**RESUMO ARTIGO CONGRESSO BRASILEIRO DE NEUROLOGIA:**

**Assessment of Respiratory Function in Friedreich’s Ataxia**

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**INTRODUCTION:**

Hereditary ataxias constitute an extensive group of clinically and genetically heterogeneous neurodegenerative diseases. The ataxias are divided into autosomal dominant, autosomal recessive, X-linked, mitochondrial ataxias, congenital and sporadic ataxias. (tese Celiana)

Friedreich’s ataxia (FRDA) is the most common cause of autosomal recessive ataxia worldwide. It was first described by Nikolaus Friedreich in 1863. It is caused biallelic GAA repeat expansion in the FXN gene, which results in reduced expression of the encoded protein Frataxin. (Machado et al, 2024). So, Friedreich ataxia (FRDA) is a rare condition that cause nervous system damage and movement problems, including muscle weakness and impaired coordination (ataxia). Heart problems, vision problems, spine problems, and diabetes can occur, too. Whitin 10 to 20 years of the first symptoms, an individual with FRDA generally requires a wheelchair. (Paridhi Jains et al, 2022). Friedreich ataxia, develop disorders of the sleep, respiratory complications and dysphagia. Aspiration pneumonia is one of the main causes of death in these patients. There are few studies on the assessment of pulmonary function in ataxias, as well as what is the form of prevention for aspiration pneumonia and ventilatory failure. The purpose of this study is, therefore, to evaluate the respiratory function in Friedreich ataxia. (tese Celiana)

**OBJECTIVES:** To evaluate the lung function of individuals diagnosed with Friedreich's ataxia through respiratory function tests and phrenic nerve conduction. Correlate respiratory assessment data with demographic data (age, duration of the disease), anthropometric data (weight, height, BMI, cervical and abdominal circumference) and data from the SARA and ICARS scales**.**

**METHODS: Cross-sectional study, including men 4 and women 12 and controls healthy men 10 and women 10. Age 13- 68 in Friedreich.**

**We used for assessment spirometry, Inspiratory and expiratory pression test, sniff test and Peak cough flow test. We used too, correlated tests with ICARS and SARA scale. RESULTS:** The Shapiro-Wilk test was used to evaluate data adherence to normal distribution. To describe the variables between the groups, means and standard deviations, and absolute and relative frequencies were used. To compare variables between groups, the Mann-Whitney test was used. To compare categorical variables, the chi-square test was used. The significance level established was 5%. Analyzes were performed in STATA (StataCorp, LC) version 18.0.

**DISCUSSION**: Patients with Friedreich’s ataxia has many respiratory difficulties comparing with healthy control. This has not been observed in follow-up studies of this patients.

**CONCLUSION:**