The Squeaky Baby
A Pediatric Provider’s Guide to Stridor in the Young Child
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Disclosure Statement
• I have no actual or potential conflict of interest in relation to this program.

Learning Objectives
• Understand the clinical approach to the young child with stridor
• Become more familiar with laryngeal pathologies contributing to pediatric stridor and respiratory distress

The infant with Respiratory Distress: Levels of pathology
• Nose/Nasopharynx – obligate nasal breathers for 6 weeks
  – Pyriform aperture stenosis, choanal atresia, rhinitis of infancy
• Oral Cavity/Oropharynx
  – macroglossia, micrognathia, vascular cyst, abscess, mass (lymphatic malformation, teratoma, etc.)
• Larynx
  – Laryngomalacia, RAP, vocal cord paralysi, laryngeal web/iatresia, sacrocyetal cyst, subglottic cyst, subglottic stenosis, laryngeal fist, paresap
• Trachea/Bronchi
  – Tracheomalacia, stenosis/complex tracheal rings, External Compression: Vascular Rings/Anomalies, bronchiolitis, Foreign body
• Heart/Lungs
  – Bronchopulmonary dysplasia, congenital diaphragmatic hernia, pneumonia, heart failure

The Larynx in Action

Laryngeal Anatomy
Stridor Versus Stertor

- **Stridor**: Sometimes harsh, high-pitched, musical sound produced by turbulent airflow through partially obstructed airway
  - Inspiratory / Expiratory / Biphasic

- **Stertor**: Low-pitched inspiratory sound from nose/pharynx (e.g. snoring)

Physics Review

- What determines the pitch of a guitar string?
  - Resonant frequency: based on the length, mass, and tension of the string

- Stridor happens at the level of the larynx and subglottis because the tissues are shorter, smaller and tighter than higher up (compare with snoring – stertor)

So why are little children so much more prone to stridor and respiratory distress than older kids and adults?

Radius and Resistance

- **Resistance**: 16x
- **Resistance**: 3x

Assessment: Type of stridor

- **Inspiratory** stridor: high-pitched
  - Supraglottic
  - Supraglottic

- **Biphasic** stridor: intermediate pitch
  - Glottis & Subglottis
  - Inspiratory component usually louder

- **Expiratory** "stridor": lower pitched
  - Wheeze or rattle – not really stridor
  - Lower cervical or Intrathoracic trachea/bronchi

Why Inspiratory/Expiratory?

Based on Differences in Surrounding Pressure

Cervical Obstruction

Thoracic Obstruction
This 2 month old patient presents to your office

Clinical Assessment
1. Assess acuity! Is this evolving rapidly?
   - Fever
   - Drooling (new onset)
   - Rapid change in cry or voice
   - New food aversion
   - New retractions/nasal flaring
   - Fatigue

Clinical Assessment
1. Assess acuity! Is this evolving rapidly?
2. Obtain Airway History/chronicity
   - Acute/Chronic? Progressive/Static/Fluctuating?
   - Prior respiratory problems or events?
     - ALTEs, Perinatal cyanosis, cyanotic spells
     - aspiration pneumonias
     - "croup"-like episodes
   - History of intubation
     - Difficult or emergent? #? Length of time intubated?
   - Other Relevant PMH
     - Prematurity, Heart, Lung, Neuro, Genetic

Clinical Assessment
1. Assess acuity! Is this evolving rapidly?
2. Obtain Airway History/chronicity
3. Review associated symptoms
   - Feeding adequacy and diet
     - Failure to thrive
   - Growth – weight AND vertical
   - Choking episodes
     - Frequency, Severity, Timing, Consistency of food causing choking?
     - Frequent spitting up/reflux history

Physical Exam
- Coloring (cyanosis), obtain O2 sat if concerning or acute onset
- Stridor/Stertor, timing
- Voice quality – strength, hoarseness
- Hold the baby and assess effect of positioning
- Work of Breathing
  - Does the child appear uncomfortable or restless?
  - Retractions: Suprasternal tugging, substernal/subcostal
- Weight/build – evidence of FTT?
- Consider watching the child feed
- Heart and Lungs

Back to our Patient
1. Assess acuity! Is this evolving rapidly?
   - Present since birth, maybe getting a bit louder over time, not otherwise ill.
2. Obtain Airway History/chronicity
   - No respiratory events, ALTEs, etc., but parents worry a bit about the noise. No intubations, uncomplicated term birth. No other medical Hx.
3. Review associated symptoms
   - Feeding seems to go fine, weight gain is good, but does seem to spit up a lot.
   - No obvious choking.
4. Physical Exam
   - High-pitched, inspiratory stridor with a crescendo pattern, comfortable, no respiratory distress, increased when supine
Laryngomalacia

- Most Common cause of inspiratory stridor in a newborn
- Sx: Classic high-pitched inspiratory stridor – "goose honk"
  - often worse during crying, feeding, or when supine
  - may have frequent regurgitation or aspiration (impaired coordination of breathing/swallowing during feeding)
- Natural Hx: usually noticed within few weeks of birth, may worsen for up to ~ 9 mo, then gradual improvement, usually resolved by 12-24 months (occasionally takes longer).

Laryngomalacia, cont’d

- Etiology: likely mixed - delayed development of laryngeal tone, and underdevelopment of supraglottic structures
  - hypotonia leads to collapse with inspiration
  - Can occasionally be seen in older children with cerebral palsy
- Dx by Hx and awake fiberoptic exam: collapse of supraglottic tissue into the airway

Laryngomalacia - Medical Treatment

- 90% of infants with laryngomalacia will outgrow it by age 2 without surgical intervention
- Monitor feeding, weight gain, and work of breathing.
- Side sleeping or sleeping propped up may be of benefit
- Reflux control
  - Very common in infants with laryngomalacia and both entities tend to make the other worse
    - Start with ranitidine 2-4 mg/kg PO BID

When to Refer: Survey of CMH ENTs

- Sometimes okay for PCP to watch? 7 Yes, 2 No
- Provider Should Refer to ENT if:
  - Required in ~10% of cases
  - Indications:
    - Recurrent ALTEs/cyanotic events
    - Recurrent ER visits
    - Poor response to medical therapy, poor weight gain/feeding
    - Persistent parental anxiety?
  - Microlaryngoscopy/bronchoscopy with Supraglottoplasty is first line surgical therapy

Surgery for Laryngomalacia

- Supraglottoplasty
  - Typically involves releasing bilateral aryepiglottic folds and trimming arytenoid tissues.
2 year old presents in ED with stridor and barky cough in setting of URI

Note features: Biphasic stridor, often louder with inspiratory phase

Viral Croup (laryngotracheobronchitis)
- Most common cause of stridor in young children after neonatal period
- Most affected are children 6 mo.-3 y.o
  - <6 months of age – high degree of suspicion for structural/anatomic anomaly
- Peak incidence 1-2 yrs. of age
- Most cases occur late fall or early winter – Parainfluenza

VIRAL CROUP
- Diagnosis typically made clinically
  - Characteristic features
  - Usually prodromal URI
- X-rays: Usually not required, but support DX
  - Obtain lateral neck films and PA CXR
  - “steeple sign” on PA
  - Sensitivity/Specificity lacking

Management: Isolated Croup
- Typically supportive
- Humidification and single dose PO steroid for milder cases
- Oral/IV Corticosteroids and nebulized racemic epinephrine can be helpful for severe symptoms
  - Try HARD to avoid intubation – risk for acquired subglottic stenosis

Recurrent Croup
- “My child gets croup every time he gets a cold!”
- Consider structural or functional ENT abnormality
  - Subglottic lesion or stenosis
  - Chronic aspiration- laryngeal cleft, vocal fold weakness/paralysis
- Most Otolaryngologists reserve bronchoscopy for severe symptoms/frequency or other suggestive history

This infant presents with recurrent croup-like episodes, but no other URI
Subglottic hemangioma
biphasic stridor

- Often looks, sounds, and acts like croup, but:
  - Usually earlier onset – progressive Sx starting in first few weeks of life.
  - Absence of any other signs of illness is another hint
  - Up to 3% of kids with surface hemangioma will also have airway hemangioma (up to 20% if beard distribution)
    - Conversely, 50% of kids with an airway hemangioma have surface hemangioma
    - Maintain high degree of suspicion in child with skin hemangioma and recurrent stridor

Propranolol

- First line for subglottic hemangioma
  - Discovered accidentally in 2008 in dermatology patients.
  - Dosing: 1-2 mg/kg/day divided TID
- Requires monitoring for potential SE’s:
  - Hypoglycemia, wheezing, hypotension, bradycardia.
- Rapid response (24 hours) seen in most cases.
  - ~15% partial response, 4% nonresponse


Additional Treatment

- Corticosteroids
  - Useful adjunct, usually as intralesional injection, or prolonged systemic for propranolol non-responders
- Microlaryngoscopy (MLB)
  - For distressed patient, intubate, start medical treatment for a few days, then repeat MLB & trial extubation
  - Steroid injection often performed during MLB, CO2 laser ablation becoming more rare
- Open resection and/or tracheostomy less common

Same Patient

- History of:
  - prematurity and intubation
  - History of intubation & RSV
  - cardiac surgery & prolonged intubation
- Or even with no history of intubation
- Flexible endoscopy and see nothing

Subglottic stenosis

Inspiratory or biphasic stridor

- Congenital: partial failure of recanalization of the airway, usually cartilaginous but occasionally just a mucosal web
- Acquired: cicatricial, usually after prolonged intubation
- Mixed: congenital stenosis exacerbated by intubation injury

Subglottic Stenosis
### Myer-Cotton Grading System

Percent Obstruction calculated based on size endotracheal tube allows air leak at 20 cm H2O pressure versus expected size tube for age.

<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>20% Obstruction</td>
</tr>
<tr>
<td>Grade II</td>
<td>20% Obstruction</td>
<td>50% Obstruction</td>
</tr>
<tr>
<td>Grade III</td>
<td>50% Obstruction</td>
<td>90% Obstruction</td>
</tr>
<tr>
<td>Grade IV</td>
<td>No Detectable Lumen</td>
<td></td>
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### Interventions for SGS

- **Endoscopic Dilation or Excision**
  - Good for immature or thin scar, granulation tissue
- **Tracheostomy** – bypass the obstruction
- **Augmentation:** Laryngotracheal Reconstruction (LTR)
  - Grade II/III
- **Resection:** Cricotracheal Resection (CTR)
  - Grade III & IV, and failed LTR

### Balloon Dilation

- Ideal for thin scar bands/webs
- Not good for thicker scars or congenital cartilaginous stenosis
- Sometimes requires several rounds of dilation

### Laryngotracheal Reconstruction

### Open Surgical Management

- **Bypass**
- **Resect**

### Other sources of Stridor post-intubation

- Vocal Fold Granuloma
- Subglottic Cyst
Other Sources of Stridor

- Vocal Fold Paralysis
  - Unilateral often causes stridor without distress, while bilateral paralysis often requires tracheostomy
  - Unilateral usually secondary to trauma (e.g. nerve injury post-cardiac surgery), while bilateral is often neurogenic (Chiari, hydrocephalus, stroke, etc.), and ~40% idiopathic
    - All new bilateral VFP patients need an MRI
- Recurrent Respiratory Papillomatosis
  - HPV 6 & 11, acquired at birth (infected mother)
  - Frequently requires MANY procedures to control

Not covered today but relevant...

- Lower Large Airway Pathology
  - Tracheal Stenosis
  - Tracheobronchomalacia
  - Complete Tracheal Rings
  - Vascular Rings/Slings
- Tend to present with wheezing/rattly breathing, with respiratory decompensation with even mild URIs
  - often confused for asthma
- But that’s for another day...

Take Homes

- The character of stridor can provide clues to the etiology
- Laryngomalacia is the most common cause of stridor in infants, most of whom will not require surgery
- Certain elements of a child’s history raise suspicions that more may be going on (prior intubation, major comorbidity, progressive symptoms, absence of URI symptoms, etc.)
- We’re here to help!

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Useful & Cited Resources