2013 Charleston Swallowing Conference

Session 10
Gastrointestinal And Swallowing Interactions In Pediatrics
10:00 - 11:30am
Saturday, Oct. 12, 2013

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Disclosures

- I have the following financial relationships relevant to the content of my presentation:
  - Cengage Learning: Royalties on sale of books
  - Pearson: Royalties on sale of books
  - Northern Speech Services: Royalties on sale of training manuals
- I have no relevant nonfinancial relationships to disclose

Dysphagia with GI Issues

- Aspiration risks: differentiate "above" from "below"
- Negative impact on interest in oral feeding
  - Vomiting
  - Esophagitis
  - Volume sensitivity/tolerance
  - Retching, gagging with fundoplication
  - Globus

State of Art & Science for Evaluation of Infants & Children with Signs/Symptoms of Dysphagia

- Clinical feeding evaluation
- FEES (often when upper airway & swallowing concerns are noted)
- VFSS
  - Stand alone examination
  - Combined with esophageal manometry

Introduction

- Predictability: the only predictable aspect for infants & young children is unpredictability
- Systematic decision making is important, with individual variability always in mind
- Sensori-motor learning & neural plasticity: useful considerations for management plans
- What else?

Criteria for Instrumental Evaluation

- Risk for aspiration by history or observation
- Prior aspiration pneumonia
- Suspicion of pharyngeal/laryngeal problem on basis of etiology
- Gurgly voice quality
- Need to define oral, pharyngeal, & upper esophageal components for management
Instrumental Swallow Evaluations

- Videofluoroscopic Swallow Study (VFSS)
  - AKA – Modified Barium Swallow Study (MBS)
  - Oral Pharyngeal Motility Study (OPMS)
  - Rehabilitation Swallow Study
  - Cookie Swallow + other names
- Fiberoptic Endoscopic Evaluation of Swallowing (FEES) + Sensory Test (FEESST)

Flexible Endoscopic Evaluation of Swallowing (FEES)

- No radiation
- Bedside exam possible
- Defines some aspects of pharyngeal physiology
- Can evaluate handling of secretions
- Sensory testing can be done

Videofluoroscopic Swallow Study (VFSS)

- Defines oral & pharyngeal phases
- Defines esophageal transit time & basic motility
- Delineates aspiration related factors
  - Before, during, or after swallows
  - Texture specificity
  - Physiologic reasons for aspiration
  - Estimate of risk

What VFSS is NOT

- To rule out aspiration or determine if child aspirates with oral feeding (important finding but not reason for exam)
- Simulation of a real meal
- Evaluation of oral skills for bolus formation
- Chewing evaluation
- Esophageal function (only upper esophagus)

VFSS Aims

- Examine intervention possibilities
  - Postural changes
  - Sensory enhancement
  - Maneuvers in children (not infants)
  - Diet modifications (texture changes)

Major Unanswered Question:

How much aspiration of what can a system tolerate before chronic lung disease becomes an issue?
Considerations for VFSS

- Medical stability
- Start with thinnest liquid possible – controlled volume prior to consecutive swallows
- Therapeutic modifications: position, utensils, texture, taste, temperature, size of bolus
- Tube feeder: pre-exam experience with PO

VF Equipment & Procedures

- 30 frames per second, not pulsed
- Radiation exposure determined by radiologist
  - Need adequate clarity
  - Minimal radiation dose
  - Coning – avoid orbits of eyes – cannot eliminate the thyroid gland
- Radiation safety always priority

Procedural Decisions

- No fixed order for presentations in pediatrics
- Preferable to start with thinnest liquid
  - Controlled bolus size to start, e.g., spoon before going to bottle or cup drinking
- Work toward thicker as needed
  - Not want residue in pharynx that may complicate interpretation with thinner later
- Exceptions: parents tell us that child will not accept anything else if he gets liquid first

Reasons to Start with Thin Liquid

- If aspirated
  - More easily expectorated – but remember young infants & those with neurologic impairment are not likely to cough
  - Small amounts of thin liquid may be absorbed by “stable” lungs (more research needed – we don’t know how much, how long)
  - Cannot block the airway

Findings & Interpretation

- Findings: objective descriptions
  - By phase of swallow per texture & volume
  - If aspiration, report must note
    - Before, during, or after swallows
    - Texture specificity
  - Interpretation: reasons for findings
    - Relate findings to underlying physiologic reasons

Findings Related to GI Issues

- Aspiration from above: oropharyngeal dysphagia
- Type 1 laryngeal cleft: may be noted when barium contrast “sneaks in to trachea from behind” rather than typical pattern
- Retrograde transit from lower esophagus: esophageal dysmotility or GER
- “Normal” findings with pulmonary problems: more likely to relate to GI tract
### Esophageal Dysphagia Diagnosis
- Dysphagia for solids > liquids, structural cause likely
- Dysphagia for solids & liquids similar, dysmotility likely cause

### Review of Findings
- Findings are reviewed with caregivers immediately following exam
- All exams are saved as permanent record
- SLP reviews in real time, slow motion, & frame-by-frame
- Radiologists & PAs also review after exam
- Call or page with questions

### Problem Areas from VFSS
- Oral phase
- Initiating pharyngeal swallow
- Pharyngeal phase
- Esophageal phase (upper)
  - Esophagram or UGI may be needed to define esophageal function
  - Impedance, manometry, or pH probe

### Manometry with VFSS
- High Resolution Manometry (HRM)
  - Closely spaced point pressure sensors at high spatial resolution
  - Measures pressure changes in pharynx & upper esophageal sphincter
- Simultaneous VFSS correlates swallow function with pressure measures
- Gives definitive information re impact of reduced pharyngeal function on UES

### HRM-Esophagus Manometry
- Multiple pressure sensors (>20)
- Spaced at 1 cm intervals
- Allows simultaneous evaluation of
  - Pharynx
  - UES
  - Esophageal body
  - LES
  - Stomach

### Solid-state HRM Promising
- Meets 2 major requirements
  - Rapid response rate
  - Insensitivity for axial movements
- Advantages
  - Objective assessment to describe physiologic mechanisms of dysphagia in pressure-flow mechanical terms
- Rommel & Omari, 2011
Management Recommendations

- Route for nutrition/hydration
- Feeding suggestions
- Therapy recommendations
- Additional suggestions
- Plans for follow-up or re-evaluation

PO Feeding: Slow tube feeding

- No sense of hunger or satiation
- Night feeds – we are not physiologically geared to be fed through the night (beyond young infancy)
- If GJ feeds, only saliva & whatever is taken orally goes through the stomach
- Highest priority: always nutrition & hydration

PO Feeding: GT + Fundoplication

- Gagging, retching not unusual
- Fundoplication may be too tight
- Procedure reduces stomach size, thus capacity would be reduced
- Fundoplication may tear or loosen, resulting in emesis & possible aspiration

Food Refusal

- Common in child with history of surgical procedures, prolonged intubation, NPO for more than a few weeks
- Memory is long after medical/surgical stability is attained – e.g., s/p EA/TEF
- Eosinophilic esophagitis (EoE) – elimination diet not always adequate for total PO
- Global sensory deficits may have impact

Feeding Aversion

- Hypothesis: in most children, aversion is actually imposed from outside
  - Pressure from caregivers & (therapists in some instances) to get children to eat/drink in ways that impose changes in bigger steps than can be managed on the basis of sensorimotor skills
  - Logical responses by child related to past experiences of pain, discomfort

Recommendations After VFSS

- Changes in route of nutrition/hydration
- Nutrition guidelines
- Position & posture changes
- Alterations of food textures, temperatures
- Utensil changes
- Changes in feeding schedule & pacing
- Oral sensorimotor program with food
- Nonnutritive oral sensorimotor program
Principles for Repeat VFSS

- Same as for initial VFSS
- Information needed for
  - Definition of etiology or diagnosis
  - Guide for management decisions
- NOT some arbitrary time interval
- Child should be at baseline

Summary

- Instrumental exam needed to define pharyngeal & upper esophageal phases of swallowing, typically VFSS
- Keep in mind: children with complex health & developmental issues may have many radiology studies throughout their lifetimes
- How will findings impact management decisions?
- A cooperative child is needed

Summary

- Inter-relationships of oropharyngeal swallowing & GI tract disorders are multifactorial & complex
- Optimal care of children with complex dysphagia & GI tract disorders requires comprehensive assessment by a team of professionals
- Monitoring over time is critical as children change

Summary Questions

- What are primary factors in determining decisions to feed orally with risks for aspiration?
- When do risks of strict NPO outweigh risks of occasional aspiration on liquid?
- What are implications of frequent deep penetration on VFSS?
- Who assumes responsibility for decisions to feed or not to feed?
Esophageal Disorders presenting as Dysphagia in Children

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I have no special interests.
I will be discussing medications used off label.

Esophageal Motility Dysfunction: Common Associated Disorders

- Eosinophilic Esophagitis
- Tracheoesophageal Fistula/Esophageal Atresia
- Cerebral Palsy
- Trisomy 21 and other genetic syndromes
- Nissen fundoplication
- Gastroesophageal Reflux
- Achalasia
- Globus – no esophageal dysfunction
Tools to Assess Esophageal Disorders that present as Dysphagia

- Esophagram
- Upper GI contrast study
- pH/impedance monitoring
- Upper endoscopy with biopsies
- Esophageal manometry

Diagnosis of GERD:
As measured by pH probe

**pH of ≤ 4:**
- Birth to 12 months:
  - >10% of time in 24 hrs
- >12 months of age:
  - >4% of time in 24 hrs
- Frequency:
  - >5 minutes duration
- Supine vs upright

**Symptom index**
- # times a symptom occurs assoc with pH < 4 divided by total number of symptoms.
  Positive ≥ 50%

Traditional pH Monitoring:
Not straightforward

**Advantages:**
- no sedation required
- functional study

**Disadvantages:**
- Difficulties with detecting reflux in patients that have continuous or frequent feeds
- Day-to-day variability
- Do you use with or without antacid?
  - Depends on what question you are asking
**Bravo™ pH Monitoring System**

**Compare Catheter pH probe with Bravo™ probe**

**Advantages:**
- Less interference with daily activities; 48hrs test

**Disadvantages:**
- Age
- Pain at the site – chest pain
- Dysphagia (68% in one study had discomfort)
- Risk of aspiration of the capsule if does not release
- Not releasing
- Requires sedation

**Impedance Monitoring**

**Definition:** ratio of voltage to current at a measuring frequency

**Air, fluid, and solid** material in the esophagus have characteristic impedance
- Intraluminal bolus movement, size, and character can be detected based on differences in conductivity to alternating current.

**Combined with pH, liquid reflux can document acid, non-acid, weakly acidic reflux**
pH/Multi-channel Intraluminal Impedance (MII) Monitor

Tracing of pH/MII probe in GERD

pH/Impedance Monitoring cont

• Advantages:
  – Tells you more physiologically and functionally what is happening
  – Improved data in infants who are being fed frequent meals.
  – Pediatric patients appear to have nonacid reflux more commonly associated with respiratory conditions

• Disadvantages:
  – The sensitivity of the study is still no more than 85% with or without medications.*
  – Day-to-day variability
  – Young infants have pH only reflux.

Esophagram

- Evaluate emptying
- Presence of appropriate stripping waves – 1°/2°
- Absence/presence of abnormal 3° waves
- Rule out obstruction

Upper GI contrast study

- Infants: rule out abnormal anatomy
  - Malrotation
  - Stenosis/Dysmotility
- Older children:
  - Hiatal hernia
  - Peptic ulcer
  - Dysmotility/Achalasia
  - Stenosis/stricture

pH Monitoring is More Accurate than Upper GI

- One study* showed detection rate of 43% with a 48% false-negative rate as compared to pH results which showed a detection rate of 83%
- But it still helps us document anatomic abnormalities

Endoscopy:
Normal Esophagus

Endoscopy:
Esophagitis

Erosive Esophagitis
“EoE” Eosinophilic Esophagitis

Esophageal manometry

- Achalasia – typically LES, but also UES
- Assess propagation of esophageal motility with swallows
- Difficult to obtain in infants and need specialty center
Case 1: TEF/EA

- 17 month old with chronic recurrent aspiration and gastroesophageal reflux
- She had been dilated for dysphagia symptoms 6 months prior.
- Follow-up UGI one month later that was normal
- New symptoms 4 months later
- Repeat UGI/esophagram and EGD confirmed proximal esophageal stricture at the anastomotic site and dilation performed.

VFSS Performed

- Upper esophageal stricture
- Could not complete VFSS because of patient refusal.
- The thin contrast passed through the narrowed area, but not the applesauce.

Esophagus Pre-Dilation
Post-Dilation

Esophagus Post-dilation
Hiatal Hernia

Post-op, patient had improved swallowing.

SML Bronchoscopy

- However, the patient’s pneumonias recurred.
- She was taken back to the OR for another diagnostic procedure: SML bronchoscopy and laryngeal cleft type 1B was diagnosed.

Eosinophilic Esophagitis (EoE)

- Presentation in an infant may be severe GERD
- Presentation in a toddler may be feeding refusal.
  - Difficult to discern between EoE and behavioral etiology to food refusal
- Upper endoscopy with biopsies of the esophagus are required to definitively diagnose EoE.
Case 2

- 2 yr old female with history of congenital heart disease s/p repair, frequent vomiting/GERD symptoms daily, and feeding refusal.
- Dysphagia symptoms by history as she would eat liquids easier than solids
- During procedure for otolaryngologist to place PE tubes, an EGD was performed.
- Severe eosinophilic esophagitis
- Treated with elemental formula and flovent and she began eating orally

Case 3

- 12 month old failing to thrive
- Feeding refusal
- Patient had no vomiting or history of GERD
- Patient had no history of feeding difficulties until the last few weeks
- EGD showed severe EE

Histopathology

White pearls seen as eosinophilic abscesses
What does eosinophilia in the esophagus mean clinically?

• **1982, Winter, et al.** correlated markers of reflux esophagitis with the presence of eosinophils
  – 18/46 pts with GERD; 12/18 had only 1 eos/hpf
  – Confirmed in adult studies
  – But Tummala, et al. found approximately 1 eosinophil in the biopsy of controls also

• Tx reflux esophagitis with positive eosinophils on biopsy aggressively → 90-95% improve

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**Eosinophilia in the esophagus, cont.**

• **1995, Kelly et al.** treated 10 pts with GERD sx, who did not respond to antireflux meds, with elemental formula

• **1993, Attwood, et al.** compared EE pts with GERD pts → 56 vs 3.3 eos/hpf

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**EE: AGA Consensus Diagnostic Guidelines***

• Clinical symptoms of esophageal dysfunction
• ≥15 Eosinophils in 1 high-power field
• Lack of responsiveness to high-dose proton pump inhibition (up to 2 mg/kg/day) or
• Normal pH monitoring in the distal esophagus
Clinical Manifestations*

- Food refusal or intolerance in infants/toddlers
- Gastroesophageal reflux symptoms
- Vomiting and abdominal pain
- Dysphagia and food impaction
- Failure to thrive
- Chest pain (17-20%)
- Diarrhea (1-24%)


Longitudinal Study in Children: Liacouras, et al. 2005

- 10 year experience, 381 children
- Most symptomatic with GERD, dysphagia
- UGI: 6% upper esophageal narrowing
- EGD: rings in 12%, 1 required dilation
- Tx: systemic steroid vs swallowed fluticasone, or dietary restriction
- Relapse rate is high once therapy discontinued

Examples of Narrowing of the Esophagus
### Intraesophageal pH Testing and Impedance

- **One impedance monitoring study** has been done in EE pediatric patients
  - Compared EE with GERD and control
  - Full column reflux nor non-acid reflux is sig in EE*
- **pH monitoring**: 9 adult and 11 pediatric studies†
  - Adults: normal in 82%
  - Pediatrics: normal in 90%

* Rosen, et al. JPGN. 2008

### Atopic Tendency in Children

- Studies show 50-80% of patients have allergic history: eosinphils vs IgE levels
- EE associated with Th 2 type immune response
  - IL-4, IL-5, IL-13 and mast cells in EE pt esophagus
- In mice, EE induced by allergen exposure in the respiratory tract after mucosal sensitization
  - May explain seasonal variation, previously reported

### Treatment for EE

- Acid-suppressive therapy may improve symptoms, but not eosinophilic infiltrate
- Swallowed fluticasone VS systemic prednisone
  - \( \rightarrow \) remission, not cure
  - Urgent relief may dictate choice
- Elimination diet: based on food RAST +/- skin prick testing and/or history
14 Years of EE:
Clinical Features and Prognosis

- Spergel, et al. JPGN 2009
- 620 patients, 333 with at least one year followup
- On presentation, 68% were < 6 years old
- GERD, feeding disorder, FTT
- 11 resolved all food allergies; 33 resolved some
- Conclusion: chronic disease: <10% resolve
- None had progression to eosinophilic gastroenteritis

Trisomy 21

- Always be aware of possible anatomic lesions of the upper GI tract from esophagus to the duodenum, especially in young children and infants, but especially those with genetic syndromes.
- 10 yo with chronic food refusal who only accepts Pediasure who has been receiving therapy her whole life for feeding.
- Started having vomiting 1 year prior that responded to prevacid.
- UGI was done and showed 2 duodenal strictures starting just a few centimeters from the bulb.
- She also had evidence of celiac disease.

Dysmotility

- In Cerebral Palsy and genetic syndromes, poor emptying of the stomach and/or slow emptying of the esophagus predispose to symptoms of food refusal that might be labeled as dysphagia or contribute to it.
- In CP, we have to be concerned for Nissen fundoplication (anti-reflux surgery) contributing to poor emptying – may contribute to aspiration pneumonia
Genetic Syndromes and Dysmotility of the GI tract

- Rett’s syndrome
- William’s syndrome
- Trisomy 21, especially if born with duodenal stenosis
- DiGeorge syndrome
- Possibly any others that might not be well-known, but present due to the increased availability and frequency of genetic testing

Questions?