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Friedreich's Ataxia: Treatment and Management for the Speech-Language Pathologist

Friedreich's ataxia is a neurodegenerative disease, and it is the most common hereditary ataxia (Eigentler et al., 2012; Richardson, Kelly, Yu, Simpkins, & Worth, 2015). Ataxia is the loss or weakness of physical movements and can present as incoordination. Characteristics of Friedreich's ataxia include vision problems, limb ataxia, dysarthria, and dysphagia (Vogel et al., 2017). Dysarthria effects the control of muscles during speech (Vogel, Folker, & Poole, 2014). Dysphagia is difficulty swallowing. Onset of the disease typically occurs during the adolescent years when one is going through puberty, but it is common to not have symptoms appear until years after the suspected onset (Poole, Wee, Folker, Corben, Delatycki, & Vogel, 2015). One out of 50,000 people are affected and one out of 720 are carriers (Richardson et al., 2015).

There are many structures in the brain that are affected by Friedreich's ataxia, but the most prevalent area is the cerebellum (Folker et al., 2012). The cerebellum integrates smooth controlled movements, as well as cognitive tasks like planning. Other areas that are affected include the corticobulbar and corticospinal tracts. These coordinate skilled voluntary movement (Folker et al., 2012).

In the field of speech-language pathology, people with Friedreich's ataxia may be seen on their caseload due to the patient having dysarthria and dysphagia. No options are available to stop the progression of the disease, but there are therapeutic interventions available to modify some of the symptoms (Cook & Giunti, 2017). Since Friedreich's ataxia is a degenerative disease, it is crucial to get a baseline when the person is first diagnosed. The use of augmentative and alternative communication devices (AAC) may be used as the disease progresses, as well as improving motor control for speech improvement and using compensation strategies (Cook & Giunti, 2017). Swallowing impairments, also known as dysphagia, is very common and put someone's safety at risk. The speech-language pathologist may need to alter diets and continue instrumental analysis for dysphagia as the disease progresses (Keage, Delatycki, Gupta, Corben, & Vogel, 2017).

The purpose of this presentation is to provide an overview of the signs and symptoms of Friederichs's ataxia and what speech-language pathologists (SLP) can do to help manage the symptoms. It will include management of dysarthria and dysphagia, and what it means for an SLP to manage a degenerative disorder.

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