

Sprengel's Syndrome

INTRODUCTION

Your child has been diagnosed with Sprengel's Syndrome, which is also called Sprengel's deformity. This condition is diagnosed when one shoulder blade (scapula) sits higher on the back. Most of the time, the elevation is mild and creates a mild asymmetric appearance to the back. This asymmetry to the back is often picked up in a scoliosis screening examination and the patient is sent for scoliosis evaluation. Careful examination will determine the diagnosis. Function is normal and the cosmetic issues are minimal. Treatment is usually not needed.

BACKGROUND

The scapula develops early in fetal development high in the neck and then moves downward with a muscular envelope onto the chest wall. Usually, the movement is symmetric on the right and left sides, but not always. Asymmetric positioning of the scapula is called congenital elevation of the scapula or more commonly Sprengel's deformity. If the Sprengel's deformity is an isolated condition, the function of the shoulder is normal. The cosmetic aspect of a mildly elevated shoulder is usually insignificant. If the scapula sits very high, there can be cosmetic issues. If there are significant cosmetic issues, surgery can be done to move the scapula down.



The etiology of Sprengel's Syndrome remains unknown, but there is a presumed arrest in fetal development and motion of the scapula, which can be associated with anomalies of the cervical spine and ribs. Physical examination and some time xrays of the upper thoracic spine and neck will evaluate for these problems.

CLINICAL FINDINGS AND DIAGNOSIS

Physical examination reveals scapular position asymmetry with fullness at the base of the neck on the involved side. The spinal exam will otherwise be normal. Shoulder motion should be normal. Radiographs of the spine and chest allow evaluation and confirmation of the Sprengel's deformity. The xrays will also look for congenital anomalies of the vertebrae and ribs. If there are abnormalities of neck motion, radiographs of the cervical spine may be done to look for congenital anomalies of the cervical spine.

TREATMENT

Generally no treatment is needed as shoulder function is normal and cosmetic issues are minimal. When the appearance of the scapula is unacceptable, it can be improved by removal of upper part of the scapula or by a Woodward procedure which move the scapula down to the level of the opposite scapula. This is a fairly involved procedure and is done for only when there are significant cosmetic issues.



MORE INFORMATION

Further information can be obtained on the internet. Two good sites for expert and peer reviewed information are the American Academy of Orthopedic Surgeons at www.aaos.org and the Pediatric Orthopedic Society of North America at www.orthokids.org.