Strategies to Mitigate Speech and Swallowing Impairments in Ataxia

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PARTNERING FOR PROGRESS
2023 Annual Ataxia Conference
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Las Vegas, NV

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2023 Annual Ataxia Conference
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PRESENTER DISCLOSURES

Allison Hilger, PhD, CCC-SLP is employed by the University of Colorado Boulder where she receives a salary. She received NIH funding to conduct research on speech in ataxia in 2019, which funded many of the studies she will talk about today.

She has no non-financial disclosures to share.

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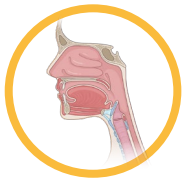
Outline



Speech and swallowing difficulties in ataxia

Overview of speech/swallowing anatomy and physiology

Current theories of causes for speech and swallowing impairments in ataxia

Speech and swallowing therapy options

At-home strategies

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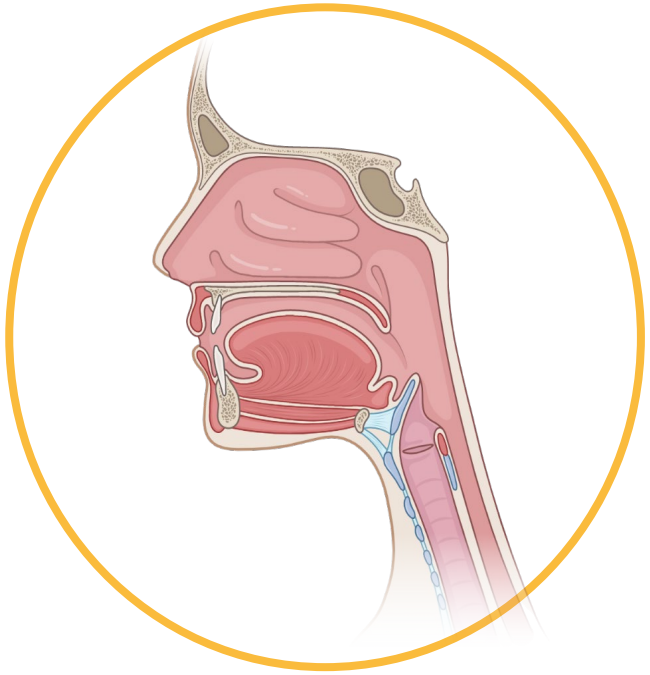
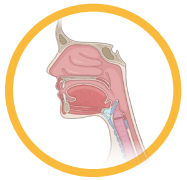


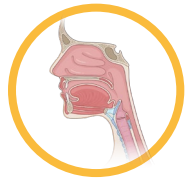
Speech and swallowing difficulties in ataxia

What we currently know



- There are **general speech and swallowing difficulties** that arise from cerebellar damage
  - **<u>Speech difficulties:</u>**
    - Inconsistent articulatory errors
    - Variable pitch and loudness
    - Reduced speech naturalness
    - Slowed rate of speech
    - Occasionally hoarse/breathy voice
  - **<u>Swallowing difficulties</u>**
    - Sometimes food/liquid penetrates the airway but is coughed out
    - Sometimes food/liquid is aspirated, meaning it is drawn into the lungs
      - This can cause aspiration pneumonia
    - Sometimes food/liquid is regurgitated nasally

Generally, all of these difficulties are caused because movements are no longer coordinated from cerebellar damage



What we currently know

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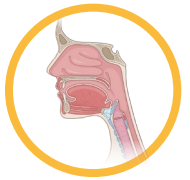
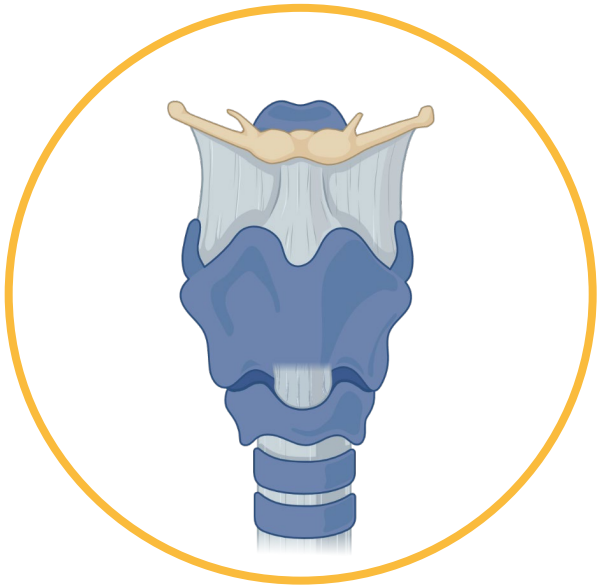
We'll walk through these terms in more detail



What we currently do not know

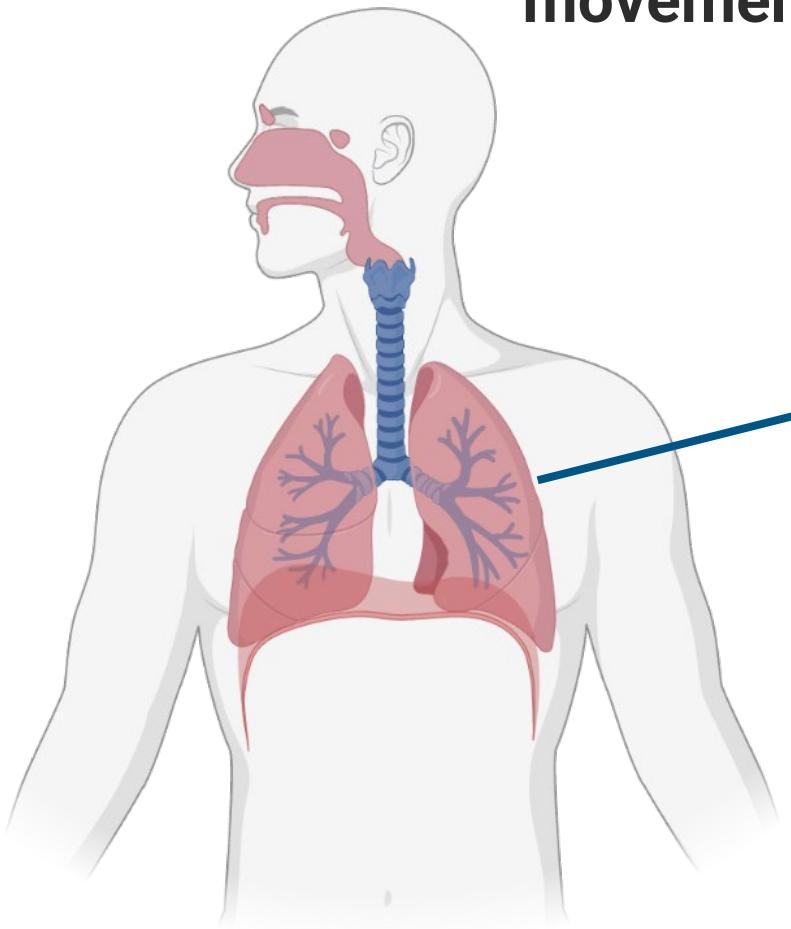
- How speech and swallowing difficulties **change by ataxia etiology and SCA subtype**
  - Some **SCA subtypes** present with different speech/swallowing characteristics than others
  - Some **ataxia etiologies** (e.g., Friedreich's Ataxia, episodic ataxia, gluten ataxia, etc) present with different speech/swallowing characteristics than others
- How speech and swallowing difficulties change by extent and location of cerebellar damage

Let’s back up…



Overview of speech/swallowing anatomy and physiology

Speech and swallowing are highly complex movements that require precise coordination



Respiration

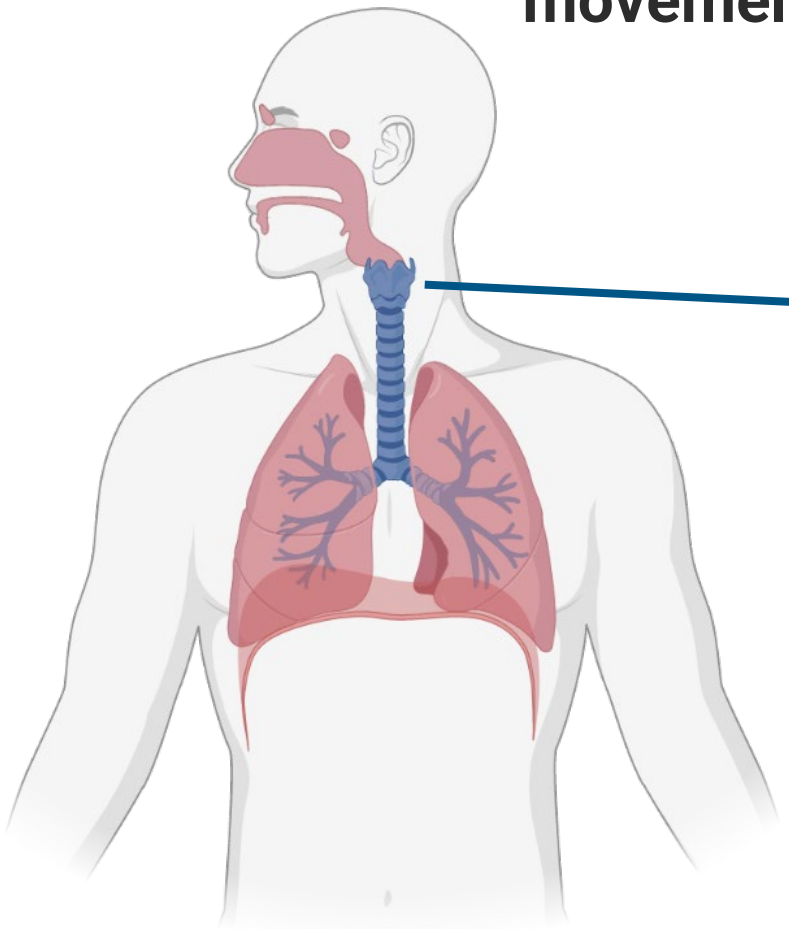
For speech:

- Driving force of speech
- We talk on the exhale
- How much we inhale before we talk influences how well we control pitch/loudness of speech and how fatiguing it is to talk

For swallowing:

- Swallowing must be coordinated with the respiratory cycle to prevent aspiration
- Best to swallow while exhaling

Speech and swallowing are highly complex movements that require precise coordination



Larynx

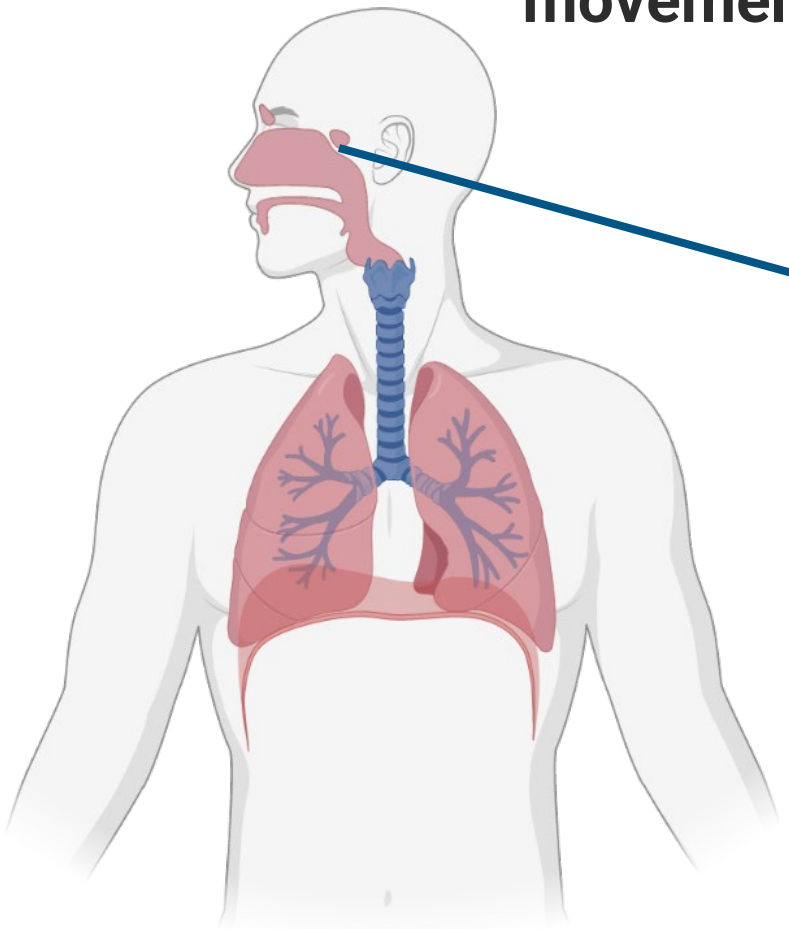
For speech:

- The location of our vocal folds which vibrate to produce our voice
- We can change the shape, length, and tension of our vocal folds to change our pitch, loudness, and vocal quality

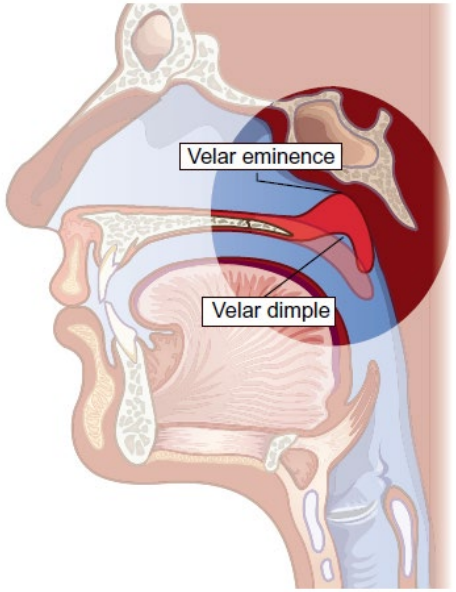
For swallowing:

- The larynx should be closed off and the vocal folds should be tightly closed while swallowing to prevent aspiration

Speech and swallowing are highly complex movements that require precise coordination



Nasal cavity



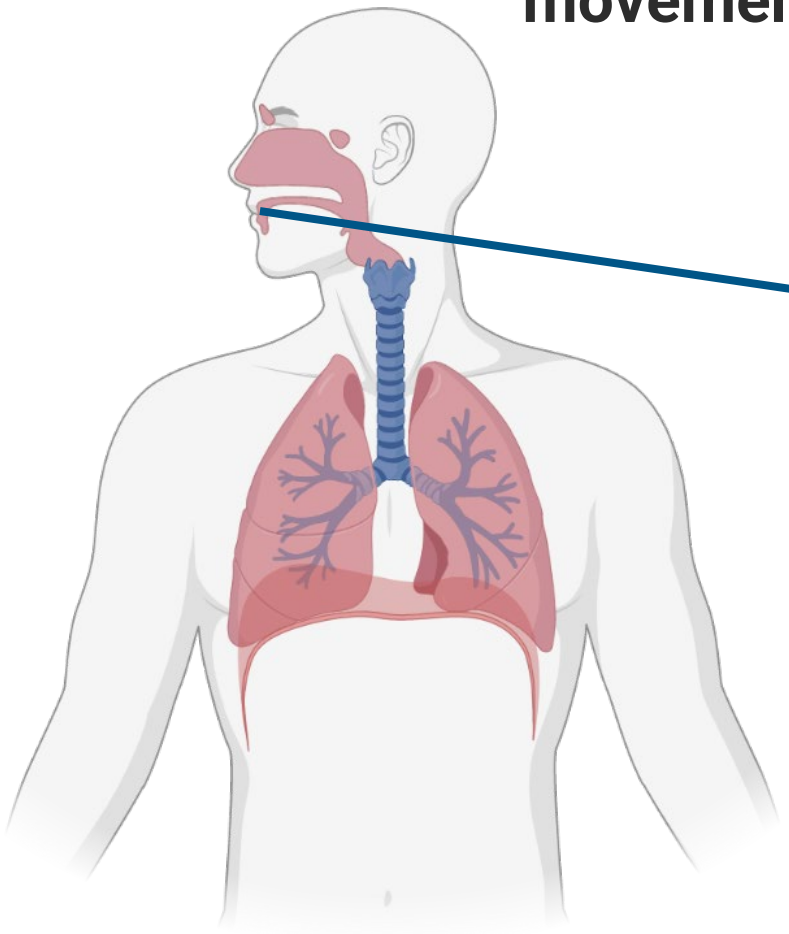
For speech:

- We can open and close our nasal cavity for nasal sounds (“m” and “n”) or non-nasal sounds
- If the nasal cavity is open when it shouldn’t be, you will sound hyper-nasal

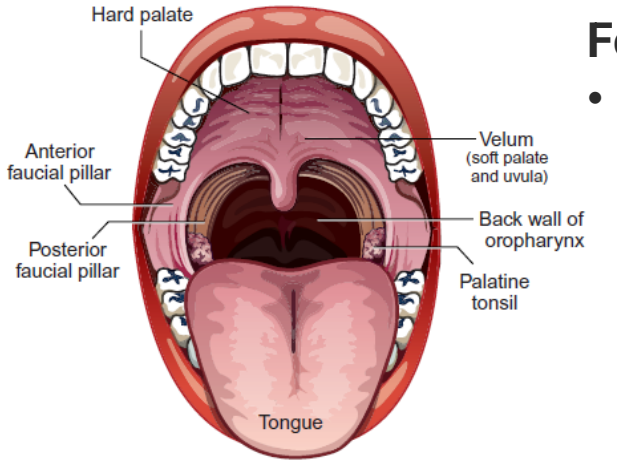
For swallowing:

- The nasal cavity should be closed off while swallowing
- If it is open, food and liquid can be regurgitated and come out of the nose

Speech and swallowing are highly complex movements that require precise coordination



Mouth



For speech:

- We articulate speech sounds by moving our tongue and lips to articulate with our teeth, hard palate, and soft palate

For swallowing:

- We chew up food and liquid to prepare for swallowing

Key terms

- Articulation
- Prosody
- Voice quality
- Resonance
- Intelligibility
- Naturalness

- Penetration
- Aspiration

Key terms

- Articulation
- Prosody
- Voice quality
- Resonance
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- Naturalness

Dysarthria = clinical term for speech impairment from neurological disorder, specifically impairment in the execution and production of speech

- Penetration
- Aspiration

In ataxia, the specific term is *ataxic dysarthria.*

Key terms

- **Articulation**
  - Using our tongue, lips, teeth, and roof of the mouth to “articulate” or create speech sounds
  - Example: putting our lips together to say “p” or “b”
- Prosody
- Voice quality
- Resonance
- Intelligibility
- Naturalness

- Penetration
- Aspiration

In ataxia:

- Movements of tongue, lips, teeth, and roof of the mouth are uncoordinated
- Results **inconsistent articulatory errors**
- This means that sometimes you produce an “r,” for example, correctly and sometimes incorrectly
- There is nothing wrong with the structures of the mouth, but rather the coordination among them is impaired
- Articulation becomes harder with longer words and with fatigue

Key terms

- Articulation
- **Prosody**
  - Changing pitch, loudness, and timing
  - Express emotion
  - Ask a question versus make a statement
  - We change prosody by modifying our breathing patterns and vocal fold movements
- Voice quality
- Resonance
- Intelligibility
- Naturalness

- Penetration
- Aspiration

In ataxia:

- Coordination of breathing patterns and vocal fold movements is impaired
- Results **variable prosody**
  - Loudness bursts when talking
  - Pitch that is too high or too low
  - Timing of words is not correct
  - Difficult to express emotion

Key terms

- Articulation
- Prosody
- **Voice quality**
  - How “clear” our voice sounds
  - Hoarse, breathy, raspy, clear, etc.
  - We control voice quality by how open or closed our vocal folds are when talking
- Resonance
- Intelligibility
- Naturalness

- Penetration
- Aspiration

In ataxia:

- Coordination of breathing patterns and vocal fold movements is impaired
- Results **variable voice quality**
  - For some people, their voice is usually/always hoarse or breathy
  - For other people, they may have breaks in voicing where they have a clear voice one second and then a hoarse voice the next second

Key terms

- Articulation
- Prosody
- Voice quality
- **Resonance**
  - How much air goes through our nose while talking
  - Air should go through our nose when making “m” and “n” sounds but not other sounds
  - If too much air goes through the nose, you will sound hypernasal
- Intelligibility
- Naturalness

- Penetration
- Aspiration

In ataxia:

- Coordination of the velum to close off the nasal cavity is impaired
- Results **variable nasality**
  - Sometimes people will sound nasal when talking and sometimes they won’t
  - Will likely worsen with longer words and fatigue

Key terms

- Articulation
- Prosody
- Voice quality
- Resonance
- **Intelligibility**
  - How many words can be understood from your speech
  - Usually measured as a percentage
  - “Patient X has 80% intelligibility. I can understand 80% of their speech”
- Naturalness
- Penetration
- Aspiration

In ataxia:

- Intelligibility is usually high despite the speech difficulties (>70%) (Hilger, Fahey, & Cloud, 2022)
- People with ataxia are generally understandable even when the speech impairment is more severe

Key terms

- Articulation
- Prosody
- Voice quality
- Resonance
- Intelligibility
- **Naturalness**
  - How natural or unnatural the speech sounds
  - Rated on a scale from normal, mild, moderate, to severe
- Penetration
- Aspiration

In ataxia:

- Naturalness is usually quite impaired (Hilger, Fahey, & Cloud, 2022)
- Likely because prosody is disrupted

Key terms

- Articulation
- Prosody
- Voice quality
- Resonance
- Intelligibility
- Naturalness

Dysarthria = clinical term for speech impairment from neurological disorder, specifically impairment in the execution and production of speech

- Penetration
- Aspiration

Key terms

- Articulation
- Prosody
- Voice quality
- Resonance
- Intelligibility
- Naturalness

- Penetration
- Aspiration

Dysphagia = clinical term for swallowing impairment

Key terms

- Articulation
- Prosody
- Voice quality
- Resonance
- Intelligibility
- Naturalness

In ataxia:

- Incoordination causes the airway to stay open sometimes while swallowing
- Resulting in food/liquid entering the airway
- Likely to happen when fatigued or distracted

- **Penetration**
  - When food/liquid enters the airway but is coughed out
  - A sign that a person could aspirate
  - Likely feels like a choking sensation that causes you to cough
- Aspiration

Key terms

- Articulation
- Prosody
- Voice quality
- Resonance
- Intelligibility
- Naturalness

- Penetration
- **Aspiration**
  - When food/liquid enters the airway and travels to the lungs
  - Can cause infection in the lungs AKA aspiration pneumonia

In ataxia:

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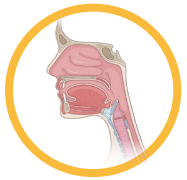
What we currently know



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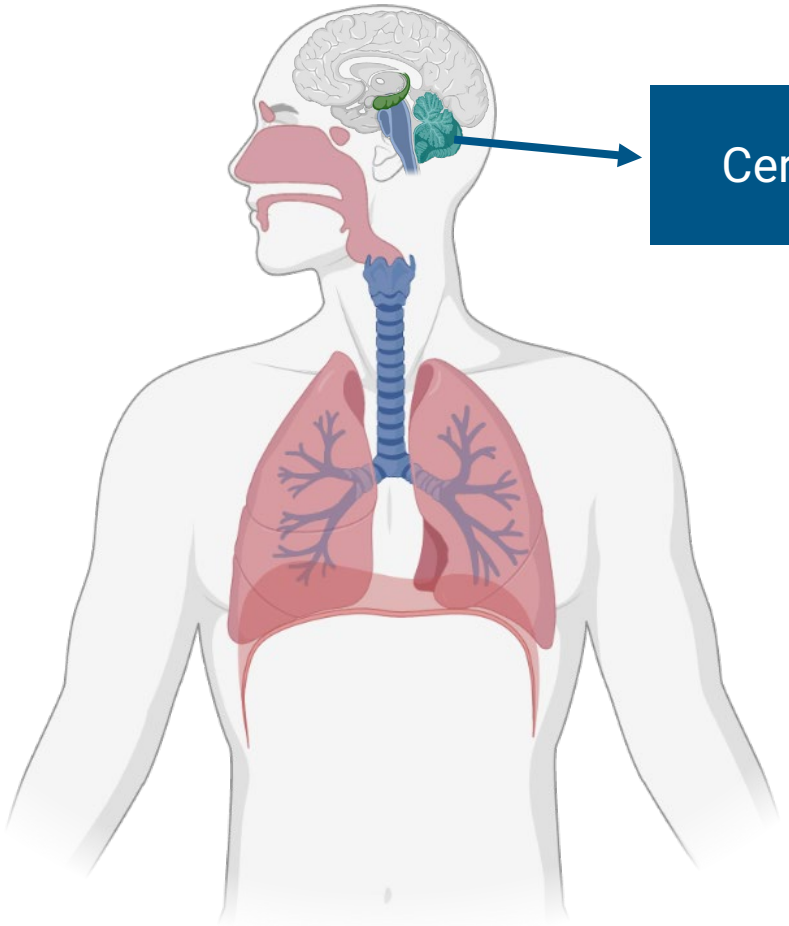
Let's walk through these terms now that we are familiar with them

Current theories of causes for speech and swallowing impairments in ataxia

Theory #1. Cerebellar Incoordination of muscles

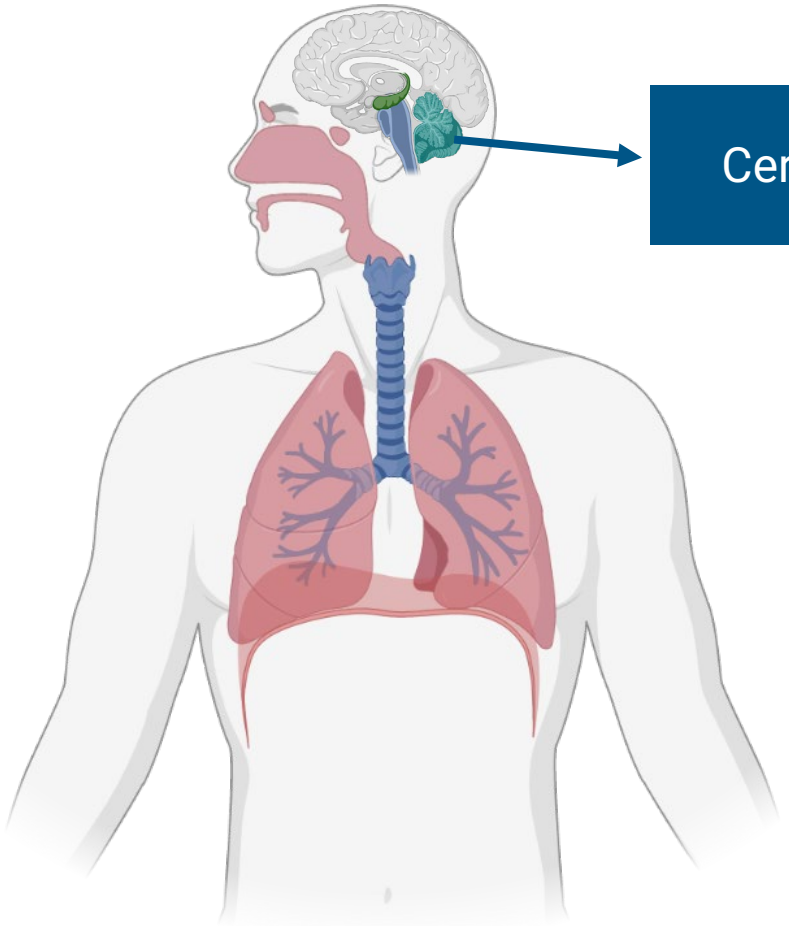


The cerebellum coordinates and sequences movements across the mouth, nose, larynx, and lungs for speech and swallowing.

If there is cerebellar damage, these movements will become uncoordinated:

- Movements of the mouth for articulation
- Vocal fold movements for pitch and loudness
- Breathing patterns for speech
- Closing off the airway for swallowing
- Sequencing the order of muscular activation for swallowing

Theory #2. Cerebellar distortion of sensory feedback



The cerebellum receives sensory feedback from the sensory organs (hearing, taste, feeling, smell, vision) and integrates that information with current movement.

It says, “I feel you that sticky peanut butter you are swallowing didn’t get all the way down so you should swallow again”

Or, “You voice sounds louder than you planned so you should talk more quietly”

If there is cerebellar damage, it won’t be able to accurately integrate this sensory information.

Causes of speech/swallowing difficulties in ataxia

Both theories are supported and complementary, meaning that there are two main cerebellar roles for speech and swallowing that are disrupted:

1. Coordinating and sequencing muscular movement
2. Integrating sensory feedback into current movement

So… what we can do about it?

Speech and swallowing therapy options

Speech Therapy Options

Speech therapy goals should be **individualized** to your specific difficulties.

The goals should be based on where in the speech mechanism is being **most disruptive to your speech**:

- Breath control
- Hypernasality
- Voice quality
- Articulation
- Pitch and loudness control

Speech Therapy Options

However, there are some more **standardized treatment protocols** that may be effective:

- Lee Silverman Voice Treatment (LSVT)
- Breath control techniques
- Melbourne Ataxia Speech Treatment
- Alternative and Augmentative Communication (AAC) Devices

Speech Therapy Options

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Speech Therapy Options

Lee Silverman Voice Treatment (LSVT)

- Developed by Cynthia Fox, Lorraine Ramig, and Shimon Sapir in the early 2000's as a speech treatment for Parkinson's Disease
- **Main concepts:**
  1. Very intensive: 16 one-hour treatment sessions across four consecutive days per week for four weeks
  2. Training to speak LOUDLY
     - Idea is the focusing solely on vocal loudness will improve vocal quality and intelligibility

Speech Therapy Options

Lee Silverman Voice Treatment (LSVT)

- Is it effective in ataxia?
  - Sapir et al (2003): case study of a woman with cerebellar dysfunction secondary to thiamine deficiency
    - **Improved voice quality, articulation, and intelligibility**
  - Lowit, Egan, & Hadjivassiliou (2020): rater-blinded, single-arm study in 18 people with FA, 1 person with SCA6, 1 person with idiopathic cerebellar ataxia, and 1 person with spastic paraplegia
    - **Improved voice quality**
    - **No changes in intelligibility and speech naturalness**

Speech Therapy Options

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    - **Improved voice quality**
    - **No changes in intelligibility and speech naturalness**

Jury is still out. LSVT may be effective for some types of ataxia, but it doesn't appear to be effective for all types.

Speech Therapy Options

Lee Silverman Voice Treatment (LSVT)

- **Pros:** straightforward treatment protocol that is insurance-approved
- **Cons:** limited evidence for being effective in ataxia and very time intensive

Speech Therapy Options

Standardized treatment protocols:

- Lee Silverman Voice Treatment (LSVT)
- **Breath control techniques**
- Melbourne Ataxia Speech Treatment
- Alternative and Augmentative Communication (AAC) Devices

Speech Therapy Options

Breath control techniques

- There have not been studies on breath control and speech outcomes yet (but stay tuned with my lab research!)
- However, it is very likely that focusing on **breath control will improve speech intelligibility and naturalness**
  - Better respiratory support makes it easier to control pitch and loudness, improves vocal quality, and paces the rate of speech better

Speech Therapy Options

Breath control techniques

- Typical goals in speech therapy for breath control:
  - Inhaling to an appropriate lung volume before speaking
  - Taking a breath at an appropriate location while speaking (at the end of sentence and not the middle of a word)
  - Increasing the number of words produced per breath
  - Taking a breath before running out of air
- The advantage of this therapy technique is that **you only focus on breath control,** and you will likely see generalized improvement to other aspects of speech (voice quality, prosody, intelligibility, etc)

Speech Therapy Options

Standardized treatment protocols:

- Lee Silverman Voice Treatment (LSVT)
- Breath control techniques
- **Melbourne Ataxia Speech Treatment**
- Alternative and Augmentative Communication (AAC) Devices

Speech Therapy Options

Melbourne Ataxia Speech Treatment

Developed by Dr. Adam Vogel, a behavioral neuroscientist at the University of Melbourne, Australia

- **Patients spend 45 minutes/day for four weeks completing a computer program exercise at home:**
  - Saying words and sentences, and reading a passage, and doing pitch and loudness control exercises
  - Provided with audio and visual feedback of results of duration, pitch, and loudness variation
  - Goal to improve intelligibility, vocal control, and prosody

Speech Therapy Options

Melbourne Ataxia Speech Treatment

- Is it effective?
  - Vogel et al. (2019): seven patients with autosomal recessive spastic ataxia of Charlevoix-Saguenay (ARSACS)
    - **Improved intelligibility and enhanced naturalness!**
  - Vogel et al. (2022): 16 patients with SCA 1, 2, 3, 4, or 6
    - **Improved intelligibility in 75% of the participants**
    - **Better pitch and loudness control**
    - **Clearer vocal quality**

- **Pros:** convenient, at-home therapy tailored to ataxia that has promising preliminary results for effectiveness
- **Cons:** currently developed for Australians and likely not beneficial for an American accent yet
- **Overall… this is a therapy tool to keep an eye on for accessing in the US in the future!**

Speech Therapy Options

Standardized treatment protocols:

- Lee Silverman Voice Treatment (LSVT)
- Breath control techniques
- Melbourne Ataxia Speech Treatment
- **Alternative and Augmentative Communication (AAC) Devices**

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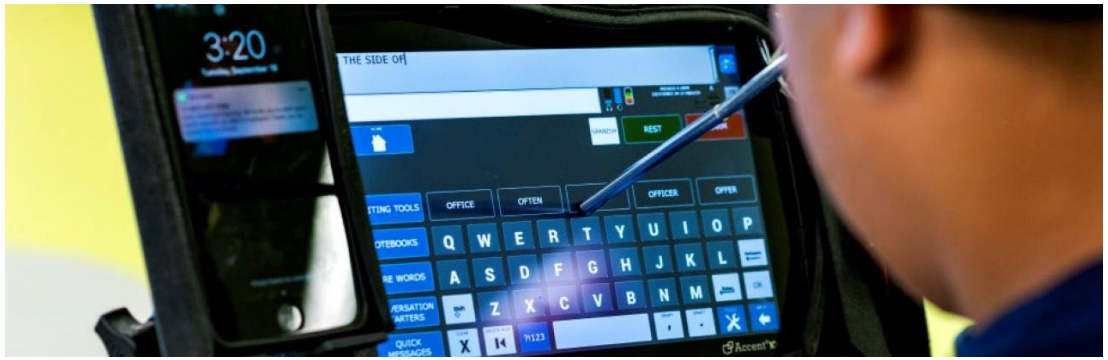
Speech Therapy Options

Alternative and Augmentative Communication (AAC) Devices

- When speech intelligibility is severely compromised, and/or speech is highly fatiguing, an AAC device can be used
- Types of AAC devices:
  - Electronic, speech generating device
    - Typically on a tablet or iPad
    - Can use a keyboard or picture symbols
    - Often funded by insurance
    - Speaker's voice can be modified



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Voice banking: creating a synthetic voice using your own voice, made by recording a large number of messages and using then using AI.

Many companies provide this service:
https://www.thevoicekeeper.com/
https://www.prc-saltillo.com/assets/uploads/PRC-Saltillo-MessageVoiceBank_091721_KCM.pdf
https://www.talktometechnologies.com/pages/voice-and-message-banking

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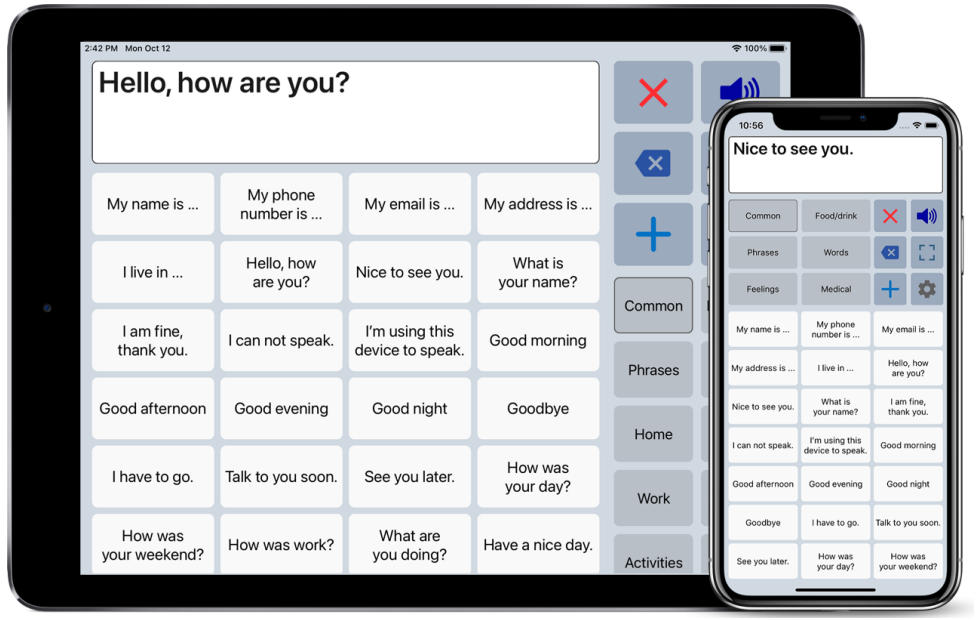
    - **Speaker's voice can be modified**
  - iPhone apps:
    - TouchChat
    - Proloquo2Go
    - QuickTalk AAC
    - iCommunicate for iPad
    - SonoFlex



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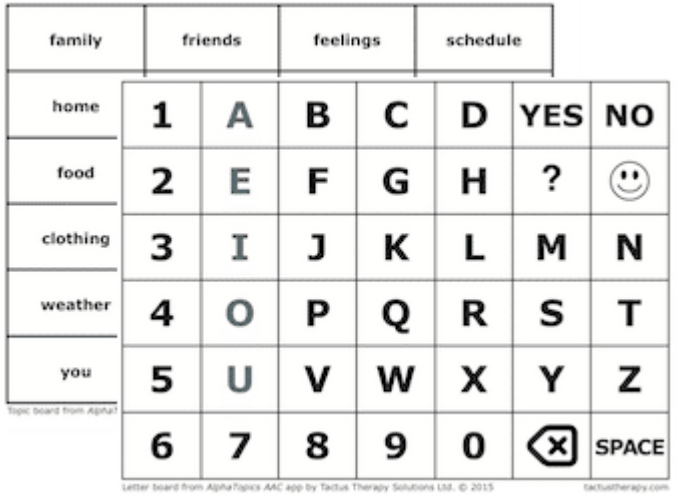

Speech Therapy Options

Standardized treatment protocols:

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Swallowing Therapy

Swallowing Therapy Options

No standard protocol for treating dysphagia in ataxia

- Goals should be individualized based on specific difficulties
- **Likely goals include:**
  - Modifying thickness of liquids to slow the swallow down for better coordination
  - Improving swallow strength and speed
  - Improving cough strength
  - Finding strategies that are effective for you (but may not be effective for everyone)
    - Chin tuck while swallowing
    - Using a straw
    - Swallowing twice each bite

What can I do at home?

At-Home Strategies

At-home speech strategies

Think of these strategies as a toolbox where you can pull out a tool that works for you when you want to use it. It will likely be tiring to use these strategies all day, all the time. Instead, you can use these strategies when you want to speak more clearly.

- Posture
- Breath control
- Overarticulation
- Slowing down (but take caution- not always effective!)

At-home speech strategies

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- Posture
  - Sitting upright makes it easier to use good breath support for speech
  - Improve vocal quality and pitch and loudness control
  - Likely enhanced speech naturalness and intelligibility
- Breath control
- Overarticulation
- Slowing down (but take caution- not always effective!)

At-home speech strategies

Think of these strategies as a toolbox where you can pull out a tool that works for you when you want to use it. It will likely be tiring to use these strategies all day, all the time. Instead, you can use these strategies when you want to speak more clearly.

- Posture
- Breath control
  - Focus on taking a solid breath before speaking (but not too large of a breath)
  - Try to say 5-10 words per breath
  - Take a breath between sentences
- Overarticulation
- Slowing down (but take caution- not always effective!)

At-home speech strategies

Think of these strategies as a toolbox where you can pull out a tool that works for you when you want to use it. It will likely be tiring to use these strategies all day, all the time. Instead, you can use these strategies when you want to speak more clearly.

- Posture
- Breath control
- Overarticulation
  - Cue yourself to exaggerate your articulation
  - Will likely be very fatiguing
  - Good tool to use for special circumstances (an important phone call, for example)
- Slowing down (but take caution- not always effective!)

At-home speech strategies

Think of these strategies as a toolbox where you can pull out a tool that works for you when you want to use it. It will likely be tiring to use these strategies all day, all the time. Instead, you can use these strategies when you want to speak more clearly.

- Posture
- Breath control
- Overarticulation
- Slowing down (but take caution- not always effective!)
  - Some people find that slowing down helps them articulate better
  - Other people find that slowing down is exhausting and not helpful

At-home swallow strategies

- Swallow strongly
- Swallow twice/bite
- Sit upright when eating and drinking
- Take small bites
- Focus on exhaling when swallowing
- Eat without distractions
- If you feel a choking sensation, use a strong cough

At-home swallow strategies

- Swallow strongly
  - Exaggerate your swallow and try to swallow strongly
  - Will help speed up your swallow and push the food/liquid past the airway
- Swallow twice/bite
- Sit upright when eating and drinking
- Take small bites
- Focus on exhaling when swallowing
- Eat without distractions
- If you feel a choking sensation, use a strong cough

At-home swallow strategies

- Swallow strongly
- Swallow twice/bite
  - Often, residue from food/liquid is left behind after a swallow
  - Swallowing twice will help clear that residue
- Sit upright when eating and drinking
- Take small bites
- Focus on exhaling when swallowing
- Eat without distractions
- If you feel a choking sensation, use a strong cough

At-home swallow strategies

- Swallow strongly
- Swallow twice/bite
- Sit upright when eating and drinking
- Take small bites
- Focus on exhaling when swallowing
- Eat without distractions
- If you feel a choking sensation, use a strong cough

Use gravity to help push the food/liquid past the airway into the pharynx

At-home swallow strategies

- Swallow strongly
- Swallow twice/bite
- Sit upright when eating and drinking
- Take small bites — Smaller bites are easier to control and coordinate during the swallow
- Focus on exhaling when swallowing
- Eat without distractions
- If you feel a choking sensation, use a strong cough

At-home swallow strategies

- Swallow strongly
- Swallow twice/bite
- Sit upright when eating and drinking
- Take small bites
- Focus on exhaling when swallowing
- Eat without distractions
- If you feel a choking sensation, use a strong cough

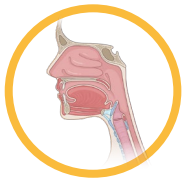
Will help close the larynx off during the swallow and prevent food/liquid from being drawn into the airway

At-home swallow strategies

- Swallow strongly
- Swallow twice/bite
- Sit upright when eating and drinking
- Take small bites
- Focus on exhaling when swallowing
- Eat without distractions
- If you feel a choking sensation, use a strong cough

Distractions while eating/drinking can make it harder to coordinate the swallow

Outline



Speech and swallowing difficulties in ataxia

Overview of speech/swallowing anatomy and physiology

Current theories of causes for speech and swallowing impairments in ataxia

Speech and swallowing therapy options

At-home strategies

NAF National Ataxia Foundation

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Any Questions?