Pediatric Aspiration and other Aerodigestive Disorders

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Disclosures

I have no actual or potential conflicts of interest in relation to this presentation.
Learning Objectives

- Recognition of signs and symptoms that indicate a possible aerodigestive disorder
- Improve understanding of types of pediatric aerodigestive patients
- Understand the multidisciplinary approach to treating pediatric aerodigestive disorders
- Provide knowledge regarding surgical and medical treatment options of pediatric aerodigestive patients

Who is an “Aerodigestive Patient”?

- There is no universally used definition
- According to the American Academy of Pediatrics:
  
  “A pediatric aerodigestive patient is a child with a combination of multiple and interrelated congenital and/or acquired conditions affecting airway, breathing, feeding, swallowing, or growth that require a coordinated interdisciplinary diagnostic and therapeutic approach to achieve optimal outcomes. This includes (but is not limited to) structural and functional airway and upper gastrointestinal tract disease, lung disease because of congenital or developmental abnormality or injury, swallowing dysfunction, feeding problems, genetic diseases, and neurodevelopmental disability.”

  Aerodigestive Care: AAP Consensus Statement, 2018

### Who is an “Aerodigestive Patient”?  

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<th>Airway</th>
<th>Pulmonary</th>
<th>Gastroenterology</th>
<th>Feeding/ Swallowing</th>
<th>Genetic</th>
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<td>GERD</td>
<td>Swallowing dysfunction</td>
<td>Trisomy 21</td>
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### Signs and Symptoms of Aerodigestive Patients

**Clinical signs and symptoms of oral dysphagia**
- weak or uncoordinated suck
- delayed or slow oral transit
- anterior oral loss

**Clinical signs and symptoms of pharyngeal dysphagia (including trigger of swallow)**
- absent swallow reflex
- suck/swallow/breath incoordination
- wet voice/breathing
- increase in work of breathing
- rattyly feel on back
- watery eyes or surprised look
- gasping
- vomiting
- reduction of oral intake
- choking


Overview of Pediatric Aerodigestive Program

► Essential construct and functions of an aerodigestive program
  ► **Core Members of Team With Input Required for All Patients:** Care coordinator, Gastroenterology, Nursing, Otolaryngology, Pulmonology, Speech-language pathology
  ► **Input for some patients or consultation:** sleep medicine, social work, dietician, respiratory therapy, pediatric surgery, allergy and immunology, anesthesia, child life, genetics, developmental pediatrics, neurology, radiology


Overview of Pediatric Aerodigestive Program

► Impact of an interdisciplinary care model for pediatric aerodigestive patients:
  ► Reduction in time to diagnosis (6 vs 150 days)
  ► Fewer required specialist consultations (5 vs 11)
  ► Significant reduction in risk:
    ► Fewer radiation exposures (2 vs 4)
    ► Fewer anesthetic episodes (1 vs 2)
  ► Reduction in total cost associated with diagnostic evaluation
    ► Significantly reduced from a median of $10,374 to $6,055

Overview of Pediatric Aerodigestive Program

Multi-D Aerodigestive Program

Consultants to the Team
- Pediatric Neurologist
- Pediatric Surgeon
- Physician Medicine and Rehab MD
- Pediatric Genetics
- Pediatric Radiologists

Pediatric Otolaryngologist
Pediatric Otolaryngology NP
Program Care Coordinator
Pediatric Pulmonologists x4
Pediatric GI Physicians x2
Pediatric Speech Therapists x5
Social Worker

Process of Aerodigestive Patients

Patient referral to Aerodigestive Program or one of the participating specialties
- From outpatient Peds
- From PICU
- From NICU
- From Peds Floor
- From ER

Evaluations by each specialty
- Pediatric Pulmonary
- Pediatric GI
- Pediatric ENT
- Pediatric Speech Therapy

Modified Barium Swallow Study and Esophagram

Aerodigestive Team Meeting
- Feeding strategies
- Medical management
- Social concerns
- Consultations, referrals, MRI, etc.
- Sleep concerns

Triple Endoscopy
- Direct Laryngobronchoscopy (ENT)
- Flexible bronchoscopy (Pulm)
- EGD (GI)

Aerodigestive Team Meeting
- Repeat additional imaging
- Modifications of feeding regimen
- Modifications of medical management
- Decision regarding airway surgery
Case Study:

Initial presentation (to primary care sick clinic):

- Six month old term male infant
  - Born at 37 2/7 weeks via C-section
    - Pregnancy complicated by pre-eclampsia, gestational diabetes, and GBS
    - Respiratory distress at birth, intubated x24 hours. Extubated to CPAP and weaned to RA over the next 24 hours.
  - 7 day NICU stay for pneumonia
  - Was started on famotidine in NICU secondary to apneas and reflux symptoms with feeds

- A few weeks after leaving the NICU, he developed chronic wet cough
  - Cough is noted to be worse with feeds
  - “Rattly” sounding breathing after feeds

Initial presentation (to primary care sick clinic):

- History of current illness:
  - Wet cough has worsened for the last 1 week with ear tugging and irritability for the last 2-3 days
  - Fever 101F for 2-3 days
  - Family reports frequent episodes of wet cough, occasional retractions, and choking/gagging with feeds

- Plan
  - Bilateral AOM, prescribed Augmentin due to being treated 2 weeks prior with Amoxicillin
  - Diagnosed bronchiolitis/viral URI
  - Referral to Pediatric Pulmonary for chronic cough
  - Referral to Pediatric ENT for recurrent AOM, 4 episodes in 3 months
Case Study:

- **Review of Systems:**
  - **Pertinent positives**: recurrent AOM, diarrhea with antibiotics, eczema, recurrent rhinitis, reflux, wet cough, choking with eating
  - **Pertinent negatives**: growth concerns/delay, no other infection types except AOM and respiratory illness, no pneumonias diagnosed, no stridor

- **Family history:**
  - **Positives**: Asthma (mother), allergic rhinitis (father), recurrent AOM s/p BMT and PE tubes (sister)
  - **Negatives**: No family history of cystic fibrosis, recurrent pneumonias, sinusitis, immune deficiencies

Pediatric Pulmonary Evaluation

**Pertinent History:**

- **Recurrent respiratory illnesses:**
  - Parents report he has not gone more than 2 weeks since birth without a “respiratory illness”
  - **ED visit:**
    - At 5 months old, he had increased work of breathing, wheezing, rhinorrhea.
    - Diagnosed with RSV
    - Treated with prednisolone and albuterol (bronchodilator not noted to improve symptoms)
  - Admitted at 6 months for acute hypoxemic respiratory failure with bronchiolitis
  - Four total courses of antibiotics for ear infections; transient improvement in cough while on antibiotics or steroids
Pediatric Pulmonary Evaluation

Chest XR

- FINDINGS: Normal size cardiothymic silhouette. **The lungs are hyperinflated.** **Mild prominence of the central bronchovascular markings.** No focal parenchymal process. No pleural effusions. No pneumothorax. No acute osseous abnormalities.

- IMPRESSION: Nonspecific findings can be seen with reactive airway disease versus viral/atypical infection. No focal airspace disease.

Pediatric Pulmonary Evaluation

**Pulmonary Differential Diagnoses**

- Dysphagia / aspiration
- Recurrent viral respiratory illnesses
- Early asthma phenotype
- Cystic fibrosis
- Primary ciliary dyskinesia
Pediatric Pulmonary Evaluation

Plan:

- **For evaluation of etiology of recurrent wheezing and chronic cough:**
  - Chest X-Ray: demonstrated airway inflammation, no other structural or focal abnormalities
  - Sweat Chloride Test: to rule out cystic fibrosis as cause for chronic cough
  - Modified Barium Swallow Study (MBSS) and esophagram:
    - Pending clinical course and findings of the above studies, an airway evaluation, immune deficiency testing, genetic testing for primary ciliary dyskinesia, or chest CT could be considered in next steps.

- **For treatment of airway inflammation (with chronic cough / wheeze):**
  - Recommended initiation of an inhaled corticosteroid: Flovent 44mcg 2 puffs BID with spacer/mask.

Pulmonary Care for Aerodigestive Patients

Pulmonary presentation of chronic aspiration:

- Common presentation with pulmonary manifestations include recurrent wheezing, chronic cough, recurrent pneumonias, “wet” sounding breathing
- Later development of severe impairment of lung function, irreversible pulmonary scarring, bronchiectasis

Chest imaging:

- CXR: Relatively insensitive to early change; normal in 14% of children with chronic aspiration. Demonstrates bronchial wall thickening and/or hyperinflation; May demonstrate infiltrates
- Chest CT: More sensitive, however more radiation and still nonspecific (bronchial wall thickening, air-trapping, bronchiectasis, ground-glass opacities, centrilobular opacities)

Tutor JD. Dysphagia and Chronic Pulmonary Aspiration in Children. Pediatrics in Review May 2020, 41 (5) 236-244
### History (pertinent positives):
- History of intubation after delivery
- Wet breath sounds and wet cough, worse with eating
- Noisy breathing at baseline described as “gurgling”, denies stridor
- Chronic nasal congestion
- 4 episodes of AOM in 6 months
- Pure tone audiogram prior to this visit normal
- Passed newborn hearing screen

### Exam (pertinent positives):
- Bilateral middle ear effusion
- Course rhonchi throughout lung fields
- Nasal stertor
- FFL completed with nonspecific findings

### Flexible Fiberoptic Laryngoscopy (FFL)
Pediatric Otolaryngology Evaluation

Findings on FFL:
- Cobblestoning of oro/nasopharynx
- Erythematous arytenoids
- Increased secretions in oropharynx
- No redundancy of arytenoids, no inward prolapse

Differential Diagnoses
- Dysphagia / aspiration
- GERD
- Tracheomalacia/bronchomalacia
- Vascular ring or sling
- Tracheoesophageal fistula
- Other airway anomaly

Treatment Plan

- Referral to remaining members of Aerodigestive Team:
  - Speech Therapy
  - Pediatric Gastroenterology
  - Modified Barium Swallow Study
  - Esophagram
    - To evaluate for dysphagia/aspiration and vascular ring/sling, esophageal dysmotility, or esophageal stricture
  - Direct Laryngobronchoscopy (DLB) with rigid bronchoscopy after evaluation by Aerodigestive team
Pediatric Speech Pathology Evaluation

Initial Video Fluoroscopic Swallow Study (VFSS), 7 months:
- Moderate-severe oropharyngeal dysphagia
  - Delayed swallow to pyriform sinuses
  - Aspiration with thin and nectar-thick liquids
  - No aspiration with honey-thick liquids
  - Occasional shallow nasopharyngeal reflux
  - Laryngeal elevation, BOTR, pharyngeal constriction within functional limits

Esophagram: Normal

Recommendations:
- Honey-thick liquids (1 tbsp oatmeal cereal per 1 oz of formula) via Doctor Brown’s Level 4 nipple
- Follow ups with concern for intolerance to oatmeal thickening, transitioned to commercially available thickener

Speech Therapy Care for Aerodigestive Patients

Incidence of dysphagia
- 1% of children in general population
- Incidence rate is higher in some populations like prematurity, congenital cardiac disease, CP, genetic and neurologic abnormalities, and airway malformations
- Causes of pediatric dysphagia are not the same as in adult population

Videofluoroscopic Swallowing Study (VFSS/MBS)
- Gold standard
- In real time, allows visualization of bolus through upper aerodigestive tract
- It shows the timing and presence of the aspiration
- Speech Therapist can assess bolus volumes, textures, and compensatory strategies effects

Speech Therapy Techniques to address aspiration
- Modified diet
- Modify bottles/nipples and other feeding equipment
- Pacing, positioning, other compensatory strategies
Pediatric Gastroenterology Evaluation

History:
- Coughing and choking with feeding
- Episodes of emesis/spit up multiple times per day
- Irritability and crying for approximately 20 minutes
- Growth remains stable (weight for length remains at 25%ile)
- Lack of improvement on famotidine after being used x2 months

Differential Diagnosis:
- Gastroesophageal Reflux
- Eosinophilic Esophagitis
- Oral motor incoordination
- Esophageal Dysmotility
- Esophageal anatomical abnormalities
- Esophageal compression or stricture

Plan:
- Proton Pump inhibitor 1.5 mg /kg/day
- pH/impedence probe to evaluate for acidic nature of reflux
- EGD for full evaluation

Aerodigestive Team Discussion
- Once patient has been evaluated by all specialties involved in Aerodigestive Team, they are presented at a team discussion
- Subsequent evaluations are discussed depending on each evaluation (i.e. Neurology, Physicians Medicine and Rehab)
- Triple Scope discussion
- Determinations regarding surgical repair of abnormalities
- Follow-up discussions after subsequent evaluations and surgical interventions
Case Study:

Summary: 6 month old with known aspiration
- Honey thickened liquids
- Proton Pump Inhibitor
- Inhaled corticosteroids
- After modifications to feeding and diet, transient improvement was noted for approximately 3-4 months.

Update to Case Study: Patient is now 12 months old
- Return of mild respiratory symptoms despite consistent use of honey thickened liquids
- Has remained on Nexium due to chronic GERD concerns
- Still cannot rule out airway anomaly → Triple Scope

Triple Scope Evaluation

- Key component of the aerodigestive evaluation is the “triple endoscopy”
  - Direct Laryngoscopy and Rigid Bronchoscopy (DLB)
  - Flexible fiberoptic bronchoscopy with bronchoalveolar lavage (flexible bronchoscopy with BAL)
  - Esophago-gastro-duodenoscopy (EGD)

- Under a single anesthetic
  - Requires significant scheduling coordination
  - Minimizes the risks and redundant costs of multiple anesthetics
  - Collaco et al estimated a 41% reduction in anesthetic episodes through the utilization of combined endoscopy

Direct Laryngobronchoscopy (DLB)

Normal findings on DLB

Classification of Laryngotracheal Cleft

DLB evaluation reveals Type 1 laryngeal cleft
Flexible Bronchoscopy with Bronchoalveolar Lavage

Upper trachea (clear secretions)

Main Carina (mucosal edema, cobblestoning, R mainstem white secretions)

Left mainstem (mucosal edema, white secretions)

Bronchoalveolar Lavage:
- **Cell Counts:**
  - RBC <3000/mcL
  - WBC 817/mcL
  - 77% Neuts, 2% Lymphs 18%, Monos/Macros, 1% Eos
- **Lipid Laden Macrophage Index:** 59
- **Culture:** *Moraxella catarrhalis*
  - >10,000 CFU/ml, Beta Lactamase positive
  - Treated with Augmentin

Pulmonary Care for Aerodigestive Patients

Bronchoscopy / BAL results
- Commonly demonstrates bacterial pathogens from oropharynx:
  - Strep pneumoniae, group A Staph aureus, Proteus species, E. coli, Moraxella, Aerobacter species, H. influenza

- Cell count differential may assist in determination of indications for treatment

- Lipid Laden Macrophage Index: Nonspecific but useful, may be elevated in setting of infection specificity 79% and sensitivity 69%


Esophagogastroduodenoscopy (EGD)

- Essentially normal EGD
  - No evidence of esophageal narrowing or strictures
  - No evidence of eosinophilic esophagitis
  - Some erythema associated with possible GERD
  - Normal biopsies

GI Care for Aerodigestive Patients

- Evidenced based relationship between GERD and recurrent pneumonia due to failure of airway protection
  - Improvement with both medical (i.e. Proton Pump Inhibitors) and surgical therapy (i.e. Nissen Fundoplication)

- Prospective study outlining causes of recurrent pneumonias:
  - aspiration with swallowing 48%
  - immunologic disorders 14%
  - GER 14%
  - congenital heart disease 9%
  - asthma 8%
  - respiratory tract anatomic abnormalities 8%

- Evidence supporting benefit of PPI against pneumonia, chronic cough, and wheezing

Surgical Intervention

Team decision by Aerodigestive Team

Timing: When symptoms are still present despite feeding modifications and medical management, typically after 1 year of age

Goal: Decrease in respiratory symptoms, decrease in chronic lung damage, improvement in diet tolerance

Realistic expectations communicated to family prior to procedure by all specialties of team

Type 1 Laryngeal Cleft Repair

Laser assisted surgical technique to denude tissue prior to approximating arytenoids

Approximation of arytenoids with 5-0 Vicryl suture
Postoperative Type 1 Laryngeal Cleft

- Pediatric ICU care following surgery
- NPO x 24 hours.
- Dexamethasone prevent laryngeal edema
- Bedside evaluation by speech therapy to determine readiness to feed postoperatively
- Remain on preoperative feeding modifications
- Continue Nexium BID x 6 weeks
- Follow up FFL in office to evaluate postoperative site
- Repeat MBSS 6 weeks after surgery to determine if can wean diet
ENT Care for Aerodigestive Patients

- In a 10 year retrospective study evaluation swallow function of children following cleft repair:
  - 57% of children had normal swallow function after repair (one study reports up to 75% of patients normalize after repair)
  - 20% showed penetration of thin liquids following repair
  - 23% of children continued to document aspiration

- Notch augmentation (ongoing research and evaluation) vs. continue to modify diet and treat symptoms recognizing defect as not a complete laryngeal cleft
  - Current practice at our center to augment laryngeal notch if with continued symptoms
    - Statistically significant improvement in diet after notch repair
    - 53% of patients achieved a normal diet after notch repair
    - 87% of patients showed improvement defined by decrease in ICS dose, decrease in diet consistency, or both

Osborn et al. (2014). Swallowing function after laryngeal cleft repair: More than just fixing the cleft. The Laryngoscope


Follow Up on Patient Presentation

Speech:
- VFSS repeat 14 months of age s/p type 1 laryngeal cleft repair
- Results: mild oropharyngeal dysphagia, delayed initiation, decrease laryngeal closure, laryngeal penetration of bolus. No aspiration visualized with thin or nectar.
- Feeding modification changes transition to thin liquids beginning with initial transition to nectar. Transition to thin was tolerated.

Pulmonary:
- Significant improvement in respiratory symptoms since laryngeal cleft repair
- Tolerating wean to thin liquids per speech therapy
- No choking, coughing episodes with drinking per parents
- No baseline cough, wheeze, shortness of breath
- No subsequent antibiotics or steroid courses. No further ED visits or hospitalizations
- Currently weaning down on inhaled medications.
Key “Take Home” Points:

- Signs of pediatric aspiration are subtle and often unrecognized.
- Refer early as early intervention prevents long term and potentially irreversible lung damage.
- An interdisciplinary team approach is required to care for these patients due to the complex nature of their swallowing issues.
- Goal of therapy is to modify diet to promote safety of swallowing and optimize medication to prevent irreversible lung damage.
- Thorough evaluation and multiple reevaluations are necessary to determine anatomical cause of aspiration and to consider all potential causes of each patient’s aspiration.
- After 1 year of age, consider surgical repair if symptoms persist and an airway anomaly is discovered.

References:


Osborn et al. (2014). Swallowing function after laryngeal cleft repair: More than just fixing the cleft. *The Laryngoscope*


Thank you!

Please do not hesitate to email me with any questions!
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