

## CASE REPORT

# Sprengel Deformity: Magnetic Resonance Imaging Findings in Two Pediatric Cases

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## ABSTRACT

The characteristics of Sprengel deformity, which is also called congenital high scapula, are malposition and dysplasia of the affected scapula, with possible omovertebral connection. The aim of the present study was mainly to present the magnetic resonance imaging (MRI) findings of two pediatric cases of Sprengel deformity. A 7-year-old girl and a 9-year-old boy with deformities in their right shoulder were studied. Plain radiographs were obtained. MRI was performed for both children. The fibrous omovertebral connection is depicted in its longest form in one plane. Omovertebral band is best screened in coronal and axial cross sections. We are introducing a new MRI sign which we named as "Ra's eye" to define the appearance of omovertebral band within the surrounding fat tissue.

**Key words:** Magnetic resonance imaging, omovertebral band, sprengel deformity

## INTRODUCTION

Sprengel deformity (SD), also called congenital elevated (or high) scapula or undescended scapula, is the most frequently seen congenital anomaly of the shoulder girdle that happens as a result of the failure in the scapula's caudal migration during early intrauterine life. This condition also involves regional muscle hypoplasia or atrophy that causes a misshaped shoulder and limits shoulder movement.<sup>[1]</sup>

The first three cases related to this condition were published in 1863 by Eulenberg with the title "hochgradige Dislocation der Scapula" (i.e., a high grade dislocation of the scapula). Sprengel described four cases of "Die angeborene Verschiebung des Schulterblattes nach oben" (congenital displacement of the scapula upward) in 1891 and suggested a theory for its formation mechanism,

relating this anomaly to high amniotic fluid pressure.<sup>[2]</sup> Later on, this clinical entity was named after him.

## CASE REPORTS

## Case 1

A 7-year-old girl was presented with a deformity of the right shoulder. The right scapula was small and elevated on physical examination but there was no impaired movement of the shoulder [Figure 1]. There was increased horizontal to vertical length of the scapula and no omovertebral bony connection was observed in the plain radiographs [Figure 2a].

Therefore, this anomaly was evaluated with magnetic resonance imaging (MRI). MRI showed a thin hypointense band extending from the superomedial angle of the

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scapula to the 7th cervical vertebra's spinous process [Figures 2b, c, and 3]. In this patient, we observed a new MRI sign that we have named as "Ra's eye" to define the appearance of omovertebral band within the surrounding fat tissue [Figure 3c]. The case was classified as grade 1 according to the Cavendish classification [Table 1] and hence we inferred that the patient did not need a surgery to correct this minor cosmetic deformity. Further, there were minimal Bifid costa, scoliosis, segmentation anomalies and spina bifida in thoracic spine were the accompanying pathologies.

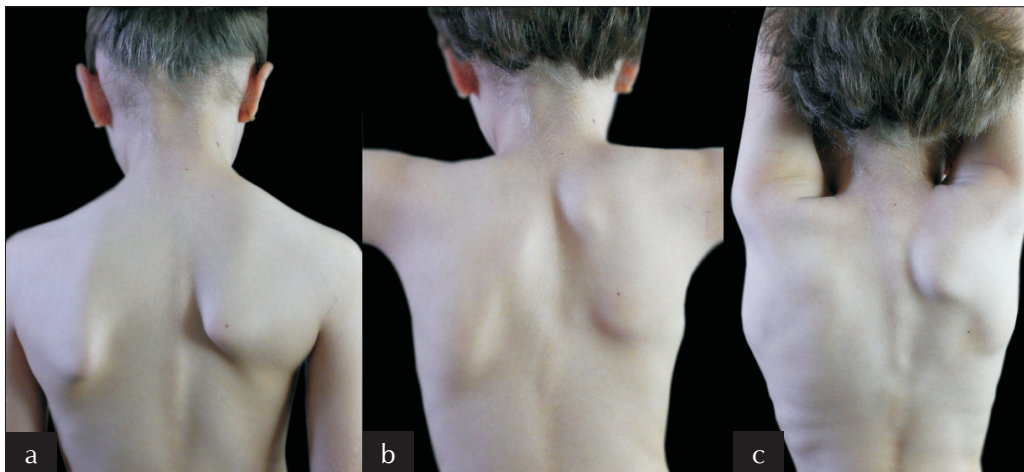
### Case 2

A 9-year-old boy presented with a deformity of the right shoulder. The right scapula was found to be raised and fixed medially on physical examination [Figures 4]. There was partially restricted movement of the shoulder joint. Three dimensional computed tomography (3D CT) examination revealed abnormality, anterior curving of the supraspinous

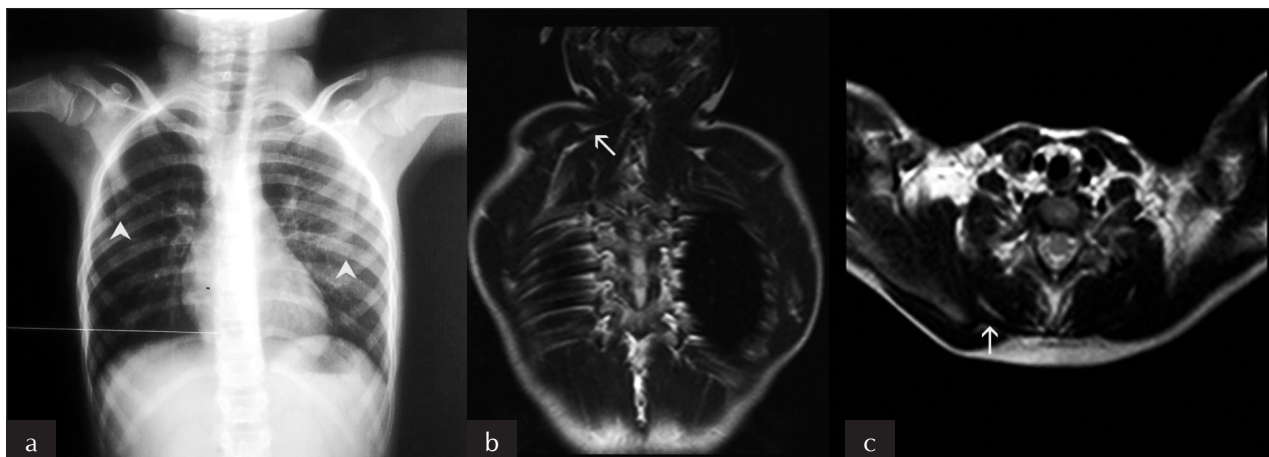
portion of the raised right scapula. The horizontal and vertical lengths were almost the same in both scapulas. There was no omovertebral bone. In the MRI, there was a thick and short hypointense band seen between the superomedial angle of the scapula and the posterior components of the 7<sup>th</sup> cervical vertebra [Figure 5]. The omovertebral band was confirmed with ultrasonography (US) [Figure 6]. The case was classified as grade 2 according to the Cavendish classification [Table 1]. The patient

**Table 1: Cavendish classification**

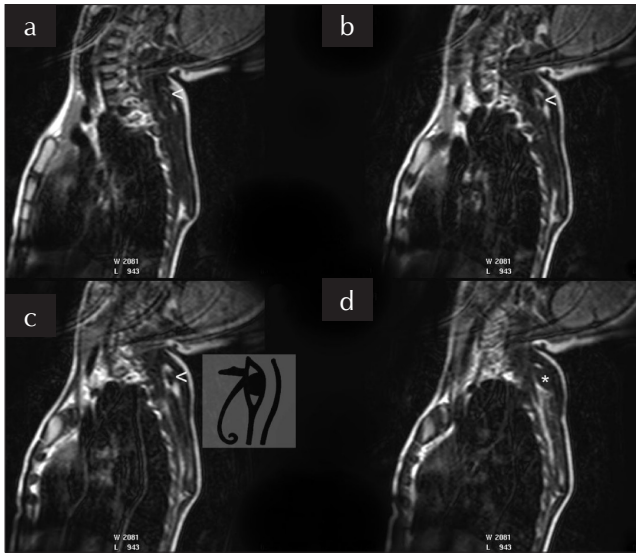
Grade	Features
Grade 1	Very mild deformity is observed. When covered with clothes the deformity is almost invisible.
Grade 2	The deformity is still mild, but appears as bump. The superomedial portion of the high scapula is convex, forming a bump.
Grade 3	Moderate deformity with 2-5 cm visible elevation of the affected shoulder compared to normal one
Grade 4	Severe deformity with > 5 cm elevation of the affected shoulder accompanied by neck webbing.



**Figure 1:** A 7-year-old girl showing Sprengel deformity on the right shoulder (a, b, c) The right scapula is raised, shorter, and larger than the contralateral one. There is no restriction in the right shoulder movements.



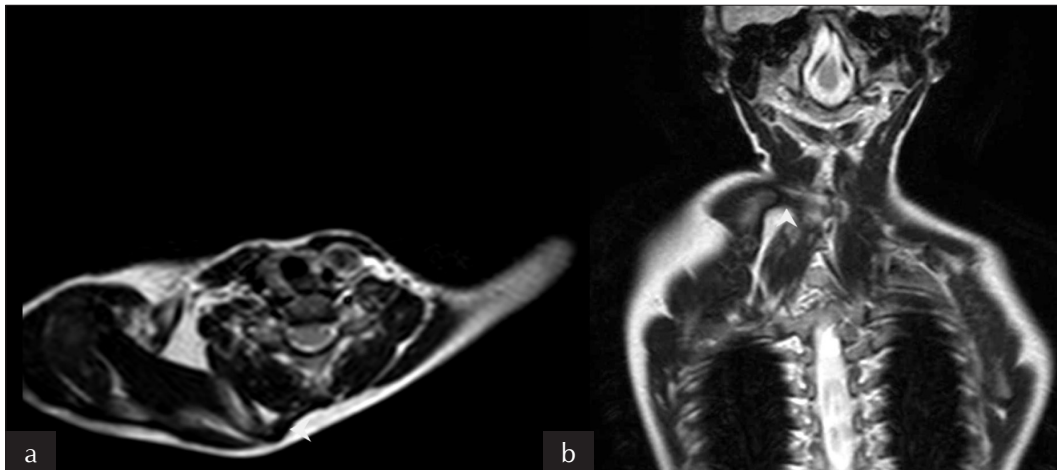
**Figure 2:** (a) Plain radiograph shows increased horizontal/vertical length of the right scapula. There is no omovertebral bony connection. (b) Coronal and (c) axial magnetic resonance (MR) images of the right scapula.



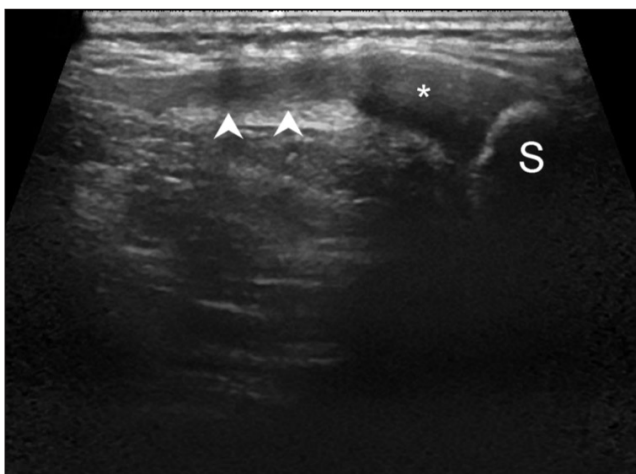
**Figure 3:** Sagittal MR image series (a, b, c, d) demonstrate the omovertebral band. (c) Omovertebral band and peripheral fat tissue both look like an eye. \*Points to the superomedial angle of the right.



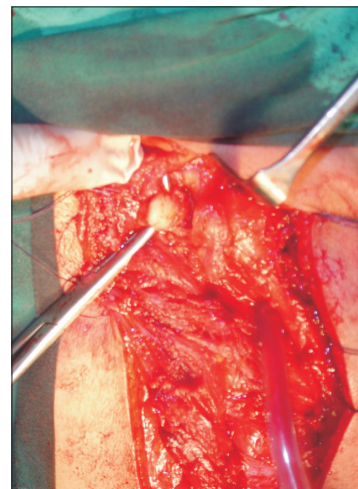
**Figure 4:** A 9-year-old boy showing Sprengel deformity on the right side. Right scapula is raised and neck appears plump and shorter on the same side.



**Figure 5:** (a) Axial and (b) coronal MR images of the right scapula show a thick and short hypointense OB extending from the superomedial angle of the scapula to the 7th cervical vertebra's spinous process.



**Figure 6:** Ultrasound image of the omovertebral band. Arrowheads point to the fibrous component and \* points the cartilaginous component. S: Scapula.



**Figure 7:** Photograph of the omovertebral band as seen during the surgical (modified "Green") procedure.



underwent surgery and the fibrous omovertebral band was resected [Figure 7]. Scoliosis, segmentation anomalies, and spina bifida in thoracic spine and bifid costa were the accompanying pathologies.

## DISCUSSION

Normally the scapula lies between the 2<sup>nd</sup> and the 7<sup>th</sup>-8<sup>th</sup> thoracic vertebrae on the posterior thoracic wall. The scapula differentiates, at about five weeks of the gestation, opposite the inferior cervical vertebrae. This anomaly occurs at about 9 to 12 weeks of fetal development, if the scapula does not migrate from the neck to its usual thoracic position.<sup>[3-5]</sup>

The characteristics of SD are malposition and dysplasia of the affected scapula with omovertebral connection. We see varying degrees of scapula elevation (unilateral or may even be bilateral) and medial rotation of the inferior pole of the scapula that makes the glenoid face downward. This situation results in limited movement of the shoulder joint, especially limited abduction. On physical examination, the neck appears plump and shorter on the affected side. There is a decreased height to width ratio of the dysplastic scapula such that it is larger than the contralateral one.<sup>[2]</sup> Omovertebral connection is seen in approximately one fourth or half of the cases.<sup>[6,7]</sup> The omovertebral connection types are fibrous tissue, bony and/or cartilaginous. Willet, in 1880, and Walsham, in 1883, were the earliest to mention the omovertebral bone in two cases with this clinical entity.<sup>[2]</sup> Omovertebral band extends from the superomedial border of the scapula to the spinous processes, lamina, or transverse processes of the lower cervical spine, most commonly of the 4<sup>th</sup> to 7<sup>th</sup> cervical vertebrae.

The condition is almost always sporadic, and rarely does it run in families with an autosomal dominant pattern of inheritance.<sup>[4]</sup> The male to female ratio is 1:3.<sup>[3]</sup> It is generally unilateral. Congenital anomalies of other systems and several other syndromes may often accompany this condition.<sup>[2,8]</sup> Klippel-Feil Syndrome (KFS) goes with scoliosis and cervical vertebral defects. SD has been found in 7–42% of the patients with KFS.<sup>[9]</sup>

The Cavendish classification is currently being used for determining the need for surgery.<sup>[10]</sup> A number of screening techniques can be used in SD. Intrauterine diagnosis can be obtained with prenatal US, so families can be prepared for any cosmetic and functional complications that come with SD.<sup>[8]</sup> If surgery is planned to correct the cosmetic deformity

and functional impairment, there are various surgical methods advocated by different authors. Currently, two of the most used methods in surgery are the “Woodward” procedure and the “modified-Green” procedure.<sup>[4,5]</sup>

The disorder should be evaluated primarily with plain radiography and CT. Malposition, dysplasia, and points of the tethering of the scapula with bony omovertebral connection can be examined best with 3D CT.<sup>[1]</sup>

In our cases, there was no omovertebral bone on plain films or on CT scans so we evaluated omovertebral connection by MRI. 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. We screened omovertebral band best in coronal and axial cross-sections. In the sagittal images, we named the appearance of omovertebral band with peripheral fat tissue as “Ra’s eye” [Figure 3c]. We believe that in the future, omovertebral band will be evaluated routinely with MRI in addition to other imaging procedures to help doctors plan treatment options and various operational procedures. In conclusion, omovertebral band needs to be routinely evaluated radiologically in patients Sprengel deformity.

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