A Short Course in Neurology for the SLP: Unraveling the Neurological Knot

"If the human mind were simple enough for us to understand, we would be too simple-minded to understand it."
Anonymous

Overview:

• Medical Speech-Language Pathology: A Brief History
• Neurological basis for swallowing and speech
• A Neuro-diagnostic algorithm
• Neurological assessment for the SLP

MEDICAL SPEECH-LANGUAGE PATHOLOGY
A Brief History

Early challenges:
• Establishing a role for the SLP in medical setting
• Education of SLPs equipped to work in medical settings
• Educating physicians and other health professionals
• Demonstrating economic viability
Treatment Guided by Neurological Models

- Anomia due to TBI
  - Neural networks (direct and indirect)
  - Address the “phonological processor”
- Aphagia due to infarct of PICA
  - Recognize relationship between cortical and subcortical systems

Neurological Basis for Swallowing and Speech

Swallowing

- Sensory-motor response
- Voluntary & reflex behaviors
- Multiple cranial nerves
- Coordinated by a central pattern generator
A Sensory-Motor Response

Motor cortex

UMNs

Motor

Sensory

Brainstem

Voluntary

Reflexive

Cranial Nerves for Swallowing

5 Jaw movement, facial sensation, oral sensation

7 Facial movement, taste

9/10 Taste, palate & pharynx movement, gag, vocal folds

12 Tongue movement

Neural Innervation

Motor cortex

Bulb

Bulbar musculature
These UMN are "direct activation pathways".

UMN = Direct and Indirect Activation Pathways

Direct UMN
- Spasticity
- Hyperreflexive
- Primitive reflexes
- Dyspraxia

Indirect UMN
- Rigidity
- Loss of stability (e.g., tremor)
- Abnormal posture

UMN vs. LMN lesions

Bilateral UMN damage necessary to cause pseudo-bulbar symptoms

Unilateral LMN damage sufficient to cause bulbar symptoms
Gag reflex

Rules of Thumb!

1. Unilateral UMN = rarely severe, lasting dysphagia
2. Bilateral UMN = spastic, supranuclear dysphagia
3. Unilateral LMN = incomplete paralytic dysphagia
4. Bilateral LMN = paralytic dysphagia or aphagia
Motor Control Systems

Motor neurons in brainstem and spinal cord

Motor Control Systems

Basal ganglia posture stability muscle tone
Cerebellum coordination timing of movement
Both communicate to motor cortex via the thalamus

Central Pattern Generator

- Located in the medulla oblongata
- Consists of 2 main groups of paired nuclei
  - Dorsal group—the solitary nucleus
    - Receives peripheral input from sensory organs (via CNs)
    - Receives supranuclear input from cortical and subcortical structures (via UMN)
  - Ventral group—the nucleus ambiguus
    - Distributes swallow drive to pools of motor neurons in various CNs
    - CNs then activate appropriate muscle fibers
    - The patterned movements of swallowing can be elicited from stimulation of either side
    - However, a unilateral medullary lesion is capable of significantly impairing swallowing
DSG (dorsal swallow group) receives input from supranuclear areas and peripheral afferents. DSG neurons activate the VSG (ventral swallow group), which in turn orchestrates actions through motor cranial nerves and cervical spinal nerves.
Sensory receptors for taste, touch, temp stimulated (CNs 5, 7, 9, 10)

Dorsal CPG integrates input

Ventral CPG orchestrates patterned motor response (via CNs 5, 7, 9, 10, 12 and C 1-3)

Supramedullary trigger

“A Most Complex and Unique Mechanism”

Conversational speech:
- ~ 100 muscles contributing
- ~ 100 neuromuscular junctions per muscle
- ~ 14 sounds per second
- = ~ 140,000 neuromuscular events per second

Doesn’t include incalculable neural activity involving cognition and language!
The Conceptual Basis for BG Function

BG operate using important neurotransmitters

- GABA (inhibitory)
- glutamate (excitatory)
- dopamine (both excitatory and inhibitory effects)
- acetylcholine (both excitatory and inhibitory effects)

The Theoretical Basis for BG Function

Too little cortical excitation = poverty of movement, slow movement, hypokinetic

= normal excitation  = normal inhibition
Theoretical Basis for BG Function

Excessive cortical excitation = hyperkinetic, involuntary movement

= normal excitation
= normal inhibition

Motor Control: Cerebellum

• Essential for motor control in timing voluntary movements
  – Lesions may cause ataxia—dysmetria and overshoot
  – “intention tremor”
• Coordinates by controlling errors
• Rapid movements most affected

http://www.mh-heinemann.co.uk/cerebellum.jpg

The motor plan

Fronto-pontine-cerebellar tract

Connections

Muscle information (position, stretch, tension,...)
Cerebellum

- Receives input from peripheral sensory systems (position, muscle tone—stretch, tension...)
- Receives input from motor cortex on intended, voluntary movement
- Compares what is intended to sensory input received
- Corrections are sent back to motor cortex

Cerebellum

- Cerebellar impulses can increase or decrease rate of movement
- Pathology can result in absence of voluntary movement—i.e., akinetic
- Cerebellar output to vestibular nuclei influences muscle tone and balance in skeletal muscles
- Cerebellar function is vital in rapid movements (speaking, running, playing a musical instrument, dancing...)

MOTOR SPEECH

Motor Control Systems
- Cerebellum
- Basal Ganglia

Frontal lobes (Broca’s) for motor speech plan
- Direct and Indirect UMN activation tracts
- LAMNs serving Cranial Nerves 5, 7, 9, 10, 12
LANGUAGE IN THE BRAIN

- Each brain consists of 100,000 miles of myelin-covered nerve fibers
- There are 86 billion neurons in the adult brain
- Each neuron has between 1000 and 10,000 synapses
- There are about 125 trillion synapses in the cerebral cortex alone

Seven Components of the Wernicke-Geschwind Model

How the Wernicke-Geschwind Model Works
• This is a modular model of language processing
  — Also called ‘box-and-arrow’
  — Knowledge is contained within a box and pathways connect boxes
  — Knowledge stored locally, and processed serially— one process at a time

1. See a cow
2. Find the word “cow”
3. Say “cow”

Modular models led to aphasia syndrome classification systems:

• Broca’s -- nonfluent, agrammatic
• Wernicke’s -- fluent with impaired comprehension
• Conduction -- “fluent” but hesitancies with word finding issues, paraphasias, sometime jargon, can’t repeat
• Transcortical motor -- nonfluent, like Broca’s, but preserved repetition
• Transcortical sensory -- fluent, like Wernicke’s, but preserved repetition
• Anomic -- not localized, impaired naming
• Global -- combined fluent and nonfluent characteristics
However, it’s not quite so straightforward!

Highly distributed network of activation

Difficulty with syndrome classification and modular processing:
- Doesn’t adequately describe linguistic details of language impairments
- People with the same syndrome can have different deficits
- Identical deficits can occur in different syndromes (e.g. anomia)
- Modular processing models offer little insight into underlying linguistic mechanisms

Alternatively….
- Consider models that apply psycholinguistic principles of processing
- Provides insight into underlying linguistic mechanisms
- Provides direction for diagnosis and treatment
- 2 important conceptual points:
  - Parallel distributed processing
  - Spreading activation (Dell 1986, Roelfs 1992)
Spreading activation example: Stimulus YELLOW

Possible limbic activation???

Parallel Distributed Processing Models

Basic principles
1. The representation of information is distributed, not local
2. Memory and knowledge for specific things are not stored explicitly, but represented in the connections between units
3. Learning occurs by gradual changes in connection strength—through experience

Consider the spreading activation that occurs when this stimulus is presented:
Spreading activation
– When a neuron is activated, it spreads to all other units connected to it
  • Consider a 2-level interactive model of naming:

Evidence: Large-scale meta-analysis

In the following slides you will see:

• Meta-analysis from 129 functional imaging studies aimed at defining the composition of the semantic, syntactic/sentence, and phonological processing networks

SEMANTICS: activation peaks elicited when S’s were asked to “provide a name”
SENTENCE / SYNTACTIC: S’s generate a grammatical sentence

VIGNEAU et al., 2006

PHONOLOGICAL: S’s verbalize in imitation

VIGNEAU et al., 2006

UNIQUE and OVERLAPPING NETWORKS
SENTENCE/SYNTACTIC, SEMANTIC, PHONOLOGICAL

VIGNEAU et al., 2006
Think about words

- In dictionaries there is an entry for each word
  - Phonological, graphic, grammatical, semantic
  - All in one place
- In the brain the situation is entirely different
  - Each word is represented as a large network
  - Different kinds of information in different locations
  - Also true for each phrase that is learned as a unit, e.g., “How was your day?” or “What’s for dinner?”

Based on work by: Sydney Lamb, Rice University
"The effective unit of operation...is not the single neuron and its axon, but bundles or groups of cells and their axons with similar functional properties and anatomical connections."

Nodal interconnections
(known facts from neuroanatomy)

• Nodes (columns or vertical networks) are connected to
  – Nearby nodes
  – Distant nodes via myelinated association pathways
• Connections to nearby nodes are either excitatory or inhibitory
  – Via horizontal axons (through gray matter)
• Connections to distant nodes are excitatory only
  – Via longer myelinated axons of pyramidal neurons

Some things that are now well established

• The brain is a network
  – Composed of neurons
    • Neurons are interconnected
  – Activity travels along neural pathways
  – Cortical neurons are clustered in columns
    – Columns come in different sizes
      – The smallest: minicolumn ~ 70-110 neurons
    – Each minicolumn acts as a unit
      – When it becomes active all its neurons are active
• Different networks in different locations for various kinds of “information”
  – Visual, auditory, tactile, motor, memories, emotion...

Uniformity of cortical structure

• What distinguishes one kind of information from another is what it is connected to
• Lines and nodes are approximately the same for all associational cortical areas
  – Same kinds of columnar structure
  – Same kinds of neurons
  – Same kinds of connections
• Different areas have different functions because of what they are connected to
What impairments are found at each level of processing?

Consider...

“In diagnostic testing and assessment, we want the patient to fail IF THEY WILL; whereas in treatment we want the patient to succeed IF THEY CAN.”
References

- http://www.ruf.rice.edu/~lngbrain/main.htm (Sydney Lamb, Rice Univ.)
A Diagnostic Algorithm for Swallowing and Speech in Degenerative Disease

Dysphagia Diagnostic Decision Tree

ESOPHAGEAL DYSPHAGIA

Food stops or sticks

ESOPHAGEAL DYSPHAGIA

Food stops or sticks

ESOPHAGEAL DYSPHAGIA

Food stops or sticks

ESOPHAGEAL DYSPHAGIA

Food stops or sticks
Dysphagia diagnostic decision tree

General Characteristics of Neuro Dysphagia

Changes in muscle:
- Tone
- Strength / force
- Range of movement
- Speed
- Coordination
- Control

Common co-impairments:
- Motor speech (dysarthria)
- Impaired mental functions
- Impairments in other motor behaviors
- Abnormal reflexes

SUPRAMEDULLARY PATHOLOGY
Characteristics of Supramedullary Conditions

- Pathology in direct and indirect activation pathways and/or motor control circuits
- Weakness, spasticity, hyperkinesia, hypokinesia, dystonia and/or ataxia
- Protective reflexes generally intact
- Primitive, pathologic reflexes
- Emotional lability
- Poor coordination for swallow
- Loss of intellectual controls

UMN lesions

Bilateral UMN damage is necessary to cause major symptoms

UMN = Direct and Indirect Activation Pathways

Spasticity
Hyperreflexive
Primitive reflexes
Dyspraxia

Rigidity
Loss of stability (tremor)
Abnormal posture
Motor Control Systems

Basal ganglia
posture
stability
muscle tone
Cerebellum
coordination
timing

Cerebellum

Ataxic Dysarthria

• Articulatory inaccuracy
  – Imprecise consonants
  – Irregular articulatory breakdown
  – Distorted vowels
• Prosodic excess
  – Excess and equal stress
  – Prolonged phonemes
  – Prolonged intervals
• Phonatory-prosodic insufficiency
LOWER MOTOR NEURON

or

Motor Unit Pathology (Neuron cell body, axon, NMJ, muscle fiber)

Common Characteristics of LMN Pathology

- Loss of protective reflexes
- Flaccid weakness
  - Atrophy and fasciculations
- Aphonia
- Respiratory support
- Cognitive and other higher level functions are intact
Unilateral LMN damage is sufficient to cause bulbar symptoms of weakness, flaccidity, atrophy, and loss of reflexes.

**LMN or Motor Unit Pathology**

- Neuron cell bodies; medullary nucleus
- Neuron axons; cranial nerve
- Neuromuscular junction; synapse
- Muscle fibers

**Mixed UMN & LMN Pathology**

- Mixed, upper and lower systems
- Spastic and flaccid weakness
- Atrophy with fasciculations
- Shranked-strangled voice
- Poor breath support
- Hyperventilation
- Slow, imprecise articulation
- Cortico-bulbar tracts and lower motor neurons

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**Brainstem**

**Bulbar musculature**
### Categories of CNS diseases and conditions

- **Supramedullary**
  - CVA (unilateral hemispheric or bilateral)
  - Parkinson's
  - Progressive Supranuclear Palsy
  - Huntington's
  - Multiple Sclerosis
  - Primary Lateral Sclerosis
  - Dementias

- **Medullary**
  - CVA (intranuclear brainstem)
  - Progressive Bulbar Palsy (MND)
  - Mixed (UMN & LMN / supramedullary & medullary)
    - ALS (MND)
    - Vascular

### Categories of PNS diseases and conditions

- Bulbar poliomyelitis
- Peripheral neuropathies
  - Diphtheria
  - Botulism
  - Rabies
  - Diabetes Mellitus
Categories of PNS diseases and conditions
- MOTOR END PLATE
  - Myasthenia Gravis
  - Eaton Lambert

Categories of MUSCLE diseases and conditions
- Muscular Dystrophies
- Polymyositis, Dermatomyositis
- Metabolic Myopathy (thyrotoxicosis, myxedema, steroid myopathy)
- Amyloidosis
- Systemic Lupus Erythematosis
Examining Patients from a Neurologic Perspective

- Making general clinical observations
- Observing gross neurological functions
- Adapting a cranial nerve examination for the SLP
  - Simultaneously judge both pathology and function
    - Define the neuromuscular type—not disease
    - Look for correlating and confirming signs

Basic Principles in S/F Examination for SLPs

1. Perform examination bilaterally; compare sides for symmetry
2. For function, look at multiple dimensions:
   - force (strength)
   - range of motion
   - speed
   - precision
   - stability
   - volitional vs. automatic/reflexive movement

A Complete Neurological Exam

1. Mental Status Examination
2. Cranial Nerve Examination
3. Motor
4. Coordination and Gait
5. Reflexes
6. Sensory
Plantar (Babinski) Reflex

- Normal response is toe flexion
- Toe extension and separation is positive Babinski sign
- Positive Babinski suggests UMN affecting LE in question

Visual field pathways

Optokinetic Nystagmus

- OKN is elicited by a series of visual stimuli moving across the subject's visual field—a rotating drum or strip of cloth with black and white stripes serving as stimuli.
- Stimuli typically elicit pursuit eye movements in the direction of target movement with rapid return saccades in the opposite direction.
- Visual field defects do not cause an asymmetry of OKN. Asymmetric OKN is evidence of a motor lesion. A horizontal asymmetry is most commonly due to a unilateral, posterior hemisphere lesion, often in the parietal association cortex.
Optic

Ophthalmoscopic Exam
- Optic disc
- Retinal vessels

Retinopathy

Papilledema

Normal Retina

Papilledema

Hypertensive Retinopathy

Diabetic Retinopathy
Pupils and Extraocular movements

- Range of motion -- paresis
- Voluntary movement -- praxis
- Nystagmus

Motor Speech Perceptions

- Respiration
- Phonation
- Resonation
- Articulation
- Prosody

Focused Speech & Swallow Exam

- Judge pathology
  - Gather multiple clues to neurological condition
  - No one finding leads to a conclusion
  - Judge pathology vs. normal variance
- Judge function
  - Consider 0-3 ratings for each element to assess change over time
Intraoral exam

Velum

Important Findings from Gag Reflex

- Asymmetrical
- Absent with other pathology
  - Nasal speech
  - Nasal regurgitation
- Loss of sensation
- Hyperactive with other bilateral UMN signs
  - Spastic dysarthria
  - Release reflexes
Gag reflex

Lingual Function

Judge:
- Praxis
- Force
- Speed
- ROM
- Coordination

Laryngeal Elevation
Laryngeal exam

Summary

- Add up the observations and signs
- Look for patterns that help to identify neurological systems involved; not disease
- One sign does not make a diagnosis; look for confirmatory findings
- Practice-Practice-Practice
- Follow degenerative disease patients closely and objectify findings