



Sprengel's deformity of the shoulder joint treated by Woodward operation

Mohamed Mansour Elzohairy¹ · Adel Mohamed Salama¹

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Abstract

Background Sprengel's shoulder deformity is a rare condition that happens because of the abnormal termination of the caudal migration of the scapula during the embryonic period.

Methods This retrospective clinical study included 10 consecutive children who underwent a Woodward operation to correct the Sprengel's shoulder deformity.

Results The average preoperative Cavendish grade for cosmetic evaluation was 3.5 (range 3–4), which decreased to 1.4 (range 1–2) at the final review (statistically significant, $p > 0.002$, Wilcoxon signed-rank test). Cavendish grade improvement and increase in abduction had a strong positive association ($r = 0.681$, Spearman correlation coefficient).

Conclusion The results showed that both functional and cosmetic outcome had a positive correlation with Woodward procedure. More cosmetic and functional improvement was seen at the last follow-up visit especially in cases where the operation was done in the younger years.

Level of evidence IV.

Keywords Sprengel's · Shoulder deformity · Woodward operation

Introduction

Sprengel's deformity is defined as a congenital failure of the descend of the scapula and was first described by Eulenburg [1] in 1863. Sprengel had reported in four cases; in 1891, so, the deformity has been described as Sprengel's deformity [2, 3]. The main pathology of this deformity is variable degrees of scapula elevation and scapula hypoplasia due to omovertebral bone, in nearly one-third of the patients which

articulates between the medial border of the scapular bone and one or more of the cervical vertebrae. At the absence of an omovertebral bony bar, a fascial fibrous sheath extends from the superior angle of the scapula to the spinous process, the lamina, or the transverse process of one or lower cervical vertebrae. This fixation leads to the high and medial position of the scapula [4–26]. The Sprengel deformity usually happens at birth and is increased with the child growth. Cosmetic and limited abduction at the shoulder is the main problem regarding this deformity. The classification of this condition depends on the grading system according to Cavendish [7], which is generally accepted as a classification system starting from grade I to grade IV depending on the severity of the deformity.

Many surgical techniques had been described to correct the Sprengel's deformity; all of them had their merits and demerits with no consensus in those few reported literature on the best method to treat this deformity. Also, there is no agreement between the authors on the ideal age for the surgical correction. Because of that, and for the paucity of these reports, we are in need to add the experience of our institute as regarding the management of Sprengel's Deformity [4–6].

Dr. Mohamed Mansour Elzohairy and Dr. Adel Mohamed Salama had shared in this work.

✉ Mohamed Mansour Elzohairy
elzohairy.me@gmail.com
<http://www.mmmansour.faculty.zu.edu.eg>
<https://scholar.google.com.eg/citations?user=XfVQ6RsAAAAJ&hl=en>
https://www.researchgate.net/profile/Mohamed_Elzohairy2

Adel Mohamed Salama
Adelsalmaa@gmail.com

¹ Orthopaedic Surgery Department, Faculty of Medicine, Zagazig University, Zagazig City, Ash Sharqia Governorate 44519, Egypt

The hypothesis of the present study is that Woodward procedure is a safe and valuable operation for Sprengel deformity correction as regarding the cosmetic and functional outcome, especially if it is performed in the earlier age of 1.5 years up to 4 years even in severe Cavendish grade IV deformity. So we try to answer the following queries as, what is the main pathology, indications, the ideal age, the complications, the postoperative functional outcomes and the advantages of Woodward procedure [8] compared to different surgical procedures that are used to correct this deformity.

The current retrospective study is aiming at the evaluation of the functional outcome and cosmetic results of children who underwent the operative correction of the Sprengel's Deformity by the Woodward procedure and to evaluate the degree of the cases and/or family satisfaction.

Materials and methods

From January 2000 to May 2014, 10 children underwent the Woodward procedure at our hospital to correct the Sprengel's shoulder deformity because of cosmetic and functional causes. There were (two boys and eight girls) with 10 involved unilateral shoulders (six left shoulders and four right shoulders). The average age of the cases was 2.9 year [range, 1.5 years (18 months) to 4 years] at the operation time. The preoperative evaluation consisted of a clinical examination, a preoperative cosmetic appearance, abduction

function of the shoulder joint, radiological evaluation of the cervical spine, and associated spinal anomalies, scoliosis, and hemivertebrae all of them have been documented as they might accompany Sprengel's deformity. Radiographs and accompanied deformities were reported. The Cavendish grading system [6, 7], which is; (1—very mild, 2—mild, 3—moderate, 4—severe deformity) had been used preoperatively and postoperatively for cosmetic evaluation, Cavendish grade 1, where the shoulders are level and deformity is not noticeable when the patient is dressed. Cavendish grade 2, where the shoulders are almost level, but the deformity is visible with the patient dressed, Cavendish grade 3, where deformity is easily visible and the involved shoulder joint is elevated by 2–5 cm, Cavendish grade 4, where the superior angle of the scapula is close to the occiput, as shown in Fig. 1, Table 1. Shoulder joints details were evaluated by CT in all (10 shoulders) and magnetic resonance imaging (MRI) in (one shoulder) only preoperatively. None of the cases had a cardiac or renal anomaly. Five patients (50%) had an omovertebral bony bar by radiological examinations. Before the operation, the high and hypoplastic shape of the affected scapula was the main feature. Neither of the cases exhibited any neurological abnormality nor associated spinal anomalies or congenital bony deformity.

The operative technique

General anesthesia was used in all patients. All patients were positioned in the prone position. The Woodward operation



Fig. 1 Sketches showing the various Cavendish grades. **a** Cavendish grade 1, where the shoulders are level and deformity is not noticeable with the patient dressed. **b** Cavendish grade 2, where the shoulders are almost level, but the deformity is visible with the patient

dressed. **c** Cavendish grade 3, where deformity is easily visible and the involved shoulder joint is elevated by 2–5 cm. **d** Cavendish grade 4, where the superior angle of the scapula is close to the occiput [6, 7]

Table 1 Cavendish grading system scale used to cosmetically classify patients with a Sprengel deformity [7]

Grade I (very mild)	Leveled shoulders and practically invisible deformity when the patient is clothed
Grade II (mild)	Shoulders practically leveled, but the deformity is visible when the patient is clothed
Grade III (moderate)	Shoulder elevated from 2 to 5 cm and the deformity is easily visible
Grade IV (severe)	Shoulder significantly elevated, with the superior angle of the scapula close to the occipital

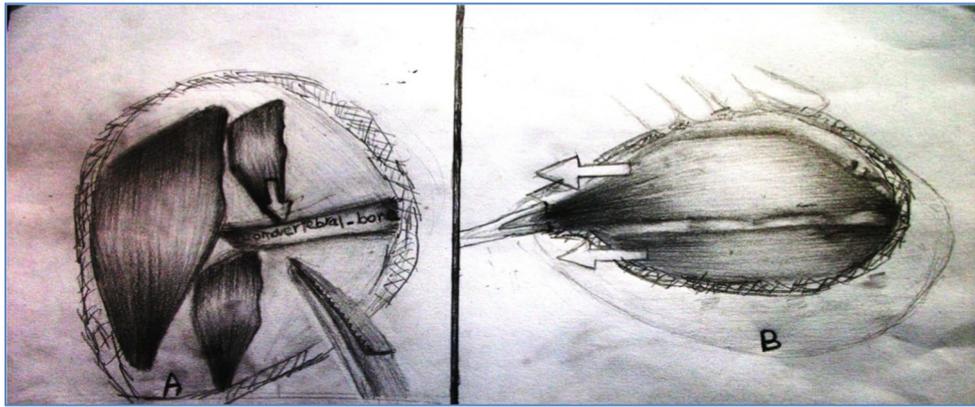
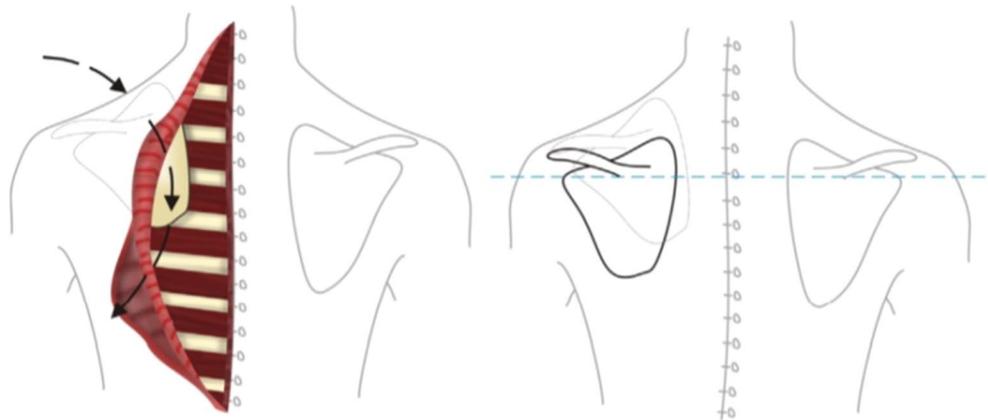


Fig. 2 **a** Excision of omovertebral bone. **b** Detachment of the origins of the trapezius and rhomboid muscles from the spinous process and caudal displacement of the elevated scapula after excision of the omovertebral bone

Fig. 3 Surgical technique by Woodward et al. After a midline incision, the vertebral attachments of the aponeurosis of the trapezoid, rhomboid, and levator scapulae muscles are detached to be able to rotate and relocate the hypoplastic left scapula more caudally on the thorax. The blue 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 [25]



[5, 6, 8, 25] which included a midline longitudinal incision that was made from the midcervical region to the lower thoracic vertebra, the detachment of trapezius and rhomboid muscles origins from the spinous process and caudal displacement of the high scapula after removing of any omovertebral bone or fibrous connection on the scapula, as shown in Fig. 2. The attached muscles from the medial and superior borders of the scapula were reflected extraperiosteally. A scapuloplasty which is excision of the superomedial border of the scapula was used to reshape the deformed scapula in all the ten patients. The scapula and detached muscles were repositioned with heavy sutures, as shown in Fig. 3. Intra-operative image fluoroscopy, as shown in Fig. 4, was used for assessment of scapular descend and correction. No clavicle osteotomy or external fixation was performed in any patient.

Postoperative rehabilitation and follow-up

Patients had Velpeau sling protection for 2 weeks, followed by progressive rehabilitation of the operated shoulder. The

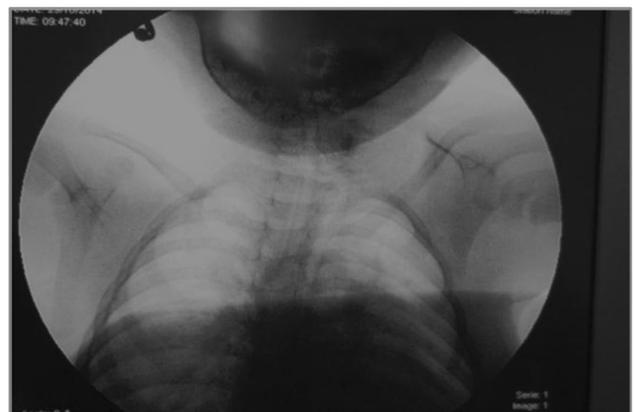


Fig. 4 Intra-operative image fluoroscopy was used for assessment of scapular descend and correction

patients were followed up clinically and radiological at 1, 2, 3, 6 months and then annually up to the last visit after surgery. The cosmetic appearance, abduction with other functions of the shoulder and the satisfaction of the patients and

parents were documented. Statistical analysis was done using SPSS v.16 software (SSPS Inc., Chicago, IL, USA). The test of Wilcoxon signed-rank was used for the paired analysis of preoperative and final results of Cavendish grades and the range of abduction. Spearman's correlation coefficient test was used to compare the relationship between cosmetic enhancement and functional improvement. A p value > 0.05 was considered to be significant.

Results

All patients have been followed at an average of 3.5 ± 1.2 years (range: 3–5 years). All the 10 cases were improved cosmetically and functionally with surgery. The mean arc of shoulder joint abduction improved from average $83^\circ \pm 11.5$ (range: 60° to 105°) before to $152.5^\circ \pm 18.3$ (range: 135° to 175°) after Woodward surgery. The increase in the degrees of the arc of motion was significant on statistical analysis ($p > 0.005$, Wilcoxon signed-rank test), indicating good functional results with the procedure. The mean preoperative Cavendish grade for cosmetic evaluation was 3.5 ± 0.3 (range 3–4), which decreased to 1.4 ± 0.2 (range 1–2) at the final review (statistically significant, $p > 0.002$, (signed-rank Wilcoxon test). The Cavendish grade improvement and increase in abduction had a strong positive association ($r = 0.681$, Spearman correlation coefficient). A typical clinically good result is shown in Figs. 5, 6. The Woodward



Fig. 5 Preoperative clinical photograph showing a Cavendish grade 4 Sprengel deformity at 1.5-year-old female child



Fig. 6 Follow-up clinical photograph at 5 years follow-up with the Cavendish grade 4 deformity being reduced to grade 1

operation achieved an average lowering of 2.6 (range 2–3.2) vertebral levels at the final follow-up in comparison to the preoperative radiographs in the current study (Figs. 7, 8). In all patients, the scapular body size remained smaller than its contralateral scapula. An unsightly surgical scar was reported in one case that had superficial wound infection which was treated after two weeks with antibiotic therapy. 50% (5/10) of our cases had an associated omovertebral



Fig. 7 Preoperative anteroposterior radiograph showing elevated scapula on the left with spine of the scapula at the level of the sixth cervical vertebrae. Also seen is the omovertebral bar (marked by arrow)



Fig. 8 Follow-up radiograph at 5 years follow-up of the same patient with spine of the scapula at the level of the first thoracic vertebra, thus, achieving a lowering of two levels with derotation of the scapula

bar which was excised. No clavicle osteotomy was done on our patients. None of the children in our series had an injury in the brachial plexus. All the parents and children were satisfied with the final results of the treatment. None

of the patients have shown loss of cosmetic appearance or range of motion of the operated shoulders, and the location of the scapula did not change with time, as shown in Figs. 8, 9, Table 2.

Discussion

The main pathology in Sprengel shoulder deformity is binding of the scapula to cervical vertebrae due to an omovertebral process that, reduces the scapulothoracic movement, further impeding the range of abduction at the shoulder joint [4, 6]. The glenoid directed inferiorly due to medial rotation of the distal angle of the scapula and, subsequently, the degrees of abduction at the shoulder joint are reduced [4]. So, in all the surgical procedures, for correction of Sprengel deformity, we aiming mainly to correct this pathology by releasing the scapula-cervical bony binding that was reported from 18 to 60% by other authors in the literature [9] and was presented in 50% of the cases of the current series. While ossified connective or cartilaginous tissue binding was presented in 50% of our patients. Jindal et al. [6] in their study had compared the outcome in two groups of cases with or without an omovertebral binding bar and they reported that the absence or presence of the

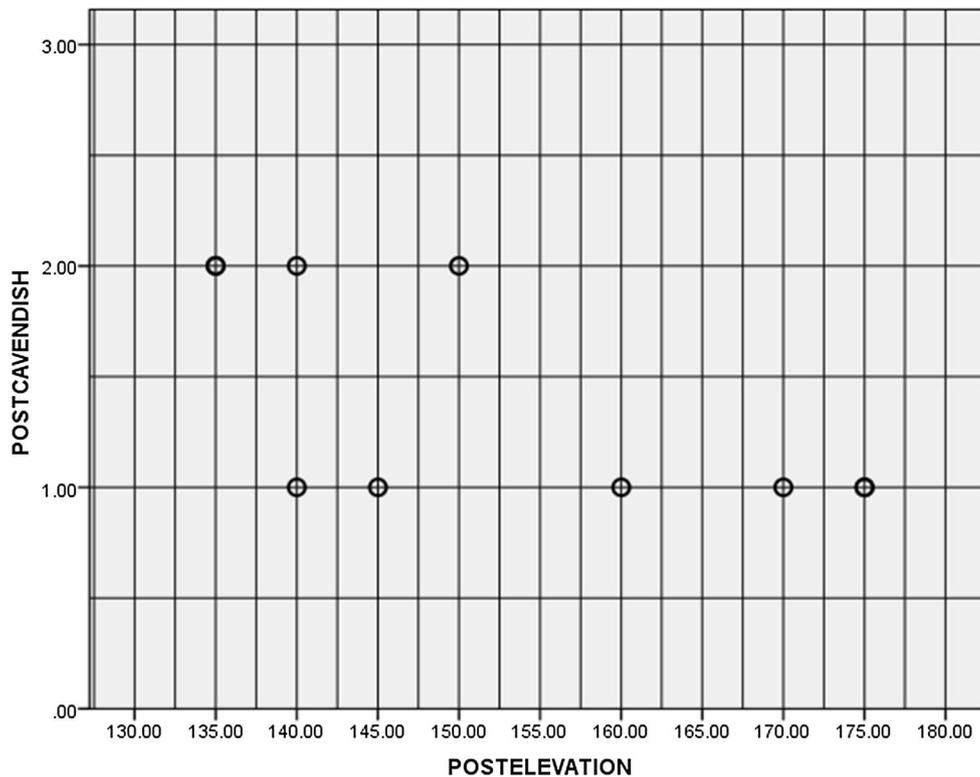


Fig. 9 Scatter diagram showing the relationship between the improvement in shoulder abduction degrees (postelevation) and cosmetic improvement (postCavendish grade)

Table 2 Summarized data from patients with Sprengel deformity operated by the Woodward technique

	Case 1	Case 2	Case 3	Case 4	Case 5	Case 6	Case 7	Case 8	Case 9	Case 10
Gender	F	F	M	F	F	M	F	F	F	F
Side	R	L	L	L	L	R	L	R	L	R
Age	1y 6 m	4y	3y	3y 3 m	2y 4 m	3y 2 m	4y	3y 1 m	3y	2y
Associated deformities	Omo vertebral bone	None	None	Omo vertebral bone	None	Omo vertebral bone	Omo vertebral bone	Omo vertebral bone	Omo vertebral bone	None
Preoperative Cavendish	4	3	3	4	3	4	4	4	3	3
Postoperative Cavendish	1	1	1	2	1	2	2	2	1	1
Preoperative elevation	60°	100°	85°	80°	100°	65°	70°	75°	90°	105°
Postoperative elevation	145°	140	175°	150°	160°	135°	135°	140°	170°	175°
Lowering of the scapula	2.4	3	3.2	2.7	2.6	2	2.2	2.1	2.8	3
Complications	None	None	Superficial infection	None	None	None	None	None	None	None
Postoperative follow-up	4	3.5	2.4	5	4.5	3	3.2	3.3	3	5
Satisfaction	Satisfied	Satisfied	Satisfied	Satisfied	Satisfied	Satisfied	Satisfied	Satisfied	Satisfied	Satisfied

M male, *F* female, *L* left, *R* right, *y* year, *m* month, *cm* centimeters

bar had no effect on the end results. The same was shown in the current series [9]. Most of the authors had an agreement that, conservative management of (Cavendish grade 1) patients is satisfactory [9]. This including; rehabilitation with strengthening of muscles in the scapular region as; the rhomboid muscles, the subcapsular muscle, the deep back and abdominal muscles, which is a standard procedure also used in the management of wrong posture, juvenile kyphosis, and non-structural scoliosis.

Surgical treatment is fully indicated for mild and moderate cases (grades 2 and 3) because it provides an improvement in both function and appearance. The chosen procedure and the length of skin incision are tailored as regarding the degree of deformity. Some authors had reported poor prospects for improvement after surgical treatment in severe cases with Cavendish (grade 4), [9, 10]. Surgical management in severe cases, Cavendish (grade 4) was considered as effective by some authors, who performed clavicular osteotomy or total scapulectomy in spite of frequent complications such as permanent or temporary brachial plexus injury [9–11]. There are various surgical procedures for elevated scapulae that reported in the literature, all offering a reasonable and comparative outcome, [5–11].

Recently, the method of choice is Woodward's operation in which is no long-term plaster immobilization is required including spica-cast for anchoring external suture and it is less painful during postoperative treatment than the previous method advised by Green [11], though it gives comparative results. Green's procedure may also frequently result in secondary healing and develop an unsightly keloid scar [5–11]. Woodward's operation is basically a gentle procedure which carries a low risk of injury to the brachial plexus and profuse bleeding [5–10, 26]. It depends on a scapular transplantation to a lower level by moving the spinal origins of the rhomboid and trapezius muscles caudally [5–10, 26]. The reported results by the majority of the authors of subtotal scapulectomy who were previously recommended for the management of the most severe cases of Sprengel's deformity were unsatisfactory regarding both function and cosmetics. Scapular osteotomy with osteosynthesis, beside the removal of the prominent part of the scapula and the cause of scapular fixation, has been shown by Wilkinson et al. [13], but many authors regarded this procedure as unnecessary [5, 8, 9]. Clavicle osteotomy might be necessary when the deformity of this bone, particularly at the sternoclavicular joint, would prevent adequate descent of the scapula and shoulder joint [13].

The osteotomy (morcellation) of the clavicle followed with the operation for lowering the scapula is considered a first surgical procedure by many authors and can be done in many severe deformities to avoid the injury of the brachial plexus [13–16]. The clavicular osteotomy with sternoclavicular osteotomy is indicated mainly in case there

is an old age to facilitate the scapula reduction and avoid subsequent brachial plexus injury. However, there is no consensus among the authors about the role of clavicular osteotomy procedure in the prevention of brachial plexus injury [6]. Although there is no direct evidence that suggests that the clavicular osteotomy prevents brachial plexus palsy, it was done only as a prophylactic measure, and it also helps to mobilize the scapula more in older children, where the joints are not supple enough to permit a good surgical descent. In the recent study, no clavicle osteotomy was done since all of our cases were ranging from 1.5 up to 4 years. Brachial plexus injury remains one of the most serious side effects of the operation. Many studies documented postoperative neurological involvement, either transient or permanent, after the Woodward operation [6, 9, 10]. McMurtry et al. [17] reported that not correcting the scapular position excessively is the best maneuver to avoid neurological complications. Fortunately, we did not report the occurrence of any neurological complications in our series. We achieved an average scapular caudal lowering of 2.1 vertebral bodies. Satisfactory caudal displacement was reported by various authors who used the Woodward procedure. However, we agree with the majority of the authors that, the significant caudal lowering of the scapula achievement may not sign a good increase in the degrees of range of motion [6, 10, 18–24]. We also agree with other authors in making, overzealous attempts at correction should be particularly avoided in order to prevent any neurological complications [5–10, 18]. In the reported literature, there is no agreement about the ideal age for surgical correction. Opinion is divided when it comes to the optimal age for correction. Surgical correction in patients less than 3 years of age technically demands more than in an older child, but as the age increases, the results tend to be worse [6]. Dendane et al. [19] stated that the ideal age for surgery to perform surgical correction to reposition the scapula are those below 4 years of age, while Greitemann et al. [20] considered the surgery to be done before 6 years old. Khairouni et al. [21] reported that the patient's age and the presence of omovertebral bone do not affect the results.

Also, Doita et al. [22] achieved a good cosmetic outcome in two patients operated in adulthood, but only used bone resection techniques without the caudal displacement of the scapula. The average patient age at the time of undergoing surgery in our study was 2.9 months, which is lower than the age at which patients have been done by the Woodward operation in various studies of cases with Sprengel's shoulder [5, 6, 8–10, 16, 25, 26]. We agree with the authors that, as age increases, the more functional outcome deteriorates but the cosmetic outcome does not. So, in the older children who underwent for Woodward procedure an improved appearance is expected but not function [6, 16]. Ross and Cruess [23] analyzed 77 operated patients and they reported

the best results regarding the elevation gain which happened with the Woodward [8] and Green [12] operation, both included the lowering of the scapula. The improvement in shoulder abduction and the Cavendish grade at final follow-up was obtained in the recent study and was quite comparable to other series managed by the Woodward procedure (Table 2). Our results were in the same scale with Jindal et al. who found that functional and cosmetic outcome had a positive correlation, an inference that was not previously reported. We also found that as the duration time of follow-up increases, further cosmetic and, especially, functional improvement is seen. In the present study, we achieved an average scapular caudal relocation of 2.4 vertebral bodies. Many series have included other side effects which may plague the operation which are an infection, unsightly keloid-ridden scar marks and winging of the scapula. In the recent study, we have only one case with superficial infection and keloid formation. Our results supported the previous reports and have documented and confirmed our hypothesis that it's a great advantage to start as early as 1.5 years (18 months) even in severe Cavendish grade IV to obtain both good functional and cosmetic results. The main limitation of the present study is the small sample size. But, with the rarely reported incidence of this disorder, 10 patients are considered an adequate sample size to give good, valuable information about Sprengel deformity treatment that will be added to the literature database.

Conclusion

The Woodward operation offers a valuable improvement of both function and cosmetic appearance of the shoulder joint for cases with Sprengel deformity. The results of the present study showed that the earlier the age in which the surgical correction done the better the functional and cosmetic outcome without any reported serious complications.

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Compliance with ethical standards

Conflict of interest The authors declare that there is no conflict of interest.

Ethical standards All human studies have been approved by the appropriate ethics committee and have therefore been performed in accordance with the ethical standards laid down in the 1964 Declaration of Helsinki and its later amendments.

Ethical approval All the procedures performed in studies involving human participants were in accordance with the ethical standards of

the institutional and national research committee and with the 1975 Helsinki Declaration as revised in 2000.

Informed consent Informed consent was obtained from all individual participants included in the study.

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