



POSNA

The Core Curriculum

Sprengel's deformity

Objectives

1. Define Sprengel's deformity
2. Discuss the reason for the high incidence of other anomalies accompanying Sprengel's deformity, and list 4 of the more common accompanying conditions
3. Discuss indications for correction of Sprengel's deformity
4. Discuss surgical approaches to Sprengel's deformity
5. Discuss complications of surgery for Sprengel's deformity and strategies to reduce their incidence

Discussion point

1. What is the omovertebral bone?

Discussion

Sprengel's deformity is an unusual condition, characterized by upward displacement of the scapula. The etiology of Sprengel's deformity is related to limb bud formation. The scapula generally descends to its thoracic location about the eighth postgestational week, having formed from paraxial mesoderm at about the level of the fourth or fifth cervical vertebra. The usual postmigrational location of the scapula is between the levels of the second and eighth posterior ribs. Since Sprengel's deformity originates in the paracervical mesoderm, it is not surprising that Klippel-Feil syndrome and congenital scoliosis are frequent concomitants. Other rib and spinal anomalies, including syringomyelia, may be present. Diastematomyelia has been noted in about 20% of patients with Sprengel's deformity. Kidney anomalies, also mesodermal in origin, are less frequent concomitants of Sprengel's deformity. The omovertebral bone, connecting the superomedial scapula to the posterior elements of the cervical vertebrae, is found about one-quarter of the time. Various other types of disruptions of limb bud formation and differentiation can be noted, including hypoplastic thumb, clavicular hypoplasia, and hemimelias. Shoulder instability may factitiously increase shoulder motion.

Clinically, Sprengel's deformity can range from minimal, with no restriction of shoulder motion; to severe, with the superior angle of the scapula near the occiput, and marked restriction of scapulothoracic motion. Glenoid version is generally normal. The omovertebral bone, not surprisingly, is associated with greater restriction of motion. The musculature of the shoulder girdle, especially the trapezius, may be hypoplastic and/or fibrosed. Operation is reserved for patients in whom the deformity is marked, or restriction of motion is severe; the two generally go together. Many surgical procedures have been described, including excision of the omovertebral bone, resection of the supraspinous portion of the scapula, subperiosteal resection or release of the

scapula, and distal transposition of the scapula by release of the superior, osteotomy of the scapula, and medial and inferior musculature from the scapula (Green) or the their origin (Woodward), with reattachment to the scapula in its corrected position. Green originally used skeletal traction to maintain the scapula in its corrected position. Modified forms of the Green and Woodward procedure are most popular at present. Complications of surgical treatment of Sprengel's deformity include loss of correction, winging of the scapula, regeneration of the excised portion of the scapula, prominent scars, and neurovascular complications resulting from compression between the clavicle and first rib when the scapula was displaced inferiorly. Strategies to reduce complications include use of a midline incision; osteotomy or morcellization of the clavicle to increase its mobility, osteotomy of the coracoid process to release the tether of the pectoralis minor, extraperiosteal dissection to reduce bony reformation, and fasciodesis of the inferior scapula to the underlying rib to prevent winging. Results are generally satisfactory if these technical factors are integrated into the surgical plan. Lesser degrees of deformity can probably be more safely treated without morcellization or osteotomy of the clavicle.

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