Dysphagia Assessment & Intervention in Neurodegenerative Diseases

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Objectives

- Brief review: Neuromuscular pathology
- Brief review: Age-related factors
- Brief review: Pulmonary defenses
- Assessment methods:
  - Clinical swallowing exam (CSE)
  - Instrumental exams
- Principles of intervention
  - Influence of medical diagnosis and prognosis
  - Evidence for selected interventions
- Time permitting – Cases will be discussed
A basic overview of

**Neuromuscular Pathology**

**Underlying Pathology**

*It is critical to understand the underlying pathology!*

- Facilitates:
  - Development of clinical hypotheses
  - Estimation of prognosis
  - Development of intervention plan

**Neuromuscular Levels of Pathology**

*Miller & Britton (2011)*

- Muscle disease/ myopathy
- Neuromuscular junction (NMJ) disease
- Peripheral nerve disease (PNS)
- Motor neuron disease (MND)
- Diseases of the central nervous system (CNS)*
Muscle disease: “Myopathy”

A change in the muscle fibers OR muscle response to a neural impulse

Many potential causes:
– Genetic defects
– Autoimmune inflammation
– Endocrine disorders
– Metabolic disease
– Infectious agents
– Toxic conditions

Neuromuscular Junction (NMJ) Disease

• Impairment of neural transmission at the NMJ
• Possible causes:
  – Myasthenia gravis (MG)
  – Lambert-Eaton Myasthenic syndrome (LEMS)
  – Blockage of Ach release due to toxins: e.g., botulism, curare
  – Tetanus

Peripheral Nerve disease

• Impairment of cranial nerves and/or spinal nerves.

• Peripheral neuropathy symptoms vary depending on the specific nerves affected.
Motor Neuron Disease (MND)

- A group of progressive disorders characterized by destruction of motor neurons.
  - Upper motor pathology, e.g.,
    - Spasticity / clonus
    - Hyper-reflexia
    - Hypertonia
    - Loss of dexterity
    - Pseudobulbar signs
  - Lower motor neuron pathology, e.g.,
    - Weakness
    - Hypotonia
    - Atrophy & fasciculations
    - Diminished reflexes

Types of Motor Neuron Disease

<table>
<thead>
<tr>
<th>Type</th>
<th>Classic Amyotrophic Lateral Sclerosis (ALS)</th>
<th>Progressive Bulbar Palsy (PBP)</th>
<th>Primary Lateral Sclerosis (PLS)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Muscle involvement</td>
<td>Bulbar &amp; Spinal</td>
<td>Bulbar only</td>
<td>Bulbar &amp; Spinal</td>
</tr>
<tr>
<td>Neurological signs</td>
<td>UMN &amp; LMN</td>
<td>UMN &amp;/or LMN</td>
<td>UMN only</td>
</tr>
<tr>
<td>Prognosis</td>
<td>Median survival 4.3 years</td>
<td>Median survival 2-3 years; typically progresses to ALS</td>
<td>Survival much longer than ALS. Higher levels of independence for years or decades</td>
</tr>
</tbody>
</table>

Table modified from Miller & Britton, 2011; References: Gordon et. al., 2006; Czaplinski et al., 2006; Leigh et al., 2003

Central Nervous System (CNS) - Demyelinating Disease

Demyelination refers to damage or loss of the myelin sheath

- PNS: e.g., Guillain-Barre syndrome, Charcot-Marie-Tooth
- CNS Examples
  - **Multiple sclerosis (MS)**
  - Progressive multifocal leukoencephalopathy (PML)
Central Nervous System (CNS) – Movement Disorders

• Basal ganglia disorders
  – Hypokinesias
    • Parkinson’s disease
    • Progressive supranuclear palsy (PSP)
    • Corticobasal ganglionic degeneration
  – Hyperkinesias
    • Huntington’s Disease (HD)
    • Dystonia
    • Tremor

• Cerebellar disorders
  – Ataxia syndromes

• Combination syndromes, e.g.,
  – Multiple System Atrophy (MSA)
  – Parkinsonism-dementia-ALS complex

Neurodegenerative Dementias

4 primary types (irreversible):
  – Alzheimer’s dementia (AD)
  – Vascular dementia (VaD) (Multi-infarct)
  – Lewy body dementia (LBD)
  – Frontotemporal Lobar Dementia (FTLD)

Frontotemporal lobar dementia (FTLD)

• Focal cortical atrophy in frontal and/or anterior temporal lobes

• Clinical syndromes:
  – Frontotemporal dementia (FTD)
  – Semantic dementia
  – Primary progressive aphasia (PPA)
Neurodegenerative Dementias
Subcortical Dementias

Overlap with movement disorders....

• Huntington’s Disease (HD)
• Parkinson’s Disease (PD)
• Progressive Supranuclear Palsy (PSP)
• Creutzfeldt-Jakob Disease (CJD)

Other conditions associated with dementia....

• Wilson’s Disease
• HIV Encephalopathy
• Wernicke’s Encephalopathy (ETOH related)
• Pugilistic encephalopathy (aka post-traumatic Parkinsonism)

Factors to Consider

• Pattern of progression
• Bulbar involvement
• Association between dysphagia and dysarthria (Yorkston et al., 2013)
• Respiratory impairments
• Age-related factors
• Cognitive impairments
Age-related factors

Presbyphagia vs. Dysphagia
Cichero & Altman, 2012; Keller 1993; Ney et al., 2009

– “Presbyphagia”: Alternations in the swallowing mechanism of otherwise healthy older adults
  • Diminished functional reserve
  • Increased risk for dysphagia
  • Increased risk for malnutrition

– “Dysphagia”: Impairment of the ability to swallow

Contributing factor: Sarcopenia
(Evans, 1995; Fielding et al., 2011; Maeda & Akagi, 2015)

• Age-related loss of skeletal muscle mass and function, characterized by reductions in...
  – Muscle mass / Cross-sectional area
  – Number &/or size of muscle fibers
  – Strength

• Independent risk factor for dysphagia in elderly individuals
• Dryness may involve mouth, pharynx and esophagus

• Reduced and altered salivary secretion
  – ↓ number of saliva producing acinar cells
  – ↑ salivary protein concentration

• Functional impact
  – Reduced lubrication and moisture for chewing / swallowing and diminished taste sensation
  – Changes to rheological (flow) properties of saliva
  – May impact food digestion process

Contributing factor: Xerostomia
(Cassolato & Turnbull 2003; Ghezzi & Ship, 2003; Nagler & Hershkovich, 2005; Thomson 2015)

• Polypharmacy / medication side effects
  – Xerostomia
  – Taste changes
  – Mouth burning
  – Reduced level of alertness
  – Mental status changes
  – Lower esophageal relaxation and reflux
  – Esophagitis

• Difficulty with swallowing pills

Contributing factor: Medications
(Ichikawa et al., 2011; Nagler & Hershkovich, 2005; Ney et al., 2009; Robbins et al., 2007)

• Taste / smell
  (Heft & Robinson, 2014; Schiffman, 1997; Schiffman & Graham, 2000)

• Intraoral tactile sensation
  (Heft & Robinson, 2014; Steele et al., 2014)

• Temperature
  (Heft & Robinson, 2014; Schiffman & Graham, 2000)

Aging and Sensory Changes
Aging – Impact on oral phase

- Reduced lingual strength and mobility
  (Clark & Solomon, 2012; Youmans et al., 2009)
- Slower oral transit
  (Shaw et al., 1995)
- Delayed initiation of the pharyngeal swallow
  (Robbins et al., 1992; Tracy et al., 1989)
- Alterations to dentition
  (Paganini-Hill et al., 2011)

Aging – Impact on pharyngeal phase

- Laryngeal & pharyngeal anatomy
  – Vocal fold changes, e.g., bowing
  – Increased laryngeal descent (Yamashiro & Kryger, 2012)
  – Increase pharyngeal lumen size (Molfenter et al., 2015)
- Weakness (Dejaeger et al., 1997)
  – Reduced tongue driving force
  – Diminished pharyngeal contractions
- Airway protection & Stasis
  – Increased frequency and extent of “penetration” (Robbins et al., 1999)
  – Pharyngeal residue: valleculae & pyriform (Dejaeger et al., 1997)
- Upper esophageal sphincter
  – Delayed and reduced extent of opening (Shaw et al., 1995)

Aging: Impact on Esophageal phase

- Reduced esophageal motility
  – Increased non-peristaltic contractions
- Inadequate esophageal clearance
  – Delayed esophageal emptying
  – Increased esophageal dilation and stasis
  – Increased risk for reflux

(Ney et al., 2009; Zboralske et al., 1964)
Aging: Impact on pulmonary function

- **Anatomic changes** (Chong & Street 2008; Lalley 2013; Ramly et al., 2015)
  - Spinal changes, e.g., kyphosis, rigidity of joints
  - Lung parenchyma, e.g., ↑ compliance, ↓ recoil
  - Respiratory muscles, e.g., ↓ diaphragm strength
  - Pulmonary defenses, e.g., mucociliary dysfunction; ↓ cough strength & effectiveness
  - Increased risk for pneumonia

- **Breathing–swallowing coordination** (Hiss et al., 2001; Shaker et al., 1992)
  - ↑ swallow apnea duration
  - Changes to respiratory phase patterns

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A Brief Overview

**Pulmonary Defenses**

Pulmonary defenses guard the lungs from infections. Pulmonary defenses include protective.....

- Anatomic features
- Mucociliary clearance
- Reflexes, including cough
- Volitional cough
- Cellular defenses

Certain conditions and interventions can impair pulmonary defenses
Pulmonary defenses

The gas exchange surface of the lung is the largest surface area in the body that is exposed to the outside environment.

Anatomic Airway Defenses

- Nose
- Nasopharynx
- Branching Tracheobronchial Tree

- Swallowing - related
  - Laryngeal closure
  - Valleculeae & pyriform sinuses
  - Epiglottic inversion
  - Laryngeal closure
  - UES contraction / closure – e.g., to prevent reflux

The Nose is a Filter

Loss of Anatomic Airway Defenses

- Mouth Breathing (Exercise)
- Trauma or Obstruction
- Intubation or Tracheostomy
Mucociliary clearance, the "Mucociliary Escalator"

Bronchial-to-Aveolar Epithelial Changes

Creative commons
Failure of Mucociliary Escalator Function
– Examples (e.g., Tilley e al., 2014)

- Genetic disorders
  - Primary Ciliary Dyskinesia
  - Cystic Fibrosis
- Acquired
  - Smoking
  - Other environmental factors, air pollution, allergins
  - Infection (e.g. Influenza virus)
  - Obstruction (e.g. endotracheal tube, trach, tumor)
  - Dehydration

Airway Defense Reflexes

Examples:
- Sneeze
- Swallow
- Gag
- Laryngeal closure
- Huff
- Cough

Cough

Rapid mass movement of secretions and foreign material out of the conducting airways
“The cough reflex is the watchdog of the lungs” (C. Jackson)

Cough receptors

- Mechanosensitive – located primarily in larynx and trachea
- Chemosensitive – located primarily in distal airways

- Cough may be volitional or reflexive
  (Lasserson et al., 2006; Stephens et al., 2003; Widdicombe & Fontana, 2006)
- Normally function - Can exert volitional control over cough – even when reflexive (e.g., Hegland et al., 2011, 2012)

Phases of normal cough

Inspiration  Compression  Expulsion
Active Vocal Fold Movements During Cough

Measurement of cough effectiveness

- Inspiration: A to B
- Compression phase: B to C
- Expulsion phase rise time (EPRT): C to D
- Peak expiratory cough flow (PECF): D
- "Volume acceleration" (VA): PECF / EPRT

Dystussia – contributing factors

- Respiratory muscle weakness
  - Aging, e.g., (Freitas et al., 2010)
  - Neurological disease, e.g., MS (Aiello et al., 2008), SCI & Neuromuscular disease (Kang & Bach, 2000; Park et al., 2010)
- Laryngeal impairments, e.g., (Britton et al., 2014; Mahajan et al., 1994)
- Reduced cough reflex sensitivity:
  - Aging: e.g., (Newnham & Hamilton, 1997)
  - Smokers: e.g., (Dicpinigaitis, 2003)
Failure of cough & other upper airway reflexes may occur due to:

- Any form of sedation
- Anesthesia
- Neurological injury or disease
- Structural disease
- Impaired glottic control
- Reduced expiratory function
  - Force
  - Flow

### Examples: Cough Airflow

**Healthy Male**

**Male with ALS**

### Associations between laryngeal and cough dysfunction in bulbar motor neuron disease (MND) – Britton et al., 2014

Airflow analyses: MND vs Controls
MND: n=12 Controls: n=12
Dystussia – Additional Clinical Implications

• Increased risk for....
  – Aspiration
    • Objective measures of volitional cough
      – Stroke: (Smith-Hammond et al., 2009)
      – Parkinson’s Disease (Pitts et al., 2008 & 2010)
    • Cough reflex testing with citric acid (Miles et al., 2013)
  – Aspiration pneumonia
    (Addington et al., 1999; Yamanda et al., 2008)
  – Respiratory distress &/or Respiratory failure
    (Hoesch et al., 2012; Su et al., 2010; Bianchi et al., 2012)

Defenses at the Alveolar Level

The alveolar macrophage!

http://www.sflorg.com/sciencenews/scn041906_01.html
Additional defenses at the alveolar / cellular level

- Surfactant
- Anti-bacterial substances
- Neutrophils
- Lymphocytes

Lowered resistance

- Immunodeficiency
- Virulent infections
- Chronic diseases: diabetes, alcoholism
- Malnourishment
- Leukopenia
- Immunosuppression

Pulmonary Defense Risk Factors for Pneumonia

- Oral Hygiene
- Loss of anatomical defenses (e.g., nose, larynx)
  - E.g., trach, endotracheal intubation
- Blunted airway defense reflexes, e.g., swallowing, cough, laryngeal closure
  - E.g., stroke, brain injury, seizure, alcohol &/or drug intoxication, anesthesia, neuromuscular disease
- Weak or absent ability to cough
  - E.g., tetraplegia, neuromuscular disease, COPD
- Impaired mucociliary clearance
  - E.g., smoking, viral infection, COPD, bronchiectasis, inherited ciliary disorders, airway obstruction, dehydration
- Weakened immunologic/cellular defenses
  - E.g., HIV, transplant, immunosuppressive therapy, alcoholism, malnutrition, diabetes, immunoglobulin deficiency

Table modified from Miller & Britton (2011); Happel et al., 2004
Assessment

Clinical Swallowing Examination (CSE)

An integral component of swallowing assessment

Utility of the CSE

- Determine underlying neuromuscular function
  - Onset, progression pattern, symptoms/signs
  - Neuromuscular observations—across systems

- Determine other factors that influence swallowing status, interventions & risk for aspiration pneumonia (Langmore et al., 1998)
- Determine need for & type of instrumental assessment
- Estimate swallowing ability, potential benefit from treatment
- Estimate risk for aspiration pneumonia and/or pneumonitis
- Monitor improvements or progression
- Estimate prognosis
- Estimate long-term outcomes (Smithard et al., 1996)
Clinical Swallowing Examination (CSE)

Includes three components:
1. History
2. Physical / oral mechanism examination
   - Including consideration of respiratory function
3. Assessment of swallowing function
   (Yorkston, Miller, Strand & Britton, 2013)

History

- Primary & concomitant medical diagnosis
- Medications
- Current speech and swallowing symptoms
- Other ancillary symptoms
- Current management of symptoms
- Treatment expectations & goals
- Advanced directives

CSE: Informal / Ancillary Observations
Miller & Britton (2011)

<table>
<thead>
<tr>
<th>INFORMAL OBSERVATIONS</th>
<th>GENERAL PHYSICAL FINDINGS</th>
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<tbody>
<tr>
<td>LOA</td>
<td>Respiration</td>
</tr>
<tr>
<td>Behavior</td>
<td>Speech/Voice</td>
</tr>
<tr>
<td>Affect</td>
<td>Upper extremity function</td>
</tr>
<tr>
<td>Nutritional state</td>
<td>Mobility</td>
</tr>
<tr>
<td>Motor speech</td>
<td>Coordination</td>
</tr>
<tr>
<td>Language</td>
<td>Muscle and bodily stability</td>
</tr>
<tr>
<td>Mental Status</td>
<td>Visual system</td>
</tr>
<tr>
<td>Independence</td>
<td>Auditory system</td>
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<tr>
<td>Hydration</td>
<td></td>
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<tr>
<td>Medical /health aids</td>
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</table>
Potential Impact of Neurodegenerative Disease on Respiratory Function

Physical / Oral Mechanism Examination

• Examination of muscles innervated by cranial nerves
  – At minimum, CNs V, VII, IX, X, and XII

• Intraoral examination

• Motor speech

• Respiratory examination

Sensory Function: Trigeminal (CN V)

From Management of Speech and Swallowing Disorders in Degenerative Diseases, Third Edition (p. 13), by K. Yorkston, R. Miller, E. Strand and D. Britton, 2013, Austin, TX: PROED. Copyright 2013 by PRO-ED, Inc.
**Facial & Jaw Muscles**

LMN Impairment  
Upper & lower facial weakness  
Weakness is unilateral to lesion

UMN Impairment  
Lower facial weakness  
Weakness is contralateral to lesion  
Miller & Britton (2011)

Odds of aspiration with incomplete facial symmetry = 0.76 times the odds of aspiration of those with complete facial symmetry. Isolated incomplete labial closure did not affect the odds of aspiration.  
(Leder et al., 2013)

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**CN IX & X**

LMN Lesion  
UMN Lesion  
Gag: Careful Interpretation Needed  
Miller & Britton (2011)

Laryngeal Examination

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**Tongue: CN 12 Hypoglossal N**

LMN Impairment  
– Right side  
Miller & Britton (2011)

UMN Impairment  
– Right side

Odds of aspiration with incomplete lingual ROM = 2.72 times the odds of aspiration of those with complete lingual ROM. Independent risk factor for aspiration.  
(Leder et al., 2013)
Intraoral Examination

Coordination of Breathing & Swallowing

Swallow apnea duration (SAD)  Respiratory-phase patterns

PATHOPHYSIOLOGY

- Increased post swallow inspiration in individuals with neurodegenerative disease
  - Motor Neuron Disease: Hadjikoutis et al., 2000
  - Parkinson’s Disease: Gross et al., 2008, Troche et al., 2011
- Increased post swallow inspiration associated with advanced age (Martin-Harris et al., 2005)
- Shorter SAD associated with hypercapnia (Boden et al., 2009)

Subglottic Pressure Support for Swallowing

- Facilitates healthy swallowing
  - May aid expiratory airflow following swallow apnea (Lang et al. 2002; Nishino & Honda, 1986)
  - May stimulate mechanoreceptors to aid laryngeal adduction (Shin et al., 1988)
  - Swallowing timing and efficiency is aided by higher lung volume (Gross, 2009)
- PATHOPHYSIOLOGY
  - Prolonged swallowing associated with lower lung volume (Gross, 2009; Gross et al., 2003)
  - Improved swallowing efficiency and timing when trached patients were on (vs. off) mechanical ventilation, and associated with higher MIP (Terzi et al., 2007)
Pulmonary function measures:
• Lung volumes
• Flow rates
• Max inspiratory & expiratory pressures: MIP/MEP
• Diffusing capacity (DLCO)

Additional Pulmonary measures:
• Arterial blood gas (ABG) or Non-invasive CO2 monitoring
  Normal PCO2: 40 +/- 3 mmHg
  >45 mmHg = respiratory acidosis (Hypercapnia)
• O2 saturations—utility—mixed in the literature. Per Wang et al. (2005)—cannot reliably detect aspiration.
• Respiratory rate: 12-20 breaths/min WNL

Pulmonary Function: Spirometry

• FEV1: volume exhaled in the first second of a forced exhalation
• FVC: forced vital capacity or the total volume exhaled during forced exhalation
• FEV1/FVC: measure of airflow obstruction

  Normal
  – FEV1 and FVC > 80% of normal
  – FEV1/FVC > 75%
  – TLC > 80% of normal
  – Published norms: Hankinson et al. 1999; CDC website

  Obstructive pattern
  – FEV1/FVC < 75%

  Restrictive pattern
  – TLC < 80% of normal

Measurement of Cough Subjective Judgment or Peak Cough Flow

• Peak cough flow (PCEF) = maximal expiratory flow rate during a cough maneuver.

<table>
<thead>
<tr>
<th>PCF Level</th>
<th>Clinical significance</th>
</tr>
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<tbody>
<tr>
<td>Greater than 500 L/min</td>
<td>Typical threshold for healthy adults; minimal risk of airway encumbrance</td>
</tr>
<tr>
<td>Less than 270 L/min</td>
<td>Increased risk for airway encumbrance</td>
</tr>
<tr>
<td>160 L/min</td>
<td>Minimum threshold to move mucous from lungs into the upper airway</td>
</tr>
</tbody>
</table>

References: Bach & Saporito, 1996; Boitano, 2006; Toussaint et al., 2009; Table from: Britton, Cleary and Miller, 2013
Symptoms & Signs of Hypoventilation

- Dyspnea on exertion or talking
- Disturbed sleep
- Morning headaches
- Excessive daytime sleepiness
- Fatigue
- Difficulty clearing secretions
- Reduced appetite
- Depression
- Poor concentration/memory
- Tachypnea
- Paradoxical breathing pattern
- Reduced chest expansion
- Weak cough
- Sweating
- Tachycardia
- Weight loss
- Hallucinations
- Dizziness or syncope
- Cyanosis
- Clubbing
- Orthopnea

Laryngeal & Swallowing Examination

Miller & Britton (2011)

Instrumental Assessments

VFSS & FEES are superior to CSE in assessing biomechanics of swallowing and detection of aspiration.

Factors to consider:

- Type of instrumental assessment
- Medical necessity—indicators for instrumental assessment
- Value—cost benefit ratio
- Timing
  - When to assess
  - How often to repeat
- Interpretation
### VFSS vs. FEES – Langmore (2006)

**Only VFSS**
- Bolus during height of swallow
- Oral and esophageal phases
- Tongue retraction
- UES opening
- Laryngeal elevation
- Extent of aspiration
- Submucosal changes, e.g., osteophytes

**Only FEES**
- Secretions
- Sensation (direct assessment)
- Surface anatomy
- Mucosal abnormalities
- Glottic closure
- Path of bolus (direct)
- Location of bolus in hypopharynx
- Effect of altered anatomy on bolus flow and airway protection.

### Key Information: Neuromuscular Condition

- Underlying pathophysiology & cause (if known)
- What is typical in this condition?
  - Pattern of progression
  - Pattern of muscle weakness
  - Other signs/symptoms
  - Medical intervention
  - Typical effects on swallowing, speech, respiration, as well as other bodily functions

### Directions for Future Research

- Relationship between the CSE and longer term outcomes, e.g., nutritional status, aspiration pneumonia, etc.
- Frequency and timing of instrumental studies in the context of progressive neurodegenerative conditions
Examples

Example: Myotonic Muscular Dystrophy (MMD)

Etiology
Pattern of progression
Pattern of muscle weakness
Other signs / symptoms
Cognitive sequelae
Medical intervention
Dysarthria or other speech symptoms
Respiration

Dysphagia in MMD

- Prevalence: 25–80% (Willig et al., 1994; Ertekin et al., 2001)
- Oral phase:
  - Oral phase impairments are typically more mild (Leonard, 2012)
  - Chewing and bolus control (Odman & Kilaridis, 1996)
- Pharyngeal phase:
  - Delayed / prolonged laryngeal elevation (Ertekin et al. 2001)
  - Valleculea and pyriform residue (Osanai, 2004)
  - Reduced strength of pharyngeal contractions (Leonard et al. 2000)
- Esophageal phase:
  - Incomplete relaxation of the PES and hypotonia of the esophagus (Marcon et al., 1998)
- Other:
  - Reduced awareness of dysphagia is common (Leonard, 2010)
Example: Myasthenia Gravis (MG)

- **Etiology**
- **Pattern of progression**
- **Pattern of muscle weakness**
- **Other signs / symptoms**
- **Cognitive sequelae**
- **Medical intervention**
- **Dysarthria or other speech symptoms**
- **Respiration**

**Dysphagia in Myasthenia Gravis**

- Dysphagia is common
- Dysphagia may co-occur with breathing, speech, voice impairments
- Presentation varies, depending on...
  - Fluctuating weakness, typical with MG
  - Specific muscle involvement
  - Timing of medications or other medical treatment
- Involves oral or pharyngeal phases of swallowing
  (Colton-Hudson et al., 2002)
- Aspiration (including silent aspiration) is common
  (Colton-Hudson et al., 2002; Kluin et al., 1996; Higo et al., 2005)

Example: Amyotrophic Lateral Sclerosis (ALS)

- **Etiology**
- **Pattern of progression**
- **Pattern of muscle weakness**
- **Other signs / symptoms**
- **Cognitive sequelae**
- **Medical intervention**
- **Dysarthria or other speech symptoms**
- **Respiration**
Dysphagia in ALS

- Dysphagia will eventually occur for most with this diagnosis: primarily oral, pharyngeal, and upper esophageal stages
  - Silent aspiration may occur
  - Weakness and reduced coordination
  - Mild initially, but progresses rapidly
  - Coughing/choking not associated with pneumonia (Hadjikoutis et al., 2000)
  - Delayed initiation for voluntary/dry swallows (Ertekin et al., 2000)
  - Saliva management impairments
  - Vocal fold impairment/laryngospasm for some (van der Graff et al., 2009)
- Malnutrition may lead to further progression of muscle weakness, immunosuppression (Heffernan et al. 2004; Worwood & Leigh, 1998)
- Respiratory impairments contribute to risk for aspiration, and lead to respiratory failure & death.
  - Reduced cough effectiveness

Example: Parkinson’s Disease

Etiology

Pattern of progression

Pattern of muscle weakness

Other signs / symptoms

Cognitive sequelae

Medical intervention

Dysarthria or other speech symptoms

Respiration

Dysphagia in Parkinson’s Disease

(Rosenbek & Jones, 2009; Yorkston et al., 2012)

- Oral phase
  - Tremor & slow initiation & speed
  - Piecemeal deglutition & tongue pumping
  - Buccal retention (aka “Pocketing”) & drooling

- Pharyngeal phase
  - Vallecular / Pyriform retention (or stasis)
  - Impaired laryngeal elevation & closure
  - Penetration / aspiration
  - Upper esophageal sphincter (UES) dysfunction

- Esophageal phase
  - Stasis
  - Abnormal contractions

- Respiratory – Swallowing incoordination
Summary

- Aging leads to higher risk for dysphagia
  - Anatomy / physiology changes associated with aging, i.e., “presbyphagia”
  - Disease prevalence increases with age

- For individuals with neurodegenerative diseases, swallowing will be impacted by...
  - “presbyphagia,” and
  - Specific patterns of degeneration associated with the specific disease

Intervention

Evidence Based Practice (EBP)

EBP is the “conscientious, explicit and judicious use of current best evidence in making decisions about the care of individual patients” (Sackett et al., 1996)
EBP: Research Challenges

Johnston et al., 2006

- Research is frequently incomplete or unavailable
- Conflicting results may be published
- Bias in published research
- Statistical significance ≠ clinical significance
- Pertains to populations (not individuals)
- Higher level RCT designs challenging
  - Multi-dimensional outcomes for rehabilitation studies
  - Ethical considerations in dysphagia research

The Art & Science of Intervention

American Thoracic Society Documents

An Official Multi-Society Statement: The Role of Clinical Research Results in the Practice of Critical Care Medicine

- Medical knowledge critical to clinical decision making:
  - Clinical research
  - Pathophysiologic reasoning
  - Clinical experience
- Other important considerations:
  - Patient and/or family preferences
  - System features or limits, e.g., coverage or resources

Intervention: Different Techniques

- **Compensation:** Interventions which offer immediate, transient, counteractive effects of the neurodegenerative condition on swallowing. E.g.,
  - Postural adjustments
  - Diet modification
  - Altered methods for eating and/or drinking
- **Rehabilitation:** Interventions that seek to improve or restore swallowing function. E.g.,
  - Skilled movements
  - Strength
  - Biomechanical support
- **Prevention & Maintenance:** Interventions that seek to maintain or preserve the ability to swallow and/or to prevent or minimize risk for aspiration pneumonia
  - In context of degenerative conditions “as long as possible.”
Principles of Intervention: Compensation
Rosenbek & Jones, 2009

1. There may be overlap between compensation and other techniques, i.e., rehabilitation, maintenance.
2. Compensatory strategies do not guarantee prevention of aspiration and its consequences.
3. Combinations of compensatory, rehabilitative (&/or maintenance) techniques are likely to have the best outcome.
4. It is best to consider and discuss duration of intervention with the patient.
5. Patients may have poor compliance.
6. Patients may decline compensatory strategies.

Principles of Intervention: Rehabilitation
Rosenbek & Jones, 2009

Rehabilitative techniques – 2 types of therapeutic targets:
1. Change the underlying physiology
   - Weakness
   - Reduced endurance
2. Increase skill

Plasticity – 3 types:
1. Muscle
2. Behavioral
3. Neural

Principles of Muscle Plasticity
Burkhead, Sapienti & Rosenbek, 2007
Rosenbek & Jones, 2009

• Intensity
  - Amount of resistance
  - Amount / Duration of training

• Specificity
• Transference
• Maintenance Plan
• Optimal timing – “Staging” in context of neuro-degenerative disease
Principles of Behavioral Plasticity

Rosenbek & Jones, 2009

• Practice schedule
  – Number of repetitions
  – Spacing of repetitions: mass vs. distributed

• Type of feedback
  – Knowledge of results
  – Knowledge of performance

• Timing of feedback

Principles of Neural Plasticity

Kleim & Jones, 2008; Robbins et al., 2008

• Use it or lose it
• Use it and improve it
• Transference
• Interference

• Factors that may influence neuroplasticity:
  – Specificity
  – Repetition
  – Intensity
  – Timing
  – Salience
  – Age

Influence of Underlying Neuromuscular Status and Medical Diagnosis on Treatment

• Diagnosis and prognosis
• Progression pattern
• Stage of disease progression
• Involvement of multiple systems, e.g.,
  – Breathing
  – Ambulation, ADLs, etc.
• Involvement of multiple healthcare disciplines
**Stage of Disease Progression:**

Disease Progression Scales

- ALS Functional Rating Scale – Revised (ALSFRS–R) (Cedarbaum et al., 1999)
  - Bulbar
  - Fine Motor
  - Gross Motor
  - Respiratory

- Expanded Disability Status Scale (EDSS) (Kurtzke, 1983); incorporates ratings of functional systems:
  - Pyramidal
  - Bowel & bladder
  - Brainstem
  - Cerebral or mental
  - Sensory
  - Cerebellar
  - Visual
  - Other

**Multi-Disciplinary Collaboration**

Team communication is needed for optimal care

- Medical
  - Neurology
  - Rehabilitation
  - Pulmonary
  - Otolaryngology
  - GI
- PT, OT, Speech
- Nutrition
- Palliative care!
- Dental

**International Classification of Functioning, Disability and Health (ICF 2001)**
Outcome: the Result of Intervention

- Impairment level
- Functional
- Activity
- Participation
- Patient reported
- Quality of life (QOL)
- Health related QOL

EXAMPLES:
- ASHA NOMS
- Functional Oral Intake Scale (FOIS) (Crary et al., 2005)
- SWAL-QOL (McHorney et al., 2002)
- EAT-10 (Belafsky et al., 2008)

Treatment Goals: Considerations

- Neurodegenerative disorders – heterogeneous
  - Prognosis, progression pattern, severity/stage
  - Systems involved
- Challenges in judging the success of intervention
- Suggest treatment goals include:
  - “Minimizing” or “lowering risk”
  - Patient/family/caregiver education
- Periodic re-assessment

Compensatory Strategies

- **DIET Modification**
  - Food texture
  - Liquid viscosity
  - Medications
  - Sensory enhancements

- **OTHER**
  - Repeated swallows
  - Slower speed
  - Energy conservation
  - Oral care

- **Positioning**
  - Upright position
  - Remain upright for awhile after eating
  - Slight neck flexion
  - Straw/short cup
  - Head rotation
  - Chin tuck

- **AMOUNTS**
  - Small sips & bites
  - One sip/bite at a time
Management Techniques to Clear Thick Mucous

- Maintain adequate hydration
- Avoid or minimize caffeine / alcohol
- Avoid or minimize milk products
- Papaya enzyme
- Club soda
- Humidifier
- Good oral care – toothbrushing; oral rinsing
- Mouth moisture (swabs/creams)
- Adjust positioning to aid swallowing
- Medications
  - Side effects: drying
  - Meds to thin out secretions, e.g., OTC Mucinex

Common Interventions for Excessive Thin Saliva

- Medications
  - Scopolamine patch
  - Tricyclic antidepressants (dryness is a side effect)
  - Atropine drops
  - Benzhexol hydrochloride
  - Glycopyrrolate (Robinul)
  Beware that excessive medication dosing might result in too much oral pharyngeal dryness

- Training with awareness & volitional swallowing
- Exercises to promote lip seal
- Other
  - Suction machine
  - Cough assist
  - Botox to salivary glands
  - Surgical interventions

- GOAL = Balance
  - Not too gurgly & wet
  - Not too dry

Preventative Maintenance: Oral Care

- Poor oral health is associated with increased risk for pneumonia (Paju & Scannapieco, 2007)

- Oral care
  - Importance
  - Precautions to reduce risk for aspiration

IMAGES: Creative Commons
www.sageproducts.com
Swallowing Maneuvers

EXAMPLES:
• Mendelsohn Maneuver
• Supraglottic swallow
• Hard / Effortful Swallow

Exercise-Based Interventions in Neurodegenerative Conditions

• Potential benefit depends
  – What is the underlying pathophysiology?
  – What is the disease stage? Earlier = better

• Three examples:
  1. Shaker exercise
  2. Lingual strengthening
  3. Respiratory muscle strength training

Shaker Exercise

IMAGE: Creative Commons
Ferdjallah et al., 2000
Inspiratory muscle training (IMT)
Expiratory muscle strength training (EMST)

• 2 types of trainers:
  1. Resistance trainers
  2. Threshold trainers

• Several products, including:
  – Expiratory Muscle Strength Trainer 150 (Aspire)
  – Ultrabreathe
  – Pflex (Respironics)
  – Powerbreathe

Benefits of Respiratory Muscle Strength Training

• Aslan et al., 2013: Increased respiratory muscle strength in individuals with slowly progressive forms of muscular dystrophy & slower decline in spirometry PFT measures
• Pitts et al., 2008; Troche et al., 2010; Sapienza et al., 2011: Increased swallowing safety & cough effectiveness in PD with EMST
• Chiara et al., 2006: Increased MEP & peak cough flow in individuals with MS and moderate disability with EMST
Respiratory Aids

- Manually assisted cough
- Mechanical In-Exsufflation (MIE)
- Lung volume recruitment (LVR)
- Belly binder
- Non-invasive ventilation

Manually Assisted Cough

- Improves expiratory cough flows
  Kirby NA, Arch Phys Med Rehabil 1966;47:705-10
- Abdominal splint prevents paradoxical motion
- Requires skilled caregiver and coordinated effort
- Inspiratory volume dependent

Mechanical In-Exsufflation (MIE)

- Non-invasive
- Non-traumatic to soft tissue
- Can clear secretions distal to the large airways
Myasthenia Gravis (MG)

- **Treatment goals:**
  - Return of functional swallowing
    - Medical treatment for MG
      - Coordination of meds with meal times
    - Monitoring function (& tracking severity of MG)
      - MG Foundation of America (MGFA) classification system
    - Avoid known triggers that may lead to exacerbation
      - Exercise intolerance
  - Management of dysphagia during exacerbations
    - Education
    - Management & Compensatory strategies
      - Energy conservation
      - Other compensatory strategies

Amyotrophic Lateral Sclerosis (ALS)

- **Treatment goals related to dysphagia may include:**
  - Maintain ability to eat for as long as possible
  - Maintain adequate nutrition
  - Reduce risk for aspiration pneumonia
  - Maintain ability to effectively cough (for as long as possible)
  - Maintain ability to manage saliva and secretions

- **Intervention will vary depending on**
  - Stage of disease progression
  - Current presentation (i.e., bodily involvement)
• Medications
  – Riluzole slows disease progression
• Nutrition
  – PEG prolongs survival time
  – Mal-nutrition contributes to progression (Braun et al., 2012)
• Respiratory
  – Non-invasive ventilation (NIV) prolongs survival, slows disease progression and improves QOL
  – Mechanical In-exsufflation (MIE) aids secretion clearance.
• Multi-disciplinary care in ALS Clinics
  – Associated with better symptom management, prolonged survival and improved QOL

ALS: Potential Benefits of Exercise

• Historically, exercise has been discouraged.
• Moderate exercise beneficial in ALS mouse model (e.g., Deforges et al., 2009)
• Limb studies – 2 small studies showed small transient gains (Bello-Haas et al., 2007, Drory et al., 2001).
  – Cochrane review 2013 concluded evidence is inadequate (Dal Bellow-Haas et al., 2013)
  – Inspiratory muscle training (IMT)
    • Small gains, but no significant differences (Pinto et al., 2012; Cheah et al., 2009)

ALS Intervention (cont.)

• Compensatory strategies
  – Positional adjustments (Solazzo et al., 2011)
  – Dietary changes
  – Repeated swallows
• Prophylactic maintenance
  – Oral care!
• Saliva management
• Respiratory intervention
  – Non-invasive ventilation
  – MIE & other assisted cough strategies
  – LVI
Multiple Sclerosis (MS)

**Cause:** unknown; involves immune system dysfunction

**Pattern of progression:** depends on type
  - Relapsing Remitting
  - Primary or Secondary Progressive

**Pattern of muscle weakness:** variable depending in part on site of lesions

**Other signs / symptoms:** visual, cognitive, autonomic impairments

**Medical intervention:** med to treat exacerbations, manage symptoms and reduce relapses.

**Speech:** may include spastic-ataxic dysarthria and/or word finding impairments.

**Swallowing:** Variable; may affect any phase

**Respiration:** Impairments, especially in primary and secondary progressive forms

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Multiple Sclerosis (MS)

- Medical management of MS symptoms
- Compensatory strategies effective for mild-mod dysphagia (Calcagno et al., 2002)
  - Energy conservation
  - Posture & diet modification
- **Exercise**
  - Moderate limb exercise beneficial (Dalgas et al., 2010)
  - IMT & EMST beneficial (Fry & Chiara, 2010; Reyes, 2013)
  - NMES: Pilot studies (Bogaardt et al., 2009; Restivo et al., 2013)
- **Impact of cognitive impairments**

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**Directions for Future Research**

- Staging of rehabilitative intervention in the context of degenerative diseases
- Application of Principles of Intervention to individuals with neurodegenerative diseases
- Benefit of compensatory strategies and rehabilitative techniques in individuals with neurodegenerative disorders