25 Validation of hSARA and first data from home-application

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Background: Clinical scales, such as the Scale for the Assessment and Rating of Ataxia (SARA), cannot be used to study the severity of ataxia at home, nor to assess day-to-day and within-day fluctuations.

Objective: To develop a video-based instrument, homeSARA (hSARA), for measuring ataxia severity that can be easily and independently applied by ataxia patients at home.

Methods: Based on the feasibility of self-application, we selected five of the eight SARA items (gait, stance, speech, nose-finger test, fast alternating hand movements) for hSARA (range: 0 – 28). We compared total hSARA with total SARA scores in 526 patients with spinocerebellar ataxia types 1, 2, 3, and 6 from the EUROSCA natural history study. To prospectively validate hSARA, we directly compared the results of hSARA with those of SARA in 50 ataxia patients. To demonstrate feasibility of independent home recordings, we performed a feasibility study in 11 ataxia patients who were instructed to obtain a video each morning and evening over a period of 14 days. Videos of the validation and feasibility study were rated offline by a trained rater.

Results: The hSARA score extracted from the EUROSCA baseline data was highly correlated with the SARA score ($r = 0.9854$, $p < 0.0001$). In the prospective validation study, the hSARA score derived from video rating was highly correlated with the conventional complete SARA score ($r = 0.9254$, $p < 0.0001$). Eight of 11 participants of the feasibility study obtained a complete set of 28 evaluable videos. Three participants obtained 13, 16 and 17 videos. The intraindividual differences between the lowest and highest hSARA score ranged from 1 to 5.5.

Conclusion: hSARA scores are highly correlated with conventional SARA scores. Application at home is feasible. There was a considerable degree of intraindividual variability of hSARA scores.