Sprengel Deformity
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History
Child with scoliosis.

Diagnosis
Sprengel Deformity

Discussion
Sprengel deformity is a congenital upward and forward displacement of one or both scapulae, which may be up to 12 cm higher than normal. A fibrous band or an "omovertebral bone" connects the scapula to either the C5 or C6 vertebrae. The shoulder girdle is usually abnormal with an absent or weak trapezius muscle, as well as hypoplastic or fibrosed rhomboid and levator scapulae muscles. Clinically, the patient’s neck is thicker and shorter than normal. Klippel-Feil syndrome, congenital spine anomalies (scoliosis, segmentation defects, dysraphism, tethered cord, syrinx and diastematomyelia), situs inversus, genitourinary anomalies and mandibulofacial dystosis may be associated with Sprengel deformity. According to Dave et al., 20-30 % of patients have associated genitourinary tract anomalies, 10% have congenital heart disease and 5 -10 % have spinal dysraphism including diastematomyelia, tethered chord and fibrous bands.

Findings
Elevation and rotation of the left scapula with fibrous attachment to the spinous processes of C5 and C6.

Reference

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