Session 17
Adult Progressive Neurological Disorders
1:30 – 3:00 pm
Saturday, Oct. 12, 2013

Authors:
Ciucci, Focht,
Plowman, Stickler
Parkinson disease, swallowing and best practices

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Overview

• Description of Parkinson disease
  – Dispelling myths
  – Describing new ways of thinking
• Describing the impact of PD on swallowing
• Concepts regarding evaluation and treatment
• Principles of Exercise and Intervention that can benefit individuals with PD

Parkinson Disease

- Bradykinesia
- Postural Instability
- Tremor
- Rrigidity
- Gait Disturbances
- Hypokinesia
- Cognitive Deficits
- Depression
- Sensory Impairment
- Dysarthria
- Dysphagia
Other systems affected by PD

- Cognition
  - Memory
  - Executive Function
  - Reasoning
- Mood/Affect
  - Depression
- Sleep
- Autonomic
  - GI
  - HR

Parkinson disease has many phenotypes

- Really described as ‘Parkinsonism’
  - Genetic causes
  - Idiopathic
  - Trauma-induced
  - Drug-induced
  - Young-onset
  - Tremor-predominant vs. Akinetic (freezing predominant)
- What does this mean?
  - Variability in presentation


Old way of thinking....

- PD is a motor only issue
  - Only neurotransmitter affected is dopamine
  - Only brain regions affected are the basal ganglia
- Doesn’t affect swallowing until later stages

New way of thinking

- PD affects entire brain
- PD affects other neurotransmitters
- PD is a problem of sensorimotor control
  - Scaling
  - Magnitude of Movement
  - Sensory feedback
- Dysphagia can occur at ANY stage

New way of thinking....

- Primary Component of Sensorimotor Control
  - Goal Directed Movement
  - Internally generated movement
  - Postural Control Adjustment
  - Skilled Movement
  - Adjust Movement to the environment
    - Bolus
Sensorimotor Control of Deglutition

Mosier & Bereznaya, 2001

Basal Ganglia & Deglutition

- Select & Initiate Motor Plans
- Sequence, Force & Timing
- Adapt to bolus changes
- Adapt general motor plans

How does this cause dysphagia?

- Decreased force
- Decreased range of motion
- Slowness
- Delay onset
- Inability to adapt to changes in bolus
  - Volume
  - Consistency
Dysphagia & Parkinson Disease

Oral Stage
- delayed oral transit
- tongue pumping
- uncontrolled bolus
- premature loss of liquid
- piecemeal deglutition
- tongue residue
- anterior/lateral sulci residue

Pharyngeal Stage:
- impaired motility
- delayed laryngeal elevation
- vallecular residue
- pyriform sinus residue
- laryngeal penetration
- aspiration
- deficient epiglottic position and ROM

Oropharyngeal Swallowing
- Difficulty with bolus propulsion and clearance resulting in
  - Residue
  - Airway compromise
  - Inefficient deglutition and inability to meet nutrition and hydration needs

Miller et al., 2006; Potulski et al., 2003; Ali et al., 1996; Bird et al., 1994; Troche, Sapienza, & Rosenbek, 2008; El Sharkawi et al., 2002; Leopold & Kapel, 1997; Fuh et al., 1997; Nagaya, Kachi, Yamada, & Igata, 1998; Potulski et al., 2003; Robbins et al., 1986; Wintzen et al., 1994
Esophageal Stage

- Weak esophageal peristalsis with food remaining in the esophagus
- Esophageal spasm
- Hiatal hernia
- Higher incidence of gastroesophageal reflux

Eadie & Tyrer, 1965; Leopold & Kupel, 1997

Morbidity and Mortality

- Weight loss
- Changes in diet
- Decreased quality of life
- Aspiration pneumonia
  - Leading cause of death related to PD

Beyer, Nerlofson, Amland, & Larsen, 2001; Clarke, 2000; D’Amelio et al., 2006

This is also a quality of life issue

- Social activities
  - Dining with friends, family and work colleagues can be negatively impacted.
  - Reluctance to eat in public due to embarrassment about drooling, slowness of eating, or fear of choking (Rosenbek & Jones, 2009).
- Patients may also have difficulty with reach-to-eat movements which can negatively impact feeding (Doan, Melvin, Whishaw, & Suchowensky, 2008).

Evaluation: Does this person have dysphagia?

• The answer is probably ‘yes’ to some degree
• Swallowing impairments do not necessarily correlate well with general ‘disease severity’
• Under-noticed by patient
  – Targeted probes may help the patient identify issues with more accuracy
• Under-noticed by practitioner
  • May not ask
  • UPDRS questions (put in examples next slide)

Variability

• Medications
• Fatigue
• Deep brain stimulation
• Disease progression

Evaluation

• Thorough history, medical history, chart review, structured interview
  – Ask the patient
  – Ask the S.O.
• Keep in mind that there are significant fluctuations in function
• Get an imaging study immediately, even if there are no significant signs of dysphagia
  – This will be your baseline
  – There may be ‘preclinical’ signs and YOU may be the first to document this
Treatment for Parkinson Disease

- Pharmacological
  - Dopamine
  - Other Neurotransmitters
- Surgical
  - Ablation
  - Transplantation
  - Deep Brain Stimulation
- Behavioral
  - Exercise

Effects of Medications on Swallowing

- Review medications
  - L-dopa, COMT or MAO inhibitor, Dopamine Agonist, other medications
  - Acknowledge side effects
  - Evaluate on and off of therapies
- No evidence for improvement, may actually diminish function

Effects of DBS on swallowing

- No improvement in the oral stage of swallowing
- Improved pharyngeal transit time, reduced pharyngeal residue and aspiration in the stimulated vs. non-stimulated condition.
- Likely, movements that require more precise/fine motor control may not be improved but the overall transfer of the bolus through the pharynx and airway protection may be improved with STN DBS.
- Impact of DBS on speech and swallowing likely depends on
  - Location of the implanted electrodes
  - Stimulation parameters


Michelle Ciucci, PhD
Adult Progressive Neurological Disorders

What can we do?

- Right now, all roads lead to.....

LSVT/LOUD

- Improved tongue base movement in oral and pharyngeal stages
- Improved OPSE

Respiration and Swallowing

- Weakness in voluntary cough is related to penetration/aspiration
  - Pitts et al., 2008 (Chest)

- Expiratory muscle strength training led to decrease in penetration/aspiration
  - Pitts, et al., 2008 (Dysphagia)
Other treatment options

Compensatory Strategies

- "Chin down" led to less aspiration
  - Logemann, et al., 2008
  - Ashford et al., 2009
- Supraglottic swallow was ineffective
  - Nagaya et al., 1998
- No evidence for effortful swallow or Mendelssohn maneuver
  - But, could we use this as a swallow-specific therapy?

Bolus Modifications

- Bigger vs. Smaller
  - Role of sensation?
- Thickened liquids = less aspiration
  - Troche, et al., 2008
  - Logemann, et al., 2008
  - Caveat: thickened liquids are association with increased pulmonary complications
Oral hygiene

• Muscle rigidity, tremor, and hypokinesia can all negatively impact the ability of PD patients to perform adequate oral care and hygiene, and PD patients have increased incidence of periodontal disease

Dysphagia Therapy and PD

• Begin to address early
• Exercise
  — LSVT/Loud
  — EMST
• Design our own treatment paradigm based on patient needs
  — Postural Changes
  — Diet Modifications
  — Frequent follow-up
  — Observe over time
  — Evaluate patients in ON and OFF states
    • DBS
    • Medications

Principles of Exercise

• Intensity
  — Intensive practice is important for maximal plasticity
  — frequency
  — repetitions
  — force/resistance
  — effort
  — accuracy

• Complexity
  — Complex movements/ environmental enrichment promote greater structural plasticity

• Salience
  — Rewarding tasks activate basal ganglia circuitry
• Use it or lose it
  • Inactivity may accelerate deficits
  • Continuous activity may slow disease progression

• Timing
  • Injury creates fertile field for plasticity


Phane et al., 2005 Plautz et al., 2000

Specifics

• Identify particular areas of difficulty
• Address that area with exercise
  – Simple cue
  – Focus on effort
  – How does that feel?
  – Intensive practice
  – Salient practice
  – Determine amount of cueing needed to maintain/carryover

Concrete Examples

• Begin intervention early and continue intervention throughout the disease process
• Practice should be task-specific
  – Example: Focus on an exercise to improve laryngeal elevation, but do this during a swallow and not during another task.
• Use a single cue
  – Minimize cognitive load
  – Example: Use one phrase, such as ‘swallow hard’ instead of multiple instructions such as ‘put the pudding in your mouth, hold it, push your tongue back forcefully, really lift your larynx and squeeze your throat, etc. The phrase you choose should elicit the behavior easily.
• Make the task highly salient and rewarding
  – Luckily, for most people, food is both salient and rewarding. Choose items that the patient wishes to eat and drink if they are safe.

• High intensity and multiple repetitions
  – Multiple repetitions, multiple times per day, for a period of weeks

• Include a sensory recalibration component
  – Example: Identify the appropriate behavior, such as the amount of tongue force to clear a bolus, and instruct the patient to ‘feel’ the amount of effort that it took to elicit that behavior. Focus on that amount of effort and although it may seem exaggerated, the patient must be trained to use that amount of effort until it becomes habitual.

Thank you!
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Multiple Sclerosis (MS)

- Overview of MS
- Epidemiology
- Pathophysiology
- Symptoms
- Swallowing impairment in MS
  - Impact of MS on swallowing function
  - Assessment of swallowing
  - Dysphagia management for patients with MS
    - Medical/surgical/pharmacological
    - Behavioral

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Neurodegenerative disease
“Many scars”
Early disease onset and progressive course
Symptom onset: 15 – 45 years
Average age at diagnosis: 29 years in females; 31 years in men
Median survival time: 40 years from diagnosis
More common in females (1.4 – 3.1 times)
More common in Caucasians
Prevalence: 58 – 95 per 100,000

Overview of MS

Incidence: 400,000
Risk of development: 1 in 750
Annual (estimated) healthcare costs: $10 billion
Annual direct and indirect costs: $62,000/person
Lifetime cost: $1 – 2 million

Overview of MS

Pathophysiology

Autoimmune disease
Dissemination of CNS inflammatory lesions
Destruction of myelin
Alters neural impulse transmission

**Pathophysiology**

- Formation of plaques (demyelinating lesions)
- Recognition of myelin basic protein and proteolipid as foreign?

**Types of MS**

- Relapsing-Remitting (RRMS)
- Progressive-Relapsing (PRMS)
- Primary progressive (PPMS)
- Secondary Progressive (SPMS)
- Benign MS

**Actual continuum of disease??**

**Common Symptoms**

- Depends on location of lesion(s)
- Cognitive dysfunction
- Pain
- Psychiatric disturbance
- Sensory impairment
- Visual disturbance
- Uroguential dysfunction
- Vertigo
- Reduced dexterity
- Muscle weakness
- Dysarthria
- Spasticity
- Dysphagia
- Ataxia and tremor
- Seizures

Smeltzer et al. (1992); Thompson, 2001; Zivadinov & Bakshi (2004)
**Fatigue**

**Multifactorial?**

**Primary**
- Inflammatory cytokines; endocrine changes; axonal loss...

**Secondary**
- Comorbid conditions, including psychiatric conditions; side effects of medications; sleep disorders...

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**Respiratory insufficiency**

- Expiratory muscle weakness
- Atelectasis
- Pneumonia

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**Impact of MS on Swallowing**

- Prevalence: 30 – 43%
- Underlying pathophysiology is variable
- Prevalence increases (65%) as disability advances
- Brainstem/cerebellar dysfunction
- ~2/3 of most disabled patients (EDSS >7.5)
- (No evidence for MS type)
- Cognitive dysfunction can disrupt swallowing

**~2% receive referral for dysphagia treatment**

- De Pauw et al. (2002); de Sa et al. (2011); Poorjavad et al. (2010); Rice & Wilkins (2013)
Impact of MS on Swallowing

- Predominantly pharyngeal dysfunction (~30%)
- Life-threatening
  - Aspiration
    - Risk increases with severe brainstem/cerebellar dysfunction and advanced disability
    - Aspiration pneumonia due to dysphagia
      - Leading cause of death
  - Malnutrition
- QUALITY-OF-LIFE!

Assessment of Swallowing

- Chart review
  - Medical/surgical/social history
  - Medications
  - Imaging
  - Structured interview

Assessment of Swallowing

- Clinical (bedside) examination
  - Neurologic (cranial nerves, CN)
    - CN V, VII, IX, X, XI, and XII
      - Sensation, range of motion, speed, symmetry, accuracy, and strength (against resistance)
  - Motor speech
    - Respiration, phonation, articulation, resonance, prosody, intelligibility
  - Reflexes
  - PO trials
Assessment of Swallowing

× Instrumental examination
× MBSS, FEES

Assessment of Swallowing

× Ask patient (caregiver) about swallowing function
× Coughing/choking
× Avoidance of food once enjoyed or modified diet
× Unintentional weight loss
× Food sticking in throat
× Previous episodes of pneumonia
× Change in speech or voice
× Difficulty managing secretions, etc.

**Must consider presence (and severity) of cognitive dysfunction**

De Pauw et al. (2002); Rice & Wilkins (2013)

Assessment of Swallowing

× Potential findings during clinical examination
× Coughing
× Positive jaw jerk reflex
× Weak cough
× Residue
× Weak tongue
× Slow movements
× Dry mouth
× Excessive saliva
× Mastication difficulty
× Vocal quality
× Labored breathing

**Must consider presence (and severity) of cognitive dysfunction**

Rice & Wilkins (2013); Thomas & Wiles (1999)
### Assessment of Swallowing
- Get instrumental evaluation of swallowing
- Baseline for comparison as disease progresses
- sEMG? Or EMG?

**Words to remember:**
- Even during periods of remittance (no acute attacks), patients with MS can have ongoing (chronic) swallowing impairments!

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### Pharmacological Management
- **Botulinum toxin A injection of cricopharyngeus**
  - Used to treat hypertonicity of CP
- Identify patients for CP myotomy
- Identify patients for vagal nerve stimulation??

**Pharmacological Management**
- Open-label pilot study (n = 25)
  - All aspirators pre-treatment (PAS > 6)
  - Pharyngeal residue and incomplete CP opening
  - Injection in each side of CP muscle
  - All had PAS scores (< 2) post-treatment
  - Mean effect duration of ~14 weeks
  - Repeat injections every 3 – 4 months

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### Pharmacological Management
- **Vagal nerve stimulation (VNS) study (n = 3)**
  - Hypothesized down-regulation of inferior olive-cerebellar circuit reduces cerebellar tremor
  - Swallowing and inferior-olive cerebellar circuit are both modulated by the nucleus tractus solitarius
  - Postural cerebellar tremor (PCT) + dysphagia
  - Timed test of swallowing: 50-mL of water
  - PCT and dysphagia improved post-VNS activation
Pharmacological Management

- Anticholinesterases (Anti-AChE)?
  - Prevents destruction of acetylcholine (Ach) by the enzyme acetylcholinesterase (AChE)
  - Transmits nerve impulses within the parasympathetic nervous system

**Limited evidence of efficacy**

Behavioral Management

- Reported in literature
  - Chin tuck
  - Thickened-liquids
    - Contraindicated in advanced disease?
  - Slow rate and small bites/sips
  - Alternating solids and liquids
  - Feeding tube

**Must consider presence (and severity) of cognitive dysfunction**

de Sa et al. (2011); Rice & Wilkins (2013); Zivadinov & Bakshi (2004)

Behavioral Management

- Thermal tactile stimulation of anterior faucial pillars
  - No evidence to support efficacy
  - Active (strength training) therapeutic program
  - Methodological flaws
  - May exacerbate symptoms
  - Avoid in patients with systematic fatigue

**Must consider presence (and severity) of cognitive dysfunction**

Festivo et al. (2013); Rice & Wilkins (2013); Zivadinov & Bakshi (2004)
Behavioral Management

Electrical stimulation pilot study (n = 25)
- 2 sessions/week x 3 weeks
- 30 Hz, 200 μsec

Post-treatment (n = 17)
- Less pooling of saliva in pyriform sinuses (n = 6)
- Less aspiration of thin liquids (n = 9)
- All patients reported improved swallowing

Behavioral Management

Electrical stimulation pilot study (n = 20)
- Low-frequency (5 Hz) intraluminal pharyngeal electrical stimulation
- Patients randomized to “real” or “sham” stimulation
- Stimulation for 10 minutes for 5 days
- Significant decrease in PAS scores
- Stimulus-induced short-term cortical plasticity?

Behavioral Management

Swallowing maneuvers?
- Mendelsohn?
- Shaker?
- Inspiratory and expiratory muscle (strength) training?

Restivo et al. (2013)

Bogaardt et al. (2013)

Rice & Wilkins (2013); Zivadinov & Bakshi (2004)
Myasthenia Gravis (MG)

Myasthenia Gravis
- Overview of MG
- Epidemiology
- Pathophysiology
- Symptoms
- Swallowing impairment in MG
- Impact of MG on swallowing function
- Assessment of swallowing
- Dysphagia management for patients with MG
  - Medical/surgical/pharmacological
  - Behavioral
### Overview of MG

- Neurodegenerative disorder
- “Grave muscular weakness”
- Symptom onset: >50 years
  - Symptom onset earlier in females
- More common in females
- Prevalence: 14 – 20 per 100,000
- Incidence: 36,000 – 60,000 cases
- Bimodal distribution
- Risk of development: 1 in 5,000

http://www.myasthenia.org/; Rosenbek & Troche (2013); Schwartz et al. (2005)

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### Overview of MG

- Estimated annual health plan paid costs for one person with MG: $15,675
- Annual MB-related pharmacy costs: $9.4 million
  - IVIg accounts for 85% of pharmacy costs

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### MG Pathophysiology

- Autoimmune disease that attacks neuromuscular junction
- MG antibodies block acetylcholine receptors
  - Fatigable weakness of skeletal muscles
- Fatigable weakness increases with activity and improves with rest

## Impact of MG on Swallowing

- Can affect any “phase” of swallowing
- Presenting symptom in 6 – 24% of patients
- Predominantly pharyngeal dysfunction
- Prevalence: 15 – 40% with generalized form
- Uncommon to be sole manifestation
- MuSK-positive MG patients tend to have more pronounced bulbar involvement
  - E.g., Lingual atrophy

**QUALITY-OF-LIFE!!**

Juan, Tou, Lo, & Wo (2010); Schwartz, Waclawik, Ringwala, & Robbins (2005)

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## Impact of MG on Swallowing

- Common symptoms reported or observed
  - Coughing/choking
  - Globus sensation
  - Regurgitation of bolus
  - Excessive saliva
  - Gagging
  - Residue
  - Avoidance of food once enjoyed
  - Weight loss
  - Aspiration pneumonia
  - Difficulty chewing
  - Weakness

**Patients may present with flaccid dysarthria, with severity increasing during prolonged speaking**

Juan, Tou, Lo, & Wo (2010); Schwartz, Waclawik, Ringwala, & Robbins (2005)

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## Impact of MG on Swallowing

- Fatigability
  - Better at beginning of meal
  - Inability to masticate or swallow
- Aspiration can trigger *myasthenic crisis*
- Poorer prognosis
- Medication side effects
  - Malnutrition
  - Dehydration

Juan, Tou, Lo, & Wo (2010); Schwartz, Waclawik, Ringwala, & Robbins (2005)
Impact of MG on Swallowing

- Response to treatment
- Symptoms may improve during treatment
- Symptoms may re-appear after completion
- Dysphagia may not improve despite an otherwise improved clinical course
- Have caregivers take a first aid course

Kuin, Bromberg, Feldman, & Simmons (1996)

Assessment of Swallowing

- Clinical (bedside) examination
- Neurologic (cranial nerves, CN)
  - CN V, VII, IX, X, XI, and XII
  - Sensation, range of motion, speed, symmetry, accuracy, and strength (against resistance)
- Motor speech
  - Respiration, phonation, articulation, resonance, prosody, and intelligibility
- Reflexes
- PO trials

Images: Kuki (2009)

Assessment of Swallowing

- Instrumental examination
  - MBSS, FEES

**Tensilon test assists in diagnosis of bulbar MG when combined with instrumental examination of swallowing function**

Kuin et al. (1996); Schwartz et al. (2005); Warnecke et al. (2008)
Assessment of Swallowing

- Assess and manage swallowing function and impairment during a myasthenic exacerbation
- Follow, assess, and treat during periods of improvement
- Minimal evaluation every 3 months
  - Manage potential risk of aspiration
  - Avoid pulmonary complications

Assessment of Swallowing

- Potential findings during clinical examination
  - Delayed laryngeal elevation
  - Delayed epiglottic inversion
  - Anterior spillage
  - Difficulty forming cohesive bolus
  - Difficulty with mastication
  - Piecemeal deglutition
  - Vallecular residue
  - Residue in pyriform sinuses
  - Penetration/Aspiration
  - Fatigability
  - Unable to hold jaw closed

Medical Management

- Anticholinesterase drugs
- Increased doses of cholinergic agents, immunomodulatory therapies, initiation of plasma exchange
- Reversal of lingual atrophy (case report)
- Prophylactic placement of feeding tube??

Kluin, Bromberg, Feldman, & Simmons (1996); Rosenbek & Troch (2013)
### Behavioral Management

- Reported in literature
- Postural techniques
  - E.g., head turn, chin tuck
- Diet modifications
- Slow rate and **small bites/sips**
- Alternating solids and liquids
- **Smaller, more frequent meals**
- Cold foods
- **Rest** prior to eating
- Feeding tube

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### Behavioral Management

- Active exercises
  - Limited by fatigability
  - Not recommended??
  - Low impact
  - Resistance training
  - Inspiratory muscle training
    - Diaphragmatic breathing
    - Pursed lips breathing

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### Research

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References and Suggested Readings


References and Suggested Readings


- http://www.nationalmssociety.org/
- http://www.myasthenia.org/

Additional Information

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**Abbreviations**

<table>
<thead>
<tr>
<th>Abbreviation</th>
<th>Definition</th>
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<tbody>
<tr>
<td>ACh</td>
<td>Acetylcholine</td>
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<tr>
<td>AChE</td>
<td>Acetylcholinesterase</td>
</tr>
<tr>
<td>Anti-AChE</td>
<td>Antiacetylcholinesterase</td>
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<tr>
<td>CN</td>
<td>Cranial nerve</td>
</tr>
<tr>
<td>CNS</td>
<td>Central nervous system</td>
</tr>
<tr>
<td>CP</td>
<td>Cricopharyngeus</td>
</tr>
<tr>
<td>CSF</td>
<td>Cerebrospinal fluid</td>
</tr>
<tr>
<td>DIS</td>
<td>Dissemination in space</td>
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<tr>
<td>DIT</td>
<td>Dissemination in time</td>
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<td>EDSS</td>
<td>Expanded Disability Status Scale</td>
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<tr>
<td>EMG</td>
<td>Electromyography</td>
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<tr>
<td>FEES</td>
<td>Fiberoptic Endoscopic Evaluation of Swallowing</td>
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<tr>
<td>FSS</td>
<td>Functional Systems Score</td>
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<tr>
<td>Ig</td>
<td>Immunoglobulin</td>
</tr>
<tr>
<td>IVIg</td>
<td>Immunoglobulin therapy</td>
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<tr>
<td>MBSS</td>
<td>Modified barium swallow study</td>
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<td>MS</td>
<td>Multiple sclerosis</td>
</tr>
<tr>
<td>MG</td>
<td>Myasthenia gravis</td>
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<tr>
<td>MRI</td>
<td>Magnetic resonance imaging</td>
</tr>
<tr>
<td>MuSK</td>
<td>Muscle specific tyrosine kinase</td>
</tr>
<tr>
<td>PAS</td>
<td>Penetration-Aspiration Scale</td>
</tr>
<tr>
<td>PCT</td>
<td>Postural cerebellar tremo</td>
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<tr>
<td>PNS</td>
<td>Peripheral nervous system</td>
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<tr>
<td>sEMG</td>
<td>Surface electromyography</td>
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<tr>
<td>VNS</td>
<td>Vagal nerve stimulation</td>
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## 2010 McDonald Criteria for Diagnosis of MS

<table>
<thead>
<tr>
<th>Clinical Presentation</th>
<th>Additional Data Needed for MS Diagnosis</th>
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<tbody>
<tr>
<td>2 attacks; objective clinical evidence of 2 lesions or objective clinical evidence of 1 lesion with reasonable historical evidence of prior attack</td>
<td>None</td>
</tr>
<tr>
<td>2 attacks; objective clinical evidence of 1 lesion</td>
<td>Dissemination in space, demonstrated by: 1 T2 lesion in at least 2 of 4 MS-typical regions of the CNS (periventricular, juxtacortical, infratentorial, or spinal cord); or await a further clinical attack implicating a different CNS site</td>
</tr>
<tr>
<td>1 attack; objective clinical evidence of 2 lesions</td>
<td>Dissemination in time, demonstrated by: Simultaneous presence of asymptomatic gadolinium-enhancing and nonenhancing lesions at any time; or A new T2 and/or gadolinium-enhancing lesion(s) on follow-up MRI, irrespective of its timing with reference to a baseline scan; or Await a second clinical attack</td>
</tr>
</tbody>
</table>

Polman et al. (2011)

## 2010 McDonald Criteria for Diagnosis of MS

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<th>Clinical Presentation</th>
<th>Additional Data Needed for MS Diagnosis</th>
</tr>
</thead>
<tbody>
<tr>
<td>1 attack; objective clinical evidence of 1 lesion (clinically isolated syndrome)</td>
<td>Dissemination in space and time, demonstrated by: For DIS: 1 T2 lesion in at least 2 of 4 MS-typical regions of the CNS (periventricular, juxtacortical, infratentorial, or spinal cord); or await a second clinical attack implicating a different CNS site; and For DIS: Simultaneous presence of asymptomatic gadolinium-enhancing and nonenhancing lesions at any time; or A new T2 and/or gadolinium-enhancing lesion(s) on follow-up MRI, irrespective of its timing with reference to a baseline scan; or Await a second clinical attack</td>
</tr>
<tr>
<td>Insidious neurological progression suggestive of MS (PPMS)</td>
<td>1 year of disease progression (retrospectively or prospectively determined) plus 2 of 3 of the following criteria: 1. Evidence for DIS in the brain based on 1 T2 lesions in the MS-characteristic (periventricular, juxtacortical, or infratentorial) regions 2. Evidence for DIS in the spinal cord based on 2 T2 lesions 3. Positive CSF (isoelectric focusing evidence of oligoclonal bands and/or elevated IgG index)</td>
</tr>
</tbody>
</table>

Polman et al. (2011)

## 2010 McDonald MRI Criteria for Demonstration of DIS

- >1 T2 lesion in at least 2 of 4 areas of the CNS:
  - Periventricular
  - Juxtacortical
  - Infratentorial
  - Spinal cord

Polman et al. (2011)
### 2010 McDonald MRI Criteria for Demonstration of DIT

- A new T2 and/or gadolinium-enhancing lesion(s) on follow-up MRI, with reference to a baseline scan, irrespective of the timing of the baseline MRI
- Simultaneous presence of asymptomatic gadolinium-enhancing and non-enhancing lesions at any time

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### Types of MS

**Relapsing-Remitting (RRMS)**
- Most common
- 85% begin with RRMS
- Acute “attacks” followed by full or incomplete recovery

---

**Progressive-Relapsing (PRMS)**
- Occurs in nearly 5% of cases
- Progressive disability from onset but also involves acute attacks or relapse
### Types of MS

<table>
<thead>
<tr>
<th>Primary progressive (PPMS)</th>
</tr>
</thead>
<tbody>
<tr>
<td>□ Progression of disability from onset</td>
</tr>
<tr>
<td>□ 10% have PPMS</td>
</tr>
<tr>
<td>□ No acute attacks</td>
</tr>
</tbody>
</table>

Zuvich et al. (2009)

---

<table>
<thead>
<tr>
<th>Secondary Progressive (SPMS)</th>
</tr>
</thead>
<tbody>
<tr>
<td>□ Initial relapsing-remitting course followed by progression of disability</td>
</tr>
<tr>
<td>□ May include occasional relapses and minor remissions and plateaus</td>
</tr>
<tr>
<td>□ Less recovery following attacks</td>
</tr>
<tr>
<td>□ Persistently worsening functioning during and between attacks</td>
</tr>
<tr>
<td>□ Fewer and fewer attacks (or none at all) accompanied by progressive disability</td>
</tr>
</tbody>
</table>

Zuvich et al. (2009)

---

<table>
<thead>
<tr>
<th>Secondary Progressive (SPMS)</th>
</tr>
</thead>
<tbody>
<tr>
<td>□ Of the 85% who start with RRMS, &gt;50% will develop SPMS within 10 years (90% within 25 years)</td>
</tr>
</tbody>
</table>

Zuvich et al. (2009)
Types of MS

- Benign MS
- Small number of relapses followed by complete recovery
- Diagnosis made only when experienced little or no disability for a period of 10 – 15 years
- Possible a relapse may occur

Zuvich et al. (2009)

Distinctive Patterns of Immune-Pathology in MS

- Pattern I: Macrophage-associated demyelination
- Pattern II: Antibody/complement-associated demyelination

Close similarities to T cell-mediated and T cell and antibody-mediated autoimmune demyelination

Denic et al. (2011)

Distinctive Patterns of Immune-Pathology in MS

- Pattern III: Distal dying-back oligodendrogliopathy
- Pattern IV: Primary oligodendrocyte degeneration

Suggestive of oligodendrogliopathy in an inflammatory background

Denic et al. (2011)
Assessment of Swallowing in MS
- Reported videofluoroscopic findings in 18 patients
  - 16 patients showed delayed laryngeal elevation and delayed pharyngeal contraction
  - 3 patients with undercoating of epiglottis
  - 3 patients with penetration
  - 4 patients with aspiration
    - All reported chronic swallowing impairment
  - 10 patients had weak tongue base retraction
  - All had residue in valleculae

Weisner et al. (2002)

Assessment of Swallowing in MS
- Reported videofluoroscopic findings in 13 patients
  - All patients showed laryngeal movement dysfunction
    - Shorter duration of laryngeal excursion
    - Longer interval between airway closure and UES opening
  - 85% demonstrated impaired epiglottic movement
    - Vallecular residue
  - 85% demonstrated impaired pharyngeal contraction
    - Middle > inferior > superior constrictor
    - Residue in pyriform sinuses
  - Penetration/aspiration
  - No relationship found between severity of dysphagia and EDSS or FSS scores

Abraham & Yun (2002)

Assessment of Swallowing in MS
- Reported manofluoroscopy findings in 30 patients
  - Delayed trigger of pharyngeal swallow
  - Bolus formation dysfunction
  - Multiple swallows for single bolus
  - Residue in valleculae and pyriform sinuses
  - Reduced pharyngeal contraction (reduced amplitude)
  - Penetration/Aspiration
  - UES dysfunction (reduced manometric relaxation)
  - Greater impairment as disability advanced

De Pauw et al. (2002)
Assessment of Swallowing in MS

- Reported endoscopic findings in 143 patients
  - Soft palate dysfunction
  - Glottal dysfunction
  - Residue
  - Aspiration
  - Risk increased with severe brainstem dysfunction
  - Compensatory strategies (e.g., posture change, altering consistency, etc) were beneficial for “mild” and “moderate” swallowing impairment
  - Eliminated aspiration
  - Greater impairment as disability advanced

Index of Pulmonary Dysfunction in MS

Adapted from Smeltzer et al. (1992)

1. Patient’s rating
2. History of difficulty handling mucus/secretions
   - No
   - Yes
3. Cough
   - Normal
   - Weak
4. Examiner’s rating
5. Strength of patient’s cough when asked to cough voluntarily
   - Normal
   - Weak
   - Very weak/inaudible
6. Value reached when patient counts aloud on a single exhalation after maximum inspiratory effort
   - ≥30
   - 20-29
   - 10-19
   - <9
7. Total Score

Kurtzke Expanded Disability Status Scale (EDSS)

- 0.0 – Normal neurologic exam
- 1.0 – No disability, minimal signs on exam
- 2.0 – Minimal disability
- 3.0 – Moderate disability, fully ambulatory
- 4.0 – Fully ambulatory without aid; Up and about 12 hours a day; Able to work a full day; Relatively severe disability; Able to walk without aid 300 meters
- 5.0 – Ambulatory without aid for 200 meters; Disability impairs full day activities
Kurtzke Expanded Disability Status Scale (EDSS)

5.5 – Ambulatory for 100 meters, disability precludes full daily activities

6.0 – Intermittent or unilateral constant assistance (e.g., cane) required to walk 100 meters with or without resting

6.5 – Constant bilateral support (e.g., cane) required to walk 20 meters without resting

7.0 – Unable to walk beyond 5 meters even with aid, essentially restricted to wheelchair; Wheels self; Transfers alone; Active in wheelchair about 12 hour a day

7.5 – Unable to take more than a few step, restricted to wheelchair; May need aid to transfer: Wheels self, but may require motorized chair for full day's activities

8.0 – Essentially restricted to bed, chair, or wheelchair, but may be out of bed much of day; Retains self-care functions; Generally effective use of arms

8.5 – Essentially restricted to bed much of day; Some effective use of arms; Retains some self-care functions

9.0 – Helpless bed patient; Can communicate and eat

9.5 – Unable to communicate effectively or eat/swallow

10.0 – Death
Kurtzke Functional Systems Scores (FSS)

**Pyramidal Functions**

- **0** – Normal
- **1** – Abnormal signs without disability
- **2** – Minimal disability
- **3** – Mild to moderate paraparesis or hemiparesis (detectable weakness but most function sustained for short periods; fatigue a problem); Severe monoparesis (almost no function)
- **4** – Marked paraparesis or hemiparesis (function is difficult), moderate quadriparesis (function is decreased but can be sustained for short periods), or monoparesis
- **5** – Paraplegia, hemiplegia, or marked quadriparesis
- **6** – Quadriplegia
- **9** – (Unknown)

www.nationalmssociety.org/

**Cerebellar Functions**

- **0** – Normal
- **1** – Abnormal signs without disability
- **2** – Mild ataxia (tremor or clumsy movements easily seen, minor interference with function)
- **3** – Moderate truncal or limb ataxia (tremor or clumsy movements interfere with function in all spheres)
- **4** – Severe ataxia in all limbs (most function is very difficult)
- **5** – Unable to perform coordinated movements due to ataxia
- **9** – (Unknown)

www.nationalmssociety.org/

**Sensory Function**

- **0** – Normal
- **1** – Vibration or figure-writing decrease only in 1 or 2 limbs
- **2** – Mild decrease in touch or pain or position sense, and/or moderate decrease in vibration in 1 or 2 limbs; or vibratory (c/s figure writing) decrease alone in 3 or 4 limbs
- **3** – Moderate decrease in touch or pain or position sense, and/or essentially lost vibration in 1 or 2 limbs; or mild decrease in touch or pain and/or moderate decrease in all proprioceptive tests in 3 or 4 limbs
- **4** – Marked decrease in touch or pain or loss of proprioception alone or combined, in 1 or 2 limbs; or moderate decrease in touch or pain and/or severe decrease in all proprioceptive decrease in >2 limbs
- **5** – Loss (essentially) of sensation in 1 or 2 limbs; or moderate decrease in touch or pain and/or loss of proprioception for most of the body below the head
- **6** – Sensation essential lost below the head
- **9** – (Unknown)

www.nationalmssociety.org/
### Kurtzke Functional Systems Scores (FSS)

#### Bowel and Bladder Function
- **0** – Normal
- **1** – Mild urinary hesitance, urgency, or retention
- **2** – Moderate hesitance, urgency, retention of bowel or bladder, or rare urinary incontinence (intermittent self-catheterization, manual compression to evacuate bladder, or finger evacuation of stool)
- **3** – Frequent urinary incontinence
- **4** – In need of almost constant catheterization (and constant use of measures to evacuate stool)
- **5** – Loss of bladder function
- **6** – Loss of bowel and bladder function
- **9** – (Unknown)

#### Visual Function
- **0** – Normal
- **1** – Scotoma with visual acuity (corrected) better than 20/30
- **2** – Worse eye with scotoma with maximal visual acuity (corrected) of 20/30 – 20/59
- **3** – Worse eye with large scotoma, or moderate decrease in fields, but with maximal visual acuity (corrected) of 20/60 – 20/99
- **4** – Worse eye with marked decrease of fields and with maximal visual acuity (corrected) of 20/100 – 20/200; Grade 3 plus maximal acuity of better eye of <20/60
- **5** – Worse eye with maximal visual acuity (corrected) <20/200; Grade 4 plus maximal acuity of better eye <20/60
- **6** – Grade 5 plus maximal visual acuity of better eye of <20/60
- **9** – (Unknown)

#### Cerebral (or Mental) Functions
- **0** – Normal
- **1** – Mood alteration only (does not affect EDSS score)
- **2** – Mild decrease in mentation
- **3** – Moderate decrease in mentation
- **4** – Marked decrease in mentation (chronic brain syndrome – moderate)
- **5** – Dementia or chronic brain syndrome – severe or incompetent
- **9** – (Unknown)
2010 McDonald Criteria for Diagnosis of MS with Progression from Onset

1 year of disease progression (retrospectively or prospectively determined)

Plus 2 of the 3 following criteria:

- Evidence for DIS in the brain based on >1 T2 lesions in at least 1 area characteristic for MS (periventricular, juxtacortical, or infratentorial)
- Evidence for DIS in the spinal cord based on >2 T2 lesions
- Positive CSF (isoelectric focusing evidence of oligoclonal bands and/or elevated IgG index)

Polman et al. (2011)

2010 McDonald Criteria for Diagnosis of MS

- If the Criteria are fulfilled and there is no better explanation for the clinical presentation, the diagnosis is "MS"; if suspicious, but the Criteria are not completely met, the diagnosis is "possible MS"; if another diagnosis arises during the evaluation that better explains the clinical presentation, then the diagnosis is "not MS."
- An attack (relapse; exacerbation) is defined as patient-reported or objectively observed events typical of an acute inflammatory demyelinating event in the CNS, current or historical, with duration of at least 24 hours, in the absence of fever or infection. It should be documented by contemporaneous neurological examination, but some historical events with symptoms and evolution characteristics for MS, but for which no objective neurological findings are documented, can provide reasonable evidence of a prior demyelinating event. Reports of paroxysmal symptoms (historical or current) should, however, consist of multiple episodes occurring over not less than 24 hours. Before a definite diagnosis of MS can be made, at least 1 attack must be corroborated by findings on neurological examination, visual evoked potential response in patients reporting prior visual disturbance, or MRI consistent with demyelination in the area of the CNS implicated in the historical report of neurological symptoms.
- Clinical diagnosis based on objective clinical findings for 2 attacks is most secure. Reasonable historical evidence for 1 past attack, in the absence of documented objective neurological findings, can include historical events with symptoms and evolution characteristics for a prior inflammatory demyelinating event; at least 1 attack, however, must be supported by objective findings.
- No additional tests are required. However, it is desirable that any diagnosis of MS be made with access to imaging based on these Criteria. If imaging or other tests (for instance, CSF) are undertaken and are negative, extreme caution needs to be taken before making a diagnosis of MS, and alternative diagnoses must be considered. There must be no better explanation for the clinical presentation, and objective evidence must be present to support a diagnosis of MS.
- Gadolinium-enhancing lesions are not required; symptomatic lesions are excluded from consideration in subjects with brainstem or spinal cord syndromes.

Polman et al. (2009)

Clinical Manifestations of MG

<table>
<thead>
<tr>
<th>Early onset</th>
<th>Late onset</th>
<th>Thyroidosis</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ocular signs 78 (50)</td>
<td>84 (36)</td>
<td>17 (12)</td>
<td>111</td>
</tr>
<tr>
<td>Positive 55 (34)</td>
<td>74 (32)</td>
<td>14 (9)</td>
<td>103</td>
</tr>
<tr>
<td>Positivity 61 (38)</td>
<td>70 (33)</td>
<td>15 (10)</td>
<td>126</td>
</tr>
<tr>
<td>Bifidus 45</td>
<td>54</td>
<td>66</td>
<td>101</td>
</tr>
<tr>
<td>Chewing 5</td>
<td>5</td>
<td>15</td>
<td>21</td>
</tr>
<tr>
<td>Salivation 7</td>
<td>7</td>
<td>12</td>
<td>24</td>
</tr>
<tr>
<td>Neck stiffness 2</td>
<td>5</td>
<td>7</td>
<td>14</td>
</tr>
<tr>
<td>Combined 23</td>
<td>20</td>
<td>43</td>
<td>66</td>
</tr>
<tr>
<td>Gastrointestinal 35</td>
<td>17</td>
<td>30</td>
<td>72</td>
</tr>
<tr>
<td>Limb ataxia 15</td>
<td>3</td>
<td>18</td>
<td>36</td>
</tr>
<tr>
<td>Bladder or feces 12</td>
<td>1</td>
<td>13</td>
<td>25</td>
</tr>
<tr>
<td>Late 49</td>
<td>6</td>
<td>50</td>
<td>85</td>
</tr>
<tr>
<td>Combined 29</td>
<td>3</td>
<td>32</td>
<td>56</td>
</tr>
<tr>
<td>Generalized 6</td>
<td>5</td>
<td>11</td>
<td>22</td>
</tr>
<tr>
<td>Respiration 4</td>
<td>2</td>
<td>6</td>
<td>10</td>
</tr>
<tr>
<td>Total 815 (59)</td>
<td>209 (81)</td>
<td>130 (91)</td>
<td>1156</td>
</tr>
</tbody>
</table>

*Partial signs of MG reported by the author’s patients. In parentheses is number of the patients who remained mostly stable during the follow-up. The late group contains only one patient with a thymoma, and 53% of patients in the onset group and 36% in the late-onset group. The mode at onset was cerebellar ataxia; 39% of all patients, 33% of the acute group, and 54% of the late-onset group. The largest number of patients had cerebellar ataxia (57% of all patients, 53% of the acute group, and 57% of the late-onset group), followed by proximal weakness, 21% (12% and 26%) and sensory loss, 16% (10% and 22%). Ocular palsy was frequent and contributed to the disability. The most frequent combined signs were 6% in the onset group and 2% in the late-onset group (12% and 2% in the late onset group). Patients with thymomas had the highest incidence of bulbar signs at onset (55% vs 30% in the early onset and 36% in the late-onset groups).

Rules (2009)
Assessment of Swallowing in MG

Colton-Hudson et al. (2002)

Reported videofluoroscopy findings in 20 patients

- Poor bolus formation (n = 7)
- Prolonged mastication
- Reduced buccal tension
- Material into lateral sulci
- Slow bolus transport (n = 13)
- Piecemeal deglutition (n = 13)
- Reside on base of tongue and soft palate (n = 16)
- Poor seal of soft palate against base of tongue (n = 15)

Assessment of Swallowing in MG

Colton-Hudson et al. (2002)

Reported videofluoroscopy findings in 20 patients

- Delay in initiation of pharyngeal swallow (n = 20)
- Reduced base of tongue to posterior pharyngeal wall (n = 17)
- Reduced epiglottic inversion (n = 12)
- Vallecular residue (n = 17)
- Residue in pyriform sinuses (n = 14)
- Penetration (n = 13)
- Aspiration (n = 7)
  - Most were silent (n = 4)

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Osserman Classification

I: Pure ocular
   - Strength of all other muscles is normal
IIA: Mild generalized
   - Prominent limb involvement
   - No pulmonary involvement
IIB: Moderate generalized
   - Prominent bulbar involvement

Osserman Classification

III: Severe generalized
   - Prominent respiratory involvement
   - Progress from ocular to severe generalized symptoms in <6 months
IV: Severe generalized (chronic form)
   - Prominent respiratory involvement
   - Progress from ocular to severe generalized symptoms in >6 months

Myasthenia Gravis Foundation of America Clinical Classification

Class I
   - Any ocular muscle weakness
   - May have weakness of eye closure
   - All other muscle strength is normal

---


Jaretzki, III et al. (2000)
### Myasthenia Gravis Foundation of America Clinical Classification

**Class II**
- May have ocular muscle weakness (any severity)
- Mild weakness affecting other than ocular muscles
  - **Class IIa**
    - Predominantly affecting limb muscles, axial muscles, or both
    - May have lesser involvement of oropharyngeal muscles
  - **Class IIb**
    - Predominantly affecting oropharyngeal muscles, respiratory muscles, or both
    - May have lesser involvement of limb muscles, axial muscles, or both

**Class III**
- May have ocular muscle weakness (any severity)
- Moderate weakness affecting other than ocular muscles
  - **Class IIIa**
    - Predominantly affecting limb muscles, axial muscles, or both
    - May have lesser involvement of oropharyngeal muscles
  - **Class IIIb**
    - Predominantly affecting oropharyngeal muscles, respiratory muscles, or both
    - May have lesser involvement of limb muscles, axial muscles, or both

**Class IV**
- May have ocular muscle weakness (any severity)
- Severe weakness affecting other than ocular muscles
  - **Class IVa**
    - Predominantly affecting limb muscles, axial muscles, or both
    - May have lesser involvement of oropharyngeal muscles
  - **Class IVb**
    - Predominantly affecting oropharyngeal muscles, respiratory muscles, or both
    - May have lesser involvement of limb muscles, axial muscles, or both

*Jaretzki, III et al. (2000)*
Bulbar MG vs. Classical MG
Adapted from Schwartz, Waclawik, Ringwala, & Robbins (2005)

<table>
<thead>
<tr>
<th>Bulbar MG*</th>
<th>Classical MG</th>
</tr>
</thead>
<tbody>
<tr>
<td>Myasthenic “snarl”</td>
<td>Ptosis</td>
</tr>
<tr>
<td>Hypernasal speech</td>
<td>Proximal limb weakness</td>
</tr>
<tr>
<td>Weakness of muscles of mastication</td>
<td>Diplopia</td>
</tr>
<tr>
<td>Neck extensor weakness</td>
<td>Neck weakness</td>
</tr>
<tr>
<td>Myasthenic crisis</td>
<td>Myasthenic crisis</td>
</tr>
</tbody>
</table>

*Bulbar findings

Weakness of striated muscles, including labial, lingual, velopharyngeal, pharyngeal, and esophageal

Flowchart of the FEES-Tensilon TEST

Start

Result of FEES Tensilon test

Jaretzki, III et al. (2000)

Myasthenia Gravis Foundation of America Therapy Status

- NT – No therapy
- SPT – Status post-thymectomy
- CH – Cholinesterase inhibitors
- PR – Prednisone
- IM – Immunosuppression therapy (other than prednisone)
- PE(a) – Plasma exchange therapy, acute
  - For exacerbations or pre-operatively

Warnecke et al. (2008)
**Myasthenia Gravis Foundation of America**

**Therapy Status**
- PE(c) – Plasma exchange therapy, chronic
  - Used on regular basis
- IG(a) – IVIg therapy, acute
  - Exacerbations or pre-operatively
- IG (c) – IVIg therapy, chronic
  - Used on regular basis
- OT – Other forms of therapy

Jaretzki, III et al. (2000)

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**Myasthenia Gravis Foundation of America**

**Post-intervention Status**
- CSR – Complete Stable Remission
  - No signs/symptoms of MG for at least 1 year
  - Received no therapy for at least 1 year
  - No weakness one examination
    - Isolated weakness of eye closure is accepted
- PR – Pharmacologic Remission
  - Same criteria for CSR except patient continues to take some form of therapy for MG
  - Use of cholinesterase inhibitors are excluded from this category

Jaretzki, III et al. (2000)

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**Myasthenia Gravis Foundation of America**

**Post-intervention Status (Change in Status)**
- I – Improved
  - Substantial decrease in pre-treatment clinical manifestations or a sustained substantial reduction in MG medications
    - In prospective studies, defined as a specific decrease in QMG score
- U – Unchanged
  - No substantial change in pre-treatment clinical manifestations or reduced in MG medications
    - In prospective studies, defined in terms of a maximum change in QMG score

Jaretzki, III et al. (2000)
Myasthenia Gravis Foundation of America
Post-intervention Status (Change in Status)

- **W – Worse**
  - Substantial increase in pre-treatment clinical manifestations or substantial increase in MG medications
  - In prospective studies, defined as a specific increase in QMG score

- **E – Exacerbation**
  - Fulfilled criteria of CSR, PR or MM but subsequently developed clinical findings greater than permitted by these criteria

- **D of MG – Died of MG**
  - Died of MG, complications of MG therapy, or within 30 days after thymectomy

Jaretski, III et al. (2000)

---

### Quantitative MG Score (QMG)

<table>
<thead>
<tr>
<th>Test items Weakness (score)</th>
<th>None (0)</th>
<th>Mild (1)</th>
<th>Moderate (2)</th>
<th>Severe (4)</th>
<th>Item score (0.1, 2, or 3)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Double vision on lateral gaze right or left (and one), seconds</td>
<td>60</td>
<td>11–60</td>
<td>1–10</td>
<td>Spontaneous</td>
<td></td>
</tr>
<tr>
<td>2. Paresis (upward gaze), seconds</td>
<td>60</td>
<td>11–60</td>
<td>1–10</td>
<td>Spontaneous</td>
<td></td>
</tr>
<tr>
<td>3. Facial muscles</td>
<td>Normal lid closure</td>
<td>Complete, weak, some resistance</td>
<td>Complete, without resistance</td>
<td>Incomplete</td>
<td></td>
</tr>
<tr>
<td>4. Swallowing 4 oz./20 mL water</td>
<td>Normal</td>
<td>Minimal coughing or throat clearing Dysarthria at Δ10–19</td>
<td>Dysarthria at Δ10–29</td>
<td>Cannot swallow (but not atropined) Dysarthria at Δ9</td>
<td></td>
</tr>
<tr>
<td>5. Speech following coughing aloud</td>
<td>None at Δ50</td>
<td>None at Δ50</td>
<td>None at Δ50</td>
<td>None at Δ50</td>
<td></td>
</tr>
</tbody>
</table>

**Quantitative MG Score (QMG) (con’t)**

<table>
<thead>
<tr>
<th>Test items Weakness (score)</th>
<th>None (0)</th>
<th>Mild (1)</th>
<th>Moderate (2)</th>
<th>Severe (4)</th>
<th>Item score (0.1, 2, or 3)</th>
</tr>
</thead>
<tbody>
<tr>
<td>6. Right arm outstretched (90° sitting), seconds</td>
<td>240</td>
<td>90–239</td>
<td>10–89</td>
<td>0–9</td>
<td></td>
</tr>
<tr>
<td>7. Left arm outstretched (90° sitting), seconds</td>
<td>240</td>
<td>90–239</td>
<td>10–89</td>
<td>0–9</td>
<td></td>
</tr>
<tr>
<td>8. Vital capacity (% predicted) mouthpiece or facemask (at best of 3)</td>
<td>≥80%</td>
<td>65–79%</td>
<td>50–64%</td>
<td>&lt; 50%</td>
<td></td>
</tr>
<tr>
<td>9. Right hand grip (best of 2)</td>
<td>Male</td>
<td>45</td>
<td>15–44</td>
<td>5–14</td>
<td>0–4</td>
</tr>
<tr>
<td>9. Right hand grip (best of 2) Female</td>
<td>Female</td>
<td>30</td>
<td>10–39</td>
<td>5–9</td>
<td>0–4</td>
</tr>
</tbody>
</table>

Wolf & Barohn (2009)
### Quantitative MG Score (QMG) (con’t)

<table>
<thead>
<tr>
<th>Task</th>
<th>Score</th>
</tr>
</thead>
<tbody>
<tr>
<td>Seat balance (best of 2)</td>
<td></td>
</tr>
<tr>
<td>Male</td>
<td>≥55</td>
</tr>
<tr>
<td>Female</td>
<td>≤55</td>
</tr>
<tr>
<td>Head, lifted (45° supine), seconds</td>
<td>120</td>
</tr>
<tr>
<td>Right leg</td>
<td>100</td>
</tr>
<tr>
<td>Left leg</td>
<td>100</td>
</tr>
</tbody>
</table>

Total QMG score (range: 0–39)

**Note:**
- A score of 30 or less indicates no weakness.
- A score of 31–39 indicates mild weakness.
- A score of 40–49 indicates moderate weakness.
- A score of 50–59 indicates severe weakness.

**Source:** Wolf & Barohn (2009)

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### Myasthenic Muscle Score (MMS)

<table>
<thead>
<tr>
<th>Task</th>
<th>Score</th>
</tr>
</thead>
<tbody>
<tr>
<td>Maintain upper limbs horizontally outstretched: 1 point per 10 s</td>
<td>0–15</td>
</tr>
<tr>
<td>Maintain lower limbs above bed plane while lying on back: 1 point per 5 s</td>
<td>0–15</td>
</tr>
<tr>
<td>Raise head above bed plane while lying on back</td>
<td>10</td>
</tr>
<tr>
<td>Against resistance</td>
<td>10</td>
</tr>
<tr>
<td>Without resistance</td>
<td>5</td>
</tr>
<tr>
<td>Impossible</td>
<td>0</td>
</tr>
<tr>
<td>Sit up from lying position</td>
<td>10</td>
</tr>
<tr>
<td>Without help of hands</td>
<td>5</td>
</tr>
<tr>
<td>Impossible</td>
<td>0</td>
</tr>
<tr>
<td>Extrinsic ocular musculature</td>
<td>5</td>
</tr>
<tr>
<td>Normal</td>
<td>10</td>
</tr>
<tr>
<td>Ptosis</td>
<td>5</td>
</tr>
<tr>
<td>Double vision</td>
<td>0</td>
</tr>
</tbody>
</table>

**Note:**
- A score of 0–15 indicates no weakness.
- A score of 16–29 indicates mild weakness.
- A score of 30–39 indicates moderate weakness.
- A score of 40–49 indicates severe weakness.

**Source:** Wolf & Barohn (2009)

---

### Myasthenic Muscle Score (MMS)

<table>
<thead>
<tr>
<th>Task</th>
<th>Score</th>
</tr>
</thead>
<tbody>
<tr>
<td>Dribbling</td>
<td>10</td>
</tr>
<tr>
<td>Complete</td>
<td>10</td>
</tr>
<tr>
<td>Mild weakness</td>
<td>5</td>
</tr>
<tr>
<td>Incomplete with corneal covering</td>
<td>5</td>
</tr>
<tr>
<td>Incomplete without corneal covering</td>
<td>0</td>
</tr>
<tr>
<td>Chewing</td>
<td>5</td>
</tr>
<tr>
<td>Normal</td>
<td>10</td>
</tr>
<tr>
<td>Weak</td>
<td>5</td>
</tr>
<tr>
<td>Impossible</td>
<td>0</td>
</tr>
<tr>
<td>Swallowing</td>
<td>5</td>
</tr>
<tr>
<td>Normal</td>
<td>10</td>
</tr>
<tr>
<td>Impaired without aspiration</td>
<td>5</td>
</tr>
<tr>
<td>Impaired with aspiration</td>
<td>0</td>
</tr>
<tr>
<td>Speech</td>
<td>5</td>
</tr>
<tr>
<td>Normal</td>
<td>10</td>
</tr>
<tr>
<td>Nasal</td>
<td>5</td>
</tr>
<tr>
<td>Stopped</td>
<td>0</td>
</tr>
</tbody>
</table>

**Note:**
- A score of 0–10 indicates no weakness.
- A score of 11–20 indicates mild weakness.
- A score of 21–30 indicates moderate weakness.
- A score of 31–40 indicates severe weakness.

**Source:** Wolf & Barohn (2009)
Myasthenia Gravis (MG) Activities of Daily Living (MG-ADL) Profile

<table>
<thead>
<tr>
<th>Activity</th>
<th>Grade 0</th>
<th>Grade 1</th>
<th>Grade 2</th>
<th>Grade 3</th>
<th>Score (0%, 10%, 20%, or 40%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Walking</td>
<td>Normal</td>
<td>Fatigued</td>
<td>Tandem gait</td>
<td>Difficult to understand</td>
<td>10%</td>
</tr>
<tr>
<td>Clenching</td>
<td>Normal</td>
<td>Fatigued</td>
<td>Ambiguous</td>
<td>Ambiguous</td>
<td>20%</td>
</tr>
<tr>
<td>Dressing</td>
<td>Normal</td>
<td>Can’t dress</td>
<td>Ambiguous</td>
<td>Ambiguous</td>
<td>40%</td>
</tr>
<tr>
<td>Breathing</td>
<td>Normal</td>
<td>Fatigued</td>
<td>Ambiguous</td>
<td>Ambiguous</td>
<td>20%</td>
</tr>
<tr>
<td>Incontinence of bladder or bowel</td>
<td>None</td>
<td>None</td>
<td>None</td>
<td>None</td>
<td>0%</td>
</tr>
<tr>
<td>Incontinence of bladder or bowel</td>
<td>None</td>
<td>None</td>
<td>None</td>
<td>None</td>
<td>0%</td>
</tr>
<tr>
<td>Bed mobility</td>
<td>None</td>
<td>None</td>
<td>None</td>
<td>None</td>
<td>0%</td>
</tr>
</tbody>
</table>

Wolf & Barohn (2009)

Myasthenia Gravis (MG) vs. Guillain-Barré Syndrome (GBS) vs. Multiple Sclerosis (MS)

<table>
<thead>
<tr>
<th>Comparison</th>
<th>Myasthenia Gravis</th>
<th>Guillain-Barré Syndrome</th>
<th>Multiple Sclerosis</th>
</tr>
</thead>
<tbody>
<tr>
<td>Autoimmune</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
</tr>
<tr>
<td>Disease</td>
<td>Disease affecting neuromuscular junction (result of anti-acetylcholine antibodies)</td>
<td>Acute inflammatory demyelinating polyneuropathy affecting the PNS</td>
<td>Autoimmune demyelinating disease affecting the CNS</td>
</tr>
<tr>
<td>Incidence Peaks</td>
<td>Bimodal peaks of incidence (Women: 20 and 30 years; Males: 60 and 70 years)</td>
<td>Highest incidence between ages 50 and 75</td>
<td>Lowest incidence between ages 20 and 50</td>
</tr>
<tr>
<td>Disease Symptoms</td>
<td>Muscle weakness begins in distal extremities and ascends upwards</td>
<td>Symptoms can develop rapidly or may take up to 3 weeks, with greatest weakness within the first 3 weeks</td>
<td>Characterized by acute attacks followed by periods of remission</td>
</tr>
<tr>
<td>Fatigability</td>
<td>Fatigability weakness increases with repetitive stimulation and improves with rest</td>
<td>Symptoms develop rapidly or may take up to 3 weeks, with greatest weakness within the first 3 weeks</td>
<td>Fatigability weakness increases with repetitive stimulation and improves with rest</td>
</tr>
<tr>
<td>Flare-ups</td>
<td>�</td>
<td>�</td>
<td>�</td>
</tr>
<tr>
<td>Bulbar weakness</td>
<td>Can occur; leading to dysarthria and dysphagia</td>
<td>Crystalline nerve involvement can occur, most often affected is CN VII</td>
<td>Lesions of cerebellum and cerebellum increase risk of dysphagia</td>
</tr>
<tr>
<td>Respiratory</td>
<td>Can cause respiratory failure</td>
<td>Respiratory muscles can be affected, which can cause respiratory failure</td>
<td>Cognition can be impaired, which can disrupt swallow function</td>
</tr>
<tr>
<td>Lesions</td>
<td>�</td>
<td>�</td>
<td>�</td>
</tr>
<tr>
<td>Treatment</td>
<td>Immunosuppressants, immunosuppressants, immunomodulating disease-modifying agents, immunosuppressants, and corticosteroids</td>
<td>Immunomodulating disease-modifying agents, immunosuppressants, and corticosteroids</td>
<td>Common treatments include plasmapheresis and IVIg</td>
</tr>
<tr>
<td>Recovery</td>
<td>Most patients make full recovery</td>
<td>Average survival is 15 to 20 years after diagnosis</td>
<td>Average survival is 15 to 20 years after diagnosis</td>
</tr>
</tbody>
</table>


Kendrea Focht, CScD, CCC-SLP

Adult Progressive Neurological Disorders

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2013 Charleston Swallowing Conference
Amyotrophic Lateral Sclerosis: 
Speech and Swallowing Management 
A Changing Historical Perspective

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www.plowmanlab.org

Overview
1. Bulbar dysfunction in persons with ALS
2. Historical management of bulbar dysfunction
3. Recent evidence:
   I. Basic Science
   II. Clinical Science
4. Application to our field
5. Conclusions

1. Bulbar Dysfunction in 
ALS
Swallow

Cough

Cough
2. Historical Management of Bulbar Dysfunction in ALS
Historical Clinical Approach

1. Surgical Interventions
2. Manage and Do not treat
3. Swallow: PEG tube placement
4. Speech: AAC Device

3. Emerging Evidence

Basic Science Data in Transgenic Mouse Model

Demonstrates that moderate intensity aerobic exercise:
1. delays disease onset
2. slows disease progression
3. increases lifespan
Clinical Data

- Limb Literature
- Respiratory Studies
- Plowman Lab pilot data and R21 Study

4. Application to Bulbar System

Conclusions
Adult Progressive Neurological Disorders

Common Progressive Neurological Disorders
- Parkinson's Disease
- Amyotrophic Lateral Sclerosis
- Multiple Sclerosis
- Myasthenia Gravis

Parkinson's Disease
- Degenerative disorder of the CNS with symptoms resulting from degeneration of dopamine producing cells in the substantia niagra in the midbrain
Motor Symptoms

- Four hallmark motor symptoms
- Tremor
- Rigidity
- Slowness of movement
- Postural instability

Tremor

- It is usually a rest tremor: maximal when the limb is at rest and disappearing with voluntary movement and sleep
- 4-6 Hz, Pill rolling
Rigidity
- Stiffness or increased resistance to the movement of a muscle due to increased muscle tone.
- With the progression of the disease, rigidity can affect the whole body and reduce the ability to move.

Bradykinesia
- Performance of repetitive or sequential movement is impaired.
- Bradykinesia is commonly a very disabling symptom in the early stages of PD.

Postural Instability
- More common in the late stages of the disease
- Results in increasing balance difficulties and falls
Amyotrophic Lateral Sclerosis

- Progressive degeneration of motor neurons
- Involves primary cortical motor neurons and anterior horn cells of spinal cord.
- Unique combination of upper motor neuron pathology and lower motor neuron pathology

UMN vs LMN Signs

**Upper Motor Neuron**
- Spasticity/clonus
- Pseudobulbar speech
- Pathologic reflexes
- Loss of dexterity
- Flexor spasms
- Weak in UMN pattern
- Brisk Jaw Jerk/Gag

**Lower Motor Neuron**
- Atrophy
- Hyporeflexia
- Fasciculations
- Weakness
- Cramps
- Flaccid speech

ALS Presentation

- Weakness
  - Asymmetric
  - Limb weakness
  - Upper > Lower extremity
  - Bulbar dysfunction
  - Dysarthria
  - Dysphagia
ALS
- Life expectancy < 5 yrs from symptom onset.
  Course < 3 yrs from diagnosis
- Mortality due to respiratory weakness and pneumonia (aspiration)

ALS
- Progressive disability related to:
  - Impairment in mobility
  - Impairment in communication
  - Respiratory insufficiency
  - Dysphagia/aspiration

Multiple sclerosis
- Inflammatory disorder that results in demyelination of CNS axons.
- Takes several forms, with new symptoms either occurring in isolated attacks (RELAPSING AND REMITTING) or building up over time (PRIMARY OR SECONDARY PROGRESSIVE)
- Between attacks, symptoms can go away completely
- However, permanent neurological problems often occur, especially as the disease advances
The disease usually begins between the ages of 20 and 50 and is twice as common in women as in men. It can present with focal dysfunction due to the focal nature of the inflammatory process.

**Treatments**

- During symptomatic attacks, administration of high dose intravenous steroids is typically administered.
- Although generally effective in the short term for relieving symptoms, corticosteroid treatments do not appear to have a significant impact on long-term recovery.
Treatments

- Eight disease-modifying treatments have been approved by regulatory agencies for relapsing-remitting multiple sclerosis (RRMS)
- In RRMS they are modestly effective at decreasing the number of attacks

MS

- Disability due to cumulative effect of the multiple demyelinating plaques.
- Significant impairment with spinal cord lesions.

Myasthenia Gravis

- Autoimmune disease
- Identifiable antibody
- Response to treatment
Essentials of Diagnosis

- Fluctuating weakness of commonly used voluntary muscles
- Activity increases weakness of affected muscles
- Short-acting anticholinesterases transiently improve the weakness

MG: Presentation

- Ocular: Ptosis, Diplopia
- Bulbar: Difficulty chewing or swallowing
- Extremity: focal or diffuse weakens
- Generalized MG: mixed picture
Examination

- Fatigable weakness
- Extraocular movement limitations
- Response to Tensilon Testing

Treatment

- Symptomatic: Cholinesterase inhibitors: Mestinon
- Thymectomy Hyperplasia/normal in young, Thymoma more likely in patients who present >50 yrs.
- Steroids: High dose initially (mg/kg) can result in remission.

- Attempt to reduce/eliminate steroids secondary to long term side-effects
- Patients may require alternative immunosuppression
  - CellCept
  - cyclosporine
  - Imuran (azathioprine)
Myasthenia Gravis

- Remission common
- Exacerbations also common