

Features of speech and swallowing dysfunction in pre-ataxic Spinocerebellar Ataxia Type 2

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Objective: To determine if objective and quantitative assessment of dysarthria and dysphagia in SCA2, specifically at pre-ataxic and early disease phases, can act as sensitive disease markers.

Methods: Forty-six individuals (16 pre-ataxic SCA2, 14 early-stage ataxic SCA2 and 16 healthy controls) were recruited in Holguin, Cuba. All participants underwent a comprehensive battery of assessments including objective acoustic analysis, clinician derived ratings of speech function and swallowing, and quality of life assessments of swallowing.

Results: Reduced speech agility manifest at the pre-ataxic stage was observed during diadochokinetic tasks, with the magnitude of speech deficit augmented in the early-ataxic stage (see Figure 1). Speech rate was slower in early-stage ataxic SCA2 compared with pre-ataxic SCA2 and healthy controls. Reduced speech agility and speech rate correlated with disease severity and time to ataxia onset, verifying speech deficits occurred prior to ataxia onset and increase in severity as the disease progresses. Whilst dysphagia was observed in both pre-ataxic and ataxic SCA2, it was not associated with swallowing-related quality of life, disease severity or time to ataxia onset.

Conclusions: Speech and swallowing deficits appear sensitive to disease progression in early-stage SCA2, with syllabic rate a viable marker. Findings provide insight into mechanisms of disease progression in early-stage SCA2 signalling an opportunity for stratifying early-stage SCA2 patients and identifying salient markers of disease onset as well as outcome measures in future early-stage therapeutic studies.

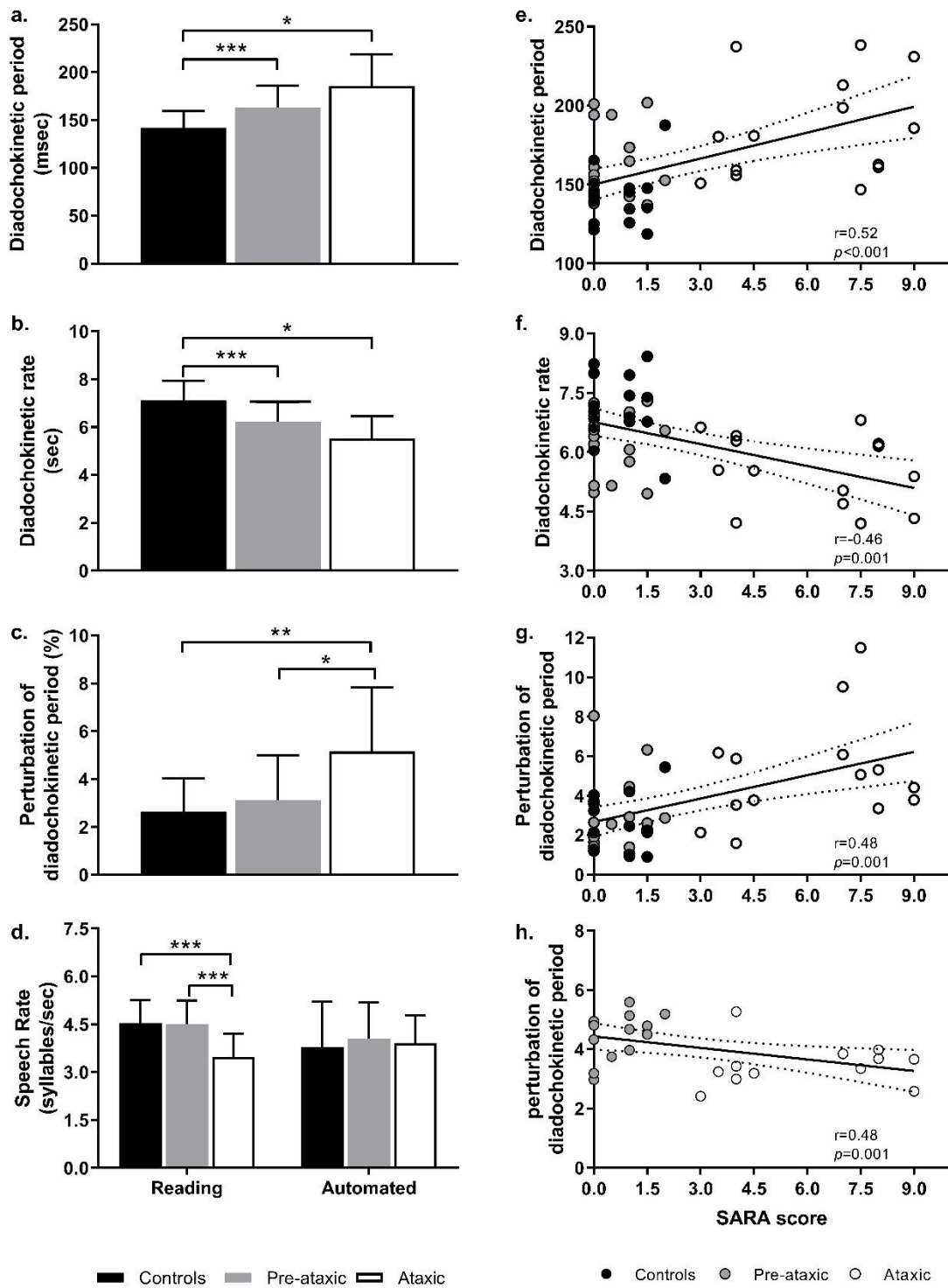


Figure 1. Mean speech agility scores (a to d; error bars represent standard error of the mean) and correlations between speech agility scores and disease severity ratings (SARA; e to h).