Congenital Laryngeal Anomalies

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Outline

- Laryngeal Anatomy, Embryology & Function
- Laryngomalacia
- Laryngoceles & Saccular Cyst
- Vocal Cord Paralysis
- Congenital Laryngeal Web & Atresia
- Congenital Subglottic Stenosis
- Laryngeal & laryngotracheoesophageal clefts
- Subglottic Hemangiomas
Laryngeal Anatomy

- Differences in Adults vs Infants
  - 1/3 size at birth
  - Narrow dimensions of subglottis and glottis
    - subglottis is the narrowest (4-5mm in diameter)
  - Higher in the neck
    - C4 at birth vs C6-7 at 15 y/o
  - Epiglottis is narrower
Laryngeal Embryology

- Laryngeal development
  - 3rd week
    - Respiratory primordium is derived from primitive foregut
  - 4th - 5th weeks
    - Tracheoesophageal (TE) septum forms by fusion of (TE) folds
Laryngeal Embryology

- Larynx develops from the 4th & 5th arches
- Primitive laryngeal aditus is T-shaped with 3 eminences
  - Hyobranchial eminence becomes the epiglottis
  - 2nd & 3rd eminence develops into the arytenoids
  - Laryngeal lumen obliterates & then recanalize by the 10th week
Laryngeal Function

- Laryngeal Function
  - Breathing Passage
  - Airway protection
  - Aid in the clearance of secretion
  - Vocalization

- Symptoms of Laryngeal Anomalies
  - Airway obstruction
  - Feeding difficulties
  - Abnormalities of Phonation
Airway Obstruction

- Symptoms
  - Stridor
  - Increase work of breathing with retraction, nasal flaring & tachypnea
  - Apnea episodes, cyanosis & sudden death

- Stridor
  - Inspiratory stridor (Supraglottic & glottic)
    - Collapse during negative inspiratory pressure
  - Biphasic stridor (Subglottic)
  - Expiratory stridor (lower tracheobronchial tree)
Airway protection

- First level - Epiglottis, aryepiglottic folds & arytenoids
- Second level - False vocal folds
- Third level - True vocal folds

Anomalies of any of these structures lead to aspiration and swallowing dysfunction.

- Symptoms - coughing, choking and gagging episodes, stasis of secretion, and recurrent pneumonia
Phonatory abnormality

- Dependent on the level of abnormality
  - Muffled cry suggest supraglottic obstruction
  - High pitch or absent cry is associated with glottic abnormalities
Laryngomalacia

- Most common congenital laryngeal anomaly (50-75%)
- Most frequent cause of stridor in children
- Male predominance 2:1
- Flaccidity of supraglottic laryngeal tissues
- Characterized by inward collapse of supraglottic structures during inspiration
Anatomic Abnormalities

- Epiglottis
  - Long tubular
  - Displaced posteriorly on inspiration
  - Inferior collapse to the vocal folds
- Short aryepiglottic folds
- Inward collapse of aryepiglottic folds (primarily cuneiform cartilages)
- Anteromedial collapse of the arytenoid cartilages
Laryngomalacia Symptoms

- Airway obstruction
  - Mild to moderate obstruction
    - Stridor exacerbated by exertion
      - Crying, agitation, feeding or supine position
  - Severe obstruction
    - Substernal retraction
    - Pectus excavatum with chronic severe obstruction
- Other complications
  - Feeding difficulties
  - GERD
  - Failure to thrive
  - Cyanosis, cardiac failure & death
Stridor in Laryngomalacia

- Inspiratory stridor
  - Intermittent low-pitched
- Starts within first two weeks of birth
- Worsens in the first few months followed by gradual improvement
- Peak at 6 months and most are symptom free by 18 to 24 months (75%)
Laryngomalacia Pathophysiology

- The cause of the collapse is unknown

**Theories**

- Derangement of supraglottic anatomy, histology or neurologic function
- Laryngeal cartilage immaturity
  - Incidence of laryngomalacia is not increased in premature infants
- Histopathology - normal microanatomy
  - Subepithelial edema
Laryngomalacia Pathophysiology

- Neurologic involvement
  - Associated with central apnea, hypotonia, mental retardation and early speech delay
- Abnormal Neuromuscular Control
  - Muscular dilation of supraglottic structures
    - Stylopharyngeus, Palatopharyngeus, Hyoglossus & Digastric
Gastroesophageal reflux

- >50% of patients with laryngomalacia
- Airway edema contributes to airway compromise
- Pathophysiology
  - Increased negative intrathoracic pressure with collapsed supraglottic leads to retrograde gastric contents
  - Edema and/or erythema of posterior supraglottic structures
Diagnosis of Laryngomalacia

- Awake flexible fiberoptic laryngoscopy
  - Visualize supraglottic anatomy and collapse
- Fluoroscopy
- Direct laryngoscopy and bronchoscopy—evaluate for synchronous lesions (27%)
Treatment of Laryngomalacia

- Observation - most cases resolve spontaneously
- Medical management for GERD
- Surgical management - severe symptoms
  - In 1922, Iglauer amputation of epiglottic redundant tissue with a wire snare
  - Supraglottoplasty (CO2 laser, microlaryngeal scissors, microdebrider)
    - Trim redundant tissue from:
      - Lateral edges of the epiglottis
      - Aryepiglottic folds
      - Arytenoids
      - Corniculate cartilages
  - Tracheotomy
Supraglottoplasty

PreOP

PostOP
Laryngomalacia

- Supraglottoplasty complications
  - Aggressive approach
    - supraglottic stenosis
    - exacerbation of dysphagia with aspiration
    - Rare- massive collapse of supraglottic framework needing tracheotomy placement

- Conservative excision minimizes the probability of postoperative complications
Laryngoceles & Saccular Cyst

- **Anatomy**
  - Saccule - cecal pouch of mucous membrane in anterior roof of the laryngeal ventricle
Laryngoceles

- Dilation or herniation of the saccule
- **Communicates** with the lumen of the larynx
- Filled by air or mucous
- Internal - extend posterosuperior into the aryepiglottic fold
- External - protrude through the thyrohyoid membrane
- Combined - External + internal
Saccular Cyst

- Congenital cyst of the larynx or laryngeal mucocele
  - **No communication** with the laryngeal lumen
  - Filled with mucous (no air)
  - Developmental- failure to maintain patency of the saccular orifice

- Two types
  - Anterior saccular cyst-
    - Protrudes into the ventricle
  - Lateral saccular cyst
    - extends into the false vocal cords and aryepiglottic folds
Laryngoceles & Saccular Cyst

- Acquired Laryngoceles
  - Increased pressure on the laryngeal lumen (player of wind instruments)

- Acquired saccular cyst
  - Occlusion of the saccular orifice (inflammation, trauma or tumors)

- Laryngopyocele
  - Infected laryngocele or saccular cyst
Symptoms

- Laryngocele
  - Intermittent hoarseness and dyspnea
  - Weak cry or aphonia

- Saccular cyst
  - Respiratory distress with inspiratory stridor
  - Inaudible or muffle cry
  - Occasionally dysphagia
Laryngoceles & Saccular Cyst

- Diagnosis
  - Flexible & rigid laryngoscopy
  - Soft tissue neck X-ray (distended with air)
  - Combined laryngcele-mass protrudes with Valsalva maneuver
- Saccular cyst- Needle aspiration confirms the diagnosis.
Laryngoceles & Saccular Cyst

- **Treatment**
  - Saccular cyst - aspiration or unroofing with cup forceps or CO$_2$ laser (recurs)
  - Endoscopic excision
    - Removing remnants CO$_2$ laser
  - Open procedures for recurrence
    - Lateral cervical approach incising the thyrohyoid membrane
    - Protect the superior laryngeal nerve
  - Intubation may be needed until edema subsides
Laser Excision of Anterior Saccular Cyst
Vocal Cord Paralysis

- Third most common congenital laryngeal anomaly producing stridor
- Unilateral & Bilateral (1:1)
- 50% are associated to other anomalies
- Acquired paralysis
  - 70% association to congenital neurologic abnormalities or neurosurgical procedure to treat them
    - (Meningocele, Arnold Chiari Malformation and Hydrocephalus)
  - Unilateral are associated to cardiovascular anomalies (PDA) and left side is more common
Vocal Cord Paralysis

- Symptoms
  - Bilateral
    - High-pitched inspiratory stridor
    - Inspiratory cry
    - Paradoxical function (pressure changes)
      - close during inspiration and open during expiration
  - Unilateral (less symptoms)
    - weak cry and occasional breathy
    - Feeding difficulties secondary to laryngeal penetration and aspiration
Vocal Cord Paralysis Diagnosis

- Awake flexible fiberoptic laryngoscopy
  - record for slow motion replay
- Direct laryngoscopy
  - Palpation of the glottis
  - Laryngeal EMG
- Imaging of head (MRI) and chest to evaluate for associated abnormalities (Neurologic & CV)
Unilateral VC Paralysis Treatment

- Watchful waiting
  - 70% of idiopathic unilateral VC paralysis resolve spontaneously
  - Most within 6 month
  - Feeding difficulties manage by thickening of liquids
  - Speech therapy consult
  - Rare surgical management

- Increased Intracranial Pressure
  - early shunting or posterior fossa decompression (better outcome)
Bilateral VC Paralysis Treatment

- Tracheotomy may be necessary (50%)
- Lateralizing one or both paralyzed vocal cords
  - Injurious to the developing larynx
- Excisional procedure
  - Tissue removed from posterior glottis
    - Endoscopic technique with laser
    - More consistent results are achieved by external approach
Congenital Laryngeal Web-Atresia

- Uncommon
- Failure of laryngeal recanalization
- Most are glottic (75%)

Symptoms
- Vocal dysfunction
  - Hoarseness
  - Aphonia if severe
- Airway obstruction

Complete laryngeal atresia is incompatible with life and need emergent tracheostomy
Laryngeal Web Diagnosis

- Flexible laryngoscopy
- Direct Laryngoscopy
- Airway films if subglottic or cricoid pathology are present
Treatment

- Thin anterior glottic web
  - Incision or dilation
- More significant glottic lesion
  - Incision and dilation with possible revision
- >75% glottic involvement and significant subglottic extension
  - Tracheotomy soon after birth
- Subglottic involvement is usually accompanied by anterior cricoid plate abnormality
  - External approach with division of the web and the cricoid plate
Congenital Subglottic Stenosis

- Second most common cause of stridor
  - in neonates, infants and children
- Incomplete laryngeal lumen recanalization
- Newborn larynx <4 mm (premature <3mm)
- Congenital less severe than acquired
- Two types
  - Membranous vs cartilagenous
Membranous Stenosis

- Circumferential & soft
- Less severe than cartilagenous
- Submucosal changes
  - Increased fibrous connective tissue layer
  - Mucous gland hyperplasia
Cartilagenous Subglottic Stenosis

- Cricoid thickening and deformation
  - Flattened cricoid
    - Smaller anteroposterior diameter
  - Elliptical appearance
    - smaller transverse diameter
    - Associated with laryngeal cleft
Congenital Subglottic Stenosis

- Symptoms
  - Upper airway obstruction predominate
  - Inspiratory stridor with progression to biphasic
    - Agitation worsens (increased air flow)
  - Mild to moderate are asymptomatic
    - URI lead to edema and symptoms of croup
    - History of recurrent or prolonged croup
  - Severe obstruction
    - Respiratory distress
    - Intubation may be needed
Congenital Subglottic Stenosis

- **Diagnosis**
  - DL & Bronch
    - Visualize the entire larynx
    - Distinction of membranous vs cartilagenous
    - Synchronous lesions
  - Measurement of the stenosis
    - ET tube placement at sequential size
Congenital Subglottic Stenosis

- **Classification**
  - Grade I < 50% obstruction
  - Grade II 51-70% obstruction
  - Grade III 71-99% obstruction
  - Grade IV no detectable lumen
Congenital Subglottic Stenosis

- Treatment of Grade I
  - Watchful waiting for growth
- >50% obstruction may require some intervention
- Soft tissue acquired lesions
  - Dilation & laser (CO$_2$ & KTP) are sometimes effective
- Most congenital stenosis are cartilagenous
  - Laser & dilation are not useful
Congenital Subglottic Stenosis

- Grade II-III treatment
  - Multiple failed extubation
  - Tracheostomy may be needed
    - Until cricoid grows for decannulation
  - Anterior cricoid split
    - Successful extubation in 66-78%
    - Decannulation rate 75-78%
Anterior Cricoid Split

- Horizontal skin incision over cricoid
- Vertical midline incision
  - Entire cricoid
  - First two tracheal rings
  - Lower 1/3 of thyroid cartilage
  - ET tube visualize
- Two Prolene sutures on each side of incised cricoid
- Intubated for 7-14 days (stenting)
Congenital Subglottic Stenosis

Grade III treatment
- Laryngotracheal decompression
  - Anterior, posterior and possible lateral
- Reconstruction
  - Costal cartilage
- Long term stenting 2-4 wks

Grade IV & few grade III
- Partial cricotracheal resection
Laryngeal & Larygotracheo-esophageal Clefts

- Rare, incidence of <0.1%
- Incomplete development of TE septum
- Communication of posterior larynx and esophagus
- Strong association with other anomalies (56%)
  - TE fistula in 25%
Laryngeal & larygoesophageal Clefts

- Laryngeal Clefting
  - Interarytenoids only
  - Partial or complete cricoid
- Laryngotracheochoesophageal clefts
  - Cervical or intrathoracic trachea
Laryngeal & Laryngotracheoesophageal Clefts

- Symptoms
  - Proportional to the length
  - Can be asymptomatic (minor)
  - Inspiratory stridor
  - Feeding problems aspiration
  - Cyanotic episodes
  - Recurrent pneumonia
Laryngeal & Laryngotracheoesophageal Clefts

- **Diagnosis**
  - CXR- pneumonia
  - Barium swallow- contrast spill over into the trachea
  - Direct laryngoscopy best single test
    - Observe and palpate the interarytenoid area
    - Relationship to the vocal cords
Laryngeal & Laryngotracheoesophageal Clefts

**Treatment**

- Supraglottic larynx
  - Conservative management
  - Swallowing therapy to prevent aspiration
  - GERD evaluation and treatment
  - Surgical approach
    - 80% success rate with Endoscopic repair

- Extension below the vocal cords
  - Surgical repair is required
Mortality

- Laryngeal clefts, rate of 11% and 46%
  - other anomalies
  - Delay in diagnosis
- Intrathoracic laryngotracheoesophageal is as high as 93%
Subglottic Hemangiomas

- Benign vascular malformations
- Histological- endothelial hyperplasia
- Female predominance 2:1
- Asymptomatic at birth
  - Stridor presents by 6 months (85%)
- Associated cutaneous hemangioma (50%)
Subglottic Hemangiomas

- Rapid growth phase in the 1st year followed by slow resolution
- Most have complete resolution by 5 years
- 30-70% mortality rate if untreated
- Priority is to maintain the airway while minimizing potential long term sequelae
Subglottic Hemangiomas

- **Diagnosis**
  - Direct Laryngoscopy
    - Compressible
    - Asymmetric, usually posterolateral
    - Bluish or reddish discoloration
  - CT & MRI
Treatment of Subglottic hemangiomas

- Systemic steroids (principal)
  - Partial regression in most patients (82-97%)
  - Risk of growth retardation and increase susceptibility to infection
  - Risk is reduced by alternate-day dosing regimen in the smallest doses
  - Also intraleision corticosteroids has been employed with successful avoidance of tracheotomy

- Interferon alpha-2a
  - 50% or greater regression of lesion in 73% of patients
  - It requires prolonged therapy, blocks various steps of angiogenesis
  - Side effects neuromuscular impairment, skin slough, fever and liver enzyme elevation
Treatment of Subglottic hemangiomas

- Tracheotomy
  - Bypass the obstructing lesion
  - Waiting for the expected involution
  - Risks of tracheostomy as well as delay in speech and language

- Laser CO2 and KTP
  - Associated with a significant risk of inducing subglottic stenosis in up to 20%
Treatment of Subglottic Hemangiomas

- Surgical excision
  - Decannulation shortly after surgery
  - Avoiding tracheostomy in 85% of patients
  - Laryngeal distortion or damage