Spinocerebellar ataxia (SCA)
Part 2: Dysphagia considerations

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As SCA progresses, swallowing function becomes progressively more impaired due to declining motor and neurological functions. The purpose of this resource is to highlight the disease progression and dysphagia assessment/treatment considerations. This resource is a brief overview intended to provide basic knowledge, and clinicians are encouraged to refer to the references provided for further reading and understanding of SCA.

UNDERSTANDING THE ETIOLOGY: WHY CAN SCA CAUSE DYSPHAGIA

Given multiple SCA subtypes, the clinical presentation of SCA is variable. However, a commonality between the subtypes is that some part of the central nervous system is affected, which can cause neurogenic swallowing dysfunction. Common structures affected include but are not limited to the cerebellum, pontine nuclei, spinal cord, peripheral nerves, cerebral cortex, and the basal ganglia (Bhandari et al., 2023). Imaging may frequently reveal atrophy, structure enlargement (such as with the ventricles), and myelin loss in the spinal cord (Bhandari et al., 2023).

DYSPHAGIA CONSIDERATIONS

Dysphagia management is of utmost importance for patients with SCA, given the significant impact on mortality. Post-mortem studies have found that aspiration pneumonia is one of the most common causes of death among people with SCA (Diallo et al., 2018; Diallo et al., 2019; Russo et al., 2015; Yang et al., 2020). The presence of dysphagia suggests a worse prognosis (Diallo et al., 2018; Diallo et al., 2019).

DYSPHAGIA PROGRESSION IN SCA

The progression of dysphagia appears to vary depending on the disease subtype; however, by moderate disease stages, most patients tend to have dysphagia (Yang et al., 2020). Thus, patients should be screened for dysphagia by moderate disease stages at the latest (and earlier screening is preferred) (Yang et al., 2020). Yang et al.'s 2020 research found a 60% prevalence rate of dysphagia among a mixed cohort of patients with SCA. Dysphagia may already exist in pre-diagnosis stages (Chien et al., 2022). Early intervention in the disease course is recommended to improve oral motor control and lessen the risks of aspiration pneumonia (Chien et al., 2022; Isono et al., 2013). Dysphagia is one of the most prominent symptoms of SCA1; studies support that 100% of individuals with SCA1 have dysphagia (Vogel et al., 2015b). Dysphagia advances to severe status within 15 years of symptom onset and is associated with higher mortality, given risks of aspiration, pneumonia, and malnutrition (Chien et al., 2022).

Dysphagia Assessment Considerations

- Chart review: note how long the patient has experienced SCA symptoms and how long they've had their diagnosis.
  - Some reports note that dysphagia generally occurs within eight years of disease onset among the majority of patients (Chien et al., 2022).

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- Be aware of relevant tools and questionnaires- one clinical tool that can be utilized is the Clinical Assessment of Dysphagia in Neurodegeneration (CADN) (Vogel et al., 2017).
    - The CADN has been studied in degenerative ataxias, compared to MBS results, and has been found to be a reliable, valid, and brief tool acceptable for use with neurodegenerative diagnoses (Vogel et al., 2017).
    - The CADN may help stratify how severe a patient's dysphagia may be and whether there's low or high importance for an instrumental swallow study to further assess current symptoms based on present complaints (Vogel et al., 2017). The CADN may be found when with the online version of the article amongst the supplemental materials (Vogel et al., 2017).
- Note common swallowing physiology impairments seen in research utilizing MBS among patients with SCA:
    - Reduced chewing movements/ oral preparation, reduced oral coordination (Abdulmassih et al., 2013)
    - Pharyngeal residue (Abdulmassih et al., 2013; Ramio-Torrentia et al., 2006)
    - Lingual coating (Ramio-Torrentia et al., 2006)
    - Laryngeal penetration and tracheal aspiration (Vogel et al., 2015a; Vogel et al., 2020),
- Note predictors of dysphagia among people with SCA:
    - Moderate ataxia characterized by Scale for Assessment and Rating of Ataxia score 15 or worse (Russo et al., 2015; Shirley Ryan Ability Lab, 2015; Yang et al., 2020)
    - Responding with the two most negative responses to the Swallowing Quality of Life (SWAL-QOL) prompts: "It takes me longer to eat than other people", "It takes me forever to eat a meal" (Russo et al., 2015)
        - Russo and colleagues (2015) suggest if the above two symptoms are noted, it should prompt MBS for objective dysphagia assessment
    - Longer SCA symptoms (3+ years) (Yang et al., 2020)
- Read head imaging reports
    - Research indicates that SCA subtypes with greater involvement of the brainstem (specifically the medulla) increases risks of sensory and motor pharyngeal phase dysphagia and aspiration pneumonia (Isono et al., 2013; Rüb et al., 2006)

DYSPHAGIA TREATMENT CONSIDERATIONS

- The SLP may utilize compensatory and/or rehabilitative efforts depending on SCA severity, dysphagia severity, cognitive status, and goals of care. Treatment techniques will vary on a case-by-case basis given the heterogeneous clinical presentation across multiple subtypes. A patient's ability to implement postural compensations and swallowing exercises is highly dependent on motor and cognitive skills.
- Compensatory measures such as changes to head positioning, diet texture, and maneuvers may be used depending on the findings of instrumental swallow studies. A compensatory strategy should not be implemented unless an instrumental study endorses its utility and safety.
- Inspiratory muscle strength training (IMST) has been found beneficial among patients with SCA (Chien et al., 2022).
    - IMST with a load set of 60% of maximum inspiratory pressure for 20 minutes per day has been found effective to improve respiratory muscle strength and breathing patterns (Chien et al., 2022).
    - It's possible that IMST treatment regimens may have additional benefits in SLP treatment: reduced shortness of breath during speech, increased glottic opening for vocal cord dysfunction, improve inspiratory drive and cough adequacy (Richardson, 2020)
- High quality research on dysphagia treatment for people with SCA is limited, however dysphagia treatments that have been studied in this population include: effortful swallow exercise, oral motor exercises, thermal-tactile stimulation (Vogel et al., 2015b).

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- The available dysphagia treatment literature places an emphasis on diet texture modification to aid with lessen risks of sequela from aspiration (Chien et al., 2022; Isono et al., 2013; Ramio-Torrentia et al., 2006)
- Note that patients with SCA may be at high risk for malnutrition, dehydration, and weight loss due to progressive dysphagia (Rüb et al., 2006). Thus, SLPs should practice in close collaboration with physicians and registered dietitians to help ensure adequate and safe oral intake.
- Ongoing swallowing re-assessments can also assist with guiding timing for PEG tube placement (Isono et al., 2013)

The next section of this resource (Part 3) will focus on dysarthria and cognitive assessment and treatment considerations.

HOW TO REFERENCE THIS RESOURCE:

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