Cerebellar Injury, Speech, & Quality of Life

Considerations for Comprehensive Care

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DISCLOSURES

Allison Hilger, PhD, CCC-SLP is employed by the University of Colorado Boulder. She received funding through the NIH and the Council of Academic Programs in Communication Sciences and Disorders to research speech impairments in ataxia. She has no nonfinancial relationships to disclose.

Caitlin Cloud, MA, CF-SLP, PCBIS recently graduated from the University of Colorado Boulder and worked with Dr. Hilger in the CO Motor Speech Lab. She has no nonfinancial relationships to disclose.

AGENDA

	Part 1	Ataxia: An introduction to the signs of cerebellar injury
	Part 2	Speech impairment in ataxia & quality of life
	Part 3	The role of speech therapy in rehabilitation
	Part 4	The big picture: Comprehensive, person-centered care
	Part 5	Q & A

LEARNING OBJECTIVES

Participants will be able to:

1. Describe the etiologies and clinical features of cerebellar ataxia
2. Identify speech characteristics associated with cerebellar ataxia
3. Summarize the effects that speech impairment may have on quality of life in persons with ataxia
4. Synthesize perceptual judgments of speech characteristics and quality of life measures in order to refer clients with ataxia for speech evaluation and intervention, as appropriate

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THE CEREBELLUM

What do you already know about the cerebellum and its functions?

THE CEREBELLUM

- Involved in the sequencing, timing, scaling, and coordination of movements
- Combines learned movements together with precise timing to produce skilled motor behavior
- Integrates sensory feedback with motor plans to adapt to specific contextual demands

Predict: What consequences could arise from cerebellar damage?

WHAT IS ATAXIA?

Definition	• A cluster of clinical signs/symptoms resulting from damage or disruption of the cerebellum • May affect gait, balance, eye and limb movements, and speech
Etiologies	• Hereditary degenerative conditions • Variety of vascular, traumatic, demyelinating, neoplastic, toxic-metabolic etiologies
Characteristics	• Incoordination/imprecision in the force, range, timing, and direction of movement • Reduced speed and fluidity of movement

WHY DO WE NEED TO TALK ABOUT ATAXIA?

(1) Impact on quality of life

"I sound like I'm drunk."
"People ask me if I've been drinking. I try to avoid speaking if I can."

(2) Relatively poor understanding of ataxia among healthcare professionals

"My doctor said that there isn't anything he can do for me."
"I was worried about my speech, but my doctor never referred me for speech therapy."
"My speech therapist had never heard of ataxia."

(3) Ataxia is under-studied in the rehabilitation literature

"Despite the widespread use of allied health care interventions in cerebellar ataxia, there is a lack of good quality studies that have evaluated such interventions. We found some support for the implementation of physical therapy and occupational therapy, but more research is needed to develop recommendations for clinical practice." (Fonteyn et al., 2014)

WHY DO WE NEED TO TALK ABOUT ATAXIA?

(4) **The etiology of ataxia can significantly impact plan of care.** Healthcare professionals should be familiar with the etiologies of ataxia, including the hereditary progressive ataxias – even if they typically treat acquired conditions (e.g. stroke, TBI)

For example: A patient arrives in your clinic for treatment after sustaining a TBI due to a fall. While completing a case history, you discover the patient had symptoms of ataxia prior to the head injury. What do you do next?

(5) **Ataxia may occur in isolation or in conjunction with many other symptoms, depending on the etiology and characteristics of the brain injury.** Recognizing the features of ataxia is one piece of the puzzle in initiating appropriate referrals and providing effective interventions following acquired brain injury, during both acute and chronic phases of recovery.

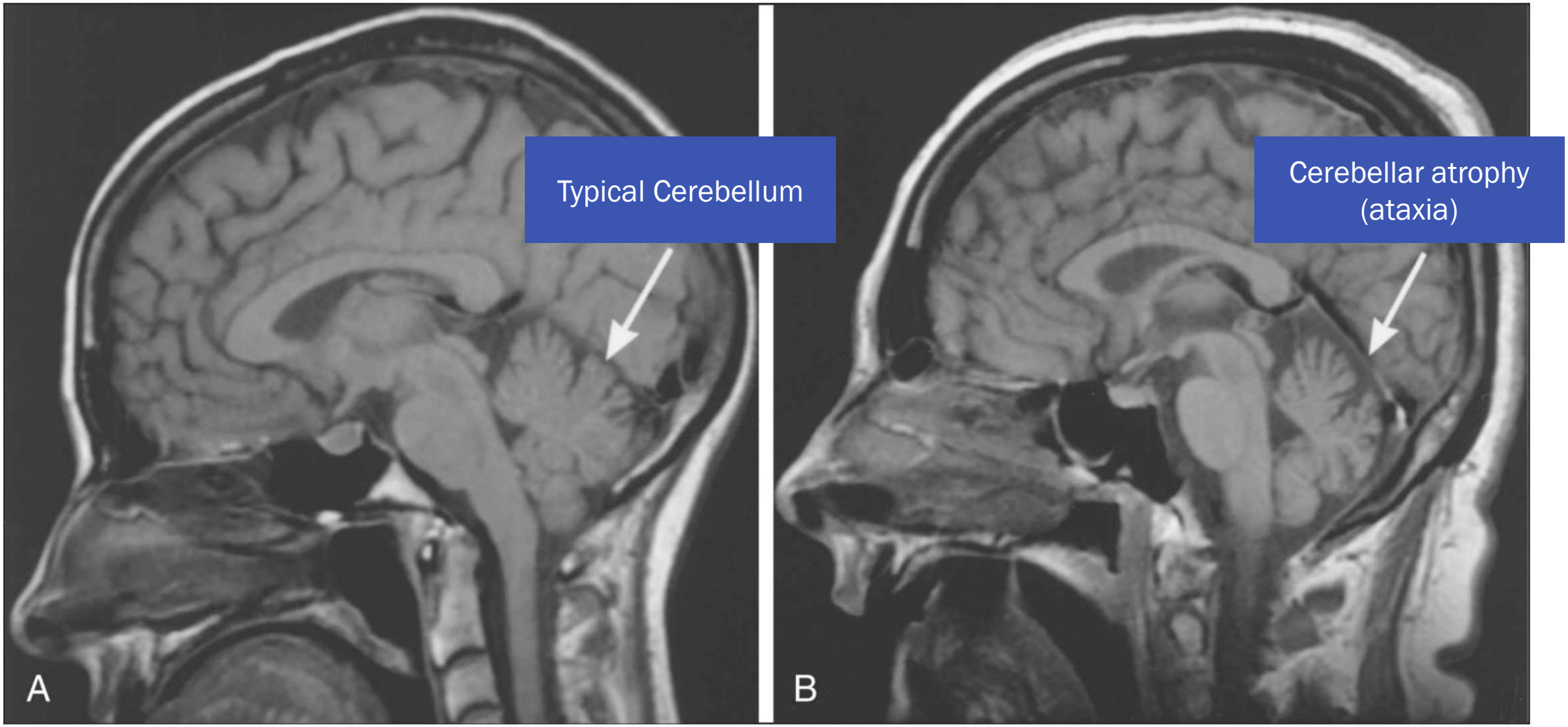
ETIOLOGIES

Degenerative diseases:

- Hereditary ataxias
  - Spinocerebellar ataxias (SCAs)
  - Friedrich's ataxia
  - Ataxia telangiectasia
  - Ataxia with isolated vitamin E deficiency
- Nonhereditary degenerative ataxias
  - Olivopontocerebellar atrophy
  - Other neurodegenerative diseases (e.g. ALS, progressive supranuclear palsy, frontotemporal dementia)

Other etiologies:

- Stroke
- Traumatic brain injury
- Postoperative
- Demyelinating conditions
- Cerebellar or brainstem tumor
- Autoimmune conditions
- Paraneoplastic cerebellar degeneration
- Toxicity (e.g., lithium, alcohol)
- Inflammatory conditions (e.g. encephalitis)
- Episodic ataxias
- Infections leading to CNS disease


Typical Cerebellum
Cerebellar atrophy (ataxia)
A
B

CHARACTERISTICS OF ATAXIA

- Gait disturbance
- Nystagmus
- Intention or terminal tremor
- Dysarthria
- Dysmetria
  - Disturbed trajectory of movement
  - Overshoot, undershoot, rebound, jerky quality
- Dyssynergia (decomposition of movement)
  - Errors in timing, speed, and fluidity of movement
  - A movement may be carried out in a series of component motions

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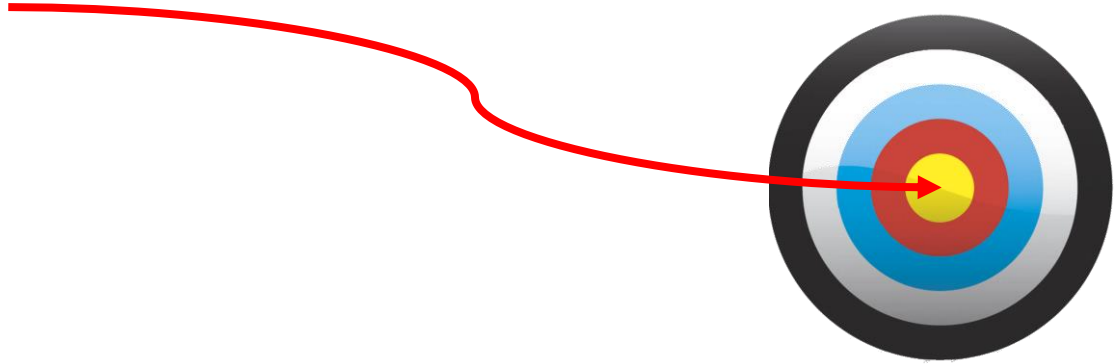

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TERMINOLOGY

Dysarthria: A neurologic speech disorder that reflects abnormalities in the strength, tone, accuracy, range, speed, and/or steadiness of movements required for respiratory, phonatory, resonatory, articulatory, and/or prosodic aspects of speech production

Intelligibility: Degree to which a listener understands the auditory signal produced by a speaker

Comprehensibility: Degree to which a listener understands a speaker based on the auditory signal plus all other information that may contribute to what has been said

Efficiency: The rate at which intelligible/comprehensible information is relayed

Naturalness: The degree to which a listener judges the speech as following normal standards of rate, rhythm, intonation, and stress patterning

A NOTE ON DYSARTHRIA

Speech is a unique, complex, dynamic motor activity

- The brain establishes an intent to produce a particular message
- The message must be converted into a code that abides by the rules of language
- The message must be organized for neuromuscular execution – this involves precise activation and coordination of respiratory and speech muscles at appropriate times, durations, and intensities

Speech allows us to express our thoughts and emotions, build relationships with others, and respond to and control our environment

- Neurologic damage or disease unmask the complex underpinnings of speech
- A motor speech disorder can significantly disrupt communicative participation and quality of life

SPEECH CHARACTERISTICS

Ataxic dysarthria may affect speech in many ways, but its characteristics are most evident in articulation and prosody

Respiration

- Initiation of utterances at low lung volumes relative to healthy speakers
- Poor timing of exhalation/phonation, resulting in air wastage

Phonation

- Harsh vocal quality
- Pitch and loudness instability

Resonance

Intermittent hyponasality

Articulation

- Vowel distortions
- Imprecise consonants
- Inconsistent, transient, and/or irregular articulatory errors
- Telescoping of syllables in multisyllabic words
- Irregular speech AMRs (rate, rhythm, precision)

Prosody

- Slow rate of speech
- Excess and equal stress
- Reduced or excessive pitch/loudness variation
- Atypical phoneme and pause durations
- Syllable durations may reflect either inflexibility or instability

SPEECH SAMPLES

Sample A

Sample B

Sample C

Sample D

How would you describe the speech in these audio samples?

QUALITY OF LIFE

Individuals with ataxia are often very **intelligible** (easy to understand).

However, the **naturalness** and **efficiency** of their speech is impaired.

From the patient's perspective:

- Patients often report that their speech sounds "slurred" or "drunk"
- They experience negative stigma due to their speech characteristics, especially when paired with other symptoms (e.g., gait disturbance)
- Patients may avoid speaking in certain situations or with certain communication partners due to the negative perceptions of others
- Even a mild speech impairment can have significant impacts on daily participation

OUR RESEARCH

Participants: 27 individuals with ataxia of various etiologies

- All participants had at least a mild form of dysarthria
- All participants were >90% intelligible during speech evaluation

Referred for speech therapy: 15 (56%)

Those with a moderate-severe impact on quality of life **directly due to speech impairment**: 22 (81%)

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THE ROLE OF SLPS IN REHABILITATION

- Assessment
- Treatment
  - Restoration
  - Compensation
  - Supplementation
- Counseling (as it pertains to a communication or swallowing disorder)
- Collaboration
- Advocacy & Education

WHEN TO REFER FOR SPEECH THERAPY

- **Does the patient report concern about their speech?**
  - Even if you think their speech sounds relatively normal/is easy to understand
- **Does the patient's speech impairment affect their quality of life?**
  - Consider activity limitations, participation restrictions, self-image, communication environments
- **Depending on the etiology of ataxia, the patient may present with other symptoms that SLPs can treat, such as:**
  - Additional speech impairments (e.g., neuromuscular weakness, strained vocal quality due to spasticity, etc.)
  - Language impairments (e.g., difficulty expressing thoughts coherently, difficulty comprehending others, etc.)
  - Cognitive impairments
  - Swallowing impairments
- **If the patient is concerned about their speech and/or presents with concerns in these other areas, make the referral!**

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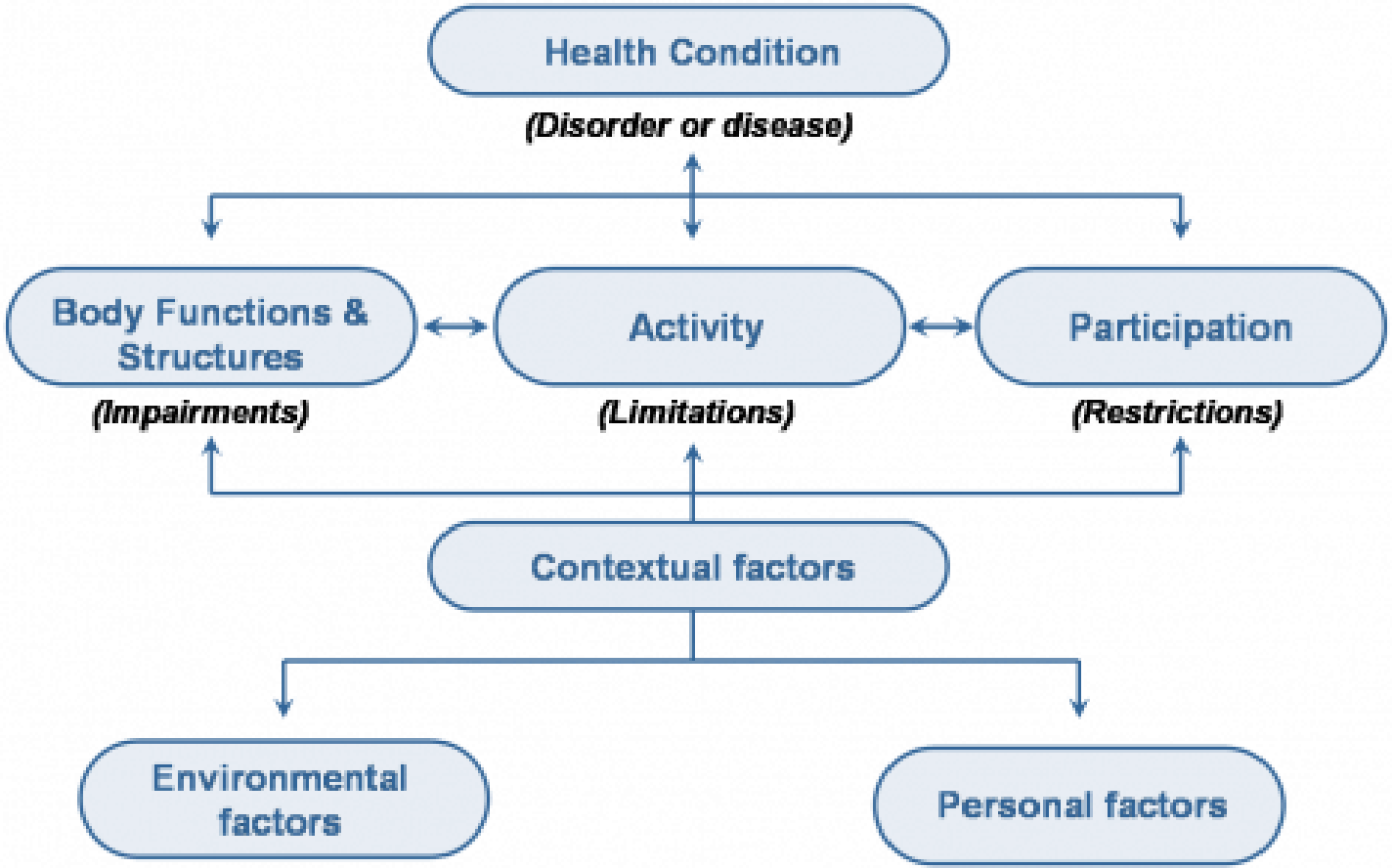

World Health Organization. (2001). International Classification of Functioning, Disability, and Health (ICF).

Based on Johnson et al., (2008)

CASE STUDY #1

Chart review:

- 35-year-old man
- MVA 2 years prior (moderate TBI)
- Patient requested appt due to fatigue when speaking and “clumsiness”

What information would you like to gather during your appointment?

CASE STUDY #1

Chart review:

- 35-year-old man
- MVA 2 years prior (moderate TBI)
- Patient requested appt due to fatigue when speaking and “clumsiness”

Clinical interview:

- Symptom onset following TBI
- Improvements in movement, balance and speech over first year
- Symptoms have been stable since then but continue to affect QoL
- Not currently receiving any therapies (e.g., PT, OT, SLP)
- Pt unsure about imaging findings

Observations & findings:

- Breathing at rest appears WFL
- Spirometry findings WFL
- Cranial nerve exam & reflexes WFL
- Limb movements have a slow, jerky quality
- Speech patterns
  - 1-2 words/breath
  - Excessively loud voice
  - Inconsistent articulatory breakdowns

What is the patient’s prognosis?

Would you make any referrals?

CASE STUDY #2

Chart review:

- 30-year-old woman
- Mild TBI due to a fall 1 month ago

Clinical interview:

- Endorses changes in balance and coordination prior to TBI
- Reports unexpected stumbling over words, especially when tired
- No w/u prior to mTBI – PCP did not refer pt for additional diagnostic testing or treatment, suspected poor sleep was the cause

What other information would you like to obtain during the appointment?

Would you make any referrals?

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https://www.colorado.edu/lab/motor-speech/

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