



Application of a Scale for the Assessment and Rating of Ataxia (SARA) in Friedreich's ataxia patients according to posturography is limited

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ABSTRACT

Background: A scale for the Assessment and Rating of Ataxia (SARA) was developed for evaluation of autosomal dominant cerebellar ataxias (ADCA) and was also recommended for clinical trials of Friedreich's ataxia patients (FRDA). FRDA, unlike ADCA, is characterized as being a sensory type of ataxia for which the disease-specific Friedreich ataxia rating scale (FARS) was developed. The objective of this study was to determine whether SARA and FARS scores are associated with posturographic parameters in FRDA patients.

Method: Adult patients with genetically confirmed FRDA ($n = 11$) and ADCA ($n = 13$) were evaluated by SARA, FARS and posturography.

Results: FRDA patients' postural stability parameters, in stance with visual control, correlated with balance impairment in FARS ($r = 0.622$; $p < 0.05$) and SARA ($r = 0.735$; $p < 0.05$). Without visual control, only FARS correlated with balance impairment ($r = 0.732$; $p < 0.05$).

Conclusion: The SARA, in FRDA patients, correlates with stance with visual control but not without visual control which emphasizes sensory ataxia. This suggests that application of the SARA in Friedreich's ataxia patients according to posturography is possible but presumably limited and FARS, although being a more time consuming scale, may have advantages over SARA in FRDA patients.

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1. Introduction

Friedreich's ataxia (FRDA) is the most frequently inherited ataxia, affecting about 1 in 30 000 individuals in Western Europe [1]. There is no treatment, although several clinical trials have been conducted in recent years. In FRDA, as in other progressive neurological conditions, accurate and efficient measurement of disease progression is necessary to evaluate the effectiveness of therapy.

Because of the heterogeneous presentation of FRDA and its slow progression it is very difficult to find a reliable instrument for measuring the changes of the FRDA's symptoms over time. The Friedreich's ataxia rating scale (FARS) [2], International Cooperative Ataxia Rating Scale (ICARS) [3], and the Scale for the Assessment and Rating of Ataxia (SARA) [4] are the recommended neurological examination-based instruments most used to assess the disease among persons with FRDA and combined forms of ataxia.

The FARS was developed as a disease-specific rating scale for FRDA patients. Initial studies showed that FARS is a more sensitive measure than ICARS [5] and has been used in clinical trials in FRDA patients. This scale examines both sensory and cerebellar symptoms [6]. The disadvantage is that it is time consuming.

The SARA, a shorter ataxia scale, was developed to measure symptoms of cerebellar ataxia in autosomal dominant spinocerebellar ataxia (ADCA). Since FRDA also has sensory (afferent) elements of ataxia, SARA was not considered to be appropriate for FRDA and was not evaluated in FRDA patients [7]. A recent study based on a comparison of the three rating scales – ICARS, FARS and SARA – indicated that SARA may be appropriate for clinical trials of FRDA patients [6]. Further, the study showed that SARA measures disease severity and progression in FRDA patients [8].

Posturographic examination has been used for measurement of postural stability and quantitative documentation of postural ataxia [9]. Increased postural instability was observed among cerebellar patients and correlated with balance impairment on a clinical ataxia rating scale [10].

Our study aimed to determine whether posturographic parameters are associated with balance impairment assessed by specified clinical ataxia rating scales among FRDA (SARA, FARS) and ADCA (SARA) patients.

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2. Patients and methods

2.1. Patients

Thirteen spinocerebellar ataxia type 2 (SCA2) patients for which the threshold number of CAG repeats in *ATXN2* gene was 33 [11,12] and 11 FRDA patients, all homozygotes for expanded GAA repeat in the *FRDA* gene, genetically confirmed by the Ataxia Center at Motol Hospital in Prague, were assessed by the SARA semi-quantitative clinical rating scale. Patients with FRDA in addition were assessed by FARS. All assessments were performed at the same time and by the same neurologist specializing in ataxia.

Patients in the study had the ability to stand with or without visual control more than 30s without support. SCA2 patients did not have any clinical signs of polyneuropathy (hyporeflexia or areflexia) or extra-pyramidal signs.

The Central Ethical Committee of the University Hospital Motol Prague approved the study. All participants gave written consent.

2.2. Rating scales

SARA is a semi-quantitative scale scored from 0 (no ataxia) to 40 (most severe ataxia) and comprised eight items: (1) gait (score, 0–8); (2) stance (score, 0–6); (3) sitting (score, 0–4); (4) speech disturbance (score, 0–6); (5) finger chase, dysmetria (score, 0–4); (6) nose–finger test; tremor (score, 0–4); (7) fast alternating hand movements (score, 0–4); and (8) heel–shin slide (score, 0–4). Limb kinetic functions (items 5–8) are rated independently on each side and the mean thereof is included in the SARA total score. Balance impairment of SARA is the sum of items 1, 2 and 3.

FARS: Clinical examination, part III of the FARS scale, includes a 117 total deficit score with a subscore for bulbar (FARS III-A, maximum score: 11); upper limb (FARS III-B, maximum score 36); lower limb (maximum score: 16); peripheral nerve (maximum score: 26); and upright stability/gait functions (maximum score: 28). Balance impairment of FARS is upright stability/gait functions.

2.3. Assessment of postural stability parameters

The multi-sensoric FOOTSCAN platform (RS scan; Belgium; 0.5 m × 0.4 m; approximately 4100 sensors; sensitivity from 0.1 of N/cm²; sensing frequency 500 Hz) was used for posturographic examination. Standard standing position with a wide base was measured according to standard practice for 30s [13] and transparent sheeting for tracing foot position was used during examination. Wide base standing position was set to the width of the hips. Standing position was the same for the standing with open and closed eyes. During the standing position with open eyes a fixed round point 0.2 m size was placed on the wall at the level of patient's eyes. Pressure on individual sensors was measured and the center of pressure (COP) calculated on the contact area. The following parameters were used: medio–lateral directional deviation (delta X); antero–posterior directional deviation (delta Y) and the whole course of total travelled way (TTW) of COP. Parameters were chosen based on the posturographic studies in patients with ataxia [9]; on work verifying different indicators of postural stability [14]; and on clinical scale affinity [14,15].

2.4. Statistical analysis

Clinical data were analyzed using parametric two-sample *t*-tests for independent samples (disease duration, age of examination) and the non-parametric Mann–Whitney *U* test for SARA and FARS. Correlation between factors was evaluated by Pearson's parametric correlation analysis. The probability of type I error (alpha) was set at 0.05 in all statistical analyses. The post-hoc (retrospective) power analysis was

performed to depict the power of each correlation in the study and was set at 0.2 (conventional value).

Statistical analysis was performed using IBM® SPSS® v21 (Statistical Package for Social Science, Inc., Chicago, IL, 2012)). Power analysis was performed using G*Power (version 3.1.9., Kiel University, GER).

3. Results

Demographic data and clinical rating scale scores are presented in Table 1. All defined postural stability parameters correlated with balance impairment on SARA and FARS among FRDA patients in stance with visual control. Total SARA and FARS scores correlated with postural stability parameters in stance with visual control. In stance without visual control, balance impairment and total score on SARA did not correlate with defined postural stability parameters, unlike FARS, where balance impairment and total score correlated with all postural stability parameters.

SCA patients' posturographic parameters correlated with balance impairment on SARA and with the total score on both scales, in stance with and without vision control, except delta Y in stance without vision control.

The post hoc power analysis depicted the power of each correlation in the study (Grey cell's background in Table 2) and confirmed correlations between SARA/SARA posture with all posturographic parameters in stance with visual control in FRDA patients; FARS posture and all posturographic parameters in stance without visual control in FRDA patients; delta Y, TTW and SARA and delta Y and SARA posture in stance with visual control in SCA2 patients; delta X and SARA and delta X and SARA posture in stance without visual control in SCA2 patients.

All correlation results between posturographic parameters and clinical rating scale scores and results of post-hoc power analysis (Grey cell's background) in FRDA and SCA2 patients are presented in Table 2.

4. Discussion

The SARA was developed to measure symptoms of cerebellar ataxia in ADCA and was not initially recommended for FRDA. In ADCA, unlike FRDA, the isolated cerebellar dysfunction causes ataxia [5]. Recent studies indicated that SARA may be appropriate for use in clinical trials among FRDA patients [6,8]. This assumption was also supported by our results in stance with visual control in which correlations between all posturography parameters and SARA total score/SARA posture were significant.

Further, association between posturographic parameters with visual control and balance impairment and total scores on FARS and SARA was found. Based on the pathophysiology of FRDA, the loss of afferent input to the cerebellum causes the cerebellar component of ataxia, and sensory ataxia severity relates to cerebellar ataxia severity. If there is an association between cerebellar and sensory ataxia and between balance impairment and posturographic parameters, it may be assumed that the total results of FARS, the sum of cerebellar and sensory ataxia, and posturographic parameters should be associated.

In SARA, unlike in FARS, in stance without visual control, we did not find any associations between posturographic parameters and balance impairment. Variable postural instability is typical for sensory ataxia, which is more pronounced with loss of visual control. Balance impairment assessment in FARS and SARA has no perceptual component and no test with eyes closed is included. However, FARS was developed to give greater weight to gait and stance [2,16], so it may be more reliable in assessment of balance impairment in FRDA patients than SARA.

In FRDA patients FARS total score/FARS posture correlated with all posturographic parameters in stance with and without vision control. These results provide the possibility to use posturographic parameters as a surrogate marker of disease severity and progression.

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