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Management of Sprengel deformity in Adolescence: A case report

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Abstract

Background: Sprengel deformity is a rare congenital anomaly characterized by failure of normal scapular descent during embryogenesis, resulting in a malpositioned, elevated scapula. It is associated with functional impairment and visible shoulder asymmetry, often necessitating surgical correction for improved mechanics and cosmesis.

Case Presentation: A 15-year-old female presented with a visible deformity of the left shoulder since birth, experiencing emotional distress but no pain or functional restriction. Examination revealed Cavendish grade 2 asymmetry with limited abduction (100°). Imaging confirmed left scapular elevation without omovertebral bar. The patient underwent the Woodward procedure with detachment and caudal reattachment of scapular muscles through a posterior midline incision.

Outcomes: Postoperative recovery was uneventful. Physiotherapy was initiated early, progressing to full activity by six weeks. At three months, shoulder abduction improved to 180°, with marked symmetry restoration and increased patient confidence. Functional and cosmetic benefits were sustained at six months.

Conclusion: The Woodward procedure offers effective functional and aesthetic improvement for moderate Sprengel deformity even in adolescence. Early postoperative mobilization is crucial for optimal outcomes. Surgical correction not only restores shoulder mechanics but also addresses psychosocial impacts in visible congenital deformities.

Keywords: Sprengel deformity, Woodward technique, deformity correction, psychosocial distress, cavendish, rigault

Introduction

Sprengel deformity represents a rare congenital shoulder girdle anomaly characterized by abnormal elevation and malposition of the scapula secondary to failure of normal caudal descent during embryonic development. This condition occurs in approximately 1 in 40,000 live births, typically presenting unilaterally with a predilection for the left side and affecting females more commonly than males. The deformity frequently associates with other skeletal abnormalities including scoliosis, Klippel-Feil syndrome, and omovertebral bars, which may significantly influence management strategies and long-term outcomes^[1].

Management approaches are dictated by deformity severity and functional impairment. Mild cases (Cavendish grades 1-2) often warrant conservative observation, while moderate to severe deformities (Cavendish grades 3-4) typically require surgical intervention to address both functional limitations and aesthetic concerns. Among surgical techniques, the Woodward procedure has emerged as a widely accepted approach, involving scapular repositioning through systematic detachment and caudal reattachment of periscapular muscles to restore shoulder symmetry and improve range of motion. Optimal timing for surgical intervention remains debated, though adolescent surgery demonstrates promising outcomes when balancing technical feasibility against psychosocial considerations that profoundly influence quality of life during formative years^[2, 3].

Case Presentation

A 15-year-old female presented to the orthopedic outpatient clinic with congenital visible deformity affecting the posterior left shoulder, present since birth. The patient reported significant emotional distress and self-consciousness regarding the asymmetry, particularly intensified during adolescence. Notably, she experienced no pain or functional limitations affecting activities of daily living.

Physical examination revealed marked shoulder asymmetry with visible elevation of the left scapula, classified as Cavendish grade 2. No muscle atrophy or tenderness was appreciated.

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Active shoulder abduction measured 100 degrees on the affected side, while other range of motion parameters remained preserved. Spinal examination demonstrated mild compensatory scoliosis without additional skeletal abnormalities.

Radiographic evaluation of the cervical and upper thoracic spine confirmed left scapular elevation and scoliosis, consistent with Rigault grade 2 classification. Computed tomography provided detailed anatomic assessment, confirming normal scapular morphology without omovertebral bar or scapular hypoplasia. Given moderate deformity severity and significant cosmetic concern, surgical correction was planned.

The patient underwent the Woodward procedure via a posterior midline incision. Following identification of the trapezius and rhomboid muscles, their vertebral origins were detached and reattached more caudally to facilitate

downward scapular repositioning. Clavicular morcellation was unnecessary due to adequate scapular mobility. The scapula was secured in the corrected position while ensuring no excessive tension on the brachial plexus.

Postoperatively, the affected limb was immobilized in a shoulder sling for two weeks. Passive and active-assisted range of motion exercises commenced after one week, progressively advancing to unrestricted activity over six weeks. No intraoperative or postoperative complications occurred, and neurological examination remained normal.

At three-month follow-up, the patient achieved 180 degrees of shoulder abduction. Shoulder symmetry improved dramatically, and the patient expressed enhanced self-confidence and satisfaction with her appearance. Both functional and cosmetic improvements persisted at six-month follow-up evaluation.



Fig 1: Sprengel deformity and abduction



Fig 2: Imaging



Fig 3: Incision

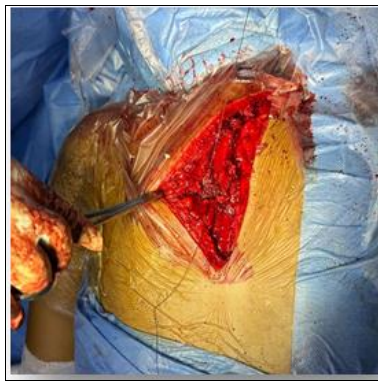


Fig 4: Deformity correction



Fig 5: post operative abduction

Discussion

The pathogenesis of Sprengel deformity involves arrested scapular descent during the third month of gestation, resulting in a high-riding scapula frequently accompanied by associated bony and soft tissue anomalies. Deformity severity is clinically graded using the Cavendish classification system, which stratifies patients from grade 1 (very mild, invisible when clothed) to grade 4 (severe, with superior scapular angle near the occiput). This classification aids surgical decision-making, with mild deformities often amenable to observation or simple procedures, while severe cases necessitate more complex surgical approaches such as the Woodward or modified Green techniques^[3].

The present case, classified as Cavendish grade 2 and Rigault grade 2, was successfully managed with the Woodward procedure—a technique involving systematic detachment of trapezius and rhomboid muscle origins from the spine, followed by caudal repositioning and reattachment to facilitate scapular descent. Contemporary literature indicates that the Woodward method provides reliable functional and cosmetic outcomes with minimal complication rates when performed by experienced surgeons, with mean improvement in shoulder abduction

ranging from 30 to 50 degrees^[3, 4]. Early postoperative physiotherapy proves crucial for restoring range of motion and preventing periarticular fibrosis; in this case, initiation of mobilization within one week contributed substantially to the excellent three-month outcome^[5].

The psychosocial impact of visible congenital deformities during adolescence can be profound, adversely affecting self-esteem, body image, and social interactions. Surgical intervention in this demographic not only addresses tangible functional deficits but also alleviates significant emotional distress. These observations align with findings from multiple studies demonstrating improved quality of life and patient satisfaction following surgical correction during adolescence^[1].

Conclusion

Surgical correction of Sprengel deformity utilizing the Woodward procedure can achieve remarkable functional and aesthetic improvement in adolescent patients. Early initiation of physiotherapy combined with meticulous postoperative monitoring are essential for maintaining shoulder mobility and symmetry. This case underscores the importance of considering surgical management not solely

for functional restoration but equally for psychosocial well-being in adolescents presenting with visible congenital shoulder deformities.

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Nil.

Conflict of Interest

The authors declared no potential conflicts of interest with regard to the submitted article. Each author certifies that he or she has no commercial associations that might pose a conflict of interest in connection with the submitted article.

Ethical standards

Institutional ethics committee approved the study.

Informed consent was obtained from the participant (legal guardian) included in the study.

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