

Sprengel's Deformity

- What are the treatment options?

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Disclosures

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Background

- First description in 1863 (Eulenberg, Aerzte Karlsbad 1863)
- Named after Otto Sprengel, a German Surgeon, who described a case in 1891
- Failure of caudal migration of the scapula (9-12th weeks of gestation).
- Scapula is hypoplastic, rotated
- Between 33% and 50% of the patients have an associated omovertebral bone (Wada et al. JPO 2013)



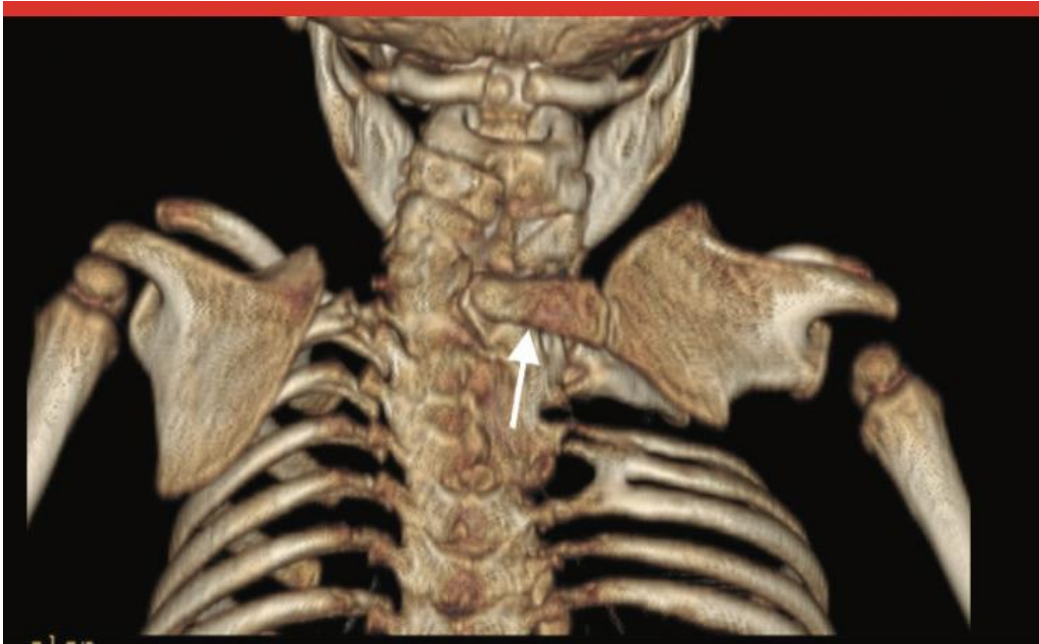
Clinical picture



- Clinical picture: High located and small scapula with limited shoulder abduction, and deformity cosmetic concern
- Associated anomalies: Klippel-Feil, Congenital scoliosis and Renal anomalies (Hensinger et al. JBJS 1974)

Courtesy of Dr. Dror Ovadia, Tel Aviv

Associated anomalies



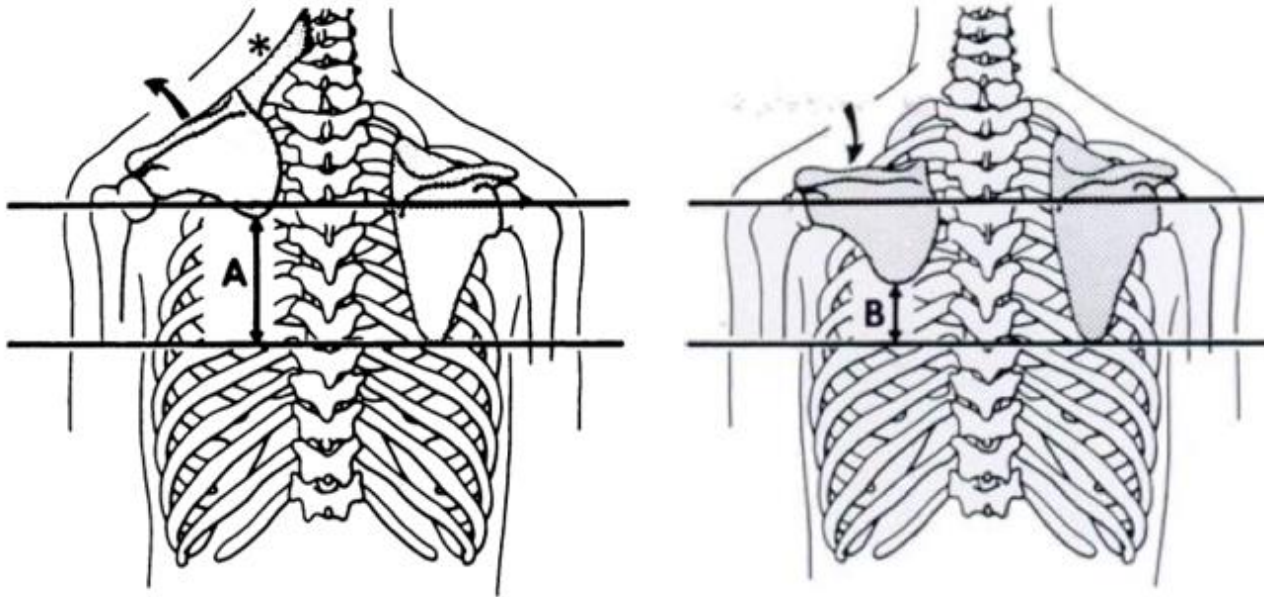
Harvey et al. JAAOS 2012

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

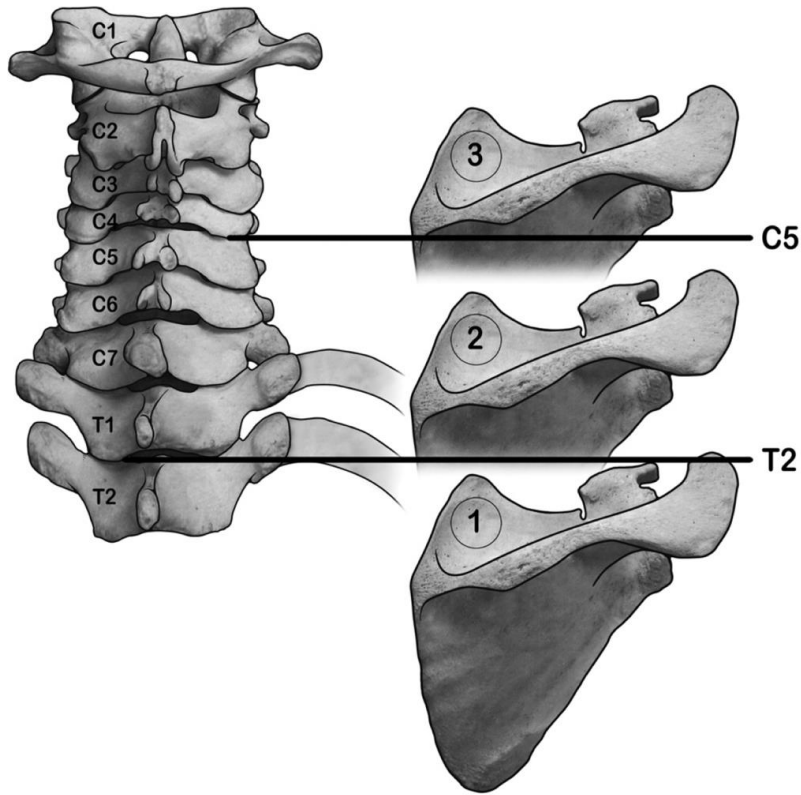
Biomechanics of functional limitation



Carson et al. JBJS
1981

- 1) Limited shoulder abduction due to external rotation resulting into downward inclination of the glenoid
- 2) Reduced scapulothoracic motion (Omovertbral bone)

Classification



Rigault's classification. Modified
from Di Gennaro et al. JBJS Br 2012

- Clinical picture: Cavendish classification (JBJS Br 1972)
 - Gr 1: no visible deformity
 - Gr 2: Bumpy aspect
 - Gr 3: Asymmetric shoulder elevation, 2-5 cm
 - Gr 4: Asymmetric shoulder elevation >5 cm
- Radiographic: Rigault's classification based on superomedial angle location (1976)
 - Gr 1: Lower than T2 but above T4
 - Gr 2: Between C5 and T2
 - Gr 3: Above C5

Imaging studies



- Cervical and Chest radiographs
- CT scans provide useful information on the omovertebral bone and position of the scapula
- MRI provides information on additional fibrous bands of the omovertebral bone between the scapula and cervical spine
- Abdominal and cardiac ultrasound to exclude associated organ anomalies

Conservative treatment



- For mild deformities (Cavendish gr 1-2)
- Physiotherapy aims to maintain shoulder ROM and prevent torticollis
- Annual follow-up until skeletal maturity
- 15 pts (17 scapulas) maintained their shoulder abduction and remained in the same Cavendish grade during 26 yr FU (Farsetti et al. JPO B 2003)

Surgical indication and timing

- Abduction of the shoulder < 90 degrees and Cavendish grade 3 or 4 deformity
- Optimal timing of the surgery between 3-4 years of age
- Surgical methods:
 - 1) Putti (subperiosteal insertion release) (Shrock JBJS 1926)
 - 2) Green (extraperiosteal muscle release) (Green JBJS 1957)
 - 3) Woodward (muscle origo release) (Woodward JBJS 1961)
with/o resection of the superomedial corner of the scapula
 - 4) Mears' scapular osteotomy (Mears JPO 2001)
- Clavicular osteotomy (morsellization) reduces the compression on brachial plexus and subclavian artery (Robinson et al. JBJS 1967)

Green's procedure



Straight incision over medial border of scapula (Green JBJS 1957)

1) Extraperiosteal release of muscles along the medial border of scapula

2) Omovertebral bone excised

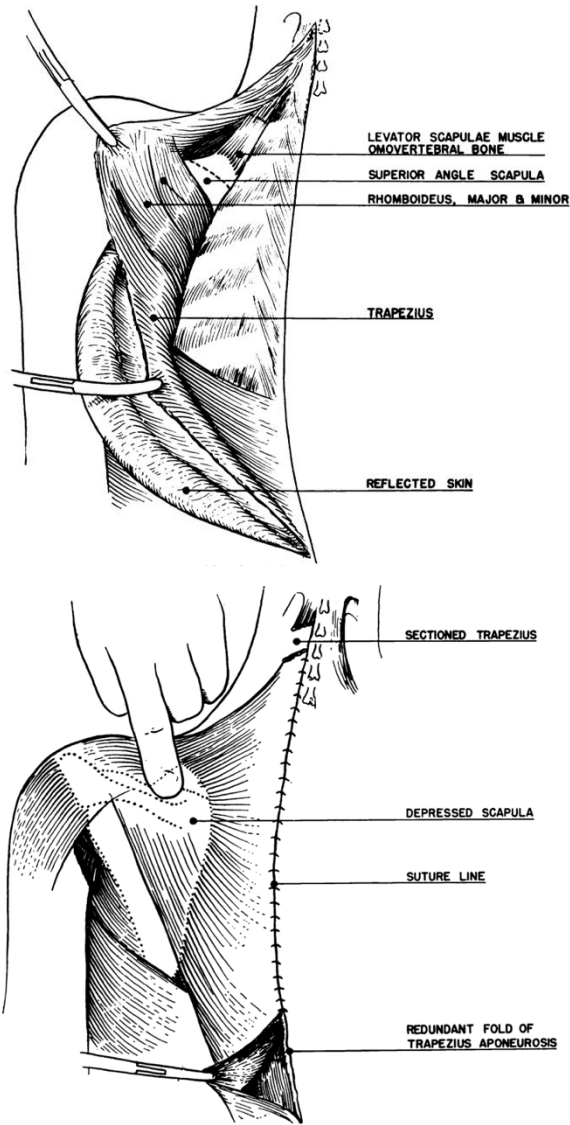
3) Superomedial corner of scapula resected

4) Relocation with lateralisation of the inferior corner, attached to the ribs



Courtesy of Dr. Dror Ovadia, Tel Aviv

Woodward procedure



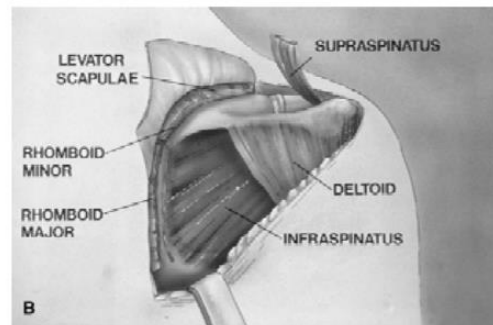
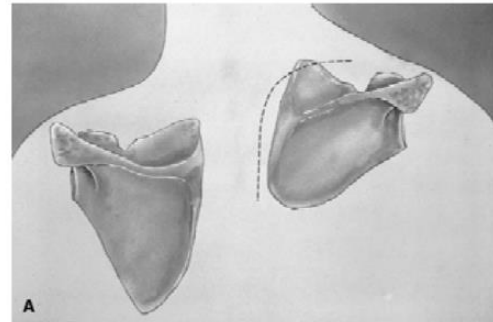
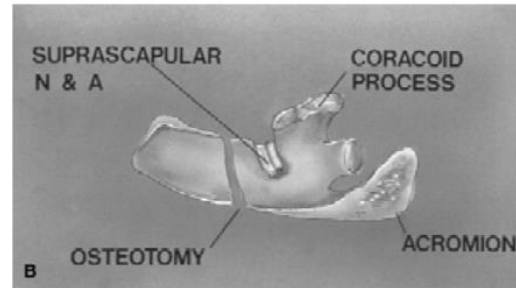
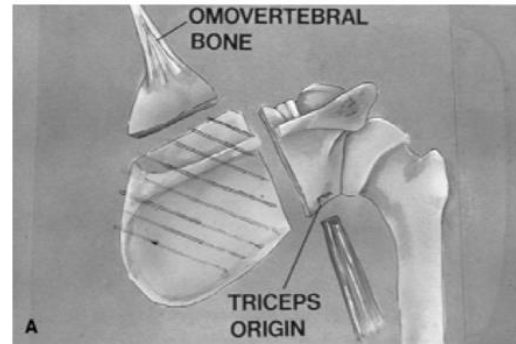
Midline incision over C4-T9 (Woodward JBJS 1961)

- 1) Origins of trapezius and rhomboid muscles released
- 2) Omovertebral bone excised
- 3) Transsection of trapezius at C4 level
- 4) Reattachment of muscles

Endoscopic technique described (Soldado et al. JPO B 2017)

Mear's Technique

- 1) Excision of superomedial angle of scapula and omovertebral bone
- 2) Oblique osteotomy of scapula & resection of part of the scapula
- 3) Release of long head of triceps



Mears DC. JPO 2001

Surgical treatment of Sprengel's shoulder

EXPERIENCE AT THE RIZZOLI ORTHOPAEDIC INSTITUTE 1975–2010

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Fig. 2a



Fig. 2b



Fig. 2c

Clinical photographs depicting the assessment of Cavendish grade⁸ in a five-year-old girl a) at presentation, showing a left Sprengel deformity with Cavendish Grade 3, b) at one month post-operatively, and c) at 11 years post-operatively, showing the deformity at Cavendish grade 1 with an acceptable surgical scar.

- 56 children treated using Green procedure at mean age of 6.4 yrs with 10.9 yrs FU
- Abduction of shoulder improved from 92° to 112° and flexion 121° to 155°
- Cosmetic improvement \geq one Cavendish grade in 54 (88.5%) shoulders
- Incomplete upper brachial plexus palsy in 3 (5%) children resolved within 7 mths

Sprengel Deformity: Pathogenesis and Management

Edward J. Harvey, MD, MSc,
FRCSC

Mitchell Bernstein, MD

Nicholas M. Desy, MD

Neil Saran, MD, MHSc, FRCSC

Jean A. Ouellet, MD, FRCSC

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

Conclusions



- Important to evaluate associated anomalies: Klippel-Feil, Congenital scoliosis and Renal anomalies
- Surgical management using both Green and Woodward techniques provides satisfactory outcomes in terms of shoulder function and deformity
- Risk of brachial plexus complications can be reduced by clavicular osteotomy