MAY RELS:
ALS Considerations for the Speech Therapist

May 3, 2022

About your presenters:

- John Novak MS MD
  - Neurologist/Neuromuscular Medicine
  - Director of OH ALS Clinic since it began in 2012. Currently held at Westerville Medical Campus and is ALS Association Certified Treatment Center of Excellence.
  - 2022 – ALSA National Care Services Committee
  - Medical Director for Neurology at Grant Medical Center
  - Disclosures – Amylyx, Argenx and ALS Association

About your presenters:

- Michelle Wilkes, MA, CCC/SLP is a licensed and certified Speech-Language Pathologist with over 25 years of experience. She has worked in the acute-care, outpatient rehabilitation, skilled nursing care, and has worked for OhioHealth at Home for over ten years. Michelle has worked with many patients with a variety of different neurological conditions over her tenure. She has also participated and presented for the Ohio Speech Language and Hearing Association conference, ALS Symposium, Central and Southern Ohio ALSA support groups, PSP support group and its National Network, as well as the Parkinson's disease Support group in Akron. Michelle holds additional certifications in VitalStim Therapy as well as Spaced Retrieval Management.

About your presenters:

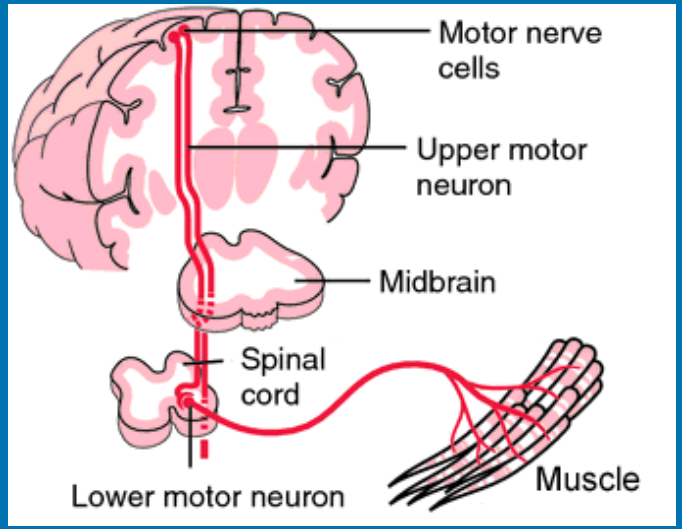
- Sara Bott MA, CCC-SLP is a licensed and certified Speech-Language Pathologist. She has experience with outpatient, private practice and skilled nursing care and has worked for OhioHealth for 2.5 years. She enjoys working with a range of diagnosis such as stroke, concussion and ALS. She is the backup SLP at the ALS clinic at OhioHealth. She has certifications in LVST LOUD and Vital stim.

ALS Background

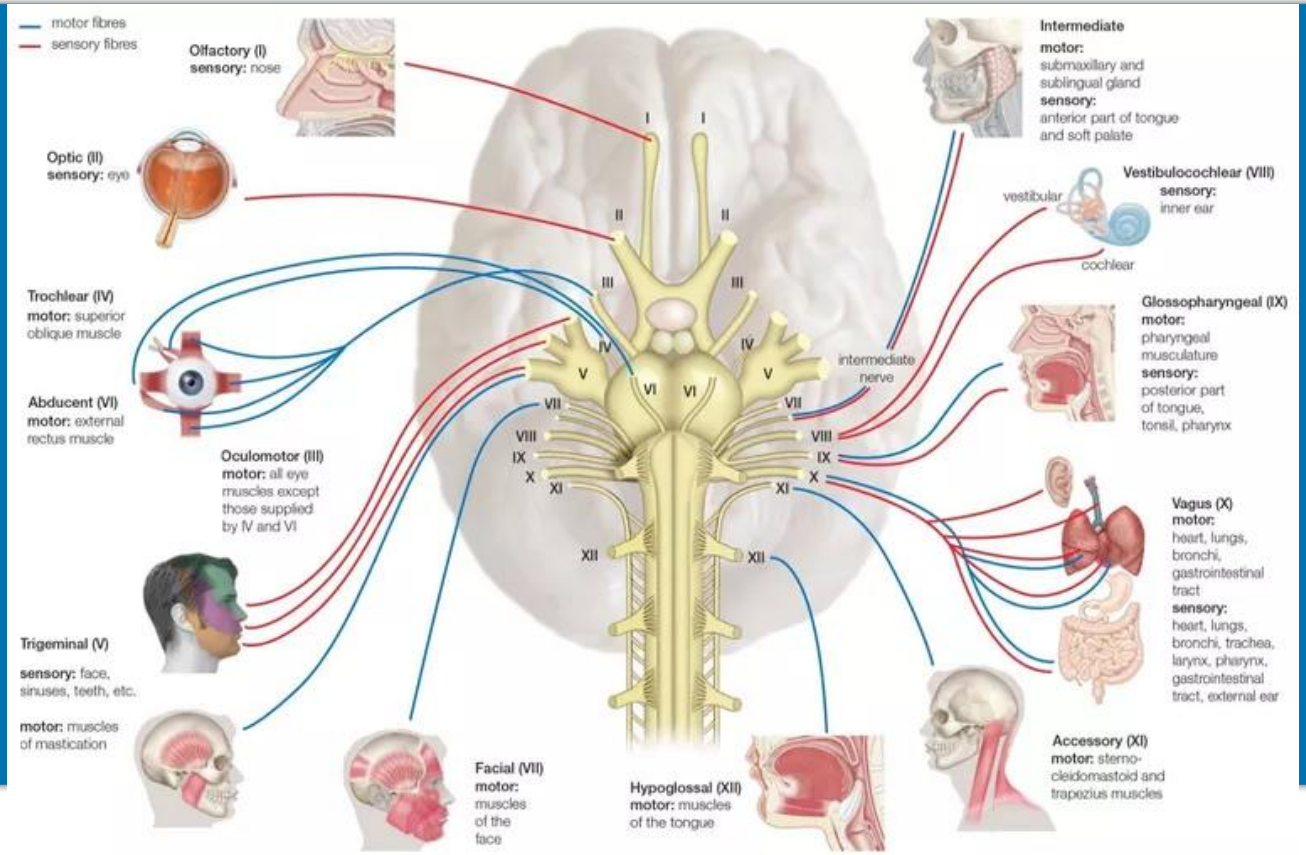
John C Novak MS MD
OhioHealth Neurological Physicians
Director, OhioHealth ALS Clinic

ALS - Amyotrophic Lateral Sclerosis

- Lou Gehrig's, Motor neuron disease
- 2-4/100K people
  - Range 20-80yo
  - Peaks in 60s
  - 10% familial
- Degeneration of Upper and Lower Motor Neurons
  - UMN – spasticity and hyper-reflexia
  - LMN – atrophy and weakness

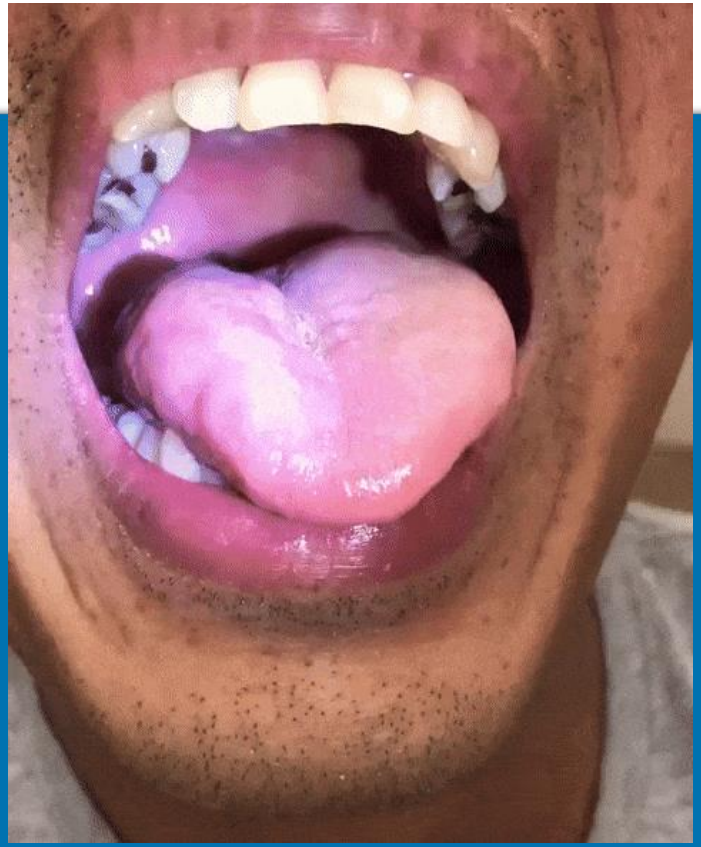


- Focal onset with progression
  - Limb
  - Bulbar
- Variable progression and spread
- Variable degrees of UMN vs LMN
- Respiratory Failure/Death in 3-5 years



Tongue fasciculations
Tongue atrophy

Dysarthria
Dysphagia
Aspiration
Sialorrhea
Facial Weakness
Head drop



- Tongue Atrophy
- Limited movement
- Fasciculations

Progressive Bulbar Palsy

- Focal Variant ALS
  - Bulbar Only
  - Longer Course?
- Possible Spread to Limbs

- Pseudobulbar Affect
  - Emotional lability/incontinence
  - Inappropriate Laughing or Crying
- Nuedexta
  - ? Benefit for swallow

Diagnostic Delay

Clinical diagnosis

- EMG and Exam
- Exclude mimics

12+ months from onset to diagnosis

- Bulbar onset can be faster
- Differential narrower

Delays treatment and therapy

Pre-dx referrals to SLP are common

Treatment

- Medical treatment may slow progression
  - Riluzole
  - Radicava
- Symptom management
  - Sialorrhea – atropine, scopolamine, glycopyrrolate, amitriptyline
  - Spasm – muscle relaxers

- Adaptation to disability

SWALLOWING AND SPEECH CONSIDERATIONS

Rehab In Reverse

- Anticipate decline with regard to DME, adaptive equipment, and activity recommendations! (Senthilkumar 2010)
  - Maximize functional ability
  - Enhance quality of life
  - Assure patient and care giver safety
  - Help people redesign their lives and life goals

- Medicare Maintenance Standard (Jimmo v.Sebelius)
  - "…does not turn on the presence or absence of a beneficiary's potential for improvement, but rather on the beneficiary's need for skilled care."
  - Skilled care may be necessary to:
    - Improve a patient's current condition
    - Maintain the patient's current condition
    - Prevent or slow further deterioration of the patient's condition

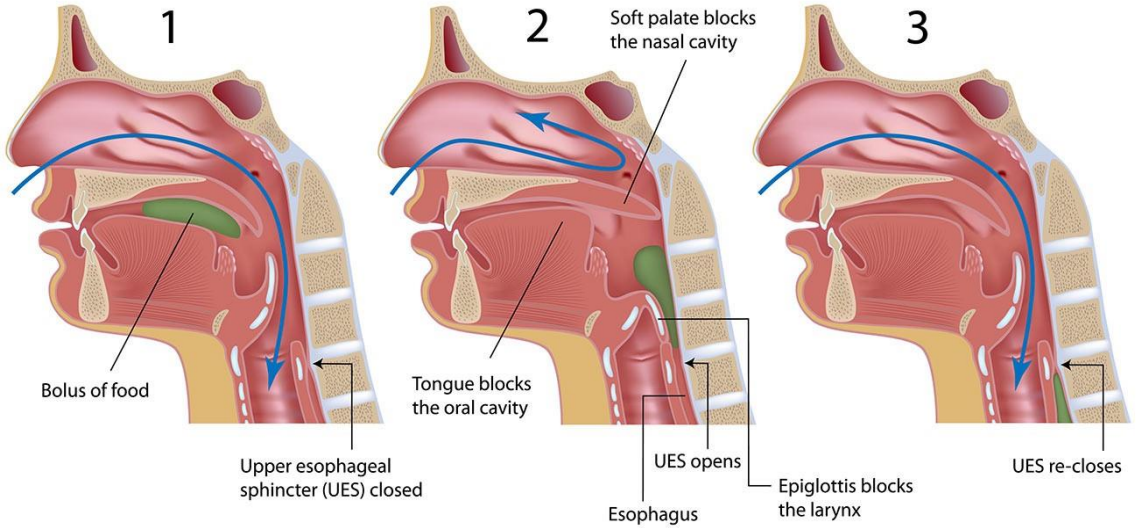
Understanding Swallowing Process

BELIEVE IN *WE* OhioHealth

Swallowing 101

- Swallowing = complex process involving 26 pairs of muscles AND 5 cranial nerves
- A swallow occurs in less than 2 seconds
- Four stages of swallowing
  - Oral preparatory stage
  - Oral stage
  - Pharyngeal stage
  - Esophageal stage

Swallowing



How ALS effects swallowing

Dysphagia and ALS

- 85% of people with ALS will experience dysphagia at some point of the disease (Chen and Garrett, 2005)
- Individuals with “bulbar onset” will likely experience dysphagia earlier in the disease
- Frequent monitoring by the SLP will help identify and intervene to keep our clients' swallow safe

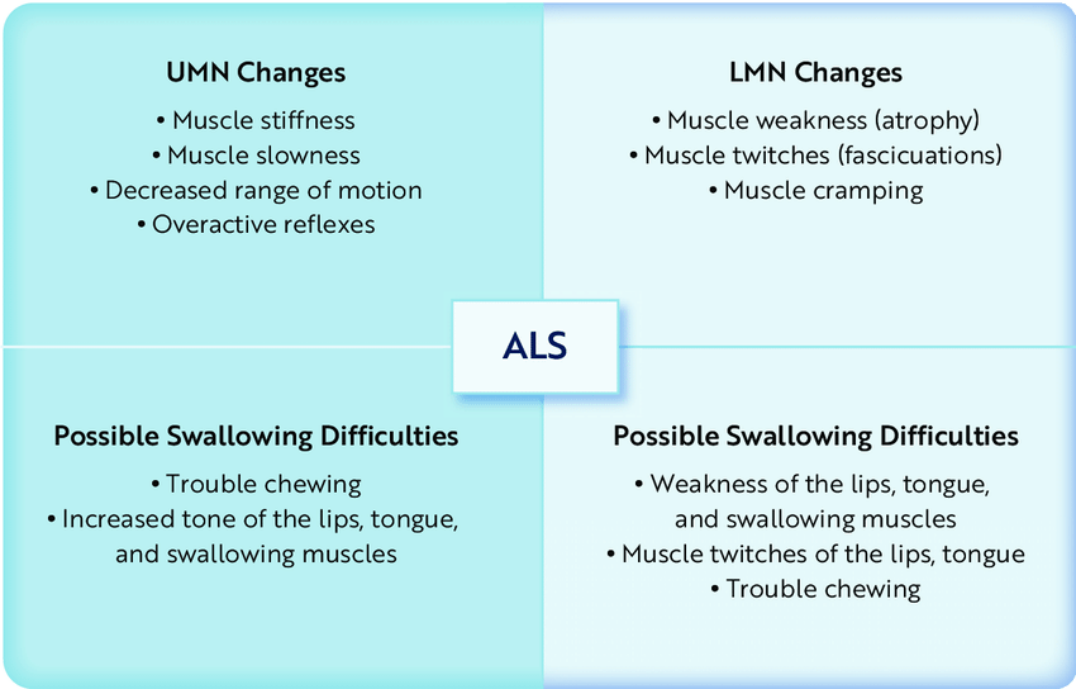
ALS and Swallowing

- People with ALS (pALS) may experience swallowing difficulty/dysphagia due to:

1. Weakness and/or rigidity of the swallowing muscles

2. Difficulty protecting the airway during swallowing

UMN v. LMN Changes



Typical Swallowing Difficulties with ALS

Stage of Swallowing	Swallowing difficulty
Oral Preparatory/Oral Stage	-difficulty managing saliva -difficulty chewing/fatigue with chewing -increased time required to eat -food/liquid spilling out through the lips -drooling -difficulty controlling food/liquid in the mouth -difficulty pushing food/liquid to back of the mouth -residue in the mouth and/or cheek
Pharyngeal Stage	-food “sticking” in the throat -food/liquid coming out the nose -coughing/choking during mealtime -shortness of breath and fatigue during mealtime -reduced cough strength and effectiveness -wet/gurgling-sounding voice
Esophageal Stage	-regurgitation of food/liquid into throat and mouth -food “sticking” in the throat/base of neck

How swallowing is evaluated?

Role of SLP in Swallow Evaluation

Monitor swallowing function and identify changes in function through disease process

- Provide recommendations to ease swallowing during mealtimes and compensate for current difficulties
  - diet modifications
  - posture changes
  - safe swallowing strategies

Swallow Evaluation Tools

- **Bedside exam** – series of questions, physical exam of face/mouth muscles, and observation of swallowing tasks
- **Modified barium swallow study** – X-ray study; gold standard for identifying swallowing impairment and airway invasion
- **Fiberoptic Endoscopic Evaluation of Swallow (FEES)** – assesses food/liquid airway entry before/after swallow only (limited)

Safe Swallowing Strategies

Common Safe Swallow Strategies

- **Effortful swallow** – squeezing all throat muscles to swallow as hard as you can
  - Helps: reduce “leftovers” or residue in throat
  - **Small bites/single sips**
  - Helps: reduce exertion and fatigue during meal
- **Chin tuck procedure** – tucking chin to chest during swallow
  - Helps: protect food/liquid from entering airway
- **Double swallow** – swallowing 2x per sip/bite
  - Helps: eliminate food/liquid left over after initial swallow
  - CAUTION: WATCH FOR FATIGUE

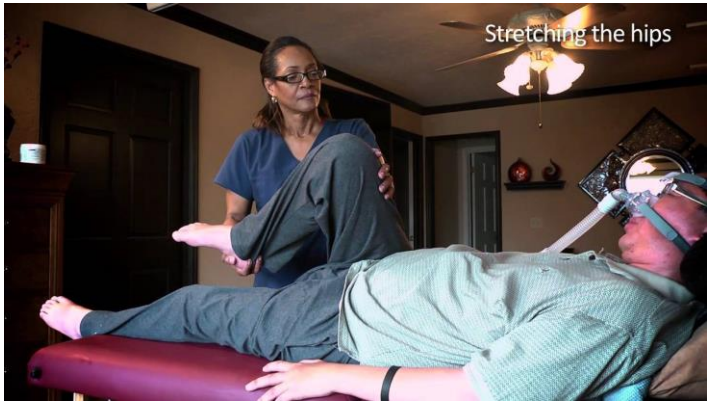
TO EXERCISE OR NOT EXERCISE ????

Aerobic Exercise Guidelines

- Systematic Review: moderate exercise is *not* associated with adverse outcomes in persons with early-stage ALS. (Lui et al 2009)
- Prior to the onset of significant weakness, atrophy, or deconditioning, moderate exercise may have physiological and psychological benefits.
- PALS may participate in an aerobic exercise program if they have the strength and energy to do so, however should be educated on precautions to avoid overexertion.

Stretching and ROM Guidelines

- Daily range of motion exercises should be encouraged to help with maintaining joint mobility and preventing contractures.
- Stretching had the best adherence when compared to strengthening and aerobic exercise (Clawson 2018).



Bulbar musculature is different!

- Speech and swallow muscles are at work 24/7 managing saliva, eating and speaking.

- Need more research in this area!

EMST ; some research…

- By Plowman (2016 pilot, 2019)
- Some improvement with speech and swallowing function
- FVC >60%
- Improved hyoid movement
- Improved cough function
- Nothing Negative happened =)

Issues with study…..

- Did not appear to give a functional change in swallow
- Improved MEP, but will it create longer term changes in speech?
- Can make expiratory muscles stronger, but in the long run will that matter?

Variables to consider

Gentle range of motion/stretching of the oral motor/articulators

pALS respiratory function

Progression rate of the disease process?

Pt motivation…

Ok to do, but don't NEED to do!

Additional variables to consider

- pALS ability to handle own secretions
- pALS energy conservation/toleration
  - Symptom management vs active therapy tolerance
  - Ex: use of EMST vs Breather vs inability to do either

Dietary Modifications

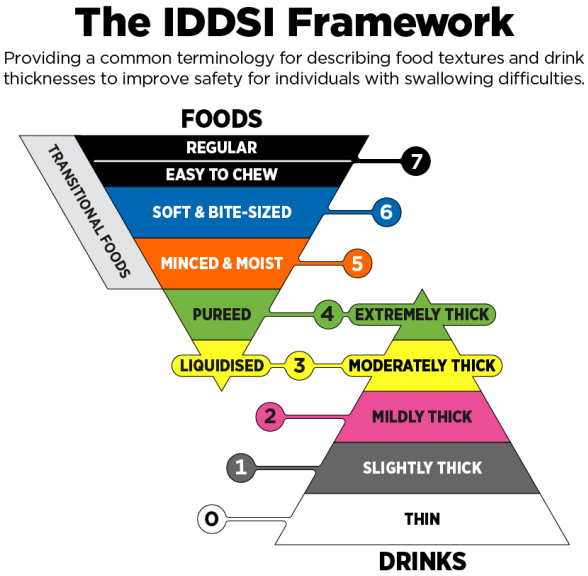
Modified Diets and Mealtime Compensatory Strategies

- **Purpose of food modification:** compensate for swallowing difficulties that our clients may be experiencing

- Altering the consistency to a more appropriate diet/liquid texture will help reduce energy expenditure during eating/chewing/swallowing as well as increase safety

- Adding sauces/gravies to foods can serve as a lubricant and ease passage of food and avoid food getting “stuck” in pALS throat/pharyngeal

International Dysphagia Diet



Why use thickeners?

- Purpose of thickeners:
  - to make regular liquids thicker, more “viscous,”
  - slow the flow rate of the liquid material

- Use of thickener to liquids allows swallowing system more time to coordinate and protect the airway

- Many different brands/types of thickeners are available – The choice is our clients!

- Consider "naturally" thickened liquids

Tips for preparing food for recommended diet levels

- If using thickened liquids, keep a large container of thickened fluid in the fridge for easy access and use throughout the day.
- Reinforce SLP-recommended safe swallow strategies
- Use a blender or Magic Bullet to soften foods that require extensive chewing

Maximizing Nutrition

Mealtime strategies to maintain nutrition

Possible Mealtime Changes	Recommended Compensatory Strategies
Longer Mealtime Duration	-Smaller/more frequent mealtimes during day -Add snacks
Difficulty Chewing	-Moisten foods with gravies and/or sauces -Take smaller bites -Eat softer foods that require less chewing
Weight Loss	-Add foods high in calories (e.g. butter) -Drink full cream milk smoothies/shakes -Supplement nutrition with foods like Boost, Ensure, or Resource Benecalorie -Add snacks
Loss of Enjoyment	-Increase taste, temperature, and textures using spices/sauces
Fatigue/Shortness of Breath	-Take smaller, more frequent meals throughout the day -Minimize exertion during meals (i.e. side conversation, distractions)

PEG considerations

- Maintaining adequate nutrition is IMPORTANT!
- Studies have shown that early placement can increase survival 4-8 months
- Consult with Physician and Dietician (GI/Radiology Doc as well). Early intervention with a team approach is key
- Hypermetabolism (burning calories faster than normal) and/or reduced caloric intake
- Weight loss/Dehydration concerns
- Respiratory considerations
  - Are they a surgical candidate?
- pALS wishes and family feelings

- ULTIMATELY IT SHOULD BE THE pALS CHOICE

Medication Management

BELIEVE IN *WE* OhioHealth

Considerations...

- 1 pill at a time
- Taking with water or thicker item depending on swallowing difficulty (thicker liquid vs puree)
- Cold vs warm presentation
- Sweet vs sour presentation
- Carbonation option
- Chin down vs neutral
- Can pill be crushed (if NOT time released)
- Can medications be altered (compounding pharmacy option)
- Can number of medications be reduced
- Standing/sitting vs lying
- Taking their time!

- EVERYONE IS AN INDIVIDUAL AND NEEDS TO BE TREATED AS SUCH

Secretion Management Issues

Which one or both?

- Sialorrhea (hypersalivation) vs xerostomia (dry mouth)
- Many factors can be the cause including medication side effects, aging issues, post radiation, pooling of secretions due to dysphagia, etc

Xerostomia

- Increasing water intake
- Lozenges (ACT)
- Dry mouth dental products: toothpaste, mouthwash, oral spay (Biotene)
- Severe cases; oil and/or spray (PAM) on toothette

Sialorrhea

- Early stages; increase awareness to swallow and wipe with tissues only if needed
- Controlling amounts of very sweet or sour foods
- Consider portable oral suction device
- Medication intervention

Medication options

- Amitriptyline (Elavil) drops under the tongue (3-4x per day)
- Glycopyrrolate (Robinul) pill every 4 hours (3-4x per day)
- Scopolamine (transdermal patch) lasts 72 hours
- Other options: botox injections and/or radiation if medication failure results

Thick secretions

- Increasing water/fluid intake if able
- Attempt the use of carbonated beverages
- Medication intervention
  - Musinex (guaifenesin): to help thin secretions (blue box)
  - Differences between Musinex, Musinex DM, and Musinex D

***ALWAYS consult physician

Cognitive considerations

Latest research

- Up to 50% of people with ALS might experience some degree of change in thinking or behavior (from mild to severe), with approximately 15% of those developing dementia

- Usually referred to neuropsychologist for cognitive evaluation

Considerations…

- Simplify communication with the affected person
- Provide supervision and accompany the person to all appointments
- Set realistic expectations for the person with ALS
- Educate providers and caregivers working with the person who has thinking/behavior impairment about where to set expectations.
- Emphasize activities they enjoy and be done safely

ALS and Speech Changes

- Changes are common with ALS over time
- Learning compensatory strategies on ways to make speech intelligible and being proactive will help clients to maintain optimal communication
- Beginning working with an SLP at the earliest sign of speech or voice changes
- Dysarthria: term used to describe slow, effortful, slurred speech, and breathy or hoarse voice. Weakening lung muscles affect speech as well.

- ALS leads to speech problems when it attacks bulbar neurons
- Nerve cells responsible for bringing messages from the lower parts of the brain to the muscles that move the lips, tongue, soft palate, jaw, and vocal folds.
- As nerves are lost to the disease, the muscles they control become weak and tight causing dysarthria
- Speaking can make patients tired, especially later in the day. They might find speaking in sentences and that projecting their voice is difficult.

Dysarthria in ALS

- ALS: the muscles that are receiving signals from the brain must compensate for the muscles at are already weak.
- This means functional muscles are doing extra work all the time to compensate and therefore require more frequent and extended rest
- Therapeutic oral exercises designed to strengthen muscles for people with other forms of dysarthria have NOT been demonstrated to improve speech for people with ALS .
- Stretch and massage may be recommended to reduce tightness or maintain range of movement

Prior to or at the First Sign of Speech Changes:

- Have a Speech Therapist measure your speech rate – assists in determining timing for interventions
- Begin Message and Voice Banking so that your own voice can be used in a speech generating device (SGD) or text-to-speech (TTS) app if ever required

Some speech compensatory strategies

- Face-to-face speaking : lip reading and other facial/gestural cues add to listener's ability to understand
- Limit communication to groups of 3 or less friends (see your face better)
- Be aware of your fatigue level which directly impacts your speech
- Plan phone conversations/gatherings for higher energy times and allow for breaks
- Avoid noisy or dimly lit rooms for important conversations
- Slow down speech and exaggerate the sounds, especially at the ends of words where they are most likely to be dropped
- Alert your partner to topic shift and name new topic; distorted speech is easier to understand once the context is established

More…

- Teach frequent communication partners to repeat back any part of message they did not understand vs saying “what?”
- If repeated word is not understood, consider spelling it aloud or using finger to spell on palm or table to help others understand
- If speech is clear but volume is decreased, consider voice amplifier and hands free mic. May help to conserve energy, but may not improve all aspects of clear speech If excess saliva is interfering with your speech, consult with Dr who can prescribe medication or a portable suction machine
- If nasal quality is distorting speech, SLP may suggest a removable, palatal lift appliance or other options to reduce excessive passage of sound through nose
- If communication partner has an uncorrected hearing loss, encourage them to seek services. A hearing aid may be helpful

Message Banking

- The process of recording phrases and expressions that are meaningful to you and those you love
- Words of affection, expressions, phrases that you often repeat, like greetings are especially important.
- Examples: Message Banking (free through Windows Store or Boston Childrens Hospital) or SGD companies

Voice Banking

- The process of creating a customized synthesized voice for you based on samples of speech
- There are different programs for voice banking that range in cost, time expenditure, and clarity of speech required
- My voice… 
- Examples: The Voice Keeper, Model Talker or Acapella

***Free when signed up with the Gleason Foundation

AUGMENTATIVE AND ALTERNATIVE COMMUNICATION

When to consider Augmentative Alternative Communication (AAC)

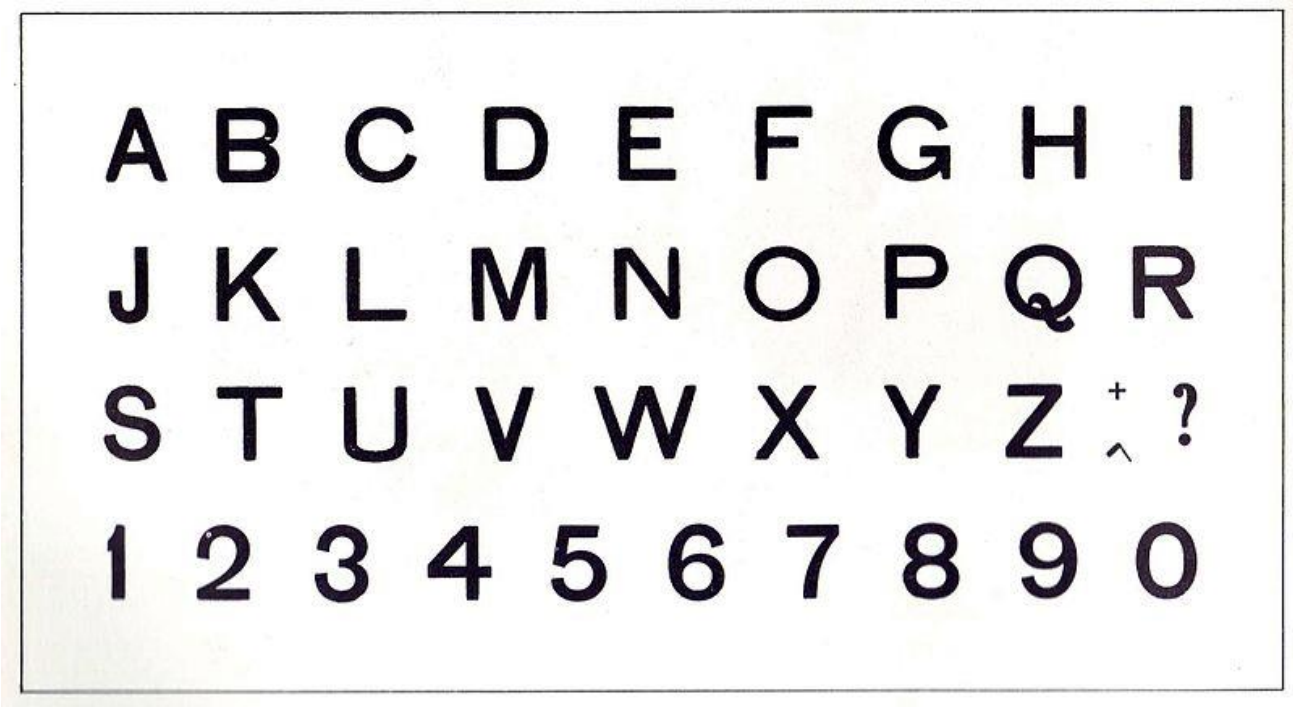
- At the point when speech is beginning to require repetitions or your rate slows significantly, a SLP will often recommend an eval for AAC
- A proactive approach to AAC involves obtaining and customizing the components of a complete AAC system PRIOR to a time when you may need to rely on it communication
- YOUR ABILITY TO COMMUNICATE AT ALL TIMES IS VITAL AND MUST BE MAINTAINED EVEN WHEN SPEECH ALONE IS NOT SUFFICIENT

Low-tech/No Tech Communication

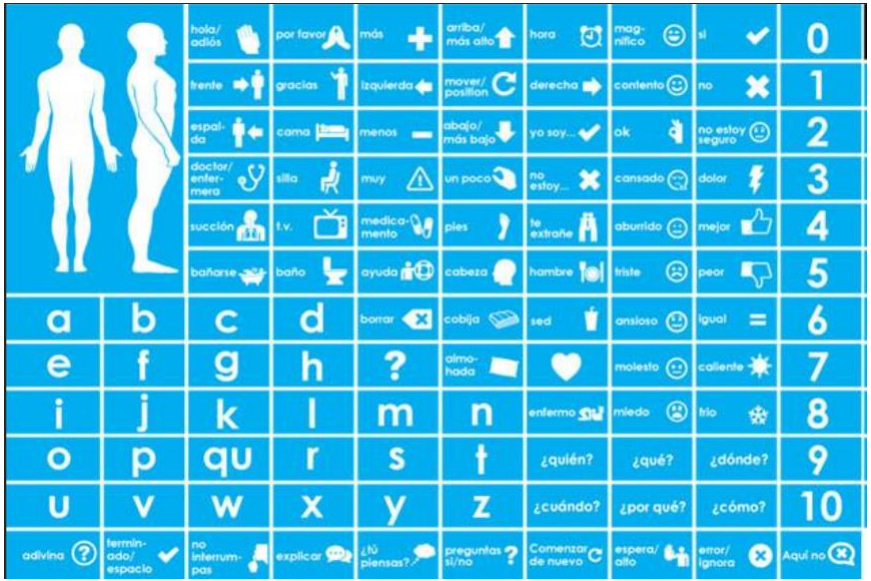
- Sometimes called rapid access communication
- Uses simple or no equipment to convey messages and is an essential part of a complete communication system
- Low-tech communication can be a backup to high-tech SGD
  - Paper communication boards
  - Alphabet boards
  - Boogie boards
  - Writing
- Many people find low-tech/no tech AAC to be preferable in some settings and situations.

- PRIMARY DIFFERENCE: THERE IS NO VOICE PRODUCED BY LOW-TECH AAC. A PARTNER IS REQUIRED TO SPEAK ALOUD THE LETTERS, WORDS, OR PHRASES YOU WRITE OR SELECT FROM A BOARD

Simple alphabet/number board



Simple alphabet/number board



Text-to-speech (TTS) and Speech generating devices (SGDs)

- Provides a means to produce voiced speech, enabling conversation, to get people's attention, talk to the phone, speech with young children, and many other everyday needs
  - TTS: Apps that can be put on phone or tablets
  - SGD: Dedicated devices through a company
- SLPs can assist with insurance and funding choices and to make sure they have the needed accessories (mounting and alternative access equipment) to make the system work best
- SLPs can also assist you installing speech or message banking to SGD or TTS app

SGD companies

- Prentke-Romich Company (PRC) / Saltillo
- Tobii-Dynavox Company
- Forbes Rehab
- Talk to Me Technologies
- Control Bionics
- Lingraphica (Does not have eye gaze)

- LOTS OF NEW COMPANIES AS WELL… EVER CHANGING

***Please consider customer service!

Funding considerations

- Funding issues need to be discussed at the outset
- VA and Medicaid typically covers 100%
- Medicare
  - Covers 80% (*However, can apply for Gleason to cover 20% co-pay*)
  - Medicare will NOT cover an SGD once an individual is in Hospice care or in a SNF
  - Medicare requires a dedicated SGD (but can be unlocked after received for an out-of-pocket expense of $25)
  - Medicare 5-year rule
- Eye gaze access
  - Have to rule out all other access options, even if eye gaze access is inevitable and/or more desirable due to efficiency issue

Text-to-speech

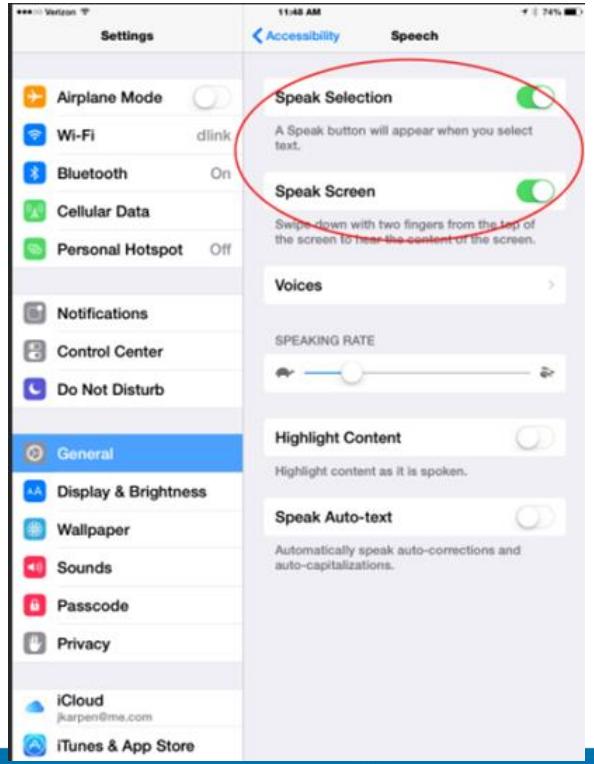
App		Price	Platform	Etc.
Vocable		Free	ios and Android	Headtracking
Jabberwocky		Free	ios and Andriod	Headtracking
TD talk Tobii		Free	ios and Android	Can save messages
Speech assistant		$14.99	ios and Andriod	Can save messages
Dialogue (Essence) PRC		$99.99	ios and Android	Can save messages

Loaning options:

- Lending Libraries:
  - Gleason Foundation
  - ALSA through our local chapter (depending on where patient lives)
    - For ex: We can borrow out SGDs to patients living in Columbus/Cincinnati area. If they live north of Mansfield they go through Northern Ohio chapter.
  - Pending availability of device
  - Companies also may provide devices to trial

Accessibility options on ipads

- Accessibility options on ipads
- –“Speak” option on ipads



Access Options

- Direct selection (hands, keyboard, stylus)
- Modifications include:
  - thick barrel pen if grip is reduced
  - increasing cursor speed of mouse so less movement is required
  - eliminating the need to double click on items to select them
- Many people with ALS find using a mouse with an onscreen keyboard rather than touch/typing can enable them to maintain use of computer, tablet or SGD.

Alternative Access (AA)

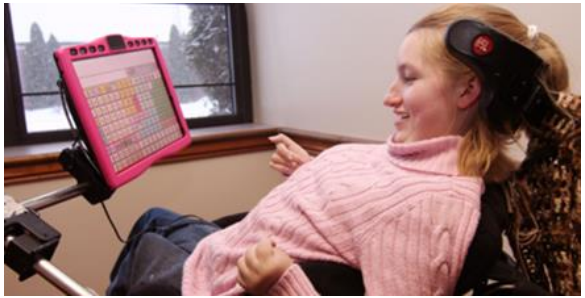
- Refers to the tools and strategies that enable people with limited hand or limb movement to continue to use their SGDs and other home modifications.
  - Tablets
  - Alerting call chimes
  - Lights
  - Locks
  - Fans
  - Remotes
  - Blinds
  - Eye drive (available on Tobii I series and newest 2020 PRC Accent devices)

Alternative Access (AA)

- Alternative Access is important for people with ALS who may lose standard ways of representing their ideas like handwriting or keyboard typing.
- For those with very weak or no arm movement, AA equipment can enable a person to operate and SGD or tablet with head, foot, or eye movement.

Alternative Access

- Head tracking
  - “Dot” on windows
  - TruDepth camera on apple



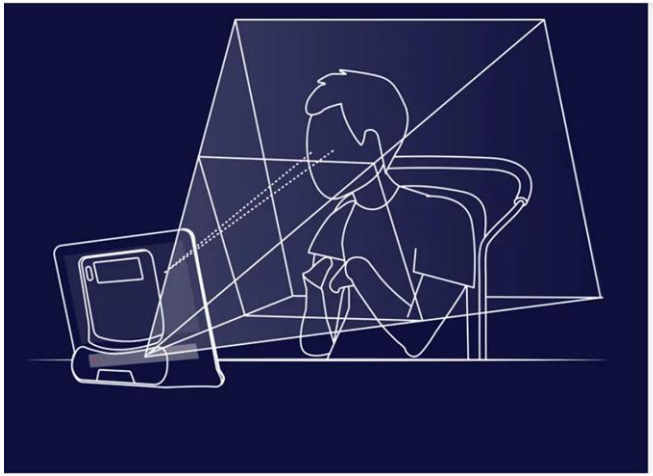
Alternative Access

- Switches
  - Typical button
  - Finger switch

Consider the changes over time pALS may experience.

Alternative Access

- Eye tracking



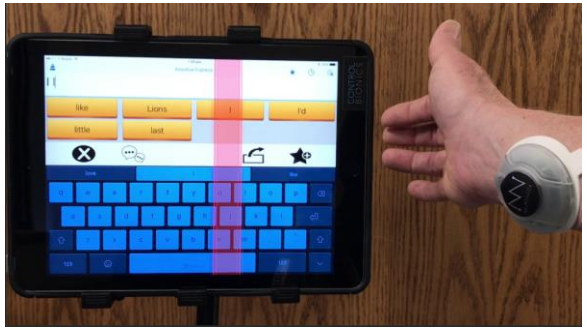
Steps

1. The eye tracker sends out near infrared light.
2. The light is reflected in your eyes.
3. Those reflections are picked up by the eye tracker's cameras.
4. Through filtering and calculations, the eye tracker knows where you are looking.

As simple as the concept may be, an enormous amount of research and development has gone into Tobii Dynavox eye trackers to make them work in a simple and unobtrusive way.

Alternative Access

- Wearable EMG control device
  - Being developed by multiple companies



Overall considerations

- Upfront with timeline and cost expectations
- Plant the seed early and develop buy-in
- Tech comfortability
  - Apple vs. Windows
- Targeting their participation
- Ongoing support and follow up
  - Outpatient to home health collaboration
  - Support from the SGD company

7 VITAL COMMUNICATION COMPETENCIES AND PARTNERING WITH YOUR SLP

- As abilities change, the SLP will continue to partner with patient and family to maintain communication.
- The SLP will make sure to have the tools and training to achieve these 7 communication competencies throughout the progression of ALS.

- 1) I can alert people in other rooms or outside the home when I have a need or emergency
- 2) My communication partners and I can use communication strategies that improve communication speech and success and reduce fatigue (using speech or AAC)
- 3) I can use a low-tech or rapid access communication system

- 4) I can produce voiced messages via speech, SGD or TTS app
- 5) I can communicate with people not in my immediate environment (email, text, phone, social media, etc)
- 6) My communication partners and I can independently set up, customize and use all of the elements of my communication system
- 7) I can describe a pro-active strategy designed to prepare for typical changes in speech and/or computer access that I may experience

- IT IS ESSENTIAL THAT THE PATIENT UNDERSTANDS THE ABILITIES THAT CAN BE EXPECTED TO BE MAINTAINED AND ALERT THEIR SLP WHEN UPDATES IN THEIR SYSTEM ARE NEEDED

Thank You!!!

Questions/Concerns? Contact me.

Michelle.Longmuir-Wilkes@OhioHealth.com
614-896-1442

Sara.Bott@Ohiohealth.com
614-441-1000

References

- Reason for Hope: ALS Resource Guide by Central & Southern Ohio Chapter
- Speech and Swallowing Issues with ALS by Massachusetts General Hospital; ALS Multidisciplinary Clinic 1/3/18
- Suggestions and information about Speech Changes and Augmentative Alternative Communication (AAC) by Amy Roman, MS, CCC/SLP, 1/3/18
- Senthilkumar J, et al. Hospice: rehabilitation in reverse. *Indian J Palliat Care*. 2010;16(3): 111–116.


BELIEVE IN WE™
OhioHealth