Sprengel deformity: An update on the surgical management

Yasser Ali Kamal*

INTRODUCTION

Sprengel deformity (SD) is a rare congenital malformation of the scapula, named after Otta Gerhard Karl Sprengel in 1891 [1], who suggested that increased amniotic fluid pressure interfered with the descent of the scapula, hence the appellation deformity [1,2]. By 1908, there were 136 published cases with SD [3]. Normally, the scapula lies superiorly at the level of the 2nd cervical vertebra, then it grows and migrates under cellular signaling pathways to determine the deformity and its association with omovertebral bone. The plan for treatment is judged by the Cavendish classification of clinical cosmetic appearance, as well as shoulder functional impairment. Mild cases need no surgical intervention, however, there are several surgical procedures preserved for moderate to severe cases with near similar postoperative outcomes. The commonly performed procedures include Green's and Woodward procedures with further modifications to enhance the early mobilization and to reduce the associated brachial plexus injury. The use of minimally invasive techniques for SD is limited but it may take its place in the future.

Keywords: Thoracic wall abnormalities; high scapula; sprengel deformity

Etiology

The embryological development of the scapula starts between the 4th and 8th weeks of gestation, where it is initially located at the level of the 4th cervical vertebra. At the final location at the level of the 7th cervical vertebra at the 6th week of gestation. Growth arrest of the scapula results in failure of the scapula to descend leaving it hypoplastic and malseated at a high level [14].

Etiologically, SD remains unclear; however, it is generally related to a developmental arrest of the intrauterine normal descent of the scapula. The deformity often coexists with other syndromes and medical conditions including Klippel-Feil syndrome, scoliosis, and rib abnormalities. The imaging evaluation is based on chest X-ray, CT with 3-D reconstruction and MRI to determine the deformity and its association with omovertebral bone. The treatment of SD should include plain radiography and computed tomography (CT) to assess the level of the scapula, determine the associated abnormalities, and delineate an omovertebral bone or band. Moreover, an omovertebral bone or band connecting the cervical vertebrae and the scapula may be found in up to 50% of cases with SD [2,4,5].

The main complaints of a child with SD include cosmetic deformity and limitation of the abduction of the shoulder. Other congenital anomalies may coexist with SD including scoliosis, rib anomalies, Klippel-Feil syndrome and diastematomyelia [6,7]. The initial imaging workup for the assessment of SD should include plain radiography and computed tomography (CT) to assess the level of the scapula, determine the associated abnormalities, and delineate an omovertebral bone or band. Moreover, the complex anatomy and omovertebral connection can be screened best with 3D-CT or MRI [8,9].

The treatment may be non-surgical or surgical according to the degree of cosmetic and functional affection by SD. Many operative procedures have been described to correct SD [10-13]; however, a controversy remains about the appropriate procedure. In this review article, we try to review the literature for the updated knowledge about the etiology, diagnosis, and treatment of SD.

Clinical features

Although SD is a rare entity, it is the most common congenital malformation of the shoulder girdle [16]. Notwithstanding the deformity is three times more predominant in male than female gender [24], it may occur equally in both sexes [25]. Unilateral location of SD is more frequent with a predilection for the left side; however, bilateral cases can exist with less disfigurement and more functional limitation than unilateral cases [26].

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The first clinical presentation of SD is often during childhood in the first or second year of life, or later during adolescence [15,27]. The main complaint in cases of SD is disfigurement due to the asymmetry of the shoulder girdle, in addition to functional impairment (Figure 1). Limited scapulothoracic motion leads partly to a reduction of glenohumeral abduction which is generally well tolerated [15].

Figure 1 A photograph of a child with Sprengel's deformity: (A) asymmetry of shoulder girdles with the higher position of the left scapula with asymmetry in the position of inferior scapular poles (dashed lines), and (B) limited right glenohumeral abduction.

The clinical presentation of SD usually includes other skeletal deformities in the thoracic rib cage and the cervical and thoracic vertebrae such as scoliosis, fused or absent ribs, chest wall asymmetry, cervical ribs, and cervical spina bifida. Moreover, SD is linked to various syndromes, including Klippel-Feil syndrome, X-linked dominant hydrocephalus, mental disturbance syndrome and diastematomyelia [21,28].

Imaging findings

Plain chest X-ray can show the abnormal position and rotation of a scapula (Figure 2). The anteroposterior view can estimate the height difference between normally situated and high scapula using the easily visualized inferior angle of the scapulae, however, chest X-ray is not accurate enough to estimate the severity of SD [25].

Figure 2 Plain chest X-ray shows the abnormal elevation of the left scapula with the prominent superior angle of the scapula in the web of the neck.

The deformity can be assessed using frontal radiographs to determine the relation of the superomedial angle to the vertebral column as proposed by Rigault [29,30]. The oblique radiographic view can be used to assess the presence of the omovertebral bone between the superomedial angle of the scapula and the vertebral column [31].

Additional radiological findings should be calculated including medialization of the scapula (the rotational difference of the two scapulae), and the anterior curvature of the supraspinous part of the scapula. The scapular medialization is calculated by measuring the distance between the medial border of the scapula and a vertical line over the C7 spinous process [32]. Computed tomography (CT) with multiplanar and 3D reconstructions images is useful in quantification of scapular dysplasia, demonstration of omovertebral bone, and assessment of the associated vertebral and rib anomalies. On CT images of the chest, both scapulae are not situated at the same level with images showing absent contralateral scapula particularly in an axial plane of the back of a child (Figure 3). Moreover, CT with 3D reconstruction images can accurately demonstrate omovertebral bone which confirms the diagnosis of SD (Figure 4) [33-35].

Figure 3 Axial CT image showing a high level of the right scapula.

Figure 4 Chest CT 3D reconstruction image, posterior view showing elevated left scapula with omovertebral bone (arrow) between the scapula and cervical spinous process.

Other imaging modalities are not routinely used but may add variable advantages when indicated. Ultrasound is useful for detection of the cartilaginous and/or fibrous structures, with the advantage of being quick, noninvasive and widely accessible. MR imaging is also useful but it is not as quick and readily available. MRI can be used to detect a cartilaginous bar or fibrous band; however, it is more useful in ruling out spinal cord abnormalities such as a diastematomyelia [9,36].
Clinical and radiological classification

The classification of SD remains based on the clinical cosmetic appearance as judged by the Cavendish classification, and the radiological scapular height compared to the vertebrae [37].

Table 1 The Cavendish clinical and Rigault radiological classification of Sprengel deformity.

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<th>Cavendish clinical classification</th>
<th>Description</th>
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<tr>
<td>I: Very mild</td>
<td>Shoulder level and deformity invisible when dressing</td>
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<tr>
<td>II: Mild</td>
<td>Shoulders almost level, lump visible in a web of the neck when dressing</td>
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<tr>
<td>III: Moderate</td>
<td>Shoulder elevated 2.5 cm, Deformity easily visible</td>
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<tr>
<td>IV: Severe</td>
<td>The superior angle of scapula near occiput, with or without neck webbing</td>
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<tr>
<th>Rigault radiological classification</th>
<th>Description</th>
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<tr>
<td>I</td>
<td>Superomedial angle lower than T2 but above T4 transverse process</td>
</tr>
<tr>
<td>II</td>
<td>Superomedial angle located between C5 and T2 transverse process</td>
</tr>
<tr>
<td>III</td>
<td>Supero-medial angle above C5 transverse process</td>
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The Cavendish classification based on the severity of scapular elevation, by assessment of the degree of the visually perceivable deformity considering the difference between the position of the deformed scapula and that of the healthy one or, in case of bilateral Sprengel shoulder, the difference between the position of the undescended scapulae compared with their healthy expected anatomical position [26,30].

On the other hand, the Rigault radiological classification estimates the level of the superomedial scapular angle compared to the vertebrae [37]. Rigault classification estimates the amount by which the scapula is raised in regards to the position of the medial angle of the scapula on standard images (Table 1): slightly raised with superomedial angle lower than T2 but above T4 transverse process (grade I), omovertebral bone generally absent; raised to the usual degree where superomedial angle located between C5 and T2 transverse process (grade II) or severely raised with superomedial angle above C5 transverse process (grade III) [30].

Treatment of Sprengel deformity

Non-operative treatment is indicated when the cosmetic deformity is mild with Cavendish grades I or II, with the minimal limitation of shoulder function. The non-operative protocol should include physiotherapy to maintain shoulder movement and prevent torticollis, as well as observation for progression of the abnormality and assessment of the associated abnormalities. The children should be encouraged to participate in sports such as swimming to maintain range of motion [14].

Surgical intervention is best indicated for moderate and severe SD with significant cosmetic deformity (Cavendish grades III and IV) and significant limitation of shoulder abduction [24]. However, some surgeons used also Rigault radiological classification as Cavendish classification remains subjective and inaccurate and it is based on morphologic and aesthetic criteria only [32].

The optimal age for surgery remains controversial and most of surgeons preferred before the age of 8 years old [16,38]; however, some surgeons recommended surgery even in older patients [39].

The presence of associated syndromes with SD may affect the functional outcome after surgery, thus these syndromes may be contraindications for surgical treatment [24].

Surgical procedures

The surgery for SD should target improvement of cosmetic appearance and shoulder function. The surgical options consist of scapular lowering, muscle detachment, scapular osteotomy, partial resection technique or a combination of these techniques [11,12,40-44]. The commonly performed surgical procedures include Green’s procedure, Woodward’s procedure and scapular osteotomy [11,14,40,45,46].

In Green's procedure, the skin incision is long, curving around the superomedial scapular corner and finishing 5 cm below the inferior angle [15]. The procedure involves the release of all muscles at their scapular insertions, resection of the supraspinatus fossa and omovertebral bone (if present), reattachment of muscles after caudal mobilization of the scapula, and muscle lengthening as needed [14,40].

A number of modifications have been added to the original Green's procedure which include; securing the inferior scapular angle into a pocket of latissimus dorsi to avoid skeletal traction by percutaneous wire [47], non-release of serratus anterior muscle to provide early postoperative mobilization [46], osteotomy of the clavicle and fixation of the inferior pole of the scapula adjacent to a rib [48], osteotomy of the base of the coracoid process and fixing of the scapula to a lower rib [4], and recently fixation of the medial portion of scapular spine to contralateral posterior iliac crest [40].

The Woodward procedure has become the most widely used technique for the management of SD, since its first description in 1961. The procedure relies on release and relocation of the origin of the muscles attached to the medial border of the scapula in addition to resection of omovertebral bone, using a midline skin incision from the spinous process of C4 to T9 [11,14].

The contemporary results of Woodward procedure showed a positive correlation of functional and cosmetic outcomes through short and long terms [45,49], with the procedure and more improvement occurs in cases of younger age at the time of surgery [45]. Moreover, the Woodward procedure may be more physiologically and biomechanically adapted than Green's procedure since it displaces more distally the origin of muscle insertion and combines glenoid varus correction with scapular lowering as the deformity does not only concern scapula but its whole region [50].

The modifications in the Woodward procedure have been proposed to include additional excision of the medial border of the scapula to prevent postoperative apparent winging of the scapula by decreasing its horizontal width [38], Anterior release of the scapula followed by its rotation to correct the varus orientation of the glenoid cavity [50], or combination of the lowering of the scapula with correction of the varus position of the glenoid by placement of an absorbable suture through the superomedial portion of the scapula [42].

Recently, Soldado et al [13] performed a minimally invasive approach endoscopic Woodward procedure in one patient with access through two small incisions at the level of the upper and lower angles of the scapula. Near-symmetrical shoulder elevation was achieved, with an excellent cosmetic result.

The vertical scapular osteotomy can correct cosmesis and shoulder function via the creation of scapular osteotomy at about 1 cm from the medial border in addition to the release of fibrous bands tethering scapula to the chest wall. The procedure associated with satisfactory results, however, the presence of complex rib anomalies may affect the postoperative outcome [44,51,52].

Another scapular osteotomy technique was described by Mears [43], including a partial superomedial scapular resection, removal of any
Omovertebral connection and release of the long head of triceps from the scapula, which results in early restoration of shoulder function with significant improvement of cosmetic appearance [43,52-54].

Post-operative complications

During the inferior migration of the scapula, there is a risk for compression and injury of the brachial plexus resulting in postoperative brachial plexus palsy; however, many of these palsies are transient [41,42]. Therefore, it is important to prevent brachial plexus injury through clavicular osteotomy in order to reduce traction on other structures, in addition to intraoperative monitoring of somatosensory-evoked potential [55].

Postoperative scapular winging is another complication after the surgical management of SD which is common after both Green’s and Woodward procedures. Winging may be a result of abnormal height to width ratio of the dysplastic scapula, or due to the mechanical or neurological failure of serratus anterior [38]. Winging does not occur with scapular osteotomy, and can be managed by excision of the medial scapular border [12,44].

Postoperative scars are common after surgical management of SD due to large incisions and extensive dissection which can result in hypertrophic or keloid scarring [42,54,56]. The type and degree of scarring differ with the surgical procedure, thus scarring after Green's procedure is usually large and prone to hypertrophy; however, it is at midline and less obvious after Woodward procedure. The scars after scapular osteotomy are smaller, prone to hypertrophy and do not extend into the neck [15,44].

CONCLUSION

Undescended scapula, or Sprengel deformity, is a rare but serious congenital abnormality of the shoulder girdle which may result in significant shoulder asymmetry and limited abduction. The deformity is usually associated with multiple syndromes and omovertebral bone. To obtain a proper diagnosis, a plain chest x-ray should be performed initially followed by CT with 3-D reconstruction or MRI to assess the degree of deformity, determine other abnormalities, and decide the plan for treatment. The mild cases with the non-significant limitation of the shoulder function need physical therapy with no surgical intervention, while surgery is reserved for moderate to severe cases before the age of 8 years-old. The surgical procedures aim to improve cosmetic and functional difficulties, by removal of the protruding parts of the scapula and any omovertebral bone in addition to the lowering of the elevated scapula. Postoperative outcomes depend on patient age, associated abnormalities and co-morbidities, with near similar results for each of the surgical procedure. Postoperative brachial plexus injury and scapular winging can be reduced by performing a clavicle osteotomy and use of scapular osteotomy procedures, respectively. The future introducing of the minimally invasive techniques in the management of undescended scapula may reduce postoperative scarring associated with contemporary large surgical incisions.

REFERENCES