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

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Disclosures

• none



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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



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"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?



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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
- Stertor
 - 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



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Definitions

- 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



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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 diamater 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

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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



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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?



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Large vs. Small Lower Airway Disease

- · Large (central) airway disease (e.g. racheomalacia):
 - 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

- - 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
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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.



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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



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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 laryngotracheoscopy/bronchoscopy





Pediatric Endoscopy

- The choice of which type of endoscopy to perform depends upon nature of child's noisy breathing
 - Snoring, stertor, 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



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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)
- - 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



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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

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Flexible Nasolaryngoscopy

- Oropharynx:
 - Tonsillar hypertrophy
- Hypopharynx
 - Tongue base collapse
- Larynx:
 - Laryngomalacia
 - Vocal fold paralysis/immobility

Laryngeal web



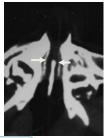
Piriform Aperture Stenosis

- 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)



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Pyriform Aperture Stenosis







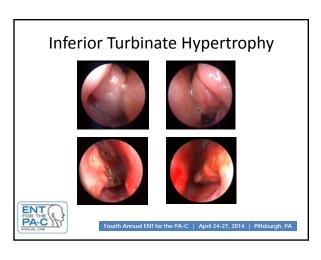
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Nasolacrimal Duct Cyst (Dacryocystocele)

- 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



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

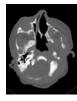


Neonatal Nasal Septal Deviation









Sinonasal Polyposis

- Rare condition
 - Cystic fibrosis accounts for 70% of cases
 - Asthma associated for 11% of cases
- - 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



Rowe-Jones et al 1996; Triglia and Nicollas 1997

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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



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Nasal Foreign Body



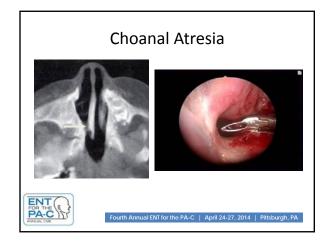


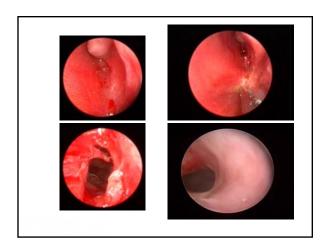


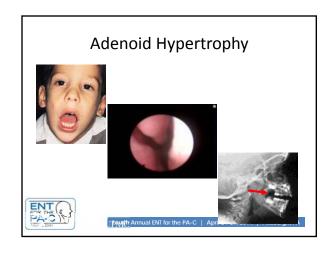
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Choanal Atresia

- Neonates are obligate 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 LangeNager's
 - Stickler's







Micrognathia

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

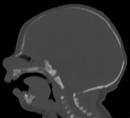




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Micrognathia

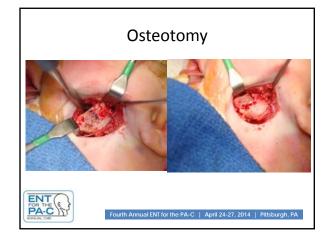




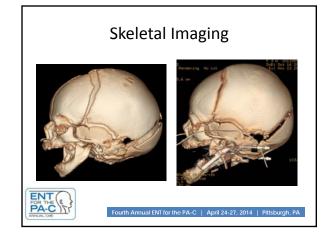
External Approach











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)

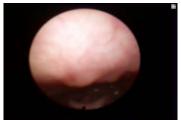


Cerebral Palsy



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Epiglottis and Tongue Base Prolapse (Grade 2)



Cornelia de Lange Syndrome

Tongue Base Collapse (Grade 3)





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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







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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: Freehagescopy with biopsy (215)
- Diagnosis: Esophagoscopy with biopsy (>15 eosinophils/HPF)
- Treatment:
 - Dietary therapy (milk, eggs, wheat, soy, peanuts, shellfish); amino acid (elemental)
 - Ingested fluticasone (MDI without spacer, x 6-8 weeks)



Laryngomalacia

- #1 congenital pediatric laryngeal disorder
 - Immaturity of supraglottic cartilages; decreased lx
- 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)

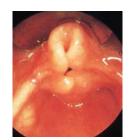
 - Supraglottoplasty: FTT, severe dysphagia, ALTEs Search for synchronous airway lesions



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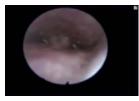
Laryngomalacia

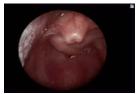




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Laryngomalacia







Laryngomalacia







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Supraglottoplasty







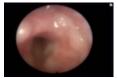
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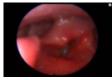
Vocal Cord Paralysis

- 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



Vocal Cord Paralysis







Bilateral Vocal Cord Paralysis

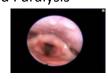
- Symptoms:
 Biphasic stridor (voice quality can be good)
- Etiology:
 - CNS: Chiari malformation
 - Idiopathic; inflammatory/metabolic
 - Palpate to rule out posterior glottic stenosis
- Management:
 - Observe for spontaneous recovery (12-18 months)
 - Approx. 50% require tracheotomy
 - Surgical options: transverse cordotomy, medial arytenoidectomy, open arytenoidpexy, VF lateralization, posterior graft LTR



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Bilateral Vocal Fold Paralysis





Posterior Glottic Stenosis

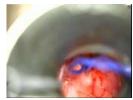




Passive mobility test

Laryngeal EMG







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CO₂ Laser Transverse Cordotomy





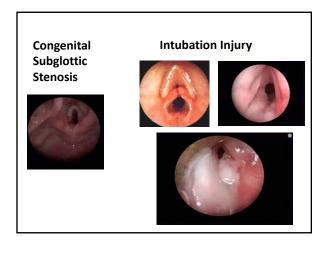
Posterior Cricoid Split with Cartilage Graft

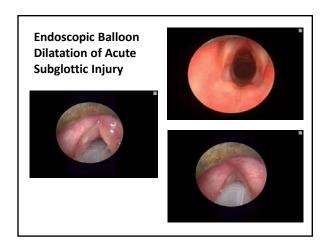


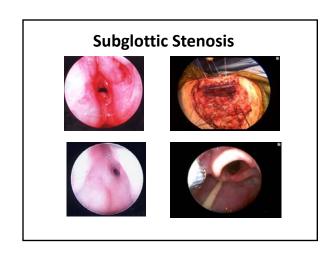
Subglottic Stenosis

- #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







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





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Acute Subglottic Stenosis

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

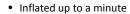


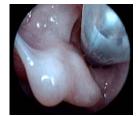


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Endoscopic Balloon Dilatation

- Balloons placed endoscopically
- Sizes from 8-14 mm
- Balloon inflated with saline up to recommended burst pressure







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





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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







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Posterior Glottic Stenosis







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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



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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%); ${\rm CO_2}$ laser (42%)

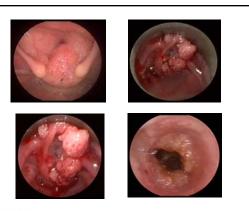
 - Mean: 22 surgical treatments/child (4.4/year)
 74% stable; 20% regress; 6% progress to distal spread (94/700)
 - Tracheotomy: 11%
- Iracneotomy: 11%
 Adjuvant therapies (21%): local or IV cidofovir injection, INF, indole-3-carbinol
 RRP-related deaths (25/700); 3/700 converted to SCCa
 HPV vaccines: 4-valent vs. 2-valent: ages 9-25

Recurrent Respiratory Papillomatosis



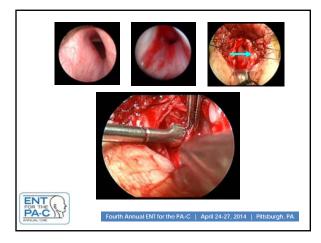






Subglottic Hemangioma

- #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
 Propranalol



Tracheobronchomalacia

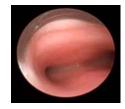
- Flaccidity of tracheal and bronchial cartilage
- Airway collapse on expiration
- Symptoms: stridor, resp. distress
- Primary tracheobronchomalacia:

 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



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Primary Tracheobronchomalacia

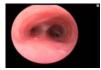






Tracheomalacia (Extrinsic Vascular Compression)





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-toend 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

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Complete Tracheal Rings

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





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Complete Tracheal Rings

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





Complete Tracheal Rings









Foreign Body Aspiration





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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
 Diapnosis:
- 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



Br	onchial Fo	oreign Body	
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Infectious Causes of Noisy **Breathing**

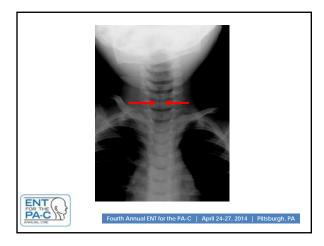


Laryngotracheobronchitis (Croup)

- Peak ages: 1-2 years old
- Usually viral (#1: parainfluenza type I)
- Clinical diagnosis (doesn't require endoscopy): low-grade fever, barky cough, biphasic stridor
- Therapy (usually outpatient; <10% hospitalized):
 Humidified air with supplemental O₂

 - 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, lifethreatening 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



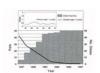
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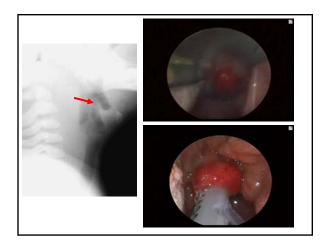
Bacterial (Exudative) Tracheitis

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

