Stridor, Stertor, and Babies that Squeak: Evaluation of Noisy Breathing in Infants

Jeffrey P. Simons, MD, FAAP, FACS
April 25, 2014

Associate Professor
Division of Pediatric Otolaryngology
Department of Otolaryngology
Children’s Hospital of Pittsburgh of UPMC
University of Pittsburgh School of Medicine

Disclosures
• none

Learning objectives
• Characterize various pediatric airway sounds and recognize their potential etiology.
• Describe the indications for flexible laryngoscopy in the office and complete upper aerodigestive tract endoscopy in the operating room.
• Develop a practical approach to the diagnosis and management of infants with noisy breathing.
“Noisy Breathing”

• Common pediatric complaint
• Implies perturbation of airflow
• Many different potential sites
• Variety of potential noises
• Goal:
  – To determine exact site and treat appropriately

“Noisy Breathing”

• Which children need endoscopic airway evaluation?
• How is pediatric airway endoscopy performed?
• What are some common endoscopic airway findings, and how are they managed?

Pediatric Airway Evaluation

• Differs greatly from that of adults:
  – Smaller diameter: minimal edema can lead to critical airway distress
  – Neonatal subglottis: 4-5mm
  – More pliable airway cartilage: “malacia” more common
  – Developmental issues related to feeding and swallowing may contribute to airway symptoms
  – Congenital anomalies and hemangiomas more likely than malignancy and autoimmune diseases
  – Patients can’t describe their symptoms
Definitions

- **Snoring:**
  - Produced by soft palate and uvula vibrating against posterior naso- and oropharyngeal wall
- **Sternor:**
  - Produced by base of tongue, hypopharynx
  - Harsh, rattling sound from pharynx
  - Typically heard during sleep
    - Decreased pharyngeal muscle tone and gravity
  - Typically heard during inspiration
    - Bernoulli effect

- **Stridor:**
  - Harsh, musical sound produced by turbulent airflow through upper airway (larynx, trachea)
  - Can be inspiratory, expiratory, or both
- **Wheezing:**
  - Continuous sound made by walls of narrowed airway vibrating against each other
  - Usually expiratory

Pathophysiology

- Bernoulli’s law:
  - As velocity increases through a constant area, pressure on wall of lumen decreases
  - In an area with anatomic narrowing due to collapse:
    - This area will collapse further with increased airflow: e.g. stridor
Pathophysiology

- Neonatal subglottis: 4 to 5 mm (coronal plane)
- Premature newborn subglottis: 3.5 mm
- Adult subglottis: 12 mm
- Airflow through subglottis governed by Poiseuille’s law:
  - Resistance related to inverse of radius to 4th power
  - Airflow resistance increases exponentially as subglottic diameter narrows
- Narrowing of infant subglottic diameter by 1 mm:
  - Decreases cross-sectional area by 75%
  - Increases airway resistance 16-fold
- Narrowing of adult subglottic diameter by 1 mm:
  - Decreases cross-sectional area by 30%
  - Increases airway resistance 2-fold

History & Physical Examination

- Can the site of airway obstruction be localized by the sound quality?
  - Snoring: nasopharynx, palate
  - Stertor: base of tongue, tonsils, hypopharynx
  - Stridor: larynx, trachea
  - Wheeze: peripheral lower airway

History & Physical Examination

- Can the site of airway obstruction be localized by the phase of respiration?
  - Inspiratory noise: glottis and above (extra-thoracic)
    - Snoring/stertor: pharyngeal soft tissue
    - Stridor: supraglottis/glottis
  - Expiratory noise (or grunting):
    - Tracheobronchial disorder (intra-thoracic)
  - Biphasic noise:
    - Fixed lesion (usually subglottic stenosis)
Diagnostic Dilemma

- Children under age 2 frequently have congenital large airway disease (tracheobronchomalacia) that masquerades as small airway disease (asthma)
- How can you tell these sources of “wheezing” apart?

Large vs. Small Lower Airway Disease

- Large (central) airway disease (e.g., tracheomalacia):
  - Presents 1st 6 mos. of life
  - Often congenital
  - Chronic “congestion”
  - Usually no hypoxemia
  - Palpable fremitus
  - Normal diaphragm pos.
  - No response to Abx
  - No response to bronchodilator

- Small (peripheral) airway disease (e.g. asthma):
  - Presents later in life
  - Often acquired
  - Produced by trigger
  - Hypoxemia
  - No palpable fremitus
  - Depressed diaphragm
  - No response to Abx
  - Good response to bronchodilator

Initial Airway Assessment

- History:
  - Birth history, age of symptom onset, duration and severity of symptoms, associated feeding/voice problems
- Physical examination:
  - Observe child breathing, phonating, feeding
  - Auscultate over nose, mouth, neck and chest
  - Examine nasal cavities, oral cavity, oropharynx, neck
  - Observe for neck/chest retractions; assess urgency of situation
- Radiography:
  - May be helpful, but rarely gives definitive upper airway diagnosis
- When is endoscopy indicated?
  - Whenever the diagnosis is in question, or during emergencies
History and Physical Exam

- Immediate assessment of urgency of situation
- Careful inspection of patient:
  - Respiratory rate, degree of distress
  - Tachypnea, fatigue
  - Nasal alar flaring, accessory neck/chest muscle use
  - Cyanosis, air hunger:
    - Patient may hyperextend neck
    - If unstable, bring immediately to O.R.

History and Physical Exam

- If child stable:
  - Auscultation:
    - Listen over nose, mouth, neck, chest
    - Observe respiratory cycle, and relationship of any stridor to phase of respiration
    - Supraglottic and glottic obstruction: prolonged inspiration
    - Bronchial obstruction: prolonged expiration
  - Place child prone: should relieve stridor from laryngomalacia, macroglossia, micrognathia, vascular compression

Respiratory Distress

- Respiratory distress with feeding:
  - Congenital nasal obstruction
  - TE fistula
  - VC paralysis
  - Laryngeal cleft
  - GERD
  - Cricopharyngeal achalasia
- Respiratory distress after surgical procedure:
  - Subglottic edema from ETT
Types of Airway Endoscopy

- Flexible nasolaryngoscopy

- Rigid laryngotraceoscopy/bronchoscopy

Pediatric Endoscopy

- The choice of which type of endoscopy to perform depends upon nature of child’s noisy breathing:
  - Snoring, stridor, inspiratory stridor, concerns regarding vocal cord paralysis:
    - Flexible endoscopy
  - Expiratory stridor, suspected intubation injury, complex symptoms/patients:
    - Rigid endoscopy
- These different techniques are often complementary

Flexible Nasolaryngoscopy

- Benefits:
  - Can be performed in office, or at bedside
  - No general anesthesia needed
  - Immediate diagnosis (don’t have to wait to schedule a procedure)
  - Allows assessment of pharynx
  - Accurate assessment of laryngeal dynamic function (true vocal cord mobility, laryngomalacia)

- Drawbacks:
  - Cannot assess subglottis or trachea
  - Less magnified image
  - Accuracy may be compromised by patient struggling
  - Rare chance of precipitating laryngospasm in uncontrolled environment
Rigid Endoscopy

- Benefits:
  - Magnified, clear view of airway
  - "Leisurely" inspection of airway with airway protected and child ventilated
  - Thorough inspection of entire airway (including subglottis and trachea)
  - Other potential interventions immediately available (e.g. lasers, optical forceps, etc.)

- Drawbacks:
  - May delay diagnosis (case often has to be scheduled)
  - Requires general anesthesia
  - Sedation and anesthesia may inadvertently suppress pharyngeal muscle tone and vocal fold mobility

Flexible Nasolaryngoscopy

- Anterior nasal cavity:
  - Piriform aperture stenosis
  - Nasolacrimal duct cyst
  - Inferior and middle turbinate hypertrophy
  - Nasal septal deviation
  - Nasal polyps
  - Nasal foreign body

- Posterior nasal cavity:
  - Choanal atresia/stenosis

- Nasopharynx:
  - Adenoid hypertrophy, other mass lesions

Flexible Nasolaryngoscopy

- Oropharynx:
  - Tonsillar hypertrophy

- Hypopharynx
  - Tongue base collapse

- Larynx:
  - Laryngomalacia
  - Vocal fold paralysis/immobility
  - Laryngeal web
**Piriform Aperture Stenosis**

- Congenital narrowing of piriform aperture:
  - Anterior nasal cavity stenosis
- Symptoms:
  - Respiratory distress, cyanosis, feeding difficulties in newborn
- Diagnosis:
  - Endoscopy: narrow nasal vestibule
  - CT: narrowing of piriform aperture <11mm; <5mm: usually requires surgery
- Management:
  - Observation for mild cases (5-11mm)
  - Surgical intervention for severe cases (<5mm)

**Nasolacrimal Duct Cyst (Dacryocystocele)**

- Cystic dilatation beneath inferior turbinate
- Etiology:
  - Congenital obstruction of distal (Hasner) and proximal (Rosenmuller) lacrimal duct valves
  - Can be unilateral or bilateral
    - Bilateral cases can be life-threatening
- Treatment: intranasal excision of cyst with lacrimal duct stenting
- Differential diagnosis: glioma, encephalocele, rhabdomyosarcoma, dermoid cyst
  - Obtain pre-op CT
Nasolacrimal Duct Cyst

Inferior Turbinate Hypertrophy

Neonatal Nasal Septal Deviation

- Occurs in approx. 1% of all births
  - Much greater incidence in vaginal births vs. C-section
- Etiology:
  - Most likely birth trauma
  - Pressure against nose in utero
- Can cause life-threatening airway obstruction
- Management:
  - If mild symptoms: observe
  - If severe symptoms:
    - Manipulate septum back onto maxillary crest (within 3 days)
    - May require nasal stenting
Neonatal Nasal Septal Deviation

Sinonasal Polyposis

- Rare condition
- Cystic fibrosis accounts for 70% of cases
  - Asthma associated for 11% of cases
- Cystic fibrosis:
  - Disorder of mucosal Cl⁻ secretion: dry respiratory mucosa; impaired mucociliary clearance
  - Infectious sinusitis, hyperplastic mucosal changes: polyps
  - Approx. 50% have sinonasal polyps
  - 22% undergo sinus surgery (avg. age: 9-11 years)
  - Medical therapy: steroids (topical, injected); antibiotics (systemic, nebulized)
  - Endoscopic sinus surgery
  - 50% change of needing surgery again prior to 24 months
  - Unrealistic to expect cure of polyposis in CF

Nasal Polyps
Intranasal Foreign Bodies

- Most common in young and developmentally delayed children
- Symptoms: unilateral fetid nasal discharge
- Most common location: below inferior turbinate, or immediately anterior to middle turbinate
- Can remain present for years
- Vegetable matter and button batteries most dangerous
- Can be inadvertently aspirated
- General anesthesia may be required for removal in some children

Nasal Foreign Body

Choanal Atresia

- Neonates are obligatory nasal breathers for 1st 6-8 weeks of life
- Nasal obstruction can be life-threatening
- Choanal atresia: failure of choana to open
  - Between posterior nasal cavity and nasopharynx
    - 1:5,000 to 7,000 live births; F>M
  - 45% bilateral:
    - Respiratory distress requiring urgent intervention; can temporize with Montgomery nipple
  - 55% unilateral:
    - Not life-threatening; unilateral nasal congestion, mucoid rhinorrhea
- Types: bony, membranous (rare), mixed (#1)
- Surgical repair: transpalatal vs. transnasal (endoscopic)
- Post-operative care:
  - Removal granulation; dilatation to prevent re-stenosis
- Can be associated with CHARGE syndrome
Tonsil Hypertrophy

Midface Hypoplasia
- Midface hypoplasia:
  - Leads to soft palate collapse
- Examples:
  - Crouzon’s syndrome
  - Treacher Collins
  - Pfieffer’s
  - Apert’s

Micrognathia
- Leads to glossoptosis (base of tongue collapse)
- Pierre Robin sequence
- Syndromes:
  - Cornelia de Lange
  - Nager’s
  - Stickler’s
Micrognathia

- Usually managed conservatively in mild cases.
- Severe cases may need surgery
  - Tongue lip adhesion
  - Mandibular distraction
  - Tracheotomy

External Approach
Pharyngeal Soft Tissue Obstruction

- Trisomy 21:
  - Small pharynx
  - Pharyngeal hypotonia
- Cerebral palsy:
  - Decreased pharyngeal muscle tone
- Hunter and Hurler’s syndrome:
  - Mucopolysaccharide storage in pharynx causes pharyngeal obstruction

Epiglottis Collapse (Grade 1)

Epiglottis and Tongue Base Prolapse (Grade 2)
Tongue Base Collapse (Grade 3)

Laryngopharyngeal Reflux (LPR)
- Symptoms associated with LPR/GERD:
  - Intermittent stertor, cyanotic spells, frequent emesis, failure to thrive, choking/gagging, nasal congestion
  - All of these symptoms significantly more common < 2 years
  - Empiric treatment with upright position, thickened feeds, reflux medication:
    - Reasonable 1st step with mild symptoms
    - Recommended if endoscopy is negative but symptoms persist

Eosinophilic Esophagitis
- Eosinophilia of esophageal mucosa
- Normal pH probe
- Lack of response to PPI
- Pediatric symptoms:
  - Dysphagia, regurgitation, abdominal pain, failure to thrive
  - Strong association with asthma, AR, eczema
  - Associated with laryngeal inflammation; LTR failure; hoarseness, cough, stridor
- Diagnosis: Esophagoscopy with biopsy (>15 eosinophils/HPF)
- Treatment:
  - Dietary therapy (milk, eggs, wheat, soy, peanuts, shellfish), amino acid (elemental)
  - Inhaled fluticasone (MDI without spacer, x 6-8 weeks)
Laryngomalacia

- #1 congenital pediatric laryngeal disorder
  - Immaturity of supraglottic cartilages; decreased laryngeal sensation
- Symptoms: inspiratory stridor, worse supine and during feeds
- Diagnosed with awake flexible endoscopy
- 100% have pharyngeal acid; 66% have esophageal acid exposure on pH probe (Matthews et al. 1999)
- Management:
  - >90% require observation only, most resolve by 18 months
  - Anti-reflux strategies (thickened feeds, medication)
  - Supraglottoplasty: FTT, severe dysphagia, ALTEs
  - Search for synchronous airway lesions
Laryngomalacia

Supraglottoplasty

Vocal Cord Paralysis

- 2nd most common pediatric laryngeal disorder
- Unilateral:bilateral = 50:50
- Unilateral
  - Symptoms: hoarseness, aspiration (occ.)
  - Etiology: cardiac surgery (#1), neck trauma
  - Management: speech tx; injection laryngoplasty, medialization laryngoplasty
Bilateral Vocal Cord Paralysis

- **Symptoms:**
  - Biphasic stridor (voice quality can be good)

- **Etiology:**
  - CNS: Chiari malformation
  - Idiopathic; inflammatory/metabolic
  - Palpate to rule out posterior glottic stenosis

- **Management:**
  - Observe for spontaneous recovery (12-18 months)
  - Approx. 50% require tracheotomy
  - Surgical options: transverse cordotomy, medial arytenoidectomy, open arytenoidectomy, VF lateralization, posterior graft LTR

---

Bilateral Vocal Fold Paralysis

---

Posterior Glottic Stenosis

- Passive mobility test
Subglottic Stenosis

- #1 acquired pediatric laryngeal disorder
- #1 pediatric laryngeal disorder requiring tracheotomy
- #3 most common congenital pediatric laryngeal disorder
- 95% acquired; 5% congenital
- Etiology: subglottic injury from ETT
- Presentation: stridor after ETT; recurrent croup; life-threatening respiratory distress; already with tracheostomy
- Management:
  - Observation (mild cases); Tracheotomy (severe cases)
  - Endoscopic techniques for acute stenosis
  - Open surgery (LTR, CTR) for mature significant stenoses
Congenital Subglottic Stenosis

Intubation Injury

Endoscopic Balloon Dilatation of Acute Subglottic Injury

Subglottic Stenosis
Post Extubation Trauma

- Incidence of acquired subglottic stenosis has been between 0-4% over last two decades
- Likely because of better ICU care of tube, treatment of GERD, decreased period of intubation
- Progression of the initial injury is variable

Acute Subglottic Stenosis

- Soft stenosis
- Look for other lesions - posterior glottis
- Treat GERD and/or infection

Endoscopic Balloon Dilatation

- Balloons placed endoscopically
- Sizes from 8-14 mm
- Balloon inflated with saline up to recommended burst pressure
- Inflated up to a minute
**Endoscopic Balloon Dilatation**

- Can be done weekly for up to 3-4 weeks
- Fairly successful with resolution in about 70% of selected cases (Sobol et al)
- Larger series required

**Late Subglottic/Tracheal Stenosis**

- Soft and thin stenosis can be managed by endoscopic dilatation with laser
- More firm and long stenosis needs open airway reconstruction

**Posterior Glottic Stenosis**
Laryngeal Web

- Failure of embryological recanalization
- Usually glottic; can extend to subglottis
- Symptoms:
  - Mild (<50% glottis): breathy hoarseness
  - Severe (>50% glottis): stridor/obstruction:
    - May need tracheotomy
- Management:
  - Incise with dilatation, mitomycin C
    - May use laryngeal keel, but might require tracheotomy
  - Open vs endoscopic

Acquired Laryngeal Web

Congenital Laryngeal Web
Laryngeal Atresia

- Associated with CHAOS:
  - Congenital High Airway Obstruction Syndrome
  - Associated with polyhydramnios and fetal hydrops
    - Fetal pulmonary secretions trapped in lungs, which expand and compress venous return to heart
  - Ex-utero intra-partum (EXIT) procedure required

Recurrent Respiratory Papillomatosis

- #1 neoplasm of upper airway in children; likely vertical transmission
- Airway: HPV types 6 and 11; cervical cancer: HPV types 16 and 18
- Prevalence: 4.3/100,000 children in U.S.; median age of diagnosis: 3
- 84% papillomas restricted to larynx
- Symptoms: hoarseness; airway obstruction (less common)
- Treatment: remove obstructing lesions: microdebrider (52%); CO2 laser (42%)
  - Mean: 22 surgical treatments/child (4.4/year)
  - 74% stable; 20% regress; 6% progress to distal spread (94/700)
  - Adjunctive therapies (21%): local or IV cidofovir injection, INF, indole-3-carbinol
- RRP-related deaths (25/700); 3/700 converted to SCCa
- HPV vaccines: 4-valent vs. 2-valent: ages 9-25

Reeves et al. 2003, Schaff et al. 2006
Recurrent Respiratory Papillomatosis

Subglottic Hemangioma

- #1 neoplasm of infant airway:
  - Grows rapidly in infancy, then slow regression
  - Presentation: progressive biphasic stridor (<6 months); 30-70% mortality if not addressed
  - 50% patients have cutaneous hemangioma (usually cervicofacial)
  - Diagnosis: endoscopy (biopsy not needed)
    - MRI if suspicion for cervical/intrathoracic extension
  - Management: individualized approach
    - Observation
    - Tracheotomy
    - Systemic steroids; injected steroids
    - Laser therapy (CO₂, Nd:YAG, KTP)
    - Open excision with laryngotracheal reconstruction
    - Interferon and vincristine: effective but toxic
    - Propranolol
Tracheobronchomalacia

- Flaccidity of tracheal and bronchial cartilage
- Airway collapse on expiration
  - Symptoms: stridor, resp. distress
- Primary tracheobronchomalacia:
  - Cartilage weakness and collapse; usually resolved by 18 months
  - Management: observation; CPAP/BiPAP if severe
- Extrinsic compression: vascular; mediastinal mass
  - Management: observation vs. great vessel pexy
- Can be acquired from tracheostomy
- Management:
  - Endoscopic vs. open repair
Tracheomalacia
(Extrinsic Vascular Compression)

Acquired Tracheomalacia
(Suprastomal Collapse)

Acquired Tracheal Stenosis

- Usually related to tracheostomy tube or cuff from endotracheal tube
- Treatment:
  - Endoscopy: laser, dilatation
  - Open: cartilage graft; resection and end-to-end anastomosis

Complete Tracheal Rings

- Normal C-shaped tracheal cartilage fused posteriorly:
  - Often associated with vascular slings
  - Other associations: Down syndrome, Pfeiffer's syndrome, TE fistula, cardiac and esophageal abnormalities
- Long-segment complete rings: presents in newborns; life-threatening
- Management:
  - Observation (if mild)
  - Complete resection with end-to-end anastomosis (<40% trachea)
  - Patch tracheoplasty (pericardium, cartilage)
  - Slide tracheoplasty (for funnel-shaped stenosis)
  - Balloon dilatation
Complete Tracheal Rings

- Neonatal
  - Increasing respiratory distress
  - Stridor and retraction
  - Apnea, cyanosis and occasionally dying spells
  - Noisy "washing machine" breathing
- Childhood
  - Insidious onset exercise intolerance with biphasic stridor
  - Incidental finding

- Slide tracheoplasty has become the treatment of choice as it has dramatically reduced the morbidity and mortality in these patients
Aspirated Foreign Bodies

- Common in 1-3 year olds (put objects in mouth)
- Accounts for 7% lethal accidents in 1-3 year olds
- Acute symptoms:
  - Coughing, choking, dyspnea
  - Can subside rapidly and spontaneously
- Long-term symptoms:
  - Persistent cough, wheezing, bronchiectasis, pneumonia
  - Inflammation/granulation develop
- Diagnosis:
  - Witnessed aspiration is most predictive
  - In absence of witnessed event, need high index of suspicion
  - CXR: may show atelectasis (25%), air trapping, consolidation (6%); FB only seen in 15%
  - Most aspirated material in kids: food (not radio-opaque)
- Treatment: rigid bronchoscopic removal
Bronchial Foreign Body

Infectious Causes of Noisy Breathing

Laryngotracheobronchitis (Croup)

- Peak ages: 1-2 years old
- Usually viral (#1: parainfluenza type I)
- Clinical diagnosis (doesn’t require endoscopy): low-grade fever, barking cough, biphasic stridor
- Therapy (usually outpatient; <10% hospitalized):
  - Humidified air with supplemental O_2
  - Oral steroids (outpatient)
  - Racemic epinephrine; IV corticosteroids
- Recurrent croup (Waki et al. 1995):
  - 50% associated with LPR; 25% associated with SGS or LM
Bacterial (Exudative) Tracheitis

- Initially described as rare, rapidly-progressive, life-threatening infection
  - Thick, membranous tracheal secretions obstruct airway
  - Average age: 4-6 years
  - Diagnosis: prompt endoscopy in O.R.
  - S. aureus: 65% cases
  - Management: suction secretions (endoscopy); IV Abx; intubation (90%)
- Recent reports describe decreasing severity
- May be part of continuum with croup
Acute Epiglottitis (Supraglottitis)

- Rapid onset inflammation of supraglottis
- True airway emergency
- Management: direct trip to O.R.; attempt intubation; IV Abx
- H. influenzae type B
- 1987: Conjugate HiB vaccine in U.S.: dramatic reduction in incidence (3,400/yr to 125/yr)
- Adult cases less severe; only 10-20% caused by HiB
- Effect of vaccine:
  - Avg. age went from 5 to 14 years
  - Overall less severe

Conclusions

- Variety of causes of noisy breathing in infants
- Important to identify type of noisy breathing
- Diagnosis based upon:
  - History and physical examination
  - Flexible and rigid aerodigestive endoscopy
- Management options:
  - Based upon severity of symptoms, diagnosis, and general health of child
  - Still searching for optimal treatment for many pediatric upper airway disorders