Laryngeal Manifestations of Neurological Disorders

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Basic Neurological Motor Pathway

- Pyramidal Motor System
  - Upper Motor Neurons (UMN)
    - Descending Pathways – Pyramidal Tracts
      - Corticospinal tract
      - Corticobulbar tract
    - Activate the lower motor neuron
  - Lower motor neuron (LMN)
    - Peripheral motor nerves
      - Spinal
      - Cranial (Bulbar)

- Neuromuscular junction
  - Neurotransmitter (acetylcholine) released from nerve terminal flows across junction and stimulates muscular contraction
- Muscle
Neuroanatomic pathways

Symptoms suggesting Neuropathology

- Speech
  - Dysarthria, hypernasality, abnormal resonance
- Voice
  - Asthenia, breathiness, instability, strain
- Swallowing
  - Oral incompetence, aspiration, nasal regurgitation, inability to initiate swallow
Clinical Assessment

- Basic head and neck exam, including cranial nerves
- Special attention to:
  - Facial and lateral jaw movements
  - Tongue fasciculations
  - Tongue strength
  - Coordination of tongue movement
  - Laryngeal elevation with swallow
  - Velar function

Clinical Assessment

- Perceptual speech and voice evaluation
- Laryngeal Exam
  - Vocal fold motion
  - Pharyngeal wall motion
- Consider: FEES or MBS
Extrapyramidal Neurologic System

- System of nerve tracts and pathways connecting the cerebral cortex, basal ganglia, thalamus, cerebellum, reticular formation, and spinal neurons in complex circuits not included in the pyramidal system
- Responsible for coordinated reflex interactions
- Affects motor function by either facilitation or suppression

Extrapyramidal Neurologic System

- Voice
  - Hypotonic – flaccid
  - Hypertonic – constricted
- Speech
  - Spastic
  - Ataxic
- Breathing
  - Vocal fold dysfunction (paradoxical motion)
- Swallowing
  - Impaired if associated with significant muscular weakness
## Associated Symptoms

<table>
<thead>
<tr>
<th>FAILURE TO SUPPRESS</th>
<th>FAILURE TO FACILITATE</th>
</tr>
</thead>
<tbody>
<tr>
<td>Tremors</td>
<td>Bradykinesia</td>
</tr>
<tr>
<td>Chorea</td>
<td>Diminished postural responses</td>
</tr>
<tr>
<td>Athetosis</td>
<td></td>
</tr>
<tr>
<td>Dystonia</td>
<td></td>
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<tr>
<td>Myoclonus</td>
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</tbody>
</table>

## Spasmodic Dysphonia

- **Voice**
  - Increased effort
  - Unreliable in different situations (Stress)
  - Whisper is normal
  - Maybe able to sing
  - Abductor and Adductor varieties
    - Patients usually aware of words and situations which make voice worse

- **Swallowing - Uninvolved**
Spasmodic Dysphonia

--- Hz --- dB

Spasmodic Dysphonia

--- Hz --- dB
Vocal Tremor

- Voice
  - Tremor
  - Strain/roughness
  - Often deny effort associated with SD
  - Not sound specific

- Swallowing
  - Unaffected
Parkinson’s Disease

- Voice
  - Weak with early fatigue
  - Breathy - soft
  - Pitch elevated

- Speech
  - “mumble”

- Swallowing – potential problems late in disease
Multiple System Atrophy

- Shy-Drager syndrome
- Progresses more quickly than PD
- Autonomic dysfunction
- Parkinsonism
- Ataxia
- Stridor and dysphagia
UMN Pathway Disruption

- Spasticity
  - spastic dysarthria
  - spastic dysphonia

- Swallowing and other vegetative functions - relatively well preserved until disruption is severe
  - Swallowing - Inability of UES to relax
  - Breathing - Inability of vocal folds to relax to produce voice or allow inspiration
LMN Pathway Disruption

- Flaccidity
  - flaccid dysarthria
  - flaccid dysphonia

- Swallowing and other vegetative functions are affected early
  - Dysphagia to liquids
  - Breathing – impaired due to lack of abduction
Associated Signs & Symptoms

- Upper motor: spasticity, hypertonia, hyperreflexia, clonus, Babinski sign
- Lower motor: flaccidity, hypotonia, hyporeflexia, atrophy, fasciculations (usually for motor neuron disease only)

Site of Lesion

- Extrapyramidal disorders
  - Parkinson’s disease
  - Cerebellar stroke
  - Spasmodic dysphonia
  - Tremor
- Upper motor neuron disorders
  - Stroke
  - Pseudobulbar palsy
  - Primary lateral sclerosis (PLS)
Site of Lesion

- Lower motor neuron
  - Brainstem stroke (e.g. lateral medullary syndrome)
  - Myasthenia gravis
  - Guillain-Barre'
  - Polio (post-polio)

- Mixed
  - TBI
  - Motor Neuron Disease
    - ALS
    - Progressive Bulbar Palsy

Motor Neuron Disease

<table>
<thead>
<tr>
<th>Type</th>
<th>UMN degeneration</th>
<th>LMN degeneration</th>
</tr>
</thead>
<tbody>
<tr>
<td>ALS</td>
<td>yes</td>
<td>yes</td>
</tr>
<tr>
<td>PLS</td>
<td>yes</td>
<td>no</td>
</tr>
<tr>
<td>PMA</td>
<td>no</td>
<td>yes</td>
</tr>
<tr>
<td>Progressive bulbar palsy</td>
<td>no</td>
<td>yes - bulbar region</td>
</tr>
<tr>
<td>Pseudobulbar palsy</td>
<td>yes - bulbar region</td>
<td>no</td>
</tr>
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Motor Neuron Disease in the Otolaryngology Clinic

- 1759 patients presented with voice, speech and swallowing complaints
  - 15/1759 diagnosed with ALS
  - Referring diagnoses included
    - Unknown neurological disease
    - GERD
    - Stroke
    - Bowing
    - SD
    - Polyp
  - Typical time between initial ENT visit and accurate diagnosis was 6 months

- 220 patients diagnosed with MND in Neurology clinic
  - 44/220 presented with bulbar signs (dysarthria, dysphagia, dysphonia)
  - 19/44 initially presented to otolaryngologist
  - 8/19 neuromuscular disease was missed initially by ENT

Treatment Options

Relief of Spasticity
Treatment Options

Relief of Spasticity - PLS

Imperovement of Glottic Closure
Spasmodic Dysphonia

Parkinson’s Disease
Role of the Otolaryngologist

- Acute observation of the presenting signs and symptoms
- Knowledge of the corresponding neuroanatomy and possible disease states
- Expedient referral to appropriate neurological evaluation
- Primary management
  - Airway safety
  - Other disorders of head and neck – atrophy, spasticity