Neonatal Stridor

Neonatology 2015

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Disclosures

• None
Goals/Objectives

• Describe embryology related to laryngeal development
• Define stridor and distinguish it from stertor
• Use the type of stridor to narrow differential in a neonate
• List and describe with the more common causes of stidor in the neonate
Embryology\textsuperscript{1,2}

- Larynx, trachea, bronchi, and lungs are derived from the foregut ventromedial diverticulum called the tracheobronchial groove
  - Day 20: primitive foregut identifiable
  - Day 22: development of laryngotracheal sulcus
  - Day 24: primitive lung buds
  - Day 28: primitive esophagus and trachea divide; caudal to cranial fusion of the tracheoesophageal septum (defect may cause TEF or laryngeal cleft)

- Separation in development of the GI and respiratory tracts
Laryngeal embryology

http://embryology4genius.weebly.com/development-of-larynx.html
Embryology Cont.

• Day 32 – mesenchymal arytenoid swellings appear from 6th arch
• Gestational week 8: Obliteration of laryngeal lumen occurs
• Gestational week 10: recanalization of the laryngeal lumen (webs, stenosis, and atresia can occur if not proper)
• Splanchnic mesenchyme creates the cartilage, connective tissue, and muscle of the trachea
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Stridor

- Unusual, extrathoracic sound heard from the airway during breathing related to airway obstruction
- Pitch related to degree of airway obstruction and velocity of flow
- Usually relates to supraglottic structures or below
- When related to the oropharynx, nasopharynx, or nasal cavity, the term stertor or even snoring is the preferred term
My rough guide…

- Stertor

- Stridor
In either case...

- Noise from partial airway obstruction with turbulent flow
- The obstruction can be fixed or variable
- Inspiration → extrathoracic pressure is negative relative to atmospheric pressure → supraglottic collapse; vice versa
- Biphasic stridor is likely related to either a fixed obstruction or two-level obstruction
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Sometimes the cause is obvious
Causes of Stertor

• Nasal obstructions (choanal atresia, pyriform aperture stenosis, nasolacrimal duct cysts)
  – Obligate nasal breathers

• Oral cavity/oropharyngeal obstructions (macroglossia [Beckwith Widemann], glossoptosis, retrognathia/micrognathia, dermoid cyst, thyroglossal duct cyst)
Narrowing the differential

• Symptomatology can help to differentiate
  – Inspiratory: at or above the vocal cords
  – Expiratory: below the vocal cords
  – Biphasic: at or below the vocal cords
History

• From birth or within the first few wks of life
• Association with position
• Changes during feeding?
• Intermittent v continuous
• Abnormal phonation
• Other congenital abnormalities
Stridor Causes

• Supraglottic
  – Laryngomalacia

• Glottic
  – Vocal cord paralysis
  – Webs

• Subglottic
  – Stenosis
  – tracheomalacia
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Laryngomalacia

• Most common cause of stridor in children
• Usually not present at birth but develops during the first few weeks
• Due to collapse of supraglottic structures during inspiration
  – Infolding of the epiglottis (omega-shaped)
  – Shortened aryepiglottic folds
  – Redundant arytenoid mucosa
• Feeding difficulties may be associated and GERD often present
• May be associated with cyanotic spells or ALTEs which are relieved when prone
Surgery for laryngomalacia

- Only 10% or less of patients require surgery
- Most grow out of the problem by 18 to 24 months of age
- Conservative management usually includes management of reflux, prone or elevated positioning (with apnea monitor), and increasing calories
- Indications for surgery include failure to thrive and cyanotic spells/obstructive events
- May be an underlying neurologic condition
Laryngomalacia
Increasing severity
Before and After
Risks with surgery

• Failure to resolve the condition/need for repeated procedures
• Supraglottic stenosis/posterior glottic stenosis
• Aspiration (usually transient)
Laryngeal Clefts

- Uncommon (1:10-20,000)
- Associated with tracheomalacia, certain syndromes, tracheoesophageal fistulas, and other defects
- Suffer recurrent aspiration
- Repair endoscopically if accessible, otherwise open surgical procedure which may require bypass
Benjamin-Inglis Classification
Type II laryngeal cleft
Repair
Saccular Cysts

• Saccular Cysts
  – Mucus filled cyst due to the obstruction of the ventricle causing airway obstruction
  – Bstruct larynx
  – Decompress endoscopically or externally
Vallecular Cysts

- Vallecular Cysts
  - Mucus retention cyst found at the base of the epiglottis
  - Present with progressive airway obstruction
  - Emergency decompression if necessary then excision
Vallecular cyst
After resection
Postop view a few wks later
Vocal Fold Immobility

- Second most common cause of neonatal stridor
- 5-10% of tracheostomies are done for this reason
- Unilateral paralysis may be noticed at birth due to weak cry/later due to limited laryngeal obstruction with stridor only during episodes of distress/aspiration
- Bilateral paralysis with significant stridor and airway obstruction
Vocal Fold Immobility

- Diagnosis made by fiberoptic laryngoscopy
- Insult may occur anywhere from laryngeal motor nuclei to intrinsic laryngeal musculature
- Investigation with direct laryngoscopy and bronchoscopy may be helpful to rule out anatomic causes of VC immobility
- With bilateral paralysis, imaging head, neck and chest usually recommended (12-30% neurogenic)
Course of vagus

Diagram showing the course of the vagus nerve, including its division into cranial and spinal nuclei and various branches such as the carotid body and sinus, common carotid artery, recurrent laryngeal nerve, superior laryngeal nerve, and pharyngeal nerve.
Vagus and RLN

Yau. Laryngeal Nerve Anatomy
Unilateral vocal cord paralysis
Chiari malformation
Vocal Cord Paralysis

• Treatment depends on symptoms and cause
• Vocal cord function may return in a matter of weeks, months or years depending on etiology
• Bilateral VC paralysis usually requires a tracheotomy
  – Daniel has demonstrated avoidance of tracheostomy in a few patients using Botox to the cricothyroid muscles
  – Often wait for return of laryngeal function
  – Other endoscopic adjunct procedures include cordotomy, arytenoidectomy, lateralization
Laryngeal Webs

- **Laryngeal Web**
  - Represents a failure of airway recanalization
  - Associated with VCFS
  - May be isolated anomaly, or have cardiac and distal airway problems
  - There are 4 grades with progressive airway narrowing, with grade 4 representing complete laryngeal atresia often with associated TEF
Laryngeal webs (cont)

• Cohen classification:
  – I: thin glottic web without subglottic extension (<35% airway obstruction)
  – II: thicker web with minimal subglottic extension (35-50% airway obstruction)
  – III: Solid web with subglottic involvement 50-75%
  – IV: Solid web with subglottic involvement and stenosis (75-90%)
Laryngeal Webs
Keel
Subglottic stenosis

• Third most common laryngeal abnormality
• Difficult to discern whether it is congenital or acquired (5% congenital of all SGS)
• Present as persistent or worsening stridor, recurrent croup, or inability to extubate
• First diagnostic exercise is flexible fiberoptic laryngoscopy
• Further investigation with diagnostic bronchoscopy
  – Newborn subglottis=4mm
  – Premature subglottis=3mm
# Myers-Cotton Grading of SGS

<table>
<thead>
<tr>
<th>Classification</th>
<th>From</th>
<th>To</th>
</tr>
</thead>
<tbody>
<tr>
<td>Grade I</td>
<td>No Obstruction</td>
<td>50% Obstruction</td>
</tr>
<tr>
<td>Grade II</td>
<td>51% Obstruction</td>
<td>70% Obstruction</td>
</tr>
<tr>
<td>Grade III</td>
<td>71% Obstruction</td>
<td>99% Obstruction</td>
</tr>
<tr>
<td>Grade IV</td>
<td>No Detectable Lumen</td>
<td></td>
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Management

• ABCs: evaluate and establish airway if necessary
• If intubation is necessary, would be good to do bronchoscopy or intubate over a telescope to evaluate; smaller tubes may be needed is SGS is suspected
• Heliox, nebulized racemic epinephrine and humidified oxygen can be used as adjuvant or temporizing treatments
• Tracheostomy when all other means of establishing airway are unsuccessful
Management

• Characterize the type of stenosis and length
• Grade I and II may be observed or more amenable to endoscopic approaches
• Therapeutic Options
  – Trach necessary for severe narrowing
  – Dilation
  – CO2 laser/cold steel
  – Endoscopic posterior cricoid split with cartilage graft +/- anterior split
  – Open
Open surgery

• Resection of stenosis (need 4-5 mm of healthy airway below cords)
• Airway expansion (laryngotracheoplasty with cartilage grafts) – more effective in young children/milder stenosis and that extending to cords (costal cartilage, auricular, and thyroid ala)
• Single stage (use of ETT as stent for 7-14 days) v. double stage (suprasomal stent) – can depend on ICU
Mild subglottic stenosis
Balloon dilatation
Afterwards
Grade III subglottic stenosis
Intubation Consequences

• Can occur even after short times of intubation
• Results in fronds of granulation tissue or furrows in the mucosa over the arytenoids
• Intubation may also cause mucosal retention cysts
• Fusion of concentric narrowing (SGS), posterior scarring (posterior glottic stenosis), or laryngeal webbing
Intubation Complications
How Do You Avoid This?

• Always use the smallest tube possible
• Do not use a cuff unless absolutely necessary
• Manage any reflux disease aggressively
• Recognize that some patient may have smaller subglottic area (ie Down syndrome)
• If there is no leak with a tube, it is too big, even for a short period of time
An example
Subglottis
Trachea
Carina
Anterior grafting
Cartilage Grafting of SGS
Tracheal resection

T = Tube; HT = High frequency jet ventilation tube
Tracheal Resection
Subglottic Hemangioma

• Natural history is progression from mild inspiratory stridor to airway obstruction (most growth between 4-6 months of life)
• Usually an eccentric lesion in the airway
• Screen if there are beard-distribution hemangiomas
• Treatment
  – Propranolol now standard of care
  – Steroids
  – CO2 laser ablation
  – Open surgical excision
  – Tracheotomy
Subglottic Hemangioma
Tracheomalacia

- Commonly diagnosed condition, but may be primary or secondary
- Normal ratio of trachea is 4.5-5:1 cartilage to pars membranosa
- Best diagnosed using bronchoscopy
- Severity based on degree of compression
  - Type I <70%
  - Type II 70-90%
  - Type III >90%
- Symptoms range from dying spells to seal-like cough
Tracheomalacia

- Treatment based on related anatomy
- Often associated with other airway pathology (TEF, laryngeal cleft)
- Must address extrinsic compression
- If intrinsic or unable to address compression trach may be necessary
- Most children will become asymptomatic after 2
- May need customized trach with appropriate length
Tracheomalacia
Bronchomalacia

- Abnormal collapsability of the bronchus
- Must determine if intrinsic or extrinsic
- Presentation may be with recurrent pneumonias with persistent secretions
- If severe enough may cause respiratory compromise and air trapping
- Treatment
  - CPAP
  - Trach
  - Stent
Endobronchial stenting

Bronchial stenting

Trachea

Tumour

Stent

Bronchus
3-D printer external stent

Courtesy University of Michigan Health
Innominate Artery Compression

- Most common vascular compression
- Can be diagnosed on bronchoscopy by placing pulse oximeter on right hand and occluding innominate
- If symptoms are recurrent or severe, aortapexy may be indicated
- Bronchoscopy at time of aortapexy ensures relief of obstruction
Double Aortic Arch

- Complete vascular ring encircles the trachea and esophagus
- Ascending aorta bifurcates and rejoins distal aorta
- Results from persistent right ventral fourth arch
- Must divide non-dominant branch
- Will have some persistent tracheomalacia
Double Aortic Arch
Posterior Tracheal Compression

• Pulmonary Artery Sling
  – Associated with complete tracheal rings
  – Right pulmonary artery passes between trachea and esophagus
  – Only cause of posterior tracheal pulsation

• Retro-esophageal Subclavian
  – Right subclavian artery passes behind esophagus
  – Causes dysphagia lusorium
  – Diagnose on barium swallow
  – Incidence 1:300
Posterior Tracheal Compression
Complete Tracheal Rings

• Tracheal cartilages are usually 4-5:1 ratio with pars membranosa
• Normally the only complete cartilagenous ring is the cricoid
• Also known as the “stove pipe” trachea
• Often associated with pulmonary artery sling
• May be associated with pulmonary agenesis
Tracheal Rings
Surgical Repair of Tracheal Rings
Summary
Key Points

• Distinguishing between stridor and stertor can help to localize problem and where to go
• Characterizing the type of stridor goes a long way with diagnosis
• Flexible endoscopy is usually used as first modality usually
• Imaging may be helpful in defining the extent of disease or identifying the cause of the problem
References