

Stridor in Infancy

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May 2011

Objectives

- Identify symptoms and associated findings of pediatric stridor
- Describe the possible sites of obstruction leading to stridor
- Understand its different etiologies
- Determine the appropriate evaluation and management of the stridorous pediatric patient

Stridor

- Harsh, high-pitched musical sound
- Produced by turbulence of airflow through a partially obstruction in the larynx
- Pathologic narrowing of the airway
- The site of the obstruction must be in the airway but the lesion may be extrinsic to the airway

Stertor

Low pitched inspiratory sound produced by nasal or nasopharyngeal obstruction

Wheeze

- A continuous sound made by the walls of a narrowed airway vibrating against one another
- Can be inspiratory or expiratory but more often expiratory
- Multiple pitches (polyphonic)

Anatomical and Physiological Considerations

Differences between the anatomy and physiology of the infant/child respiratory system and that of an adult

- Airway size and shape
- Rate of oxygen consumption
- Lung capacity
- Compliance of the lung

Infant Airway





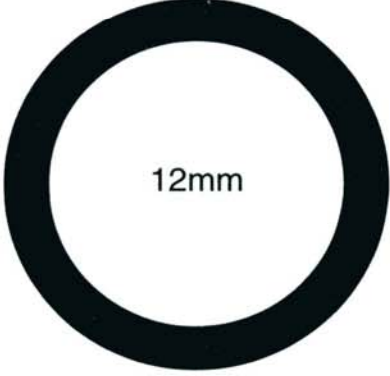
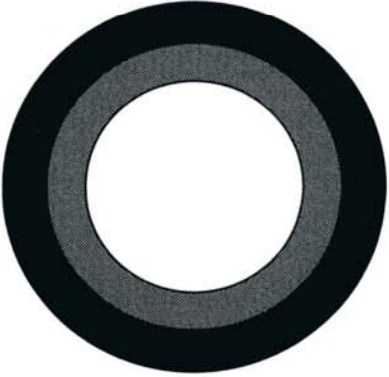
- At birth the infant larynx is approximately one third the size of the adult larynx
- The vocal cords are 6 to 8 mm long, with the posterior aspect composed of the cartilaginous process the arytenoid
- The subglottic diameter measures approximately 4.5 by 7mm
- A diameter of less than 3.5 mm suggests a marginal subglottic airway and is consistent with subglottic stenosis

Infant Airway

- Epiglottis is proportionally narrower than that of the adult and assumes either a tubular form or the shape of the Greek letter omega
- The lumen of the cricoid ring is systematically smaller than the trachea from birth to 3 years of age and its small size may correlate with infants at risk for early infant death
- Circumferential mucosal edema of 1 mm within the larynx of an infant causes a glottis to narrow by over 60%.



The Larynx, Trachea, Bronchi, Lungs, and Esophagus

	Normal	Edema 1mm	Resistance	X-Section Area
Infant	 4mm		↑ 16X	↓ 75%
Adolescent	 8mm		↑ 3X	↓ 44%
Adult	 12mm		↑ 2X	↓ 30%

Clinical Manifestations of laryngeal anomalies

- Respiratory Obstruction
- Stridor
- Weakened or Abnormal Cry
- Dyspnea
- Tachypnea
- Aspiration
- Sudden Death

Location of Obstruction

Stridor can be localized to discrete areas of the airway according to the nature of the sound in relationship to the phase of breathing

Location of Obstruction

These discrete regions can be divided into three zones

1. Supraglottic and supralaryngeal zone which includes the pharynx
2. Extrathoracic tracheal zone including both glottis and subglottis
3. Intrathoracic tracheal zone which includes primary and secondary bronchi

Location of Obstruction

- Supraglottis- Inspiratory and high-pitched
- Glottis and Subglottis (extrathoracic tracheal zone)- Biphasic of intermediate pitch
- Intrathoracic tracheal/bronchial zone- Expiratory often confused with wheezing

Stridor

- Detailed History
- Good Physical Exam
- Sudden Stridor ----> Urgent
- Mild Stridor ----> Outpatient

History

- Duration and presence of any respiratory distress
- Time of onset- at birth, gradual, progressive, etc
- Relationship to feeding
- Past medical history- History of intubation
- Characteristics of the cry
- Trauma
- Foreign body- Laryngeal or esophageal FB
- Associated symptoms

Physical Exam

- CAREFUL INSPECTION OF THE PATIENT IS THE FIRST PRIORITY
- Respiratory rate and degree of distress
- Tachypnea and onset of fatigue
- Flaring of nasal alae, retractions and other signs of respiratory distress
- Auscultation- Sequential listening over the nose, open mouth, neck and chest
- Respiratory cycle
- Stridor as it relates to infant positioning
- Quality of voice or cry

Radiologic Evaluation

- Plain views of the soft tissues of the neck and chest provide information about airway patency and the presence of mass lesions
- Video Fluoroscopy to ascertain respiratory effort and segmental ventilation
- Barium swallow- Vocal cord paralysis, posterior laryngeal cleft, external compression from vascular structures
- CT/MRA- Vascular compression of tracheobronchial tree
- Ultrasound used in infants with VC paralysis

Flexible Endoscopy

- Transnasal Flexible Endoscopy
- Performed while awake
- Can be performed in the office/clinic
- Examination of the nose, choana, nasopharynx, hypopharynx, supraglottis and glottis
- Vocal cord mobility, laryngeal masses, laryngomalacia and other laryngeal problems

Rigid Endoscopy

- Indicated when:
 - Diagnosis remains in question
 - The previous evaluation suggests a subglottic lesion
 - A second significant distal lesion in the airway is suspected in addition to the diagnosis of a more obvious proximal lesion
- Sequential inspection of the pharynx, larynx, trachea and bronchi
- Lumen size, VC mobility and the presence of dynamic compression or infection

CONGENITAL MALFORMATIONS OF THE AIRWAY

Congenital Malformations of the Airway

Congenital laryngeal
anomalies in 1:10,000 to
1:50,000 live births (Van den
Broek and Brinkman 1979)

SUPRAGLOTTIS

CONGENIAL FLACCID LARYNX LARYNGOMALACIA

- Accounts for about 60% of laryngeal problems in the newborn
- Flaccidity or incoordination of the supralaryngeal cartilages, especially the arytenoids
- Stridor is typically noted in the first few weeks of life and is characterized by fluttering, high pitched inspiratory sounds.

Symptoms

- Intermittent, high-pitched inspiratory stridor is the hallmark of laryngomalacia
- Symptoms usually appear within the first two weeks of life
- An increase in the severity of stridor over the initial few months usually is followed by a gradual improvement
- Symptoms are usually at their worst at 6 months of age
- Most patients are symptom free by 18 to 24 months of age

Symptoms

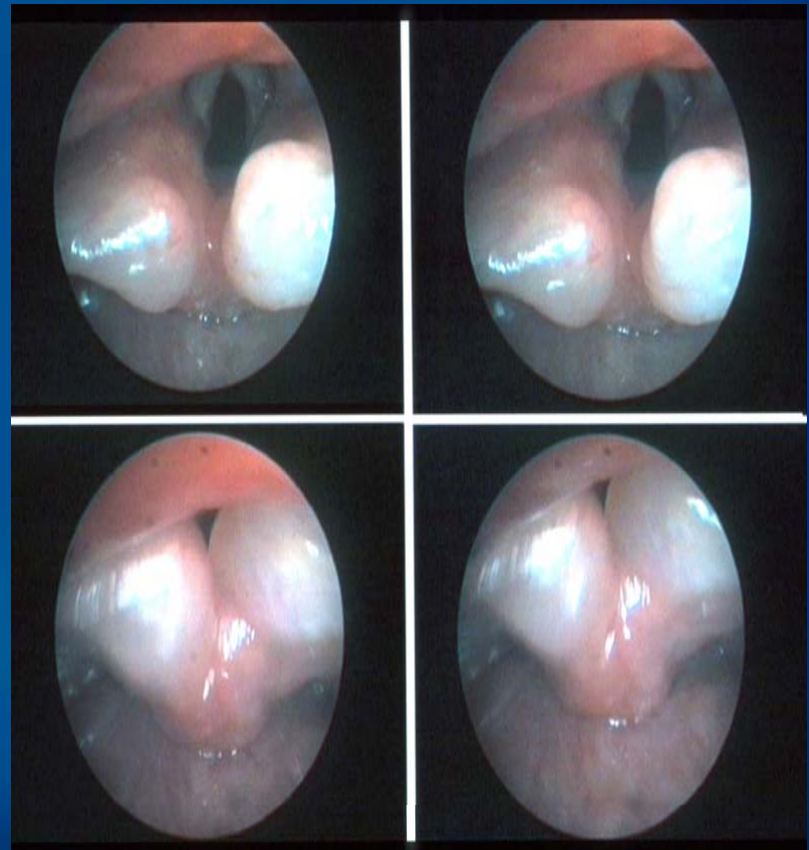
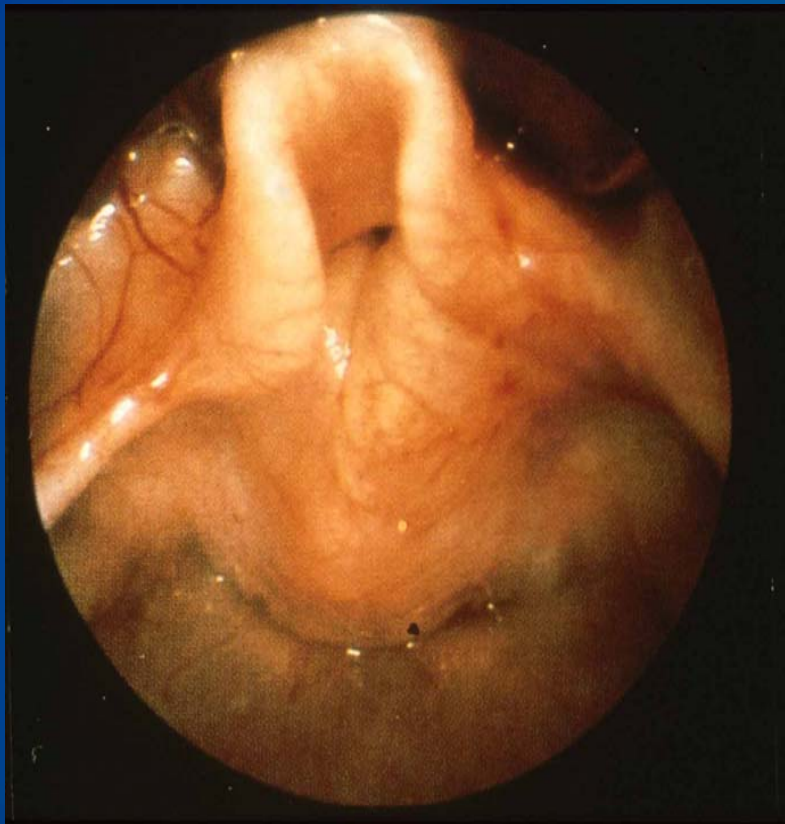
- Stridor is exacerbated by exertion
 - Crying, agitation, feeding or supine positioning
- Moderate to severe cases maybe complicated by feeding difficulties, gastroesophageal reflux, failure to thrive, cyanosis, intermittent complete obstruction or cardiac failure

Diagnosis

Diagnosis in the neonate can be confirmed only by direct observation of movement of the supraglottis during respiration

- Awake fiberoptic laryngoscopy
- Direct laryngoscopy and rigid bronchoscopy for severe symptoms and to evaluate the possibility of synchronous lesions (which exist in up to 27% of patients with laryngomalacia)

Laryngomalacia



Treatment

- Expectant observation is suitable for most cases of laryngomalacia
- Most patient's symptoms resolve spontaneously without intervention
- Medical treatment of any primary or secondary gastroesophageal reflux

Surgical Intervention

- Apparent life threatening events
- Feeding difficulties
- Failure to thrive
- Stridor with cyanosis
- Apnea
- Cor Pulmonale

Surgical Intervention

Historically tracheostomy was the standard therapy

- Currently treated by “supraglottoplasty”
Trimming of mucosa from the lateral edges of the epiglottis, the aryepiglottic folds and the arytenoids as necessary depending on the site and degree of obstructive tissue

Other supraglottic causes of stridor

Supraglottic Hemangioma

- Rare and less common than those of the subglottic area
- Treatment is centered around airway protection
- Spontaneous resolution usually occurs by 2 years of age

Others:

- Laryngocele
- Saccular cysts
- Bifid/Absence of Epiglottis
- Lymphangioma
- Supraglottic Web

GLOTTIS

Vocal Cord Paralysis (VCP)

- VCP is the 3rd most common congenital laryngeal anomaly producing stridor in infants and children
- Unilateral and bilateral vocal cord paralysis occur with equal frequency
- Of those cases of congenital VCP 50% are associated with other anomalies
- Of those acquired VCP nearly 70% are secondary to congenital neurologic abnormalities (meningomyelocele, Arnold Chiari malformation and hydrocephalus) or the neurosurgical efforts to treat them

Symptoms

- Bilateral VCP of the vocal folds typically produces high-pitched, inspiratory stridor
- Unilateral VCP produces much less prominent symptoms in the neonate
 - Weak cry
 - Breathiness
 - Feeding difficulties secondary to laryngeal penetration and aspiration

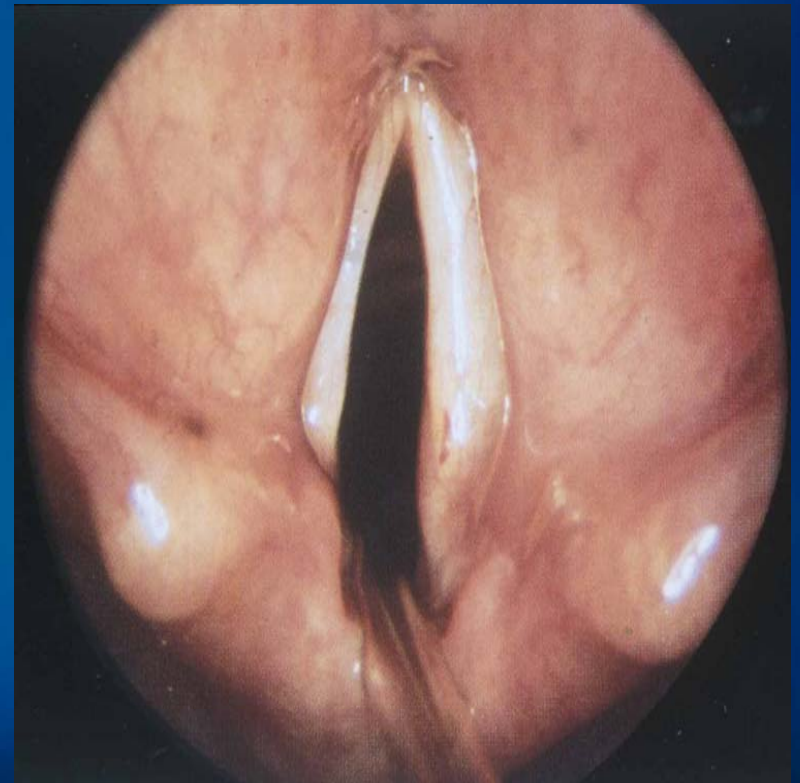
Diagnosis

- Usually made by awake flexible laryngoscopy
- Thorough investigation for the underlying cause should include imaging of the head and chest to evaluate for possible associated cardiovascular or neurologic anomalies

Vocal Fold Paralysis

Unilateral

- Difficult Delivery
- Weak Cry
- Aspiration
- Spontaneous Resolution

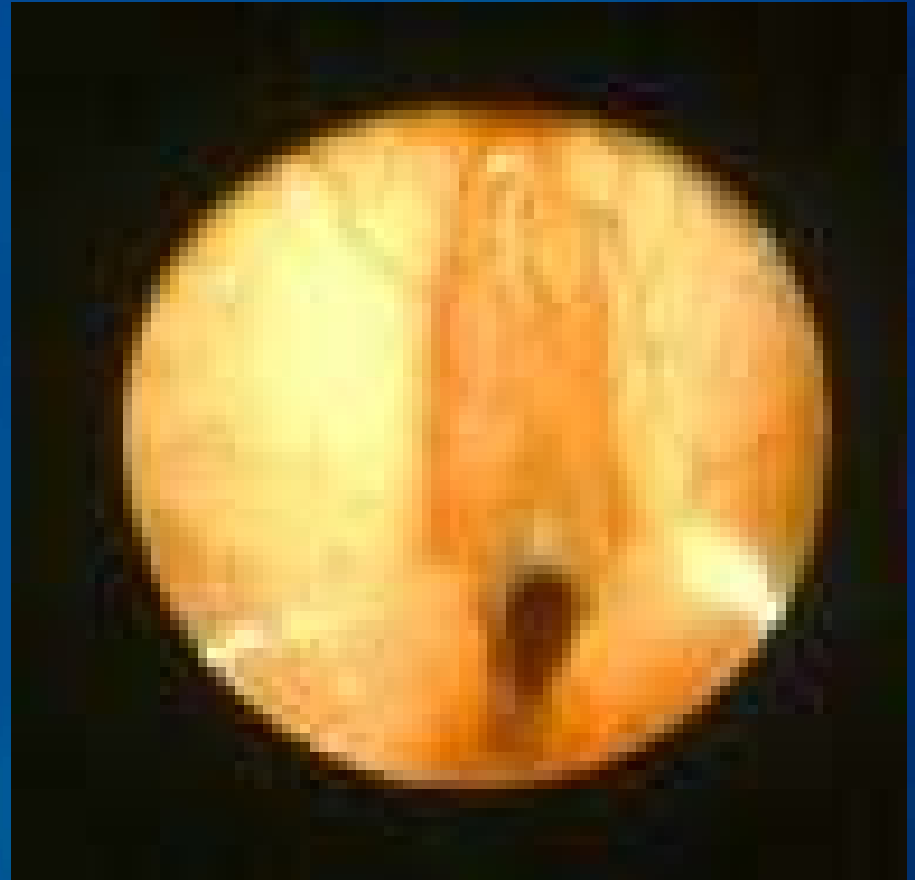


Treatment

- If treated early, paralysis due to increased intracranial pressure often responds to cerebrospinal shunting or posterior fossa decompression
- VCP in infants usually resolves within 6 to 18 months
- Unilateral VCP rarely requires surgical intervention
- Bilateral VCP a temporary tracheostomy is usually, but not always, necessary

Laryngeal Glottic Web

- 75% of laryngeal webs occur at the level of the glottis
- Anterior with posterior concave opening
- Most are thick and fibrous with subglottic extension



OTHER GLOTTIC CAUSES OF STRIDOR

- Cri du Chat Syndrome
- Anterior Laryngeal Cleft
- Duplication of Vocal Cord
- Neurofibromatosis of the Larynx
- Amyloidosis of the Larynx
- Congenital Neuromuscular Disorders
- Laryngoptosis
- Laryngeal Atresia

SUBGLOTTIS

SUBGLOTTIC STENOSIS

- Congenital vs. Acquired
- Airway lumen in the region of the cricoid measuring <4.0 mm in diameter in a full term newborn (3.0 mm in premature infant)
- Mild cases can present as recurrent “croup”
- Grades I-IV
- Congenital subglottic stenosis is often associated with other congenital lesions and syndromes
- Treatment depends on Grade

Congenital Subglottic Stenosis

- Second most common cause of stridor in neonates, infants and children
- Involves narrowing of the subglottic lumen in the absence of trauma (intubation)
- Incomplete recanalization of the laryngeal lumen during embryogenesis
- Can be divided by histologic criteria into cartilaginous and membranous

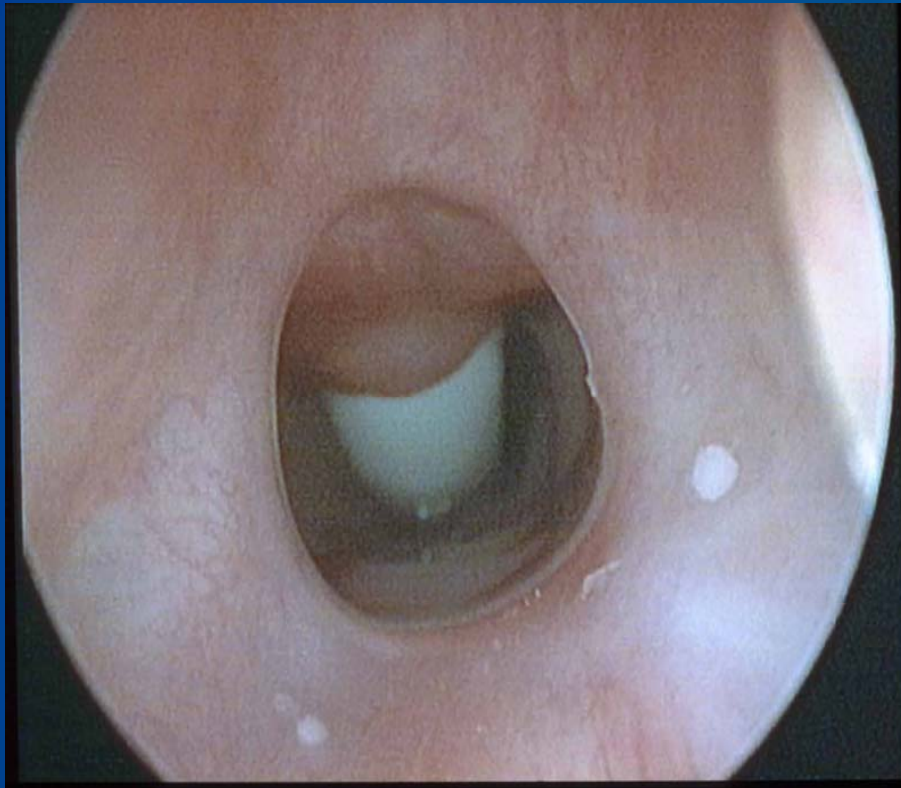
Acquired Subglottic Stenosis

- Low Birth Weight
- Prematurity
- Systemic Infections
- Prolonged Ventilation
- Bronchopulmonary Dysplasia

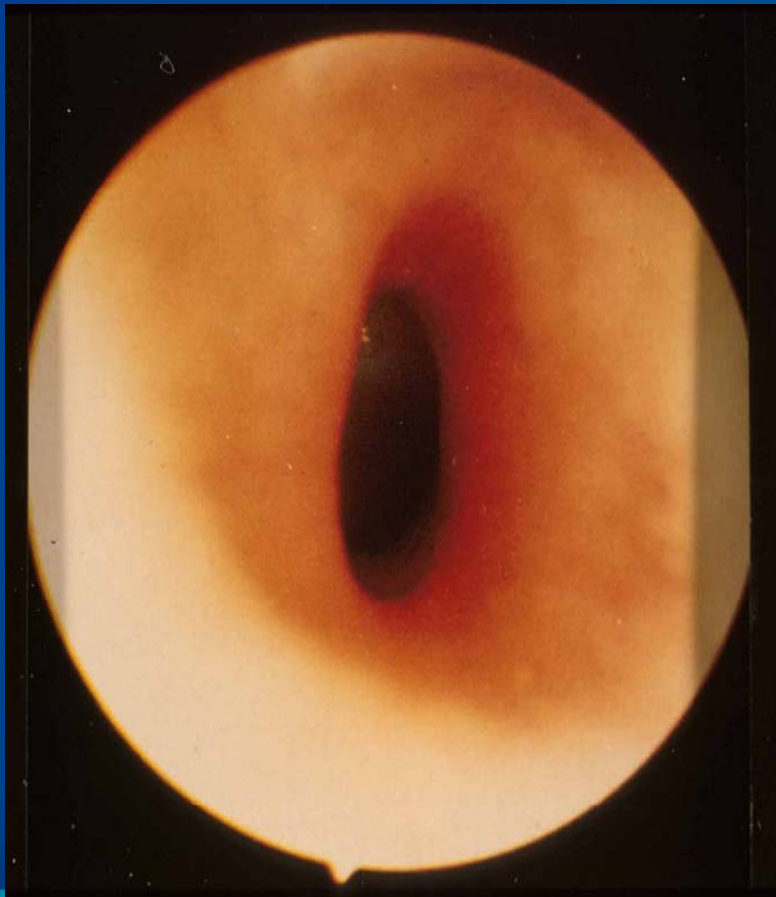
Symptoms




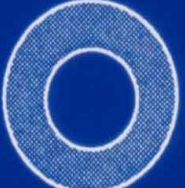
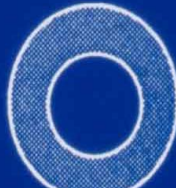

- Mild to moderate stenosis maybe asymptomatic until an upper respiratory tract infection results in additional subglottic edema
- Patients with this condition often have a history of recurrent and prolonged croup episodes
- With severe obstruction during the neonatal period intubation is often necessary

Subglottic Stenosis



Subglottic Stenosis



Classification	From	To
Grade I	 No Obstruction	 50% Obstruction
Grade II	 51% Obstruction	 70% Obstruction
Grade III	 71% Obstruction	 99% Obstruction
Grade IV	No Detectable Lumen	

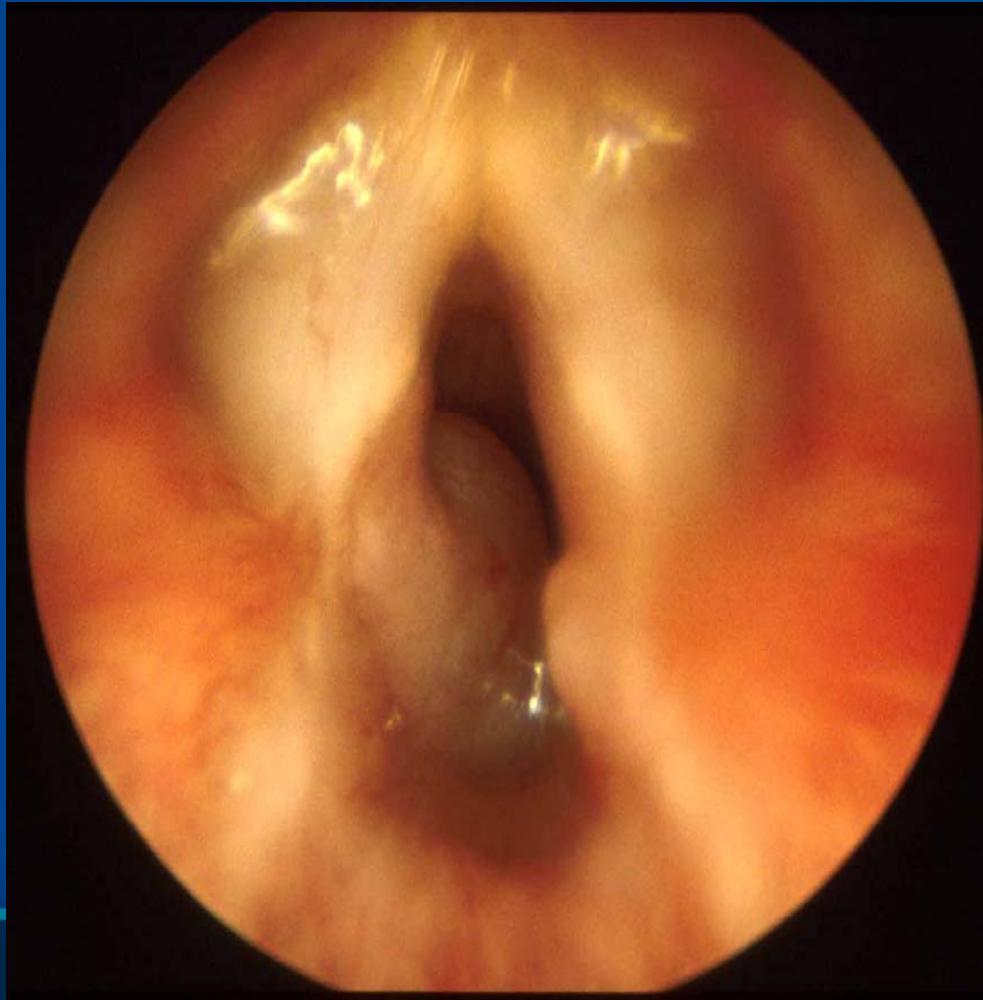
Treatment

- Individualized to each patient depending on degree of stenosis, extension out of the subglottis and the patient's medical condition, swallowing ability, age and weight
- Congenital subglottic stenosis are often less severe than acquired stenosis
- Grade I stenosis (<50% obstruction) can usually be treated with supportive care until sufficient laryngeal growth is spontaneously achieved
- Most patients with >50% obstruction require at least some level of intervention
- Options for surgical management include endoscopic techniques, expansion procedures and partial cricotracheal resection

Subglottic Hemangioma

- Biphasic stridor after 3rd week of life
- 85% present in the first 6 months
- Stridor exacerbated by crying and URTI
- Harsh cry and dyspnea
- Asymmetric subglottic narrowing is the classic finding on soft tissue neck radiographs
- 50% have an associated cutaneous hemangioma
- Natural course of the disease is for growth for 6-18 months followed by gradual regression

Subglottic Hemangioma

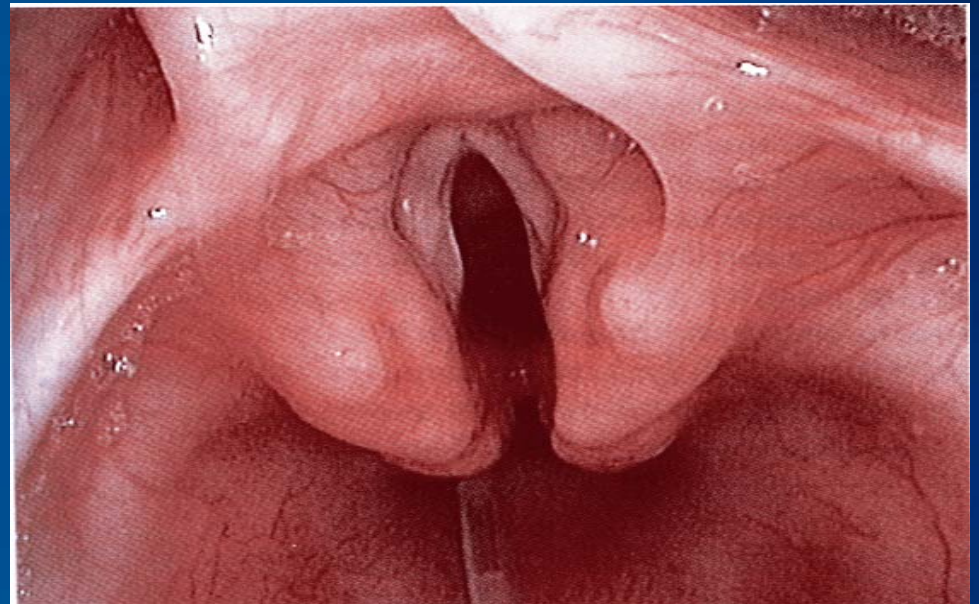


Treatment

- Subglottic hemangiomas are associated with a 30% to 70% mortality rate if left untreated
- Treatment is aimed at maintaining the airway while minimizing potential long-term sequelae of the treatment itself
- Current management options include laser partial excision, open surgical resection, systemic or intralesional steroids, systemic interferon alpha-2A, and tracheotomy

Posterior Laryngeal Cleft

- Failure of rostral development of the tracheoesophageal septum
- Absence of the septum prevents the proper formation of the cricoid cartilage ring
- 30% infant history of maternal polyhydramnios
- 20% with tracheoesophageal fistula



Other Causes of Subglottic Stridor

- G Syndrome (Opitz-Frias)
 - Laryngeal cleft
 - Also includes abnormal facies, hypertelorism, wide anterior fontanelle, low set ears, hypospadias, cleft lip and palate
- Subglottic Web
 - can often mimic a deformity of the cricoid cartilage or subglottic stenosis
 - About 7% of laryngeal webs are in the subglottic region
 - Generally anteriorly based with a small opening posteriorly

CONCLUSION

- Stridor is a symptom of turbulent airflow within the airway
- Further evaluation of the patient is determined based on history and physical exam
- Laryngomalacia is the most common cause of neonatal stridor and is usually treated conservatively
- Acute onset of stridor usually necessitates emergent airway evaluation and management