Augmentative and Alternative Communication in Motor Neuron Disease - ALS Type

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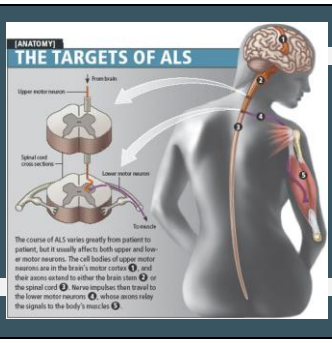
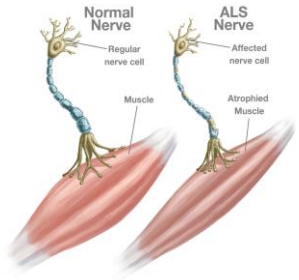
Learning Objectives

- To describe the neurological symptoms of ALS
- To describe the dysarthria in ALS
- To describe the speech pathologist's role as ALS progresses
- To review the role/types of AAC from low tech to high tech



ALS Overview

- Upper Motor Neuron
  - Motor Cortex
  - Brain stem
- Lower Motor Neurons
  - Brain stem
  - Spinal cord



Epidemiology

- Incidence: ~2 per 100,000/year
- Prevalence: ~5–7 per 100,000
- Typical age of onset: 55–75
- Median survival: 2–5 years after diagnosis
- Estimated ~$250,000 out-of-pocket cost for care
- ~25–30% present with bulbar onset (speech/swallowing first)
- ALS is not a notifiable disease

Epidemiology

10% inherited through genetic mutation

90% Sporadic

- Environmental toxins
  - Beta-methylamino-L-alanine (BMAA)
  - Heavy metals (lead, manganese, mercury, zinc, and copper)
  - Pesticides, herbicides, insecticides
  - Solvents (benzene, formaldehyde, and methylene chloride
- Brain & spinal cord injuries
- Military service
- Occupations
  - Military
  - Professional sports
  - Manufacturing, welding, mining
  - Painting, construction
  - Electricians, mechanics, train drivers
- Smoking*

Bulbar Onset vs Spinal Onset ALS

Bulbar Onset	Spinal Onset
• Dysarthria • Dysphagia • Emotional lability • Faster speech decline	• Limb weakness • Gait problems • Speech/swallowing issues later in disease

Clinical Features

Upper Motor Neuron	• Spasticity • Muscle weakness/slowness • Hyperreflexia
Lower Motor Neuron	• Muscle weakness • Atrophy • Twitches/fasciculations • Hypotonia • Areflexia

Bulbar Symptoms	Signs of Frontal lobe dysfunction	Changes in Emotional Control
• Motor speech changes or dysarthria • Diminished reflexive swallow & drooling • Abnormal swallow or oropharyngeal dysphagia	• Frontotemporal dementia • ~50% • Language, cognitive, memory changes	• Pseudobulbar affect

Dysarthria in ALS

Damage of the direct and indirect pathways of the CNS (UMN system)

Clinical Characteristics

Pathologic oral reflexes (sucking, snout, jaw jerk reflex)
Lability
Hypertonia
Hyperactive gag reflex

Motor Speech Characteristics

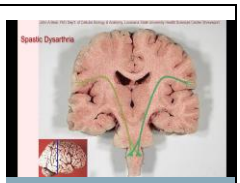
Respiration
- Paradoxical breathing

Phonation
- Harsh, strained
- Pitch breaks

Resonance
- Hypernasality

Articulation
- Imprecise articulation (voicing of voiceless phonemes)
- Slow movement/rate
- Slow & regular AMRs

Prosody
- Reduced stress
- Monopitch
- Monoloudness
- Short phrases



Spastic Dysarthria

- https://www.youtube.com/watch?v=P7lx1F6Qyqk
- https://www.youtube.com/watch?v=ZlKon8qZA4E

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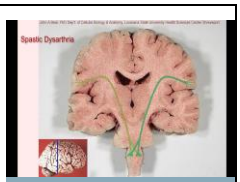


Spastic Dysarthria

Injury/damage to cranial and/or spinal nerves

Lower Motor Neuron

Clinical Characteristics

Flaccidity
Fasciculations
Atrophy
Hypoactive gag reflex
Deterioration that improves with rest

Motor Speech Characteristics

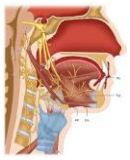
Respiration
- Reduced breath support for phonation
- Reduced vocal intensity

Phonation
- Breathy & hoarse

Resonance
- Hypernasality
- Nasal emission

Articulation
- Imprecise articulation
- Slow rate
- Imprecise AMRs

Prosody
- Excess and equal stress
- Monopitch
- Monoloudness
- Prolonged phonemes



Flaccid Dysarthria

https://www.youtube.com/watch?v=jPFepAOQg0M

Mixed Spastic-Flaccid Dysarthria & Anarthria

https://www.youtube.com/watch?v=DqiqrQerMaQ&embeds_referring_euri=https%3A%2F%2Fneurosigns.org%2F&source_ve_path=MzY4NDIsMzY4NDIsMjc2NjY

- Individuals with ALS will develop mixed spastic-flaccid dysarthria
- Signifies damage to bilateral upper motor neurons and lower motor neurons
- As motor neurons deteriorate further, individuals can eventually become anarthric

Speech Intelligibility

DYSARTHRIA SEVERITY RATING SCALE

(Severity rating is based on acoustic characteristics of functional speech.)

0 = normal: (100%)

Speech is highly intelligible, there are no motor speech problems.

1 = minimal: (95%)

Functional speech is consistently intelligible, although patient may experience subjective difficulties.

2 = mild: (90%)

Functional speech is intelligible but speech does not sound normal.

3 = mild-to-moderate: (80%)

Functional speech is usually intelligible, but repetition may be required.

4 = moderate: (70%-60%)

Production of phrases and short sentences is usually intelligible. Intelligibility decreases as length of output increases.

5 = moderately-severe: (50%-30%)

Functional speech is usually unintelligible, but improves with context, decreased rate, exaggerated articulatory movements or other compensatory techniques.

6 = severe: (<20%)

Unintelligible functional speech; inability to modify speech due to severe motor impairment.

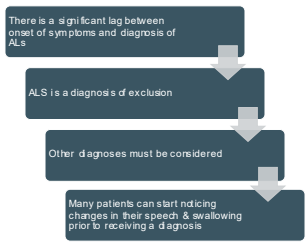
7 = Aphonic/Mute: (0%)

Motor Speech Considerations

- Communication is a human right
- As a neurodegenerative condition, dysarthria is expected to worsen
- Therefore, the individual's needs for compensation will change overtime
- Having reliable access to communication is important
- The intelligibility of someone's speech does not

Differential Diagnosis

Pre-Diagnosis



Pre-Diagnosis SLP's Role

- SLPs should recognize progressive speech symptoms
- Thorough case history and patient interview are critical
- Perform oral motor & motor speech evaluations
  - Assess the various subsystems of speech
  - Diadochokinetic rates
  - Spontaneous speech sample
- Document perceptual speech features and intelligibility

- Avoid recommending intensive speech exercises without diagnosis
- Some progressive neurodegenerative conditions may worsen with exercise-based treatment
- Focus on assessment and monitoring of symptom progression

Condition	Onset Pattern	Speech Characteristics	Key Neurologic Signs	Distinguishing Features
Amyotrophic Lateral Sclerosis	Gradual, progressive	Mixed spastic–flaccid dysarthria	Fasciculations,tongue atrophy, UMN + LMN signs	Speechand swallowing progressively worsen; nosensory loss
Stroke	Sudden	Flaccid or unilateral UMN dysarthria	Cranial nerve deficits, unilateral weakness	Abrupt onset with focal neurologic deficits,can improve with time
MyastheniaGravis	"Crisis"/Fluctuating	Flaccid dysarthria	Ptosis, diplopia, muscle fatigue, myasthenic crisis	Symptoms improve with rest or medication
Parkinson's Disease	Gradual	Hypokinetic dysarthria (monotone, reduced loudness)	Bradykinesia, rigidity, tremor	Speechcan improve with therapy - LSVT ,etc
Primary Lateral Sclerosis	Gradual	Spastic - Slow rate, strained voice, reduced pitch variation	Pure UMN signs (spasticity, hyperreflexia)	No LMN signs(no fasciculations or muscle atrophy)
Huntington's Disease	Gradual	Hyperkinetic - Irregular rate, variable fastness, suddenvoice stoppages	Chorea, cognitivedecline, behavioral changes	Involuntarymovements strongly affect speech

Post Diagnosis

- Multidisciplinary clinic can provide access to various services

- Neurology, Pulmonary, RN coordinator, Social worker, Registered dietician, Physical therapist, Occupational therapist, Speech-Language Pathologist, Respiratory therapist, Mobility/Wheelchair specialist

- Individuals can be at various stages of their disease and dysarthria when they finally receive the diagnosis

- Patient and family education matters

ALS & Treatment

- Neurodegenerative disease = progressive
- ALS is a motor neuron disease so deterioration is expected
- Fatigue can lead to worsened decline
- Research on the benefit/detriment of exercise is limited
- Current guideline is moderate, low-intensity exercise is recommended

SLP's Role Post-Diagnosis

- Speaking & swallowing as exercise
- EMST®
- Provide education about expected speech changes
- Voice/Message Banking
- Counsel patients and families
- Introduce strategies for energy conservation
- Begin proactive communication planning

Augmentative & Alternative Communication (AAC)

AAC

Augmentative Communication: Supplementing existing speech

Alternative Communication: Used in place of speech that is absent or non functional

Low-tech AAC

- Alphabet boards
- Picture/message boards
- Pen & paper
- Boogie board

High-tech AAC

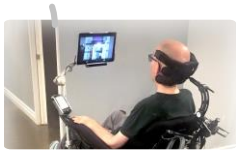
- Tablet devices
- Speech Generating Devices (SGD) - Durable Medical Equipment (DME)

Selection Methods

- Direct
- Switches
- Puff
- Eye Gaze

Speech Generating Device

- Durable Medical Equipment
- Coverage under insurance
- Medicaid/Medicare coverage and requirements vary by state
- Direct vs eye gaze selection coverage can also vary
- Requires an AAC evaluation
- Device is meant to cover all the individual's communication needs
- Communication program, E-mail, Social media, etc.
- Reliable access to communication, **anywhere**

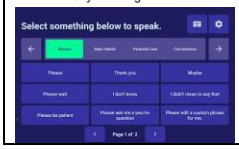


Mounting Systems

- Table mount
- Rolling mount
- Wheelchair mount

Communication Apps

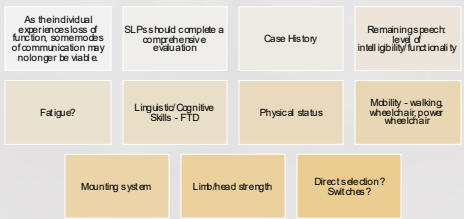
- Good for individuals who want to use a handheld device
- Less complex form of high tech AAC
- Text to speech
- Categories/phrases
- Head/eye tracking



Voice Banking & Message Banking

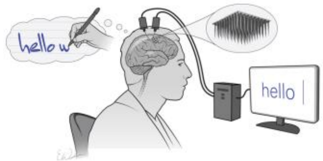
- Banking to be able to use at a later date/on a SGD
- Message banking - records a phrase/expression in the individual's "current voice"
  - Same idea as you create voice memor
- Voice banking - custom synthesized voice. Speech sample gets synthesized into a "new" voice that is able to say novel phrases
  - Requires a certain level of speech intelligibility
  - Some invidiuals with ALS may not have intelligible speech by the time of diagnosis
- Voice cloning/Voice banking with prior recordings
- Varying costs - Educate on resources (Team Gleason, ALS Associations)

AAC Considerations in the ALS Population



Higher-tech AAC (Brain Computer Interface)

- Severe dysarthria/anarthria
- Does not require the individual to learn eye gaze



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