



Laryngeal Manifestations of Neurological Disorders

Katherine C. Yung, MD
Assistant Professor, Division of Laryngology
Dept. of Otolaryngology-Head and Neck Surgery

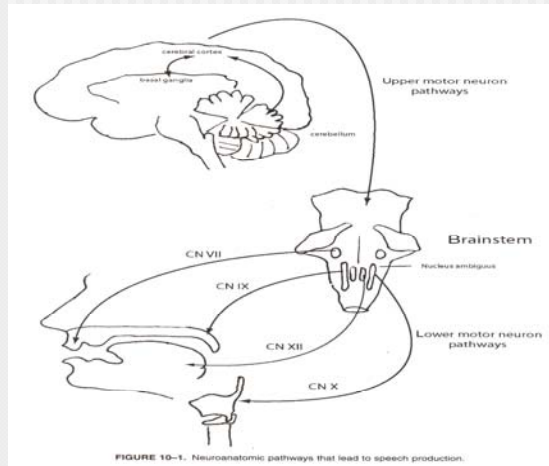


Basic Neurological Motor Pathway

- 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



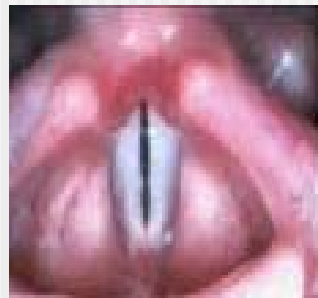
Clinical Assessment

- 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

FAILURE TO SUPPRESS

- Tremors
- Chorea
- Athetosis
- Dystonia
- Myoclonus

FAILURE TO FACILITATE

- Bradykinesia
- Diminished postural responses



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



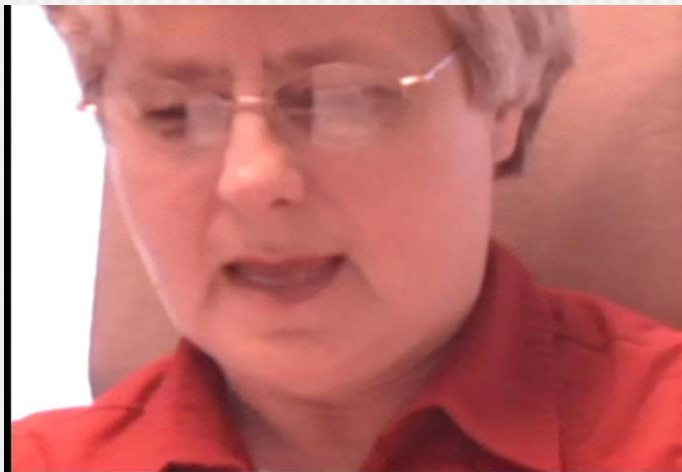
UCSF Department of Otolaryngology - Head and Neck Surgery
San Francisco, California

Spasmodic Dysphonia



UCSF Department of Otolaryngology - Head and Neck Surgery
San Francisco, California

Spasmodic Dysphonia





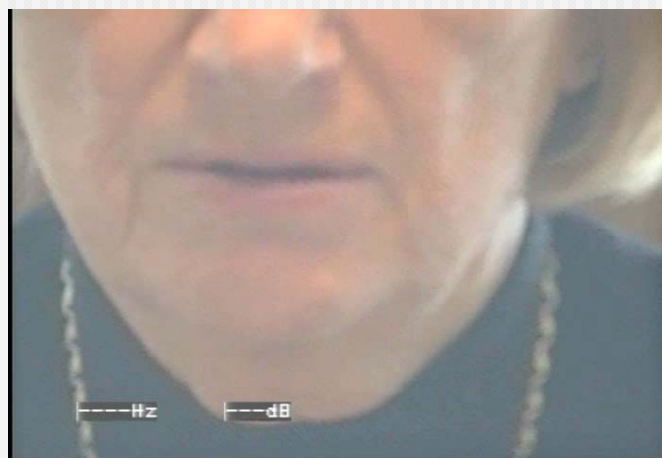
Vocal Tremor

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

- Swallowing
 - Unaffected



Tremor





Parkinson's Disease

- Voice
 - Weak with early fatigue
 - Breathy - soft
 - Pitch elevated
- Speech
 - "mumble"
- Swallowing – potential problems late in disease



Parkinson's Disease





Multiple System Atrophy

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



Multiple System Atrophy





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



UMN Pathway Disruption





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



LMN Pathway Disruption





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

Type	<u>UMN</u> degeneration	<u>LMN</u> degeneration
<u>ALS</u>	yes	yes
<u>PLS</u>	yes	no
<u>PMA</u>	no	yes
<u>Progressive bulbar palsy</u>	no	yes - bulbar region
<u>Pseudobulbar palsy</u>	yes - bulbar region	no



Motor Neuron Disease in the Otolaryngology Clinic

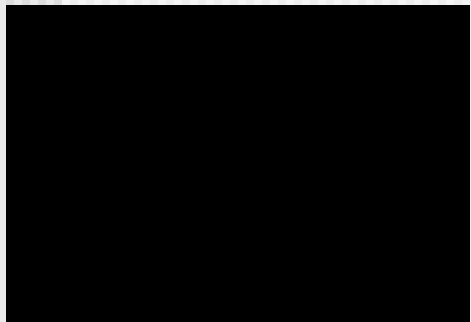
- 1759 patients presented with voice, speech and swallowing complaints
 - 15/1759 diagnosed with ALS
 - Referring diagnoses included
 - Unknown neurological disease
 - GERD
 - Stroke
 - Bowing
 - SD
 - Polyp
 - Typical time between initial ENT visit and accurate diagnosis was 6 months
- 220 patients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

Chen, A, Garrett CG. Otolaryngol Head Neck Surg. 2005 Mar; 132 (3):500-4.



Treatment Options

Relief of Spasticity





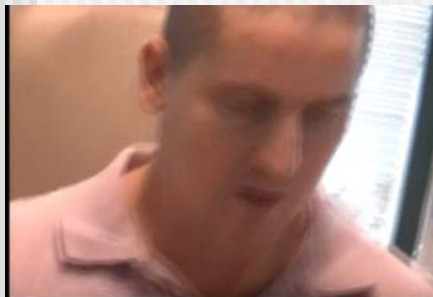
Treatment Options

Relief of Spasticity - PLS



Treatment Options

Improvement 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