MOHAMMED V UNIVERSITY - RABAT
FACULTY OF MEDICINE AND PHARMACY - RABAT-

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SPRENGEL'S DEFORMITY :
Management of Neglected Cases
A case report and literature review

THESIS

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BY

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FOR THE

DEGREE OF MEDICINE DOCTOR

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تَوَكَّلْتُ وَإِلَيْهِ أُنِيبُ﴾

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RABAT

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INTRODUCTION

Congenital elevation of the scapula also known as Sprengel's deformity (SD) is a complex congenital anomaly of the shoulder girdle, characterized by upward displacement and rotation of the hypoplastic scapula due to failure of descent during embryonic development. First described by Eulenberg in 1863 [1] and Sprengel in 1891[2], Sprengel's deformity often is associated with other anomalies of the skeletal system and other systems. [3]

Though rare, it is the commonest anomaly of the shoulder girdle. Though both shoulders can be affected, the left shoulder seems to be affected more often. Among the sexes, it is more common among females with the female: male ratio being about 3:1. [4]

SD is usually recognized in childhood [3], due to cosmetic distress and disfigurement (shoulders asymmetry) or functional deficit by limitation of movement of the shoulder.

Surgical management of this anomaly typically is performed during childhood, between 3 and 8 years of age. [3] [5]

Thus, most of the literature on the treatment of SD has focused on pediatric cases. Few reports have described the management of previously untreated adolescent and adult cases [4].

The aetiology of SD is yet to be fully elucidated. A molecular biological study has implicated the axial skeleton of neural crest origin in the development of SD.

Neural crest migration anomalies have been implicated in some defects associated with deletion 22q11.2 syndrome (del22q11.2) [6] [7] [8].

It is the result of a scapula blocked from its normal descent during the embryonic period: a disruption of the migration of the scapula occurs from the embryonic limb bud level, which also causes failure of normal bone and soft tissue development in the shoulder girdle. [7]

In 20-30% of the patients, an omovertebral ‘‘bone’’ is found, either consisting of bone, cartilage, or fibrous tissue, that attach the affected scapula to the cervical spine and makes abduction of shoulder beyond 90 degrees virtually impossible [3][9][10]

Surgical treatment of deformity seeks to improve aesthetics and function, and may consist of osteotomies, bone resections, muscle releases with repositioning of the scapula, or a combination of these. [11] [12] [13] [14] [15] [16]

Our study consists in a case report of neglected Sprengel deformity (a 22 years old male with untreated SD), in addition to a review of literature, that allowed us to collect some adolescent and adult cases, in order to illustrate epidemiological, clinical, radiological, and especially the therapeutic aspects of this deformity when not treated in the recommended age.

HISTORY

No discussion of this deformity would be complete without mentioning that it would be more accurately named ''Eulenburg's deformity.'' Indeed, Moritz Michael Eulenburg's first descriptions in 1863 of three cases [1], as well as Willet's and Walsham's anatomical dissection and therapeutic omovertebral bone excision in 1880 and 1883 [17] [18] (by reporting 2 cases with anatomic descriptions of this clinical entity) antedate Otto Gerhard Karl Sprengle's four cases in 1891 by several years. Kolliker[19] in 1891, reporting two cases later in the same year and in the same journal, referenced Sprengel's notable article in which Sprengel denied an awareness of previous descriptions of the deformity and gave the condition its eponym, the Sprengel deformity [2]. It was in Kolliker's report that Sprengel was credited with the somewhat undeserved eponymous memorial.

Nevertheless, Sprengel, unlike those who preceded him, recognized that the deformity was caused by the failure of the scapula to descend [1].

Analyses of groups of patients not exceeding 20 cases predominate among subsequent reports and publications concerning SD. The description of the most numerous group containing 112 patients from 7 research centres in Great Britain was published by Cavendish [20]. Due to characteristic symptoms, the disease is diagnosed and treated prior to completion of 10 year of life.

To the best of our knowledge, Doita was the first to described two cases of surgical treatment of SD at adulthood [21].

The descriptions of Richard III suggest that he may has had Sprengel's deformity (the King Richard's physical affliction). He is described as "a valiant crook-back prodigy", of slight build, for which he compensated by undertaking vigorous exercise, especially fighting. He did not appear functionally compromised. [22]

ANATOMOPATHOLOGY

The embryonic primordium of the scapula appears during the fifth week of intrauterine life and acquires the final morphology by the end of the eighth week (end of embryonic period). It initially appears at the level of the fourth to fifth cervical vertebrae. During the same period, the brachial plexus gives rise to the peripheral nerves of the upper limb, which operates as a stimulus for development of the upper limb muscles. During its growth, the scapula descends over the upper five ribs to reach the correct anatomical position that holds at birth. This developmental migration of the scapula reflects a progressive adaptation of the bone, which serves for brachiation and not for bearing weight. The increased demands for greater range of movement and flexibility in humans have altered the morphology and dynamics, both of the shoulder joint and the scapula. The scapula acquired a spine, enlarged in size, especially in the part beneath its spine, widened (high breadth to length ratio), and moved backward, standing at an angle of approximately 45°relative to the midline (see Fig 1).

Failure of scapula to descend leads to Sprengel deformity, in which case the bone sits 2–10 cm higher than expected. The maintenance of high position of the scapula in the process of skeletal development leads to a series of other musculoskeletal defects including hypoplasia, medialization, and adduction of the scapula, prominence of its upper angle, distal rotation and lateral angulation of the glenoid cavity, changes in the position of the clavicle, anomalies of the cervicothoracic vertebrae and ribs, and muscular hypoplasia or atrophy of the shoulder musculature. A fibrous structure may bridge the cervical spine with the undescended scapula, which when ossified is called the omovertebral bone (omo derives from the Greek word for shoulder "ώμος"). The result in practice is limitation of the abduction at the shoulder joint and "growing out" of the shoulder blade. [5] [23] [24]

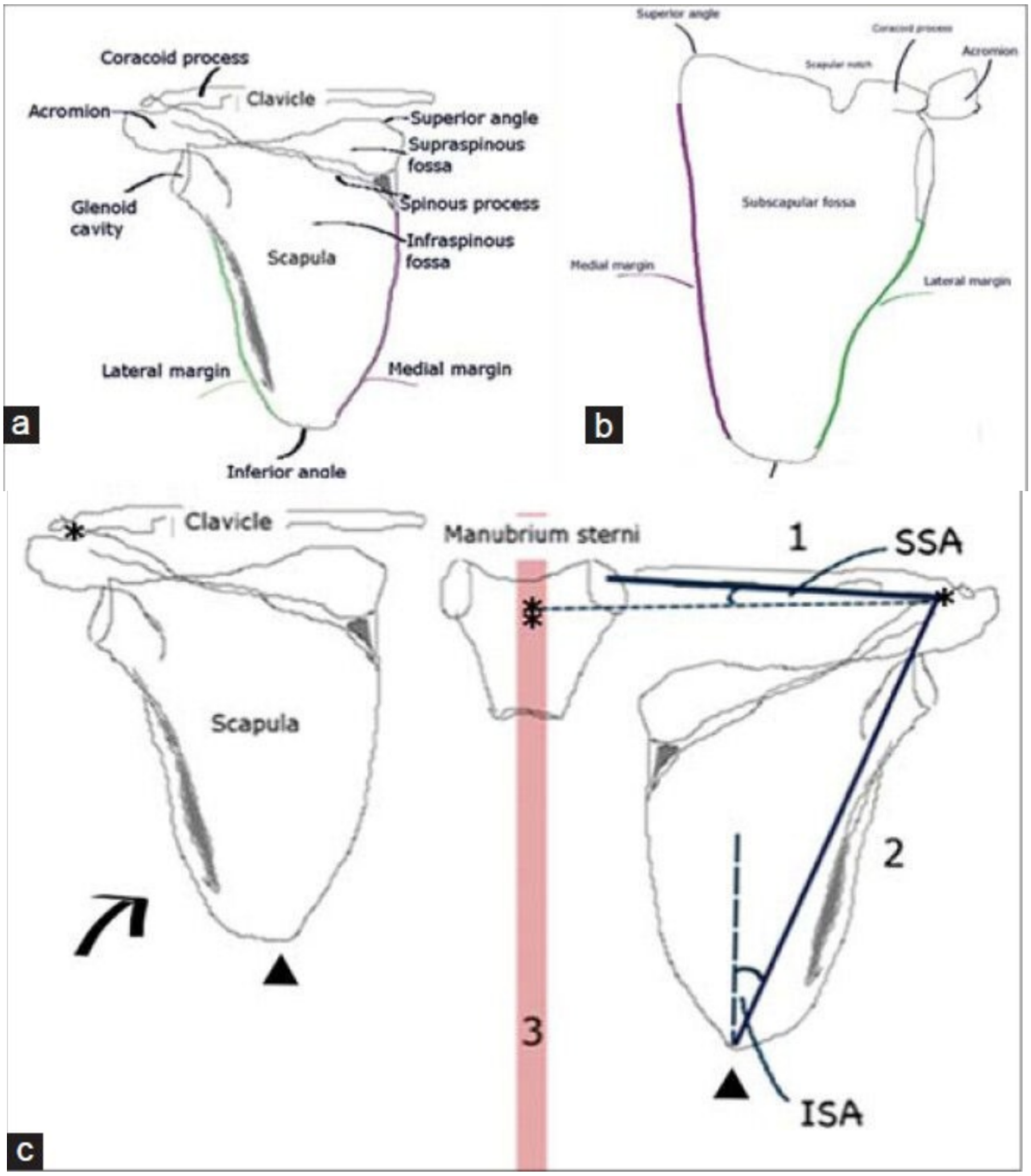


Figure 1: Line drawings show normal anatomy, (a) dorsal and (b) ventral views of the left scapula, and (c) Sprengel deformity. The affected scapula (arrow) sits 2-10 cm higher than normal in medial rotation: 1, the line from the midpoint of the acromioclavicular joint (asterisks) to the midpoint of the sternoclavicular joint (double asterisk); 2, the line connecting the middle of the acromioclavicular joint (asterisks) to the inferior angle of the scapula (arrowheads); and 3, a line along the spinous processes of the vertebrae (red line). SSA: Superior scapular angle, ISA: Inferior scapular angle [85].

PHYSIOLOGY

Shoulder is composed of four joints, namely glenohumeral joint, acromioclavicular joint, sternoclavicular joint and scapulothoracic joint.

Shoulder joint is the most mobile joint in human body and responsible for movements of arm and scapula.

It is very mobile and for attaining so much mobility, the stability of shoulder joint has been compromised by the body. This instability is compensated for by rotator cuff muscles, tendons, ligaments, and the glenoid labrum.

Movements of the scapula are brought about by the scapular muscles. The scapula can perform six actions:

- Elevation: upper trapezius and levator scapula; This movement allow the shoulder girdle to move upwards as in shrugging the shoulders.
- Depression: lower trapezius; This brings a reverse of the elevation movement. The pectoral girdle and entire shoulder downwards.
- Retraction (adduction): rhomboids and middle trapezius; This movement pulls the scapula back towards the rib cage. Pure adduction is not possible, associated with a retro-drive or ante-drive can reach 40 °.
- Protraction (abduction): serratus anterior; This is moving the shoulder blade (scapula) forwards, away from the rib cage and spine.

The first 0-15 degrees of abduction is produced by the supraspinatus. The middle fibres of the deltoid are responsible for the next 15-90 degrees. Past 90 degrees, the scapula needs to be rotated to achieve abduction, it can reach 180 °. that is carried out by the trapezius and serratus anterior.

- Upward rotation: upper and middle trapezius; This movement Lift the arms overhead; scapulae will follow and rotate upward.
- Downward rotation: rhomboids; This is rotating the lower scapula towards the rib cage as in moving the arm behind the back.

In Sprengel deformity, passive glenohumeral abduction, external and internal rotational motions are normal, while scapulothoracic joint movements can be very severely affected.

The report of glenohumeral / scapulothoracic joint use in the abduction movement was measured by Popen and Walker at 5/4 after thirty degrees of abduction, which means that the humerus moves by 5 °, when the scapula slides 4 ° on the thorax. But from 0 to 30 °, most of the movement is of glenohumeral origin. For a combined abduction of more than 90 °, the scapula must be free to rotate. Thus, fixation and medial rotation of the scapula limits abduction by at least 100 ° in many patients with congenital elevation of the scapula. The limitation of the abduction is, in general, proportional to the severity of the deformity and in particular to the degree of the medial rotation of the scapula. The glenoid cavity looks down, which is an additional cause of the limitation of the shoulder's abduction. If this bone is located high in the cervical region, it can also limit the rotational movements of the neck, finally another cause of limitation of the movements is represented by the muscular abnormalities and the anterior curvature of the supraspinatous fossa [25] [26] [27] [28] [29].

PATHOPHYSIOLOGY

The scapula is a cervical appendage that normally differentiates opposite the fourth, fifth, and sixth cervical vertebrae at about 5 weeks' gestation.

From the 6th week, it migrates caudally to reach its definitive physiological position at the 12th week. The inferior angle of the scapula is then situated between the 6th and 8th thoracic vertebrae. During this caudal migration, the morphology of the scapula changes to become adapted to the prehensile function of the upper limb

From the 12th week of inter-uterine life, the morphology of the scapula is similar to that of an adult scapula, higher than it is wide; any impediment to its descent results in a hypoplastic, elevated scapula, known as the Sprengel deformity [6][7].

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. 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 location 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 [30]. Other authors have suggested an autosomal dominant inheritance. 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. Most cases are sporadic, and the aetiology remains unknown [19] [31].

MATERIALS & METHODS

I. MATERIAL:

A. The objectives of this thesis:

- To draw the epidemiological profile of neglected Sprengel deformity, from our case report and published cases in the literature.
- To know the clinical and radiological aspect, and the evolution of the deformity.
- To study the different therapeutic approaches and their results in neglected cases (adolescents and adults).

B. Selection of cases:

- Our case report:

A 22 years old male with no previously treated Sprengel deformity.

- Literature review:

We searched for indexed <u>full articles and abstracts,</u> in the search engines Pubmed, Google Scholar, EMC, Ovid and EBSCO. We were interested in articles in French, English, German, Spanish and other languages which were published until 2017, according to the following key words: Sprengle's deformity in adults, Neglected Congenital High Scapula, Sprengel disease, Omovertebral bone, Klippel-Feil syndrome, Woodward, Green procedure, Surelevation congenitale de l'omoplate negligée/chez l'adolescent / l'adulte, Os omovertebral, Choroba Sprengla, Kość omowertebralna, deformidad de Sprengel con adolescentes y adultos.

As a result, we could obtain 22 publications; 19 are in English, 01 in German, 01 in Spanish and 01 in Korean language.

Of all the publications founded: 18 articles are isolated case, while 4 are series of cases.

II. METHODS:

From our case report and from a review of the literature, we will try to describe the main epidemiological, clinical and radiological aspects of the deformity and especially its surgical management, while highlighting the results of our study.

So, we researched Sprengel deformity in adolescents and adults, basing on an International database using the following exploitation sheet:

- Gender
- Age
- Affected Side
- Main Symptoms
- Preoperative Clinical & Plain Radiological features
- Associated anomalies
- Management
- Postoperative Results

Only cases that meet at least 6 of our inclusion criteria, were considered.

A. Inclusion criteria:

- The age of patients must be higher or equal to 15 years old.
- No-previously treated Sprengel deformity
- At least 6 parameters of the exploitation sheet must be exploitable.

B. Exclusion criteria:

- Pediatric cases
- Previously treated patients
- Parameters of the exploitation sheet not available.

Unfortunately, we had difficulties to use all the database because of limited access in our country, so we eliminated all articles that only the abstract is available, because of lack in features. Nevertheless, after application of inclusion & exclusion criteria, besides our case report, we were able to retain 15 cases of neglected SD from the literature; 3 adolescents, 10 adults and 2 elderly cases. 5 cases from a series of 9, a series of 2 cases, and the rest were isolated cases.

In total, we have 18 shoulders to study, including our report and two patients with SD in both shoulders.

we regrouped the cases obtained from literature in Table1.

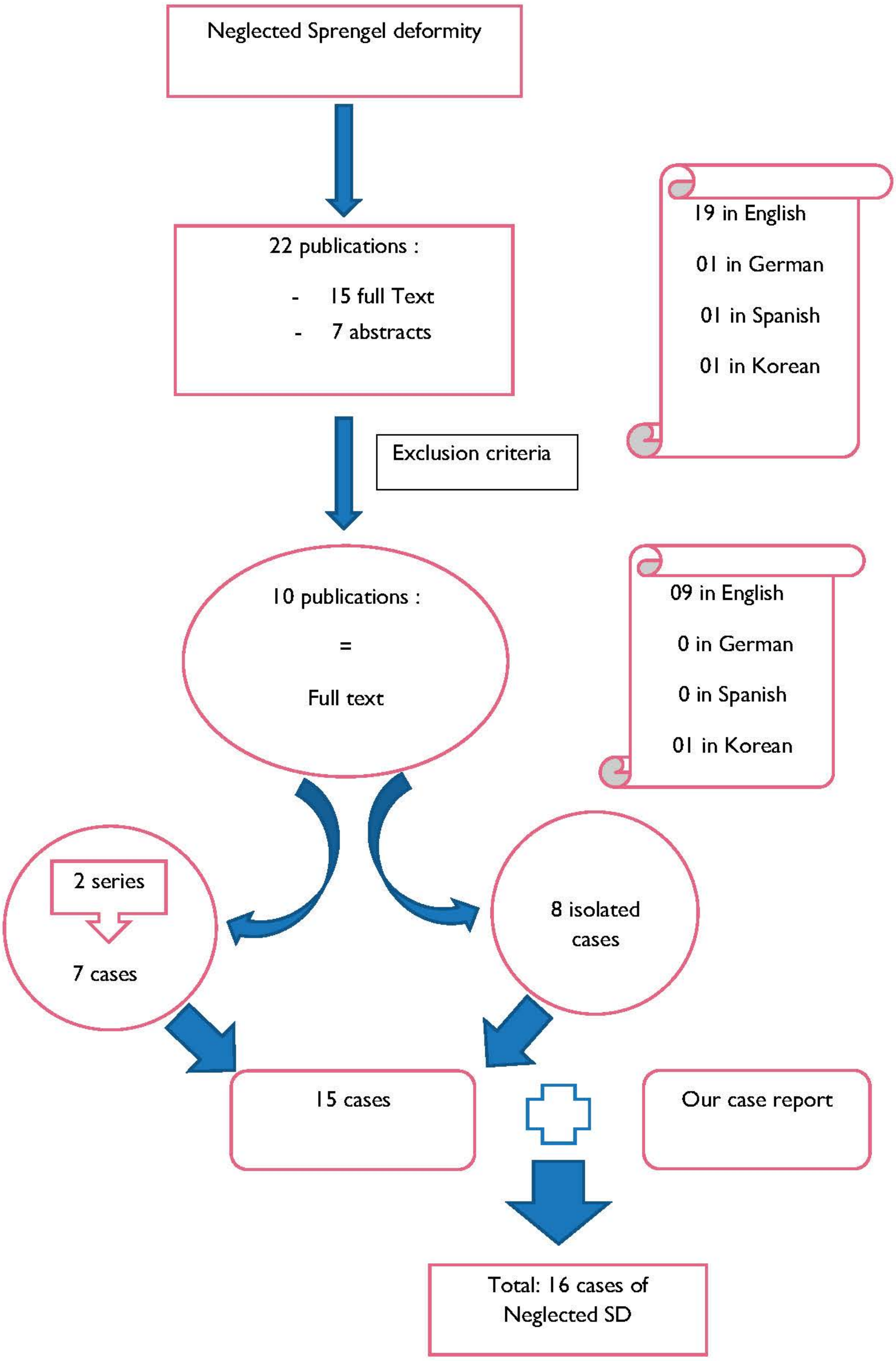


Figure 2: Review of the literature

Table I: Neglected Sprengel deformity cases from literature.

Author(s)	**Department / Article**	**Nombre of Adult cases**		**Gender**	**Age**	**Family history present**	**Side**	**Main Symptoms**		**Preoperative Clinical & Radiological features**			**Computed Tomography &/or Magnetic Resonance Imaging findings**	**Associated anomalies:** Cervicothoracic(a), Renal(b), Myelopathy(c) Syringomylia(d) Cleft lip (e)	**Surgical technique**	**Results**			
								Neck/ Peri scapular pain	Restriction of motion	Cavendish grade (A)	Abduction (B)	Rigault grade (C)				**Function** B	**Function** C	**Aesthetics/ (A)**	**Pain**
R. Dhir et al.	*Shoulder Unit, Royal National Orthopaedic Hospital, Stanmore, Middlesex,* ***UK*** */* The congenital undescended scapula syndrome: Sprengel and the cleithrum: a case series and hypothesis	**5**	1:	F	17	No		+	+	4	90	2	- Presence of cleithrum between thoracic spine and medial scapula in rhomboids region with hypoplastic body	a + b (unilateral,Vacterl)	- Bone excision	150	1	1	-
			2:	F	21	No	Left	+	+	4	100	2	- Bone growth arising from left scapula extending to neck and thoracic cage	a	- Nonoperative treatment	100	2	4	+
			3:	M	21	No		+	+	4	90	2	- Prominent bony sheet (cleithrum) arising from superomedial angle of scapula, fused to it; multilevel congenital fusion of cervical spine consistent with Klippel-Feil syndrome	a + b (unilateral renal agenesis)	- Bone excision	160	1	2	-
			4:	M	28	No	Left	+	+	4	110	2	- Accessory cleithrum linking scapula to vertebral column; anterior bone fusion at C2-3 and C3-4 levels and incomplete spina bifida	a	- Bone excision	160	1	1	-
			5:	M	32	No		+	+	4	100	3	- Omovertebral bar and deformed spinous process	a	- Bone excision	100	3	4	-
L. Füllbier et al.	*Departments of Neurosurgery and Neuroradiology, Klinikum Stuttgart, Katharinenhospital, Stuttgart,* ***Germany*** / Omovertebral bone associated with Sprengel deformity and Klippel-Feil syndrome leading to cervical myelopathy	**1**	6:	F	34	-	Left	+ -Progressive gait ataxia -Urinary incontinence	+	4 -Neurological exam: an impairment for pain and light touch in both legs, more on the right side, hyperreflexia of the left knee and ankle jerk and mild gait ataxia based on tandem and blind walking.	?	2	- atypical bone configuration of the left shoulder with elevation and dysplasia of the scapula and an aberrant bony structure extending from the superomedial border of the scapula to the C-6 transverse process leading to constriction of the spinal canal	a (KFS) + c	-Partial resection of the omovertebral bone and decompression of the spinal canal	+/- - neurologically unchanged - urinary incontinence resolved		+	_

B. Gillespie et al.	University of Colorado School of Medicine and Memorial Hospital, Colorado Springs, **Colorado** / Surgical Excision of an Omovertebral Bone in an Adult with Untreated Sprengel Deformity	**1**	7:	F	25	?	Left	+	+	4	90	2	- omovertebral bone solidly fused to the C5, C6, and C7 vertebrae at the left laminae and was adjacent to the vertebral foramen . There was an underlying posterolateral fusion at the left C5, C6, and C7 vertebral levels, consistent with Klippel-Feil syndrome.	a(KFS)	-Bone exicion and the medial supraspinous region of the scapula.	150	?	1	-
M.Doita et al.	the Department of Orthopedic Surgery, Kobe University School of Medicine, Kobe, **Japan**. / Surgical Management of Sprengel's Deformity in Adults	**2**	8:	F	20	?	Left	+	+	4	95	3	- omovertebral bone arose from the cervical spinous process(C3-C4) and terminated in a pseudarthrosis with the scapula. Spina bifida at C5. Congenital block vertebrae between CS,C6, and C7 & between T2, T3,and T4.	a (KFS, Spina bifida)	- Resection of an omovertebral bone and the medial supraspinous region of the scapula.	100	?	1	-
			9:	F	26	?	bilateral	+	+	4	100	3	- an S shaped left ornovertebral bone that extended from the C3 to the left scapula + a right omovertebral bone that extended from the C7 vertebra to the right scapula	a + d + e	- Resection of an omovertebral bone and the medial supraspinous region of the scapula.	160	?	1	-
P. TEDERKO et al.	Rehabilitation Department of the Medical University of Warsaw, **Poland**. / Omovertebral bone. Case description	**1**	10:	F	33	?	right	+	+	2		2	- deformation of vertebral arches and presence of OVB at the level of C5-C6	a + c	-Subtotal resection of OVB		?	1	-
										-Scoliosis -Increased tone of paravertebral muscles within lumbar and cervical regions									
H. O. Ong'ang'o et al.	Orthopaedics Department, Kenyatta National Hospital, Nairobi, **Kenya**. / SPRENGELS SHOULDER AT THE KENYATTA NATIONAL HOSPITAL; A CASE REPORT	**1**	11:	M	17	?	left	+	+	4	?	2	-omovertebral bone originating from the transverse process of C6 vertebrae and tethering the medial angle of the scapula		-Woodwards procedure	+/-		4	-

C. Kafadar et al.	Department of Radiology GATA Haydarpasa Training Hospital, Istanbul,**Turkey/** Omovertebral bone associated with Sprengel deformity presented with chronic cervical pain	**1**	12:	F	63	?	left	+	+	?	?	2	-elevation and rotation of the left scapula, partial fusion of the C2–C7vertebrae, and cervical scoliosis. An abnormal osseous connection between the superomedial border of the scapula and the left lamina of C6 vertebra	a	?	?			
SARKAR, A; et al	Sub-Divisional Hospital, Ghatal, **India**/ Sprengel's deformity and unilateral absent kidney: A case report	**1**	13:	M	30	No	Left	+	+	4	90°	?	-OVB	b (absent left kidney)	-Conservative management	?			
Mirhosseini, Seyyed Ahmad et al.	Department of Neurosurgery, Isfahan, **Iran** "Sprengel Deformity and Klippel-Feil Syndrome Leading to Cervical Myelopathy Presentation in Old Age."	**I**	14:	F	50	No	Left	+	+	4	?	3	-Fused vertebral body of C5-C6, an aberrant bony structure extending from the superomedial border of the scapula to the C5 transverse process leading to constriction of the spinal canal	a (scoliosis, KFS) +c	-Resection of the omovertebral bone and decompression of the spinal canal	+/-		1	-
								-Progressive gait ataxia		Short neck, low occipital hairline -Neurological examination: impairment in pain sensory and light touch in both legs, prominent on the right side, hyperreflexia in the left knee, ankle jerk,						- neurologically unchanged			
Yoon GW, Chung SH.	Department of Orthopedic Surgery, Kosin University College of Medicine, Busan, **Korea**/ Sprengel Deformity with Bilateral Huge Omovertebra.	**1**	15:	F	17	No	bilateral	+	+	4	Left: 155° Right: 170°	Left: 2 Right: 2	-Fused vertebral body of C5-T1, an OVB extending from the superomedial border of left the scapula to the C6, a larger OVB extending from the superomedial border of right the scapula to the T1	a	Woodward procedure + excision of OVBs	Left: 170° Right 180°	Left: 2 Right: 2	?	-

OUR CASE REPORT

The patient was informed that data concerning the case would be submitted for publication, and he provided consent.

I. MEDICAL HISTORY:

When the patient was 1 year old, his mother noticed a prominence on his left scapula. In addition to that, as he was growing up, he was experiencing limitation in left shoulder function noticed by his family, especially when he was reaching forward and upward.

When he was 5 years old, he was referred to the regional Hospital of Nador, for the management of his condition. Thus, the deformity was identified as an elevated left scapula. Surgical management was indicated but the patient's parents declined surgery because of financial constraints.

When the patient was 22 years old, he became concerned about the cosmetic appearance of the deformity, and he reported posterior neck pains, intensified within last 2 years (70/100 in VAS scale). Applied treatment, which included administration of oral non-steroid anti-inflammatory drugs, resting and periodical use of orthopaedic collar, did not bring satisfactory effects.

Family history was unremarkable for any congenital disorder.

II. PHYSICAL EXAMINATION

Physical examination revealed an adult in fair general condition. There was an elevated left scapula with a bony prominence extending from the left shoulder to the neck (see Fig. 3), in the projection of superior edge of right trapezius muscle, palpable on the height of C4 spinous process. This cosmetic aspect could be considered grade 4 on the Cavendish scale.

He also had a short neck, low posterior hairline and diminished cervical motion. These features represent the classic clinical triad of Klippel-Feil syndrome.

No muscular atrophy was detected. The left clavicle appeared shortened and there was fullness in the supraclavicular fossa.

There were no neurologic deficits or other physical anomalies.

However, the range of motion (ROM) of the left shoulder was restricted, with only 145 degrees in abduction (see Fig. 4).

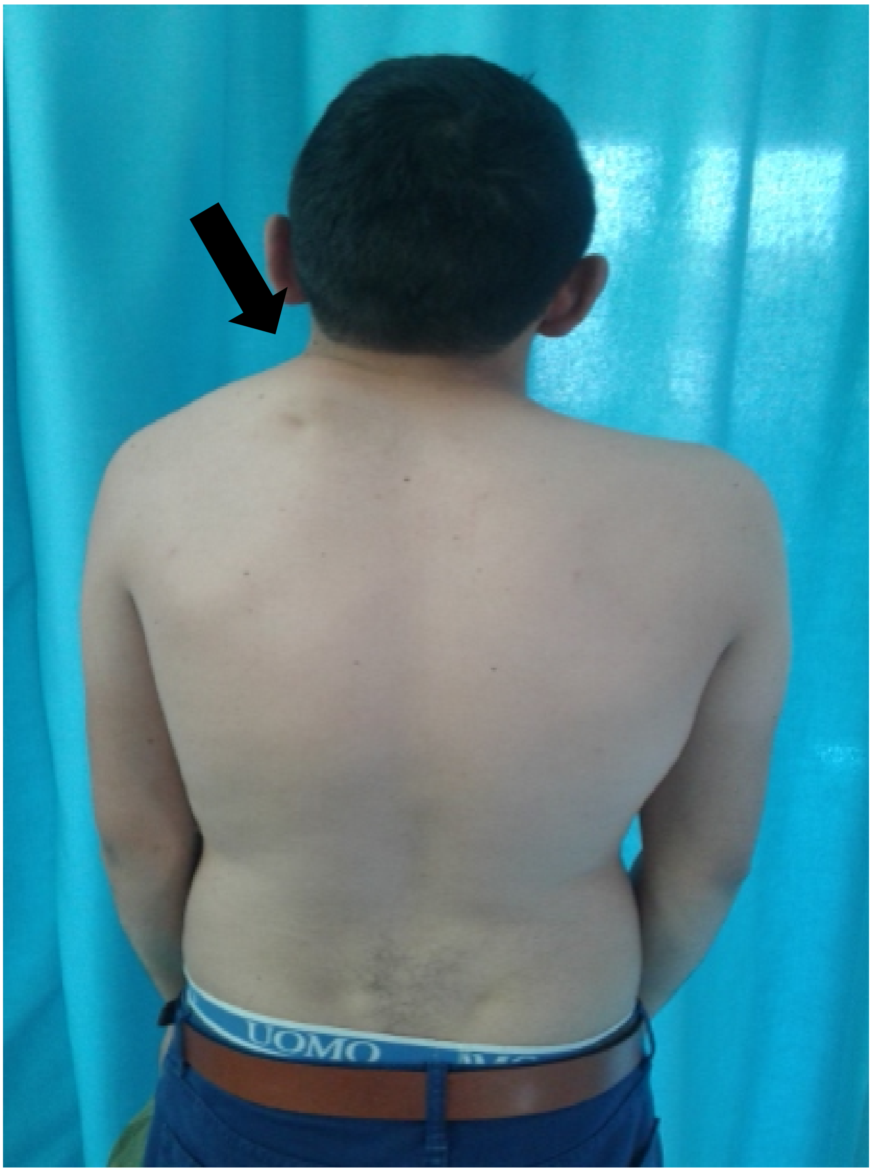


Figure 3: Sprengel's deformity of left shoulder. The left shoulder is elevated to higher level than right shoulder with short neck.

Figure 4: Preoperative clinical photograph demonstrating maximal active shoulder abduction.

III. RADIOLOGICAL EXPLORATION:

A. Plain X-ray examination:

Plain radiographs revealed (Fig. 5):

- Shoulder asymmetry; high left scapula,
- The presence of a large omovertebral bone extending from the cervical vertebras to the left scapula forming a pseudo joint with its supero-medial border, and
- Deformed left clavicular

The deformity is classified grade 3 on the Rigault scale.

Figure 5: Anteroposterior radiograph showing elevated scapula left. Also seen is the omovertebral bar between the supermedial angle of scapula and cervical spine (asterisk) forming a pseudo joint (arrow), as well as, deformed left clavicular.

B. CT scans features:

A computed tomography (CT) scan with 3D reconstruction showed (Fig. 6, 7, 8, 9, 10, 11, 12):

1. On the anterior view:

- Fusion of cervical vertebrae from C3, C4 to C7
- High left scapula attached to the cervical spine with an aberrant bony structure
- Deformed and shortened left clavicular

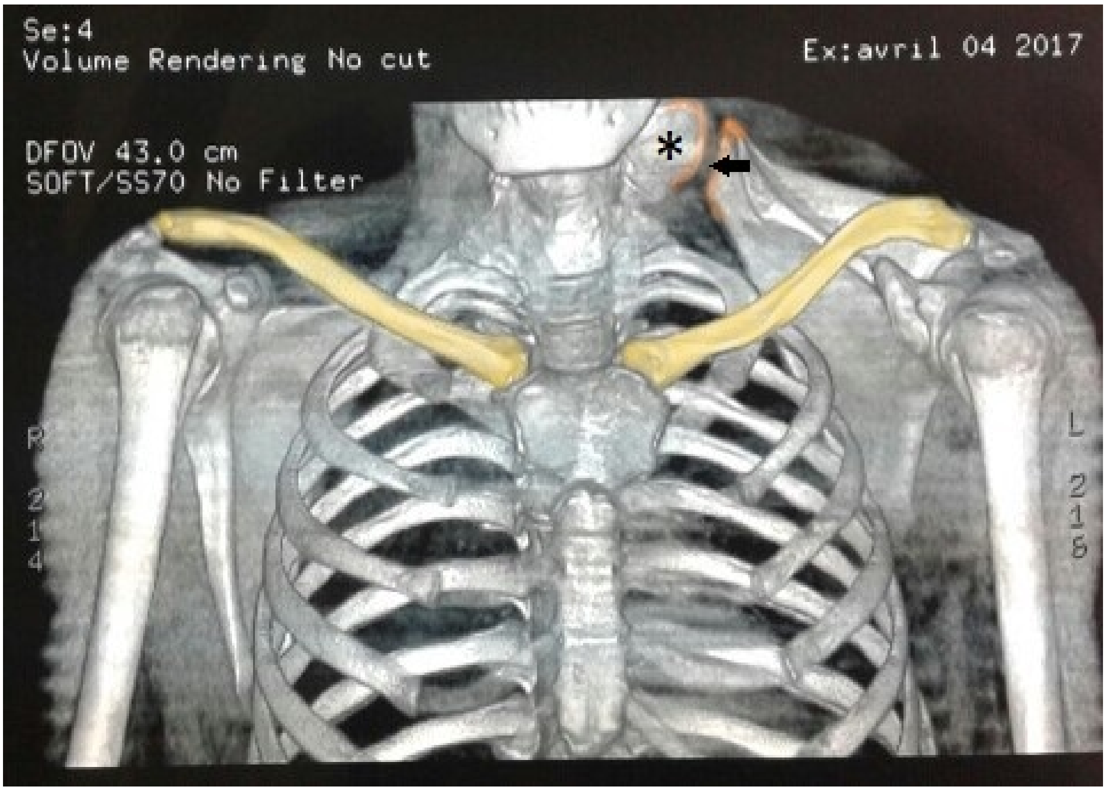
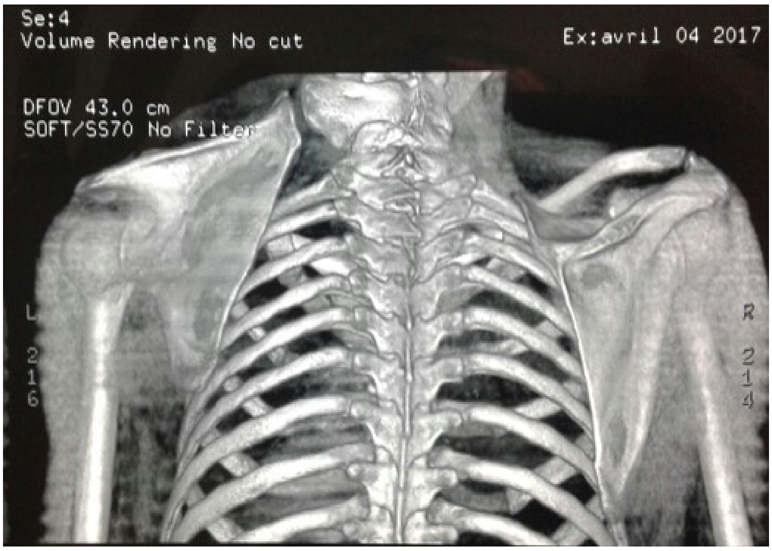
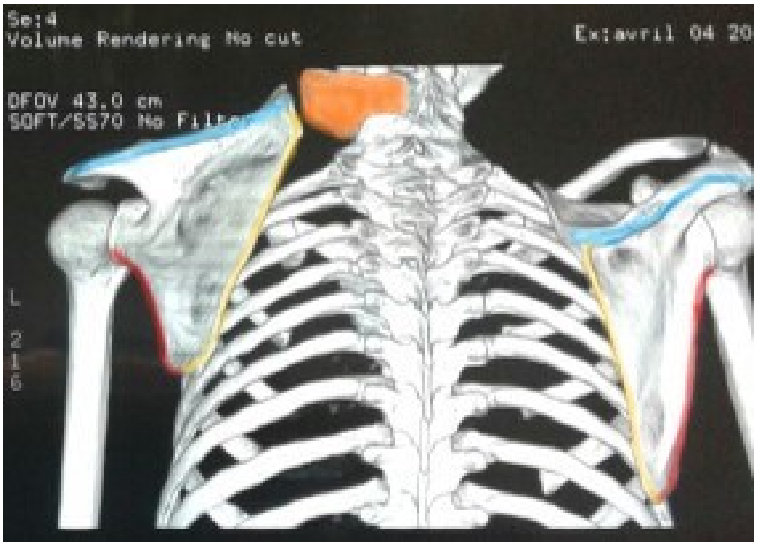


Figure 6: Reconstruction in Volume Rendrering (Anterior view) showing: High left scapula attached to the cervical spine with an omovertebral bone (asterisk). The pseudo-joint between the OVB and scapula is indicated with the arrow. The left clavicular is deformed**.**

2. On the posterior view:

- Large omovertebral bone arose from left lamina of C3-C4-C5 and terminated in a pseudo-joint with the superomedial border of the scapula.
- High, hypoplasic and rot   ated left scapula





Figures 7 & 8: Posterior view, 3-D reconstruction in volume Rendering showing: High hypoplasic left scapula attached to the cervical spine with a large aberrant bony structure (colored in orange).

3. On the axial view:

- No muscular abnormality
- Intact Glenohumeral joint
- Degree of version of glenoid cavity: 40° in the right vs 25° in the left

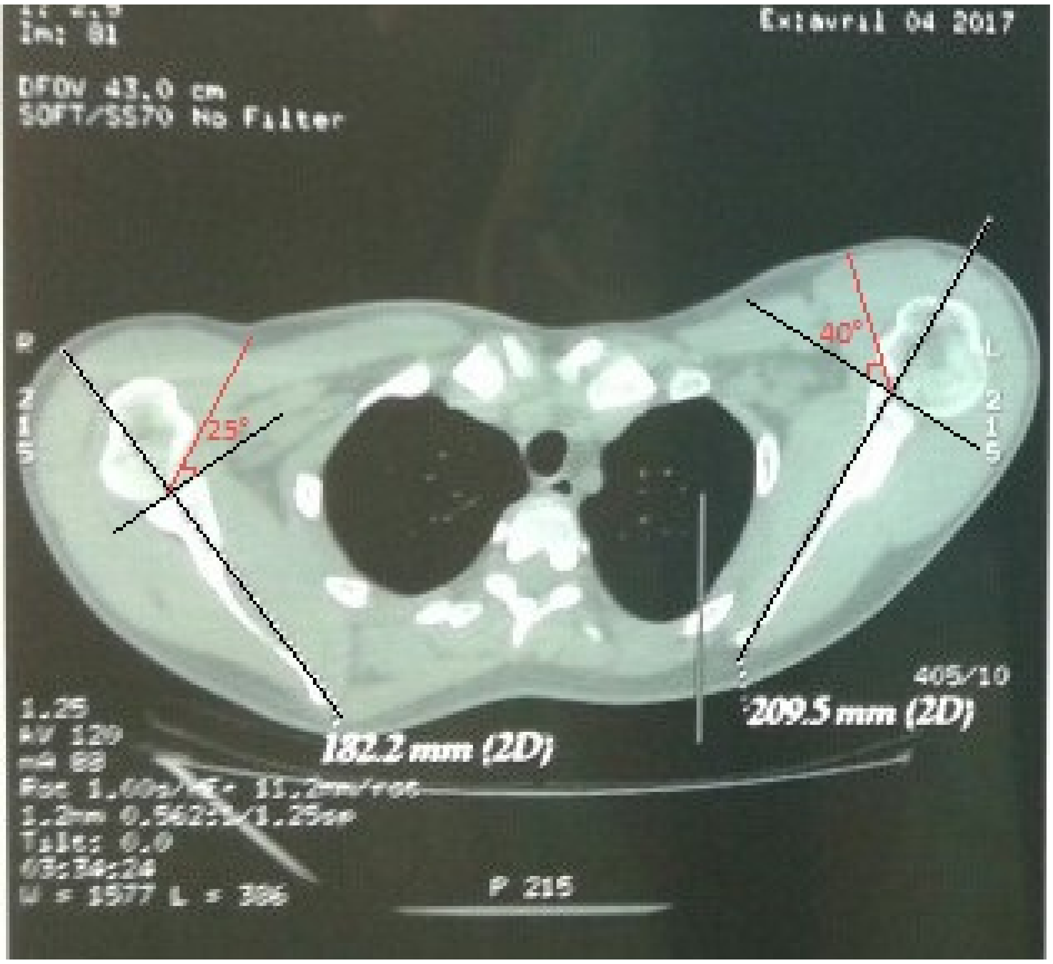


Figure 9: Axial CT showing the degree of version of glenoid cavity on both shoulders**.**

4. On the upper view:

- Omovertebral bone solidly fused to the C3-C4-C5 vertebrae at the left laminae with a large base of implantation and terminated in a pseudarthrosis with the superomedial angel of the left scapula.
- Deformed left clavicular; it is shortened, curved and has an excavation in the centre, with an opposite direction of orientation.
- Narrowed space between the left clavicular and the spine of the left scapula: 14mm vs 78mm (see Fig. 11 & 12).

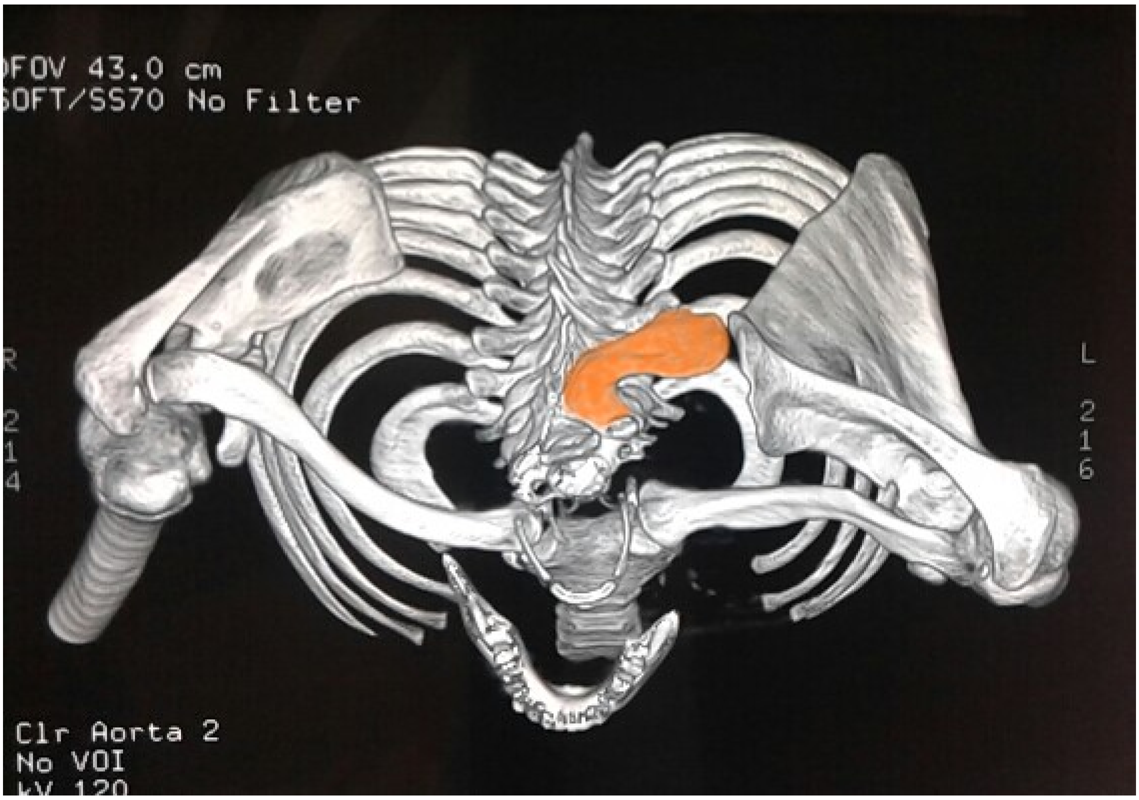


Figure 10: Trunk 3D CT scan; Upper view: Omovertebral bone is colored in orange**.**

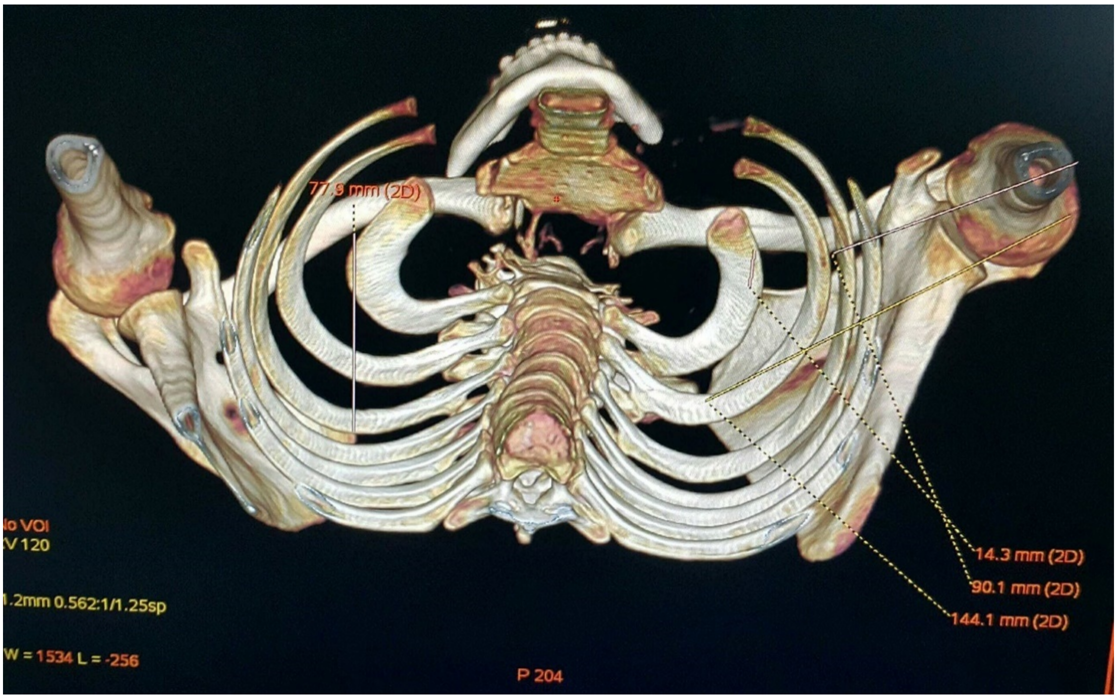


Figure 11 + Figure 12: Trunk 3D CT scan; Upper view showing narrow space between the left clavicular and the spine of the left scapula: 14mm vs 78mm.

IV. SURGICAL MANAGEMENT:

The decision was made to proceed with surgery to remove the omovertebral bone, to get the scapula free of any attachment as well as inferiorly shifting its location if possible, with the goal of decreased neck and shoulder pain while improving shoulder motion.

- Under general anesthesia
- Position: The patient was in semi-prone position with the affected limb draped freely to allow movement of the arm and manipulation of the scapula
- Incision: A straight midline incision was performed over the spinous processes, from C2 to D8.
- Technique: simple excision of the OVB & the superomedial border of the scapula

The omovertebral bone was exposed completely and was observed to extend from the C3, C4 and C5 vertebra to a pseudo-joint at the superomedial border of the scapula (see Fig 13). The trapezius was dissected from its spinous process origin in continuity cranially with the rhomboids. The superomedial portion of the scapula as well as the omovertebral bone were dissected extraperiosteally then excised.

Hemostasis was assured and a closed suction drain was placed before layered closure.

The arm was immobilized using a Velpeau sling.

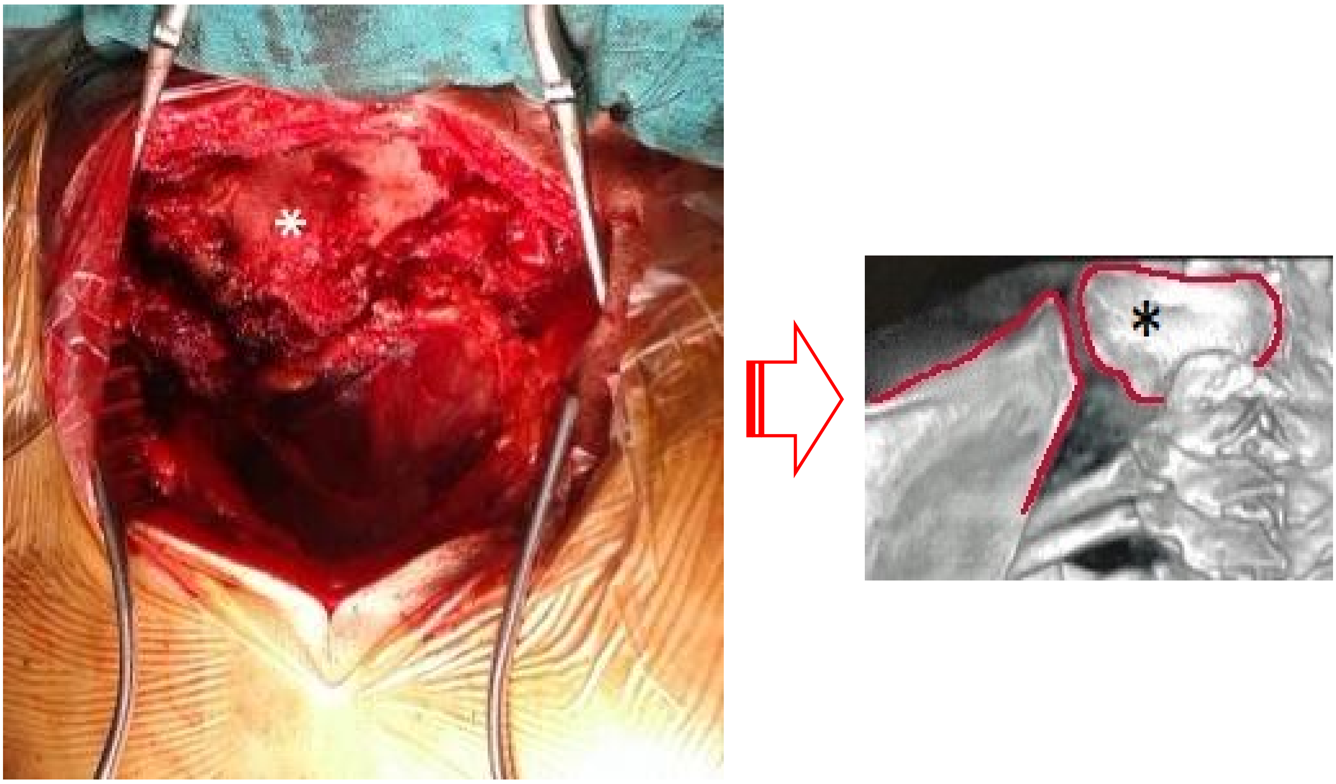


Figure 13: intraoperative view with corresponding radiologic image.
The omovertebral bone (asterisk) is visible between the medial border of the scapula (arrow) and the cervical spinous processes.

V.FOLLOW UP:

Postoperative course was uncomplicated; the patient was discharged from hospital 24h after surgery (after removing the Redon drain).

The patient received Velpeau sling protection for tree week, followed by progressive rehabilitation of the operated shoulder.

Three months later, the ROM in the left shoulder showed significant improvement in abduction; from 135 degrees to 170 degrees (Fig. 14), and the dull pain that had been present at the nape of the neck was significantly reduced.

At a medium-term follow-up period of nine months, improved abduction of 170° was maintained and global motion of the shoulder has progressed, neck pain has disappeared, and formation of keloid scar was observed (Fig. 15).

There was no change in his neurological state.

The patient and his family were satisfied with the final result, especially the correction of the cosmetic deformity.

Figure 14: Surgical scar nine months postoperatively**.**

Figure 15: Abduction improved from 145°to 170°and was maintained at nine months postoperatively.

RESULTS

In the light of our case and the review of the literature, the results were as following:

I. EPIDEMIOLOGY:

A. Gender:

This series included 10 females and 6 males, with a sex ratio F/M: 1,6 .

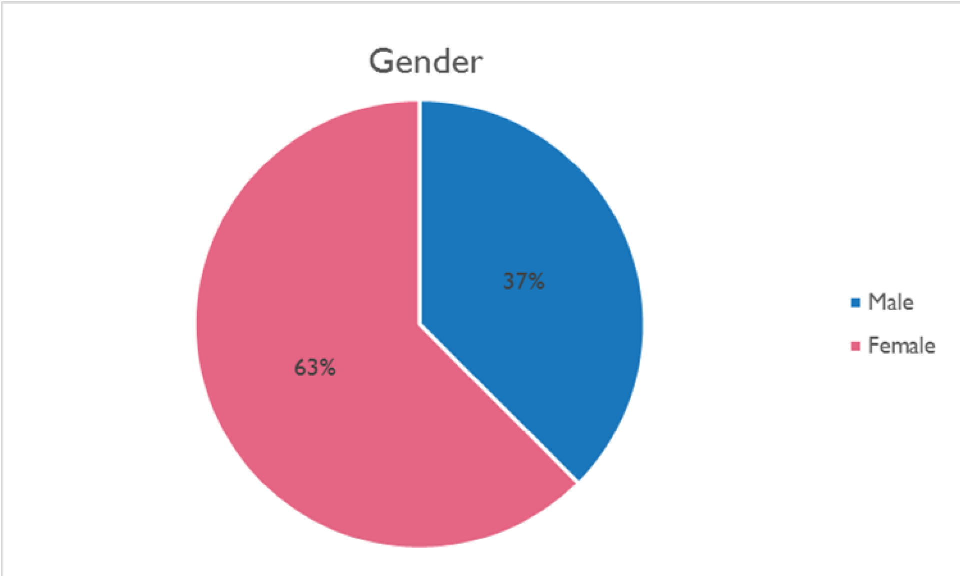


Figure 16: Distribution of Sprengel deformity by sex.

B. Age:

The patients included in our study were divided by age groups to assess the mean age of the management of the deformity in its neglected form.

We noted that the maximum of cases in our study were managed in adulthood, between 20 and 40 years old, with 10 patients of 16, so a percentage of 62,5.

Four patients were managed in adolescence, so a percentage of 25.

The mean age at management was 26,8 years (range, 17-63 years).

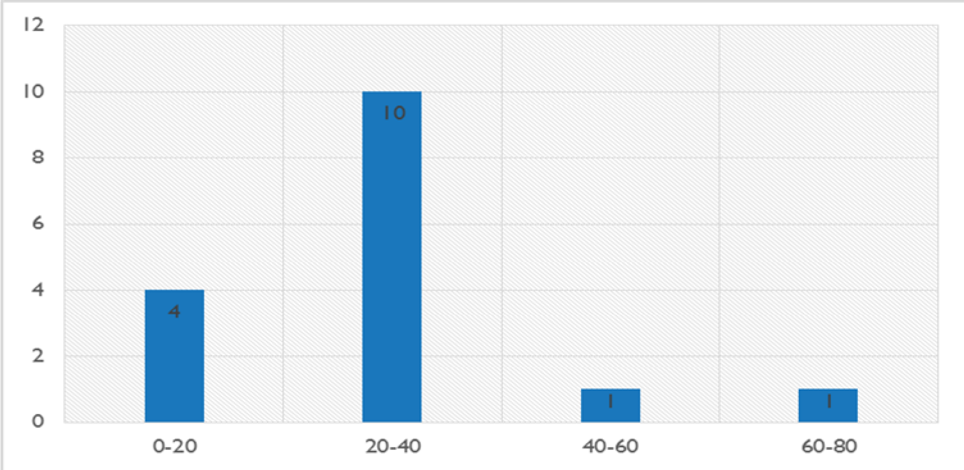


Figure 17: Number of managed cases of neglected Sprengel deformity according to age groups.

C. Affected side:

Left shoulder was affected in 13 patients, while the right side was affected in only 1 patient, and two cases had SD in both shoulders.

Thus, the left shoulder was more affected than the right side, with 81% versus 13%.

Bilaterality was present in our series with 6% (2 patients).

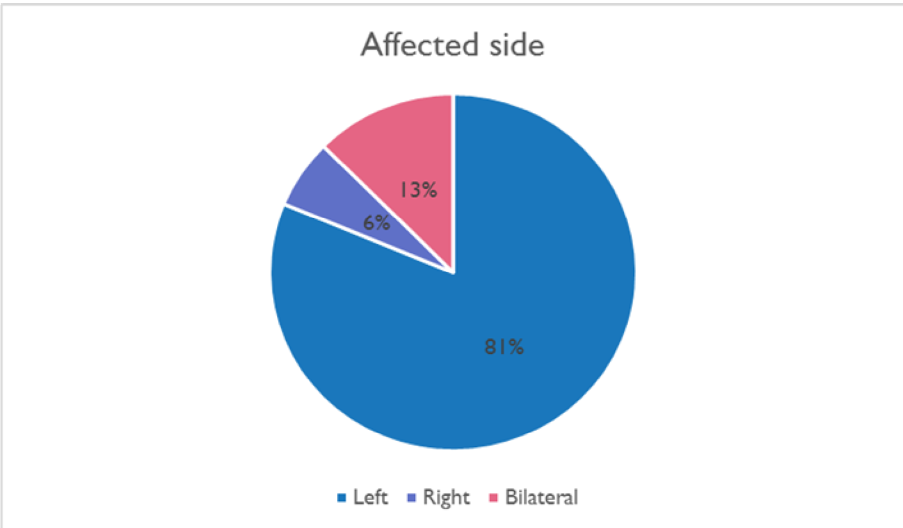


Figure 18: Distribution of neglected Sprengel deformity according to the affected side.

II. MAIN SYMPTOMS:

Neck &/or periscapular pain was present in all cases, as well as restriction of neck and shoulder motion. Neurological signs (secondary to constriction of the spinal canal) were present in two of total cases.

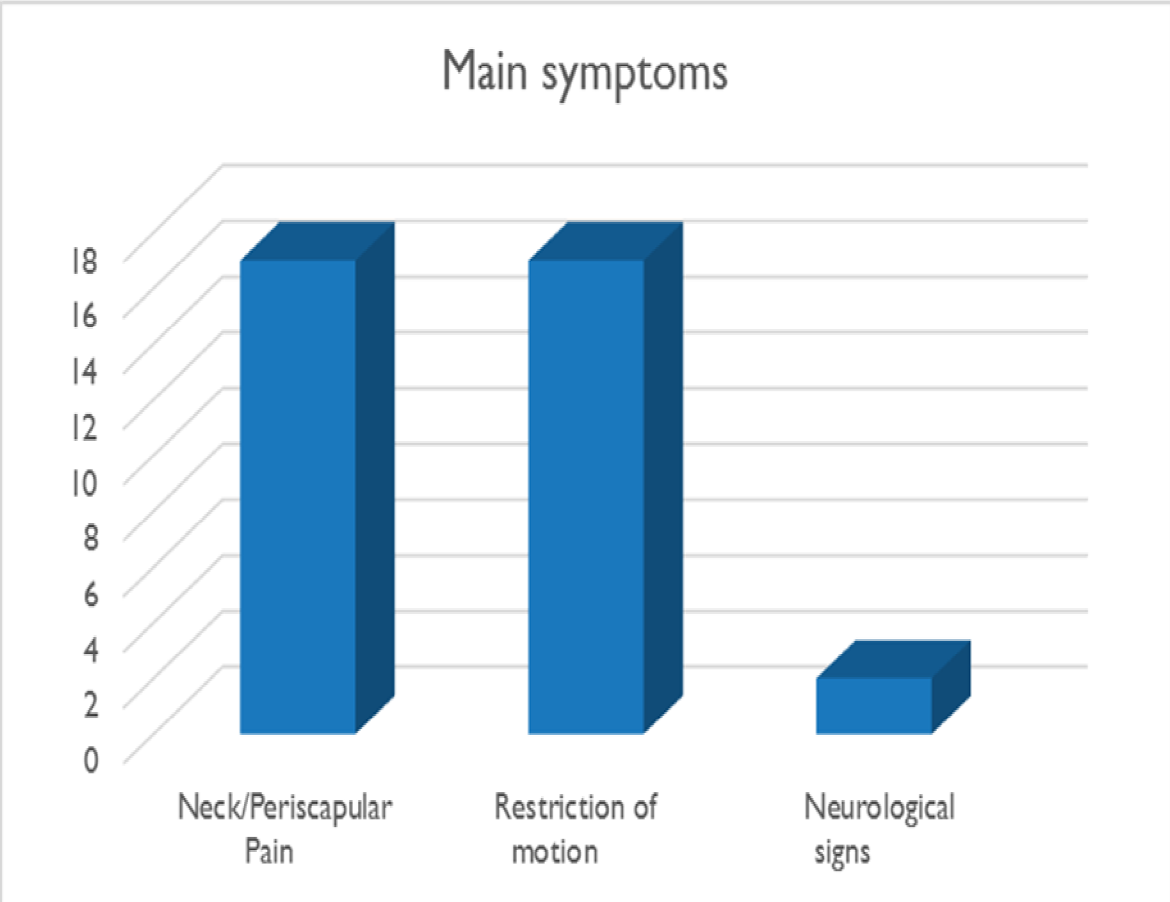


Figure 19: Main symptoms reported in the series.

III. PREOPERATIVE CLINICAL & RADIOLOGICAL FEATURES

A. Clinical features:

1.The cosmetic appearance:

It was classified according to the Cavendish system (a method of grading of appearance suggested to assist in selecting cases for operation), 14 of 15 patients were assigned to grade 4 on the Cavendish scale and 1 was grade 2 (one Author did not mention the exact grade of his patient, but cosmetic impairment has been reported).

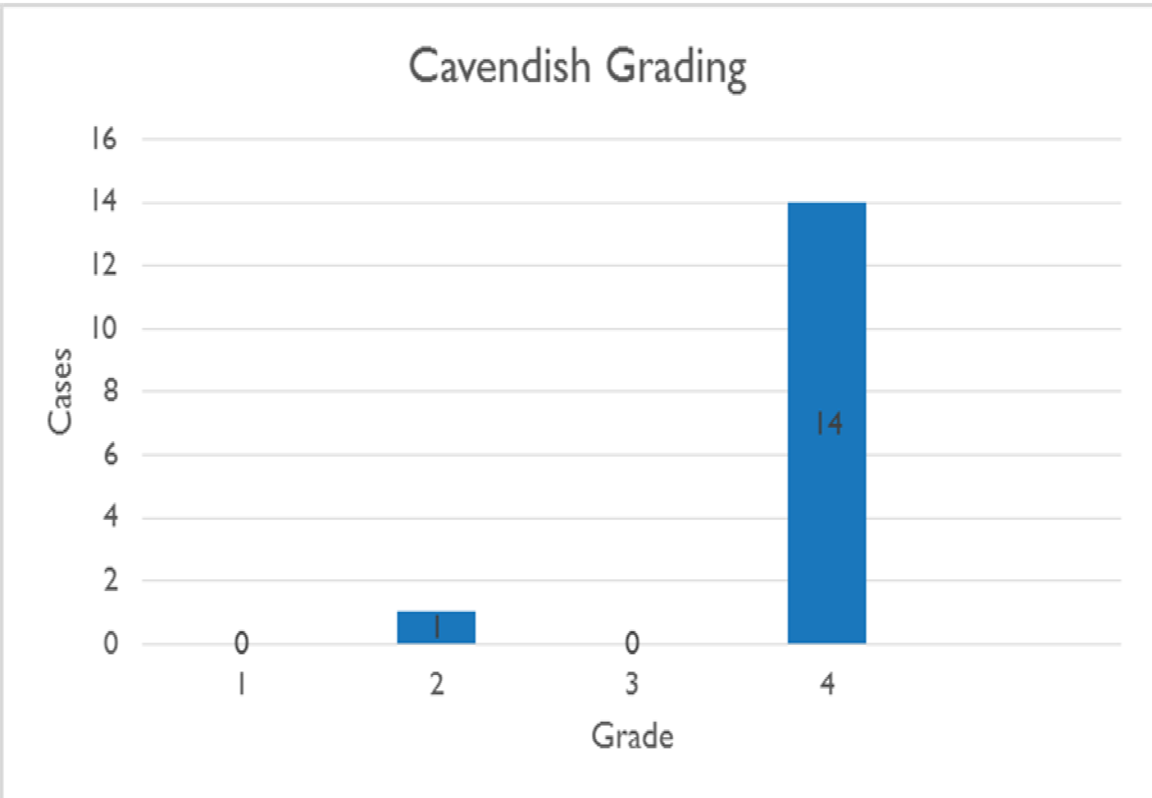


Figure 20: Preoperative Cavendish grading in our series.

2.The functional impairment:

Decreased shoulder function was reduced here in "Shoulder Abduction"; it was limited in most of the cases to ≤100° (in 70% of all cases).

It has been taken into consideration only cases whose shoulders abduction degrees were specified in the articles (13 shoulders).

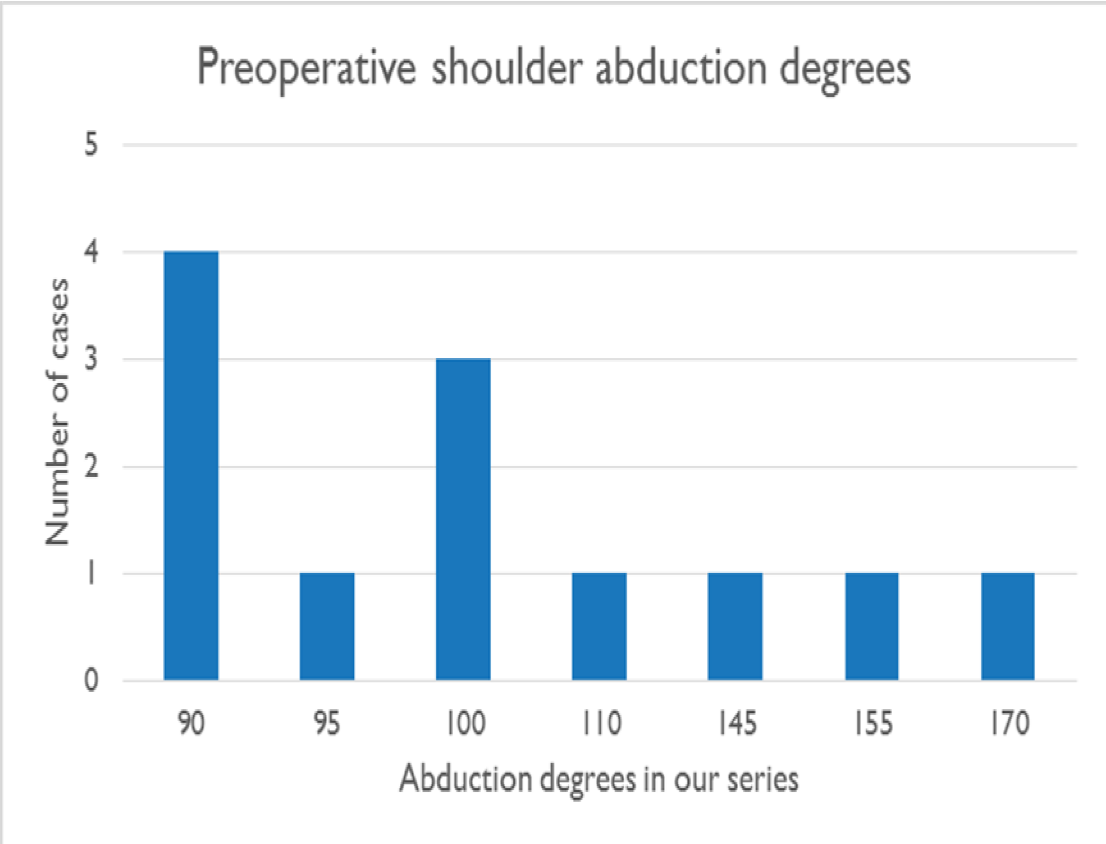


Figure 21: Preoperative shoulder abduction degrees in our series with the corresponding cases.

B. Plain Radiological features:

Radiologic features were classified according to Rigault systems; 10 shoulders of 18 were grade 2, and 6 were grade 3 on the Rigault scale.

(Two Authors did not mention the exact grade of their patients).

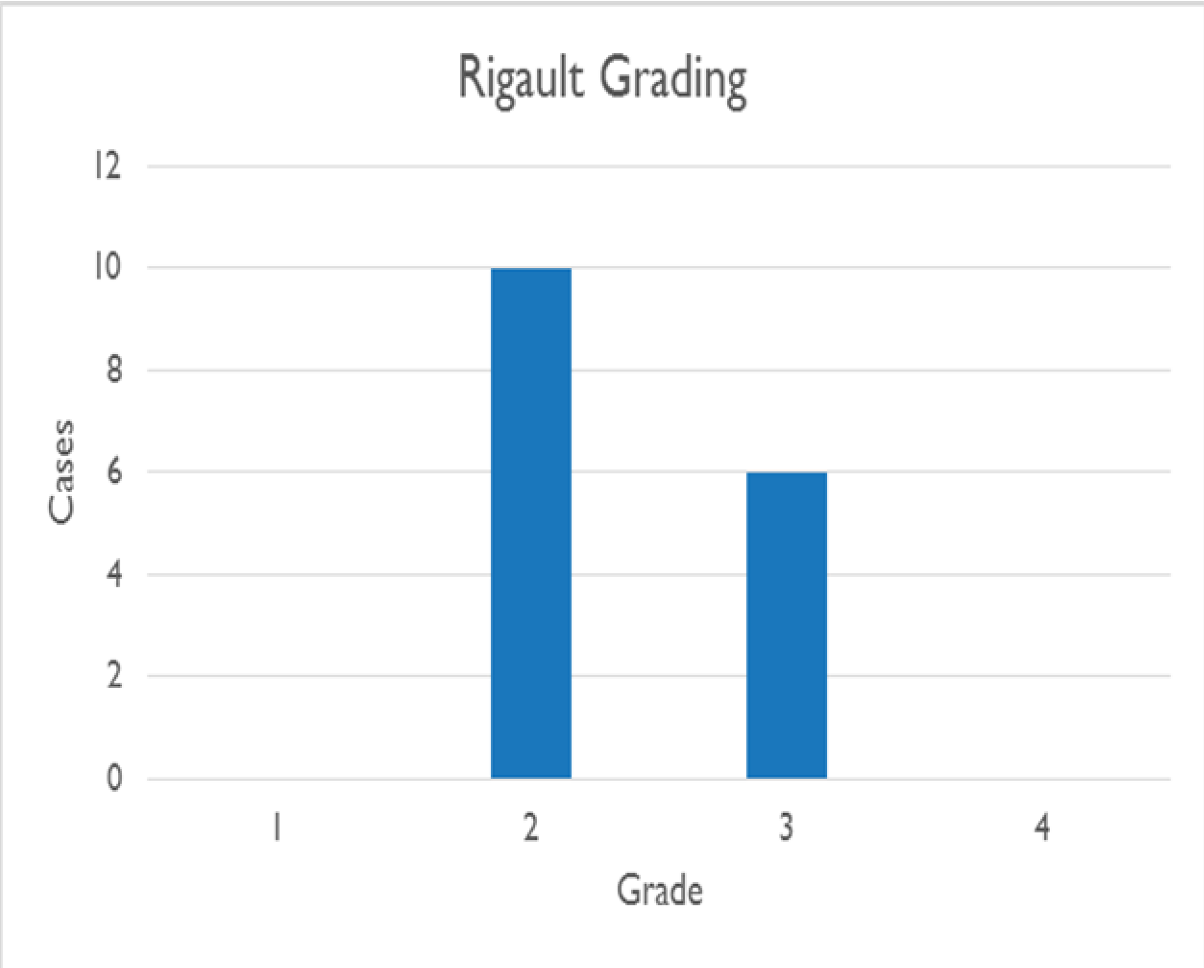


Figure 22: Preoperative Rigault grading in our series.

IV. ASSOCIATED ABNORMALITIES:

Omovertebral bone was found in all cases.

All 16 cases had associated cervicothoracic spinal anomalies; with Scoliosis accounting for 68 per cent, Kippel-Feil syndrome was found in 25% of the patients, spinal dysraphism with Spina bifida in 2 patients and Syringomyelia in 1 patient. Muscular abnormalities were found in 37% of the cases. There were two cases of cord compression.

Renal abnormalities reported in 4 patients (agenesis). Other abnormalities were found in about 6 per cent of the patients (cleft lip and palate).

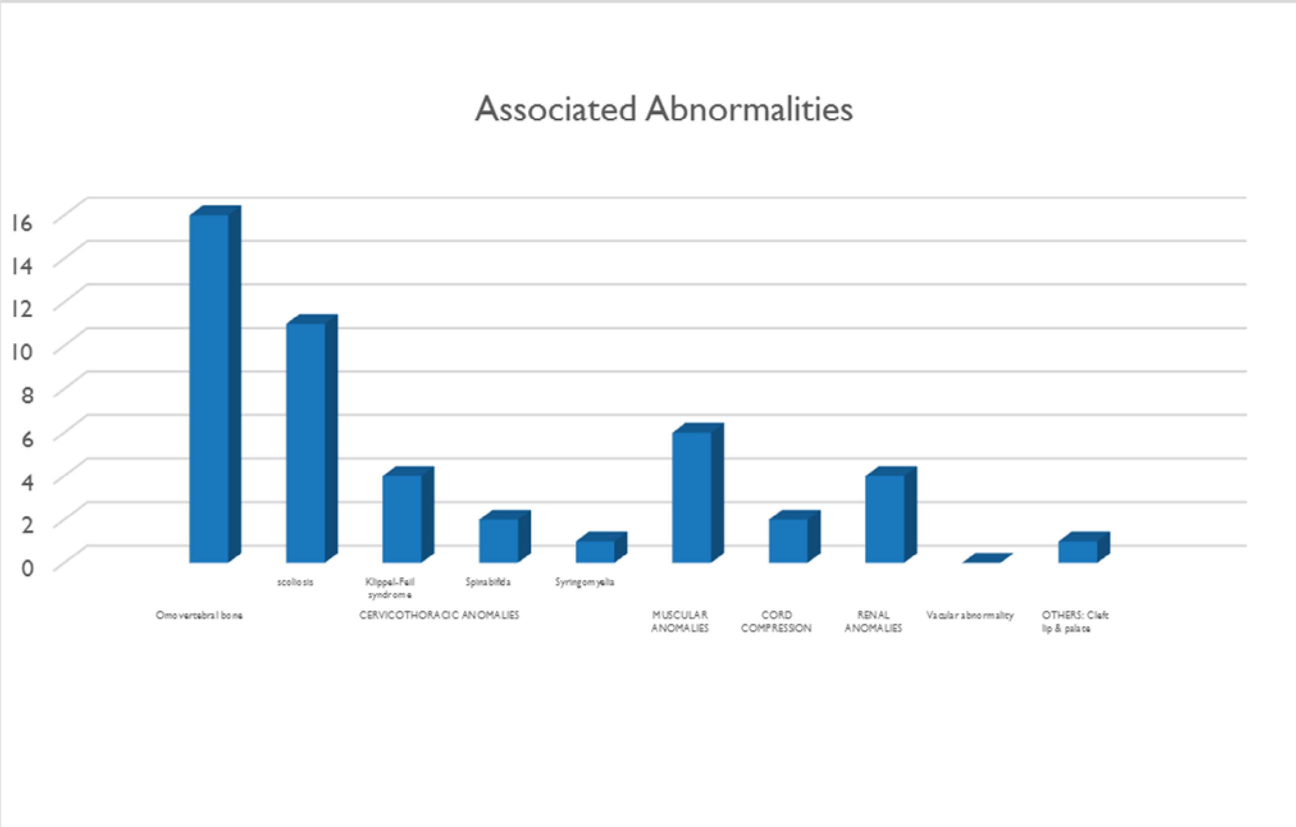


Figure 23: Associated abnormalities reported in our series.

V. MANAGEMENT (SURGICAL PROCEDURES AND NON-OPERATIVE TREATMENT):

11 shoulders were treated by simple bone excision (including our case report), four underwent Woodward procedure. Mears technique was performed on one shoulder.

Green's procedure wasn't used on any of the patients.

Two patients had nonoperative treatment.

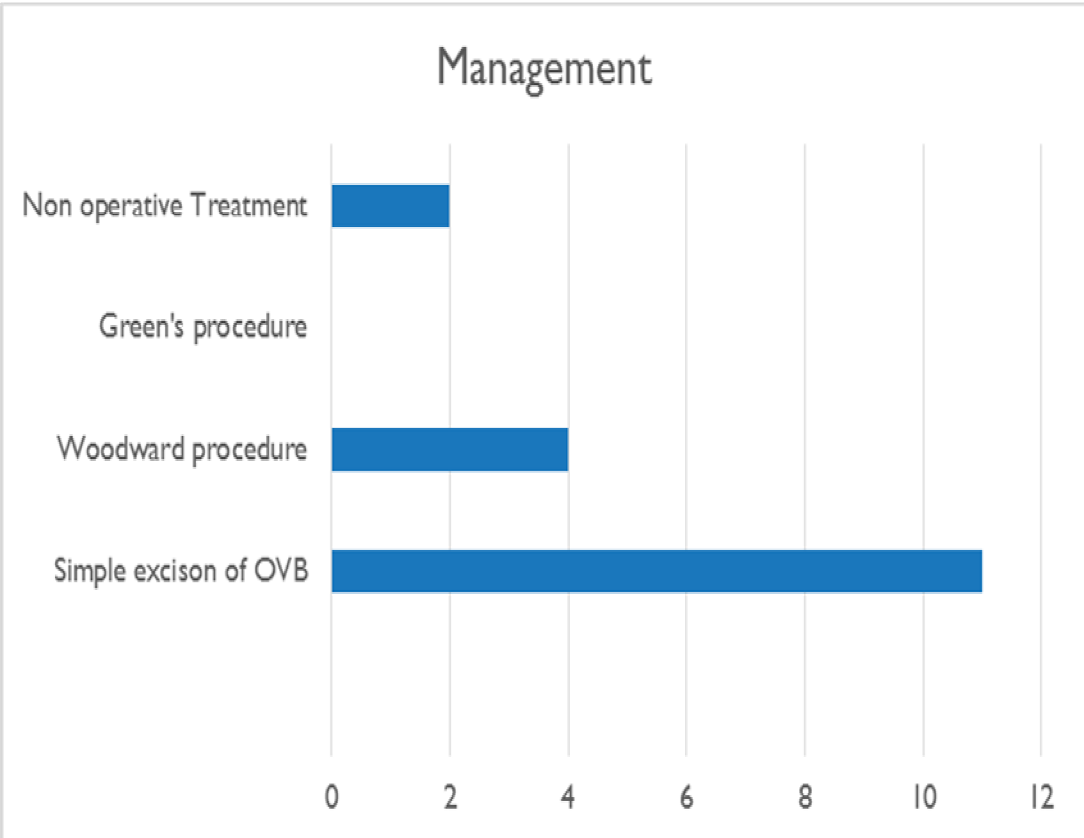


Figure 24: Operative and non-operative management of the cases in our series.

VI. POST-OPERATIVE RESULTS:

A. Clinical features :

1. The cosmetic appearance:

The median Cavendish score improved from 4 to 1.

The Cavendish score has improved from grade 4 preoperatively to grade 1 postoperatively in 7 patients and one patient improved from grade 2 to grade 1.

Two of the postoperative patients still had a score of 4, as well as the two other patients who had nonoperative treatment.

In tree cases, the postoperative grade wasn't determinate (the preoperative grade was 4 and improvement has been reported).

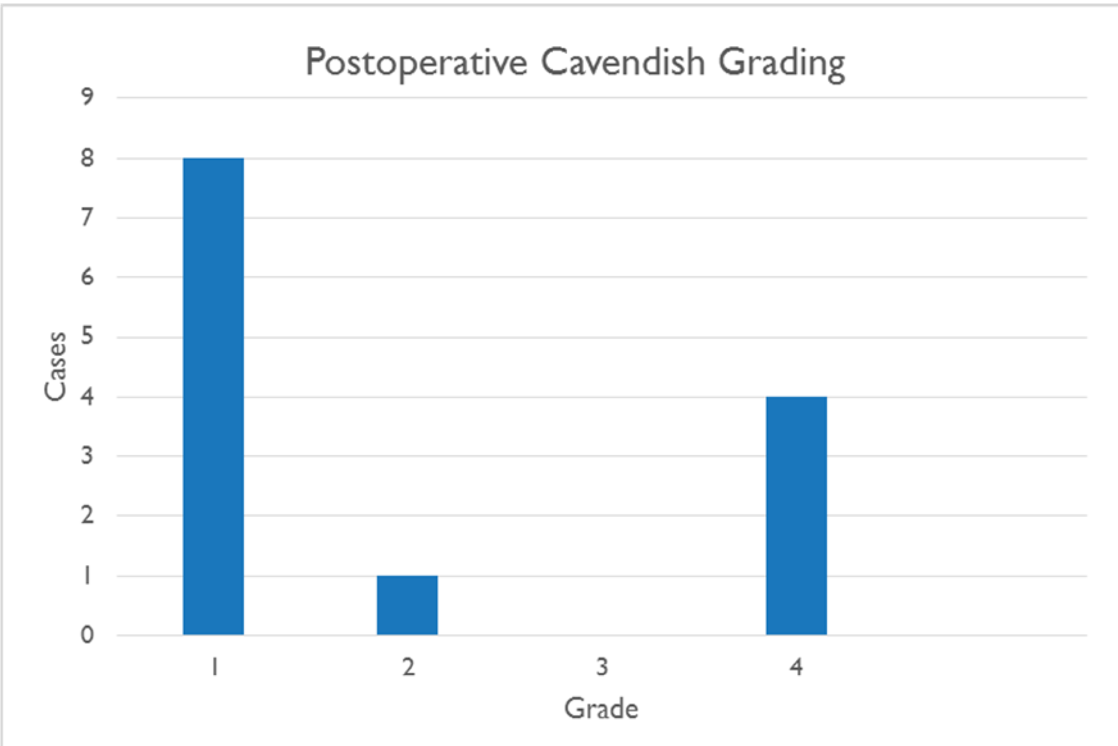


Figure 25: Postoperative Cavendish grading in our series.

2. The functional impairment:

Mean abduction improved significantly from 108° (90°-170°) to 152° (100°-180°), an improvement of 44° (±standard error). It has been taken into consideration only cases whose degrees were specified.

9 of 13 shoulders had an abduction degree ≤100° in preoperative, after surgery, they improved to higher or equal to 150°. In two cases, the preoperative degree was >150°, they only showed an improvement in average 15°.

In two cases, there was almost no improvement.

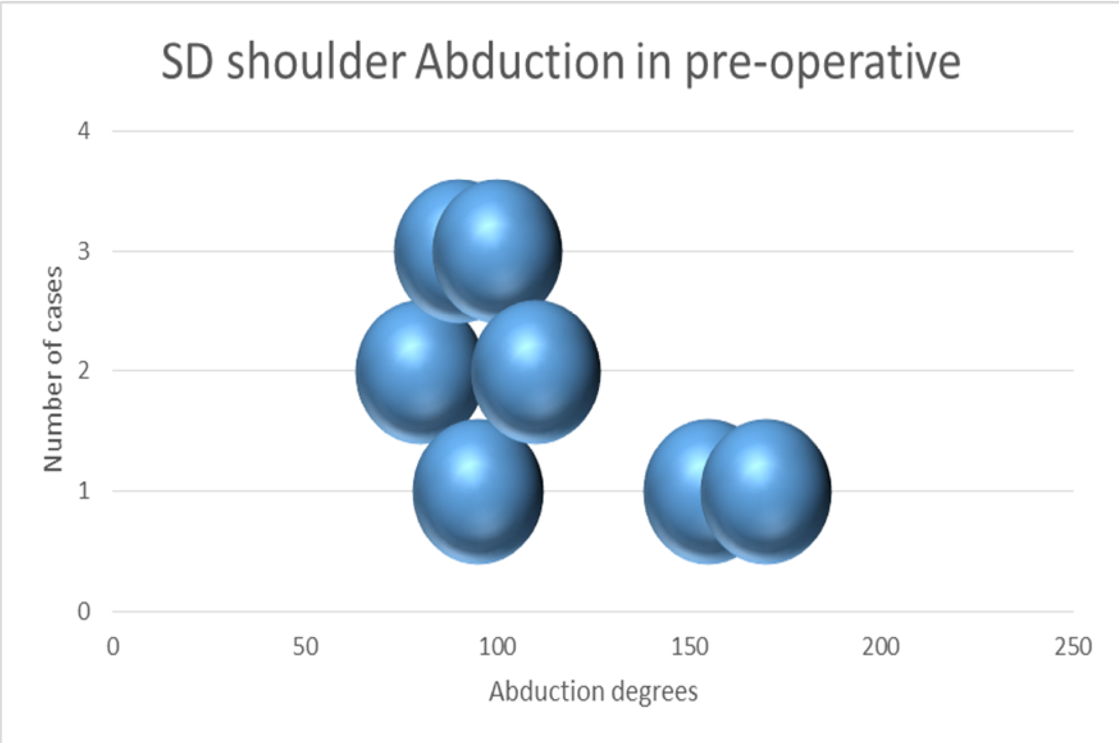


Figure 26: Shoulder Abduction degrees in pre-operative.

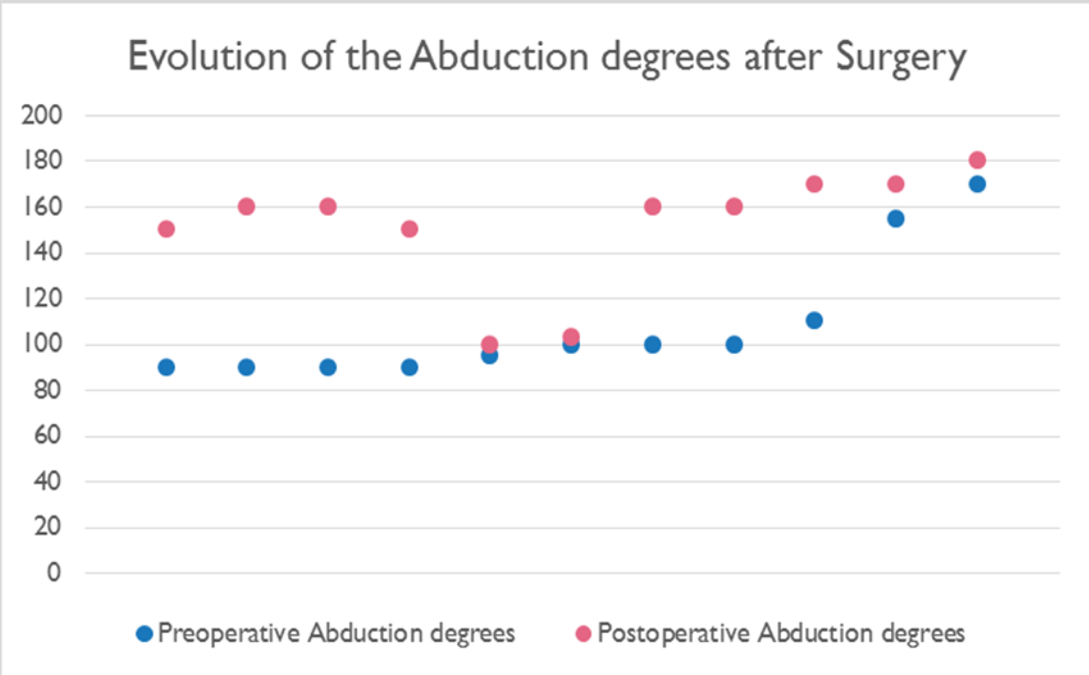


Figure 27: Evolution of the Abduction degrees after Surgery.

3. Pain and neurological signs:

All patients who underwent surgical management of their SD reported less neck and/or periscapular pain.

The two patients who had myelopathy, remained neurologically unchanged.

B. Plain Radiological features:

Five patients improved from grade 3 to grade 2, and two patients improved from grade 3 to grade 1, and one patient stayed grade 3 after surgery.

Tree patients improved from grade 2 to grade 1.

Four patients still had grade 2 after surgery and the two patients with nonoperative treatment didn't improve.

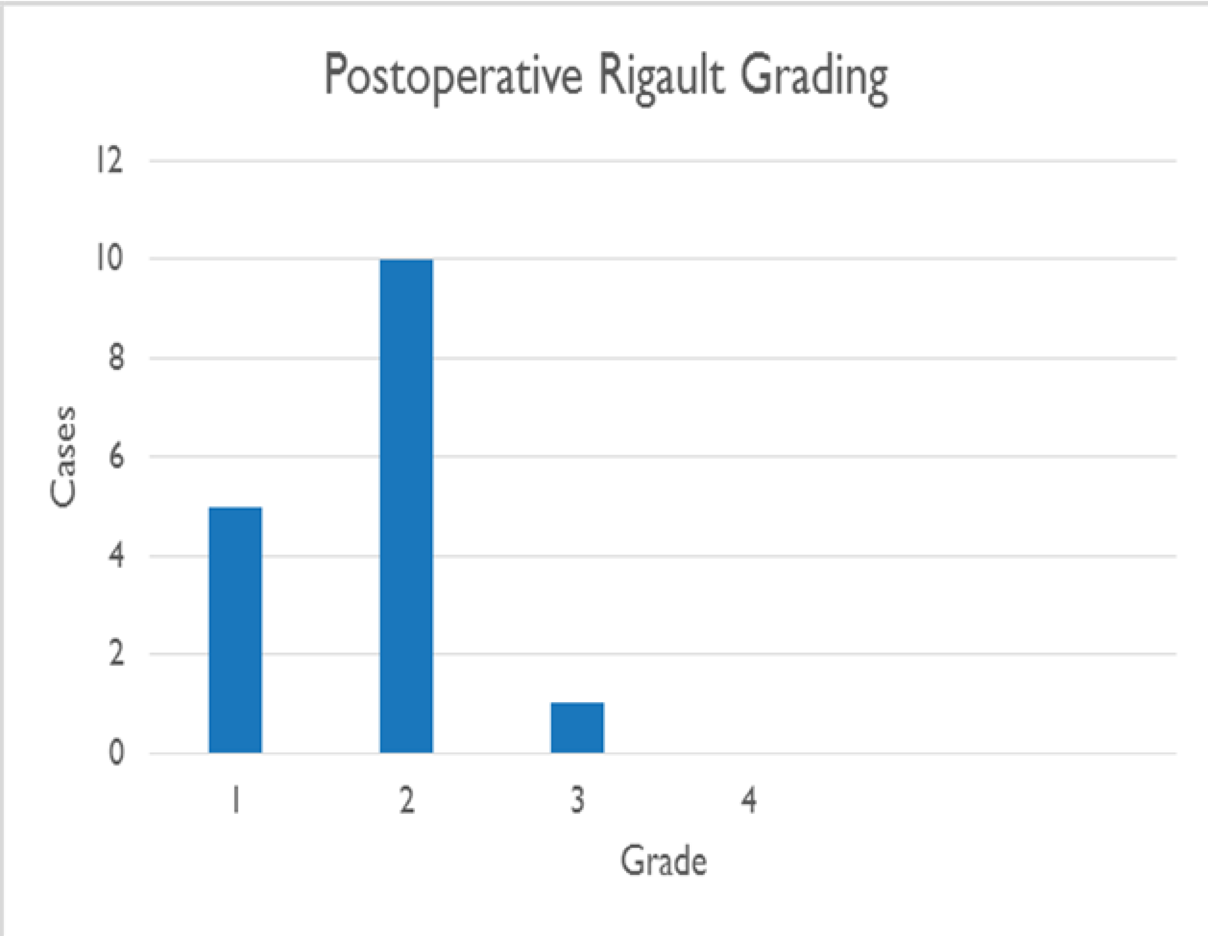


Figure 29: Postoperative Rigault Grading.

DISCUSSION & REVIEW OF THE LITERATURE

A. EPIDEMIOLOGY

1. Prevalence:

The prevalence of neglected or adult Sprengel deformity was not well described in the literature, currently no accurate information on the incidence and prevalence of this malformation in either adults or children. Although the condition is rare, it is the most common congenital shoulder girdle deformity. [4] [31] [32]

2. Age and Gender:

The deformation is usually observable at birth and it intensifies with a child growth [33].

Our patient has been treated at the age of 22 years old, the mean age at operation in our series was 26,8 years (range, 17-63 years). In fact, adolescent and adult cases represent the neglect aspect of the deformity.

In our study, 64,7% of all cases, were managed in adulthood, between 20 and 40 years old, this could be explained by the unaesthetic disfigurement (scapular asymmetry, posterior thoracic swelling corresponding to the projection of the tip of the scapula) and its psychological impact, the progression of limited active abduction of the shoulder as well as the emergence of pain at the level of the neck or sometimes in the periscapular region. [34][35]

In general, there is predominance for the female gender with a female to male ratio of 3:1.

In other series, there is male predominance[20] [21] [31][33].

3. Localisation:

The congenital elevation of the scapula is usually unilateral (in 88% of cases); with or without left or right predominance depending on the author. Bilateral involvement has been described, in which case it is functionally more disabling, but much more cosmetically acceptable. The frequency of the bilateral forms found is generally from 5 to 18%.[3][36]

In our series, left side was more affected than the right side. (it was involved in 81%) and two patient had bilateral form (6%).

4. Genetics:

Usually arising sporadically, a few familial forms of Sprengel deformity have been described and suggest the possibility of genetic transmission. [37]

There was no family history of SD, neither in the literature group or in our report.

B.CLINICAL FEATURES:

1. **The primary ‘’deformity’’ is cosmetic: asymmetric magnification of neck contour** and shoulders asymmetry.

The varied clinical presentation of the patient with Sprengel deformity is a composite of the deformities produces by the undescended scapula and the associated cervicothoracic abnormalities that are almost always present. For this reason, as Cavendish has pointed out, the same scapular deformity, even when bilateral, ‘’may vary widely in ugliness.’’

In its failure to descend, the scapula remains higher than normal, on the posterior chest wall, by around two to ten centimetres. It is adducted and is smaller than normal in the vertical plane and appears larger than normal horizontally. The inferior angle is medially rotated and abuts the thoracic spinous processes, causing the glenoid to face inferiorly. The higher the scapula lies, the less it is rotated. A visible lump in the suprascapular region is characteristic due to the upwardly rotated superomedial angle of the scapula, which causes the ipsilateral side of the neck to appear fuller. The prominent lump in the web of the apparently short neck is noted in infancy or early childhood by the parents (figure 30).

The superior part of the scapula may be prominent and curved forwards over the apex of the thorax leading to deformity of the clavicle. [21] [31]

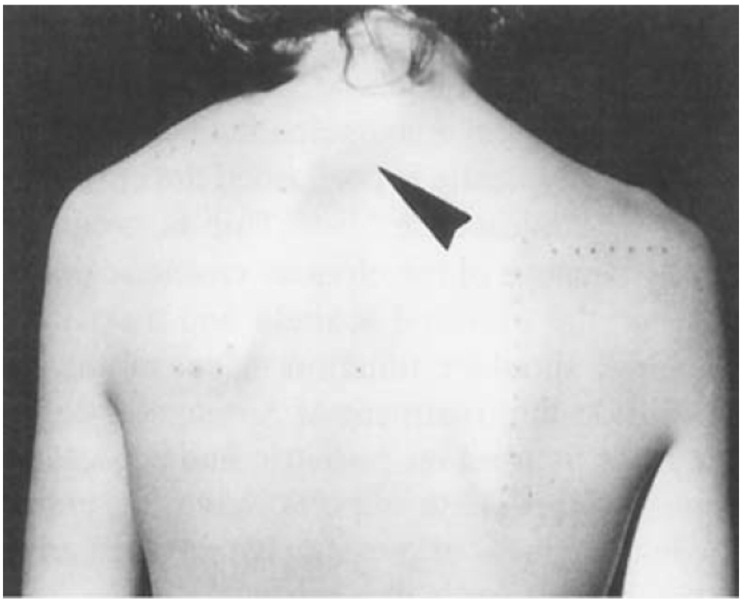


Figure 30: Photograph showing elevated left scapula, and superomedial prominence because of an omovertebral bone. The black arrow shows the bony prominence extending from the shoulder to the neck. “Doita M, Iio H, Mizuno K. Surgical management of Sprengel’s deformity in adults. A report of two cases.”

An omovertebral connection exists in about one third of cases. This is a rhomboid or trapezoid shaped structure that usually lies in a strong fascial sheath, which extends from the superomedial angle of the scapula to the spinous process, lamina or transverse process of the cervical vertebrae (most commonly the fourth to seventh cervical vertebrae). It is palpable as a chondro-osseous bar, rather than as a fibrous cord, in 20 to 30 per cent of all patients. [33] [38]

Crha [39] underlines that in the diagnostics of omovertebral formations main role is plaid by digital palpation and not by radiography because calcification of these structures occurs only in one patient for four.

The omovertebral connection is usually unilateral. Baulot [40] described a case of SD with double unilaterally situated OVBs; the first one within elevator muscle of scapula and the second one in rhomboid muscle. It is the primary cause of restricted shoulder motion in patients with Sprengel's deformity. It is associated with a fixed, elevated scapula and has a major role in determining the shape and the malpositioning of the scapula. Williams cited one documented a case of OVB occurrence without characteristic features of SD [38].

The prominent may be masked by contralateral thoracic scoliosis or exaggerated by an ipsilateral curve. Brevicollis that accompanies the often associated Klippel-Feil syndrome also exaggerates the deformity. Clearly, one might best think of the condition as a deformity of the cervothoracic spine and shoulder girdle.

The spinoscapular muscles are also adversely affected. The trapezius, rhomboid, serratus anterior, levator scapulae, pectoralis major, latissimus dorsi or the sternocleidomastoid muscle may be absent, hypoplastic or contain multiple fibrous adhesions. The trapezius muscle is the most commonly affected muscle. If the serratus anterior muscle is weak, winging of the scapula may occur [41] [42].

The cosmetic aspect of the deformity has been classified by Cavendish into four grades in an attempt to simplify indications for treatment.

The Cavendish classification (Cavendish, 1972): A method of grading of appearance suggested to assist in selecting cases for operation.is based on the degree of scapular elevation, which is separated into four grades. According to this classification, grade 1 (very mild) asymmetry cannot be observed if the patient is dressed; grade 2 (mild) refers to a shoulder that is almost in the correct position and deformity is visible as lump in the web of the neck, even when the patient is dressed; in grade 3 (moderate), the deformity has a shoulder elevation of 2–5 cm and can be easily seen; grade 4 (severe) refers to a shoulder that is markedly elevated, and the superior angle of the scapular lies at the level of the occiput [20] (Figure 37).

Table II: Cavendish grading

Grade	Severity	Clinical features
1	Very mild	Shoulder joints level Deformity invisible when patient is dressed
2	Mild	Shoulder joints level Deformity visible when patient is dressed
3	Moderate	Shoulder joint elevated 2-5 cm Easily visible deformity
4	Severe	Gross elevation (superior angle near occiput) ± neck webbing or brevicollis

About 94 per cent of the patients in our series, were grade 4 on the Cavendish scale, this could be explained by the progressing of the impairment, because the fibrotic tissue will not grow accordingly in the developing child.

In postoperative, the median Cavendish score improved from 4 to 1.

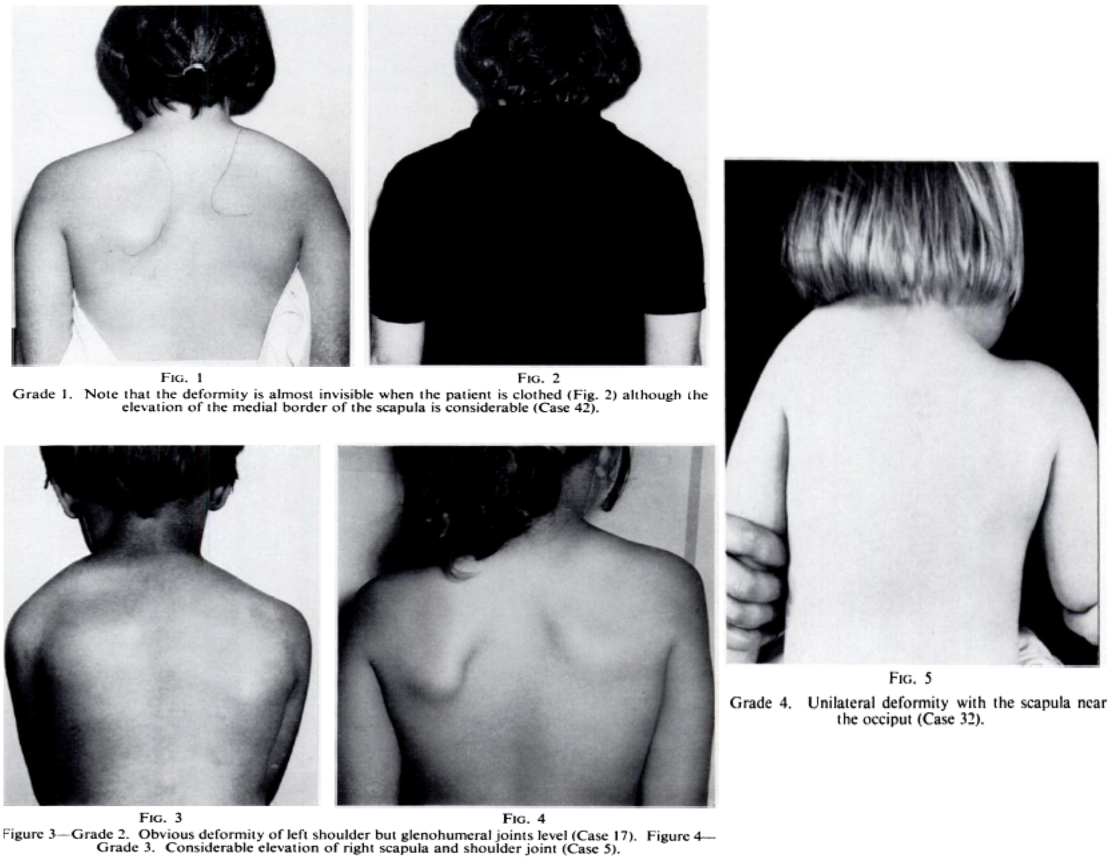


Figure 31: Cavendish grading: Doita M, Iio H, Mizuno K. Surgical management of Sprengel's deformity in adults.

2. Functional impairment:

The functional aspect of the deformity has been attributed to a forward curvature of the superior angle of the scapula over the apex of the thorax, abutment of the medial scapular border against the spinous processes of adjacent vertebrae and the omovertebral bone.

Affected patients have varied degrees of functional limitation resulting from decreased range of motion (ROM) in the shoulder joint.

On examination, passive movement of the glenohumeral joint, including initial abduction, external and internal rotation may be normal. However, scapulothoracic movements are usually limited (by fibrous bands beneath the subscapularis, associated muscular deficiencies, scapulospinous abutment at the inferior angle, and binding of the curved superior angle over the chest). Although glenohumeral motion is normal, the medial scapular rotations face the glenoid downward, such that the normal arc of abduction is similarly rotated and thus not fully available. The most common limitations are abduction and forward flexion. In 40% of patients, abduction is limited to less than one hundred degrees (because of minimal scapulothoracic motion and an inferiorly rotated glenoid) [43]. This is more likely if an omovertebral bone is present. (Fig. 38)

These limitations affect many activities of daily living and it could be observed early in childhood, as the child is listing when reaching forward and upward.

In our series, shoulder Abduction was generally less than or equal to 100 degrees (in 70%).

Mean abduction improved significantly from 108° (90°-170°) to 152° (100°-180°), an improvement of 44° (±standard error).

Sometimes instability of acromioclavicular articulation with inferior subluxation increases mobility of upper limb [44].

Figure 32: Preoperative clinical photograph demonstrating maximal active shoulder abduction; Our case report.

3. Pain:

Generally, Sprengel's deformity tends to be painless and many patients are not diagnosed until adolescence [3].

The pseudarthrosis is the source of the dull pain in the neck and shoulder girdle. It is reasonable that resection of the omovertebral bone could reduce neck pain [5].

The omovertebral bone can be ankylosed at either junction or remain a fibrous pseudarthrosis that may become locally painful [43].

Also, chronic multiregional pain ailments coexist with scoliosis and malformations of numerous vertebrae in an adult person [45].

Neck and/or periscapular pain was among the main symptoms in all neglected SD cases in our series.

C. RADIOGRAPHIC FEATURES

There have been various indices proposed for the study of the morphology of the scapula and the estimation of the degree of scapular deformity. The scapular index is an indicator of the relationship between the breadth and the length of the scapula. It measures the width along the base of the scapula (length of spinous process), the length between its superior and inferior angles, and it is expressed as a percentage (100 × scapular breadth/scapular length). The infraspinatus index describes the width of the scapula relative to the length of the infaspinatus fossa. Both of these indices are estimated radiologically.

1. Plain radiograph

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[46].

Sprengel's deformity is best seen on an anteroposterior radiograph of the chest and both shoulders. The scapular displacement can be measured by a method described by Leibovic et al. [47], which uses three lines drawn on an anteroposterior radiograph, to calculate the superior scapular angle and the inferior scapular angle which give the viewer some idea about scapular rotation [3].

However, the contours of the scapula are difficult to evaluate by plain radiography because of its position, rotation, and shape [32].

Radiographs are also helpful to determine the presence of associated abnormalities (eg, scoliosis, rib abnormalities, omovertebral bone.

The severity of the deformity can also be graded using a radiographic classification devised by Rigault [48] (table, figures):

Table III: Rigault grading

Grade	Severity	Radiologic features
1	Mild	Superomedial angle lower than T2 but above T4 transverse process
2	Moderate	Superomedial angle located between C5 and T2 transverse process
3	Severe	Superomedial angle above C5 transverse process

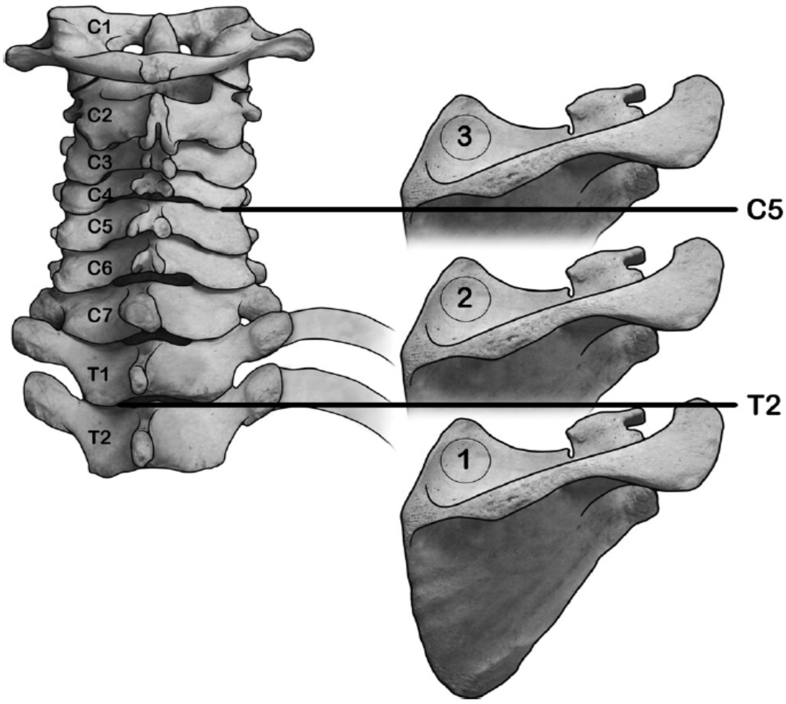


Figure 33: Diagram showing Rigault's radiological classification for Sprengel's deformity of the shoulder. Grade 1, superomedial angle lower than T2 but above T4 transverse process; Grade 2, superomedial angle between C5 and T2 transverse process; Grade 3, superomedial angle above C5 transverse process [49].

In our series, 62% of Sprengel's shoulders were grade 2 (moderate) on Rigault scale, while 38% were grade 3 (severe).

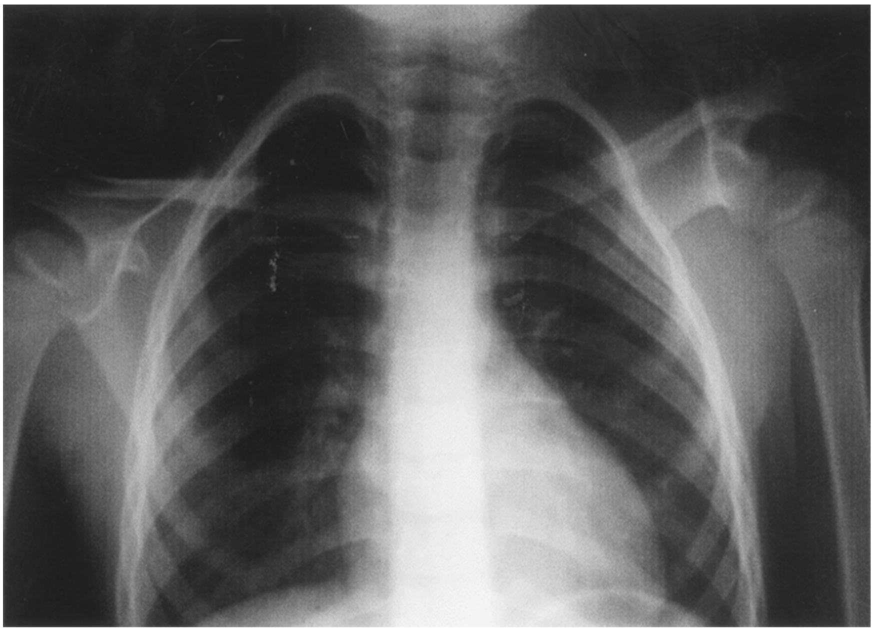


Figure 34: The radiological appearance of Sprengel's deformity [15].

Figure 35: Anteroposterior radiograph showing elevated scapula on the left with spine of the scapula at the level of the sixth cervical vertebra. Also seen is the omovertebral bar (marked by arrows) [50].

Figure 36: Anteroposterior plane X-ray of cervical spine. Note a rightsided omovetebral bone expanding down from the level of C5-6 [5].

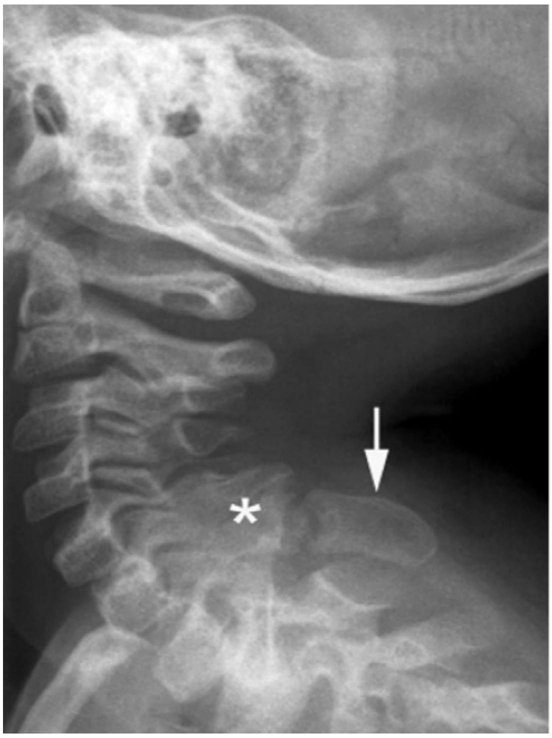


Figure 37: X-ray image (cervical spine in profile). Note the omovertebral bone (arrow) and the C5-C6 zygapophyseal ankylosis (asterisk) [5].

Figure 38: X-ray of a patient with bilateral Sprengel's and omovertebral bones [48].

2. Computed tomography scan (CT)

CT scans may be performed to visualise the affected region, study in detail the anatomy of the affected shoulder(s) and delineate an associated omovertebral bone, its size and tethering points, and especially its attachments and relation to the spine [3]. It is also warranted for severe cases that require surgical management.

Moreover, three-dimensional CT reconstructions are necessary to further evaluate the complex pathoanatomy; it can be very useful for evaluating the position of the scapula in space (lateral tilt in the frontal plane, anterior tilt in the sagittal plan and medial tilt in the transverse plane) and the degree of scapular dysmorphia. It can also help detecting associated bone abnormalities, particularly of the vertebrae, and for planning the surgical procedure, as well as detecting an intraspinous lesion before surgery. [46]

CT scans play a prominent role in preoperative evaluation but cannot adequately assess any non-ossified components or the presence of fibrous or cartilaginous structures [5] [46] (see fig. 39; 40; 41; 42).

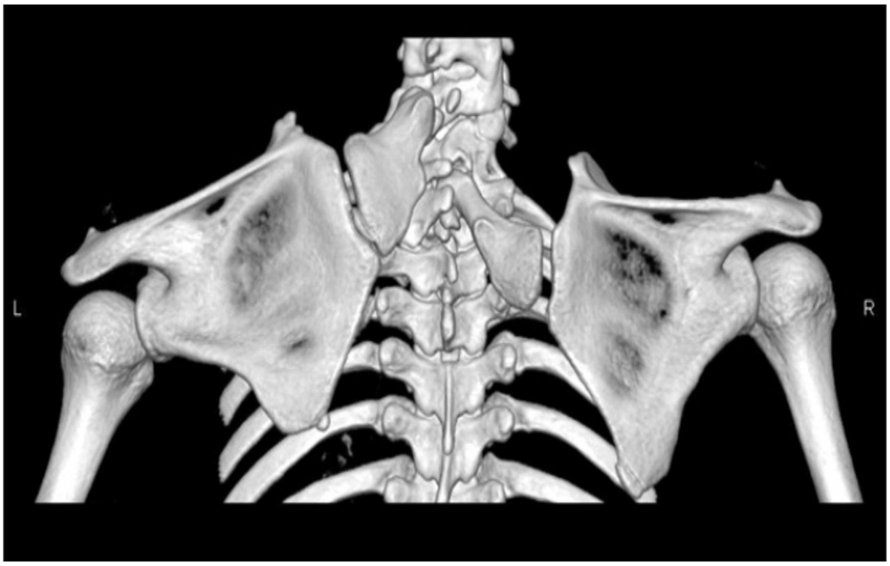


Figure 39: Computed tomography of the upper chest showed cervical fusion and bilateral Sprengel's deformity with omovertebral bone [51].

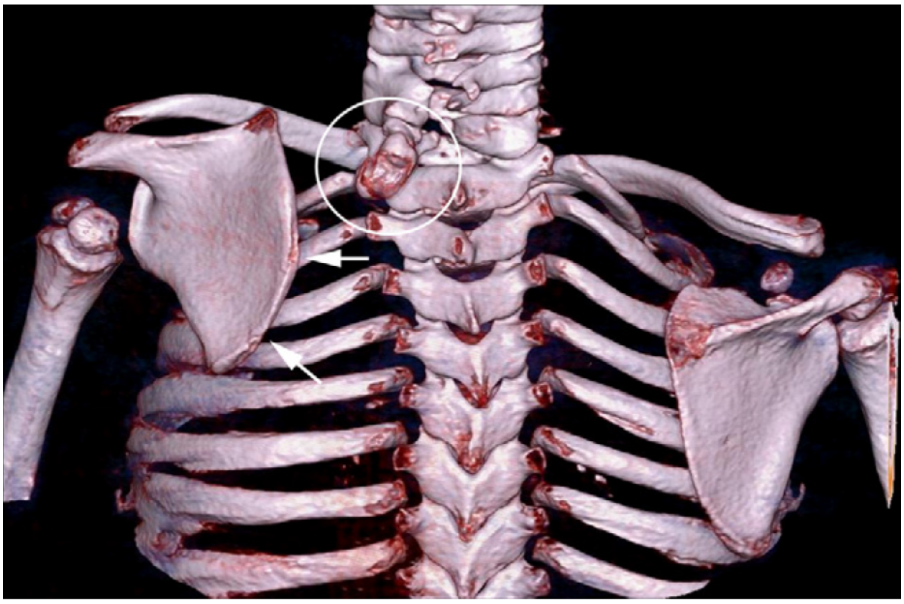


Figure 40: CT scan (posterior view in 3D VRT reconstruction). The omovertebral bone is clearly visible. Note the scapular dysmorphia (arrows), the raised ipsilateral clavicle, and the cervical spina bifida occulta [5].

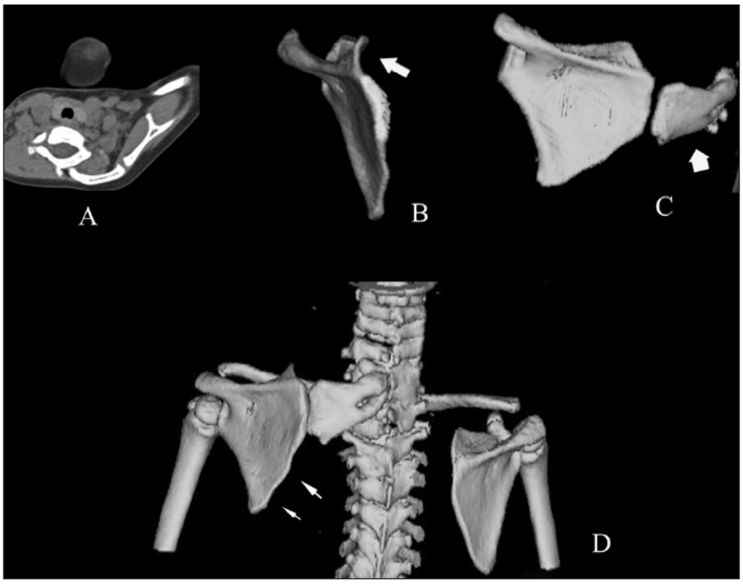


Figure 41: CT scan (A) and 3D reconstruction (B, C and D) show the omovertebral connection (thick arrow) arising from the medial border of the scapula and the vertebral column. (B) shows anterior curving of the supraspinous portion of the affected scapula (arrow). (D) shows the convex medial border and the concave lateral border of the affected scapula (multiple arrows) [52].

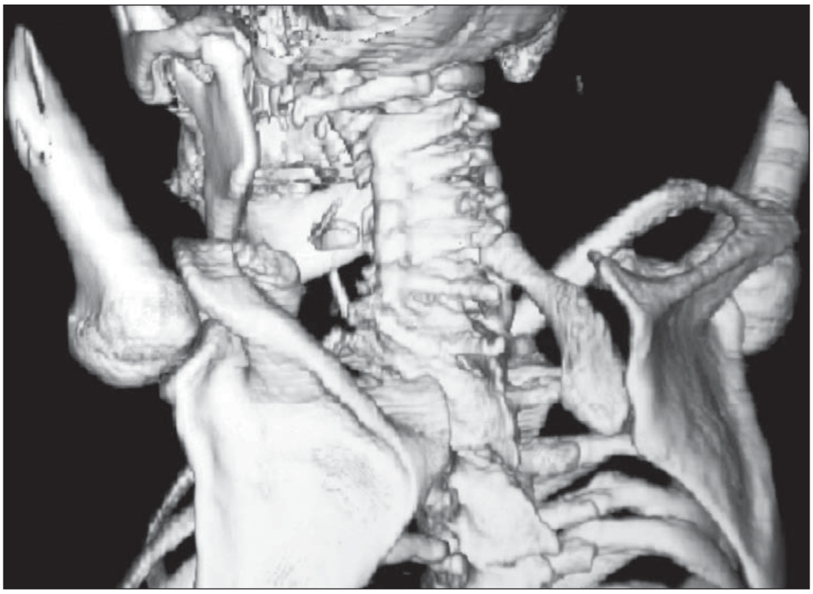


Figure 42: Tomographic imaging with 3-dimensional reconstruction reveals the entire omovertebral bone with distal part expanding along of the medial margin of the scapula [46].

3. Magnetic resonance imaging scans (MRI)

MRI can be useful for analysing fibrous and/or cartilaginous components and for detecting associated medullary abnormalities (diastematomyelia) [5] [31]. It is easy to perform in adult unlike young children who may require sedation or general anaesthesia to avoid movement artefacts.

In case there was no omovertebral bone on plain films or on CT scans, omovertebral connection needs to be evaluated by MRI (fig. 43).

A recent study [53] has reported specific positioning for detecting non-ossified structures as well as possible; The patient must be properly before starting MRI scans. The patient's neck is to be in a lateral flexion position to the opposite side; the same side is lifted up by supporting the shoulder from beneath. Thus, fibrous omovertebral connection is depicted in its longest form in one plane. Omovertebral band is screened best in coronal and axial cross-sections. In the sagittal images, they named the appearance of omovertebral band with peripheral fat tissue as “Ra's eye” (See fig. 44)

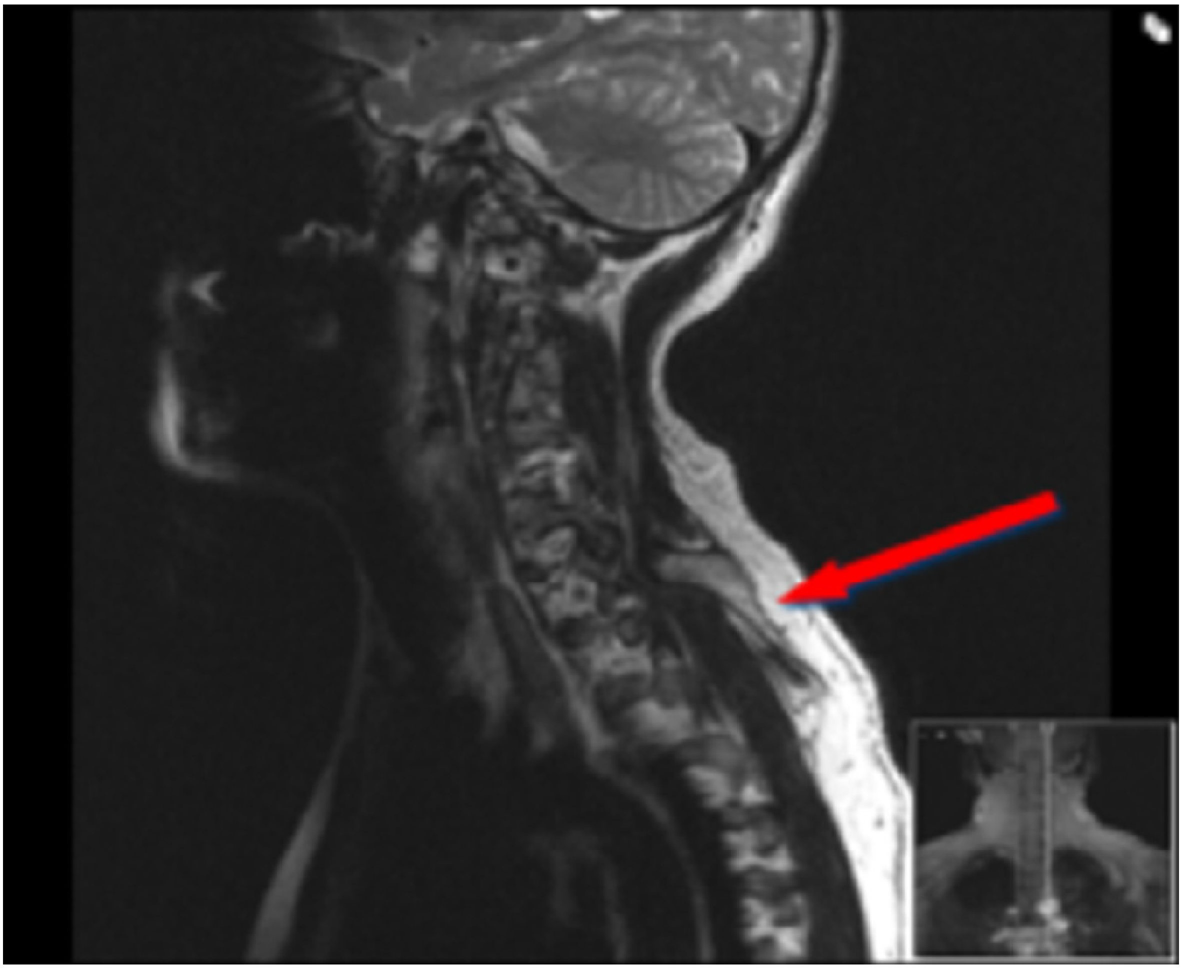


Figure 43: MRI of cervical spine.The cleithrum (arrow) and pseudarthrosis with the thoracic spine are shown. No medial parascapular muscle is visible because of endomuscular ossification [54].

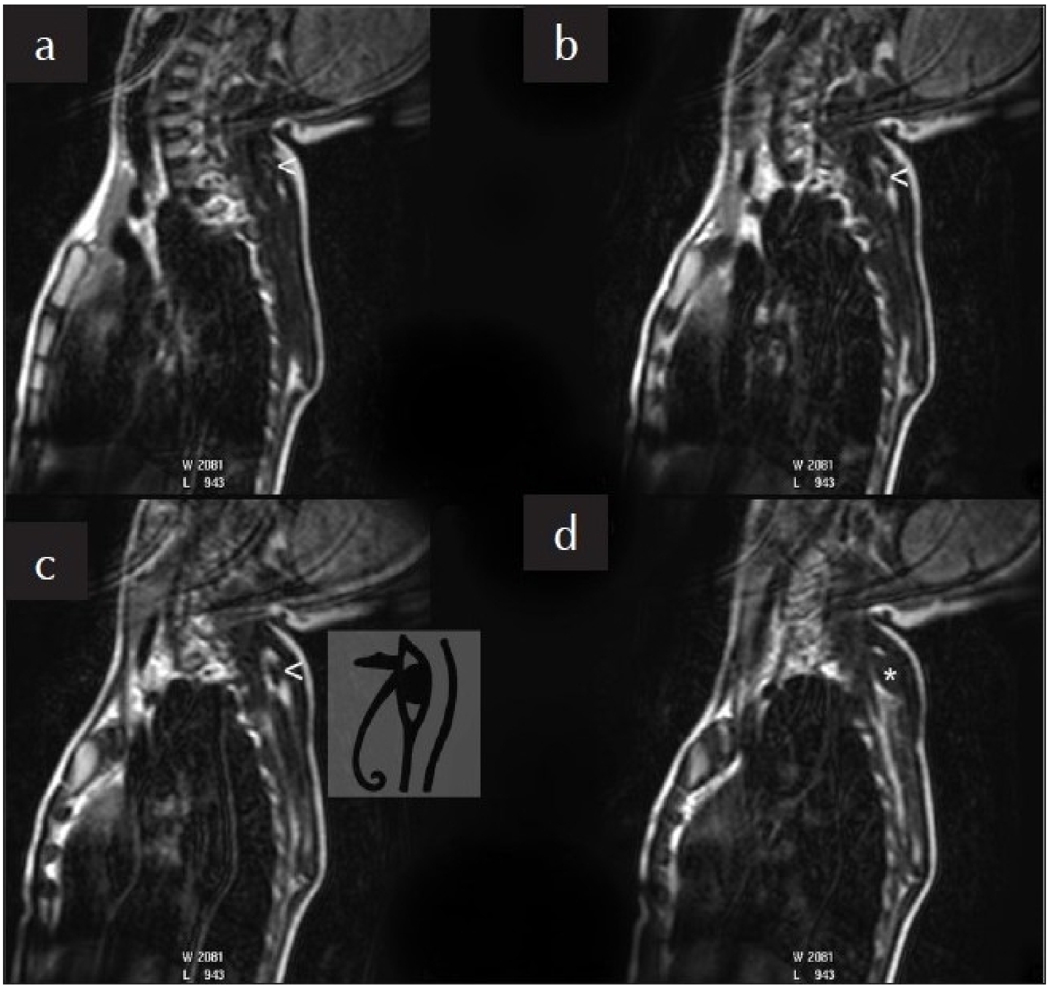


Figure 44: Sagittal MR image series (a, b, c, d) demonstrate the omovertebral band. (c) Omovertebral band and peripheral fat tissue both look like an eye. * Points to the superomedial angle of the right.

4. Ultrasound examination:

An ultrasound examination is, on the other hand, simple, easy to use, and allows easy evaluation of the ossified and non-ossified components of a superficial supernumerary structure. It could be performed in the case of SD where no omovertebral bone can be detected on X-ray images and might be preferable to MRI in young children. In ultrasonography, cartilaginous components have a characteristic appearance: they are hypoechoic, well defined, rounded or oval structures. The only ultrasound observation reported in the literature concerns a structure which was both fibrous and cartilaginous, the morphological appearance differing with the tissue concerned: a hypoechoic, well defined, oval structure (cartilage), and a hypoechoic, well defined, linear structure (fibrous tissue) [5].

5. Prenatal ultrasonographic diagnosis of Sprengel's deformity [55]:

Intrauterine diagnosis can be obtained with prenatal ultrasonography, so families can be prepared for any cosmetic and functional complications that come with SD.

Chinn DH reported a case of prenatal ultrasonographic detection of Sprengel’s deformity; a 33-year-old G1, P0 woman in whom second-trimester showed a prominent nuchal fold predominately left sided, the scapula was elevated, a suspected omovertebral bone was present, articulating between the medial border of the scapula and the cervical spine. Associated abnormalities have also been detected: scoliosis, multiple thoracic vertebral body segmentation abnormalities, supratentorial arachnoid cyst, ambiguous genitalia and absence of right kidney.

Klippel-Feil syndrome was suspected. Amniocentesis showed a normal male karyotype, 46,XY. The amniotic fluid α-fetoprotein level was normal.

The foetus was delivered at 39 weeks. Postnatal evaluation including physical examination, radiographs, magnetic resonance imaging, and ultrasonography confirmed the prenatal observations of Sprengel's deformity with an omovertebral bone and the associated anomalies. The constellation of congenital features was not diagnosed as representing a specific syndrome.

Note that, the elevated scapula was shown best in the axial plane. That only one scapula was visible on a true axial view indicated that the imaged scapula was elevated. One should be cautious in the observation of an elevated scapula, however, because an off-axis axial plane could simulate this finding. In this case, careful observation of the 2 symmetric lamina of the adjacent cervical vertebral body ensured that the image obtained was truly axial to the vertebral body (Fig. 45).

The determination that the nuchal soft tissue thickening was asymmetric was made by careful comparison with the unaffected side in the coronal and axial planes (Fig. 46).

The sonographic detection of the omovertebral bone in its expected location, interposed between the superior scapular margin and the cervical spine. It is conceivable that a cervical rib might simulate the appearance of an omovertebral bone; however, an elevated scapula would not be expected with an isolated cervical rib.

The importance of detecting Sprengel's deformity prenatally lies in attempting to narrow a difficult diagnosis in an anomalous fetus. Furthermore, identification of this entity will permit more specific counseling concerning the orthopedic implications of this anomaly.

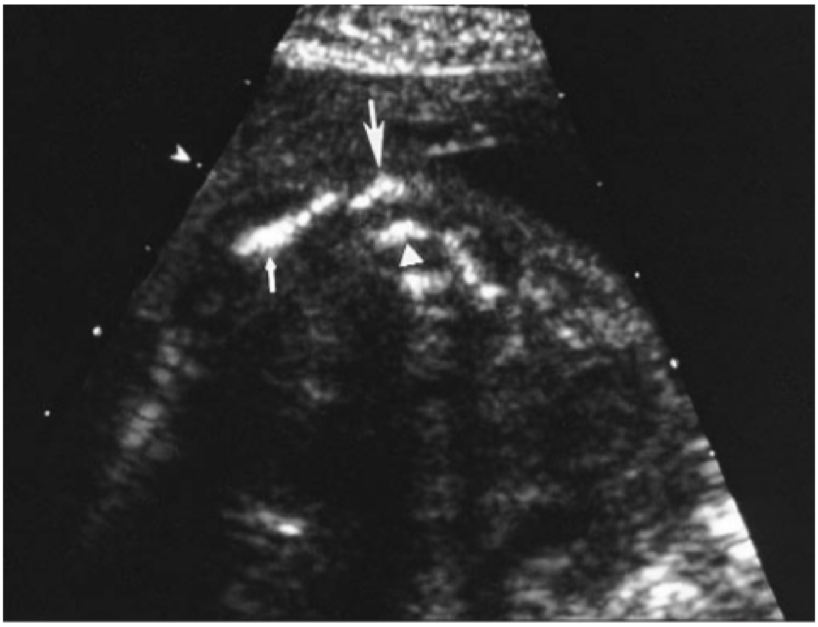


Figure 45: Axial ultrasonogram of the fetal low cervical spine. Note the symmetric lamina of the depicted cervical vertebral body. The scapula is elevated (short arrow) , and the omovertebral bone (long arrow) is interposed between the scapula and the cervical vertebra (arrowhead)

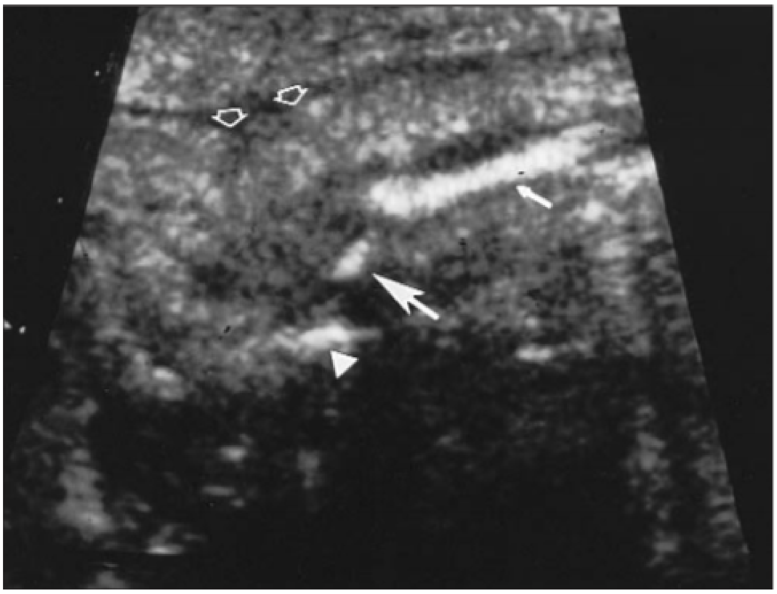


Figure 46: Coronal ultrasonogram of the elevated left scapula. Note the omovertebral bone (long arrow) interposed between the scapula (short arrow) and the lamina of the cervical vertebra (arrowhead). Also note the prominent soft tissue (open arrows) overlying the elevated left scapula.

Figure 47: Frontal chest postnatal radiograph showing the elevated left scapula (short arrow) and the omovertebral bone (long arrow) interposed between the superior scapula and the cervical spine. Also note the scoliosis, multiple thoracic vertebral segmentation abnormalities, and multiple hypoplastic upper left ribs

D. ASSOCIATED ANOMALIES:

Sprengel's deformity almost never occurs as an isolated malformation. It is usually accompanied by various other anomalies, especially in the cervicothoracic vertebrae or the thoracic rib cage.

It is important to note that these associated abnormalities could be diagnosed or suspected by the physical examination.

1. Musculoskeletal abnormalities [36] [45] [56].:

a. Vertebral, Costal and Clavicular abnormalities:

SD is frequently associated with other bone abnormalities which can be costal (aplasia, hypoplasia, synostosis, bifidity, absence, supernumerary ribs, cervical ribs etc.), clavicular (abnormalities in shape and/or position) and, above all, vertebral (Klippel-Feil syndrome, cervical and/or thoracic spina bifida occulta, hemivertebrae, secondary scoliosis, etc.) (see fig. 48; 49; 50; 51).

All 16 cases had associated cervical vertebral anomalies; with Scoliosis accounting for 68 per cent, Kippel-Feil syndrome was found in 25% of the patients, spinal dysraphism with Spina bifida in 2 patients and Syringomyelia in 1 patient.

Only one case had clavicular shape abnormality (Our report).

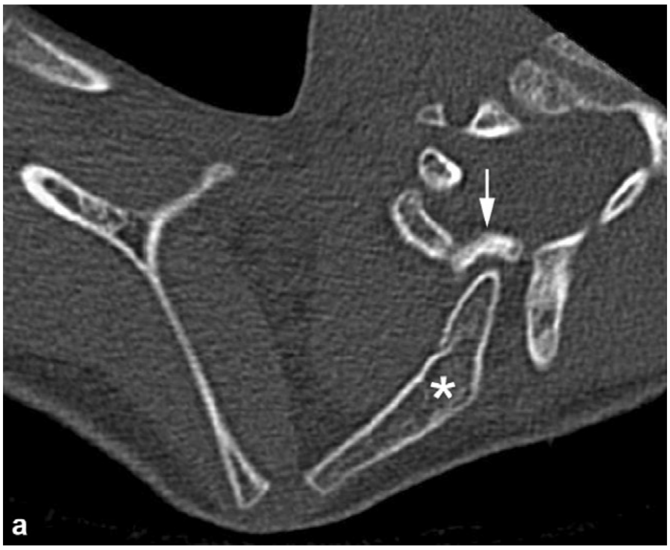


Figure 48: CT scan (transverse slice in the bone window). The omovertebral bone (asterisk) is visible between the lamina of a cervical vertebra and the medial border of the scapula. Note the associated bone scalloping (arrow); [5].

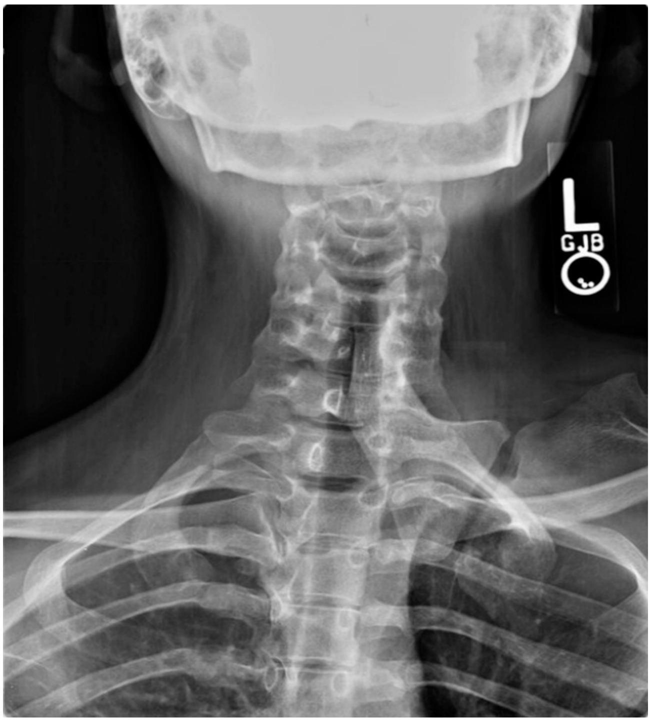


Figure 49: X-Ray imaging demonstrating the omovertebral bone between the posterolateral spinal elements of the C5, C6, and C7 vertebrae and the superomedial border of the scapula. Also note the congenital fusions between the C5, C6, and C7 vertebrae [43].

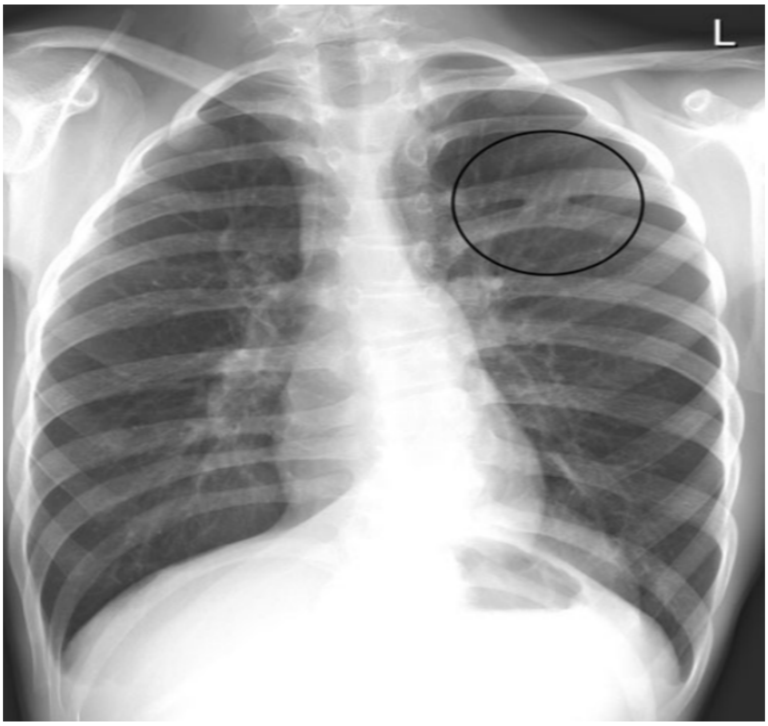


Figure 50: Chest radiograph illustrating left-sided fused fourth and fifth ribs (encircled) and a sinistroconvex thoracolumbar scoliosis [56].

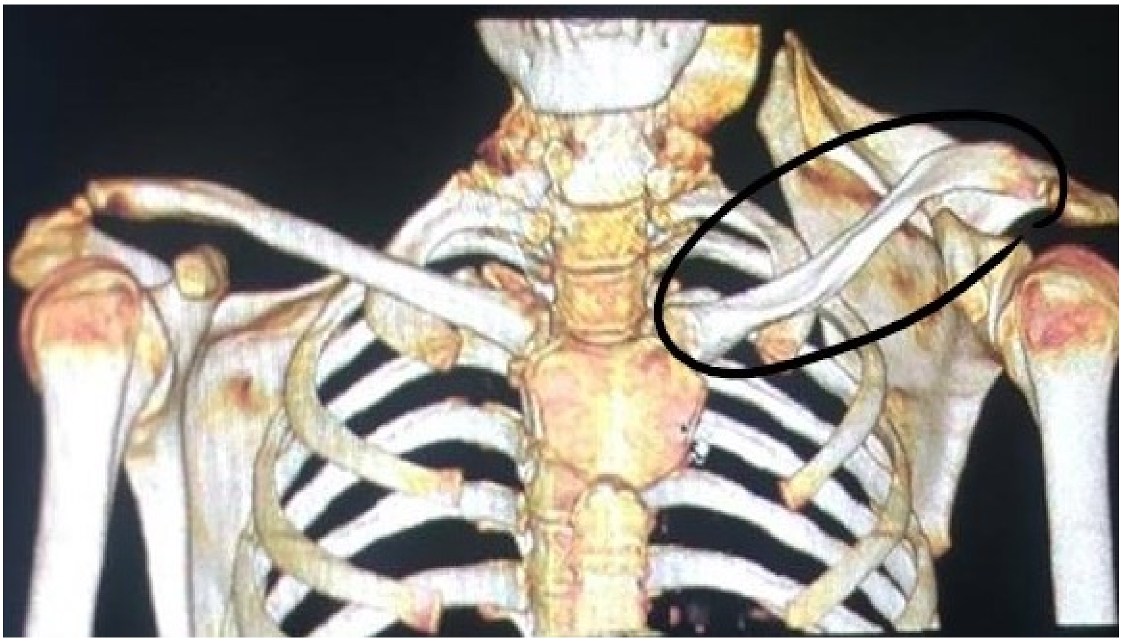


Figure 51: Volume rendering reconstruction anteroposterior view showing deformed and shortened left clavicular. (Our case)

b. Muscular abnormalities:

Hypoplasia of the cervical muscles; The trapezius, rhomboid, or levator scapulae may be absent, hypoplastic, or contain multiple fibrous adhesions. The serratus anterior may be weak, leading to winging of the scapula. Other muscles, such as the pectoralis major, latissimus dorsi, or the sternocleidomastoid torticollis, may be hypoplastic and similarly involved.

In our series, muscular abnormalities were found in 37% of the cases (partial endomuscular ossification of medial scapular suspension muscles [54]).

We are comparing the frequency of musculoskeletal abnormalities in different series (Table III).

6. Neurological abnormalities [57]

Neurological problems of Spinal Cord and/or Medulla insufficiency presenting in infancy are related to the compromise of the Cranio-vertebral junction.

Cervical fusions at the lower levels (as in SKF), if they are not massive, typically does not manifest until late in life (Tethered cord syndrome) [34], when degenerative changes or instability of adjacent segments develops leading to narrowing of the Spinal Canal.

The case of an omovertebral bone in association with Sprengel deformity and KFS as a cause of cervical myelopathy is unique in the adult population. [35]

However, it should be considered in the setting of neurological symptoms characteristic of cervical myelopathy in patients with obvious skeletal dysmorphias of unknown etiology.

Early diagnosis and surgical treatment may prevent secondary neurological damage.

A relationship between a Sprengel deformity and diastematomyelia [58] (a condition in which a part of the spinal cord is split, usually at the level of the upper lumbar vertebra) has been shown and may be linked to Sprengel deformity in the anomalous embryonic development.

Association of Sprengel’s deformity with other multiple neural tube defects (spina bifida, Klippel-Fiel syndrome, diastometamyelia) must always be kept in mind and comprehensive neurological examination should be carried out in a patient with Sprengel's deformity.

In our series, two patients had cervical myelopathy (12%), they also had as associated abnormalities; an omovertebral bone and KFS.

7. Other abnormalities [59] [60]

A multiple of other abnormalities is found, including but not limited to congenital dislocated hip, talipes equinovarus, pes planus, radial club hand, congenital dislocation of the radial head, symbrachydactylia, pterygium colli, cleft palate, facial asymmetry, aortic arch, congenital heart disease, sinus inversus viscerum, absent thumb, tethered cord, and atlantoxial instability, urinary tract and renal abnormalities (absent, ectopic kidneys) (fig. 52; 53).

In our series, renal abnormalities were reported in 4 patients (agenesis), and one patient had cleft lip and palate, for which he was operated in childhood.

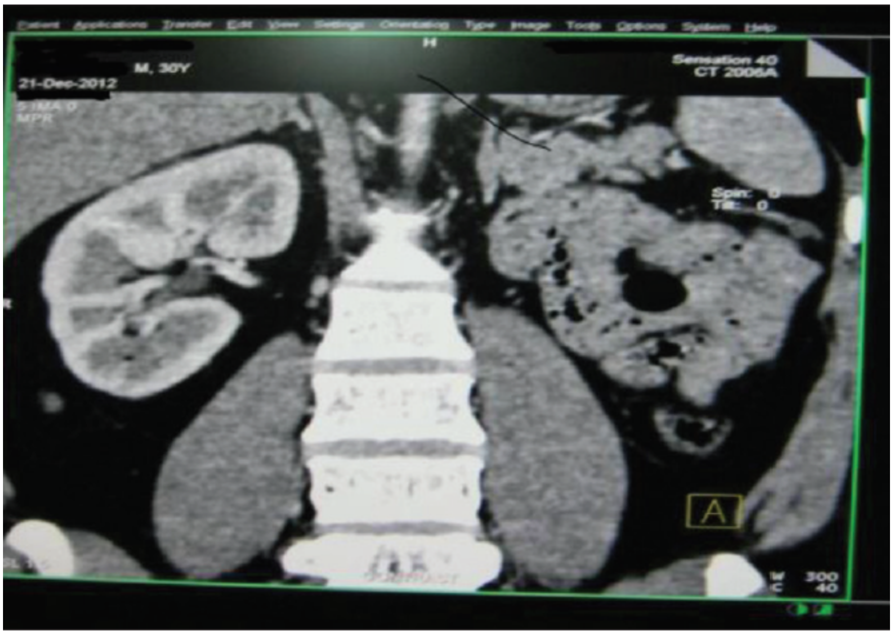


Figure 52: 3-D Computed Tomographic Scan showing absent kidney on left [59].

Figure 53: A and B, Volume rendering reconstructions with anteroposterior view (A) and left-lateral view (B) showing a bovine aortic arch variant (white arrow) and a U-shaped dorsal running loop of the left subclavian artery. [60]

The posterior course of the proximal part of the left subclavian artery entering the intraspinal space through the left-sided fused vertebrae at the T1 level with the same-level dorsal exiting is clearly seen .

2. Associated Syndromes:

- Klippel-Feil syndrome (characterised by the congenital fusion of any two of the seven cervical vertebrae)
- Greig syndrome (characterised by polydactyly, cutaneous syndactyly, ocular hypertelorism, macrocephaly and a high, prominent forehead)
- Poland syndrome (characterised by hypoplasia or absence of the pectoralis on one side of the body and cutaneous syndactyly of the ipsilateral hand)
- VATER association (characterised by vertebral defects, imperforate anus, tracheoesophageal fistula, radial dysplasia and renal dysplasia)
- DiGeorge Syndrome (characterised by heart defects, cleft palate, autism, learning disabilities, recurrent infections and hypocalcemia)
- Floating-harbor syndrome (characterised by short stature, delayed bone growth, delayed communication skills and distinct facial features)
- Goldenhar syndrome (characterised by incomplete development of the ear, nose, soft palate, lip, and mandible)

These associated conditions are of great clinical significance for several reasons: Parents and patients must be made aware of the deformities that will remain unchanged after surgical correction of the undescended scapula (such as scoliosis with Klippel-Feil syndrome) so that their expectations are realistic.

Tethered cord and diastematomylia should be ruled out prior to considering scoliosis surgery. Ipsilateral rib cage abnormalities may predispose these patients to thoracic outlet syndromes with scapular replantation. Midline dissection should be avoided in patients with thoracic spinal dysraphism. Prior to surgery renal status should be evaluated with ultrasonography, if not intravenous pyelography, because of the high incidence of associated renal abnormalities [3][31][58].

Table IV: Musculoskeletal abnormalities in different series. [61]

AUTHOR	Nbr of cases	Vertebral abnormalities			Scoliosis	Costal	Muscular	OVB	Diastemato-mylia
		High	Medium	Low					
Alaaeldin Azmi Ahmad	11	6 (54%)	1 (9%)		3 (27%)	2 (18%)		7 (63%)	
Eulenburg	3						3		
Willet	1							1	
Sprengel	4				1	4			
Horwits	136		22 (16%)		52 (38%)	22 (16%)	46 (34%)	34 (25%)	
Hayashi	126	14 (11%)	5 (6%)	6 (6%)	58 (46%)	14 (11%)	11 (9%)		
Fairbank	18	3 (17%)	7		10	4	8		
Smith	50	2 (4%)	12 (24%)		8 (16%)	20 (40%)		14 (28%)	
Junge	121	7 (6%)	52 (43%)	25 (21%)	56 (46%)	30 (25%)	23 (19%)		
Jeannopoulos	35	4 (11%)	23 (66%)	7 (20%)	20 (57%)	17 (49%)	1 (3%)	11 (31%)	
Green	15		12			12		6	
Cavendish	100	24 (24%)	39 (39%)	28 (28%)	39 (39%)	25 (25%)	14 (14%)	19 (19%)	
Ross	77		21 (27%)	3 (4%)	42 (55%)	23 (30%)		16 (24%)	
Abdalimuhsin	15	2			5		1		
Our series	17	5 (30%)	3 (18%)		11 (65%)		5 (30%)	17 (100%)	

3. Sprengel's deformity and Klippel-Feil syndrome: [62] [63] [64]

Klippel-Feil syndrome, initially reported in 1884 by Maurice Klippel and André Feil, is characterized by congenital fusion of two or more cervical vertebrae.

It occurs in a heterogeneous group of patients unified only by the presence of a congenital defect in the formation or segmentation of the cervical spine, and may be associated with other organ system anomalies, in addition to their spine abnormalities.

These anomalies may include: Scoliosis, spina bifida, hearing difficulties, eye abnormalities, cleft palate, dental problems, genitourinary problems such as abnormal kidneys or reproductive organs, visceral anomalies, heart malformations, or lung defects that can cause breathing problems.

Approximately 35% of patients with Klippel–Feil syndrome also have Sprengel's deformity.

Klippel-Feil syndrome is associated with both spontaneous and progressive neurologic sequelae, secondary to the progression of the skeletal pathology (unstable fusion pattern, craniocervical anomalies, and associated spinal stenosis) and could be aggravated in case of neck trauma. [65]

This heterogeneity requires comprehensive evaluation of all patients and treatment regimes that can vary from modification of activities to extensive spinal surgeries. Furthermore, it is unclear whether Klippel–Feil syndrome is a unique disease, or if it is one part of a spectrum of congenital spinal deformities.

Although once thought that KFS patients presented with the classic clinical triad of manifestations of a low posterior hairline, short neck, and limited neck range of motion, studies have shown that 34% to 74% of KFS patients present with such findings.

However, the “hallmark” of this condition is the improper segmentation of cervical motion segments, discernible as congenitally fused vertebrae with or without associated manifestations.

Various authors have proposed classification schemes to elucidate on the patterning of congenitally fused vertebrae (cervical, thoracic, and lumbar combined), risk of neurologic injury, genetic disposition, and intersegmental mobility on the development of symptoms or associated abnormalities. However, according to some studies, neurologic compromise is independent of various fusion patterns in KFS. Conversely, others contend that change in mobility in the lower cervical spine, attributed to the abnormal vertebral fusion associated with KFS, contributes to altered stress forces and/or a degenerative cervical disease process.

Although several authors have stressed that cervical spine-related symptoms in KFS patients are pronounced in adulthood, critical review of the literature reports up to 50% of pediatric KFS patients to be symptomatic .

In our series, Klippel-Feil syndrome represented 30% of all cases, included our report. In fact, the diagnosis was suspected in our patient basing on the clinical features: short neck, posterior low hair implantation and restricted neck range of motion (figure).

Anteroposterior plain radiographs were reviewed to assess the presence of congenitally fused segments.

A computed tomography (CT) scan with 3D reconstruction confirmed the presence of the KF syndrome showing fusion of cervical vertebrae from C3, C4 to C7.

Congenitally fused segments were defined as any presence of bone bridging across the interbody space or posterior elements, and the absence of motion on flexion and extension.

Figure 54: Photograph of our patient showing short neck and low posterior hairline related to KFS.

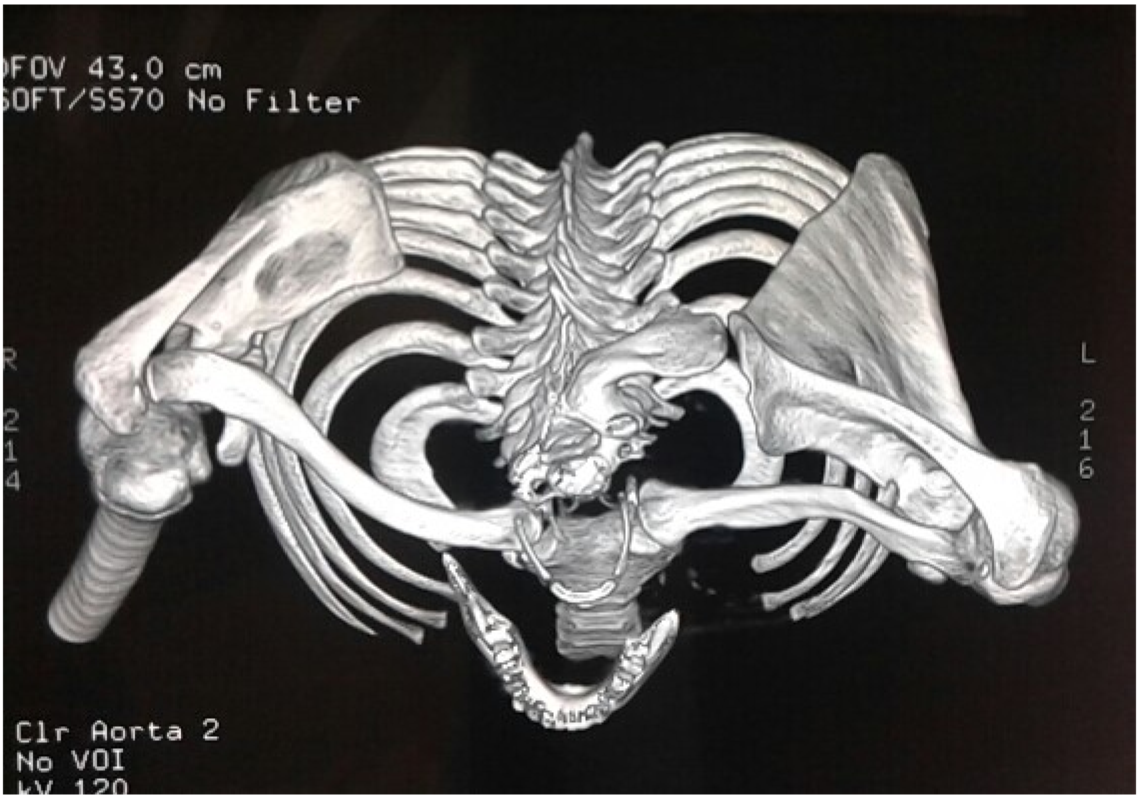


Figure 55: computed tomography (CT) scan with 3D reconstruction (upper view) showing cervical fused vertebrae**.**

Feil subsequently classified the syndrome into the following three types (Table IV, Fig. 55):

- **Type I** - Massive fusion of the cervical spine
- **Type II** - Fusion of one or two vertebrae
- **Type III** - Presence of thoracic and lumbar spine anomalies in association with type I or type II Klippel-Feil syndrome

Since the original description, other classification systems have been advocated to describe the anomalies, predict the potential problems, and guide treatment decisions.

In a series of articles, Samartzis et al., suggested their own classification system [62], which stratified patients as follows:

- **Type I** - Single-level fusion
- **Type** II - Multiple, noncontiguous fused segments
- **Type III** - Multiple, contiguous fused segments

Gray et al [63] described 462 patients with Klippel-Feil syndrome and found that the level of fusion did not greatly affect the incidence of neurologic symptoms. The most frequent level they identified was a defect of the occiput to C1, C2, and C3. These produced the most symptoms; lesions below C3 and 4 were slightly less likely to cause symptoms. Twenty-seven percent of symptoms occurred in the first decade.

Nagib et al [64] described three types and related the incidence of neurologic symptoms to each type as follows:

- **Type I** - Two sets of block vertebrae with open intervening spaces that can sublux gradually or with acute trauma
- **Type II -** Craniocervical anomalies with occipitalization of the axis and basilar invagination; this causes increased mobility at the craniocervical level and can lead to foramen magnum encroachment; it can be associated with Arnold-Chiari malformation and syringomyelia
- **Type III** - Fusion of one or more levels with associated spinal stenosis

Table IV: Radiographic Classification Scheme of Congenitally Fused Cervical Segments in Patients with Klippel-Feil Syndrome

Classification Type	Criteria
I	Single congenitally fused cervical segment
II	Multiple non-contiguous, congenitally fused segments
III	Multiple contiguous, congenitally fused cervical segments

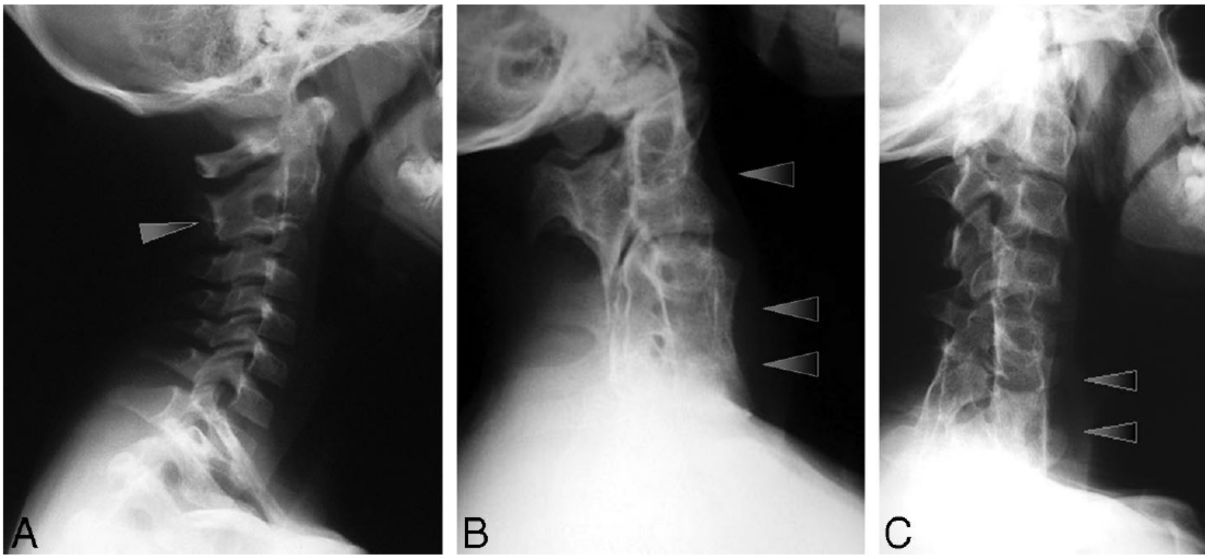


Figure 56: Klippel-Feil classification types of congenitally fused cervical patterns: A, Type I; B, Type II; and C, Type III [62].

E. MANAGEMENT OF SPRENGEL DEFORMITY:

1. Conservative management:

The two main problems encountered in Sprengel's deformity are cosmetic malformation and impairment of shoulder abduction. Conservative treatment has no positive effect on this disorder at advanced age.

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, but should be followed until skeletal maturity because the deformity and functional loss can progress according to some Authors [43][62]. At some institutions, this management consist of annual observation, to assess for progression of shoulder abnormality and associated deformities [66].

In adults, nonoperative management of Sprengel deformity provides no significant improvement in either cosmetic deformity and improvement in extremity function. For some Authors, conservative treatment is not acceptable in SD.

In our series, two patients had nonoperative treatment. They were advised for conservative management with analgesic.

Because the condition is uncommon, it is difficult to develop a consensus of management. The patient's preference for operative and nonoperative intervention is considered in the decision-making.

2. Surgical management:

Even though surgery for Sprengel deformity in adults has been performed, no series has short-term or long term follow up results to guide the chirurgical approach.

a) Objectives:

The main goals of surgery are to improve the cosmetic appearance and to improve the scapular function as well as decreasing neck pain.

b) Principle:

The basic surgical technique is to first release the cause of the scapular binding and secondly to relocate the scapula if possible.

c) Indications:

The indications for surgery are functional impairment, neck or periscapular pain and cosmetic dissatisfaction caused by shoulders asymmetry. The Cavendish scale, based on cosmetic criteria, is popular but subjective, and should be used in combination with Rigault's classification.

Surgical treatment is fully indicated in mild and moderate cases (grades 2 and 3) because it provides improvement in both function and appearance. When symptoms are minimal (The presence of only a mild deformity with minimal restriction of movement), no intervention is required.

When considering surgery, the associated congenital abnormalities, medical co-morbidities should be taken in consideration. The final appearance is dictated by the pre-operative state and the presence of associated spinal deformities has a marked negative impact. When surgery is for cosmetic gains alone, the patient and carer's expectations must be carefully managed. Concurrent deformities will likely persist along with an unsightly surgical scar.

d) Surgical techniques:

Corrective operations range from simple excision of the superior angle of the scapula [20] [67] [68] to subtotal scapulectomy [69] [70]. Contemporary surgical procedures are based on one of three operations and their modifications: Green's procedure, Woodward's procedure and Scapular osteotomy, as well as resection of omovertebral bone if present.

Traditional procedures rely on intact and normal musculature to facilitate scapular transposition.

The Green or Woodward procedures generally are indicated for patients 3 to 8 years old, when the surrounding soft tissue remains supple, after this age, they become less pliable, and because the risk of brachial plexus injuries from stretching or compression by the clavicle increases with age.

In adults, resection of an omovertebral bone, the superomedial portion of the scapula or both, without lowering the scapula, has been recommended. This treatment is safe, technically is not demanding, and can provide improved shoulder function and improved cosmetic appearance [21] [43].

- ❖ **Excision the superomedial portion of the scapula, Operative technique: [20]**

A transverse incision, to avoid spreading ofthe scar, is made just above the spine of the scapula, from which the upper fibres of trapezius are detached as far as the acromion. Medially, the fibres of trapezius are split beyond the medial border of the scapula. The trapezius is then retracted, exposing the upper part of the scapula and the omo-vertebral bone. The levator scapulae and rhomboids are divided along their scapular attachments down to the lower border of the rhomboids.

The amount of scapula excised varies slightly from case to case; it should always include most of the supraspinous part with the attached parts of supraspinatus and subscapularis, the medial end of the spine and any prominent knob on the medial border which might impinge on the spinous processes of the vertebrae. This extraperiosteal excision prevents regeneration which would spoil the result. The omo-vertebral bone may be excised subperiosteally.

- **Green's procedure: [71] [72] [73] [74]**

In the Green's procedure (Fig. 57; 58; 59), muscles are detached from their scapular insertion, the trapezius muscle is then elevated extraperiosteally and reflected medially, thus exposing underlying muscles and the scapula. The supraspinatus muscle is then detached from the scapula along with its periosteum. The omovertebral bone, when present, is excised. The insertions of the levator scapulae muscles on the superior angle of the scapula and of the rhomboid muscles on the medial border of the scapula are dissected. The supraspinous fossa of the scapula is resected, taking care to avoid injury to the suprascapular neurovascular bundle. The scapular attachments of the latissimus dorsi and serratus anterior muscles are detached from the anterior aspect of the scapula. The scapula is then displaced distally down to the level of the normal side using the superiomedial angle of the scapula as a landmark rather than its tip because of constant scapular hypoplasy. Once the scapula is in its corrected position, the muscles are reattached to it, the supraspinatus muscle to the base of the scapular spine, rhomboid muscles should be reinserted without compromising the new position of the scapula in the frontal plane by lengthening it if necessary, in order to achieve good orientation of the glenoid and prevent the glenohumeral articulation from drifting back into varus, and therefore increase in the range of abduction.

Postoperative resisted pendular shoulder exercises are used for several weeks, after which active ROM exercises are encouraged.

Some modifications were brought to the initial Green's procedure: a clavicular osteotomy is performed first to reduce the risk of brachial plexus palsy (BPP); the insertion of the serratus anterior is dissected from the spinal border of the scapula and scapula adherence to the thoracic wall is separated; insertion of the supraspinatus muscle is resected; the inferior pole of the scapula is sutured to the thoracic cage into a pocket of the latissimus dorsi muscle.

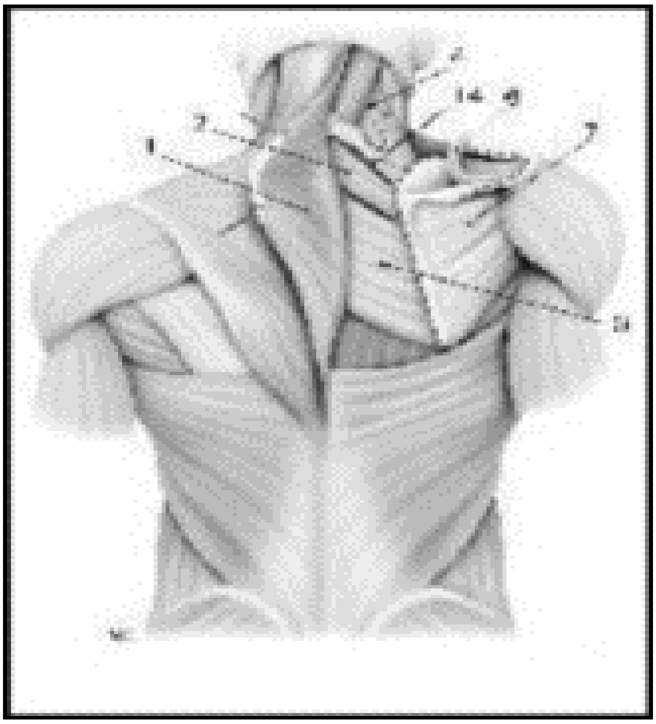


Figure 57: Trapezius muscle disinsertion step from its scapula and clavicle attachments (from EMC: Techniques chirurgicales, Damin J.-P.).

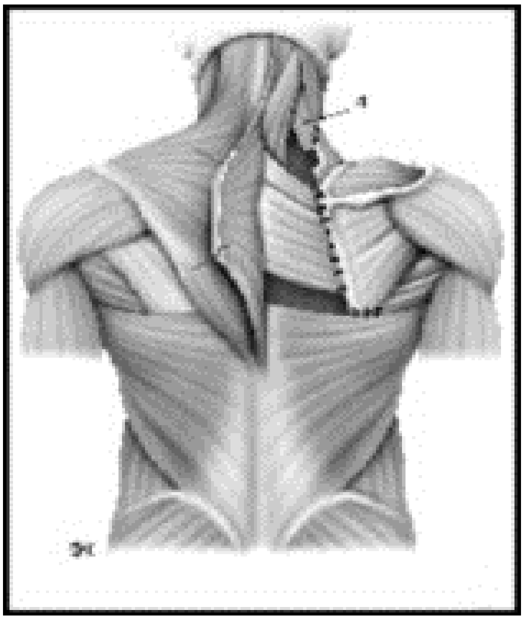


Figure 58: Schematic representation following: supraspinatus fossa bone resection, omovertebral bone resection, figure of L type lenghtening of levator scapulae, global lowering, rhomboid muscles reattachment at a higher site and distal tip scapula fixation (from EMC: Techniques chirurgicales, Dasmin J.-P.).

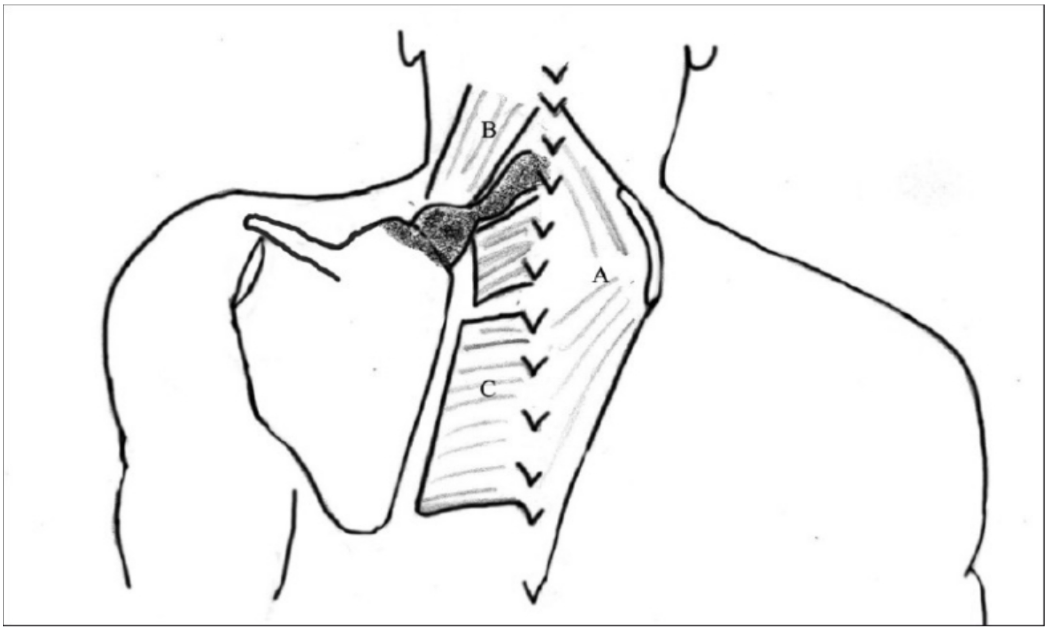


Figure 59: Diagrammatic representation of Green's procedure. Shaded region indicates the area of resection (omovertebral bar, supraspinous fossa). (A – Dis-inserted trapezius; B – Levator scapula; C – Dis-inserted rhomboids – major and minor) [71].

❖ Woodward's procedure: [11] [41] [66].

The patient is placed in lateral decubitus position with the entire arm and shoulder girdle included in the operative field. A subperiosteal exposure of the clavicle can be developed through a transverse incision above the middle third. A segment approximately 2 cm long is removed at the midportion, morcellized, then replaced in the periosteal tube, which is carefully repaired and the skin closed. The patient is then rolled into the prone position without redraping. The head is supported on a craniotomy headrest with the neck slightly flexed. A midline incision is then made from C4 spinous processes to the T9 spinous process. The skin and subcutaneous tissues are undermined on the involved side out to the vertebral border of the scapula to allow better visualization of the musculature and to allow scapular lowering without skin tension. The lateral border of the distal trapezius is identified and should be bluntly separated from the latissimus dorsi. The origin of the trapezius is then sharply dissected from the spinous processes. The origins of the rhomboids major and minor are identified and divided. The rhomboids and the upper portion of the trapezius are separated from the serratus posterior superiorly and the sacrospinalis by the deep fascial layer. This tissue plane must be gained, as the intact aponeurosis is essential for scapular fixation. When this fascial layer has been removed from the spinous processes to the upper limits of the exposure, the sheet of musculature can be retracted laterally to expose the omovertebral connection, when present.

When removing an omovertebral bone, it is best to do so extraperiosteally. A bone-biting rongeur can be used to remove the bone from the scapular angle and from its connection to the cervical spine. The spinal accessory nerve on the undersurface of the trapezius and along the vertebral border of the scapula must be protected, especially when removing the omovertebral bone. The transverse cervical artery must also be protected during the dissection. The supraspinous portion of the scapula should be removed extraperiosteally when its anterior curvature limits downward caudal mobilization or when it remains as a prominence after the scapula is relocated.

The shoulder girdle and combined trapezius and rhomboid muscular sheet are then moved caudally until the spine of the scapula on the affected side comes down to the same level as the scapular spine on the uninvolved shoulder. Lowering the inferior angle to the level on the normal side is overcorrection and shoulder be avoided.

While maintaining the position of the scapula, reattach the aponeurosis of the trapezius and rhomboids. The redundant tissue of the trapezius and aponeurosis is folded over and sutured in a pants-over-vest fashion.

The skin and subcutaneous tissue should be closed meticulously to minimize the spreading of the scar in the superior aspect of the incision. A Velpeau bandage is then applied and worn for approximately two weeks [25].

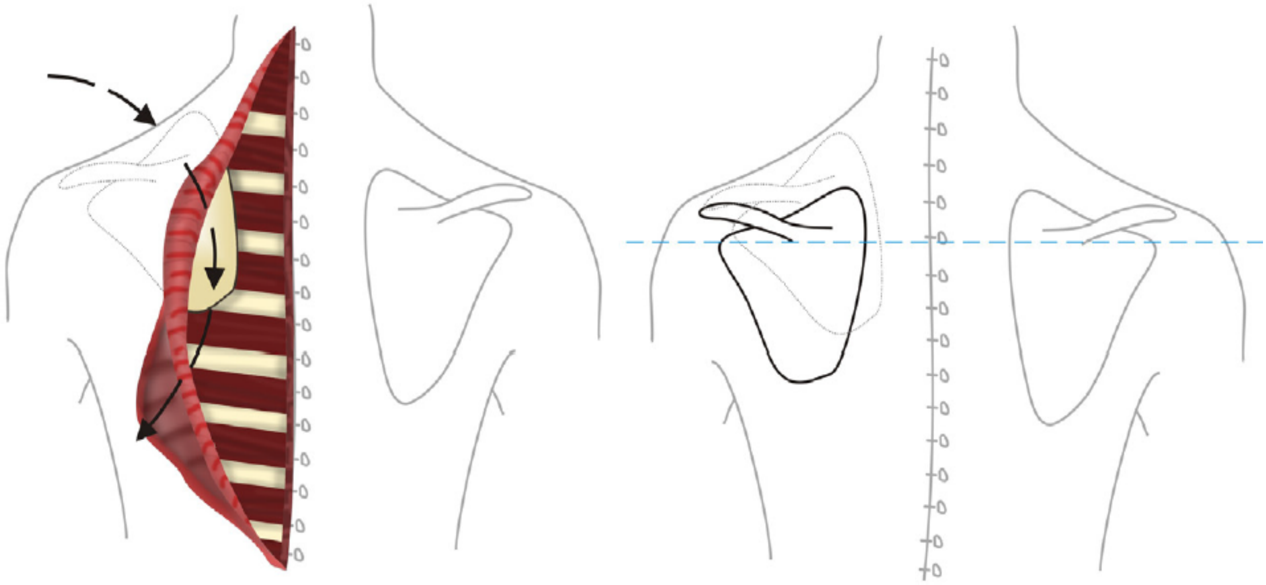


Figure 60: Surgical technique by Woodward et al. After a midline incision, the vertebral attachments of the aponeurosis of the trapezoid, rhomboid, and levator scapulae muscles are detached to be able to rotate and relocate the hypoplastic left scapula more caudally on the thorax. The blue line represents the reference point for the position of the scapular spine of the hypoplastic scapula at the same level as the normal controlateral scapula [41].

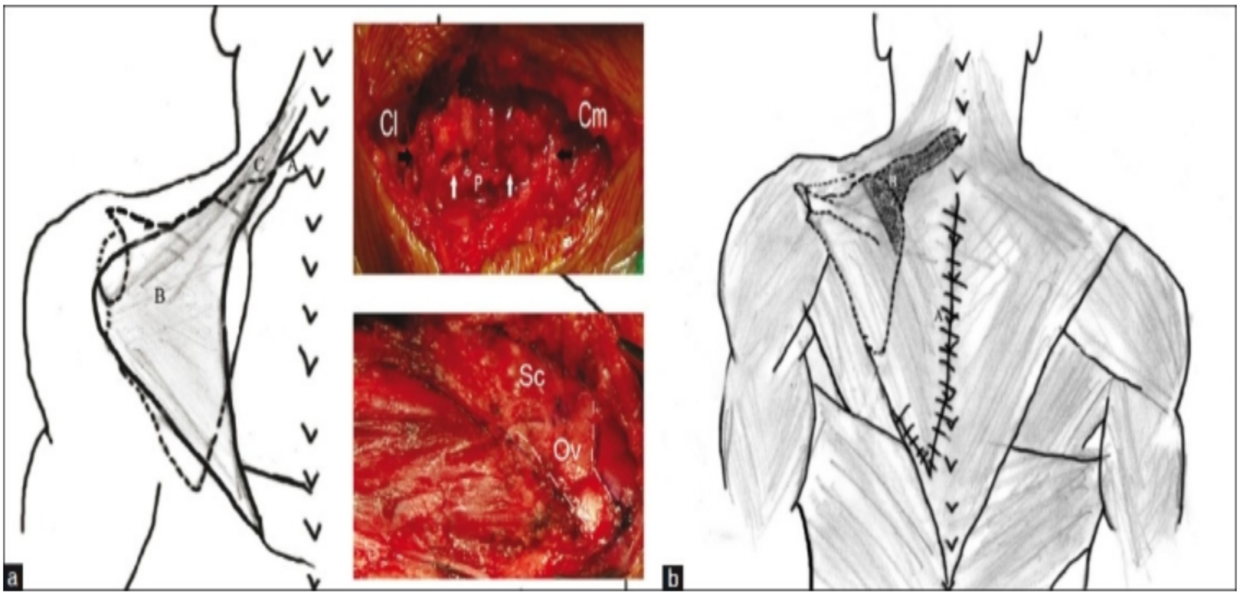


Figure 61: (a) Diagrammatic representation of Woodward's procedure: The origins of trapezius and rhomboids (B) are resected from the spinous processes (A – Omovertebral bar; B – Elevation of trapezius and other scapular musculature; C – Levator scapulae). Inset – Top right shows morselization of the clavicle; 'Cl' and 'Cm' – The lateral end and the medial end of the clavicle, respectively; P – Periosteum sutured over the morselized part; Black arrows – Extent of the morselization; White arrows – Sutured periosteum.; (b) Shaded area indicates extraperiosteal excision of the omovertebral bar and the supraspinous portion of the scapula. The aponeurosis of trapezius and rhomboids (A) are sutured over the scapula in the corrected position [44].

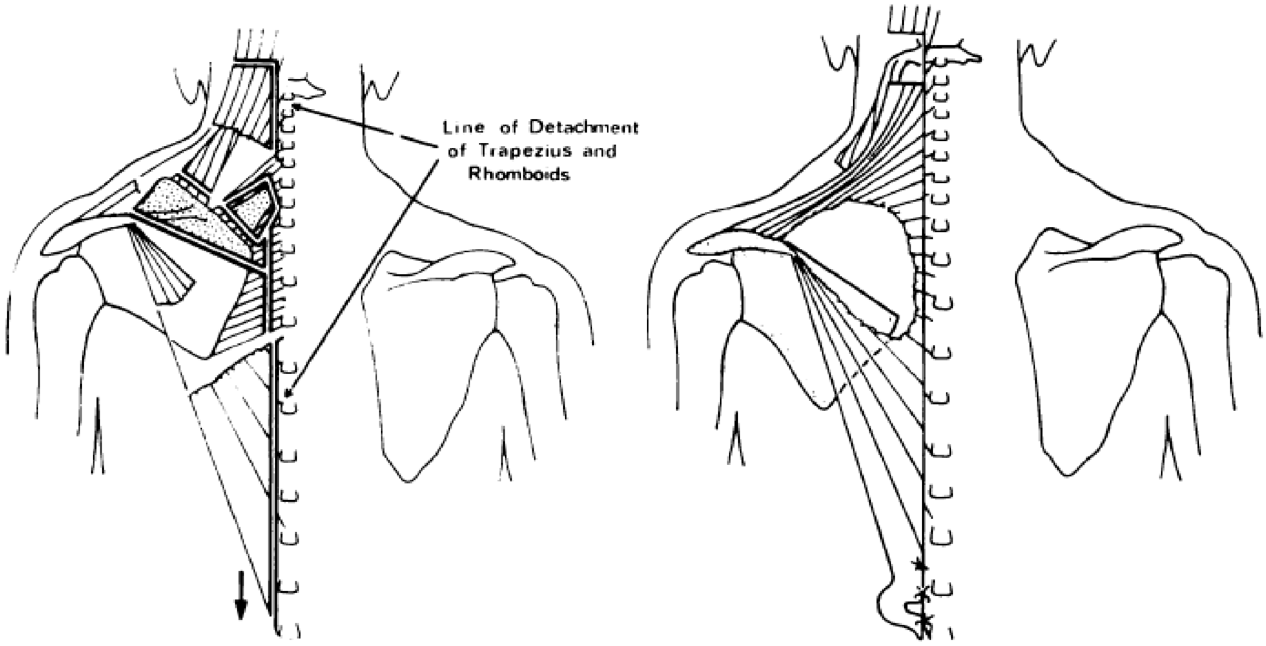


Figure A Figure B

Figure 62: Drawings to show Woodward type scapular transplantation. Stippled areas show excised bone. Figure A: Before trapezius slide. Figure B: Showing displacement of scapula after trapezius slide. [52]

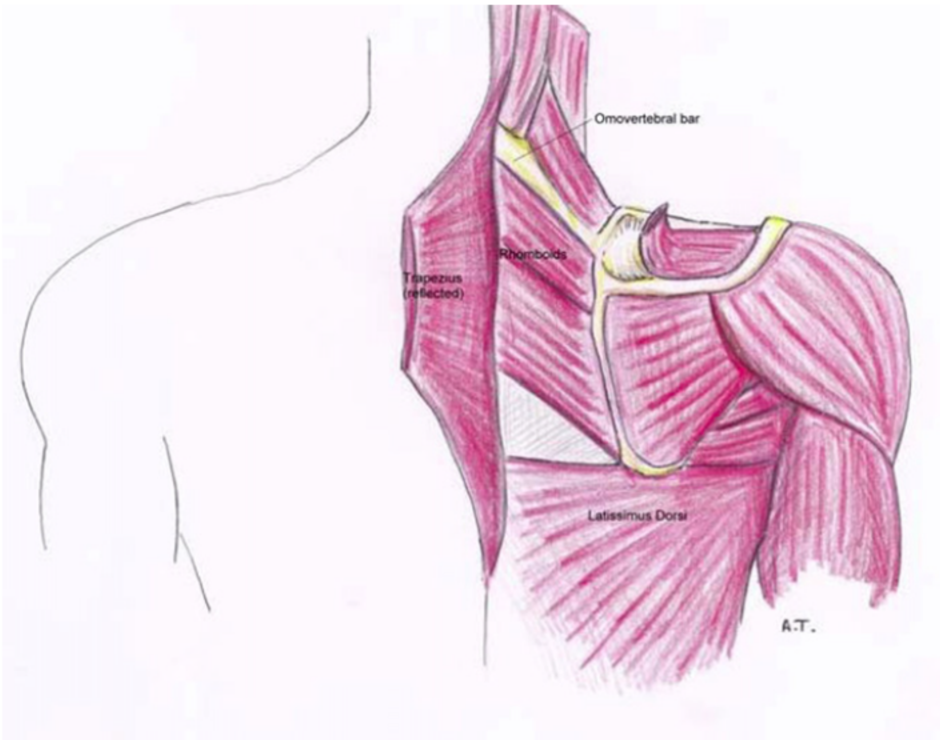


Figure 63: Posterior view of omovertebral bar [50].

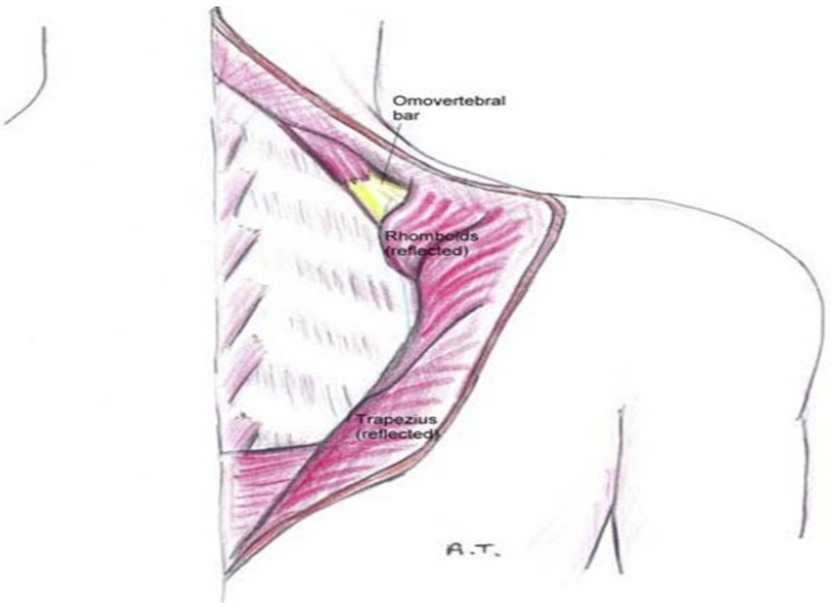


Figure 64: Medial dissection showing omovertebral bar [50].

❖ Scapular osteotomy:

A further option is a vertical scapular osteotomy which was first described in 1914 by König [70], a case report in which he performed a medial vertical osteotomy allowing a 4cm downward displacement. He fixed the body of the scapula in its new position by drilling a hole at its base, then looping a tube of latissimus dorsi through it and fixing it to itself. Later in the paper he considered this fixation was a mistake as it limited rotation of the bone and thus abduction of the arm. There were no further reports of the operation performed until that of Wilkinson and Campbell in 1980 [13]. They reported on vertical displacement osteotomy at the vertebral border of the scapula, finding it to be reliable and safe (see fig. 65).

Operation technique: 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 free 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 [21].

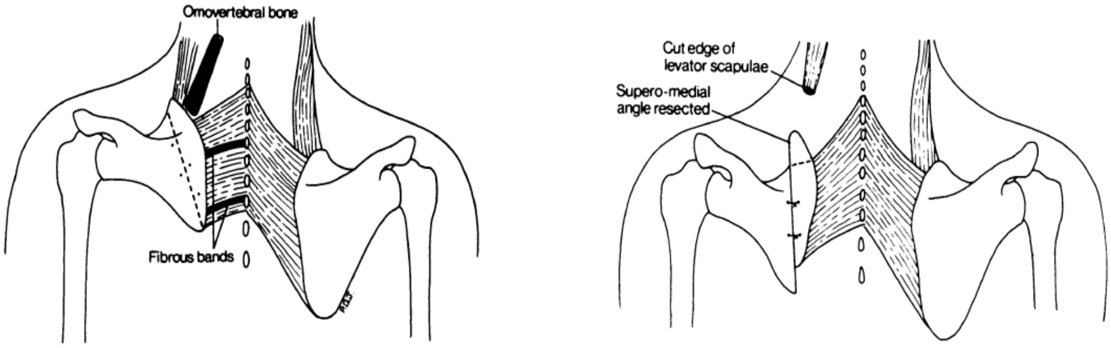


Fig A Fig B

Figure 65: (A) Fibrous bands are commonly found within the rhomboid muscles. In the absence of an omovertebral bone, a fibrous band may also be found attached to the superior angle of the scapula. The position of the osteotomy and offset drill holes are shown. (B)Position following displacement of the osteotomy, release of levator scapulae and division of fibrous bands. The redundant superior angle of the scapula has been resected. [22]

McMurtry et al [15], 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 [71] 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 scapula. 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.

Recently, Masquijo et al [75] reported statistically significant improvements in flexion and abduction in a prospective cohort study involving 21 patients treated with this technique ($P< 0.001$). Cosmetic 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 [73]. 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 [76] 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 [21].

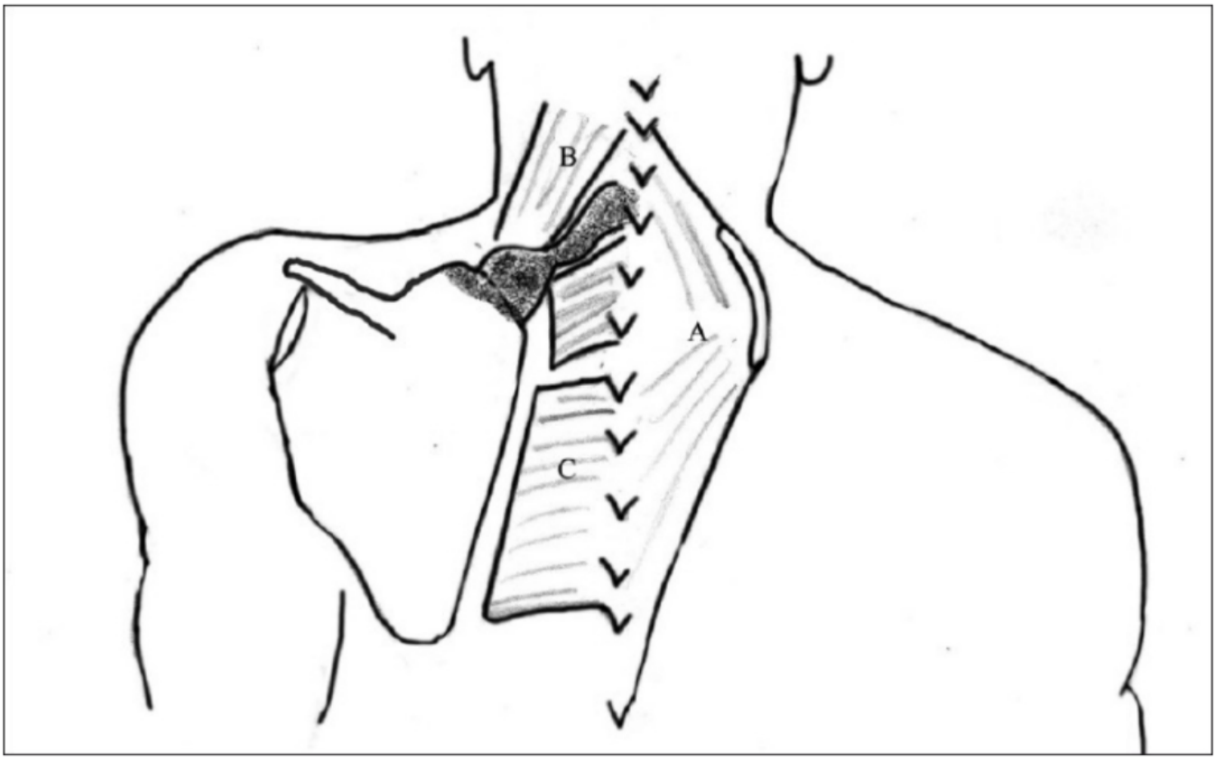


Figure 66: Diagrammatic representation of Mears' procedure. The shaded region represents the area to be osteotomized (A – Reflected trapezius; B – Rhomboids; C – Levator scapulae; T – The detached triceps) [52].

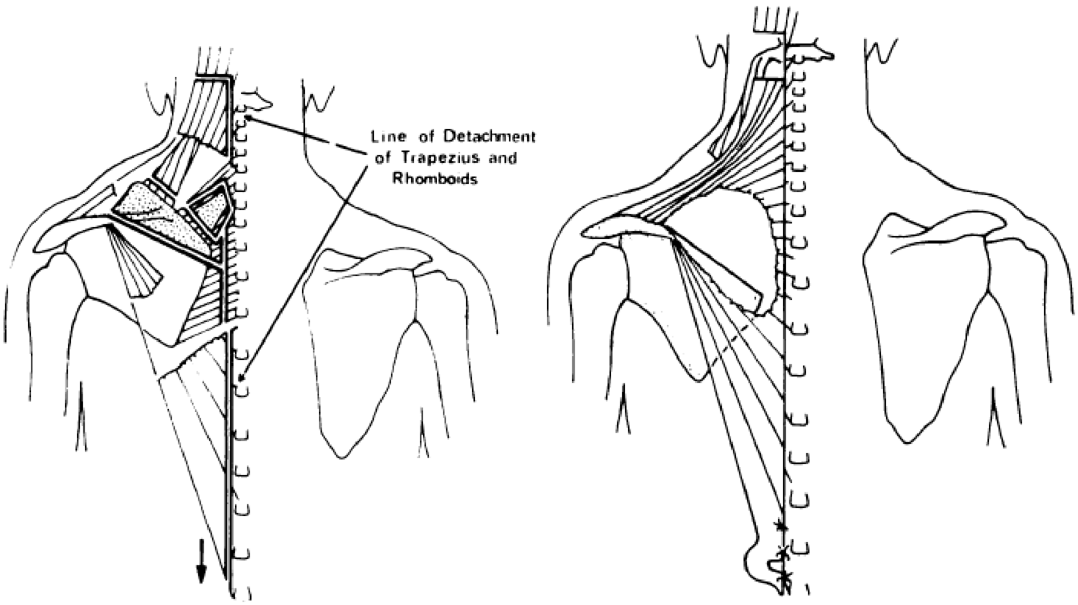


Figure A Figure B

Figure 66: (A) Drawing to show subtotal scapulectomy. Stippled areas show excised bone. (B) Drawing to show a “Putti-Schrock” type of scapular transplantation. Stippled areas show excised bone [24].

❖ Osteotomy of the clavicle (morcellation):

As an adjunct to any of the above procedures, it may be necessary to perform subperiosteal osteotomy of the clavicle, in severe cases and because of deformity of this bone, particularly at the sternoclavicular joint, to avoid compression of the brachial plexus or subclavian vein after inferiorly shifting the location of the scapula (Schrock 1926 [73]; Robinson, Braun, Mack and Zadek 1967 [78]) .

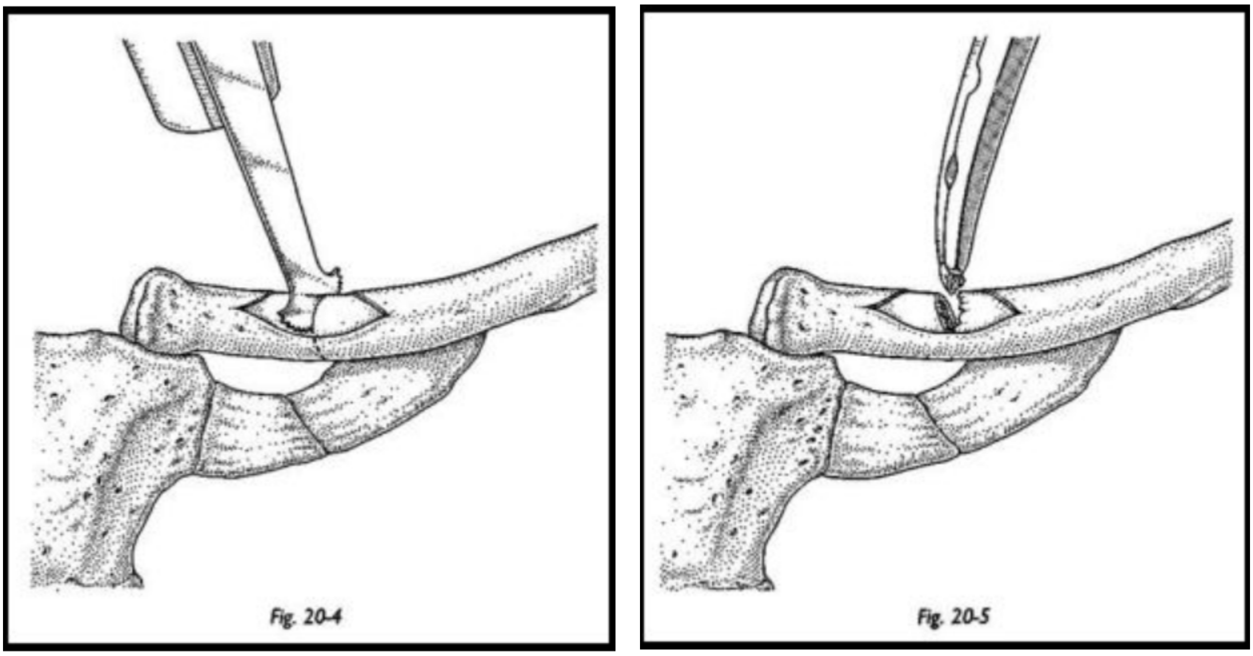


Figure 67: (A) With the patient lying supine on the operating table, a 2-cm transverse incision is made over the anterior aspect of the middle third of the clavicle. [79]

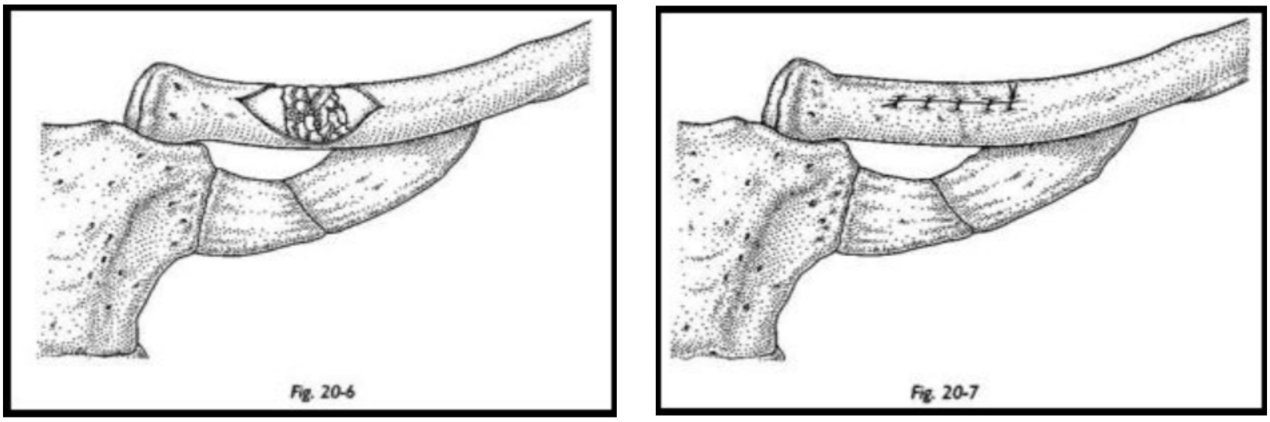


Figure 67: (B) The clavicle is exposed subperiosteally, and an oblique osteotomy made. The ends of the bone are then morcelized with bone nibblers, and the bone is left in situ the periosteal sleeve is then closed, and the -wound is sutured. [79]

e) Complications of surgical treatment:

The most frequently listed complications of surgical treatment of DS are: loss of correction and failure of functional improvement, protruding shoulders, paralysis of brachial plexus, compression of subclavicular artery, and hypertrophic/keloid scarring (in 30% of operated patients) [33].

The dorsal scapular nerve, spinal accessory nerve and suprascapular nerve can be injured during surgery. The dorsal scapular nerve runs close to the superomedial border of the scapula and can be injured during dissection of the periscapular muscles at the superomedial angle of the scapula. Boon paid attention to the risk of dorsal scapular nerve damage [80].

The spinal accessory nerve is located between the trapezius and rhomboid muscles and is, therefore, at risk theoretically; however, injury is uncommon when these muscles operate as a single unit. The suprascapular nerve runs in the suprascapular notch of the scapula and may be injured if the dissection is carried too far laterally when the superior portion of the scapula is resected. By staying at least one centimetre medial to the notch, the risk of injury can be reduced [1].

Although brachial plexus lesion is the most important complication cited in the literature, this complication did not occur in our patient and was not observed in any of adult cases in the literature.

The brachial plexus is at risk of compression intraoperatively due to scapular transfer, as it is migrated inferiorly and to the close vicinity of the clavicle to the neurovascular structure. Several authors have reported brachial plexus palsy after surgery for Sprengel deformity; however, many palsies were transient. In fact, children who are 8 years of age and older are known to be at risk for this complication due to the distorted and often fibrotic anatomy with very limited elasticity left.

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. In children, clavicular osteotomy was recommended by Andrault et al. as a ‘‘must’’ systematically if intraoperative monitoring is not available [72].

Following strategies are presented aiming in reduction the percentage of complication occurrence: application of longitudinal incision near midline, collar bone osteotomy, caracoid process osteotomy to reduce tension of pectoralis minor muscle, fasciodesis of inferior angle of the scapula .

Most surgical techniques involve large incisions and extensive dissection, which can lead to hypertrophic and keloid scars.

Other postoperative outcomes include scapular winging, resulting from incomplete reattachment of the serratus anterior muscle, regeneration of a bony omovertebral, regrowth of the superior pole of the scapula and recurrence of the deformity, as well as prominence of the sternoclavicular joint [49][52].

As SD can occur concurrently with KFS, any surgical intervention that implies scapular descent can increase the chance of operative vascular injury if a vascular anomaly is present. Thus, vascular assessment is imperative when surgical correction of SD is needed in a patient with concomitant KFS.

There is no constant correlation of anatomical and cosmetic results with recurrence of the deformity, age of the patient and functional outcome.

f) Postoperative management:

Early postoperative mobilisation is important to restore movement before adhesions form. Some authors reported that the shoulder is placed in a shoulder immobilizer for 6 weeks. Others reported that the shoulder is immobilized for 4 weeks. Bellemans and Lamoureux [81] described that gentle active and passive exercises are started 2 days after surgery, and the arm is held in a sling for 3 weeks. Ahmad [82] recommends immediate postoperative mobilization with physiotherapy.

Our patient Velpeau sling protection for tree week, followed by progressive rehabilitation of the operated shoulder.

g) Surgical outcomes:

Most of the good surgical outcomes have been reported for children. However, the surgical outcome for adult Sprengel's deformity rarely has been reported [21] [83].

Results are presented with different outcome measures, such as patient satisfaction, cosmetic, function, or radiologic classification, and involve various surgical methods;

The final appearance is dictated by the pre-operative state. The size, shape, and position of the omovertebral bone may predict improvement of shoulder motion after resection.

However, in our review of literature concerning SD in adolescent and adult patients, the surgical procedures most commonly used include excision of the omovertebral bone and the prominent superior angle of the scapula in 61% of all shoulders, and Woodward procedure in 22%.

Although surgery did not restore normal appearance or function because of the complexity of the abnormal anatomical structures in combination with other anomalies, shoulder abduction and cosmetic appearance has significantly improved; there was cosmetic improvement by at least one Cavendish grade in all shoulders, functionally, the improvement of abduction was excellent with a mean gain of 44°, the position of scapula improved by at least one Rigault grade and significant reduction in neck pain.

These results represent the eligible published data we used in this study concerning neglected SD. We found that surgery was successful in older children and in adults and at higher Cavendish grades.

The 61% of shoulders managed by simple bone excision, were represented in 6 studies:

_ Doita et al. (in 2000): the authors reported good results after surgical correction in 2 adults, following omovertebral bone excision for previously untreated Sprengel deformity, and they recommended surgery even in older patients.

*Technique: resection of an omovertebral bone and the medial supraspinous region of the scapula.

*Results: The first patient experienced significant reduction in her neck pain and a good cosmetic result because the bony prominence disappeared. However, this patient did not experience significant improvement in shoulder motion (shoulder abduction had only slightly improved from95° to 100°). The second patient experienced greater improvement in shoulder motion (the abduction had improved from 100° to 160°), a significant reduction in her neck pain, and a good cosmetic result because the scapula dropped to the level of the opposite side. The bony prominence also disappeared.

_ Fullbier et al. (in 2010): were the first to report a case of an adult patient with omovertebral bone, associated with unilateral Sprengel deformity and KFS, leading to cervical myelopathy.

They recommended isolated resection of the omovertebral bone as an adequate and safe procedure, feasible to treat older patients with good outcome.

*Technique: resection of the intraspinally located bony fragment, C-6 laminectomy, and partial resection of the omovertebral bone.

*Results: reduction in neck pain, improvement in neck motion, and a good cosmetic result but she had only slight improvement in gait ataxia and shoulder motion. Furthermore, the patient's urinary continence has resolved.

Until now, a case of an adult patient with omovertebral bone leading to cervical myelopathy has not been reported. Patients with such complex bone deformities usually become symptomatic during childhood; others may present later in life due to mild functional impairment or insufficient medical health care.

Early diagnosis and surgical treatment may prevent secondary neurological damage.

_Tedreko et al. (2013): presented a case an adult woman with multifocal spinal malformations who present with pain and range of motion limitation related to a large omovertebral bone.

*Technique: partial excision of the omovertebral bone (a fragment close to scapula was remained to save rhomboid muscle).

*Results: The patient showed improvement at the discharge from hospital after 3 weeks of rehabilitation; improvement in neck motion, reduction of neck pain and was satisfied with cosmetic result

_ Gillespie et al. (in 2013): illustrate in their case report, functional improvement following simple excision of an omovertebral bone in an adult without prior treatment for Sprengel deformity.

- Technique: simple excision of omovertebral bone
- Results: patient had greatly improved shoulder motion (abduction improved from 90° to 150°) and felt less pain.

_ Mirhosseini, Seyyed Ahmad et al. (2013): reported a 50-year-old woman with omovertebral bone, associated with unilateral Sprengel deformity and KFS, leading to cervical myelopathy. *Technique: resection of the intraspinally located bony fragment, C-5 laminectomy, and partial resection of the omovertebral bone.

*Results: the patient was neurologically unchanged, but she experienced a significant reduction in her neck pain, improved range motion of the neck, and the cosmetic result was good.

_ Dhir et al. (2014): described 9 patients treated for Sprengel deformity with a distinguishing pathoanatomic feature: partial endomuscular ossification of medial scapular suspension muscles (the cleithrum). Four of them were older than 15 years, Cavendish grade 4, and treated successfully by operation.

They demonstrated that simple excision of the bone permits greater scapular motion, improved scapular position, and enhanced comfort and function of the patient (mean abduction improved significantly from 97,5° to 142,5°) in the short term.

These results are concordant with what we achieved with our case report, attesting that excising the omovertebral and the superomedial part of the scapula is a safe and effective technique in patients with severe neglected Sprengel deformity.

Woodward procedure was performed on 3 shoulders (in 22% of all cases) with modest functional outcomes.

_H. O. Ong’ang’o et al. (2012): reported a case of 17 years old male with severe Sprengel deformity and omovertebral bone. He was managed by the Woodward’s procedure, but the results were uneventful.

_Gi Woon Yoon et al. (2013): report on a case of Sprengel’s deformity with a huge bilateral omovertebral bone and KFS. Surgery was performed because of discomfort and cosmetic problems according to the Woodward procedure. The patient experienced satisfying cosmetic result and moderate functional improvement.

CONCLUSION

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.

Sprengel disease is usually diagnosed and treated at developmental age.

In adult patients, resection of superiormedial part of scapula and omovertebral bone excision (if exist) is the surgical technique of choice and appears feasible at any age and in higher Cavendish grades of displacement.

By the reason of slenderness of clinical reports about treatment of Sprengel deformity in adult patients, it is difficult to scan natural course of the disease, especially in relation to omo-vertebral structures behaviour.

Early diagnosis and institution of appropriate therapy is believed to prevent fixation of incorrect movements and posture stereotypes and to allow for achievement of better therapeutic results especially within movements range of cervical spinal segment and upper limbs functions.

Beneficial, functional and cosmetically satisfactory effect of bone resection in case of our patient confirms observations of Doita and other authors, concerning such method choice in the treatment of adult patients, comparing to other techniques that include mobilization of the scapula.

SUMMARY

SUMMARY

Title: Management of Neglected Sprengel's deformity: case report and literature review.

Author: LAGHRICH Imane

Key words: Sprengel's deformity, Elevated scapula, Omovertebral bone, Surgical management.

Objectives: _ To Know the clinical and radiological aspect

_To draw the epidemiological profile of neglected Sprengel deformity

_To study the different therapeutic approaches and their results

Materials & Methods:

Our study about neglected Sprengel Deformity included 15 cases from a review of literature and a case report.

Results:

The mean age at management was 26,8 years, with a sex ratio Females/Males of 1,6. The left shoulder was affected in 13 patients and two cases had bilateral Sprengel deformity.

The main symptom was shoulders asymmetry, its severity was evaluated according to the Cavendish system; 15 patients were assigned to grade 4. The functional deficit was evaluated by shoulder abduction which was limited mostly to ≤100°. All patients reported neck pain.

Plain radiographs/ CT scans were used to identify the presence of omovertebral bone and other associated abnormalities, assess the severity of the deformity on Rigault scale and aid in surgical planning.

The management of neglected Sprengel deformity consisted on surgery; 11 shoulders (including our report), were treated by simple bone excision, involving resection of the elevated portion of the scapula and removal of omovertebral bone, four patients underwent Woodward procedure, and Mears technique was performed on one shoulder. Only two patients had nonoperative treatment.

Discussion:

Most patients, including our case report, were treated by Simple bone excision that showed good outcomes: improved scapular position, greater range motion of the neck and shoulder, with mean improvement of almost 50° in shoulder abduction, as well as satisfying cosmetic result. Neck pain was significantly reduced or disappeared. Other procedures showed uneventful results with moderate functional improvement.

Conclusion:

Simple bone excision is the treatment of choice in Neglected Sprengel deformity.

RESUME

Thèse: Prise en charge de la déformation de Sprengel négligée: à propos d'un cas et revue de littérature.

Auteur: LAGHRICH Imane

Mots clé : déformation de Sprengel, surélévation congénitale de l'omoplate, l'os omovertebral, traitement chirurgical.

Objectifs : _Connaître l'aspect clinique et radiologique

_Tracer le profil épidémiologique

_Étudier les approches thérapeutiques et leurs résultats chez les cas négligés

Matériels et méthodes :

Notre étude menée sur 16 patients :15 cas d'une revue de la littérature et à propos d'un cas.

Résultats :

L'âge moyen est de 26,8 ans, avec un sex-ratio F / H à 1,6. La localisation gauche retrouvée chez 13 patients. La bilatéralité retrouvée chez deux patients.

Le principal symptôme était l'asymétrie des épaules, évaluée selon le système de Cavendish ; 15 patients assignés au grade 4. Le déficit fonctionnel évalué par l'abduction de l'épaule souvent limitée à ≤100°. Tous les patients rapportaient des cervicalgies.

La radiologie utilisée pour visualiser l'os omovertébral et les malformations associées, évaluer la gravité sur l'échelle de Rigault et faciliter la planification chirurgicale.

11 épaules (y compris notre cas), traitées par simple excision osseuse : résection de la partie élevée de l'omoplate et l'ablation de l'os omovertébral, quatre patients par procédure de Woodward, et un par technique de Mears. Deux patients ont eu un traitement conservateur.

Discussion :

La plupart des patients, y compris notre cas, traités par une simple excision osseuse, rapportant de bons résultats ; l'amélioration de la position du scapula avec mobilité plus large, objectivé par l'amélioration du degré de l'abduction, avec un gain de presque 50°, avec résultat esthétique satisfaisant. La douleur était significativement réduite voir disparue. Les autres procédures ont montré des résultats modestes.

Conclusion :

La résection du bord supéro-medial de l'omoplate et de l'os omovertebral constitue le traitement de choix dans la déformation de Sprengel négligée.

الملخص

العنوان: التكفل بتَشَوُّه سبرينغل (ارتفاع الكتف الخلقي) المنسي بصدد حالة وبعض المراجع الطبية.

المؤلفة: الغريش إيمان

الكلمات ألأساسية : تَشَوُّهُ سبرينغل - ارتفاع الكتف الخلقي - العظم الكَتِفِي الفِقْوِي – العلاج الجراحي

الأهداف:

- معرفة المظهر السريري والإشعاعي
- رسم المظهر الوبائي لتَشَوُّه سبرينغل بتَشَوُّه سبرينغل المنسي
- دراسة مختلف المقاربات العلاجية ونتائجها.

الوسائل والمنهجيات: همت دراستنا 16 مريضا: 15 مريضا من محتلف المقالات الطبية وحالة واحدة.

النتائج:

بلغ متوسط الأعمار 26.8 سنة بنسة جنسية تقدر ب1.6. تموضع التشوه على اليسار عند 13 مريضا، وعلى الجانبين عند مريضين.

كان العرض الرئيسي لاتناظر الكتفين ، حيث قيمت وخامته بنظام كافانديش حيث سجلنا 15 مريضا بالدرجة الرابعة. أما التحدي الوظيفي فقد قيم بتبعيد الكتف الذي كان محدودا في غالب الحالات بأقل من 100°. عان جميع المرضى من آلام على مستوى العنق.

استعمل التصوير الإشعاعي المعياري والمفراس الثلاثي الأبعاد لتحديد وجود العظم الكتفي الفقري وتشوهات أخرى مصاحبة، ولتقييم وخامة التشوه حسب سلم ريغولت ولتسهيل التخطيط الجراحي.

يعتبر العلاج الجراحي الحجر الأساسي في التطفل بتشوه سبرينغل المنسي، فلقد عولجت 11 حالة (من بينها حالتنا) باستئصال عظمي بسيط يتضمن قطع الجزء المرتفع من عظم الكتف و استئصال العظم الكتفي الفقري. استفاد 4 مرضى من إجراء وودوارد، أما تقنية ميبرس فقط استعملت على كتف واحدة، بينما حصل مريضين على علاج محافظ.

المناقشة:

عولج معظم المرضى بما في ذلك حالتنا باستئصال عظمي بسيط، وقد أظهر ذلك نتائج جيدة خاصة تحسن تموضع الكتف بحركية أوسع للعنق والكتف، وقد سهم كل هذا في تحسن درجة تبعيد الكتف مع كسب ما يقارب 50 درجة، ونتائج جمالية سارة. نقص الإحساس بالألم أو اختفاؤه بشكل كبير. وقد أظهرت إجراءات أخرى نتائج متواضعة مع تحسن وظيفي طفيف.

الخلاصة:

يشكل قطع العلوي الإنسي للكتف و العظم الكتفي الفقري العلاج الأمثل لتَشَوُّه سبرينغل المنسي.

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Hippocratic Oath

At the time of being admitted as a member of the medical profession:

I solemnly promise that I will devote my life to serve humanity.

I will give to my teachers the respect and gratitude that is their due.

I will practice my profession with conscience and dignity.

The health of my patient will be my first consideration.

I will not betray the secrets that are confided in me.

I will maintain by all the means in my power, the honor and the noble traditions of the medical profession.

My colleagues will be my brothers.

I will not permit considerations of religion, nationality, race, party politics or social standing to intervene between my duty and my patient.

I will maintain the utmost respect for human life from the time of conception.

Even under threat, I will not use my medical knowledge contrary to the laws of humanity.

I make these promises solemnly, freely and upon my honor.

Serment d'Hippocrate

Au moment d'être admis à devenir membre de la profession médicale, je m'engage solennellement à consacrer ma vie au service de l'humanité.

Je traiterai mes maîtres avec le respect et la reconnaissance qui leur sont dus.

Je pratiquerai ma profession avec conscience et dignité. La santé de mes malades sera mon premier but.

Je ne trahirai pas les secrets qui me seront confiés.

Je maintiendrai par tous les moyens en mon pouvoir l'honneur et les nobles traditions de la profession médicale.

Les médecins seront mes frères.

Aucune considération de religion, de nationalité, de race, aucune considération politique et sociale ne s'interposera entre mon devoir et mon patient.

Je maintiendrai le respect de la vie humaine dès la conception.

Même sous la menace, je n'userai pas de mes connaissances médicales d'une façon contraire aux lois de l'humanité.

Je m'y engage librement et sur mon honneur

قسم أبقراط

بسم الله الرحمان الرحيم

أقسم بالله العظيم

في هذه اللحظة التي يتم فيها قبولي عضوا في المهنة الطبية أتعهد علانية:

بأن أكرس حياتي لخدمة الإنسانية.

وأن أحترم أساتذتي وأعترف لهم بالجميل الذي يستحقونه.

وأن أمارس مهنتي بوازع من ضميري وشرفي جاعلا صحة مريضي هدفي الأول.

وأن لا أفشي الأسرار المعهودة إلي.

وأن أحافظ بكل ما لدي من وسائل على الشرف والتقاليد النبيلة لمهنة الطب.

وأن أعتبر سائر الأطباء إخوة لي.

وأن أقوم بواجبي نحو مرضاي بدون أي اعتبار ديني أو وطني أو عرقي أو سياسي أو اجتماعي.

وأن أحافظ بكل حزم على احترام الحياة الإنسانية منذ نشأتها.

وأن لا أستعمل معلوماتي الطبية بطريق يضر بحقوق الإنسان مهما لاقيت من تهديد.

بكل هذا أتعهد عن كامل اختيار ومقسما بالله.

جامعة محمد الخامس – الرباط
كلية الطب والصيدلة بالرباط

سنـة : 2018 أطروحة رقم: 324

التكفل بتشوه سبرينغل المنسي:
بصدد حالة وبعض المراجع الطبية

أطروحة

قدمت ونوقشت علانية يوم :20 يوليوز 2018

من طرف

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لنيل شهادة الدكتوراه في الطب

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تحت إشراف اللجنة المكونة من الأساتذة

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