process [5,16], but none offers convincing evidence. However, it is clear from the embryology that the deformity should be referred to as an "undescended" scapula rather than the popular term "elevated" scapula. It should also be appreciated that it is more than an isolated problem of the scapula.

Sprengel deformity is the most common congenital deformity of the shoulder [6]. It is believed to occur as a result of interruption of the normal caudal migration of the scapula, probably occurring between the ninth and twelfth week of gestation [5,11]. The scapula is hypoplastic, and the ratio of its horizontal width to its vertical height is increased. The muscles involved with scapular movement, especially the trapezius, elevator scapulae, and rhomboids are often small, weak, and known to be infiltrated with fibrofatty tissue [15]. The resulting deformity can be a cosmetic concern and is sometimes associated with functional impairment.

The incidence and aetiology are unknown. Most cases occur sporadically, although there may be an autosomal dominant trait [3]. The deformity is due to a failure of descent of the scapula between the ninth and 12th weeks of intrauterine life. The scapula is originally located as a sagittally orientated structure in the neck. It then migrates to a coronally aligned posterior thoracic position. Failure of descent leaves it in an elevated, malrotated position. The periscapular muscles are hypoplastic; fibrous bands tether the scapula to the chest wall and further limit rotation. The bone is small, broad and rotated with the inferior angle medialised and the glenoid directed caudally. The superior angle is attached to the cervical spine by an omovertebral bone or band, Omovertebral bone: 5 (35.7%) in this study (Table 1). Associated abnormalities of the cervical spine are commonly found.

Soccurs as the result of an interruption of the normal prengel's deformity is an uncommon disease, which caudal migration of the scapula during development, most likely between the ninth and twelfth weeks of gestation [4]. 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 [1].

Classification

Cavendish grading is widely used and recommended for classification of the universe in the deformation of Sprengel. Grade 1 is the lightest, where shoulder is almost level and it cannot be noticed with clothes on. Grade 2 is also light, but the superficial portion of the jaw bone can be seen as a lump. In grade 3, the distortion is moderate, visible and the shoulder is affected by two to five centimeters higher than the normal shoulder. In grade 4, the distortion is severe, the jawbone is very high, with a supernatural angle in the bud, with antique cloth and brevicollis. One limitation of this classification is difficult to apply in bilateral cases.

Cavendish classification

Sprengel's distortion often appears with impaired function and aesthetics. In accordance with current literature, we use the Cavendish classification [17] for objective evaluation of surgical treatment (Table 4).

Vendish Score	Sprengel's deformit
Grade 1 - Very mild.	Shoulder joints are level. Deformity is invisible when patient is dressed.
Grade 2 - Mild.	Lump in the web of the neck. Deformity is visible when dressed
Grade 3 - Moderate.	Shoulder elevation 2-5 cm. Deformity is easily visible.
Grade 4 - Severe	Superior angle of the scapula is near the occiput with/without neck webbing or brevicollis

Table 4: *Cavendish classification [6].*

This classification is easier to use and more accurately evaluate the number of distortions and improvements after surgery. Cavendish in a series of 100 cases from the hospital series in the United Kingdom showed moderate to severe distortion in 41 and mild deformity in 59 cases. Errors in the diagnosis can occur although the main features of the condition are known. Klippel-Feil syndrome, polio and scoliosis of Erb are some of the conditions that cause diagnostic difficulties. It can be quite easily differentiated if appropriate clinical and radiological tests are performed.

The children affected by the Sprengel distortion have reduced the range of motion (ROM) and different functional limits due to shoulder joint. Lack of movement of Scapulothoracic junction and poor reversal results reduce decreased shoulder abduction. Shoulder abduction are usually limited to 90°. These Limitations affect many activities of daily life. It is not easy to classify a distortion when it is two faces and defects of the same level. To simplify the signs For treatment and description of the results, the following classes were constructed by Cavendish in 1972. Grade 1 - Very light, shoulder joint is level, deformity is Invisible when the patient is dressed. Grade 2 - Small tumor on the neck and Deformation can be seen when wearing clothes. Layer -3 will have a shoulder height of ~ 2 - 5 cm and visible deformation. Grade -4 is serious and shoulders are very high so the upper scapula of the scapula near the occiput, with or without the collar or brevicollis. Cavendish is used as a measure of improvement and cosmetic range Kidney abduction is a measure of function [17]. Scoliosis is found The most common abnormalities in the two large patient populations included 112 patients and 75 patients (35% and 55% respectively). Flat ribs are also common, with reports rates of 16% to 48% in patients with Sprengel deformities. Other links Disorders include diastematomyelia.

Rigualt Score

Any patient with Sprengel's deformity requires an initial radiation assessment evaluation. It should be obtained initially to assess the extent of the scapula involved vertebrae and face to face. It is also useful to determine the presence of related abnormalities (e.g. scoliosis, abnormal ribs, omovertebral bone). Rigault's classification is described by Rigualt., *et al.* based on the supersonic projection on the X-ray [18,19]. It is used to assess the degree of scapular radiographically verifiable pre-op and post-operatively. It's a simple, easy-to-calculate and widely used score. This score does not have any correlation with the Cavendish score

Radiological evaluation includes measurement of superior transposition rates and height-to-width ratio of the jaw in the rear posterior view from preoperative images such as chest X-rays compared to post-op images. Scapula location and degenerative disease will be assessed by radiography. The "Scapular Index" proposed by Broca describes the relationship between the vertical length and the horizontal width of the scapula. This index is expressed by the formula (100 width/length) and is calculated to be between 63 - 71 [20].

Plain radiography may be used to assess the distortion and presence of omovertebral communication, and to note postoperative correction. The distance between the lower and higher angles of the jaw and spine, and the angle of inclination of the glenoid, to evaluate the postoperative outcomes described by Armitage [21] are not widely used. Based on the forward radiographic image and the relationship of the superior angle to the spine, Rigault proposed a classification to assess the distortion between the superior angle of the jaw and spine.

A radiofrequency classification for Sprengel distortion was developed by Rigault., *et al.* Based on radiography - the projection of the subliminal angle of the jaw. Deformities are classified based on the relationship between the subliminal angle of the jaw and the relative level of life. Grade 1 deformation deals with superlative angles under T1, Grade 2 deformation lies between T1 and C5, and Grade 3 deformation lies on C5.

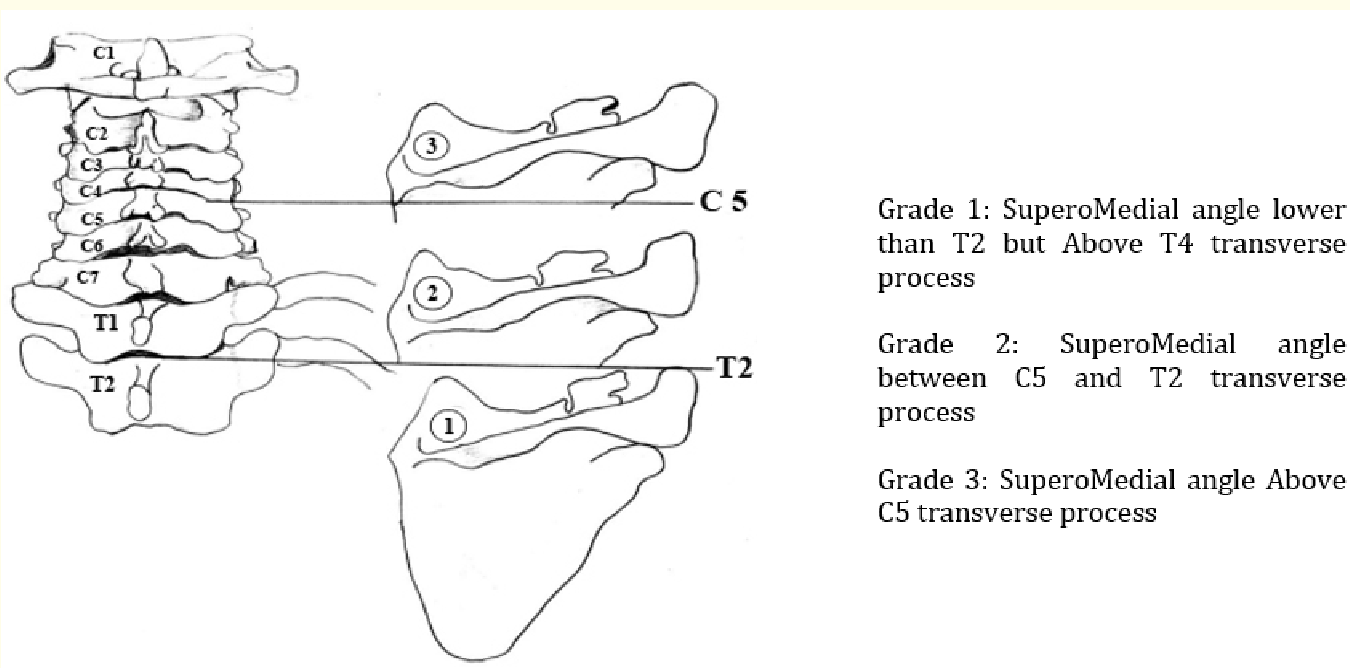

Figure 8: *Rigault’s radiological classification of the Sprengel’s deformity of the shoulder [13].*

Another classification described by Ross and Cruess measured shoulder elevation based on the level of the center of the humeral head in relation to the vertical axis trunk. Leibovic., *et al.* Sprengel's deformity has a rotational component, and scapular displacement is based on the vertical positioning of the scapula and the rotational component, assuming that center of rotation of the scapula was through acromioclavicular joint [15].

To reveal the scapula position, (early) degenerative changes, and the anatomical differences between both shoulders standard radiographs of antero-posterior view of both shoulders with chest X-ray was done. In patients who had an analgesia, the measurements of 'Superior displacement ratio' was done by drawing two reference lines. Line 1 will be drawn from the center of the glenoid cavity of the affected shoulder perpendicular to the vertebral axis line, and line 2 was drawn from the normal shoulder.

Examination of the shoulders and both upper limbs, spines, and lower limbs were done by giving attention to scapular bony prominences, scapular winging, scoliosis, chest asymmetry, or any associated morphological abnormalities. All patients were examined for the grade of deformity. The forward flexion and abduction movements of the shoulders were examined, as well as looking for a range of motion especially of the scapulo-thoracic motion, to determine whether the scapula was anchored to the spine or not.

Treatment

Nonsurgical management is reserved for children with mild deformity, minimal shoulder dysfunction, and little cosmetic deformity (e.g. Cavendish grades 1 and 2). The documentation data is set to the ROM and prevent torticollis. Do their we can thiệp, management include the observation of year to visit when the human to the major to bone, for evaluation of the progression of shoulder abnormality and associated deformities.

The current export is disclaim the display font, and special note must be returned for development of young. Farsetti., et al. [22] performed a retrospective of 15 bệnh nhân not implemented (17 scapulas). Three were Cavendish grade 1, ninth is grade 2 and five grade 3. The mean preoperative combined shoulder abduction was 125° (range, 95° to 170°). The tracking medium 26 year (interval of 10 to 55 years), average medium abduction is 125° (range, 90° to 160°), and all human variable the class 1 and 2 of the Cavendish is deformity remained in their respective categories

The age of surgical intervention was discussed by different authors [4,6,11,23]. Some authors have suggested that surgery in patients under 3 years of age is technically more difficult [4,23]. Other authors [4,6] have stated that patients over 6 years of age are not eligible candidates for a scaled-down procedure. The recommended age for surgery is 3 to 8 years. However, this is not proven by the available data due to the limited number of cases in most reporting sequences. In our study, it is difficult to determine the criteria for determining the optimal age to adjust effects. The age of our patients at the time of procedure varies from 4 to 17 years (mean, 8 years 3 months). We did not find any significant difference in any of the indicators evaluated when comparing our patients over 7 years of age with those under 7 years of age at the time of surgery.

The recommended age for adjusting the impact of this distortion is 3 - 5 years according to Woodward (1961) and under six years according to Jeannopoulos [4] and Cavendish [6]. It has been suggested that in patients younger than three years anatomy is more difficult to identify and procedures therefore become technically more difficult. Elderly patients have less soft and hard tissue, less adaptable to changes in position. In addition, older children may have become socially and psychologically hurt.

Optimal age to adjust effects is not proven in the current series. However, it has been shown, that age is no longer a specific sign. Successful implantation with good long-term outcomes in children up to age 15. Other authors have stated that children over the age of six are not suitable for such procedures [4,6]. However, we recommend that you make edits between the ages of three and eight years; At this age, surgery is fully developed and a bio-plastic state is still available to accommodate the changes caused by the operation.

Optimal age to adjust effects is not proven in the current series. However, it has been shown, that age is no longer a specific sign. Tracheal implantation has been successfully performed with good long-term outcomes in children up to 15 years (Figures 10 and 11). Other authors have stated that children over the age of six are not suitable for such procedures [4,6]. However, we recommend that you make edits between the ages of three and eight years; At this age, surgery is fully developed and a bio-plastic state is still available to accommodate the changes caused by the operation.

Eulenberg has been noted as the first to describe the congenital elevation of the scapula bone, commonly referred to as the Sprengel deformity of the shoulder [16]. This is an uncommon deformity that occurs due to interruption of normal jaw movement in the developmental process [4]. Several hypotheses have been proposed to explain this failure during development, but the exact cause is not known [5]. Cosmetic deformation is a major concern and sometimes distortion accompanies functional decline. Abduction of the involved shoulder usually limited to the severity of anger, but functional defects are usually minimal.

Various procedures have been described to improve the appearance and, in some cases, the role of the shoulder [4,9,24-27]. Putti [26] describes a process in which the upper projection of the jaw bone is removed and pulled down after dividing the attachment of the muscle. These principles were modified by Schrock [8] and then by Green [7] to describe a new process for correcting deformation. The result is disappointing when the deformity recurs either by returning the Scapula to its former position or by regenerating the bone after subperiosteal removal [6]. McFarland [25,27] recommends removal of Scapula, especially in severe cases; However, technical difficulties, problems with scars, and functional impairment are major complications. Woodward [10] describes a process in which correcting distortion is obtained by moving the origin of the trapezium and rhombus down the spine. He reported the results of nine cases with follow-up ranging from 9 months to 5 years. The initial results are encouraging and few complications.

Apparently from reviewing the documentation that the short-term outcome of the Woodward procedure is satisfactory; However, long-term studies on the topic are lacking. When surgery is usually done during childhood, the effect of growth on corrective surgery is a great concern. Grogan., *et al.* [15] reported a series of 20 patients with congenital healing of osteoarthritis treated by the Woodward procedure. Only five adults are adults and 14 years older. Carson., *et al.* [12] reported on 11 cases of Sprengel deformities that were handled by Woodward procedures and again tracked as past matures with only five patients.

A further option is a vertical scapular osteotomy, first described in 1914 by König [28] as a case report, in which he performed a vertical cut to the bone less than 4 cm. He fixed the Scapula bone's body in its new position by drilling a hole at its base, then repeating a latissimusdorsi tube through it and fixing it into itself. Later in the article, he considered this fixation a mistake because it limited bone turnover and thus kidnapped the arm. No other report of the activity was made until Wilkinson and Campbell in 1980 [29].

Deformity of Sprengel is the most common congenital deformity of the shoulder [9]. Since 1908, various procedures have been described to improve Sprengel's disfigurement and shoulder function. Previous surgical outcomes are summarized in table 1, along with patient data from this study. It has been noted that the Woodward surgical technique [17] is most common for use with Sprengel's deformed patients. Borges [30] described a modified Woodward procedure, which included the removal of the medial border of the Scapula bone to allow for better orientation of the shoulder. This method produces significant improvement in appearance, assessed on the Cavendish scale, but does not seem to significantly improve shoulder function. We modified the Woodward procedure with the removal of the Supero-Medial bone with the Omovertebral bone removed. Improvements in ranged from an average of 93.13° to 147.5° (Table 3).

Deformities in cosmetics are a major concern and are often associated with functional impairment, but not all patients with Sprengel's disfigurement guarantee surgical intervention. In clinical applications, according to the Cavendish classification, not class 1 also minimizes the function of ensuring an operation. For grade 2, if aesthetic deformities are not accepted, surgical treatment is indicated. Grafts 3 and 4 often require surgical intervention because of severe functional impairment and aesthetic decline. Rigault's classification [12] is based on radioactivity, and seems more consistent and objective than the Cavendish classification system. For aesthetic reasons, we prefer to use the Cavendish class rather than the Rigault classifier in the patient's clinical judgment because on radiography, it is difficult to measure deformity. Although the Cavendish classification is too subjective and not accurate enough, it is morphologically, and is proven to be easy to use for evaluating overall improvement in our patients.

When considering the timing of surgical intervention, the recommended age for early treatment (before age 6) according to the Woodward procedure is no pelvic surgery or improved method, suitable for corrective surgery and the effect of moderate to severe Sprengel deformation.

Author	Case	Mean age at operation (years)	Function before operation (°)	Function after operation (°)	Grade before operation	Grade after operation	Operative technique
Cavendish [6]	34	7.2	140	154	3.1	1.7	All methods in common use
Woodward [10]	9	8.1	140	172			Woodward procedure
Borges [30]	15	8.3	115	150	3	1.53	Modified Woodward procedure
Wilkinson [3]	12	8.4	118	164	3	1.33	Scapular osteotomy
Corha [32]	18	7	145	151.6	2.55	1.88	Woodward procedure
McMurtry [37]	12	8.1	88	132	3	1.5	Scapular osteotomy
Grogan [15]	20	6.5	110	147			Woodward procedure
Jung [33]	9	4.7	82	160	3.56	1.33	Woodward procedure
Hung (this study)	14	7.8	95	147.85	3.07	1.28	Woodward and Modified Woodward procedure

Table 5: *Some authors, operative techniques and results.*

Cases number: 9-34; Mean age at operation(years): 4.7-8.4; Function before operation (°): 62-145; Function after operation (°): 132-171; Grade before operation: 2.55-3.56; Grade after operation: 1.25-1.7; Operative technique: All methods in common use.

Some authors [6,31] recommend clavicular osteotomy as a routine part of the procedure to reduce the risk of neurovascular compression that can occur between the collarbone and the distended chest wall. We agree with Grogan., et al. [15] As long as the adjustment was not too strong, the operation of the bone with the clavicle was not necessary to achieve significant improvement. However, in our series, clavicle osteotomy was used in a patient who developed a brachial plexus failure after procedure. This patient had a severe cosmetic deformity (Class IV Cavendish) and moderate functional impairment of the shoulder abduction.

We do not recommend a clavicular surgery as a regular part of the procedure. Although the collarbone is a component of deformity, it is often not necessary to osteotomies it to achieve significant improvement. Our series shows that as long as the correction attempt is not too strong, it works quite safely. We used clavicle osteotomy in patients who needed more adjustment, and an additional step was indicated to reduce the risk of neurovascular compression that may occur between the collarbone and deformed chest wall [31]. We didn't performing clavicular surgery in this study,

Complications

Complications due to surgical management of Sprengel deformities are largely due to hypertrophic scars, recurrence of the maxilla of the scapula bone, brachial plexus Injury and winged arms. Most surgical techniques involve large incision and extensive dissection, which can lead to hypertrophic scars and keloids. In the long term follow-up study by Farsetti., *et al.* [22] which included 22 patients, 4 of 8 patients treated surgically was found to have an unsightly scar.

Ahmad [34] reported keloid scarring in 4 out of 11 patients. The brachial plexus is at risk of compressing intraoperatively as the scapula is moved inferior. Some authors have reported plexus brachial paralysis after surgery for Sprengel deformity; however, many palsies is transient [14,35,36]. However, we think that the clavicle osteotomy should be considered to prevent brachial plexus injury. Intraoperative somatosensory-evoked potential monitoring may help to prevent such injury [37]. Other postoperative outcomes include re-winging [31] of deformity, and failure of functional improvement.

There was One patient was felt to have had an exaggeration of the scapular winging, had been present before operation; no specific information could be gleaned from the notes as to any injury during operation. This complication has also been reported by Carson., *et al.* [12] who recommended that scapular winging before operation should be considered a relative contra-indication to this Woodward procedure. There were no infections.

Conclusion

Sprengel deformity is the most common congenital malformation of the shoulder joint and can cause significant functional impairment and functional decline. Proper diagnosis is necessary because the condition can be manifested by a variety of abnormalities associated with significant morbidity, if not properly managed. Initial evaluation should include plain radiography as well as two and three dimensional CT to assist in surgical planning and to determine the presence of omovertebral bone. Non-surgical management is appropriate for mild cases, with surgery for severe cases. Some surgical techniques have been developed with the goal of improving cosmesis and shoulder function. In general, these procedures involve removing the protrusion of the scaphoid bone along with any omovertebral bone as well as the scapula bone under a caudad position.

Woodward procedures are the most widely reported procedures. Hypertrophic scars can occur as a result of large incisions and large scars. Good clinical results can be achieved after surgery, with significant improvement in appearance and shoulder movement. Prior to treatment, the surgeon should talk to the family about the expected results and potential complications

Based on the review of the literature and the results of this series, the Woodward procedure appears to be the best surgical procedure available to correct congenital scapula bone.

Limitations of the Study

Limitations of this study include the small cohort of patients and the postoperative results was too small to perform parametric statistical analysis. Second, this is an interim outcome report as most patients were not followed-up until skeletal maturity. Third, the subjects of this study were not homogenous in terms of the data used. Fourth, compare results between cases with and without modified Woodward procedures all equally value.

Bibliography

1. Eulenberg M. "Beitrag zur dislocation der scapula. Amlicht ber deutscher N aturforsch". *Aerztinnen-Karlsbad* 37 (1863): 291-294.
2. Sprengel RD. "Dieangeborene vershiebung des schulterblattes nach oben". *Langenbecks Archiv fur Klinische chirurgie* 42 (1891): 545-549.

3. Wilson MG., *et al*. "Dominant inheritance of Sprengel's deformity". *Journal of Pediatrics* 79.5 (1971): 818-821.
4. Jeannopoulos CL. "Congenital elevation of the scapula". *Journal of Bone and Joint Surgery-American Volume* 34.4 (1952): 883-892.
5. Horwitz AE. "Congenital elevation of the scapula-Sprengel's deformity". *The Journal of Bone and Joint Surgery* 6 (1908): 260-311.
6. Cavendish ME. "Congenital elevation of the scapula". *Journal of Bone and Joint Surgery-British Volume* 54B.3 (1972): 395-408.
7. Putti V and Beitrag ZA. "Pathogenese und Behandlung des angeborenen hochstandes der schulterblattes". *Fortschr Geb Rontgenstrahlen* 12 (1908): 328-349.
8. Schrock RD. "Congenital elevation of the scapula". *Journal of Bone and Joint Surgery-American Volume* 8.1 (1926): 207-215.
9. Green WT. "The surgical correction of congenital elevation of the scapula (Sprengel's deformity)". *Journal of Bone and Joint Surgery-American Volume* 39 (1957): 1439.
10. Woodward JW. "Congenital elevation of the scapula: Correction by release and transplantation of muscle origins. A preliminary report". *Journal of Bone and Joint Surgery-American Volume* 43 (1961): 219-228.
11. Carson WG., *et al*. "Congenital elevation of the scapula. Surgical correction by the Woodward procedure". *Journal of Bone and Joint Surgery-American Volume* 63.8 (1981): 1199-1207.
12. Rigault P., *et al*. "Sur elevation congenital de l'omoplate chez l'enfant. Etude anatomopathologique et therapeutique a propos de 27 observations". *Revue de Chirurgie Orthopédique et Réparatrice de l'appareil Moteur* 62 (1976): 5-26.
13. Ross DM and Cruess RL. "The surgical correction of congenital elevation of the scapula: A review of seventy-seven cases". *Clinical Orthopaedics and Related Research* 125 (1977): 17-23.
14. Leibovic SJ., *et al*. "Sprengel deformity". *Journal of Bone and Joint Surgery-American Volume* 72.2 (1990): 192-197.
15. Grogan DP., *et al*. "The congenital undescended scapula. Surgical correction by the Woodward procedure". *Journal of Bone and Joint Surgery-British Volume* 65.5 (1983): 598-605.
16. Engel D. "The etiology of the undescended scapula and related syndromes". *Journal of Bone and Joint Surgery* 25.3 (1943): 613-625.
17. Susan Standring. Gray's Anatomy: The Anatomical Basis of Clinical Practice, Expert Consult-40th Edition (2008).
18. Bianca S., *et al*. "Isolated congenital amastia: a subclavian artery supply disruption sequence?" *American Journal of Medical Genetics Part A* 152A.3 (2010): 792-794.
19. Khairouni A., *et al*. "Congenital high scapula". *Journal of Pediatric Orthopaedics B* 11.1 (2002): 85-88.
20. Rutherford NC. "A Contribution to the Embryology of the Fore-Limb Skeleton". *Journal of Anatomy and Physiology* 48.4 (1914): 355-377.
21. Armitage BM., *et al*. "Mapping of scapular fractures with three-dimensional computed tomography". *Journal of Bone and Joint Surgery-American Volume* 91.9 (2009): 2222-2228.
22. Farsetti P., *et al*. "Sprengel's deformity: Long-term follow-up study of 22 cases". *Journal of Pediatric Orthopaedics B* 12.3 (2003): 202-210.
23. Jeannopoulos CL. "Observations on congenital elevation of the scapula". *Clinical Orthopaedics* 20 (1961): 132-137.
24. Ehrlich MG. "Shoulder problems in children". In: Rowe CR, ed. The shoulder. New York: Churchill-Livingstone (1988): 455-470.
25. McFarland B. "Congenital deformities of the spine and limbs". In: Platt H, ed. Modern trends in orthopaedics. London: Butterworths (1950): 107-137.
26. Putti V. "Beitrag zur atiologie, pathogenese und Behandlung des angeborenen hochstandes der schulterblattes". *Fortschr Geb Rontgenstrahlen* 12 (1908): 328-349.
27. Smith AD. "Congenital elevation of the scapula". *Archives of Surgery* 42 (1941): 529-536.

28. König F. "Einc neue operations des angeborenem sculter blattochstandes". *Zentralblatt Fur Chirurgie* 40 (1914): 530-537.

29. Wilkinson JA and Campbell D. "Scapular osteotomy for Sprengel's shoulder". *Journal of Bone and Joint Surgery-British Volume* 62B (1980): 486-490.

30. Borges JL. "Modified Woodward procedure for Sprengel deformity of the shoulder: long-term results". *Journal of Pediatric Orthopaedics* 16.4 (1996): 508-513.

31. Robinson RA., *et al*. "The surgical importance of the clavicular component of Sprengel's deformity". *Journal of Bone and Joint Surgery-American Volume* 49 (1967): 1481.

32. Crha B. "Surgical treatment of Sprengel's deformity of the scapula". *Scripta Medica (BRNO)* 74.4 (2001): 245-254.

33. Jung WS., *et al*. "Clinical Experience of the Woodward Procedure in Sprengel's Deformity". *Formosan Journal of Surgery* 43 (2010): 81-86.

34. Ahmad AA. "Surgical correction of severe Sprengel deformity to allow greater postoperative range of shoulder abduction". *Journal of Pediatric Orthopaedics* 30.6 (2010): 575-581.

35. Andrault G., *et al*. "Green's surgical procedure in Sprengel's deformity: Cosmetic and functional results". *Orthopaedics and Traumatology: Surgery and Research* 95.5 (2009): 330-335.

36. McMurtry I., *et al*. "Osteotomy for congenital elevation of the scapula (Sprengel's deformity)". *Journal of Bone and Joint Surgery-British Volume* 87.7 (2005): 986-989.

37. Shea KG., *et al*. "Somatosensory evoked potential monitoring of the brachial plexus during a Woodward procedure for correction of Sprengel's deformity". *Muscle Nerve* 41.2 (2010): 262-264.

Volume 10 Issue 2 February 2019