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General neurologic findings in SCAs: a comparison between NESSCA and INAScount

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Introduction: Spinocerebellar ataxias (SCAs) due to CAGexp display heterogeneous clinical manifestations. The most used scale in SCAs is SARA, which measures ataxia severity. Other neurologic manifestations have been measured by NESSCA and INAScount. Our aim was to describe their baseline characteristics among clinical and preclinical stages of SCA3/MJD carriers.

Methods: Molecularly diagnosed subjects and at 50% risk relatives were recruited. CAGexp, SARA, NESSCA and INAS were obtained. Ages at onset (AO) was defined by the onset of gait ataxia SCA3/MJD. Models to predict mean age at onset (AO) for each CAGexp length were developed. Time to onset (TTO) was the difference between present age and predicted AO. To put all carriers in the same time perspective, TTO was combined to disease duration (from symptomatic carriers) in the variable TTO/DD. Significance threshold was 0.05.

Results: 22 symptomatic and 15 presymptomatic SCA3/MJD carriers were recruited. In SCA3/MJD, NESSCA (rho=0,808) and INAScount (rho=0,705) correlated to TTO/DD without roof effect; SARA correlated with NESSCA (rho=0,865) and INAS count (rho=0,810); NESSCA correlated with INAS count (rho=0,762).

Conclusion: There were incomplete correlations between NESSCA and INAScount in all scenarios, due to differences in items contents and/or weights. Overall, correlations obtained for NESSCA were stronger than those obtained for INAScount. Follow-up evaluations will allow obtaining the best parameters to compare these scales.

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