CONGENITAL LESIONS OF LARYNX

- Laryngomalacia (congenital laryngea l stridor)
- Congenital vocal cord paralysis
- Congenital subglottic stenosis
- Laryngeal web
- Subglottic haemangioma
- Laryngo-oesophageal cleft
- Laryngocele
- Laryngeal cyst

1. Laryngomalacia (congenital laryngeal stridor).

It is the most common congenital abnormality of the larynx.

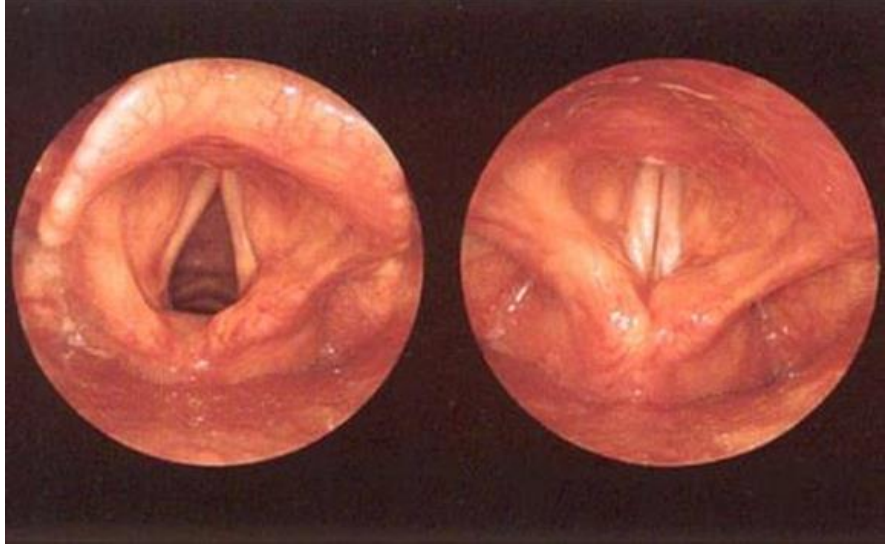
It is c haracterised by excessive flaccidity of supraglottic

larynx which is sucked in during Inspiration producing stridor and sometimes cyanosis.

Stridor is increased on crying

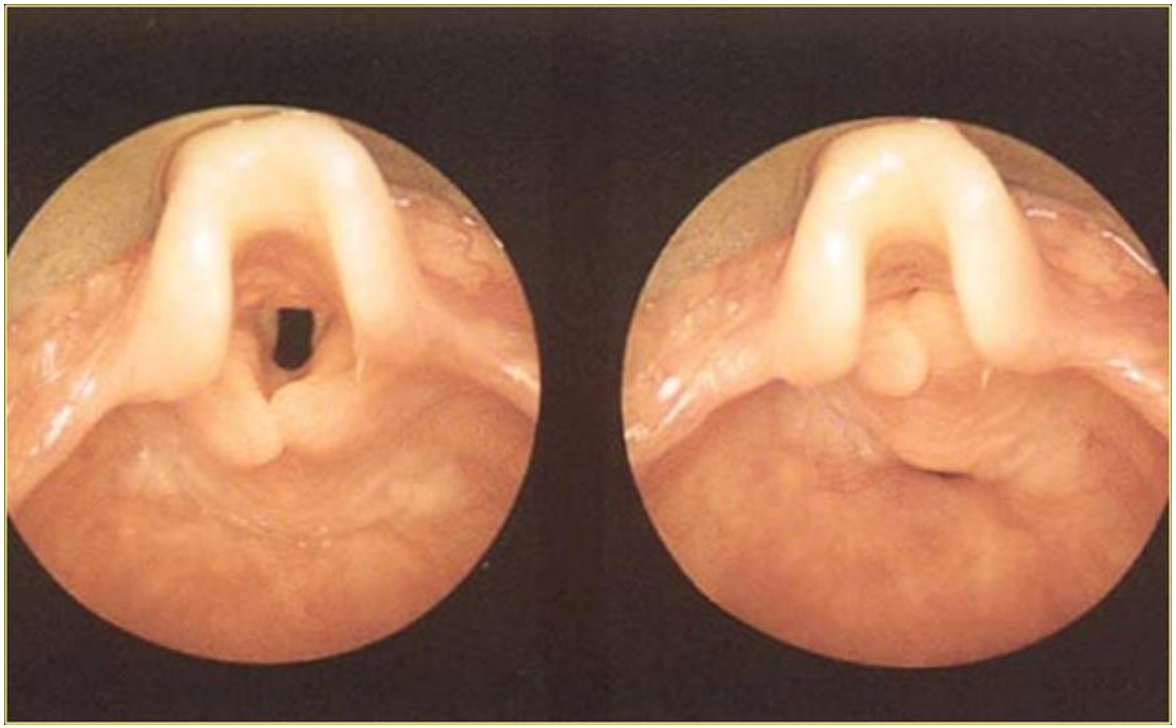
but subsides on placing the child in prone position; cry is normal.

The condition manifests at birth or soon after, and usually disappears by 2 years of age.



Normal La*ry*nx

Collapse of arytenoid mucosa; shortened aryepiglottic folds; tubular epiglottis with posterior collapse



The supraglottic structures are pulled into the lumen around a vertical axis with inspiration

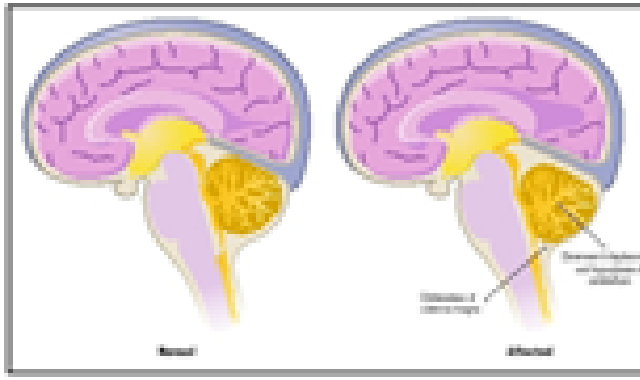
- Condition arises from a continued immaturity of the larynx, as if the fetal stage of laryngeal development has persisted.
- The abnormality appears to be flaccidity or in- coordination of the supra-laryngeal cartilages, especially the arytenoids that is expressed when the infant is stressed by excitation with an increased respiratory rate.
- Stridor is typically noted in the first few weeks of life and is characterized by fluttering, high-pitched inspiratory sounds.

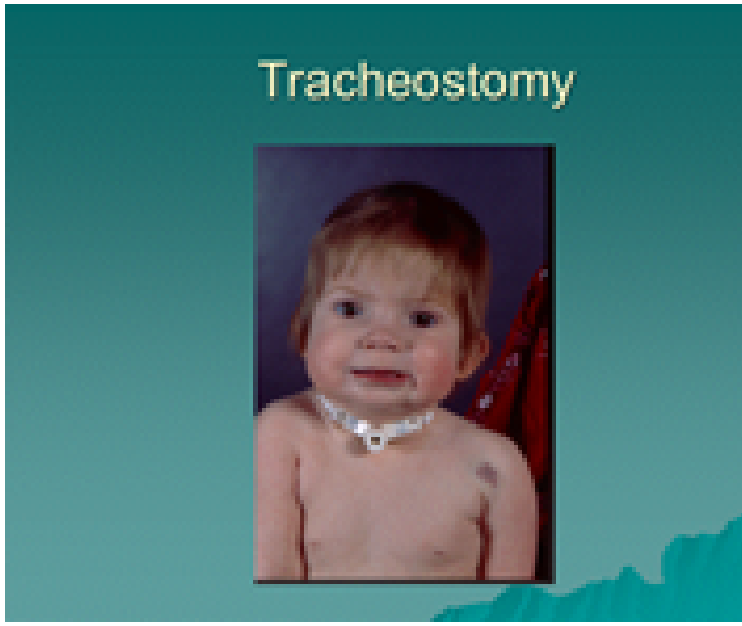
- Therapy consists of confirming the diagnosis by flexible laryngoscopy and reassuring the parents that the prognosis for the child is favorable.
- Position changes of the infant may help alleviate the stridor as it typically worsens in the supine position.
- In the past, tracheotomy was the surgical procedure of choice for severe cases.
- Supraglottoplasty has proven successful for the correction of supraglottic obstruction and is now the surgical procedure of choice.

Congenital Vocal fold paralysis

- Vocal fold paralysis has long been recognized as a significant cause of stridor and hoarseness in infants and children.
- It is the second most common cause of stridor in the newborn behind laryngomalacia. Laryngeal paralysis may be present at birth or may manifest itself in the first month or two of life.
- The neurologic impairment reflects an injury to the vagus nerve.

- The lesion can occur anywhere from the brain through the neck into the chest and into the larynx.
- Many paralyses are idiopathic in up to 47% of cases,
- The most common causative factors include entities such as:
  - Arnold Chiari malformations,
  - Hydrocephalus, neonatal hypotonia,
  - Multiple peripheral paralysis (myasthenia gravis).
  - Other causes include birth trauma and cardiac anomalies.
- Associated laryngeal lesions such as clefts and stenosis are also commonly often found.





Symptoms

- Any or all of the normal laryngeal functions may be abnormal in the pediatric patient with laryngeal paralysis.
- The most common symptom is stridor.
- Ineffective cough, aspiration, recurrent pneumonia, and feeding difficulties are also commonly reported.
- Consistent stridor, cyanosis, and apnea are frequent.
- Voice and cry, however, may be normal particularly in cases of bilateral vocal cord paralysis.
- Hoarseness and dysphonia are common in cases of unilateral vocal fold paralysis.

Management strategies depend on the child's underlying condition.

- Children with bilateral vocal fold paralysis frequently require surgical intervention.
- The airway is often markedly compromised and in over 50% of cases a tracheotomy is required.
- Multiple lateralization options include CO2 laser cordotomy and open arytenoidectomy, artenoidpexy, arytenoid separation with cartilage grafting or laser arytenoidectomy and cordectomy

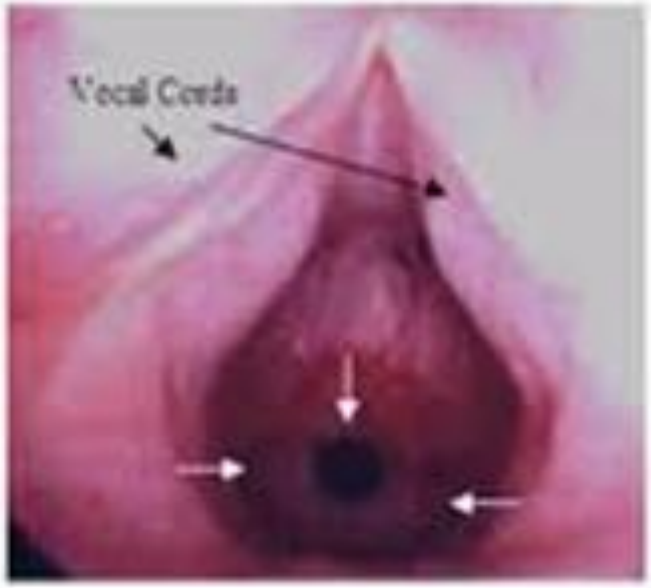
- The management of unilateral vocal cord paralysis in children is usually less urgent than that of bilateral paralysis.
- Children adjust well to persistent unilateral vocal cord paralysis with few sequelae. A weakened cry may result but an adequate airway is the typically maintained.

Subglottic Stenosis

- May be classified as either acquired or congenital.
- Although congenital subglottic stenosis is uncommon, accounting for 5% of all cases, it is the third most common congenital airway problem (after laryngomalacia and vocal cord paralysis).
- Congenital SGS is thought to be secondary to failure of the laryngeal lumen to recanalize properly during embryogenesis.
- SGS is considered congenital if there is no history of endotracheal intubation or other forms of laryngeal trauma.

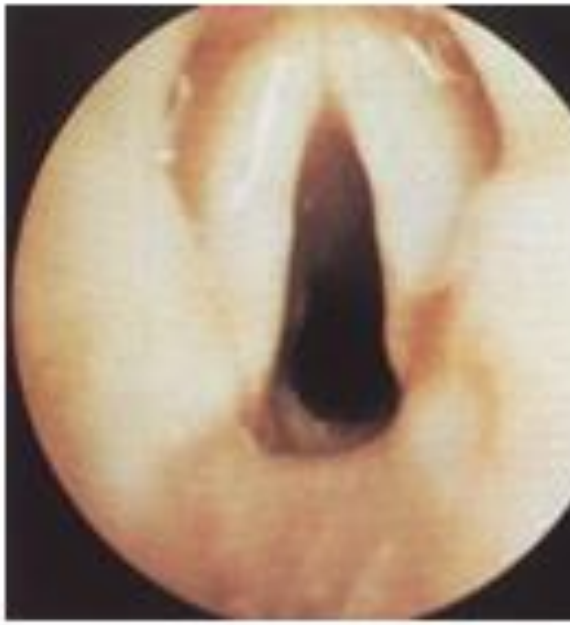
- Subglottic stenosis is defined as a subglottic lumen 4.0 mm in diameter or less at the level of the cricoid in a full term infant.
- The normal newborn subglottic diameter is 4.5 – 5.5 mm and in premature neonates around 3.5 mm.
- Congenital SGS is divided histopathologically into membranous and cartilaginous types.
- Membranous SGS is usually circumferential and consists of fibrous soft-tissue thickening.
- The cartilaginous type usually results from a thickened or deformed cricoid cartilage

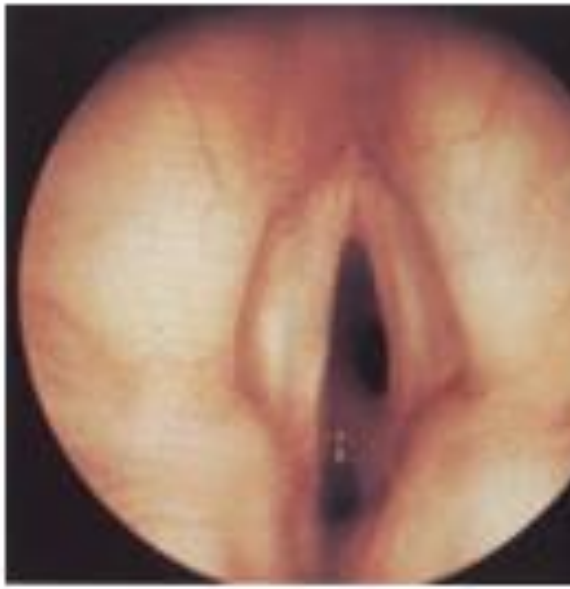
- The severity of congenital subglottic stenosis depends on the degree of SG narrowing.
- Children with subglottic stenosis usually present with stridor and/or respiratory distress.



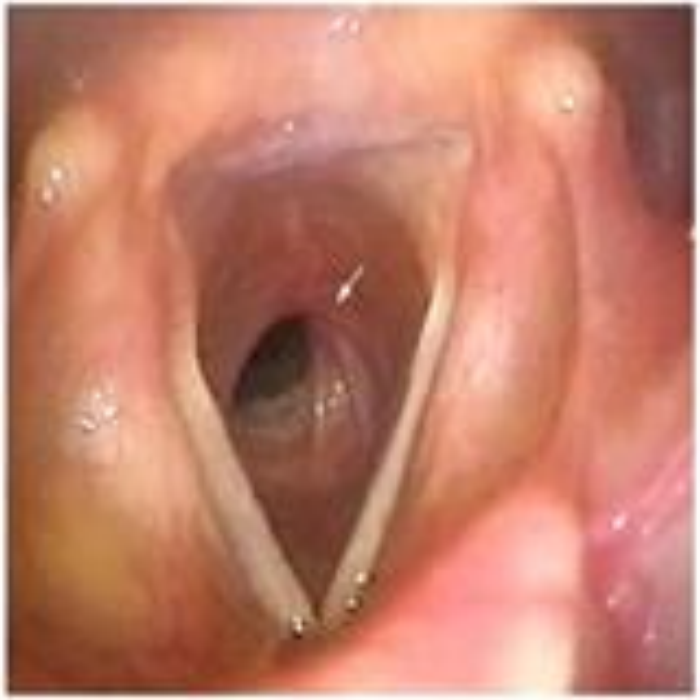
Classification

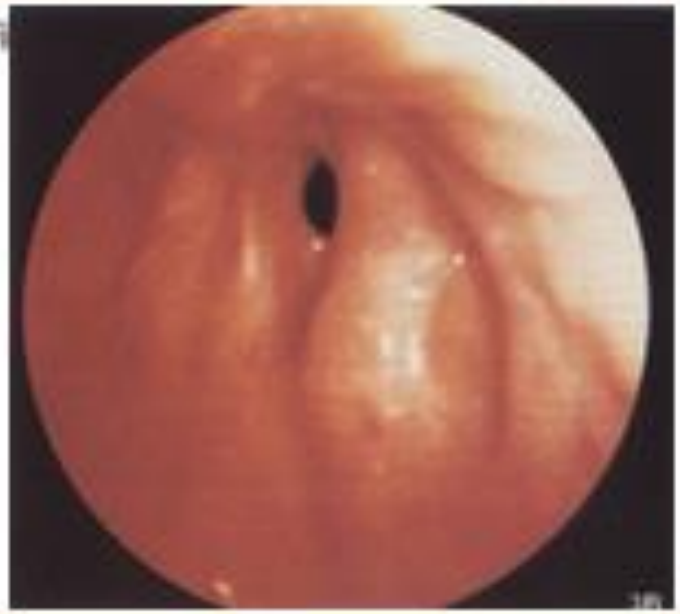
- The McCaffrey system classifies laryngotracheal stenosis based on the subsites involved and the length of the stenosis.
- Four stages are described:
  - Stage I lesions are confined to the subglottis or trachea and are less than 1cm long
  - Stage II lesions are isolated to the subglottis and are greater then 1 cm long





- Stage III are subglottic/tracheal lesions not involving the glottis
- Stage IV lesions involve the glottis





Treatment of congenital SGS is tailored to the symptoms and grade of the stenosis.

- Symptoms are typically less severe in congenital SGS than in the acquired form.
- Congenital SGS also improves as the child grows, and less than half of children with this disorder will require a tracheotomy.
- For those children who do require surgical intervention, several options are available.

Subglottic haemangioma. Though congenital, patient is asymptomatic till 3-6 months of age when haemangioma begins to increase in size. About 50% of the children have associated cutaneous haemangiomas. Patient may present with stridor but has a normal cry. Agitation of the patient or crying may increase airway obstruction due to venous filling.

Direct laryngoscopy shows reddish blue mass below the vocal cords. Biopsy is sometimes,
not always, associated with haemorrhage. Some patients have associated mediastinal haemangioma. Depending on individual case,

treatment
(a) Tracheostomy and observation, as many haemangiomas involute spontaneously.
(b) Steroid therapy. Dexamethasone 1 mg/kg/day for1 week and then predniso lone 3 mg/kg in divided doses for one year.
(c) CO2 laser excision if lesion is small.

. Laryngo-oesophageal cleft. It is due to failure of the fusion of cricoid lamina. Patient presents with repeated aspiration and pneumonitis. Coughing, choking and
cyanosis are present at the time of feeding

Laryngocele. It is dilatation of laryngeal saccule and extends between thyroid cartilage and the ventricle.

It may be internal, external or combined . Treatment is endoscopic or external excision.

8. Laryngeal cyst. It arises in (aryepiglottic fold and appears as bluish, fluid-filled smooth swilling in the supraglottic larynx. Respiratory obstruction my necessitate tracheostomy. Needle aspiration or incision and drainage of cyst provides an emergency airway. Treatment is de roofing the cyst or excision with CO2 laser

Laryngeal webs

Laryngeal webs occur in the glottic level and affect the vocal cords

More than 90% of laryngeal webs are located anteriorly and extend toward the arytenoids.

The webs vary in thickness from a thin structure to one that is thicker and more difficult to eradicate.

Other types include the posterior glottic web, causing interarytenoid vocal cord fixation; subglottic webs, which may occur with or without cricoid cartilage involvement and subglottic stenosis; and supraglottic webs.

Laryngeal webs occur due to incomplete recanalisation of larynx.

Treatment depends on thethickness of the web. Thin webs can be cut with a knife or CO2 laser.

Thick ones may require excision via laryngofissure and placement of a silicon keel and subsequent dilatations.