TITLE: Cognition and its correlates in SCA 1, 2, 3, and 6

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**ABSTRACT:**

**Background and objectives:** There is growing evidence of cognitive deficits in spinocerebellar ataxias, with the Cerebellar Cognitive Affective Syndrome Scale (CCAS-S) an increasingly common measure of this dysfunction. There remain ongoing questions as to how scale performance relates to day-to-day cognitive function and other features of SCAs. We aimed to evaluate CCAS-S performance amongst individuals with SCA1, SCA2, SCA3, and SCA6 relative to matched controls, and to investigate relationships between performance and demographic factors, ataxia severity, psychomotor performance, and non-motor functions.

**Methods:** Using data collected remotely via an online and teleconference-based approach, we evaluated performance on the CCAS-S amongst individuals with SCA1 (n=14), SCA2 (n=16), SCA3 (n=18) and SCA6 (n=26) relative to control groups matched for age, sex, and education level. We examined associations between CCAS-S performance and a) age and education, b) measures of ataxia symptom severity, c) performance on computerised finger-tapping and reaction time tasks d) self-rated cognition, depression, emotional regulation, psychosocial function, and fatigue.

**Results:** CCAS-S performance was significantly reduced in SCA2, SCA3, and SCA6 compared to controls. Better CCAS-S performance was associated with more years of education and lower ataxia symptom severity. Better CCAS-S performance was also associated with both average psychomotor task speed (motor tapping speed, simple reaction time, decision reaction time) and variability in psychomotor performance across task trials (motor tapping speed, motor timing, decision time). Finally, CCAS-S performance was significantly associated with self-reported day-to-day cognitive function.

**Discussion and conclusion:** The CCAS-S is a useful cognitive measure in SCAs that maps on to self-reported cognitive function as well as motor function. This study motivates further investigation into the manifestation of cognitive dysfunction in these conditions and the role ataxia severity motor symptoms and psychomotor variability play in the development of these deficits.

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