Congenital Laryngeal Anomalies

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Introduction

- Wide range of problems
- Anatomy
- Embryology
- Diagnosis

Types: supraglottis, glottis, subglottis

- Presentation/diagnosis
- Management
Normal Anatomy

- Larynx
  - Ventilates and protects lungs
  - Clears secretions
  - Voice

- Differences in adults and infants
  - 1/3 size at birth
  - Narrow dimensions (subglottis vs. glottis)
  - Higher in neck and more pliable
  - Epiglottis narrower
Embryology

- Respiratory primordium
  - Third week – 26 days
- Respiratory primordium separated by tracheoesophageal folds
  - Fuse to form septum (4-5 weeks)
Embryology

- Larynx from 4\textsuperscript{th} and 5\textsuperscript{th} arches
- Primitive larynx altered by hypobranchial eminence, epiglottis, arytenoids
- Laryngeal lumen obliterated and recanalized
Clinical Manifestations

- Respiratory obstruction
- Stridor
- Weak cry
- Dyspnea
- Tachypnea
- Aspiration
- Cyanosis
- Sudden death
Clinical Diagnosis

- History
  - Premature, medical problems
  - Birth records, intubation history
  - Symptom frequency, feeding

- Physical exam
  - Observation
  - Voice
  - Flexible exam
Clinical Diagnosis

- Radiography
  - Neck films, chest films
  - Barium swallow
  - CT/MRI
- Endoscopy in OR
  - Gold standard
Supraglottic Anomalies

- Laryngomalacia
  - Most common (60%)
  - Boys > girls
  - Inspiratory stridor: *not always at birth
  - Benign, self-limiting
  - May be severe
  - Immature larynx
Laryngomalacia

- Diagnosis: flexible laryngoscopy
- Occasional endoscopy
- Treatment = expectant, reassurance
  - Position changes
  - Close follow up
- Severe cases = surgery
Supraglottic Anomalies
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Supraglottic Anomalies

- Results of supraglottoplasty
  - Largest series: 53% complete regression
    - 2 needed tracheotomy, pharyngomalacia
    - ?BiPAP
  - Other studies: 77-100%
  - Complications: stenosis, CA fixation, PGS
  - GERD association?
Supraglottic Anomalies

- Saccular cysts
  - Similar to laryngoceles
  - Filled with mucous
  - May need immediate trach/intubation*
  - Endoscopically vs. open
Supraglottic Anomalies

- Laryngofissure
- External approaches
- Recurrences if entire cyst not removed
Supraglottic Anomalies

- Laryngocele
  - Dilated sac filled with air (ventricle)
  - Internal vs. external
  - May present at birth—stridor*
  - Difficult to diagnose—CT?
  - Endoscopic or open procedures
  - Recurrences low
Supraglottic Anomalies

- Vascular and lymphatic malformations
  - Hemangiomas
    - 30% birth—grow in first 6-18 months
    - Dyspnea, stridor, feeding problems later*
    - Endoscopic evaluation
    - Multiple treatment options
  - Lymphangiomas
    - Compress epiglottis—airway distress at birth*
    - Symptoms varied
    - Endoscopic evaluation: CO2 laser
Supraglottic Anomalies
Supraglottic Anomalies

- Supraglottic webs – rare
- Anomalous cuneiform cartilage
- Bifid epiglottis
  - Pallister-Hall syndrome (hypothalmus, polydactaly, laryngeal)
Glottic Anomalies

- Laryngeal webs
  - Failure of recanalization of larynx
  - 75% at glottic level
  - Most anterior with subglottic involvement
  - Four types—increasing severity
  - May present at birth*
  - Diagnosis: flexible laryngoscopy
    - Airway films helpful with subglottis
Glottic Anomalies
Glottic Anomalies

- Treatment dependent on type and symptoms
- Simple division
- Local flaps
- Staged dilations
- Endoscopic or open keel insertion
Glottic Anomalies
Glottic Anomalies

- Laryngeal Atresia
  - Most severe process from failed recanalization
  - Always present at birth*
  - Only survive if TEF or immediate trach
  - Later LTR
  - Other anomalies
Glottic Anomalies
Glottic Anomalies

- Congenital High Upper Airway Obstruction (CHAOS)
  - 1994—ultrasound with large lungs, flat diaphragms, dilated airways, fetal ascites
  - EXIT procedure (ex utero intrapartum treatment)
- Multidisciplinary team
  - C-section, maintain placental blood flow, quick tracheotomy
Glottic Anomalies

- Vocal cord paralysis
  - Second most common cause of stridor
  - 10-15% of laryngeal pathology
  - Unilateral vs. bilateral
  - Vagus nerve damage
  - Idiopathic (47%)
  - ACM, hydrocephalus, trauma, cardiac problems
Glottic Anomalies

- Vocal cord paralysis
  - Poor cough, aspiration, pneumonia
  - Cry or voice (?normal)
  - Stridor most common
  - Airway control imperative
    - History and PE
    - Flexible laryngoscopy
    - Airway films, U/S, barium swallow, CT/MRI, endoscopy
Glottic Anomalies

- Bilateral vocal cord paralysis
  - Tracheotomy in 50%
  - Present at birth*
  - ACM—posterior fossa decompression/shunt
  - Serial endoscopy/EMG
  - 60% return with ACM
  - If not, lateralization procedures (over one year)—Woodman arytenoidectomy, laser cordotomy/arytenoidectomy/cordectomy, open procedures, reanimation, electrical pacers
Glottic Anomalies
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Glottic Anomalies

- Unilateral TVC paralysis
  - Less urgent
  - Do not present at birth usually
  - Weak cry, airway adequate
  - Speech therapy
  - Thyroplasty?
Subglottic Anomalies

- **Subglottic hemangioma**
  - Congenital vascular lesion—variable symptoms
  - 30% at birth—most in 6 weeks-18 months
  - Growth phase, involution phase
  - Biphasic stridor* later
  - Cutaneous involvement (50%)
Subglottic Anomalies

- Diagnosis
  - History, PE
  - Radiographs
  - Rigid endoscopy
    - Compressible, blue-red mass, posterior-lateral wall of subglottis
Subglottic Anomalies
Subglottic Anomalies

- Subglottic hemangioma
  - Tracheotomy
  - Laser ablation— CO2 vs. KTP
  - EBR, cryotherapy, sclerosing agents
  - Corticosteroids
  - Open excision
Subglottic Anomalies

- Posterior laryngeal cleft
  - Failure of tracheoesophageal septum development (rostral portion)
  - 6% with TEF have PLC
  - Pallister-Hall syndrome
  - May present at birth*
  - Respiratory distress with feeds, cyanosis
  - Aspiration, pneumonia, death
Subglottic Anomalies

- Posterior laryngeal cleft
  - Chest radiographs
  - Barium swallow
  - Endoscopy important
    - Relationship of cleft to cricoid
    - Four types
Subglottic Anomalies
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Subglottic Anomalies

- Posterior laryngeal clefts
  - GERD control
  - Endoscopic, open (2 layer closure)
  - Sternotomy
  - Overall mortality 43%
  - Type IV clefts: 93% mortality
Subglottic Anomalies

- Subglottic stenosis
  - Acquired or congenital
  - Failure of laryngeal lumen to recanalize
  - Membranous vs. cartilaginous
  - Other anomalies
  - Less than 4.0 mm (3.5 mm)
Subglottic Anomalies

- Subglottic stenosis
  - Respiratory distress at delivery to recurrent croup
  - Usually not at birth*
- History and PE (biphasic stridor)
- Endoscopy
  - Cotton grading system

<table>
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<th>Classification</th>
<th>From</th>
<th>To</th>
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<tr>
<td>Grade I</td>
<td>No Obstruction</td>
<td>50% Obstruction</td>
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<tr>
<td>Grade II</td>
<td>51% Obstruction</td>
<td>70% Obstruction</td>
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<td>Grade III</td>
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<td>Grade IV</td>
<td>No Detectable Lumen</td>
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Subglottic Anomalies

- Subglottic stenosis
  - Most conservative*
  - Dilation or laser not useful
Subglottic Anomalies

- Subglottic stenosis
  - ACS
  - Ant split with cartilage
  - Ant/post split with cartilage
  - Four quadrant split
  - Cricotracheal resection