

controlled study to settle the important issue.

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Sprengel's Deformity

Sprengel's deformity is a rare congenital anomaly of the shoulder resulting from inadequate caudal movement of scapula during development(1). The disorder occurs almost always as a sporadic event even though a few familial cases with possible autosomal dominant inheritance have been reported. The deformity produces cosmetic and functional impairment. We report a case of unilateral Sprengel's deformity associated with thoracic and lumbar vertebral anomalies in a 3-year-old boy.

A 3-year-old boy was seen by us for abnormal position of neck. He was born to non-consanguineous couple and his birth and attainment of developmental milestones were normal. He weighed 11 kg and his height was 80 cm (both less than 5th percentile on NCHS charts). His neck was

short and extension and rotation movements were restricted. He held his head always flexed. Abduction at left shoulder was only of 120°. The left shoulder was at a higher level. Roentgenological examination revealed abnormally highly situated, hypoplastic left scapula (*Fig. 1*). Its superior angle was rotated towards the vertebral column. There were hemivertebrae at T₁ T₃, L₃ and crowding of ribs on the left side.

Sprengel's deformity presumably results when there is interference with the descent of the scapula from the neck to the thorax(1). Normally, at about five weeks the fetal scapulae differentiate opposite sixth vertebrae. It undergoes caudal migration to its usual thoracic position between 9-12 weeks. The shape of scapula also undergoes change as the initial horizontal diameter which exceeds the vertical, starts decreasing until the mature dimensions are reached. In case of Sprengel's deformity, there is not only failure of caudal migra-

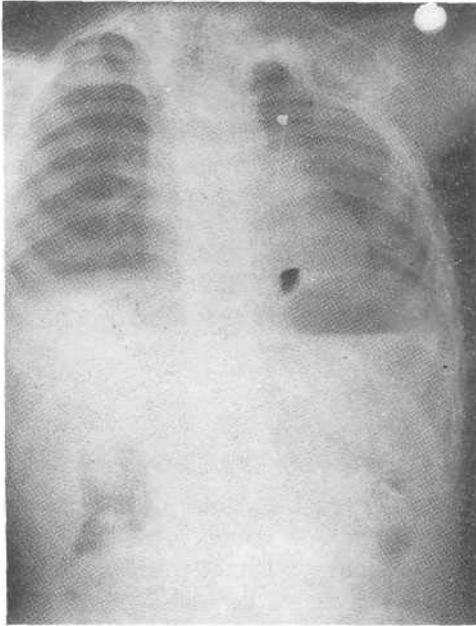


Fig. 1. Sprengel's deformity on left side.

tion, the embryonic dimensions of scapulae are also retained(3).

The scapulae remain high on the chest wall, the inferior pole adducted and the superior angle prominent in the web of the neck. Sometimes the superior aspect may be curved over the apex of the chest wall anteriorly(3). An omovertebral band which may be composed of fibrous tissue cartilage or bone (or both) connects the superior angle of the scapula to the cervical spine as seen in 25-30% of cases(3). Other connections may also be found like a fibrous connection between scapula and chest wall, and osseous bridge extending from the clavicle to spine of scapula(2,4). The movements at the shoulder become limited by these anomalous connections and by the rotational component of the deformity

which makes the glenoid face downwards resulting in decreased abduction.

The deformity that results in normal scapular descent will subsequently also lead to abnormal development of bone and soft tissue affecting the entire shoulder girdle. Congenital anomalies of other systems are also commonly associated. These include diastomata myelia, tethered cord, lumbosacral lipoma, renal anomalies(2), abnormalities of thoracic ribs(1), absence of ribs(5), cervical spinabifida, syringomyelia, platybasia, etc. Cervical vertebral anomalies are often reported(3), but abnormalities of thoracic and lumbar vertebrae as noted in our case are not reported earlier. Sprengel's deformity is unilateral in about 90% of cases(2). Sometimes Sprengel's deformity can be seen as a part of other syndromes like Klippel Feil syndrome(6).

Those with minor cosmetic deformity or minimal functional impairment need no surgery as the benefit may be minimal(3). Various surgical methods have been advocated by different authors. The "Woodward" procedure and the modified "Green" procedure are the currently most used procedures(1,3). Although the ideal age suggested for surgical correction is 3-8 years, age is no longer a specific contraindication(3).

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Dopamine Infusion: A Simpler Formula

Dopamine, a potent sympathomimetic amine is the drug of choice in management of neonatal shock unresponsive to volume expanders, adrenaline and sodium bicarbonate administration. Since the drug has to be administered in continuous infusion, it is imperative to have a simple formula for dosage calculations. The American Heart Association (AHA) has provided the following formula, multiply 6 times the weight in kg times the desired dose in $\mu\text{g}/\text{kg}/\text{min}$, divided by the desired amount of fluid to infuse in ml/h which provides the amount of dopamine in mg to add to each 100 ml solution prepared.

This formula is not easy to remember as evidenced by a small survey in 50 pediatricians, of teaching hospitals in Delhi. The awareness regarding AHA formula was 100% but only 4 out of 50 could correctly reproduce it. Rest of the doctors were using their own improvised formulae. All agreed

that there was a need for simpler formula.

We derived a formula for calculating the dopamine to be infused to ml/hour. This also takes into account the concentration of dopamine available in marketed solution, *i.e.*, 1 ml containing 40 mg of dopamine so that there is no need of further calculations from mg to ml. The derived formula is ml of dopamine/hour = $0.0015 \times \text{wt (kg)} \times \text{dose } (\mu\text{g}/\text{kg}/\text{min})$.

The advantage of this formula lies in its simplicity, easy reproducibility, absence of any division factor involved at any stage, dosages being directly calculated in ml instead of meg or mg. One has to remember only one constant *i.e.*, 0.0015. The formula does not take into account the amount of fluid to be infused, thus also avoiding confusion and multiple calculations if two simultaneous drips are being set up.

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