

## IMAGING CASES

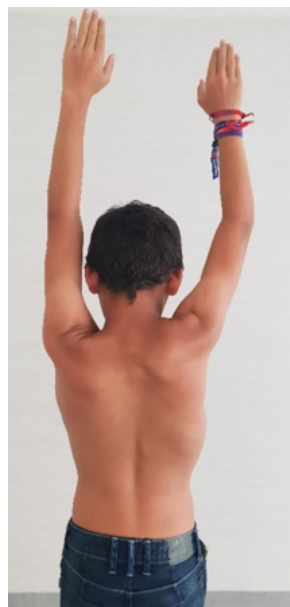
### MISALIGNED SHOULDERS

#### OMBROS DESALINHADOS

Íris Santos Silva<sup>1</sup>, Aníbal Correia<sup>2</sup>, Rita Santos<sup>3</sup>, Catarina Aleixo<sup>4</sup>, Mafalda Santos<sup>4</sup>

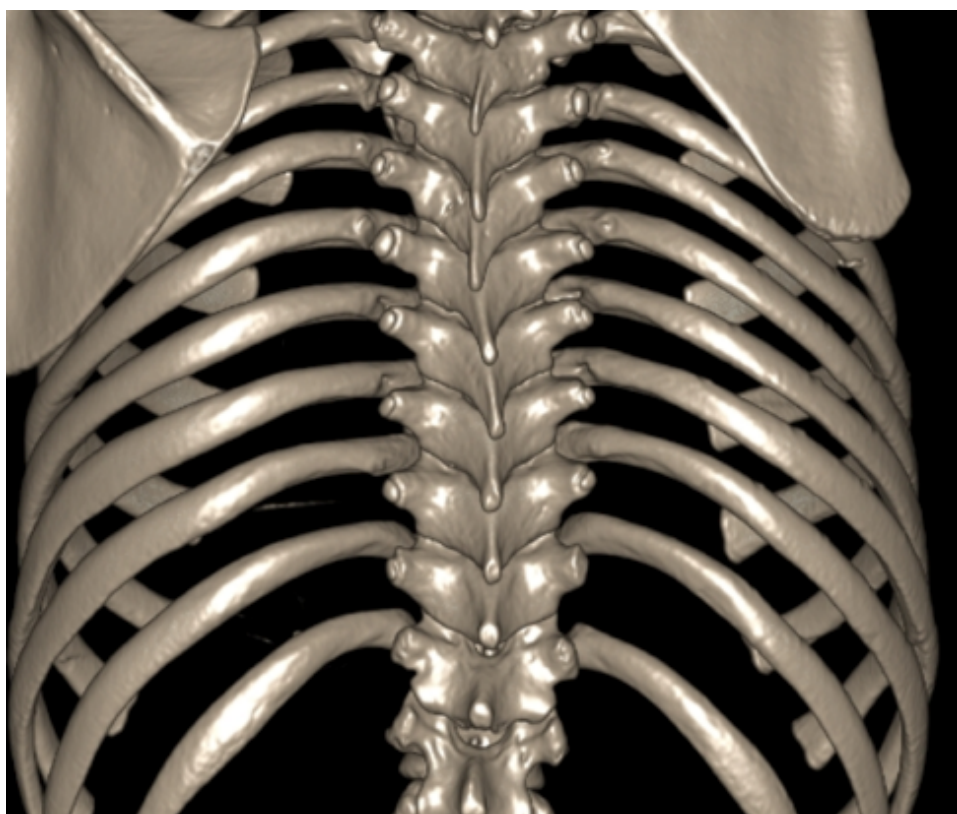
A nine-year-old boy without relevant past medical history was referred to the Orthopedic Department due to shoulder asymmetry. He denied pain, other symptoms, or functional limitation in daily activities. On physical examination, the right scapula was at a higher level compared with the left and slightly limited abduction was noted on the right shoulder. Spine x-ray was performed, revealing mild scoliosis, and computed tomography (CT) scan revealed the upper location of the right scapula with ipsilateral clavicle verticalization.

**What is your diagnosis?**



**Figure 1 and Figure 2** - Asymmetry in the position of scapular poles, with slightly limited abduction on the right shoulder.

1. Department of Pediatrics, Hospital Sousa Martins, Unidade de Saúde Local da Guarda. 6300-035 Guarda, Portugal. irissantossilva.92@gmail.com
2. Department of Pediatrics, Unidade 2, Centro Hospitalar Vila Nova de Gaia/Espinho. 4400-129 Vila Nova de Gaia, Portugal. anibalalvescorreia@gmail.com
3. Department of Orthopedic, Hospital Distrital de Santarém. 2005-177 Santarém, Portugal. rita.santos.d@gmail.com
4. Department of Orthopedic, Unidade 2, Centro Hospitalar Vila Nova de Gaia/Espinho. 4400-129 Vila Nova de Gaia, Portugal. catarinabpaleixo@gmail.com; mafalda.orto@gmail.com



**Figure 3** - 3D CT Scan

## DIAGNOSIS

Sprengel Deformity

## ORIENTATION

Surgical correction was not performed, since the boy had no functional limitation. Two annual follow-ups were conducted, with no changes in signs or symptoms.

## DISCUSSION

Sprengel deformity is a congenital elevation of the scapula. Although rare, it is described as the most common congenital abnormality of the scapula.<sup>1-3</sup> It has a male predominance (3:1) and may be associated with congenital anomalies in the family.<sup>3</sup> The condition can occur on both sides concurrently, but is most often unilateral, with a predilection for the left side.<sup>4</sup>

The scapula develops embryologically during the fifth week in the upper dorsal/lower cervical region and descends up to the final anatomical position by 8-12 weeks of gestation. The pathology in

Sprengel's deformity, which sits the bone 2-10 cm higher, probably represents a continuation of the scapula fetal form joined by failure in descending. Maintenance of a high scapula position can lead to other musculoskeletal defects, such as hypoplasia, medialization, adduction and prominence of the upper angle of the scapula; distal rotation and lateral angulation of glenoid cavity; changes in the position of the clavicle, cervicothoracic vertebrae and ribs; and atrophy of the shoulder musculature.<sup>5,6</sup> Moreover, other pathoanatomical anomalies can be found, such as omovertebral bone or costovertebral defects (spina bifida and kyphoscoliosis).<sup>5,7</sup>

Some syndromes are associated with Sprengel's deformity, the most common being Klippel-Feil Syndrome, present in about 19–27% of patients with Sprengel's deformity. Other linked syndromes include teratological conditions, such as anencephaly (a triad of occipital defect, spina bifida of cervical vertebrae, and fixed retroflexion of the head), X-linked dominant hydrocephalus, mental disturbance syndrome, and diastematomyelia.<sup>4,8</sup> In this case report, no evidence of any syndrome was found.

Among differential diagnoses, scapular winging should be considered, in which scapula alignment compared side to side for asymmetry reveals a declination of the affected shoulder, being a painful and debilitating condition. The most common reported causes are dysfunction of the serratus anterior or dysfunction of the

trapezius.<sup>9</sup>

Treatment can be conservative or surgical. Surgery is considered in patients with severe limitation of scapular function, neck pain, or to improve cosmesis.<sup>5,8</sup> Surgery should be performed between the ages of three and eight years, due to risk of nerve impairment below the age of eight. Surgical procedures involve a combination of scapular lowering with displacement of the origin or insertion of the scapular muscles on the spine/scapula and resection of the superomedial border. Associated congenital anomalies may be a contraindication to surgery.<sup>5</sup> Since the present child had no symptoms, surgical correction was not performed. The patients had two annual follow-ups, during which no changes in signs or symptoms were noted.

## ABSTRACT

Sprengel deformity is a congenital elevation of the scapula. Herein is reported the case of a child with the right scapula at a higher level compared to the left and limited abduction of the right shoulder. Other pathoanatomical anomalies can be associated. Treatment is usually conservative, since surgery is only considered in patients with severe scapular function limitation, neck pain, or to improve cosmesis.

**Keywords:** conservative treatment; elevation of the scapula; Sprengel deformity

## RESUMO

A deformidade de Sprengel é uma elevação congénita da omoplata. É descrito o caso de uma criança com a omoplata direita num nível mais elevado do que a esquerda e com abdução limitada do ombro direito. Outras anomalias ou síndromes podem ser associadas a esta deformidade. O tratamento é geralmente conservador, uma vez que a cirurgia é reservada a doentes com severa limitação funcional, dor no pescoço ou para melhoria estética.

**Palavras-Chave:** deformidade de Sprengel; elevação da omoplata; tratamento conservador

## REFERENCES

1. Harvey EJ, Bernstein M, Desy NM, Saran N, Ouellet JA., Sprengel deformity: Pathogenesis and management. *J Am Acad Orthop Surg.* 2012; 20:177–86.
2. Grogan DP, Stanley EA, Bobechko WP, The congenital undescended scapula: Surgical correction by the Woodward procedure. *J Bone Joint Surg Br* 1983; 65:598-605.
3. Durrani MYK, Sohail AH, Khan I, Hashmi P. Sprengel's deformity.

*J Ayub Med Coll Abbottabad* 2018; 30:135–7.

4. Kariki E, Vasiliadis K, Tsitouridis I, Bindoudi A., The Rare Sprengel Deformity: Our Experience with Three Cases. *Journal of Clinical Imaging Science*, 2014, 4:55.
5. Kadavkolan AS, Bhatia DN, DasGupta B, Bhosale PB. Sprengelen deformity of the shoulder: Current perspectives in management. *Int J Shoulder Surg.* 2011; 5:1–8.
6. Gonen E, Simsek U, Solak S, Bektaser B, Ates Y, Aydin E. Long-term results of modified green method in sprengel's deformity. *J Child Orthop.* 2010; 4:309–14.
7. Tsirikos AI, McMaster MJ. Congenital anomalies of the ribs and chest wall associated with congenital deformities of the spine. *J Bone Joint Surg Am.* 2005; 87:2523–36.
8. Rashid M. Sprengel's Deformity. A Review Of The Literature. *Internet J Orthop Surg* 2009; 15:1-6.
9. Didesch JT, Tang P. Anatomy, Etiology, and Management of Scapular Winging. *J Hand Surg Am.* 2019; 44:321-30.

## CORRESPONDENCE TO

Íris Santos Silva  
Department of Pediatrics  
Hospital Sousa Martins  
Unidade de Saúde Local da Guarda  
Avenida Rainha Dona Amélia 19,  
6300-035 Guarda  
Email: iris.silva@ulsguarda.min-saude.pt

Received for publication: 20.12.19

Accepted in revised form: 07.01.21