Title: Home speech and coordination measurement in spinocerebellar ataxias and multiple system atrophy

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Background: Current clinical measures of spinocerebellar ataxias (SCA) are subjective and typically not suitable for frequent at-home administration, resulting in imprecise measurements of disease severity. This study aimed to evaluate the construct validity and test-retest reliability of at-home assessments to monitor disease severity in SCA and possible/probable Multiple System Atrophy-cerebellar type (MSA-C) patients. Developing accessible tools to precisely measure disease severity will support clinical trials and care for individuals with ataxia.

Methods: Subjects participated in a remote study to collect speech biomarkers, Hevelius computer mouse task performance, and wrist and ankle accelerometer data. Participants were mailed a standardized laptop, computer mouse, and 2 GENEActiv devices to wear continuously for one week. A physician-administered examination was conducted for each participant via Zoom video conference. During this session, BARS, SARA, and UPDRS components that could be completed remotely were performed by two neurologists. Following the clinical assessment, participants completed speech surveys and Hevelius computer mouse tasks biweekly for 4 weeks and 5 quality of life and daily function questionnaires, including PROM-Ataxia and Neuro-QOL, at baseline and post-study.

Results: Enrollment is projected to complete in August 2022. 16 subjects with SCA/MSA-C and 7 healthy controls have completed the study. Analysis of construct validity showed that out of 32 Hevelius features, 25 demonstrated a moderate-strong correlation with either BARS or SARA scores (r=0.60-0.75) and 4 demonstrated a moderate-strong correlation with PROM-Ataxia arm scores (r=0.65-0.69). Of those highly correlated features, 6 showed good-excellent test-retest reliability (ICC=0.797-0.904). The assessments demonstrated high feasibility, with participants completing 94% of Hevelius tasks.

Discussion: These data demonstrate the potential use of Hevelius in tracking disease severity in SCA and MSA-C patients. We plan to extend this study longitudinally over a period of 12 months following the initial testing. Our final analysis will include additional modalities, such as data from GENEActiv devices and speech surveys.