



## What is Ataxia?

While the term “ataxia” is used to describe symptoms of incoordination that may occur in a variety of disease processes, the term “Ataxia” is also used to label a heterogeneous group of neurodegenerative diseases. The degenerative ataxias (referred to herein as Ataxia) result from hereditary or sporadic genetic mutations that ultimately lead to degeneration of the cerebellum, cerebellar pathways, and/or the spinal cord.

## Key Characteristics

### Overview of Common Symptoms:

Symptoms vary by person and type of Ataxia. Age of onset and rate of progression varies as well. Symptoms may worsen slowly, over decades – or quickly, over mere months. Common symptoms of Ataxia include impaired coordination, dysarthria, dysphagia, deterioration of fine motor skills, gait and balance abnormalities, eye movement abnormalities, tremors, heart problems, vision changes, and cognitive changes. Speech-language pathologists (SLPs) are key allied health providers in the management of speech, swallowing, and cognitive dysfunction resulting from Ataxia.

### Motor Speech Disorders in Ataxia

Ataxia primarily affects the cerebellum, the brain region responsible for the coordination and fluidity of skilled motor behavior. Cerebellar degeneration often leads to ataxic dysarthria, a motor speech disorder characterized by:

- Atypical respiratory patterning
- Imprecise articulation with irregular articulatory breakdowns + telescoping
- Prosodic abnormalities – difficulty modulating pitch, loudness, and speech rate
  - May have overly rigid prosody (scanning cadence) or highly variable prosody (explosive loudness bursts, excessive variation, pitch instability)
  - Rate of speech may be slow or variable overall, and often produce atypical segment/pause durations which influence perception of speaking rate

While ataxic dysarthria is the most common motor speech disorder observed in Ataxia, patients may also exhibit mixed dysarthrias. Clinically, you may see evidence of weakness, spasticity, rigidity, or abnormal movements during speech production. Such features point to mixed dysarthria presentations. The presence of mixed dysarthria suggests that the patient either (a) has a subtype of ataxia that leads to more widespread degeneration of neural tissue, or (b) may have a comorbid neurological condition warranting further investigation, if the observed impairments are not consistent with the patient's medical history.

### Swallowing Disorders in Ataxia

Ataxia can lead to oropharyngeal dysphagia. Swallowing symptoms may be variable, as incoordination does not necessarily manifest with every swallow or under all conditions. Swallow safety impairments tend to be more common than efficiency impairments, consistent with incoordination as the underlying pathophysiology of Ataxia. Muscle tone, range of motion, and strength as they relate to swallowing are often intact, unless neural degeneration extends beyond the cerebellum.

### Cognition and Language in Ataxia

Recent evidence has shown that the cerebellum is involved in aspects of cognition, language, and emotion. People with Ataxia may experience changes in emotional regulation, memory, attention, and linguistic processing.

## The SLP's Role: Recommendations & Current Evidence

As you review the evidence and recommendations below, keep in mind that there are many subtypes of Ataxia, and every patient is unique. Always focus on the patient in front of you and be mindful of potential bias that might affect your interpretations if you are (or aren't!) aware of the patient's neurological diagnosis.

### Evaluation & Management: Dysarthria

1. Conduct a comprehensive motor speech assessment including connected speech tasks, diadochokinetic rate tasks, phonatory tasks such as maximum phonation time, and an oral mechanism exam.
2. During evaluation, look for patterns of speech characteristics as well as salient features that don't fit the pattern. Careful differential diagnosis can contribute meaningfully to a patient's medical work-up, regardless of whether or not they already have an Ataxia diagnosis.
3. Specific treatment strategies to consider include:
  - a. Improving lung volume initiation consistency to improve prosodic control
  - b. Using acoustic feedback to visualize pitch and loudness contours
  - c. Focusing on durational control of words and syllables to improve signalling stress
  - d. Practicing multisyllabic word production

### Evaluation & Management: Dysphagia

1. Regular screening for dysphagia signs/symptoms is recommended, with further clinical and instrumental evaluation as indicated
2. When seeing a patient for speech or language concerns, be sure to monitor for early signs of dysphagia. One of the earliest emerging and most common symptoms reported by people with Ataxia is coughing/choking on liquids, but they may not perceive that this symptom reflects a swallowing problem.
3. Conduct instrumental assessment if there is clinical suspicion for dysphagia. This is critical for characterizing swallowing pathophysiology and monitoring disease progression.
4. Always conduct an instrumental assessment before providing intervention. Many dysphagia interventions are unlikely to be effective for those with Ataxia, as many current interventions target strength over coordination.
5. Even mild dysphagia can impact quality of life. Many patients benefit from counseling pertaining to the impacts of swallowing impairments on daily life.
6. Specific treatment strategies to consider include:
  - a. Target motor patterning/coordination rather than strength (in most cases)
  - b. Use sensory approaches to enhance sensory feedback and sensorimotor integration
  - c. Implement biofeedback (e.g., sEMG, FEES, spirometry)
  - d. Skill training (e.g., respiratory-swallow coordination training, cough skill training)
  - e. Compensatory approaches: these should be explored alongside exercise-based approaches given the progressive nature of Ataxia. It is often a combination of rehabilitative and compensatory approaches that are most effective.

### Evaluation & Management: Cognition & Language

1. Complete cognitive screening with evaluation/intervention as indicated
2. Patients may or may not develop cognitive-communication symptoms – these are more likely to develop later in disease progression and may also be associated with comorbid conditions.
3. If the patient exhibits rapidly progressive cognitive decline or a sudden change from baseline, be sure they are referred for further evaluation as such presentations are not typical of Ataxia.

### Referral

As with other neurodegenerative conditions, you may need to refer to specialty providers depending on your patient's clinical presentation. You may refer to providers in neurology, neuropsychiatry, counseling psychology, gastroenterology, nutrition, physical therapy, occupational therapy, or other medical and allied health providers.

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