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Validity and reliability of the International Cooperative Ataxia Rating Scale (ICARS) and the Scale for the Assessment and Rating of Ataxia (SARA) in multiple sclerosis patients with ataxia

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ABSTRACT

Background: Ataxia is an extremely common problem in multiple sclerosis (MS) patients. Thus, appropriate scales are required for detailed assessment of this issue. The aim of our study was to investigate the reliability and validity of the Turkish version of the International Cooperative Ataxia Rating Scale (ICARS) and Scale for the Assessment and Rating of Ataxia (SARA), which are widely used in ataxia evaluation in the context of other cerebellar diseases.

Method: This cross-sectional study included 80 MS patients with Kurtzke cerebellar functional system score (C-FSS) greater than zero and slight pyramidal involvement. The Expanded Disability Status Scale (EDSS), C-FSS, and Berg Balance Scale (BBS) were administered. SARA and ICARS were assessed on first admission by two physical therapists. Seven days later, second assessments were repeated in same way for reliability.

Results: Intra-rater and inter-rater reliability were found to be high for both ICARS and SARA ($p < 0.001$) The Cronbach's α coefficients were 0.922 and 0.921 for SARA (reviewer 1 and reviewer 2 respectively) and 0.952 and 0.952 for ICARS (reviewer 1 and reviewer 2, respectively). There were no floor or ceiling effects determined for either scale except for item 17 of ICARS ($p = 0.055$). The EDSS total score had significant correlations with both SARA and ICARS (rho: 0.557 and 0.707, respectively). C-FSS had moderate correlation with SARA and high correlation with ICARS (rho: 0.469 and 0.653, respectively). BBS had no significant correlation with SARA and ICARS. (rho: –0.048 and –0.008 respectively). According to the area under the curve (AUC) value, ICARS is the best scale to discriminate mild and moderate ataxia. (AUC: 0.875). Factor analyses of ICARS showed that the rating results were determined by five different factors that did not coincide with the ICARS sub-scales.

Conclusion: Our study demonstrated that ICARS and SARA are both reliable in MS patients with ataxia. Although ICARS has some structural problems, it seems to be more valid given its high correlations with EDSS and C-FSS. SARA also can be preferred as a brief assessment.

1. Introduction

Ataxia is characterized by incoordination and balance dysfunction in movements in the absence of muscle weakness (Bastian, 1997; Mariotti et al., 2005). It occurs due to lesions in the cerebellum (Ghez, 2000) and its connection. Both genetic and acquired etiological factors are responsible for ataxia (Klockgether, 2010; Tallaksen, 2008). Acquired factors include vascular, demyelinating, neoplastic, autoimmune, toxic, degenerative, and infectious etiologies (Ashizawa and Xia, 2016; Todd and Taylor, 2001; Nachbauer et al., 2015). Multiple sclerosis (MS) is a common cause of acquired ataxia, where up to 80% of MS patients suffer from ataxia at some point during their disease (Swingler and Compston, 1992). Moreover, severe ataxic symptoms have been reported in 32% of MS patients, resulting in limited functions (Weinshenker et al., 1996).

Surgical and pharmacological treatments or physical therapy and rehabilitation modalities are commonly employed to manage ataxic symptoms (Siva et al., 1999; Kesselring and Beer, 2005). However, the use of a valid and reliable assessment tool is extremely important for testing new therapeutic approaches or goal setting. Currently, different

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performance-based clinical scales are available for ataxia rating; the most widely used are the International Cooperative Ataxia Rating Scale (ICARS) and the Scale for the Assessment and Rating of Ataxia (SARA) (Trouillas et al., 1997; Schmitz-Hubsch et al., 2006a; Lynch et al., 2006; Schmahmann et al., 2009; du Montcel et al., 2008).

ICARS rates ataxia-related symptoms through 19 items and 4 subscales (posture and gait disturbances, kinetic functions, speech disorders, oculomotor disorders). Although it is semi quantitative, relies on subjective rating by clinicians (Trouillas et al., 1997). Similarly, SARA is semi-quantitative, but it is much simpler and takes less time in clinical use than ICARS (Schmitz-Hubsch et al., 2006a). To our knowledge, these two scales have not yet been tested for validity or reliability in MS patients, and Turkish versions of these scales are not available. Thus, the aim of our study was to create Turkish versions of the ICARS and SARA scales and demonstrate their reliability and validity in MS patients with ataxia.

2. Methods

2.1. Participants

This research was carried out at the Physiotherapy and Rehabilitation Department, Neurologic Rehabilitation Unit, Hacettepe University, Turkey. The local ethical committee approved the study (Hacettepe University Non-Interventional Clinical Research Ethic Boards, Go: 16/618). In this cross-sectional study, a total of 178 patients were screened for eligibility in November 2016–February 2017. Eighty volunteer MS patients of both sexes with ataxia symptoms were included in the study, while 96 patients were excluded. Patients with a Kurtzke cerebellar functional system score (C-FSS) greater than zero were defined as having cerebellar dysfunction. All participants signed an informed consent form prior to participation. The inclusion criteria were as follows: a) patients with definite MS diagnosed by a neurologist according to the McDonald criteria (Polman et al., 2005), b) patients in the age range of 18–50 years, c) patients who were clinically stable during the 3 months prior to enrollment in the study, d) no acute exacerbation within 3 months, e) cerebellar signs and symptoms with slight pyramidal involvement (C-FSS $\geq$ 1, pyramidal system score of EDSS $\leq$ 2), and f) ability to walk at least 10 m with or without an assistive device (Kurtzke, 1983). The exclusion criteria were as follows: a) having other systemic, orthopedic, or neurological disease, b) having moderate or severe pyramidal symptoms, c) having balance problems due to peripheral vestibular issues, and c) patients with EDSS scores > 6.5.

2.2. Translation of ICARS and SARA into the Turkish Language

Permission was obtained from the corresponding author, Thomas Klockgether, for SARA and the *Journal of Neurological Science* editor, John England, and the author Allen Bryer for ICARS to develop Turkish versions. The scales were translated into Turkish by two physiotherapists and a neurologist who speak fluent English and who specialize in ataxia and MS. A consensus was provided on one Turkish version (version T). Back-translation was carried out by two professional interpreters who were native English speakers (version E1 and version E2). Then, the translation team held a meeting and appraised the version T and version E1/E2. All discrepancies were corrected in version T by comparing the original text and version E1/E2. Consensus versions of the scales were employed by five different physiotherapists for 20 ataxic MS patients. At another meeting, minor revisions were made by discussing the difficulties faced by physiotherapists, and then the final versions of SARA-T and ICARS-T were created.

2.3. Measurements

2.3.1. First admission

The EDSS, SARA-T, ICARS-T, and the Berg Balance Scale (BBS) were administered to the patients after recording their demographic data (age, gender, weight, height), the clinical course of the disease (MS type, MS duration, last exacerbation), and the drugs used for MS treatment. To avoid confusion, a 1-h rest interval was set between the SARA-T and ICARS-T assessments.

2.3.2. Second admission

Seven days after the first admission ICARS-T and SARA-T were repeated with a 1-h interval in between.

2.3.3. Expanded disability status scale

EDSS assessments were carried out by a licensed physiotherapist (Y.S.) to select ataxic patients and determine the level of disability.

2.3.4. Turkish version of international cooperative ataxia rating scale

ICARS-T consists of four separate parts, each of which is used to assess different aspects of cerebellar function. The points obtained from posture and gait (0 − 34), kinetic function (0 − 52), speech disturbances (0 − 8), and gaze disturbance (0 − 6) sub-scores are combined for a maximum of 100 points. A greater score represents greater severity of the ataxia (Trouillas et al., 1997).

Patients performed the ICARS-T scale items only once in a day according to one physical therapist's instructions (H.K.). During the evaluation, two other researchers (Y.S. and A.F.) scored each item on the scale by monitoring patients' performance. The researchers were blinded to each other's scoring. Seven days later, the same patients were reevaluated for the inter-test reliability of the test.

2.3.5. Turkish version of the Scale for the Assessment and Rating of Ataxia

SARA-T consists of 8 items examining the following: 1) gait (0 − 8), 2) stance (0 − 6), 3) sitting (0 − 4), 4) speech disturbance (0 − 6), 5) finger chase (0 − 4), 6) the nose-finger test (0 − 4), 7) fast alternative hand movement (0 − 4), and 8) heel–shin slide (0 − 4). Items 5–8 are related to limb kinetic function and rated bilaterally; the mean scores for both sides are added to the total score (Schmitz-Hubsch et al., 2006a). Like ICARS-T, SARA-T assessment was carried out by two raters. Raters were familiar with the scales and had been using the English version of the scales before the study.

2.3.6. Berg Balance Scale

The BBS consists of 14 items measuring the ability to maintain the balance in different positions, postural changes and movements. Each item is scored between 0 and 4 points, and as the recorded score increases, the balance disorder increases (0 − 56). BBS is a valid and a reliable assessment scale in MS patients (Cattaneo et al., 2007, 2006).

3. Data analyses

SPSS software package (version 21, SPSS Inc., Chicago, IL) was used for statistical analyses. Variables were defined as Mean ± standard deviation (SD) for numerical data and as frequency (%) for categorical data. Floor/ceiling values were computed for SARA-T and ICARS-T. Reliability and validity analyses were carried out using the guideline of Consensus Based Standards for the Selection of Health Status Measurement Instruments (COSMİN) (Mokkink et al., 2010).

In a sample of 80 participants responding to 19 ICARS-T items and 8 SARA-T items, the scales achieved 98% power to the detect the difference between the coefficient of Cronbach's α under the null hypothesis of 0.90 and the alternative hypothesis of 0.80 using a two-sided F test, with a significance level of 0.05.