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Differential Diagnosis of the Dysarthrias

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Today's objectives

- Detail the underlying neuropathology of the dysarthrias.
- Review the importance of differential diagnosis of the dysarthrias.
- Identify salient features that differentiate the dysarthrias including neuropathology, perceptual features, and physical characteristics.

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Differential diagnosis

- Evolution of a discipline
  - A need to better understand and define clinical populations.
  - Evaluation
    - More accurate diagnosis
  - Treatment
    - Guide our treatment decisions

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Background

- Darley, Aronson, & Brown
  - Pivotal articles (1969)
  - “Motor Speech Disorders” text (1975)
  - Defined and identified the dysarthrias
    - Initial sample = 212 patients
      - Lesion site
      - Perceptual characteristics
      - Physical characteristics

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Mayo system classification

- Medical/etiology-based method of classification
- Which aspects of nervous system are affected and what perceptual speech behaviors result?
- Basic principles
  - Not all people with dysarthria sound the same
  - The dysarthrias can be recognized by how they sound
  - Similarities in types of dysarthria reflect lesion loci and common underlying pathophysiology

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Dysarthria types

- Flaccid
- Spastic
- Unilateral upper motor neuron (UUMN)
- Ataxic
- Hypokinetic
- Hyperkinetic
- Mixed

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The Dysarthrias

"A collective name for a group of *neurologic* speech disorders resulting from abnormalities in the strength, speed, range, steadiness, tone, or accuracy of movements required for control of the respiratory, phonatory, resonatory, articulatory, and prosodic aspects of speech production. The responsible pathophysiologic disturbances are due to central or peripheral nervous system abnormalities and most often reflect weakness; spasticity; incoordination; involuntary movements; or excessive, reduced, or variable muscle tone."

(Duffy, 2020 p. 3)

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Perceptual ratings

- Overall intelligibility
- Respiratory support
- Voice quality
- Articulatory precision
- Resonance balance
- Prosody

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Physical Characteristics

Structure & Function

- Weakness
- Muscle tone
- Confirmatory signs
- Movements
  - Slow
  - Fast
  - Variable
  - Irregular

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Prevalence

The dysarthrias comprised over 47% of referrals to the Division of Speech Pathology over a 7-year span (2009-2016).

- Total sample size 9,430
- Encompassed acute care, acute rehabilitation and outpatient diagnostic and multidisciplinary clinics.

Referral sources:

- Neurology
- Neurosurgery
- Physical medicine and rehabilitation (PM&R)
- Ear, nose, and throat (ENT)
- General internal medicine
- Other

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Benefits & Drawbacks

- Assists with confirmation of lesion site
  - Can rule out other potential diagnoses
- Clinical “shorthand”
- Guides treatment decisions
  - Understanding the underlying pathology results in selecting the appropriate treatment
- Reliability has been questioned
- Corresponding lesion data
  - Clinical knowledge

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Take home message…

- Clinically useful
  - Evolution continues
- Our understanding of neuropathology will never be static
  - New discoveries
  - New procedures
- For now, the system is a useful one that will continue to be refined
  - Hyperkinetic
    - Myriad of movement disorders

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Definition revisited

- Key features of the dysarthrias
  - Neurologic basis
  - Disorder affecting movements
  - Categorized based on
    - Lesion
    - Physical characteristics
    - Speech characteristics
- Cheat sheet…

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Dysarthrias

- Flaccid Dysarthria
  - Results from injury/damage to cranial and/or spinal nerves
    - Final common pathway
    - Lower motor neuron
  - Reflexive, automatic, and voluntary activities are ALL affected when the lesion resides in the FCP.
- Can impact all subsystems of speech
  - Primary characteristics can be traced to weakness
  - Which of the Cranial and/or spinal nerves might be implicated?

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- Etiologies
  - Surgical
  - Nonsurgical
  - Neuropathies
  - Tumor
  - Myasthenia gravis**
  - Degenerative
  - Vascular
  - Anatomic malformation
  - Demyelinating
  - Other

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Lesion analysis

- Trigeminal
  - Innervation properties
    - Three branches (sensory)
    - Motor component
- Signs
  - Sensory loss
  - Jaw deviation (unilateral)
  - Jaw may hang open (bilateral)
- Speech signs
  - Imprecision in connected speech
  - Most affected with bilateral lesions

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- Facial
  - Innervation properties
    - Sensory
    - Motor
  - Which would be most devastating to speech?
- Physical Signs
  - Facial droop - hemiface
  - Drooling (possibly)
  - Pocketing
- Testing
- Speech signs
  - Imprecision in connected speech
    - Related to facial weakness
      - What sounds would be most affected?

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- Glossopharyngeal
  - Innervation properties
    - Sensory
    - Motor
  - Physical signs
  - Speech signs

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- Vagus
  - Innervation properties
    - Sensory
    - Motor
  - Physical Signs
    - Velar signs
    - Laryngeal signs
  - Speech signs
    - Phonation
    - Respiration
    - Resonance

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- Spinal Accessory
  - Innervation properties
    - Motor
  - Physical Signs
    - Shoulder weakness
  - Speech signs

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- Hypoglossal
  - Innervation properties
    - Motor
  - Physical Signs
    - Tongue deviation
    - Atrophy
  - Speech signs
    - Imprecision with articulation
    - Compensation with the mandible

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- Spinal nerve lesions
  - Damage to spinal nerves C1-5
  - Primary damage to respiration
    - How does that interfere with speech?
  - Physical Signs
    - Rapid/shallow breathing
    - Exaggerated thoracic expansion
  - Flaccid dysarthria related to isolated damage to spinal nerve lesions is rare

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Primary Characteristics

- Weakness
- Hypotonia and reduced reflexes
- Fasciculations and fibrillations
- Progressive weakness with use**

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Spastic Dysarthria

- Secondary to damage of the direct and indirect pathways of the CNS (UMN system)
  - Direct pathway
    - Pyramidal tract
    - Corticobulbar tracts
      - Interacts with cranial nerves
    - Corticospinal tracts
      - Interacts with spinal nerves
    - Facilitatory action
      - Leads to skilled discrete movements

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- Indirect pathway
  - Extrapyramidal
  - Regulates reflexes, maintains posture, tone
  - Inhibitory nature

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- Difficulty with fine, discrete movements
  - Initially decreased tone
  - Weakness
  - Develops into increased tone, spasticity
    - Those muscles utilized in fine movements most affected
  - Decreased reflexes initially, then more pronounced
  - Babinski sign
  - Oral motor reflexes
    - Suck
    - Snout
    - Jaw jerk

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Etiologies

- Vascular
  - Brainstem stroke
    - Bilateral effects
    - Cerebral stroke can't have the same impact, unless there is previous damage, or perhaps midline shift/compression from a hemorrhagic stroke
  - Lacunar infarcts
  - Multi-infarct dementia
    - Not a common problem with DAT

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Oral Mechanism

- Dysphagia
- Drooling
- Pseudobulbar affect
- Normal jaw strength
- Articulatory/facial movements may be slow
- Hyperactive reflexes
- Sluggish velar movement
  - Difficulty sustaining elevation

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- Inflammatory
  - Leukoencephalitis
- Degenerative disease
  - Amyotrophic lateral sclerosis
  - Primary lateral sclerosis
  - Progressive supranuclear palsy
  - Multiple system atrophy
- Traumatic Brain Injury
- Multiple Sclerosis
- Neoplasm

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Speech Characteristics

- Impairments noticed most by looking at muscle movement rather than isolated muscles
- ↓ speed
- ↓range
- ↓force
- ↑muscle tone

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- Imprecise artic
- Monopitch
- Reduced stress contours
- Voice changes
- Monoloudness
- Slow rate
- Hypernasality
- Short phrases
- Distorted vowels
- Pitch breaks
- Excess and equal stress

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Ataxic Dysarthria

- Cerebellum
  - Two cerebellar hemispheres
  - Vermis
  - Ipsilateral contribution
    - Unlike the cerebral hemispheres which have contralateral contributions
    - With median lesions, effects may be bilateral

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Cerebellar “circuitry”

- The “circuit”
  - Notice of movement-cerebral hemispheres
  - Monitors the movement via feedback from the muscles/joints that conduct the movement.
  - Regulates movement in an ongoing fashion based on both ends of this circuit.

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- All related to timing and force of motor movement/execution
  - Ataxic dysarthria typically manifests with diffuse cerebellar damage
    - Any signs/symptoms from unilateral lesions tend to improve rapidly
  - Disordered gait
  - Truncal instability, titubation
  - Rotated head postures
  - Occular motor abnormalities

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- Degenerative Diseases
  - Hereditary Ataxias
  - Multiple Sclerosis
    - Paroxysmal Ataxic Dysarthria
- Vascular Disorders
  - Aneurysms
  - AVMs
  - Occlusion in the vertebrobasilar system
- Neoplasm
- Trauma

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- Toxic/metabolic conditions
  - Alcoholic cerebellar degeneration
  - Severe malnutrition
    - Vitamin deficiencies
  - Pharmacological
    - Antiseizure drugs
    - Lithium
    - Valium
  - Hypothyroidism
  - Normal Pressure Hydrocephalus

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Oral Mechanism

- Most aspects of the OME can be entirely normal
  - Size
  - Strength
  - Symmetry
  - Face, jaw, tongue, palate
  - Rest, and during sustained postures
  - Gag
  - Reflexive swallow

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OME abnormalities

- Irregular AMRs
  - Observe the regularity of movement of the tongue, lips, and jaw
  - AMRs and connected speech
- Systemic problem
  - Affects the entire system rather than isolated impairment from muscles/muscle groups

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- Imprecision with articulation
- Irregular articulatory breakdown
- Distorted vowels
- Excess and/or equal stress
- Prolonged phonemes/pauses
- Slow rate
- Harshness
- Monopitch
- Monoloudness
- Reduced/irregular breath groups
- Telescoping of syllables

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Hypokinetic Dysarthria

- Pathology of the basal ganglia control circuit
  - Muscle rigidity
  - reduced force/range
- Decreased mobility a/o range leads to the term “hypokinetic”
- The only dysarthria that has “rapid” speech rate as one of the perceptual characteristics
- Depletion or reduction of the neurotransmitter, dopamine
- Parkinson’s disease
  - best reflects hypokinetic dysarthria

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Basal Ganglia Control Circuit

- Functions
  - regulate muscle tone
  - movements that support goal directed activity
  - postural adjustments
  - adjust movements to the environment
  - assists in learning new movements
- Damage results in reduced movement a/o a failure to inhibit involuntary movement
- Hypokinetic dysarthria most reflects the reduced movement

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Clinical Characteristics

- Resting tremor
  - 4-7 Hz
  - Limbs
  - Pill-rolling
  - Jaw, lips, and tongue
- Rigidity
  - resistance to passive stretch
  - decreased movement
- Bradykinesia or Akinesia
  - slow movement
  - freezing
  - shuffling gait
  - micrographia
  - masked facies
- “He always looks like he is mad at me”

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- Reduction in
  - arm movement when walking
  - gestures
  - blinking
  - head movement
  - swallowing frequency
- Loss of postural reflexes
  - reduced “righting” response
- Stooped posture
- Difficulty initiating movement
  - while lying down
  - sitting to standing
- FOG – freezing of gait

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Etiologies

- Damage to the basal ganglia
  - degenerative
  - vascular
  - Traumatic
  - Inflammatory
  - Neoplastic
  - Toxic
  - Metabolic disease

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Oral Mechanism

- Immediate revealing features
  - Masked facies
    - flat affect
  - Respiratory signs
  - Infrequent swallowing
    - accumulation of saliva/drooling
  - Size, strength, and symmetry may be normal
  - AMRs may be slowly initiated a/o rapid and restricted in range

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Speech Characteristics

- Monopitch
- Monoloudness
  - reduced in intensity
  - breathy quality
- Reduced stress
- Short phrases
- Variable rate
- Short rushes of speech
- Imprecise consonants

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Hyperkinetic Dysarthria

- Another dysarthria type that manifests secondary to damage to the basal ganglia control circuit
- May affect all or isolated speech subsystems
- Involuntary movement is the hallmark feature
  - rhythmic
  - irregular
  - fast
  - slow

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Abnormal

- What constitutes abnormal?
- Involuntary movements when steadiness is expected
- Hyper in this case does not mean fast
  - excessive/extra
- Many forms
  - classified on speed and periodicity

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Classification

- Dyskinesia
- Myoclonus
- Tics
- Chorea
- Ballism
- Athetosis
- Dystonia
- Spasm
- Tremor

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Etiologies

- Degenerative
- Vascular
- Traumatic
- Inflammatory
- Toxic
- Metabolic Disease

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Upper Motor Neuron Dysarthria

- Unilateral lesions of the upper motor neuron system
  - UMN system
    - Direct and indirect pathways
      - Direct signs
        - Hemiplegia-paresis
        - Lower facial weakness
          - Contralateral
        - Possible unilateral weakness of the tongue, palate, larynx
      - Indirect signs
        - Increased muscle tone
        - Spasticity
          - Occurs over time

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Etiologies

- Unilateral
  - Stroke
  - Tumors
  - Focal trauma
  - Lacunar infarcts
- Severity
  - Often improves rapidly
    - Dependent upon lesion characteristics
    - Left side lesions can co-occur with language deficits
- OME findings
  - Contralateral facial weakness
    - UMN pattern, or “central” to differentiate from LMN pattern.
  - Unilateral lingual weakness

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Mixed Dysarthrias

- Combination of dysarthrias
  - two or more
  - diffuse damage
- Etiologies
  - Degenerative
    - ALS
    - MS
    - PSP
  - Toxic metabolic
    - Wilson's disease
    - Hepatocerebral degeneration
    - Hypoxic encephalopathy
  - Vascular disorders
  - Trauma
  - Neoplasm
  - Infectious diseases

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Nonorganic Speech Disorders

- A group of speech disorders that can by all appearances, present as a dysarthria (or at times, another speech disorder).
  - Complex symptomatology
  - Complicated history
- Review the medical work-up
  - Is there evidence to support a neurological basis/etiology
- Speech examination
  - Do the symptoms “fit”
  - Are the symptoms distractible
  - Fluctuations in performance
    - Nature and severity of fluctuations

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- In line with physiologic expectations?
- Can the symptoms be modified?
  - Reports of intermittent “normal” speech?
  - Improve with trial of therapy?

- Caveat: Be aware, and beware of bias. A functional label can also lead down the wrong path.

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