

Congenital disorders of the Larynx

By

William Kennedy M.D.

UCLA Head & Neck Surgery Program

Congenital disorders of the Larynx

Organogenesis

At birth the larynx is located high in the neck between levels C1- C4

By age 2 the larynx begins to descend inferiorly

By age 6 the larynx reaches the adult position between levels C4 through C7

Congenital disorders of the Larynx

The larynx develops from the endodermal lining and the adjacent mesenchyme of the foregut between the fourth and sixth branchial arches.

At 20 days' gestation, the foregut is first identifiable with a ventral laryngotracheal groove

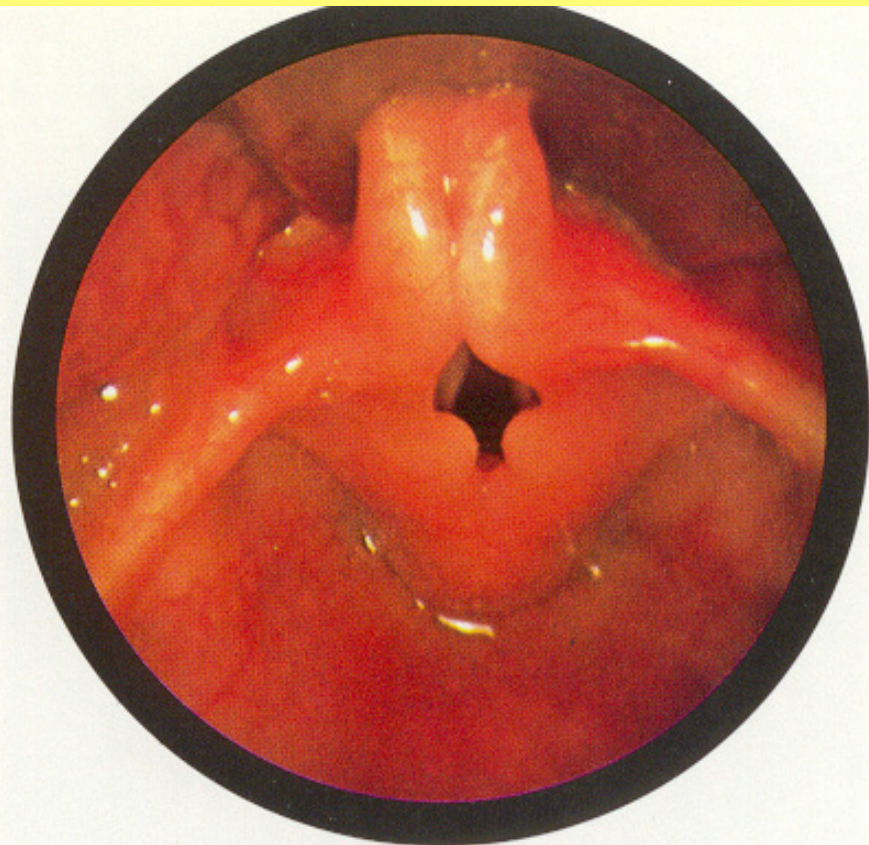
The laryngotracheal groove continues to deepen until its lateral edges fuse. By day 26, this tube descends caudally, where the trachea becomes separated from the esophagus by the tracheoesophageal septum with a persistent slitlike opening into the pharynx.

Laryngomalacia

- Most common congenital anomaly of the larynx
- characterized by partial or complete collapse of the supraglottic structures on inspiration.

It is the most common cause of congenital stridor, accounting for 60% of all cases

Males 2x > female



Laryngomalacia

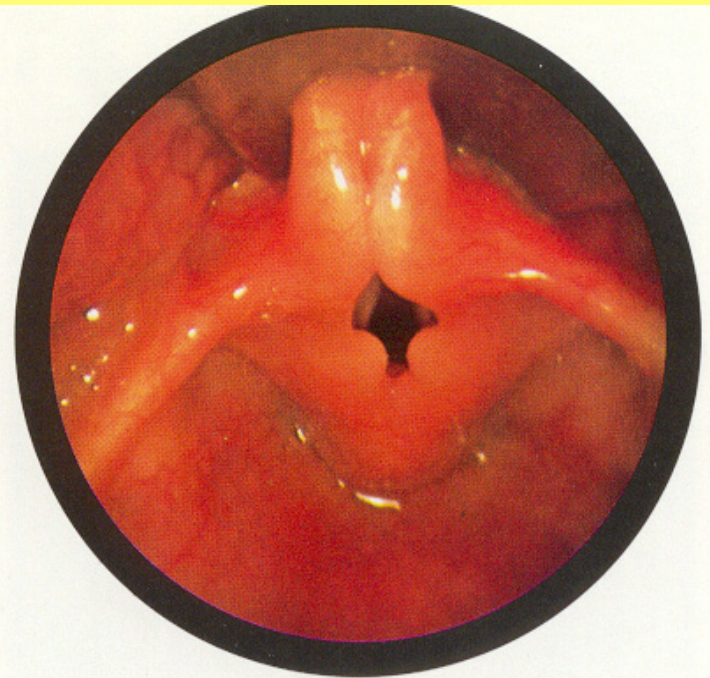
Etiology: Unkown

theories: GERD, Immature neuromuscular control

Epiglottis is derived from the 3rd and 4th branchial arches.

Overgrowth of the 3rd results in elongation of the structure and the observed laryngomalacia

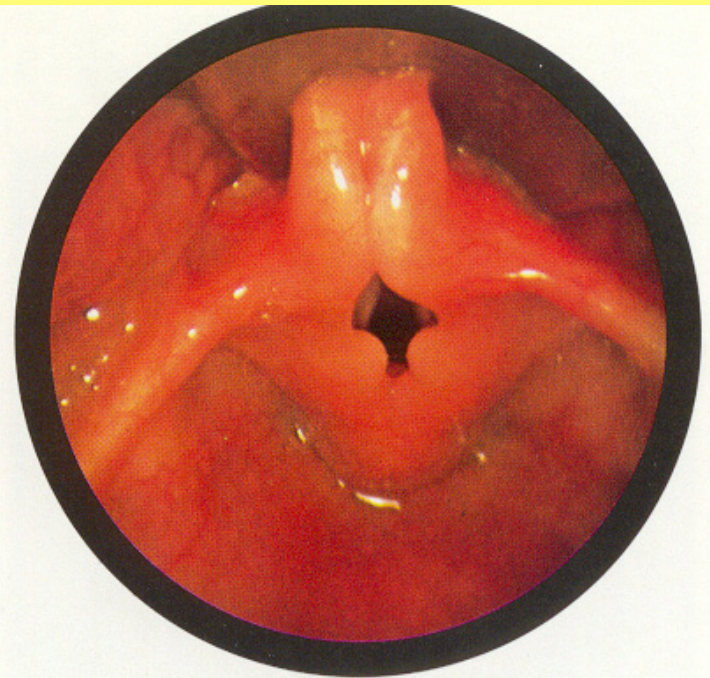
However histological studies do NOT demonstrate a difference between the quality of the cartilage between normal/abnormal



Laryngomalacia

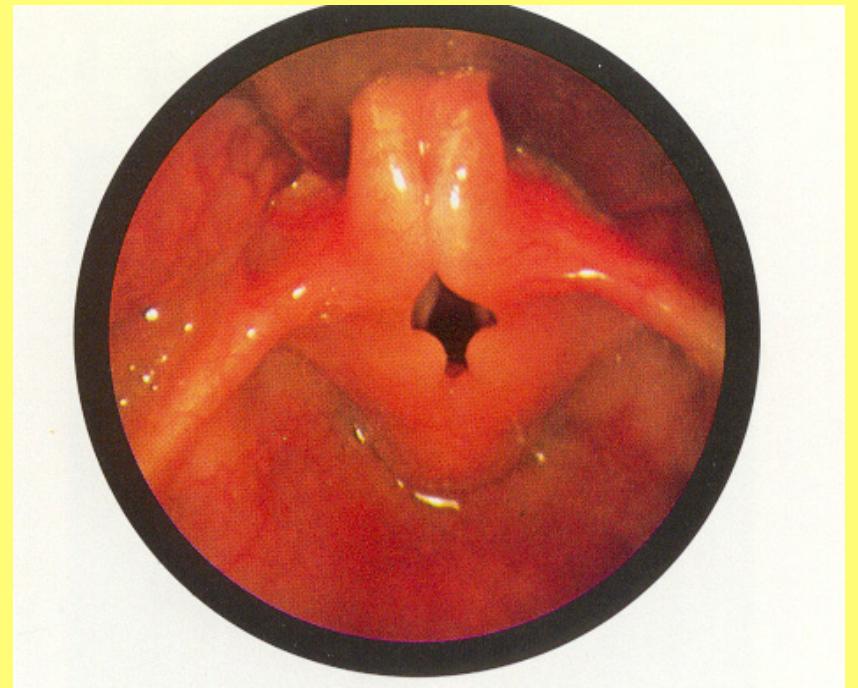
Immature neuromuscular control may be responsible for the arytenoid prolapse observed in laryngomalacia, although an increase in the incidence of laryngomalacia does not occur in premature infants who have classic hypotonicity.

Histological studies of the neuromuscular structures of the larynx do not reveal a difference between children with and without laryngomalacia.



Laryngomalacia

Gastroesophageal reflux (GER) may play an etiological role in laryngomalacia. Histological evidence of gastroesophageal reflux laryngitis has recently been documented from aryepiglottoplasty specimens. The most important feature discovered was a band of inflammation of variable intensity beneath the epithelium with edema deep to it.



Laryngomalacia

Clinical presentation

The most common symptoms inspiratory stridor

- that is worse in the supine position, feeding, and agitation
- that is relieved by neck extension and prone positioning.

Phonation is characteristically normal.

Timeline of Symptoms

Usually absent at *birth*, begin within the *first few weeks of life*, increase over several months, and resolve by *18-24 months of life*.

Less commonly, the child may experience feeding difficulties; however, failure to thrive is rare. Respiratory distress and cyanosis are rare.

Laryngomalacia

Physical exam

Laryngomalacia can best be diagnosed with a flexible fiberoptic laryngoscopy in a patient who is awake.

If the patient is sedated and undergoes flexible fiberoptic bronchoscopy, the topical lidocaine typically used can cause increased collapse of the arytenoids and folding of the epiglottis during inspiration—possibly leading to overestimation of the severity of laryngomalacia

Laryngomalacia

Physical exam

Elongation and lateral extension of the epiglottis (omega shaped) that falls posteroinferiorly on inspiration

Redundant bulky arytenoids that prolapse anteromedially on inspiration

Shortening of the aryepiglottic folds, which results in tethering of the arytenoids to the epiglottis

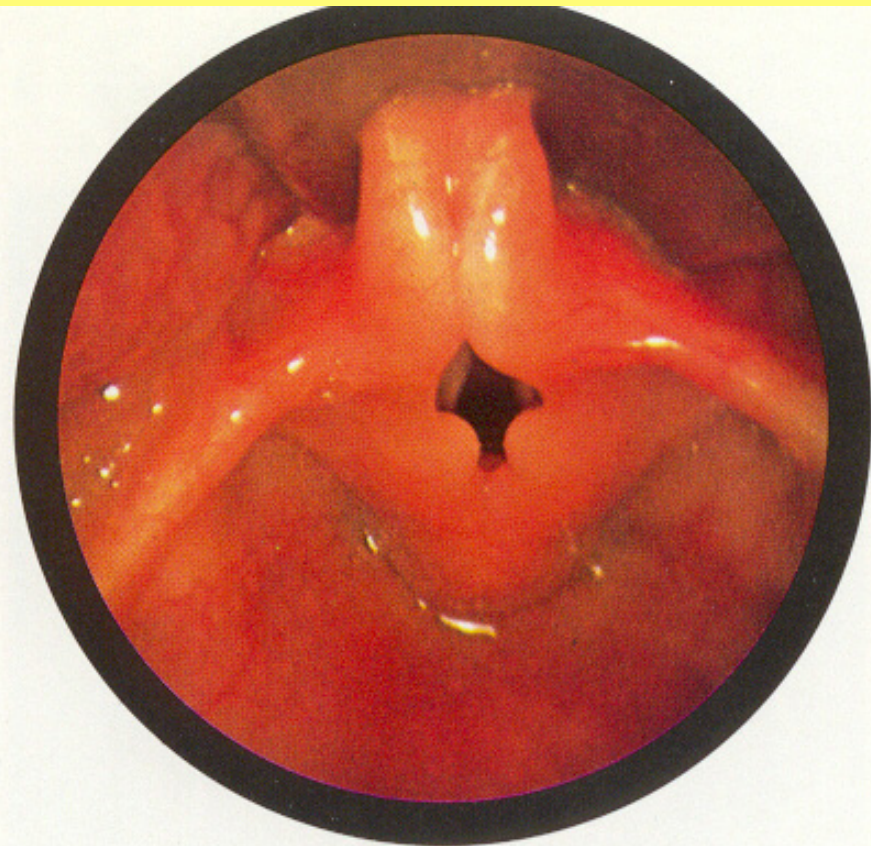
Inward collapse of the aryepiglottic folds (cuneiform cartilages) on inspiration

Visualization of the vocal cords, which are normal in structure and function

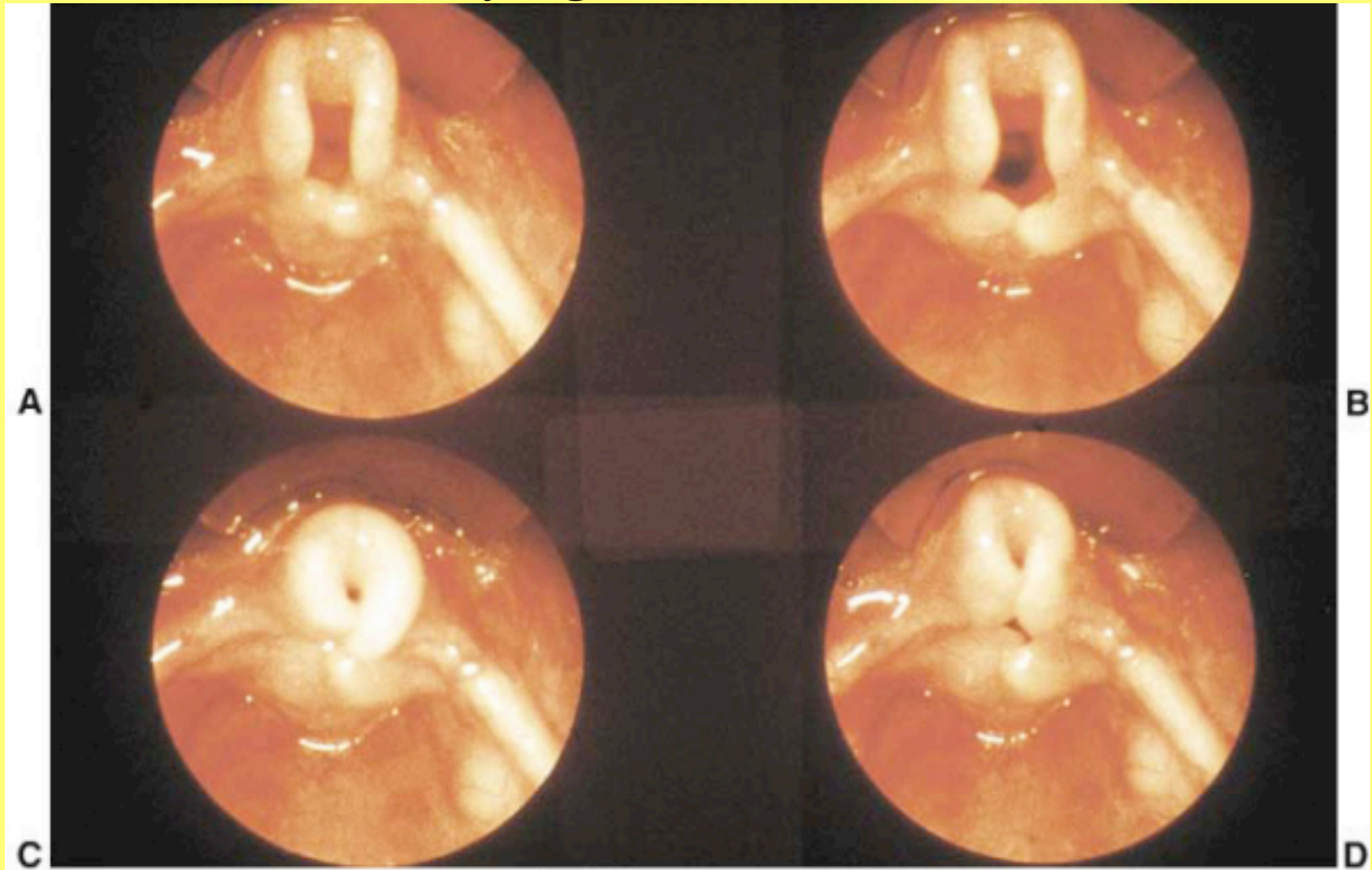
Laryngomalacia

Flexible endoscopy

Elongation and lateral extension of the epiglottis (omega shaped) that falls posteroinferiorly on inspiration



Laryngomalacia



Laryngomalacia: Progressive airway obstruction on inspiration. Note omega-shaped epiglottis. (From Benjamin B: *The pediatric airway*, slide #26, Slide Lecture Series. American Academy of Otolaryngology-Head and Neck Surgery, 1992.)

Laryngomalacia

Treatment

In approximately 90% of reported cases, the condition is mild, no intervention is needed and the parents can be reassured accordingly (Lane et al. 1984)

Medical management should include treatment of documented gastroesophageal reflux disease (GERD) because this condition is known to contribute to laryngomalacia

Laryngomalacia

Treatment

Surgical management is indicated in rare instances if respiratory complications develop if there is serious respiratory obstruction with substantial sternal and intercostal recession, feeding difficulties that may be compounded by reflux enhanced by the high negative intrathoracic pressures generated, and consequent failure to thrive.

Tracheotomy can be performed for emergent episodes of distress and should be left in place until the supraglottic pathology resolves with age.

Laryngomalacia

Treatment

Surgical management

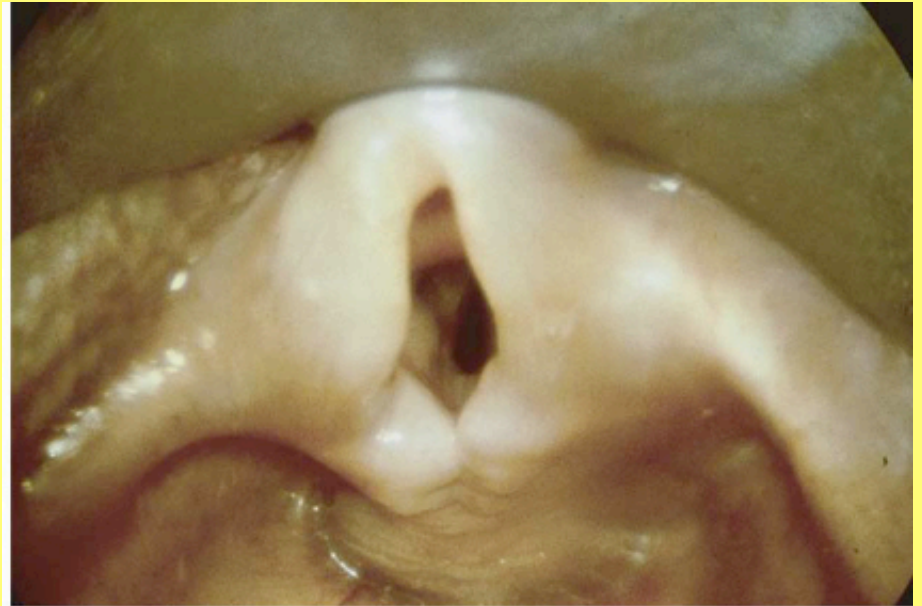
-Restoration of an adequate airway can be achieved by performing an endoscopic aryepiglottoplasty (sometimes termed a supraglottoplasty; Jani et al. 1991).

Supraglottoplasty

- 1) Using cup forceps and microscissors (or alternatively the carbon dioxide laser), each aryepiglottic fold is first divided to release it from the edge of the epiglottis,
- 2) the redundant mucosa and submucosal tissue are then excised from over the arytenoids, together if necessary with part or all of the cuneiform cartilages. The stridor is usually improved immediately following the surgery,

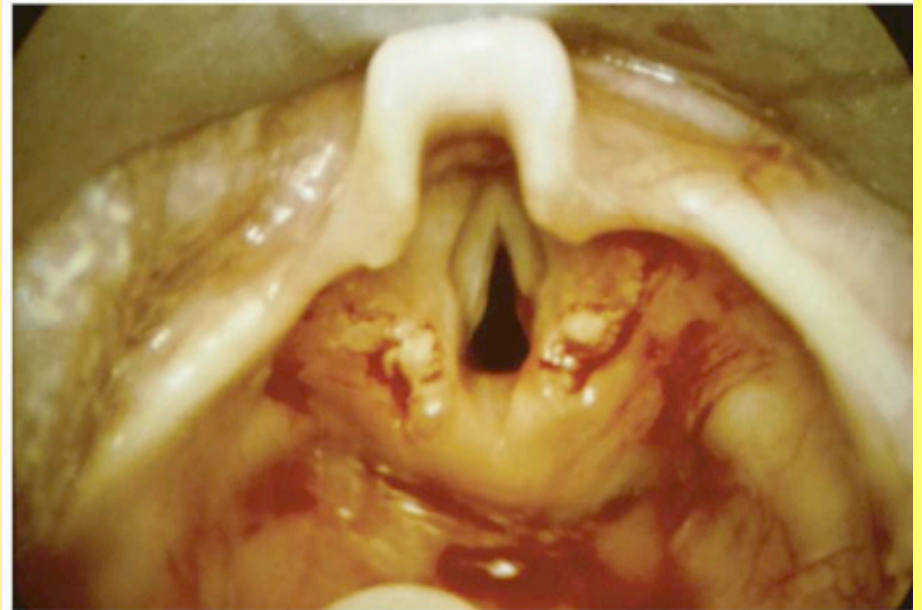
Laryngomalacia

Pre-supraglottoplasty



A

Post-supraglottoplasty.



B

Vocal Fold Paralysis

Epidemiology

Vocal fold paralysis is the second most common congenital anomaly of the larynx, accounting for 15-20% of all cases.

No gender difference exists in the prevalence of this anomaly.

Up to 45% of patients have other, coexisting airway pathology, and so a formal microlaryngoscopy and bronchoscopy is essential.

Vocal Fold Paralysis

Diagnosis

If the child is stable, flexible endoscopy may be performed to demonstrate the paralysis.

Perform rigid bronchoscopy to confirm the diagnosis and to assess the airway for other anomalies.

Bilateral TVC Paralysis

Etiology

usually idiopathic.

may also occur secondary to central neuromuscular immaturity. lesions in the central nervous system, including Arnold-Chiari malformation, hydrocephalus or myelomeningocele,

Birth trauma that causes excessive strain to the cervical spine may cause transient bilateral vocal fold paralysis lasting 6-9 months.

Clinical presentation

Bilateral vocal fold paralysis manifests as an inspiratory stridor at rest that worsens upon agitation in children with near-normal phonation and progressive airway obstruction.

Aspiration is common with bilateral vocal fold paralysis, often resulting in recurrent chest infections and a failure to thrive

Bilateral TVC Paralysis

Management

The vocal cords spontaneously will become mobile in up to 50% of patients with bilateral vocal-cord paralysis. Prognosis is better if there are no associated anomalies. Most recovery of vocal cord function occurs within 24 to 36 months

Patients with bilateral vocal fold paralysis may need urgent airway intervention, which can usually be achieved by endotracheal intubation.

Tracheotomy is necessary to relieve the obstruction. and should remain in place for 2 years to allow for spontaneous recovery, which occurs completely in more than half of patients.

Bilateral TVC Paralysis

Management

If recovery does not occur, consider

- 1) vocal cord lateralization procedures in an effort to decannulate the patient.
- 2) Transverse laser cordotomy has had early success in allowing decannulation in older children and adults.

Unilateral TVC Paralysis

Etiology

usually idiopathic.

Lesions in the mediastinum, such as tumors or vascular malformations, Iatrogenic injury to the left recurrent laryngeal nerve can occur during surgery for cardiovascular anomalies

Clinical presentation

Unilateral TVC paralysis may manifest during the first few weeks of life, or it may go unnoticed. The most common symptoms are a hoarse, breathy cry that is aggravated by agitation. Feeding difficulties and aspiration may also occur.

Unilateral TVC Paralysis

Management

Approximately 70% of noniatrogenic unilateral vocal fold paralyses will resolve spontaneously, most within the first 6 months of life

Most cases of unilateral vocal fold paralysis can be managed by observation, ensuring that respiratory and feeding difficulties do not develop.

Unilateral TVC Paralysis

Management

Upright positioning is usually sufficient to alleviate aspiration difficulties.

Rarely, the child with unilateral vocal fold paralysis has problems with severe aspiration and dysphonia. In these children, injection of the vocal fold with absorbable gelatin sponge (Gelfoam) or polytetrafluoroethylene (Teflon) can be helpful

Congenital Subglottic Stenosis

Epidemiology

is the third most common congenital anomaly of the larynx, accounting for 15% of all cases.

This condition is the most common laryngeal anomaly that requires tracheotomy in infants.

Males affected 2x > as females.



Congenital Subglottic Stenosis

Etiology

Incomplete recanalization of the laryngotracheal tube during the third month of gestation leads to different degrees of congenital subglottic stenosis, with complete laryngeal atresia being the extreme form

Classification CSS can be classified into 2 types.

1) **Membranous CSS** is the result of circumferential submucosal hypertrophy with excess fibrous connective tissue and mucus glands. This type is the most common and mild form of congenital subglottic stenosis.



Congenital Subglottic Stenosis

Classification CSS can be classified into 2 types.

2) **Cartilaginous CSS** results from an abnormal shape of the cricoid cartilage. The cartilage usually narrows laterally but may also develop generalized thickening or excessively large anterior or posterior laminae.

While the lumen at the midportion of the cricoid cartilage is normally elliptical, In some infants with congenital subglottic stenosis, an elliptical cricoid is present with a transverse diameter that is significantly smaller than the anteroposterior diameter,

Congenital Subglottic Stenosis

Clinical presentation

- usually appear in the first few months of life
- stenosis is typically not evident until the child develops an acute inflammatory process, which further compromises the subglottis

Biphasic stridor with or without symptoms of respiratory distress is the most common presenting symptom.

The child may have a barking cough, but the cry is usually normal.

Suspect congenital subglottic stenosis when these symptoms are recurrent or if they are prolonged beyond the normal duration of infectious croup (1-3 d).

Congenital Subglottic Stenosis

Diagnosis

A history of recurrent croup usually suggests CSS.

Perform a *rigid bronchoscopy* to

- to confirm the diagnosis

- to assess the airway for other anomalies.

- evaluate the stenosis in terms of its *length* and *diameter*.

- 1) Passing a scope or endotracheal tube through the stenosis may adequately assess the length and diameter.

- 2) The largest tube or scope to pass through the airway is a good measure of the lumen diameter.

Congenital Subglottic Stenosis

Diagnosis

CSS defined as:

- term infant = lumen diameter is < 4 mm
- preterm infant = lumen diameter $<$ than 3 mm.

The stenosis is then **graded**:

- grade I = less than 50% obstruction;
- grade II = 52% to 70% obstruction;
- grade III = 71% to 99% obstruction;
- grade IV = no detectable lumen.

Congenital Subglottic Stenosis

Management

In patients with grade I stenosis, a watchful waiting approach can be taken and the child will likely grow out of the problem as the cricoid cartilage grows.

If the stenosis is grade II or III, a tracheotomy will likely be needed until the cricoid grows to a sufficient size to allow decannulation, or a laryngotracheal reconstruction performed to expand the cricoid ring.

Subglottic Hemangioma

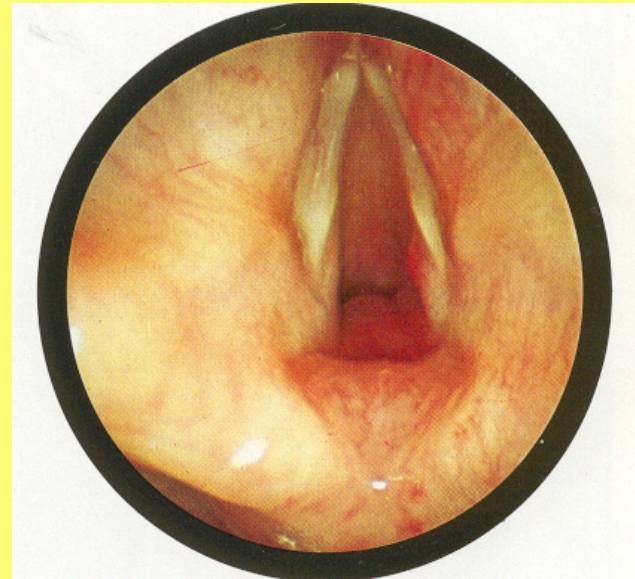
Epidemiology

Subglottic hemangiomas account for 1.5% of all congenital anomalies of the larynx.

Females are affected 2x > males.

Etiology and pathogenesis

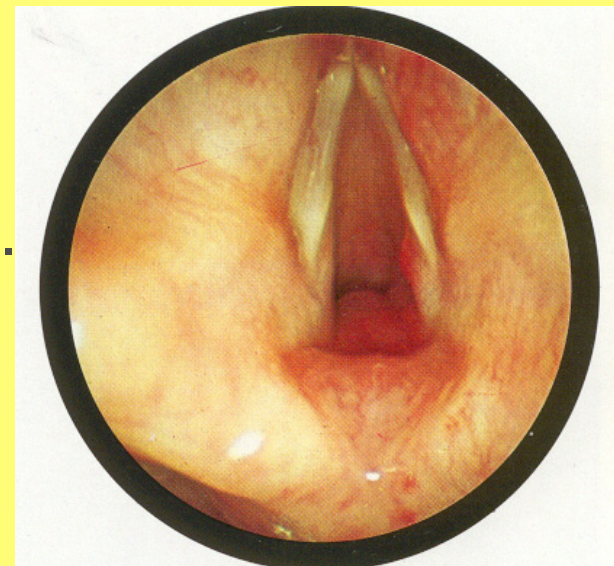
Subglottic hemangiomas develop as a result of a vascular malformation derived from the mesenchymal rests of vasoactive tissue in the subglottis.



Subglottic Hemangioma

Clinical presentation

- Usually asymptomatic at birth.
- As the lesion rapidly grows from age 2-12 months, the child develops progressive respiratory distress that is initially intermittent and then continuous.
- Symptoms manifesting with biphasic stridor, barking cough, normal or hoarse cry, and failure to thrive.
- Most patients develop airway obstruction significant enough to necessitate intervention.



Subglottic Hemangioma

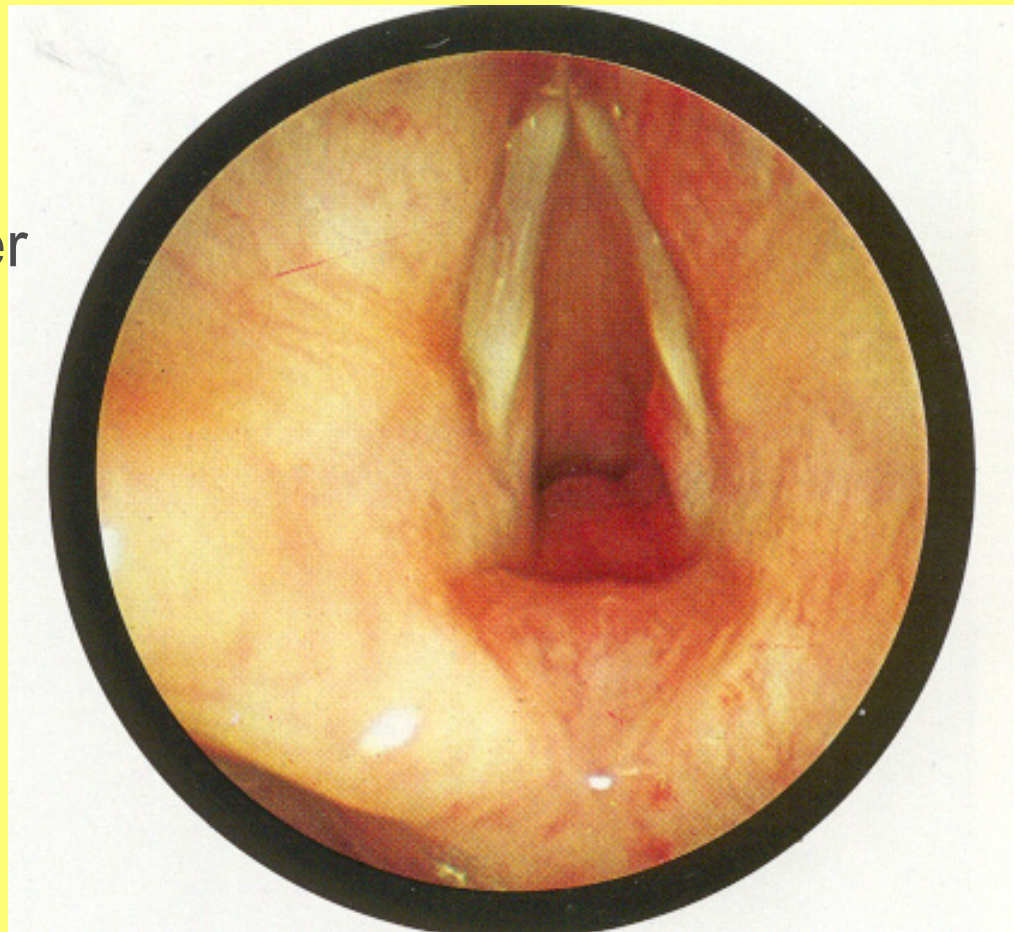
Diagnosis

Rigid bronchoscopy is necessary to establish a diagnosis of subglottic hemangioma.

The lesion is usually located posterolaterally in the submucosa in the subglottis.

It may be unilateral or bilateral, or it may be located in the upper trachea.

The lesion is pink-blue, sessile, and easily compressible

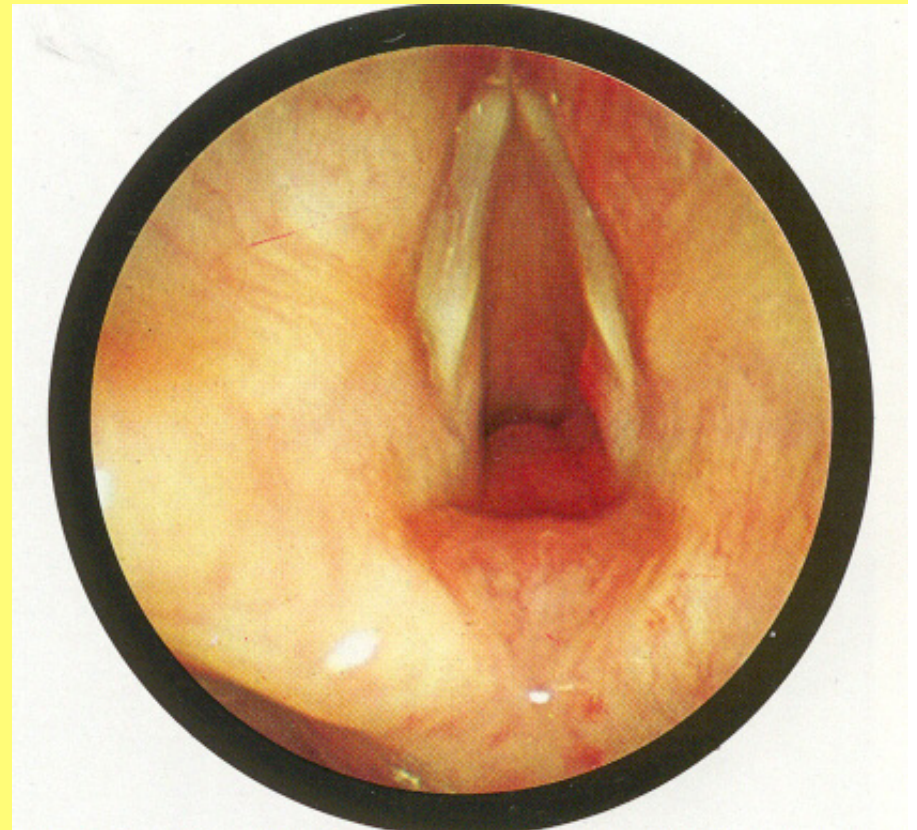


Subglottic Hemangioma

Diagnosis

When the diagnosis is unclear, perform biopsy of the lesion with caution because of the risk of significant hemorrhage.

Plain radiographs of the neck may show an asymmetric narrowing of the subglottis, which may aid in establishing the diagnosis prior to endoscopy.



Subglottic Hemangioma

Management

- 1) Observation usually suffices for small lesions, which do not cause significant airway obstruction. Tracheotomy is typically necessary to secure the airway until the lesion spontaneously regresses, usually by age 5 years.
- 2) Steroid injection into small- or medium-sized hemangiomas may precipitate involution secondary to suppression of estradiol stimulation of the lesion and increased responsiveness to vasoconstrictors.
- 3) Systemic Steroid injection In most children, systemic steroids result in at least partial regression of the hemangiomas. Long-term therapy lasting several months is needed if the hemangioma is to be controlled, leading to possible severe side effects such as Cushing's syndrome, hypertension, diabetes, and growth retardation

Subglottic Hemangioma

Management

3) Carbon dioxide laser ablation The carbon dioxide and potassium titanyl phosphate (KTP) lasers have been used in an effort to control the hemangiomas.

Using a CO₂ laser has been reported to be successful, but others have reported that the laser did not result in earlier tracheotomy decannulation.] It has been proposed that the KTP laser may be the superior instrument for treatment of the hemangiomas because the KTP beam is preferentially absorbed by hemoglobin.

The most significant complication associated with the laser is subglottic stenosis, which has been reported in up to 18% of cases.

Congenital Laryngeal Webs

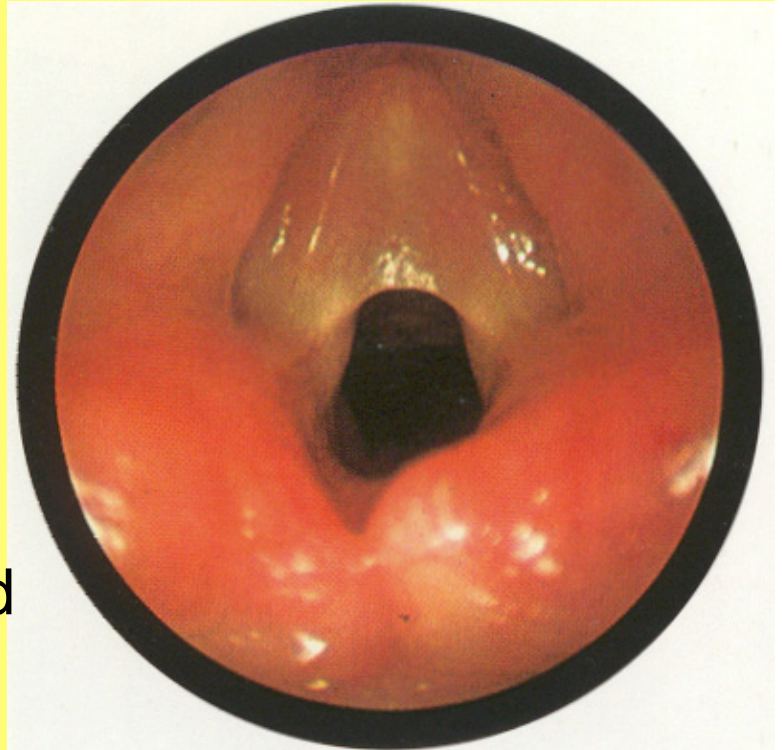
Epidemiology

Laryngeal webs are rare congenital anomalies of the larynx.

Etiology and pathogenesis

Incomplete recanalization of the laryngotracheal tube during the third month of gestation leads to different degrees of laryngeal webs.

The most common site of development of laryngeal webs is at the level of the vocal folds anteriorly, although they may occur in the *posterior* interarytenoid or in the subglottic or supraglottic area.



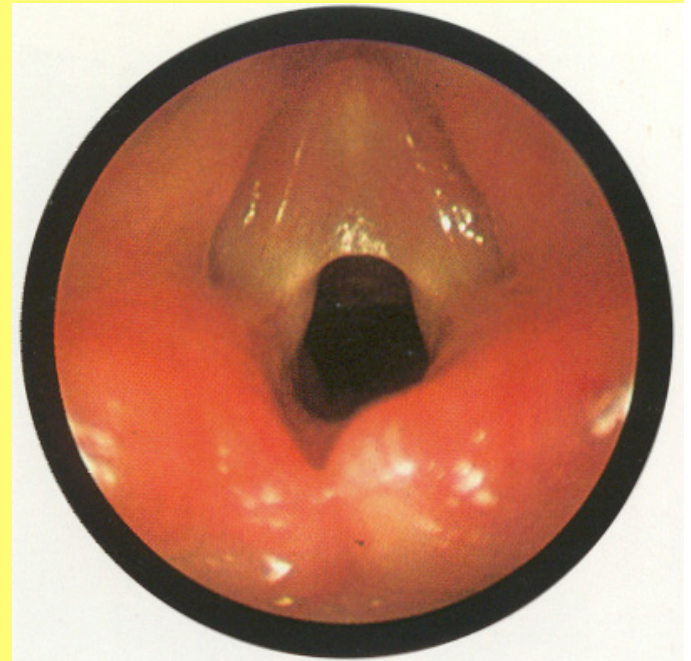
Congenital Laryngeal Webs

Clinical presentation

Laryngeal webs may manifest with symptoms ranging from mild dysphonia to significant airway obstruction, depending on the size of the web.

Stridor is rare except in patients who have a posterior interarytenoid web.

A third of children with laryngeal webs have anomalies of the respiratory tract, most commonly subglottic stenosis.



Congenital Laryngeal Webs

Posterior Webs

Apparent bilateral vocal cord paralysis may occur if an interarytenoid web is present in the posterior **larynx** causing limited vocal fold abduction.

This type of **congenital** web is rare and often necessitates a tracheotomy in the early years of life.

Stridor is the major presenting clinical feature, but patients can also present with obstructive cyanosis at birth or episodes of apnea.



Congenital Laryngeal Webs

Posterior Webs

Diagnosis is made at the time of DL with palpation of the interarytenoid area. Posterior webs may be associated with subglottic stenosis.

Management options include long-term tracheotomy with probable decannulation after 3 to 5 years, division of the web (frequently unsuccessful), arytenoidectomy, or open airway reconstruction with a posterior cartilage graft.



Congenital Laryngeal Webs

Anterior Webs

Anterior **congenital** laryngeal webs are also rare anomalies that, if severe, are diagnosed in the neonatal period after an investigation for the source of aphonia and stridor.

Occasionally a minor web will not be diagnosed until the child is older and undergoes an evaluation for chronic hoarseness



Congenital Laryngeal Webs

Anterior Webs

Management Successful treatment of an anterior laryngeal web most often demands that an open approach be used so that the subglottic narrowing can be corrected. Laryngotracheal reconstruction or laryngofissure and silastic keel are used to correct the defect.

If laryngotracheal reconstruction is performed, the approach can sometimes be single-staged.

In rare cases, where the web is limited, endoscopic laser treatment may be successful.



The End