Airway Protection: Clinical Management of Dysphagia and Dystussia in Neurodegenerative disease

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MESPA Conference
April 13, 2019
No conflicts of interest or disclosure to report
**Background: Airway Protection**

- Protection of the lower airways involves a continuum of behaviors

![Diagram showing the continuum of airway protection behaviors ranging from Swallowing to Cough, with intermediate steps including Glottal stop/LAR, Expiration reflex, Expiratory effort, and Throat clear.}
A Framework to Understand Airway Protection

First came Eccles 2009
Then modified by Hegland et al., 2012
Then refined again by Troche et al., 2014

Urge to act
Sensation of stimulus
Dynamic Sensorimotor pathway
Brainstem control centers

Voluntary control
Cerebral cortex
Conscious control

Spinal cord
Respiratory muscles
Oropharyngeal/Laryngeal muscles

Vagus Nerve
(Facial Nerve)
(Glossopharyngeal Nerve)
Peripheral stimulus
Background: Cough

Cough

Voluntary

Reflex

Facemask in-line with a pneumotachograph

Digitized (PowerLab) and recorded (Chart 7, ADInstruments) to computer

Irritant delivery port

Irritant delivery port

(Smith Hammond et al., 2001; 2009; Pitts et al., 2008; Troche et al., 2014; Hegland et al., 2014; Miller et al., 1996)
Researchers have found that reflex and voluntary cough dysfunction is predictive of swallowing function in PD and stroke (Pitts et al., 2008; 2010; Troche et al., 2016; Smith Hammond et al., 2001; 2009)
Background: Reflex Cough

- Reflex cough is particularly important for airway protection as it detects sensory stimuli in the airway and then forcefully ejects the material.

- The existing research has identified a cognitive motivational component to reflex cough whereby individuals can volitionally modulate the reflexive behavior.
Background:

Modulation of Reflex Cough

- Anecdotally, modulation of reflex cough is experienced when individuals suppress, or modify reflex cough output based on internal and external factors (i.e. environment, verbal cueing, etc.)
  - Hutchings et al., 1993
  - Leow et al., 2012
Background:

Modulation of Reflex Cough

- Hegland and colleagues (2012) evaluated the ability of healthy young adults to modulate reflex cough airflow based on verbal cues
  - Participants volitionally up-regulated reflex cough airflow with verbal cues to “cough long”, “cough short”, or “cough normally”

<table>
<thead>
<tr>
<th>Participants</th>
<th>Cough Airflow Measures</th>
<th>Natural Cough</th>
<th>Cough with cueing</th>
</tr>
</thead>
<tbody>
<tr>
<td>Healthy young adults (n=20)</td>
<td>CPD</td>
<td>.42 seconds</td>
<td>.57 seconds</td>
</tr>
<tr>
<td></td>
<td>PEFR</td>
<td>4.63 L/s</td>
<td>5.63 L/s</td>
</tr>
<tr>
<td></td>
<td>CVA (PEFR/PEFRT)</td>
<td>85.59 L/s/s</td>
<td>109.51 L/s/s</td>
</tr>
</tbody>
</table>
Background:
A role for cough rehabilitation?

- The ability to behaviorally modulate reflex cough airflow is important because populations with neurodegenerative diseases, such as Parkinson’s disease (PD) frequently develop dystussia
  - Hegland et al., 2014
  - Troche et al., 2014

- Brandimore et al., (2017) identified that HOAs and people with PD can improve reflex cough airflow with cueing
Cough Rehabilitation
Brandimore et al., 2017

- Evaluated the impact of simultaneous visual and verbal cueing for the immediate up-regulation of reflex and voluntary cough effectiveness
## Methods:
### Participant Demographics

<table>
<thead>
<tr>
<th></th>
<th>HOAs</th>
<th>PD</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Participants</strong></td>
<td>n=28</td>
<td>n=16</td>
</tr>
<tr>
<td><strong>Sex</strong></td>
<td>M=14; F=14</td>
<td>M=9; F=7</td>
</tr>
<tr>
<td><strong>Age (years)</strong></td>
<td>M=69.63 (8.3); F=71.33 (5.6)</td>
<td>M=74.22 (7.1); F=72.14 (4.2)</td>
</tr>
<tr>
<td><strong>Height (inches)</strong></td>
<td>M=69.56 (2.8); F=65.56 (2.6)</td>
<td>M=68.17 (2.3); F=62.79 (2.2)</td>
</tr>
<tr>
<td><strong>Weight (pounds)</strong></td>
<td>M=201.06 (40.6); F=145.67 (31.6)</td>
<td>M=182.0 (26.8); F=137.29 (22.0)</td>
</tr>
<tr>
<td><strong>BMI</strong></td>
<td>M=27.68 (4.3); F=25.69 (5.8)</td>
<td>M=27.48 (3.2); F=24.21 (4.1)</td>
</tr>
</tbody>
</table>
Methods:

Baseline Cough Testing

- **Voluntary Sequential Cough Testing**
  - Instructed to: “Cough like something went down the wrong pipe” (3x)

- **Reflex Cough Testing**
  - Obtained baseline reflex cough airflow at 200 µM capsaicin (3x)
  - Instructed to: “Cough if you need to”

Facemask in-line with a pneumotachograph

Irritant delivery port; 200 µM capsaicin

Digitized (PowerLab) and recorded (Chart 7, ADInstruments)

to computer

**Irritant delivery port:** 200 µM capsaicin

**Facemask in-line with a pneumotachograph:**

**Digitized (PowerLab) and recorded (Chart 7, ADInstruments):**

**to computer:**
Methods:
Modulated Reflex and Voluntary Cough Testing

- Cough Modulation Testing
  - Randomized presentations of 0 and 200 μM capsaicin

Cough Modulation Testing ("Cough as hard as you can and hit that green line")

<table>
<thead>
<tr>
<th>Block 1</th>
<th># coughs</th>
<th>Block 2</th>
<th># coughs</th>
</tr>
</thead>
<tbody>
<tr>
<td>0 μM</td>
<td>_______</td>
<td>0 μM</td>
<td>_______</td>
</tr>
<tr>
<td>0 μM</td>
<td>_______</td>
<td>200 μM</td>
<td>_______</td>
</tr>
<tr>
<td>200 μM</td>
<td>_______</td>
<td>200 μM</td>
<td>_______</td>
</tr>
</tbody>
</table>
Summary of Findings

- We identified that HOAs and people with PD can improve both reflex and voluntary cough effectiveness
  - Visual and auditory cueing to ‘cough hard’ effectively increased PEFR and CEV for both cough types
  - Modulated coughs > Baseline coughs
    - PEFR: Increased up to 50%
    - CEV: Increased up to 100%
Summary of Findings

- There were clearly different mechanisms by which HOAs and people with PD modulated reflex and voluntary cough
  - Whereas HOAs increase IV and decrease CPD; there were no significant differences in IV and CPD for people with PD
Discussion: Reflex vs. Voluntary Cough

- During modulation, voluntary cough = reflex cough for PEFR and CEV
  - May highlight differences in task execution
    - Effort
    - Stereotypic response
Discussion:

Conclusions

- The results of this research suggest that people with PD (and likely others) are amenable to up-regulation of reflex and voluntary cough function via cueing strategies.

- Certainly gives us one more treatment target for dystussia.

- May contribute to improved airway protective outcomes.
Clinical Implications

Acute Care

• Incentive spirometry
  • Expands pulmonary tissues and may promote a more even bacterial kill during recovery
  • For patients who cannot tolerate mild exercise
  • No resistance

• Peak expiratory airflow meters
  • Inexpensive
  • Proxy for cough effectiveness
  • Voluntary only
Clinical Implications

Acute Care

• Perceptual measures
  – Laciuga et al., 2015
  – UTC

• Cough Screening
  – Hegland et al., 2016
  – Active NIH research investigating the utility of FOG and capsaicin for the identification of dysphagia in neurodegenerative diseases
Clinical Implications

Outpatient

- EMST
  - PD
  - Stroke
  - MS
  - COPD

- Results:
  - *Increased* maximum expiratory pressure
  - *Increased* cough airflow and effectiveness
  - *Increased* perceived magnitude of the UTC
  - *Improved* swallowing function
Clinical Implications

Outpatient

• IMST
  – Rehabilitation program aimed to strength inspiratory muscles and decrease work to breathe
  – Populations: asthma, emphysema, restrictive pulmonary disorders, ventilator dependent

• Cough Biofeedback
  – Verbal
  – Visual
  – Education
  – Frequency
Clinical Implications: Procedures

• Cough evaluation
  • Voluntary single and sequential cough production
  • Reflex cough with FOG or Capsaicin
    • Assess UTC
    • Research only

• Videofluoroscopic Rehab Barium Swallowing evaluation
  • Presentations of barium: thin, nectar, pudding, and pill
  • Assessment of UTC
Evaluation

No Treatment

Patient and caregiver counseling and education

Compensatory

Speech: Voice amp, prosthetics, environmental modifications (caregiver training), AAC (low and high)

Swallow: Postural changes, diet modifications, PEG

Cough: Preventative cough or throat clear during meals

Treatment

Rehabilitation

Speech: LSVT, MPT, rate control therapy, IMST, ARCS, Lips, VNeST

Swallow: Masako, effortful swallow, Mendelsohn, Supraglottic, Shaker, EMST

Cough: UTC awareness training, cough modeling, biofeedback

Combined rehabilitation and compensation
Case 1: Corticobasal Syndrome

History: June 2018

- 69 year-old female presents to your clinic with a diagnosis of CBS with symptom onset in Spring 2014 (right arm tremor).

- The patient serves as primary historian and is well-known to the clinic.

- PMH = August 2017: Our evaluations revealed a moderate hypokinetic dysarthria and a moderate oropharyngeal dysphagia characterized by consistent penetration to the level of the vocal folds with thin liquids (PA = 5), moderate residue throughout mechanism, tongue pumping, and pre-swallow spill of all consistencies to the pyriform sinuses.
  - At that time, the chin tuck was unsuccessful at improving airway protection: several effortful swallows were required.
  - The patient began therapy at your clinic: maximum performance (i.e. LSVT), traditional swallowing exercises, and EMST.
Case 1: Corticobasal Syndrome

• Today the patient reports no changes in symptom status.

• She has no history of pneumonia, diet modifications, or weight loss.

• However, she is reporting coughing with thin liquids.

• She is ambulatory; however notes increased falls resulting in a broken left elbow.

• The patient believes that therapy was helpful, but has lost her EMST device and is no longer performing exercises.
Case 1: Parkinson’s disease

Let’s watch...
Case 1: Swallowing Evaluation

Please rate your urge-to-cough

<table>
<thead>
<tr>
<th>Rating</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>None at all</td>
</tr>
<tr>
<td>1</td>
<td>Very slight</td>
</tr>
<tr>
<td>2</td>
<td>Slight</td>
</tr>
<tr>
<td>3</td>
<td>Moderate</td>
</tr>
<tr>
<td>4</td>
<td>Somewhat severe</td>
</tr>
<tr>
<td>5</td>
<td>Severe (heavy)</td>
</tr>
<tr>
<td>6</td>
<td>Very, very severe</td>
</tr>
<tr>
<td>7</td>
<td></td>
</tr>
<tr>
<td>8</td>
<td></td>
</tr>
<tr>
<td>9</td>
<td></td>
</tr>
<tr>
<td>10</td>
<td>Very, very, very severe</td>
</tr>
<tr>
<td></td>
<td>almost maximal</td>
</tr>
</tbody>
</table>
Cough Assessment
Case 1 Results: Cough

Voluntary sequential cough

PEFR = 1.9 L/sec
CPD = 0

<table>
<thead>
<tr>
<th>Pulmonary Function</th>
<th>Expected Performance</th>
<th>Patient's Performance (45%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>FVC</td>
<td>3.34</td>
<td>1.53</td>
</tr>
<tr>
<td>PEF</td>
<td>6.28</td>
<td>3.49</td>
</tr>
<tr>
<td>FEVI</td>
<td>2.57</td>
<td>1.33</td>
</tr>
</tbody>
</table>
Case 1: Results

Results:
Speech evaluation – Moderate-severe hypokinetic dysarthria (previously moderate)

Swallowing evaluation – Advanced to moderate-severe oropharyngeal dysphagia. Characterized by intermittent silent aspiration with thin liquids (PA = 8), and consistent penetration to the vocal folds. The patient was cued to cough; however, cough was ineffective to clear material from airway. UTC = 0.

Cough evaluation – Ineffective voluntary cough production. PEFR = 1.9 L/sec of predicted 5 L/sec
Case 1: Treatment Plan

**Treatment plan:**
Speech: MPT and rate control therapy

Swallowing: EMST, and swallowing exercises
   - Masako, effortful swallow, Mendelsohn, super-supraglottic, etc.

Cough: Cough modeling/biofeedback

Compensations:
1.) Temporarily down-grade to nectar-thickened liquids
2.) Small bites/sips of solids/liquids
3.) Chew food thoroughly
4.) Maintain good oral hygiene
5.) Produce a preventative cough followed by an effortful swallow when material is sensed near the airway.
Case 1: Progress

- The patient re-initiated therapy at our clinic and has received 5 sessions (~once/2weeks). She remains motivated.

- Additionally, she participates in PT and a dance class offered on Thursday nights.

- Improved coordination and efficiency with EMST and swallowing exercises

- Cough remains ineffective, but improved ability to produce voluntary cough on command
Case 2: Parkinsonism

**History: May 2018**

- 70 year-old male presents to your clinic with a diagnosis of PD with symptom onset in 2008.
- The patient and his wife provide history.
- PMH = July 2017: Our evaluations revealed a moderate hypokinetic dysarthria and a moderate oropharyngeal dysphagia characterized by intermittent penetration to the level of the vocal folds with thin liquids (PA = 5), and intermittent silent aspiration (PA = 8) with moderate-severe residue in the valleculae and moderate residue in the pyriform sinuses, tongue pumping, and pre-swallow spill of all consistencies to the pyriform sinuses.
  - Chin tuck, effortful swallow and dry swallow were somewhat successful to at improving airway protection
  - The patient began therapy at your clinic: Traditional swallowing exercises, and EMST.
Case 2: History

• Today the patient reports maintenance of swallowing function.

• However, he acknowledges random coughing and choking during meals with fatigue, has modified diet to include softer foods, and complains of a wet vocal quality.

• He has no history of pneumonia but notes a 23 pound weight loss over the last year.

• The patient believes that therapy was helpful, but has lost his EMST device and is no longer performing exercises.
Case 2: Swallowing QOL

EAT-10 Survey
EAT-10 Score: 21/40

1. My swallowing problem has caused me to lose weight: 2
2. My swallowing problem interferes with my ability to go out for meals: 3
3. Swallowing liquid takes extra effort: 2
4. Swallowing solids takes extra effort: 2
5. Swallowing pills takes extra effort: 3
6. Swallowing is painful: 2
7. The pleasure of eating is affected by my swallowing: 3
8. When I swallow, food sticks in my throat: 1
9. I cough when I eat: 0
10. Swallowing is stressful: 3
Case 2: Cough Results

Voluntary sequential cough

Voluntary cough evaluation: PEF = 2 L/sec

What can we say about the overall organization of cough?
Case 2 Results: Swallowing
Case 2: Results

Results:
Swallowing evaluation: Moderate-severe sensorimotor oropharyngeal dysphagia consistent penetration and silent aspiration to the level of the vocal folds with thin liquids (PA = 8), reduced aspiration with nectar-thickened liquids, moderate-severe vallecular residue and moderate pyriform sinus residue.

Urge to cough associated with swallowing (UTC = 2).

Cough evaluation: Extremely ineffective voluntary cough = 1.9 L/sec of expected.
Case 2: Treatment

Treatment Plan: Closer to home
Assess reflex cough on re-evaluation

Rec: Soft solids diet with nectar-thickened liquids, small bites and sips

Re-initiate behavioral exercises to include: effortful swallow and Masako and EMST

Cough modeling and biofeedback

Counsel regarding UTC and the necessity of producing a cough

Contact the treating clinician