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Transfers of the sternal head of pectoralis major and latissimus dorsi to treat Sprengel's syndrome

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Sprengel's syndrome, characterized by the congenital failure in the descent of the scapula to its regular position, leading to a hypoplastic, elevated, and malrotated scapula.[5,6] It has long posed challenges in orthopedic medicine due to its rarity and the complexity of its presentation.[3,8] Another common alteration in this deformity is the presence of the omovertebral bar, found in about a third of Sprengel's syndrome cases; this bone connects the scapula to the vertebral column. Sprengel's deformity appears either as a single defect or in association with other abnormalities, most commonly Klippel-Feil syndrome, but also scoliosis, spina bifida, hemivertebrae, rib segmentation abnormalities, clavicular abnormalities, and underdevelopment (hypoplasia) of neck or shoulder muscles. A conventional anteroposterior chest X-ray, including both shoulders, is a simple and effective means of diagnosing Sprengel's deformity. Computed tomography with 3-dimensional reconstruction and magnetic resonance imaging are now examinations that assist in diagnosing coexisting abnormalities and treatment planning. Traditional surgical and nonsurgical treatments have aimed at cosmetic and functional correction, but often with limited success and significant risk of complications. The introduction of new surgical techniques[2,4] and a deeper understanding of the syndrome's pathophysiology has opened the door for innovative approaches.[9]

This technique description and case report presents a novel procedure developed to address the core anatomical and functional deficits associated with Sprengel's syndrome. Building upon the foundational principles of orthopedic surgery and incorporating advanced techniques in muscle and bone repositioning, this procedure aims to offer improved outcomes in terms of range of motion, pain reduction, and aesthetic normalization.

By examining the case of a patient treated with this new technique, this technical note seeks to explore the efficacy, safety, and potential implications of this approach. The procedural innovation lies in its muscle transfer's nature, using a series of transfers and releases to provide the most functionally effective correction possible for Sprengel's syndrome and remove the omovertebral bar.

Surgical technique

The patient is positioned laterally under general anesthesia, with evoked potential monitoring to track any neurological changes in the brachial plexus. This setup is critical for ensuring the safety and effectiveness of the procedure.

Clavicle morcellation

The first surgical step involves an approach over the middle third of the clavicle to perform clavicle morcellation. This process is crucial for 2 main reasons: it facilitates the downward movement of the scapula, essential for correcting the elevated position characteristic of Sprengel's syndrome, and it prevents potential impingement of the brachial plexus, which could occur due to the downward movement of the scapula with the clavicle. The clavicle

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periosteum is dissected and released from the clavicle to allow its suture after the middle portion of the clavicle osteotomy and subsequent morcellation, creating more space and allowing the clavicle to fit better over the brachial plexus without causing impingement, thereby preventing nerve damage, and ensuring smoother postoperative recovery. Such an impingement could lead to significant pain and functional limitations for the patient (Fig. 1).

Posterior longitudinal approach and muscle release

Following the clavicle morcellation, the procedure continues with a posterior longitudinal approach just medial to the scapula's border. This phase involves releasing any fibrous bands or omovertebral bones, crucial for rectifying the atypical positioning and limited movement characteristic of the syndrome.

Attention is then turned to the meticulous release of specific muscles, including the often-abnormal lower trapezius, levator scapulae, rhomboid minor, and rhomboid major. The gentle digital release of the space between the subscapularis muscle and the thoracic wall is also performed, ensuring minimal trauma to surrounding tissues and creating space for improved scapula mobility.

Extreme caution is exercised with the suprascapular and superior subscapular nerves to prevent postoperative complications.

The serratus anterior muscle is generally atrophic and dysfunctional, having the need of the transfer of the sternal head of the pectoralis major muscle to restore the scapula's protraction and balance. The transfer of the latissimus dorsi can also be useful in moving the scapula downward and assisting in its retraction.

Muscle transfers

The concluding stage involves transferring the sternal head of the pectoralis major and the latissimus dorsi to the inferior aspect of the scapula. This is preceded by a lower deltopectoralis approach. Careful attention is paid to the tendon of the sternal head of the pectoralis major, located just beneath the clavicular head. Release of the clavicular head of the pectoralis major from the humerus is done. Then the release of the sternal head along with its periosteum and a small bone fragment from the humerus is done to allow better healing of the muscle transfer. The latissimus dorsi needs also to be harvested through the same approach. It is just beneath the sternal head of the pectoralis major, care must be taken because beneath the latissimus dorsi is the teres minor, and sometimes the release between these 2 muscles will not be easy. This transfer is meticulously performed through a space created between the scapula and the thoracic wall with special care with radial nerve. This positioning is key to achieving the desired anatomical correction and functional improvement. The suture of the latissimus dorsi and sternal head of the pectoralis major in the inferior aspect of the scapula is performed through a small incision just inferior to the scapula, with special care taken to avoid damaging the radial nerve. The clavicular head of the pectoralis major is then reinserted through the space created during the release of the sternal head (Figs. 2 and 3). Patient is left in a regular sling for 5-6 weeks.

Case report

A, 4-year-old patient presenting marked elevation of the left scapula, with the superomedial angle reaching the level of the C3/C4 vertebral bodies, approximately 5 cm above the upper limit of the contralateral scapula, marked lateral rotation of the scapula, T4 vertebra with a 'butterfly' appearance, and left, convex dorsal scoliosis changes detailed in a spine study of the same date.

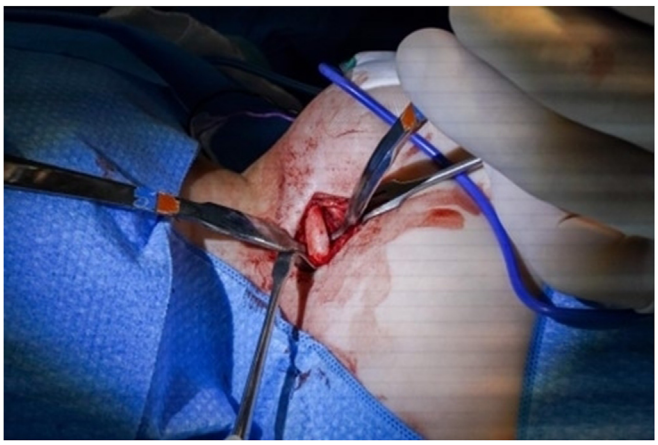

Figure 1 Longitudinal approach over the middle third of the clavicle, preserve de periosteum.

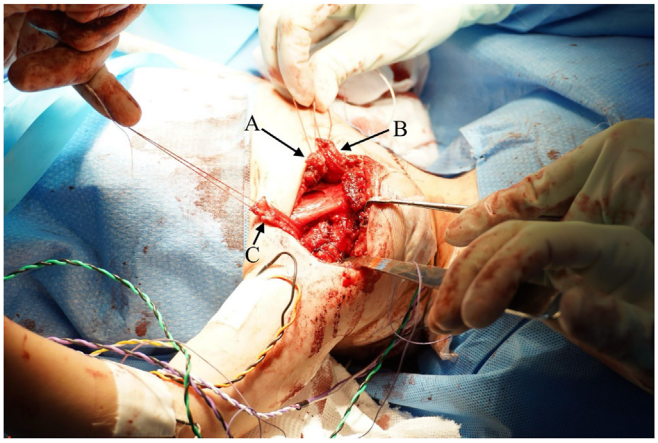

Figure 2 Deltopectoralis approach; (*A*) Pectoralis Major Sternal Head, (*B*) Pectoralis Major Clavicular Head, and (*C*) Latissimus Dorsi.

Fusion of the first 3 left costal arches, causing deformity in their contours and reduction of the thoracic cavity volume (Figs. 4 and 5). Atrophy of the anterior serratus muscle with congenital impossibility of scapular protraction and elevation. Patient is left in a regular sling for 5/6 weeks and was free for movements. After this initial period, 3-month postsurgery elevation improved from 30° to 90°.

Deltoid strength was better, indeed; during the surgical procedure, the evoked potential demonstrated improvements in the axillary nerve response that can be the cause of this improvement, associated with the better positioning of the scapula. Patient had a cosmetic improvement as follows in Figure 6.

Discussion

Sprengel's syndrome is a genetic disorder characterized by an elevated scapula above normal levels, leading to restricted movements of the upper limb and associated neurological impairments that significantly limit arm movements, including elevation and abduction. Consequently, any therapeutic approach that could partially restore scapular mobility would be highly beneficial for patients with this condition. Our observations indicate that scapular movements associated with the pectoralis minor, and the middle and upper trapezius muscles, appear to be unimpaired.

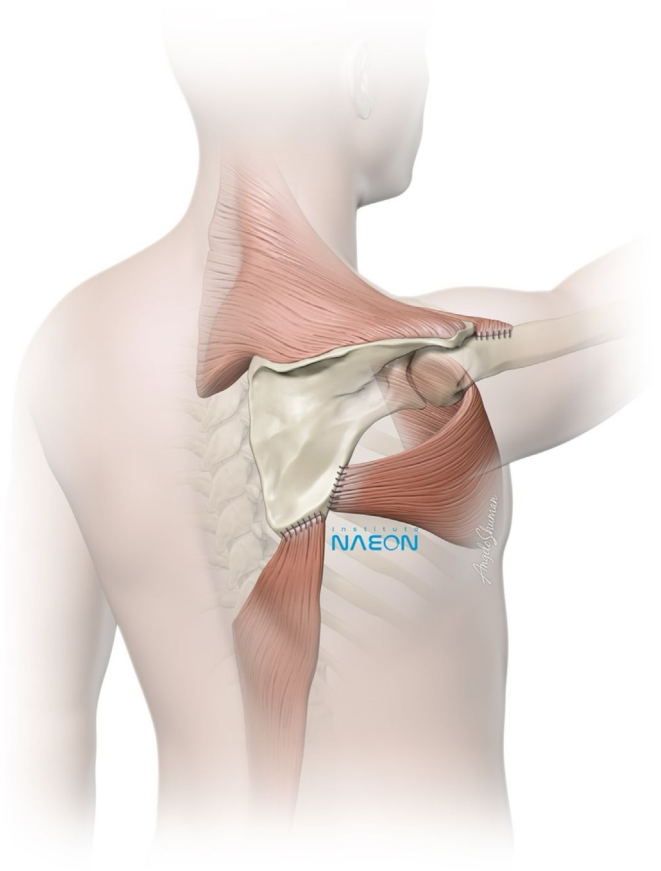

Figure 3 Illustrations showing the scapula after the transfer (used with permission of NAEON Medical Education and Research. All rights reserved).

Therefore, we propose that emphasizing the enhancement of proactive and active downward scapular movements may offer therapeutic advantages for individuals affected by this syndrome (Fig. 3). In our procedure, we observed a greater inferiorization of the scapula, superior abduction, and increased contact of the scapula with the rib cage and a smaller incision required for the procedure, compared to the Woodward and Green technique.[1,7] The rationale for the muscle transfers we propose is based on facilitating the full range of scapular movements. This includes scapular retraction, supported by the middle and superior trapezius muscles, and the latissimus dorsi; protraction facilitated by the pectoralis minor and the sternal head of the pectoralis major; and upward rotation solely by the sternal head of the pectoralis major. Superior-medial movements are associated with the intact superior and middle trapezius muscles, whereas downward and medial movements are executed by the latissimus dorsi, attached to the inferior part of the scapula.

In certain cases where the latissimus dorsi appears lax, its insertion onto the scapular spine can be adjusted similarly to the procedure used for accessory nerve lesions, positioned where the lower trapezius is attached.[4] This approach seems more logical, particularly considering the atrophy or functional limitation of the serratus anterior muscle, which limits its utility in patient rehabilitation.[2] In our hands, previous techniques seem to present less effectiveness in achieving lateralization of the scapula and interiorization of the clavicle compared to the method presented in this paper.

The most common major complications in this surgery are related to wound problems, such as keloids, hypertrophic scars, and seroma, in addition to brachial plexus nerve paralysis in special radial nerve.

Figure 4 CT-scan 3-dimensional image. *CT*, computed tomography.

Figure 5 CT-scan 3-dimensional image. *CT*, computed tomography.

Conclusion

The proposed procedure in this case involves transferring the sternal head of the pectoralis major and the latissimus dorsi to a lower region of the scapula. For further inferiorization of the scapula, we perform morcellization of the clavicle, in addition to removing the omovertebral bar. This technique has shown excellent results in scapular retraction and improved movement. Despite the excellent outcome, we need more cases for a better conclusion and assessment of the effectiveness of this technique.

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Figure 6 (**A**) Patient preoperative deformity in left shoulder. (**B**) Patient postoperative after the transfers corrects the deformity, and the incision in left shoulder.

and their guardians have signed consent forms for publication for scientific study purposes, aware that they would not receive any remuneration. The authors declare that consent has been granted by the responsible parents of the patient.

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