

# Treatment of congenital undescended scapula with Woodward operation, the functional and cosmetic outcomes

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## Abstract

**Background:** Although congenitally undescended scapula (Sprengel's deformity) reported to be rare, yet many families still seek medical help for their children. Cases with congenitally undescended scapula were collected, evaluated, and the majority subjected to surgical treatment with Woodward operation.

**Objective:** Is to demonstrate the functional in addition to cosmetic improvements obtained with Woodward procedure used for Sprengel's deformity.

**Methods:** Between December 2007 and November 2009, fifteen patients with Sprengel deformity were evaluated (all have unilateral deformity) and treated surgically with Woodward procedure. The study included 9 males and 6 females, their age range from 5 to 11 years; right side is affected in 8 patients and left side in 7 patients. The preoperative grade of the deformity (according to Cavendish classification) and the maximum shoulder

abduction were reported to assess severity and to be compared with the postoperative results.

**Results:** All the patients get improvement in scapular elevation by about 2 grades and the increase in the maximum combined abduction was 20.33° in average. Two patients developed transient brachial plexus palsy. An omo-vertebral connection was found in 10 cases, in 3 cases the connection was bony, another 3 cases was cartilaginous, and 4 cases were fibrous.

**Conclusions:** children with grade 2 and more and especially if abduction is much affected need to be treated with surgery, Woodward operation seems to be effective in handling both the cosmetic concern and the functional impairment.

**Keywords:** Sprengel deformity, undescended scapula, Woodward.

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## Introduction

Congenital Undescended Scapula is also widely known as Sprengel's deformity, it is uncommon congenital anomaly<sup>(1,2)</sup>. In this deformity, the scapula is elevated, hypoplastic, and usually somewhat adducted. Almost without exception, there are associated anomalies of the cervical and thoracic spine, as well as the ribs<sup>(3)</sup>. Attention was first directed to this deformity by Eulenberg, in Germany, in 1863. In 1891, Sprengel, in Germany again, drew attention to this deformity by describing another 4 cases.

Kolliker, who also described 4 cases in 1891, gave the condition its eponym, Sprengel deformity<sup>(4-6)</sup>. Recently a historical study on a mummified fetus, from ancient Egyptian civilization believed to be the daughter of king Tutankhamun, have shown to have scoliosis, spina bifida, and Sprengel deformity<sup>(7)</sup>.

## **Frequency**

Although Sprengel deformity is the most common congenital malformation of the shoulder girdle and some refers to prevalence of <1 per 10,000, the exact prevalence is not known<sup>(6, 10-13)</sup>. The male to female ratio is variable with different studies but most of these refer to equal involvement, however some attribute the slight female predominance to their parents' cosmetic concerns.<sup>(14,15)</sup> Right and left sides affection is of

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variable figures in different articles, and bilateral involvement occurs in 10% to 30% of the cases<sup>(16-18)</sup>.

### ***Etiology***

The exact causation is not known but some hypotheses are available like: The exogenous theory, the bleb theory, and Subclavian artery supply disruption sequence hypothesis. The condition is sporadic. Rarely, it may run in families (autosomal dominant pattern of inheritance)<sup>(8, 9, 18, 21, 22)</sup>.

### ***Presentation***

The hallmarks of the Sprengel deformity are shoulder asymmetry and restriction of shoulder abduction. Clinically, the affected scapula is usually elevated 2-10 cm and is adducted, and its inferior pole is medially rotated. Due to this rotation, the glenoid faces inferiorly. A prominence in the suprascapular region is characteristic due to the upwardly rotated superomedial angle of the scapula, which causes the ipsilateral side of the neck to appear fuller and its normal contour to be lost. The scapula is hypoplastic, and the length of the vertebral border is decreased. Occasionally, some anterior bending of the supraspinous portion is present. The condition may sometimes be bilateral, in which case, although it is cosmetically much more acceptable, functionally, it is more disabling<sup>(23, 24)</sup>.

### ***Classification***

More than one classification has been introduced for Sprengel deformity but the Cavendish classification is the most currently used. Sprengel deformity can be classified as follows:

- **Grade 1:** very mild. The shoulder joints are level and the deformity is invisible, or almost so, when the patient is dressed.
- **Grade 2:** mild. The shoulder joints are level or almost level but the deformity is visible when the patient is dressed, as a lump in the web of the neck.
- **Grade 3:** moderate. The shoulder joint is elevated 2 to 5 centimetres. The deformity is easily visible.
- **Grade 4:** severe. The shoulder is much elevated, so that the superior angle of the scapula is near the occiput, with or without neck webbing or brevicollis<sup>(9)</sup>.

### ***Patients and methods***

Between December 2007 and November 2009, fifteen cases of congenital undescended scapula treated with Woodward procedure for correction of the deformity, 2 cases from Al-Kadhemiyah Teaching Hospital and another 13 cases from Al-Wasity Teaching Hospital for Reconstructive Surgeries. Table 1 show the distribution of the patients and their age, sex, and side of involvement.

**Table 1: Distribution of the patients and their age, sex, and side of involvement. The numbering of the cases was according to the alphabetical order of their first names. M for male, F for female, R for right, and L for left.**

Case №	Age (year)	Sex		Side	
1	6		F		L
2	5		F	R	
3	7	M			L
4	5	M			L
5	11	M			L
6	6		F	R	
7	5	M		R	
8	6		F		L
9	6	M			L
10	5	M		R	
11	11	M		R	
12	10	M		R	
13	7		F		L
14	6		F	R	
15	10	M		R	
		9	6	8	7

### ***Preoperative evaluation***

Preoperatively the patients were evaluated by detailed history and examination, taking in consideration the family history of similar problem and other skeletal anomalies and any problems during gestation. Examination of the shoulders and both upper limbs, spines, and lower limbs were made giving attention for scapular bony prominences, scapular winging, scoliosis, chest asymmetry, or any associated morphological abnormalities. All the patients were examined for the grade of the deformity. Movements of

the shoulders were examined as well as of elbow, wrist, and hand; looking for range of motion especially of the scapulo-thoracic motion, whether the scapula is anchored to the spine or not. Full neurological examination of both upper limbs was performed, comparing the sensation and muscle power of both sides. All of the patients subjected to anteroposterior view of both shoulders or Chest X-Ray, some needs radiograph of the cervical, thoracic, and lumbar spines. Most of the patients have associated anomalies (Table 2).

**Table 2: The associated deformities recorded in the 15 patients.**

<b>Associated deformity</b>	<b>Number of cases</b>
<b>Rib anomalies</b>	11
<b>Scoliosis</b>	5
<b>Toricollis</b>	3
<b>Spina bifida</b>	2
<b>Cervical spine fusion</b>	2
<b>Accessory nipple</b>	1
<b>Umbilical hernia</b>	1

### **Results**

All the patients have improvement of their shoulder deformities ranging from partial to complete correction in regard to shoulder level or bony prominence with an average of improvement by 2 grades, (Table 3). Most of the patients get significant improvement in abduction by an average of 20.33 degrees (0 – 45 degrees). Patients who had the omo-vertebral connection and especially if it is not fibrous were associated with much abduction limitation. This connection found to be 66.67 % of our patients, the bony and cartilaginous bar was 40 % (Table 4). We asked the parents to put their satisfaction regarding the improvement of their children after surgery in one of 4 levels (excellent, good, fair, and poor), (Table 5). The excellent and good satisfaction was 86, 67%; fair satisfaction was 13.33%, and no poor contentment.

Regarding the complications, one patient (case № 13) developed loss of

active abduction and loss of sensation over C5 dermatome in the involved side with preservation of sensation and motor activity of other regions of the same limb. This neurological deficit resolved spontaneously and completely within 6 weeks. Another case (case № 4) developed complete brachial plexus palsy, necessitate urgent clavicular morcellization (morcellation) next day. The patient have full recovery in 7 months during which vigorous physiotherapy was underwent, the resolution started proximally and proceeded distally. Nine patients have had wide scars with no keloid or hypertrophy. One case developed multiple discrete superficial infections treated with oral cefalexin and changing dressings. Some of the patients included in the study are shown below with their postoperative results (Figure 1, 2, 3, and 4).

**Table 3: The preoperative and postoperative values for elevation grade and maximum abduction (and the obtained gain) in addition to the average of these values.**

Case №	Omo-vertebral connection	Grade of undescending		Abduction (in degrees)		
		Before	After	Before	After	gain
1	Fibrous	4	2	100	130	30
2		4	2	120	130	10
3	cartilaginous	4	1	115	160	45
4	Fibrous	3	2	140	140	0
5	Fibrous	3	1	150	160	10
6	Cartilaginous	2	1	135	150	15
7	cartilaginous	3	1	120	140	20
8	Bony	3	0	90	135	45
9		3	2	130	140	10
10	Bony	3	1	85	130	45
11		3	2	125	135	10
12		3	0	135	145	10
13	Bony	3	0	115	155	40
14	Fibrous	3	1	135	140	5
15		3	1	140	150	10
Average	-	3.13	1.13	122.33	142.66	20.33

**Table 4: The percentages of all omo-vertebral connections and of their types.**

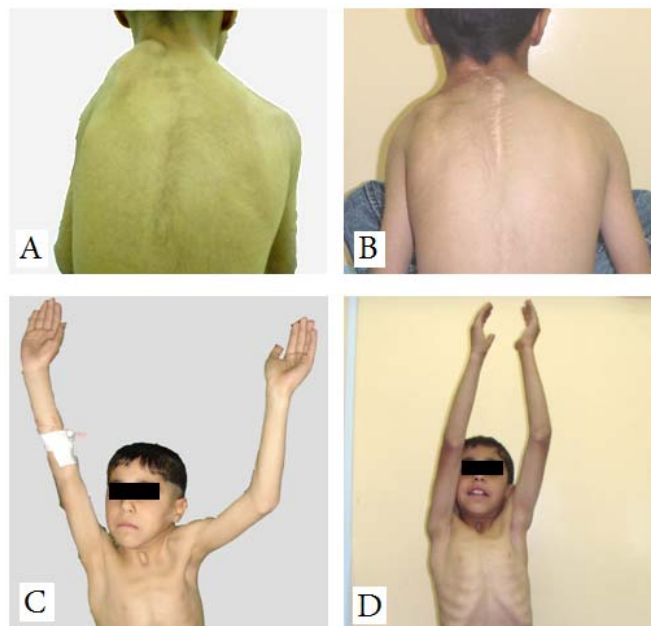
Type of connection	Number of cases	Percentage	
Bony	3	20	40
Cartilaginous	3	20	
Fibrous	4	26.66	
Total	10	66.66	

**Table 5: table of the parent satisfaction in 4 levels with no poor contentment revealed.**

Level of satisfaction	Number of parents	Percentage	
Excellent	4	26.67	86.67
Good	9	60	
Fair	2	13.33	13.33
Poor	0	0	



**Figure 1:** A, a preoperative X-ray of a patient with grade 3 deformity, note the level of the left scapula and the superomedial portion of the scapula before operation. B, 14 months postoperatively with correction of the elevation and rotation of scapula. Note also the left clavicle after morcellation (in B).



**Figure 2:** Case with left side elevated scapula reaching up to the occiput with bony prominence; B. post operative correction of the shoulder level, note the widening of the scar in its upper half; C. abduction of left arm is limited just preoperatively; D. the post operative improvement is obvious.



**Figure 3:** A and B showing the pre and postoperative photographs respectively with correction of the elevation, the dotted lines drawn on the patient refer to the level of the scapular spines which are parallel.



**Figure 4: A. grade 3 left sided elevation in the preoperative photo, B. nearly normal shoulders postoperatively, C. preoperatively there is limitation of abduction, D. there is much improvement postoperatively.**

### Discussion

Most of the studied cases were from Baghdad. Although all references refer to the deformity of being rare, most of them did not mention its prevalence in community. Because the diagnosis is clinical in the first step and the unawareness of the mild cases and because of the many and may be more attracting associated anomalies, its exact prevalence is difficult to be estimated. Many articles refer to different figures of predominant side of affection and of gender mostly affected. Most said that right to left ratio is 1 – 2:1<sup>(13, 16, 21)</sup> and Aydinli et al said it is 1:2, also most said that the females are affected more. This study showed nearly equal side involvement (R: L ratio 1.1:1) and more affection of males with a ratio 1.5:1. We have not found any family history of same problem even in the 2<sup>nd</sup> and 3<sup>rd</sup> degree relatives. Also no gynaecological problems are reported for the mothers of the patients during gestation. The Woodward procedure has been praised in having the ability to provide better both cosmetic and functional results<sup>(13, 25, 26)</sup>. Woodward operation is usually

straight forward (taking in consideration the regional anatomy) in cases of minimal complexity. In many severely affected cases it could be difficult and dangerous, especially when ribs are deficient and the field of surgery is separated from the pleura by thin sheath of delicate muscles. When there are deficient or deformed ribs, the scapula is the only protecting bony structure of the posterior upper chest and it is seated in a relatively deep bed making its lower down positioning more difficult, more bulging outward, and less congruent with the new rib cage articulation, and so less smooth and smaller range of scapula – thoracic movement. Much lowering of the scapula is not necessary associated with good function or good appearance as Cavendish noticed, so we did not rely on the mere radiological lowering of the scapula in assessing cosmetic appearance postoperatively and we think that Cavendish classification is still valid. Woodward, Grogan et al, Carson et al, and Dendane et al report a gain in abduction postoperatively ranging from 32.2 to 40 degrees. This study showed

20.3 degrees gain in abduction. This may be attributed in part to the short follow up period in our study. Even in achieving good scapular descent (clinical and radiological) there may be little improvement in function due to associated anomalies or due to less remodeling reserve in older children. The best results obtained in patients aged 5 – 7 years while patients above 10 have less improvement. The patients who are in greater expectance of obtaining more functional improvement are those with more severe affection and those with omo-vertebral bars. Obviously the follow up period of 1 to 24 months is not sufficient to evaluate long term results and remote complications. During the follow up period no worsening of the obtained improvement and no recurrences were occurred. The reviewed available articles with long time follow up showed no recurrence or worsening over long follow up.<sup>(26-28)</sup> Robinson et al and Carson et al advice routine clavicular osteotomy while Grogan et al not recommend routine osteotomy. Our data cannot either support routine clavicular osteotomy nor predict vulnerable patient to neurovascular compromise. Good closure technique of the skin using non absorbable synthetic sutures can greatly affect the scar formation and even better results were obtained by subcuticular skin closure. Apart from the case with complete brachial plexus palsy and the case of transient focal neurological deficit, the complications in general are within the expected range.

In conclusions:

1. The primary indication for correction is cosmetic concerns and the surgery is justified further for the more severe cases.

2. The improvement in abduction can be expected in and especially needed for the more severely affected children.

3. Woodward procedure seems to be especially effective in obtaining both the cosmetic and functional gain at the same time.

4. The procedure needs good deal of attention in the handling of the delicate muscles and tissues of the child with good experience and anatomical knowledge.

5. The family satisfaction about the operative results can be encouraged further by reporting the grade and function preoperatively and comparing them with the post operative reports especially by photographs.

Accordingly we recommend the following:

1. Longer period of follow-up and more patients are needed to be included in such a study to assess the results and whether relapse or other complications can occur later in life.

2. We advice to operate patients before the age of 10 years.

3. Our study used Woodward operation for Sprengel deformity but other available procedures need to be evaluated by further studies to compare results and reveal the best procedure.

4. Most of the patients with congenital undescended scapula have other congenital malformations and especially skeletal malformations which may need further assessment and management.

5. It seems to be beneficial to found pediatric orthopaedic centers to offer greater care and better evaluation for congenital and developmental disorders including data collection, management, and conducting researches.

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