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Long term results after correction of Sprengel's deformity using the Woodward procedure

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Abstract

Introduction: Sprengels deformity is a congenital anomaly of the shoulder with superior displacement and rotation of the hypoplastic scapula. The functional impairment and misshapen appearance of the shoulder can be minimized with different surgical techniques. The aim of this study was to evaluate the long term results after correction of Sprengel's deformity using the Woodward procedure.

Materials and Methods: In this study a total of 16 children with a congenital elevation of the scapula surgically treated for Sprengel deformity by Woodward's procedure were enrolled. Outcome assessment was done by Range of motion, Constant score, DASH (disability of the arm, Shoulder and Hand) score and simple shoulder test (SST) were obtained to evaluate shoulder function. Scapula placement and degenerative disease were assessed by radiographic examination. Cavendish grading was used to evaluate cosmetic appearance at final review and were compared to those before surgery.

Results: Abduction improved by 41^{0} in the first year after surgery and with final improvement of 56^{0} at long term follow-up. At the latest follow-up evaluation, the mean constant score was 85 points, the DASH score 14.59 points and the SST 9.5 points. Radiographs showed superior displacement of the involved scapula in all cases, with no signs of degenerative disease of the shoulder. Cavendish grade improved from grade 3 (pre-operatively) to grade 1 or 2 at the latest follow-up examination.

Conclusion: Woodward procedure shows to be an effective surgical procedure to improve shoulder function as well as cosmetic appearance in patients with Sprengels deformity.

Keywords: Sprengel deformity, cavendish grade, scapula, woodward's procedure

Introduction

The most typical congenital defect of the shoulder girdle is called Sprengel deformity (SD), also known as congenitalhigh scapula or congenital undescended scapula^[1]. Back in 1891, Sprengel was the first to describe the abnormal elevation of the scapula; thus, the condition was termed with reference to his name ^[2]. It is distinguished by a protrusion at the scapula's upper medial border and a restriction of shoulder abduction. The scapula is rotated medially and adducted in addition to its elevated localization, which is particularly noticeable on the superior medial border ^[1, 3]. Further, it is the most common shoulder girdle developmental anomaly. Sprengel deformity severity differs on a scale that ranges from very mild, mild, moderate, and severe based on the Cavendish classification with surgical management being indicated only for severe cases.

A plethora of surgeries aimed at correction of the deformity have been described. Putti^[4] proposed excision of the protruding part of the scapula and lowering of the rest by dividing muscle attachments. This procedure was modified by Green^[5] and Woodward^[6]. Mears^[7] described a surgical technique which involved scapular osteotomy, partial excision of the scapula and release of the long head of triceps. The relative merits and demerits of each procedure exist with proponents of each, and since the deformity is quite rare, there has been no consensus on the optimal management method.

Woodward's procedure is a preferred approach technique and has a decreased potential risk of iatrogenic brachial plexus injury and less bleeding. This prospective study is aimed at the evaluation of cosmetic and functional results in children of Sprengel's shoulder treated by the Woodward procedure.

Materials and Methods

This retrospective study was conducted from 2016 to 2023. In this study a total of 16 children with a congenital elevation of the scapula surgically treated for Sprengel deformity by Woodward's procedure were enrolled.

The children were evaluated for deformity using the Cavendish grade ^[8] as a measure of cosmesis and range of abduction at the

shoulder as a measure of function. According to the Cavendish system of grading, which has been universally accepted, Sprengel's shoulder can be divided into four grades, depending on the level of the shoulders and visibility of the hump. The operative indications in our patients were severe restriction of range of abduction at the shoulder, interfering with activities of daily living or a cosmetically displeasing deformity, or both.



Fig 1: Clinical pictures of a 6 year old boy. A (Inspection of the patient from behind, there by revealing the deformity), B (Difficulty in performing overhead abduction compared to the other limb, Note the scapulothoracic motion occurring at the affected limb) and C (Internal rotation)

Procedure

All patients were put in a prone position. Under general anaesthesia, midline incision was given over cervical spinous process to T 10 vetebra (Figure 1A). Trapezius was identified and seperated freed from origin (Figure 1B). Rhombiod were identified and freed from origin. Omovetebral bar if present was removed. Fibrosed band of levator scapula if present was

excised. Scapula was brought down and muscles were reattached more inferiorly .In 4 cases SS wiring was done to maintain the position of scapula because these 4 patients presented lately at the age of 8-12 years to maintain the position of scapula (Figure 1C).wound was closed and velpeau bandage was applied and in 2 cases u-slab was given.

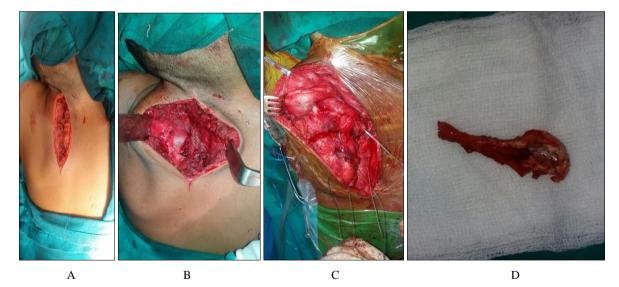


Fig 2: A (Midline incision over cervical spinous from C4 to T 10 vetebra), B (Trapezious muscle identified and separated from origin), C (Fixing scapula with SS wires to spinous processes of vertebra) and D (Excised omovertibal bar)

Post-operative rehabilitation and follow-up

Postoperatively, the limb was immobilized in a Velpeau sling and range of motion exercises begun at 4-6 weeks postoperatively. Outcome assessment was done by Range of motion, Constant score, DASH (disability of the arm, Shoulder and Hand) score and simple shoulder test (SST) were obtained to evaluate shoulder function. Scapula placement and degenerative disease were assessed by radiographic examination. Cavendish grading was used to evaluate cosmetic appearance at final review and were compared to those before surgery.

Results

The mean age of the study population was 8 (range 2-16) years.

In this study there were 10 (62.50%) boys and 6 (37.50%) girls. 9 (56.25%) children had right shoulder deformity, 7 (43.75%) children had left shoulder deformity.

Average follow-up was 14.7 (range 8-26) months. The average preoperative cavendish score was 3.5 which decreased to around 1.8 -2.5 on the final follow up. The decrease of cavendish score was found to be clinically and cosmetically significant. Abduction improved by 41⁰ in the first year after surgery and with final improvement of 56° at long term follow-up. At the latest follow-up evaluation, the mean constant score was 85 points, the DASH score 14.59 points and the SST 9.5 points. Radiographs showed superior displacement of the involved scapula in all cases, with no signs of degenerative disease of the shoulder. Cavendish grade improved from grade 3 (pre-

operatively) to grade 1 or 2 at the latest follow-up examination.

No long term complication has occurred.

С В D А

Fig 3: A (Radiograph of chest with bilateral shoulder revealing the deformity and asymmetry of scapula levels). B (Inspection of the patient from behind, there by revealing the deformity), C and D (Clinical pictures at the follow-up of 3 weeks)



Fig 4: Clinical pictures. A (At the follow-up of 3 weeks), B (Deformity being reduced to Cavendish grade 1 at the follow-up of 1 year) and C (No difficulty in performing overhead abduction at the follow-up of 1 year)

Discussion

Congenital elevation of the scapula was first described by Eulenberg^[9] in 1863, but the eponym Sprengel's shoulder was coined by Kollicker after a description of four cases in 1891^[10]. Sprengel's shoulder arises as a result of arrest of normal caudad migration of the scapula during the organogenesis phase of intrauterine life. In addition to the difference in height, Horwitz ^[11] identified four major differences that an elevated scapula has from its counterpart; changed relation of diameters, curving of the superior border or bending forward of the supraspinous portion, prolongation or rounding of the superior median angle, and presence of exostosis or articulation with the vertebral column. The incidence of the latter is highly variable, ranging from 25% to as high as 88.9% $^{[11,\ 12]}$.

Different surgical techniques are available for the management of Sprengel deformity, including Woodward's procedure, Green's procedure, Shrock's procedure, Mear's procedure, and partial scapulectomy, all offering a reasonable and comparative outcome. However, the preferred method has been the Woodward procedure, since it does not require a postoperative spica cast for suture anchor and entails a much lower risk of bleeding and brachial plexus injury.

In our study, we used Woodward's procedure. Ryabykh et al. in their study on 18 patients with Sprengel deformity used Woodward's procedure. The patients had good correction of the deformity, and no neurological deficit was noted postoperatively ^[13]. One of the aims of Woodward's procedure is to increase shoulder abduction. In the present study, we were able to achieve an average increase of 56° in shoulder abduction. Elzohairy et al. [14] reviewed 10 patients who underwent Woodward's procedure. The mean shoulder abduction improved from 83° to 152.5°, and the cosmetic appearance based on Cavendish grade has improved. Moreover, no complications have been observed except for one patient who had an unsightly scar.

Carson et al. [15] in his respective study noted significant

improvement in shoulder abduction after Woodward's procedure. Our results were similar to Carson *et al.* who achieved an average increase of 50° in shoulder abduction. Walstra *et al.* ^[16] examined the data of seven patients treated with Woodward's procedure; they have reported postoperative improvement of 56° in the shoulder abduction and a mean Cavendish grade improvement from grade 3 to 1-2.

The mean patient age at the time of undergoing surgery in our study was 8 years, which is near about equivalent to the age at which cases have been operated by the Woodward procedure in various series of patients with Sprengel's shoulder ^[15]. The improvement in shoulder abduction and the Cavendish grade at final follow up in our study was quite comparable to other series managed by the Woodward procedure.

We found that functional and cosmetic outcome had a positive correlation, an inference not previously reported. We found that, as the duration of follow up increases, further cosmetic and, especially, functional improvement is seen. In our study abduction improved by 41^0 in the first year after surgery and with final improvement of 56^0 at long term follow-up.

Opinion is divided regarding the optimal age for correction. Surgical correction before 3 years of age is technically more demanding than in an older child ^[17], but as age increases, the results tend to diminish. We found that, as age increases, the functional outcome deteriorates but cosmesis does not. The same was found by Borges *et al.* ^[18], who noted, in two older children, that a modified Woodward procedure improved appearance but not function.

Conclusion

Woodward procedure shows to be an effective surgical procedure to improve shoulder function as well as cosmetic appearance in patients with Sprengels deformity.

Conflict of Interest

Not available

Financial Support

Not available

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