Sprengel Deformity: Pathogenesis and Management

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Review Article

Sprengel Deformity: Pathogenesis and Management

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Abstract

Sprengel deformity (ie, congenital elevation of the scapula) is a rare clinical entity. However, it is the most common congenital anomaly of the shoulder. Sprengel deformity is caused by abnormal descent of the scapula during embryonic development. Sprengel deformity is associated with cosmetic deformity and decreased shoulder function. Diagnostic confusion with limited scoliosis can be dangerous to the patient because it may delay proper treatment of other abnormalities that may be present with even mild cases. Sprengel deformity is commonly linked to a variety of conditions, including Klippel-Feil syndrome, scoliosis, and rib anomalies. Nonsurgical management can be considered for mild cases. Surgical management is typically warranted for more severe cases, with the goal of improving cosmesis and function. Surgical techniques are centered on resection of the protruding portion of the scapula and inferior translation of the scapula. Recent long-term studies indicate that patients treated surgically maintain improved shoulder function and appearance.

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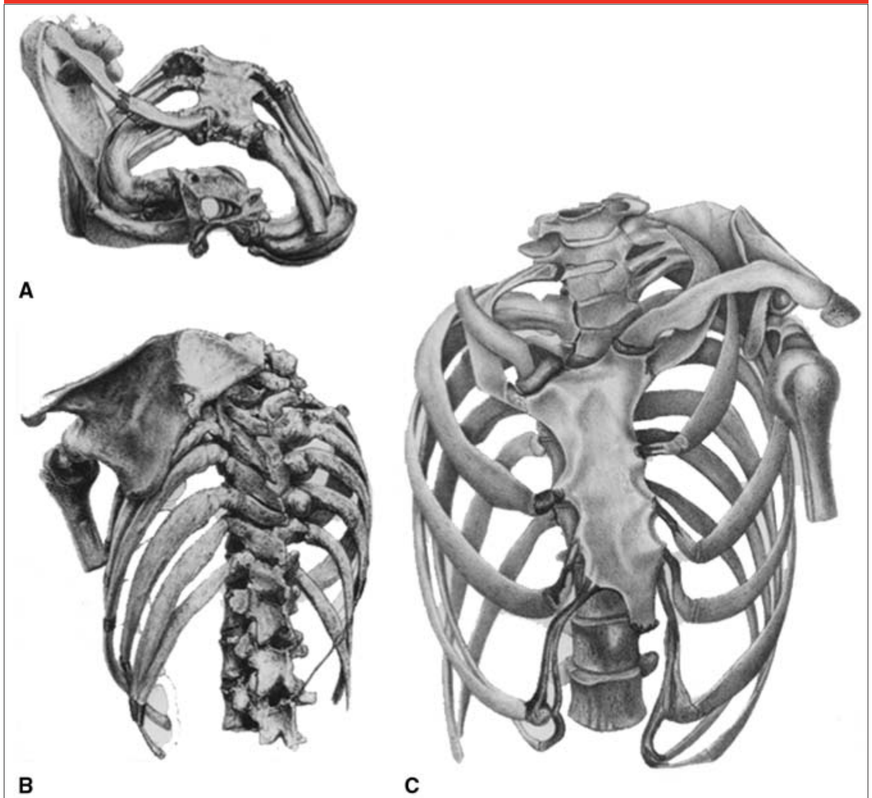
Sprengel deformity, that is, congenital elevation of the scapula, is the most common congenital abnormality of the shoulder girdle.[1,2] This rare condition is often found in association with more common disorders such as Klippel-Feil syndrome.[3,4] Recognition of cervical spine and upper extremity abnormalities is critical; such findings often indicate anomalies in other organ systems. Prompt, accurate diagnosis is essential.

Eulenberg[5] is credited with publishing the first description of congenital elevation of the scapula, in 1863. In 1891, Sprengel[6] described four cases of upward displacement of the scapula, and the condition became associated with his name. Several small case series and retrospective reviews were subsequently published describing the deformity and elucidating possible associated anomalies[7-17] (Figure 1). The disorder varies in severity from simple limited shoulder abduction and mild cosmetic deformity to more pronounced shoulder dysfunction and severe clinical abnormalities. The deformity can progress, and it is important to identify and follow even the patients with mild abnormalities. All patients warrant a detailed history and physical examination, taking care to evaluate for associated conditions in the spine and other organ systems. Surgery is indicated in severe cases. The Woodward and Green procedures are the most commonly performed surgical techniques.[9,10]

Etiology

Sprengel deformity is a developmental condition, and the associated

Figure 1



Illustrations from the year 1880 demonstrating dissection of a cadaver with Sprengel deformity and an omovertebral bone. This was the second case reported in the literature of a Sprengel deformity and the first of an omovertebral bone. The axial (**A**), posterior (**B**), and anterior (**C**) views clearly show the elevated, malrotated, and malformed scapula along with a bony connection to the spine. (Adapted with permission from Willett JA, Walsham WT: An account of the dissection of the parts removed after death from the body of a woman the subject of congenital malformation of the spinal column, body thorax and left scapula arch; with remarks on the probable nature of the defects in development producing the deformities. *Med Chir Trans* 1880;62:256-280.)

pathophysiology is related in particular to the embryology of the upper extremity. Fundamental to normal growth and development in the human embryo is a complex series of interactions between inducting cells (ie, germ layers) and responding tissues. Phenotypic mutations can occur as a result of lack of signaling molecules or ineffectiveness due to absent cellular receptors. Similar to the way in which acetabular dysplasia can cause proximal femoral abnormalities, Sprengel deformity often has an effect on surrounding structures, which require a normal scapula for development. Elevated scapula is the most obvious presenting sign, but hypoplasia of the scapula and surrounding musculature is also seen.

The scapula forms through a process of germ layer differentiation during the embryonic period between the third and eighth weeks of pregnancy. At this time, the scapula lies at the level of the fourth and fifth cervical vertebrae. Differentiation of the mesoderm into the axial and appendicular skeleton requires continued cellular signaling from surrounding tissues. The scapula is part of the appendicular skeleton and develops according to the programmed mesenchymal cell pathway. Pluripotent mesenchymal cells differentiate into skeletal tissues under the influence of various cell-signaling molecules such as bone morphogenetic proteins and fibroblast growth factors. The periphery of the limb bud also contains a group of specialized mesenchymal cells, the apical ectodermal ridge, which directs underlying limb outgrowth. Multifaceted cellular signaling pathways guide scapular growth and development, which in turn directs the growth and development of surrounding muscles, bones, and nerves. Altered or premature growth arrest in one or more tissues leads to a collection of abnormalities.

In normal development, the scapula migrates caudally during weeks 3 through 5 and continues to develop. At 6 weeks, the scapula further enlarges and approaches its final loca-

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Table 1

Associated Abnormalities in Persons With Sprengel Deformity[16,26,27]

Abnormality	Prevalence (%)
Scoliosis	35–55
Klippel-Feil syndrome	16–27
Rib anomalies	16–48
Omovertebral bone	20–50
Spina bifida	20–28
Torticollis	4
Clavicular abnormalities	1–16
Humeral shortening	6–13
Femoral shortening	1
Talipes equinovarus	1–3
Congenital dislocation of the hip	1–4
Pes planus	1–3
Other	1–3

tion at the level of the seventh thoracic vertebra. The scapula forms bone via intramembranous ossification. Normal muscle development occurs by expressing cellular receptors whose ligands originate mainly in surrounding tissues. With Sprengel deformity, normal muscular phenotype cannot be expressed because the conventional scaffolding is absent. The scapula fails to descend to its normal position, which leaves the hypoplastic scapula elevated and malrotated. Sprengel deformity, Klippel-Feil syndrome, Poland anomaly, and Möbius syndrome are some of the conditions that result from pathologic lesions early in embryonic gestation. Because of the cooperative growth of the skeletal system, such conditions lead to multiple phenotypic abnormalities. Vascular lesions arising from the subclavian artery distribution have been proposed to be a cause of these syndromes.[18-21] Other authors have suggested an autosomal dominant inheritance.[22] It is unclear at this time whether a vascular lesion or autosomal inheritance, or both, is responsible for Sprengel deformity, but they may not mutually exclusive.[22-25] Most cases are sporadic, and the etiology remains unknown.

Initial Assessment

Clinical Evaluation

Patients with Sprengel deformity typically present with cosmetic and functional impairment. In a series of 100 patients, Cavendish[26] found mild deformity in 59 and moderate to severe deformity in 41. The cosmetic abnormality is caused by the elevated and malrotated scapula. Typically, the inferior pole of the scapula is rotated medially, often approaching the midline. This results in a downward-facing glenoid cavity. The suprascapular region appears fuller secondary to the upward rotation of the superomedial angle of the scapula. The extent of resulting neck fullness varies depending on how proximal the scapula lies.

Affected children have varied degrees of functional limitation resulting from decreased range of motion (ROM) in the shoulder joint. Typically, shoulder abduction is mainly affected by lack of motion of the scapulothoracic junction and an inferiorly rotated glenoid. Shoulder abduction is usually limited to ≤90°.[16] These limitations affect many activities of daily living.

Patients may present with one or more associated abnormalities (Table 1). Scoliosis was found to be the most common associated abnormality in two large series of 112 patients and 75 patients (35% and 55%, respectively).[16,26] Rib anomalies are also common, with a reported prevalence of 16% to 48% in patients with Sprengel deformity.[16,26,27] Other associated disorders include Klippel-Feil syndrome[3,4,16,26,27] and diastematomyelia.[28]

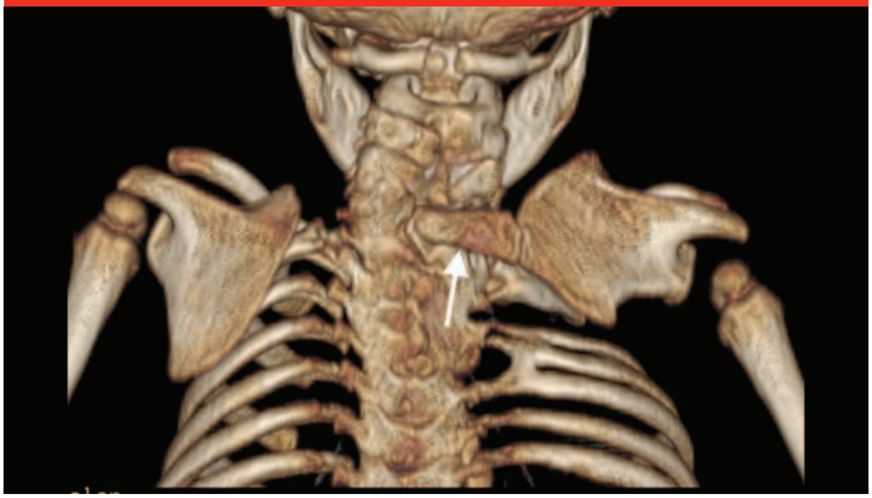
Approximately half of patients with Sprengel deformity have an omovertebral bone, which connects the scapula with the vertebrae, typically at the level of C4 through C7. The connection can be fibrous or cartilaginous, or it can be formed of mature bone. The omovertebral bone typically extends from the superomedial aspect of the scapula to the cervical spine, usually the spinous process, lamina, or transverse process of the associated vertebra. The omovertebral bone usually appears as an outgrowth from the scapula; however, it may form a pseudoarticulation. Recognition of an omovertebral bone is essential because it interferes with shoulder motion and may interfere with scapular descent. A variation of the omovertebral bone, extending from the superomedial aspect of the scapula to the base of the occiput, has been found in case reports.[29,30] In one patient, the omovertebral bone entered the spinal canal, causing myelopathy.[31]

Imaging

Radiographic evaluation is required in the initial assessment of any patient with suspected Sprengel deformity. Plain radiographs should be obtained initially to assess the level of the scapula in relation to the vertebrae and the contralateral side. Radiographs are also helpful to determine the presence of associated abnormalities (eg, scoliosis, rib abnormalities, omovertebral bone).

CT is used to identify associated abnormalities such as congenital scoliosis and cervicoscapular connections. It is also warranted for severe cases that require surgical management. CT can help delineate an associated omovertebral bone, especially its relation to the spine. Moreover, three-dimensional CT reconstructions are necessary to further evaluate the complex pathoanatomy and

Figure 2



Three-dimensional CT scan demonstrating a complete view of an elevated, malrotated, and malformed right scapula and omovertebral bone (arrow). Rib anomalies can be seen, as well. The right glenoid cavity is tilted inferiorly.

Table 2

Cavendish Classification of Sprengel Deformity[26]

Grade	Description
1 (very mild)	Shoulders are level. Deformity is not visible when the patient is dressed.
2 (mild)	Shoulders are almost level. Deformity is visible as a lump in the web of the neck when the patient is dressed.
3 (moderate)	Shoulder is elevated 2–5 cm. Deformity is easily seen.
4 (severe)	Shoulder is elevated. The superior angle of the scapula lies near the occiput.

to help develop the surgical strategy[32] (Figure 2).

Classification

Many attempts have been made to classify Sprengel deformity to aid in guiding management and evaluating outcomes. The Cavendish classification, which is based on cosmetic appearance (ie, severity of scapular elevation), is the most widely used classification system[26] (Table 2).

Rigault et al[33] developed a radiographic classification of Sprengel deformity based on the projection of the superomedial angle of the scapula on radiographs. Deformity is graded based on the relationship between the superomedial angle of the scapula and the associated vertebral level. Grade 1 deformity involves the superomedial angle below T1, grade 2 deformity lies between T1 and C5, and grade 3 deformity lies above C5.

Ross and Cruess[16] measured shoulder elevation based on the level of the center of the humeral head in relation to the trunk vertical axis. Sprengel deformity has a rotational component, and Leibovic et al[34] measured scapular displacement based on the vertical positioning of the scapula and the rotational component. They assumed that the center of rotation of the scapula was through the acromioclavicular joint. The superior and inferior scapular angles were developed to quantitatively measure and compare rotation on preoperative and postoperative radiographs. However, this measurement is difficult to use and has not been widely adopted.

Management

Nonsurgical

Nonsurgical management is reserved for children with mild deformity, minimal shoulder dysfunction, and little cosmetic deformity (eg, Cavendish grades 1 and 2). Physical therapy is instituted to maintain shoulder ROM and prevent torticollis. At our institutions, management consists of annual observation until the patient reaches skeletal maturity, to assess for progression of shoulder abnormality and associated deformities. Appearance of the shoulder is the typical presenting complaint, and particular attention should be paid to the child's psychological development.

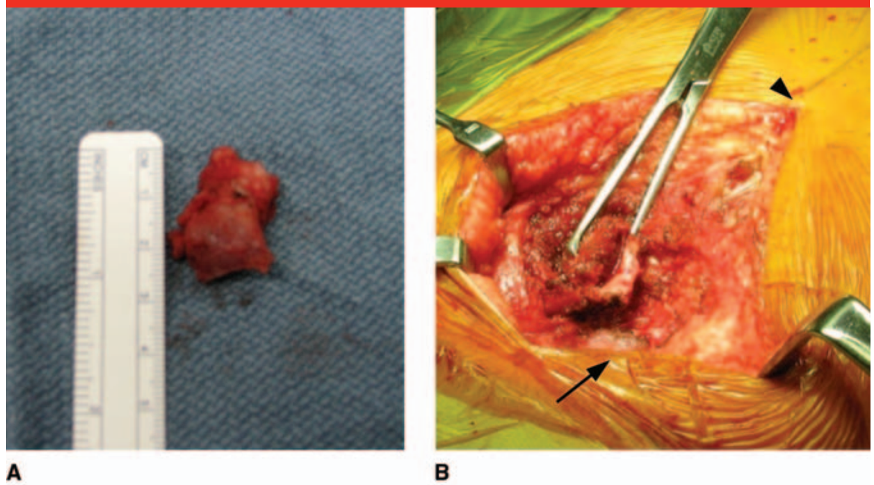
Farsetti et al[35] performed a retrospective review of 15 patients treated nonsurgically (17 scapulas). Three were Cavendish grade 1, nine were grade 2, and five grade 3. The mean preoperative combined shoulder abduction was 125° (range, 95° to 170°). At a mean follow-up of 26 years (range, 10 to 55 years), the mean shoulder abduction was 125° (range, 90° to 160°), and all patients with Cavendish grade 1 and 2 deformity remained in their respective categories.

Surgical

Surgery is the mainstay of treatment for all but mild cases, and it is performed to improve cosmesis and shoulder function. Surgical techniques involve resection of the elevated portion of the scapula, removal of omovertebral bone (Figure 3), and mobilization of the scapula to a more caudad position. To prevent brachial plexopathy when mobilizing the scapula, the surgeon must reference from the contralateral scapular spine and not from the inferior margin on the affected side because the scapula is malrotated and hypoplastic. In patients aged >8 years, a 2-cm mid clavicular resection osteotomy is recommended to decompress the brachial plexus and the first rib before scapular reduction.[7] The resected portion can be morcellized and placed back into the periosteal sheath. In younger patients, a 1-cm mid clavicular resection osteotomy is performed without replacing the bony gap.

Several surgical techniques have been developed to manage Sprengel deformity (Table 3). The Green procedure is a classic surgical technique for improving cosmesis in patients with Sprengel deformity.[7-9] The child is positioned prone. During draping, care should be taken to expose both scapulas to allow comparison during reduction. A midline skin incision is made overlying the spinous processes. The medial border of the affected scapula is identified, and the muscles are detached from their insertions. Extraperiosteal dissection identifies the trapezius muscle, which is then reflected medially. This exposes the underlying medial and superior scapular musculature (latissimus dorsi, serratus anterior, levator scapulae, supraspinatus, rhomboids), which also are resected extraperiosteally. The supraspinatus fossa is excised carefully to avoid injury to the neurovascular bundle laterally. Once free, the scapula is mobilized distally to the level of the unaffected scapula, using the superomedial angle of the scapula as a landmark. Once the scapula is in its new position, the muscular insertions are reattached to it, lengthening each muscle as needed

Figure 3



A, Photograph of omovertebral bone resected from a patient who underwent surgery for Sprengel deformity. **B,** Intraoperative photograph of the same patient with the omovertebral bone removed. The superomedial angle of the scapula (held in the Alice clamp) is seen clearly without any attachment to the vertebrae of the cervical spine. The scapula can also be moved more easily following removal of the omovertebral bone. The arrow points to the upper cervical spine, and the arrowhead indicates the inferior portion of the skin incision.

Table 3

Surgical Procedures Used to Manage Sprengel Deformity

Technique	Description
Green	1. Extraperiosteal release of all muscles at their scapular insertions 2. Resection of the supraspinatus fossa and omovertebral bone (if present) 3. Reattachment of muscles after caudad mobilization of the scapula 4. Muscle lengthening as needed
Leibovic modification of the Green technique	The reduced scapula is secured in a pocket developed in the latissimus dorsi muscle
Bellemans and Lamoureux modification of the Green technique	1. Serratus anterior muscle is not released 2. Immediate postoperative mobilization encouraged
Woodward	1. Resection of the superomedial portion of the scapula and omovertebral bone (if present) 2. Osteotomy of the clavicle 3. Detachment of the trapezius and rhomboid muscles 4. Mobilization of the scapula caudad 5. Reattachment of muscles back to vertebrae with the scapula in its new position
Mears	1. Subperiosteal release of muscle insertions on the superomedial aspect of the scapula 2. Osteotomy of the supraspinatus fossa 3. Release of the long head of the triceps and part of the origin of the teres minor 4. Inferomedial resection of the scapula to 160° of shoulder abduction

in order not to alter the corrected position. Care must be taken to prevent the glenohumeral articulation from drifting back into varus as the final reattachments are completed. Shoulder abduction is examined. Postoperative resisted pendular shoulder exercises are used for several weeks, after which active ROM exercises are encouraged.

Leibovic et al[34] modified the Green technique by suturing the reduced scapula into a pocket formed in the latissimus dorsi muscle. In 16 shoulders (14 patients), at a mean follow-up of 6.5 years, mean shoulder abduction was reported to be 148° (range, 100° to 180°), compared with 91° preoperatively (range, 60° to 120°). Although the initial results were encouraging, scapular rotation was not maintained at longer-term follow-up. Six scars were deemed to be hypertrophic, and symmetry was good to excellent in 12 patients. There were no nerve injuries or infections. Bellemans and Lamoureux[9] modified the technique by omitting the serratus anterior dissection and encouraging immediate postoperative shoulder mobilization. The authors reported a 77° increase in shoulder abduction in seven children.

The Woodward procedure, which was first described in 1961,[36] is perhaps the most widely used technique for the management of Sprengel deformity.[1,10,11,37-39] This procedure focuses on the release and relocation of the origin of the muscles attached to the medial border of the scapula as well as on resection of any omovertebral bone. The surgery is performed with the patient in the prone position, using a midline skin incision from the spinous process of C4 to T9. The lateral border of the trapezius is identified at the lower end of the incision and is separated from the latissimus dorsi. Sharp dissection is used to remove the origin of the trapezius muscle from the vertebral spinous processes. As the dissection is advanced cephalad, the rhomboids are removed from their attachments to provide a well-defined layer of deep fascia. Any omovertebral bone is removed extraperiosteally. The superomedial portion of the scapula may also be excised if it is prominent. Next, the shoulder girdle and muscle sheet can be moved inferiorly until the scapular spine is brought to the same level as that of the opposite side. The aponeurosis of the trapezius muscle and rhomboids is attached to its new position along the vertebrae. Clavicular osteotomy has also been advocated to prevent brachial plexus injury resulting from inferior displacement of the scapula. Intraoperative somatosensory-evoked potential monitoring has been shown to aid in preventing brachial plexus injury.[38] We have achieved satisfactory outcomes using the Woodward procedure combined with clavicular osteotomy (Figure 4).

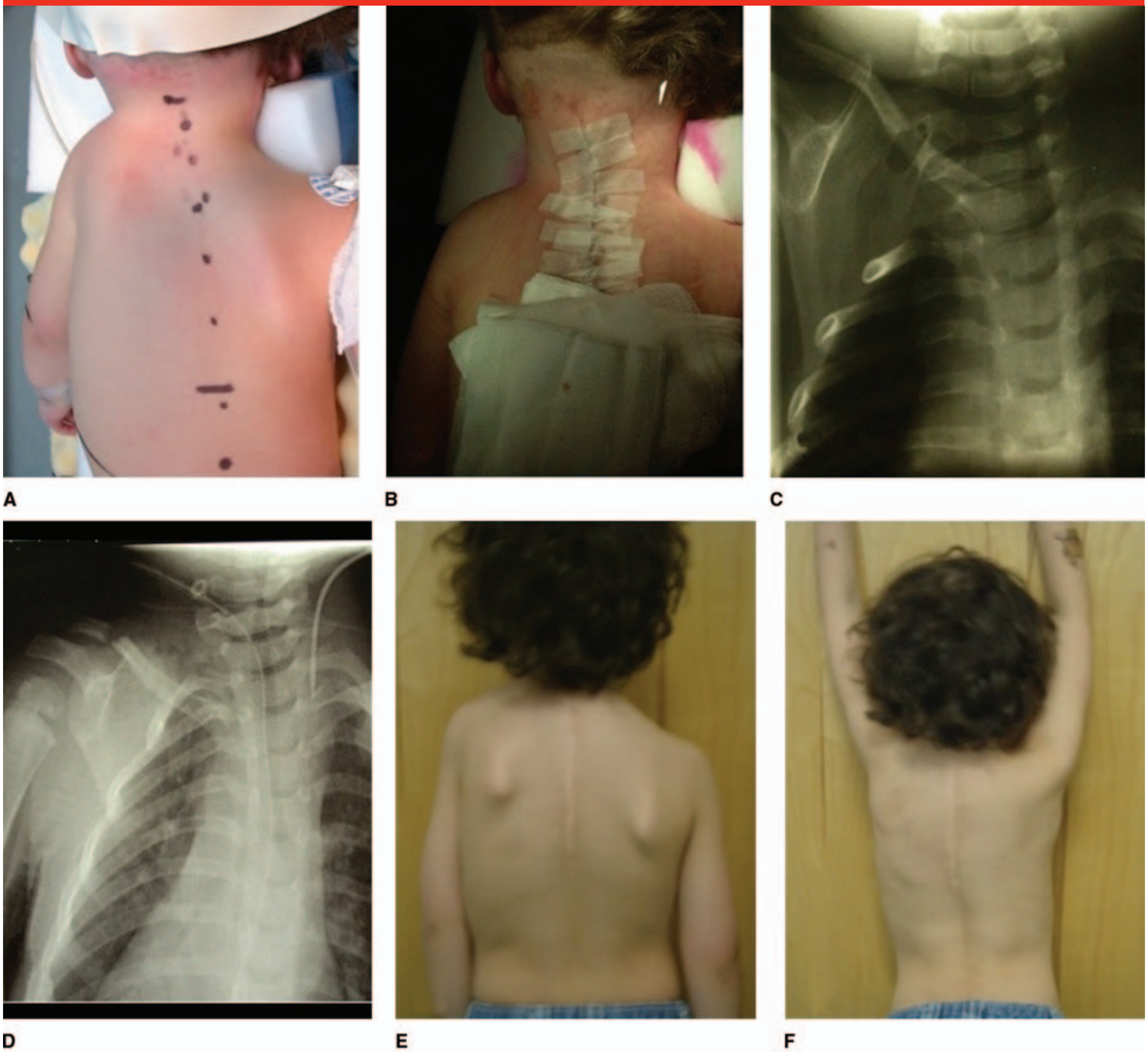
Recently, the Woodward procedure has been modified to improve the varus position of the glenoid.[12,40] Ahmad[40] placed an absorbable suture through the superomedial portion of the scapula at the junction of the upper and middle thirds. The suture was then tied at a 45° angle to the spinous process of either T11 or T12. All cases improved cosmetically and gained a mean of 49° abduction.

Scapular osteotomy to improve glenoid position has been described.[14,17] In this technique, initially described by Wilkinson and Campbell,[17] the patient is placed in a semiprone position with the affected limb facing up. A vertical incision is made slightly lateral to the vertebral border of the scapula, and dissection is done through the overlying fascia covering the infraspinatus muscle. The periosteum is then incised and reflected to either side. With the bone exposed, an osteotome is used to mark the planned osteotomy 1 cm from the vertebral border of the scapula. Offset holes are drilled into the bone along the osteotomy line. An osteotome is used to complete the osteotomy, starting at the inferior scapular angle and moving upward. All muscle attachments and fibrous bands are freed extraperiosteally from the superomedial angle of the scapula, which is then excised along with any omovertebral bone. After excision, the remaining medial portion is pulled inferiorly with forceps. Any fibrous bands between the subscapularis muscle and chest wall are divided using blunt dissection. The two opposing scapular edges are joined using silk sutures passed through the drill holes. The incision is closed after hemostasis is achieved. The patient remains in a sling for 6 weeks, after which full ROM is allowed.

McMurtry et al[14] reported adequate results in 12 patients who were followed for a mean of 10.4 years. The authors reported a mean increase in shoulder abduction of 53° and improved cosmetic appearance (mean, 1.5 Cavendish levels). These results did not deteriorate over time.

Mears[15] described a technique similar to scapular osteotomy. This procedure involves resection of the superomedial portion of the scapula and any existing omovertebral bone, scapular osteotomy with inferior descent, and release of the long head of the triceps muscle. The patient is placed in a prone position. Exposure is performed through a curvilinear incision, inverted L-shaped incision, or straight transverse incision centered over the scapular spine. The supraspinatus, levator scapulae, rhomboid, and subscapularis muscles are elevated subperiosteally from the scapula. Next, the subscapularis, serratus anterior, and infraspinatus muscles are elevated subperiosteally from the medial border of the scap-

Figure 4



A, Preoperative photograph of a boy with Cavendish grade 4 Sprengel deformity and an omovertebral bone. Note the elevation of the left shoulder compared with that of the right shoulder. **B,** Intraoperative photograph following the Woodward procedure using a midline skin incision. The left shoulder is depressed and lies at the same level as the opposite side. **C,** Preoperative posteroanterior radiograph demonstrating the elevated left scapula. **D,** Postoperative posteroanterior radiograph demonstrating the scapula repositioned at the proper anatomic level. Left clavicular osteotomy was also done to prevent brachial plexus injury during scapular reduction. **E,** Photograph obtained 3 years postoperatively demonstrating slight proximal migration of the scapula. However, cosmesis was still significantly improved compared with the preoperative state. **F,** The patient achieved normal range of motion and significantly improved abduction.

ula. An osteotomy through the supraspinatus fossa is performed from the remaining scapula, separating it into medial and lateral components. Any existing omovertebral bone should be resected. The long head of the triceps muscle and part of the teres minor are removed from the scapula. The inferomedial portion of the scapula is resected progressively until impingement-free ROM occurs to 160º of shoulder abduction. Gentle ROM exercises are allowed early on the second day postoperatively and are continued for 6 weeks. Re-

Table 4

Complications of Surgical Correction for Sprengel Deformity[10,11,14,16,36,40]

Complication	Reported Prevalence (%)
Hypertrophic scar	26–64
Brachial plexus injury	6–11
Regrowth of the superior pole of the scapula	30
Scapular winging	4–17

cently, Masquijo et al[13] reported statistically significant improvements in flexion and abduction in a prospective cohort study involving 21 patients treated with this technique ($P < 0.001$). Cosmesis also improved by an average of 2 levels on the Cavendish scale. Two patients developed keloid scar, and two others required a second surgery to remove a remaining exostosis.

Partial scapulectomy has also been advocated.[41] The patient is positioned prone, and the involved shoulder and upper limb are draped free. An inverted L-shaped incision is made that exposes the muscles along the medial side of the scapula (ie, trapezius, levator scapula, rhomboids). Subperiosteal dissection is done to expose the supraspinatus fossa, and the superior spine of the scapula is resected as far lateral as the acromial region. The superior border of the operated scapula is compared with the unaffected side to confirm adequate reduction. Any existing omovertebral bar is resected extraperiosteally. Postoperatively, no ROM is allowed for 2 weeks, and the shoulder is not immobilized. Zhang et al[41] reviewed 26 patients at a mean follow-up of 3.9 years. The rate of shoulder abduction improved 59%, and abduction improved from a mean of 110° preoperatively to 150° postoperatively. One patient had Cavendish grade 4 involvement, 10 had grade 3, and 15 had grade 2. No neurologic or scar complications were reported. An omovertebral bar was found in 12 shoulders.

Authors' Preferred Treatment

We typically manage mild cases (ie, Cavendish grades 1 and 2) nonsurgically and encourage the children to participate in sports such as swimming to maintain ROM. For moderate to severe cases (ie, Cavendish grades 3 and 4), we encourage activity and counsel the family about the eventual need for surgical management. We prefer to operate at an early age (4 to 6 years); however, we are unsure whether it is easier to operate on a younger or an older child (ie, surgical timing). In addition, we are unsure whether a relationship exists between the incidence of recurrence and timing of surgery.

Our preferred technique is the modified Woodward procedure with a clavicular osteotomy. Both procedures are typically performed with the patient in a prone position. A long midline incision is made from approximately C4 to the lower thoracic spine to adequately isolate the trapezius muscle (Figure 4). Suprafascial dissection is performed to gain access to the clavicle by tunneling under the subcutaneous layer through the posterior wound until the clavicle is reached. At that point, subperiosteal dissection of the clavicle is performed to protect the adjacent soft-tissue structures, and a Kerrison rongeur is used to resect a 1- to 2-cm portion of the mid clavicle. In patients <5 years of age, we resect approximately 1 cm; in patients >8 years of age, we resect 2 cm of the clavicle. Next, the trapezius and rhomboid layers are elevated off the midline spinous processes as a single layer. Proximally, the levator scapula muscle is transected and any existing omovertebral bone or cartilaginous/fibrous tether is resected extraperiosteally. The scapula is everted and mobilized distally and out of varus. Typically, we leave the supramedial border of the scapula unmorcellized; however, morcellization can be done to improve cosmesis. The inferior tip of the scapula is tethered to two ribs using a No. 5 braided nonabsorbable suture to help secure the scapula in its new position. The trapezius and rhomboid muscles are transposed and reattached distally on the spinous process to accommodate the new position. The redundant trapezius at the inferior edge of the wound is incised and repaired in pants-over-vest fashion to further secure the repositioned scapula.

Postoperatively, the patient is placed in a shoulder immobilizer to assist with postoperative discomfort. Formal physical therapy, consisting of passive and active ROM exercises, is begun at 6 weeks.

Complications

Complications resulting from the surgical management of Sprengel deformity largely include hypertrophic scarring, regrowth of the superior pole of the scapula, brachial plexus injury, and scapular winging (Table 4). Most surgical techniques involve large incisions and extensive dissection, which can lead to hypertrophic and keloid scars. In the long-term follow-up study by Farsetti et al,[35] which included 22 patients, 4 of the 8 patients treated surgically were found to have an unsightly scar. More recently, Ahmad[40] reported keloid scar in 4 of 11 patients.

The brachial plexus is at risk of compression intraoperatively as the scapula is migrated inferiorly. Several authors have reported brachial plexus palsy after surgery for Sprengel deformity; however, many palsies were transient.[7,14,40] Even so, we think that clavicular osteotomy should be considered for brachial plexus injury prevention. Intraoperative somatosensory-evoked potential monitoring may help to prevent such injury.[38] Other postoperative outcomes include scapular winging,[40] recurrence of the deformity, and failure of functional improvement.

Summary

Sprengel deformity is the most common congenital abnormality of the shoulder girdle and can cause significant cosmetic and functional impairment. Proper diagnosis is essential because the condition can present with multiple associated abnormalities and lead to significant morbidity, if not properly managed. Initial assessment should include plain radiographs as well as two- and three-dimensional CT to aid in surgical planning and to identify the presence of omovertebral bone. Nonsurgical management is appropriate for mild cases, with surgery reserved for severe cases. Several surgical techniques have been developed with the goal of improving cosmesis and shoulder function. In general, these procedures involve removal of the protruding portion of the scapula along with any omovertebral bone as well as translation of the scapula inferiorly to a more caudad position. The Green and Woodward techniques are the most widely reported procedures. Osteotomy of the clavicle can be performed as well to prevent brachial plexus injury. Hypertrophic scarring can occur as the result of large incisions and extensive dissection. Good clinical outcomes can be achieved postoperatively, with significant improvement in physical appearance and shoulder movement. Prior to treatment, the surgeon should talk with the family about expected outcomes and potential complications.

References

Evidence-based Medicine: Levels of evidence are described in the table of contents. In this article, references 16 and 27 are level III studies. References 1-15, 17-26, 28-31, 33-36, and 38-41 are level IV studies.

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