SPRENGEL SHOULDER

Sprengel shoulder is also called as high scapula or scapula elevata. It is a congenital abnormality of the shoulder girdle characterized by an abnormally raised scapula on one side or both sides. The movement of the scapula is marked by limitation due to fibrous bands or a bony bar, as the muscles of the scapula are poorly developed. The reported range of displacement of the shoulder blade is 2-10 centimeters. It is named after Otto Gerhard Karl Sprengel (1852-1915) , a German surgeon who described four cases in 1891.

SIGNS AND SYMPTOMS :

- Restricted ROM of shoulder and arm on the affected side.
- Limitation of shoulder abduction and elevation.
- Limited or restricted movement of the cervical spine.
- Neck deformities like mild tilting (torticollis) to severe spine deformity.
- Asymmetry in the shoulder alignment.
- An elevated shoulder blade may cause a lump in the base of the neck.
- Underdeveloped muscles in the surrounding area.

CAUSES :

- The exact underlying cause is unknown. Researchers believe that the disorder occurs early during fetal development. During the third month of pregnancy, the shoulder blade moves or 'migrates' down to its normal position. This migration fails to occur in case of sprengel deformity. The developing shoulder blade remains too high and often fails to fully form.

- Sprengel deformity may be associated with Klippel-Feil syndrome, clavicular abnormalities, rib abnormalities, scoliosis, spina bifida, hemivertebrae, underdevelopment (hypoplasia) of neck or shoulder muscles.

CLASSIFICATION:

The Cavendish classification of grading:

Grade I - Very mild deformity is observed when covered with clothes the deformity is almost invisible.
Grade II - The deformity is still mild but appears as a bump the superomedial portion of the high scapula is convex, forming a bump.

Grade III - Moderate deformity with 2-5 cm of visible elevation of the affected shoulder.

Grade IV - Severe deformity with >5 cm elevation of the affected shoulder, accompanied by neck webbing.

DIAGNOSIS:

X-ray: The x-ray shows bony and cartilage deformity or abnormality.

Computed tomography (CT): Computed tomography (CT) scans are used to identify associated abnormalities like scoliosis, cervical and scapular abnormalities.

Magnetic resonance imaging (MRI): Magnetic resonance imaging (MRI) helps to identify any bony, cartilaginous, or muscular weakness or defects.

TREATMENT:

CONSERVATIVE: Very little improvement can be offered by way of corrective exercises. The children should be encouraged to participate in sports such as swimming to maintain ROM.

SURGICAL: Moderate to severe cases require surgical approach. It consists of excision of the fibrous band or bony bar. Woodward techniques and Green are the most commonly used procedures. It involve removal of the protruding portion of the scapula and omovertebral bone as well as translation of the scapula inferiorly to a more downward position.

PHYSIOTHERAPY AFTER SURGERY:

- Thermotherapy should be used to relax the muscles before performing the exercise program.
- Suitable pain reducing modality like TENS and hydrocollator pack may be used to reduce pain.
- Gradual relaxed PROM exercises should be applied to shoulder joint.
- Special attention should be given to early mobility of scapula and shoulder abduction and elevation.
- Overall mobilization of shoulder girdle and affected joints should be done.
- Strengthening of all affected muscles.
- Maximum possible correction of posture and its maintenance is important.