2013 Charleston Swallowing Conference

Session 18
Stable And Progressive Pediatric Neurological Disorders
1:30 – 3:00 pm
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2013 Charleston Swallowing Conference
Lung / Swallowing Issues in Children with Neuromuscular Problems

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Faculty disclosures & focus
• I am a full-time faculty member at MUSC and have no conflicts of interest of any kind to disclose.
• No treatments will be discussed off-label without notice.
• This is intended to be a practical clinical discussion from a pediatric pulmonary perspective, with time for questions. It is being given as part of a multi-disciplinary workshop, so it is limited in scope / perspective.
• I apologize in advance for any errors or imprecise use of swallowing terms such as dysphagia or incoordination that I may use.

Learning objectives
After this session, learners will be able to:
• Recognize neuromuscular problems that may affect aspiration risk and severity.
• Recognize the progression of pulmonary symptoms in children with neuromuscular impairment,
• Identify outcomes of weakness with aspiration,
• Understand the importance of cough and chest symmetry in lung protection, and
• Enumerate key aspects of therapy for a child with neuromuscular issues and aspiration.
**Why is this important?**
- Demonstration of respiratory compromise in children requires muscle strength
  - Tachypnea, respiratory effort, retractions, distress
- Cough is extremely important for lung protection
- Children with neuromuscular impairment “fool us” about the severity of their respiratory problems and often can’t adequately protect their lungs
- Non-verbal children can’t express distress
- Must anticipate problems and coach families about recognition and prevention
- Pneumonia is a common cause of death

**Who are we talking about?**
- Children with neuromuscular issues
- Static or progressive
- Cerebral palsy and related problems
- Mitochondrial diseases
- Muscular dystrophies
- Spinal muscular atrophy
- Also applies to many other children
- Chronic care; children of varying ages
- Wide spectrum of manifestations

**Airway protection**
- GI and respiratory tracts have common origin, easy to aspirate
- Careful coordination to allow breathing and eating / drinking at the same time
- Poor coordination early in life and with muscle weakness; GE reflux concerning
- “Everyone aspirates” – substance, amount, and frequency influence severity
- Pulmonary protection – apnea, cough
Neurodevelopmental impairment
• Tachypnea; anatomical problems (e.g., choanal atresia, cleft palate); acute problems (e.g., bronchiolitis); chronic problems
• Loss of consciousness; seizures
• Swallowing coordination and success – textures, volume, rate of feeding
• Distraction / startle
• Sleep
• Weakness of pharyngeal muscles

Acute cough with clearance (or not)
• Airway inflammation > mucus production
• Bronchospasm (acute, chronic)
• Tachypnea; hypoxia – if marginal
• Bronchitis > pneumonia > atelectasis > bronchiectasis > fibrosis
• Everything is additive (amount, frequency)
• Goal is to minimize injury and maximize healing; importance of cough / airway clearance

Drooling as a sign of poor swallowing
• Bronchoscopy shows that such children are breathing in and out through a puddle of saliva
• Sialorrhea – how many bibs a day?
• Potential therapies to decrease salivary volume
• Impact of a g-tube bypassing pharynx
• Impact of a tracheostomy – not "sucking in" pharyngeal secretions with every breath, but liquid can drip through vocal cords.
Aspiration of noxious materials

- Saliva
- Saliva + increased bacteria (poor oral hygiene)
- Milk / formula
- Solid food
- Stomach acid
- Gastric contents (marked risk if massive amount)
- Foreign bodies (may be startle)

Common impairments

- Hypotonia of upper airway
- Obesity (no activity, parental guilt, etc.)
- Upper airway resistance vs. respiratory muscle strength (variable among diseases)
- Non-compliant / stiff chest and lungs
- Hypoventilation, especially with sleep
- GE reflux > upper airway inflammation
- Scoliosis may limit position changes; worsen cough effectiveness
- Can progress to pulmonary hypertension and even death

Management

- Minimize aspiration – from above and below;
- Promote airway clearance – chest physiotherapy (CPT); cough assist device; chest compression vest; bronchodilators
- Avoid insults – individual; seasonal; vaccines
- Minimize lung dysfunction
  - Antibiotics – acute or chronic (enteral, inhaled)
  - Mucolytics – hypertonic saline, domase alfa; N-acetyl cysteine
  - Anti-inflammatories – inhaled corticosteroids
- Anticipate, diagnose and treat respiratory failure
  - Blood gases; respiratory muscle strength
  - O2; CPAP / BiPAP / ventilation
Diagnostic evaluations

- Chest films -- scattered infiltrates; peri-bronchial cuffing; no reliable location
- Timing of episode(s) – changes can be delayed
- CT scans -- scattered (dependent) infiltrates may be more obvious
- BAL – lipid laden macrophages (not specific)
- Cultures – oral flora, anaerobes
- Pulmonary Function Testing (PFTs) – restriction, bronchospasm, muscle weakness – evolution over time
- Sleep studies – most critical time!

Diagnostic evaluations, II

- Swallowing -- observation by speech pathologist
  - Multi-texture modified barium swallow
- Gastroesophageal reflux
  - Multi-channel impedance probe study – include diary of events
- UGI used to evaluate anatomy – not function – limited time of observation
- Gastric emptying (additive problem to reflux)
- Radionuclide scans for lung deposition
- Blood gases, especially PCO₂ and base excess
- Gastric pH measurements

Take-home messages

- Aspiration is common in patients with nm issues;
- Cough is crucial for airway protection;
- Respiratory symptoms require muscle strength for recognition and assessment;
- Recurrent aspiration leads to parenchymal lung damage, hypoxemia, restrictive changes;
- Progressive muscle deterioration leads to weak cough, hypoventilation and respiratory failure;
- Mechanical airway clearance and non-invasive ventilation treatments are often helpful;
- Lung insults are additive and progressive.
References


References, II

Neurology of Pediatric Swallowing Disorders

Stephen Kinsman, MD
Pediatric Neurology
MUSC

Disclosures

- None

Issues to consider

- Localization
- Etiology/Time course
- Team functions
- Risk factors
- Assessments
- Treatments
- WHO disability model
- Family-centered care
Localization

- Anatomy
- History
- Examination
- Imaging
- Electrophysiology
- Other studies

Cranial Nerves

Central Pattern Generator of Swallowing
Motor Cortex Reorganization after Stroke

CNS - Acute

- Acute
  - Hypoxic-ischemic encephalopathy
  - Cerebral infarctions
  - Intracranial Hemorrhage
  - Infections (meningitis, encephalitis, poliomyelitis, botulism, syphilis)
  - Acute bilirubin encephalopathy
  - Metabolic encephalopathies
  - Neonatal withdrawal syndrome
  - Trauma
    - Brain and brainstem
    - Upper cervical cord

CNS – Chronic Progressive

- Chiari malformation and/or syringobulbia
- Intracranial malignancies
- Degenerative diseases
  - White matter (Metachromatic Leukodystrophy, Adrenoleukodystrophy, Krabbe)
  - Grey matter (Leukodystrophy, Tay-Sachs, Mucopolysaccharidoses)
- Mitochondrial Disorders
- Peroxisomal Disorders (Zellweger)
- Purine and pyrimidine disorders (Lesch-Nyhan)
- Disorders of copper metabolism (Wilson disease, Menkes disease)
- Spinocerebellar disorders
- Ataxia-Telangiectasia
- The Dystonias
- Multiple sclerosis
- Amyotrophic Lateral Sclerosis
- HIV encephalopathy
CNS – Chronic Static

- Genetic Syndromes
  - Velocarofacial
  - Smith-Lemli-Opitz
  - Prader-Willi
  - Trisomy 21
  - Rett syndrome
- Cerebral palsy
- Chronic bilirubin encephalopathy
- Congenital brain malformations
- Mobius syndrome
- Familial dysautonomia (Riley-Day syndrome)

PNS - Acute

- Acute inflammatory polyradiculopathy
- Hypermagnesemia
- Polymyositis
- Dermatomyositis

PNS – Chronic Progressive

- Spinal Muscular Atrophies
- Polyneuropathies
- Myasthenia Gravis
- Metabolic myopathies
  - Glycogen storage disease
  - Mitochondrial disorders
- Muscular dystrophies
PNS – Chronic Static

- Polyneuropathies
- Congenital myopathies (nemaline rod)
- Myotonic dystrophy
- Congenital muscular dystrophy
- Infantile fascioscapulohumeral dystrophy

Table 1: Definition of the neuroaxis exclusive neuroimaging findings categories

<table>
<thead>
<tr>
<th>Neuroaxis</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>Brainstem</td>
<td>Abnormal or absent neural stem structure, including bulbar areflexia and altered cranial nerve function</td>
</tr>
<tr>
<td>Spinal cord</td>
<td>Abnormal or absent neural stem structure, including areflexia and altered cranial nerve function</td>
</tr>
<tr>
<td>Basal ganglia</td>
<td>Abnormal or absent neural stem structure, including areflexia and altered cranial nerve function</td>
</tr>
<tr>
<td>Central nervous system</td>
<td>Abnormal or absent neural stem structure, including areflexia and altered cranial nerve function</td>
</tr>
</tbody>
</table>

Table 1: Feasible feeding problems in children with genetic syndromes

<table>
<thead>
<tr>
<th>Problem</th>
</tr>
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<tbody>
<tr>
<td>Dysphagia</td>
</tr>
<tr>
<td>Motor weakness</td>
</tr>
<tr>
<td>Taste disturbance</td>
</tr>
<tr>
<td>Abnormal sensory function</td>
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<tr>
<td>Weight loss</td>
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<tr>
<td>Upper airway obstruction</td>
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Conclusions

- Dysphagia is common in Pediatric Neurological conditions of both the CNS and PNS
- Proactivity and family education is critical
- Multidisciplinary care is the cornerstone.
- Trajectory thinking about conditions like cerebral palsy can help providers with family education and anticipatory guidance
- A nutritional focus is as important as a safety focus and key in maintaining a family-centered approach
Management of Pediatric Neurogenic Dysphagia

Pathology • Presentation • Treatment

Charleston Swallowing Conference
October 12, 2013
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Support and Disclosures

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- NIH NIDCD RO1/DC011290 Standardization of Videofluoroscopic Swallow Studies for Bottle-Fed Children, 2010-2015

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- Northern Speech Services, Standardizing the MBSS. http://www.northernspeech.com/MBSSimP
- Mark and Evelyn Trammell Voice and Swallowing Trust

Introduction

Management of dysphagia in the pediatric neurogenic population is arguably the most difficult population to serve.
**Oral Domain**

**Bolus Preparation**

**Physiologic Impairment**
- Impaired rotary mastication
- Imprecise lingual manipulation

**Functional Impairment**
- Reduced bolus mastication
- Incomplete bolus formation
- Risk of airway occlusion from laryngeal bolus entry

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**Oral Domain**

**Soft Palate Depression/Elevation**

**Physiologic Impairment**
- Incomplete SP to TB seal
- Incomplete velopharyngeal closure

**Functional Impairment**
- Reduced suction for bolus expression
- Posterior bolus escape
- Nasopharyngeal regurgitation
- Reduced pharyngeal bolus propulsion

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**Oral Domain**

**Lip Closure**

**Physiologic Impairment**

**Functional Impairment**

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**Pharyngeal Domain**

**Bolus Preparation**

---

**Functional Impairment**

---

**Soft Palate Depression/Elevation**

---

**Physiologic Impairment**

---

**Esophageal Domain**

---

**Physiologic Impairment**

---

**Functional Impairment**

---

**Laryngeal Vestibular Closure**

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**Pharyngeal Contraction**

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**Pharyngoesophageal Segment Opening**

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**Esophageal Clearance**

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Oral Domain
Initiation of Pharyngeal Swallow

**Physiologic Impairment**
- Delay in swallow initiation

**Functional Impairment**
- Pharyngeal bolus entry prior to laryngeal closure
- Laryngeal bolus entry before/during the swallow

Pharyngeal Domain
Pharyngeal Stripping Wave

**Physiologic Impairment**
- Reduced strength, displacement, and stripping during pharyngeal contraction

**Functional Impairment**
- Reduced esophageal bolus propulsion
- Pharyngeal residue following the swallow
- Laryngeal bolus entry following the swallow

Pharyngeal Domain
Laryngeal Elevation
Anterior Hyoid Excursion

**Physiologic Impairment**
- Reduced trajectory of motion and duration of elevation

**Functional Impairment**
- Incomplete laryngeal closure
- Laryngeal bolus entry during the swallow
- Incomplete duration and extent of PES opening
Pharyngeal Domain
Pharyngoesophageal Segment Opening

Physiologic Impairment
- Reduced extent of opening
- Reduced duration of opening

Functional Impairment
- Incomplete bolus passage into the esophagus
- Pharyngeal residue
- Laryngeal bolus entry following the swallow

EFFECTIVENESS OF TREATMENT


- Only three studies met inclusion criteria
  - Randomized controlled trial
  - Ages 0-18 with dysphagia of acquired, developmental, degenerative or genetic origin
  - Direct and indirect interventions targeting impairment, activity/participation level and environmental factors
  - Primary outcomes: physiological function, pulmonary status, diet consistency
  - Secondary outcomes: changes in growth, level of participation, caregiver stress

TREATMENT PARADIGM

- Identify physiological impairment
- Target intervention
- Set measurable objectives
- Ongoing assessment
- Team collaboration
PHYSIOLOGIC IMPAIRMENT

- ORAL Impairment
  - Poor lip closure
  - Poor lingual control
  - Weak suck
  - Poor mastication
  - Delayed initiation of swallow
- PHARYNGEAL Impairment
  - Reduced soft palate elevation
  - Reduced hyolaryngeal elevation and excursion
  - Reduced pharyngeal stripping wave
  - Reduced tongue base retraction
  - Reduced pharyngoesophageal segment opening

TYPES OF INTERVENTION

- Direct
  - Involving use of food/liquid during swallowing tasks
- Indirect
  - Motor/sensory/pharmacological interventions
    without the use of food/liquid
- Compensatory
  - Altering environmental restrictions, improving participation

(From World Health Organization, 2001)

Treatment:

NEUROLOGICAL DISORDERS

- Considerations when determining treatment plan
  - Static v progressive
  - Caregiver goals
  - Overt v silent aspiration
  - Pulmonary implications
  - Anatomical changes
Treatment: STRATEGIES

**ORAL domain**
- Increased flow rate
- Reclined position for poor bolus transfer
- Straw drinking
- Bolus intervention (Lau, C., Smith, E.O.) - administration of a single bolus via syringe for premature infants with oropharyngeal impairment; may be done with a pacifier
- Pacing

Treatment: STRATEGIES

**PHARYNGEAL domain**
- Reclined position for pharyngeal weakness
- Increased bolus viscosity
- Alternating liquids/solids
- Thickened liquids
- Chin tuck
- Head turn
- Maneuvers

Treatment: OBJECTIVES

- Target physiological impairment
- Participation objectives
- Caregiver objectives
- Nutritional outcomes
ONGOING ASSESSMENT

- Set measurable objectives
- Assess treatment effectiveness on an ongoing basis
- Adjust objectives as needed

TEAM COLLABORATION

- Ability to meet nutritional needs
- Pulmonary status and implications
- Progression of neurological status
- Caregiver goals
- NPO status

PROGRESSIVE DISORDERS

- Consider the same variables, but often a more conservative approach due to the declining mechanism and risk for severe pulmonary implications
- Team collaboration is key
REFERENCES


