Multiple Sclerosis and Related Disorders 18 (2017) 135–140

Contents lists available at ScienceDirect

Multiple Sclerosis and Related Disorders

journal homepage: www.elsevier.com/locate/msard

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

Yeliz Salcı[a,*], Ayla Fil[a], Hilal Keklicek[a], Barış Çetin[a], Kadriye Armutlu[a], Anıl Dolgun[b], Aslı Tuncer[c], Rana Karabudak[c]

[a] Department of Physiotherapy and Rehabilitation, Faculty of Health Sciences, Hacettepe University, Ankara, Turkey
[b] Mathematical Sciences, RMIT University, Melbourne, Australia
[c] Department of Neurology, Faculty of Medicine, Hacettepe University, Ankara, Turkey

ARTICLE INFO

Keywords:
Multiple sclerosis
Ataxia
Assessment
Cooperative Ataxia Rating Scale (ICARS)
Scale for the Assessment and Rating of Ataxia (SARA)

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

* Correspondence to: Hacettepe University Hospital, Neurology Department, 06100 Sıhhıye, Ankara, Turkey.
E-mail address: fztyeliz@hotmail.com (Y. Salcı).

http://dx.doi.org/10.1016/j.msard.2017.09.032
Received 10 March 2017; Received in revised form 26 September 2017; Accepted 27 September 2017

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.

3.1. Inter-rater reliability, intra-rater reliability, and internal consistency

Inter-rater and intra-rater reliability were examining with Intraclass correlation coefficient (ICC) and internal consistency was calculated with Cronbach's α coefficient. Cronbach's α ranges between 0 and 1; values around 0.8 are considered to be good close inter-item relationships, while values below 0.6 are poor or unacceptable.

3.2. Structural validity

To analyze structural validity of ICARS-T, a principal component extraction analyses was performed using varimax rotation with Kaiser normalization.

3.3. Hypothesis testing

It was performed by making the following correlations with Spearman's rank correlation coefficient (rho):

- Correlation between ICARS-T, SARA-T, and BBS (convergent validity);
- Correlation between ICARS-T, SARA-T and -C-FSS (convergent validity);
- Correlation between ICARS-T, SARA-T, and total EDSS (external validity).

3.4. Predictive validity

It was measured by using receiver operating characteristic (ROC) curves and area under the ROC curves (AUC). Patients were divided into two groups according to the C-FSS to determine whether the scales were discriminant for mild to moderate ataxia (group 1: C-FSS ≤ 2, group 2: C-FSS ≥ 3). An AUC value of 0.50 represented non-sensitivity, while a value of 1.00 represented perfect sensitivity and specificity (Altman, 1990).

4. Results

Patients' demographic data and clinical characteristics are presented in Table 1. There were no floor or ceiling effects determined for either scale except item 17 of ICARS-T ($p = 0.055$; Tables 2–3).

4.1. Reliability and internal consistency

ICC analyses showed perfect correlation between intra- and inter-observer tests for both ICARS-T and SARA-T (Tables 4 and 5). The Cronbach's α coefficients used to evaluate the internal consistency were found to be 0.922 and 0.921 for SARA (reviewer 1 and reviewer 2, respectively) and 0.952 and 0.952 for ICARS-T (reviewer 1 and reviewer 2, respectively).

Table 1
Demographic data and physical characteristics of patients.

Age (years)[a]	34.75 ± 9.36
Disease duration (year)[a]	7.33 ± 7.10
Gender F:M[b]	39 (48%)/41 (51%)
EDSS, Median (interquartile range)	3,5 (3–4)
Type of MS (RR/PP/SP)[b]	54 (67.5%)/13 (16.3%) /13 (16.3%)

F: Female; M: Male; RR: Relapsing Remitting; PP: Primary Progressive; SP: Secondary Progressive; SD: Standard Deviation.
[a] Expressed as mean ± SD.
[b] Expressed as number of patients (%).

Table 2
Floor - ceiling effect of ICARS.

Icars		Mean	Std. Deviation	p value
icars item 1	Floor (n = 22)	0,95	0,79	< 0.001
	Ceiling (n = 22)	3,68	1,52	
icars item 2	Floor (n = 22)	0,45	0,60	< 0.001
	Ceiling (n = 22)	2,36	0,85	
icars item 3	Floor (n = 22)	0,77	0,69	< 0.001
	Ceiling (n = 22)	2,64	1,18	
icars item 4	Floor (n = 22)	0,41	0,50	< 0.001
	Ceiling (n = 22)	1,91	1,02	
icars item 5	Floor (n = 22)	0,27	0,46	< 0.001
	Ceiling (n = 22)	2,18	1,10	
icars item 6	Floor (n = 22)	0,77	0,69	< 0.001
	Ceiling (n = 22)	2,73	1,03	
icars item 7	Floor (n = 22)	0,00	0,00	< 0.001
	Ceiling (n = 22)	0,50	0,80	
icars item 8 (right)	Floor (n = 22)	0,55	0,60	< 0.001
	Ceiling (n = 22)	1,73	0,77	
icars item 8(left)	Floor (n = 22)	0,59	0,50	< 0.001
	Ceiling (n = 22)	1,91	0,75	
icars item 9 (right)	Floor (n = 22)	0,50	0,51	< 0.001
	Ceiling (n = 22)	1,64	1,09	
icars item 9 (left)	Floor (n = 22)	0,59	0,50	< 0.001
	Ceiling (n = 22)	1,91	0,92	
icars item 10 (right)	Floor (n = 22)	0,64	0,49	< 0.001
	Ceiling (n = 22)	1,59	0,73	
icars item 10 (left)	Floor (n = 22)	0,73	0,46	< 0.001
	Ceiling (n = 22)	2,00	0,98	
icars item 11 (right)	Floor (n = 22)	0,77	0,43	< 0.001
	Ceiling (n = 22)	1,64	0,66	
icars item 11 (left)	Floor (n = 22)	0,91	0,43	< 0.001
	Ceiling (n = 22)	1,91	1,02	
icars item 12 (right)	Floor (n = 22)	0,50	0,60	< 0.001
	Ceiling (n = 22)	1,59	1,01	
icars item 12 (left)	Floor (n = 22)	0,41	0,59	< 0.001
	Ceiling (n = 22)	1,77	1,27	
icars item 13 (right)	Floor (n = 22)	0,55	0,51	< 0.001
	Ceiling (n = 22)	1,36	0,73	
icars item 13 (left)	Floor (n = 22)	0,73	0,46	< 0.001
	Ceiling (n = 22)	1,91	1,02	
icars item 14	Floor (n = 22)	0,14	0,35	< 0.001
	Ceiling (n = 22)	1,45	1,22	
icars item 15	Floor (n = 22)	0,05	0,21	< 0.001
	Ceiling (n = 22)	1,45	0,80	
icars item 16	Floor (n = 22)	0,00	0,00	< 0.001
	Ceiling (n = 22)	1,09	0,68	
icars item 17	Floor (n = 22)	0,64	0,58	0,055
	Ceiling (n = 22)	0,95	0,49	
icars item 18	Floor (n = 22)	0,09	0,29	0,006
	Ceiling (n = 22)	0,45	0,51	
icars item 19	Floor (n = 22)	0,14	0,35	0,003
	Ceiling (n = 22)	0,55	0,51	

4.2. Structural validity

Factor analysis revealed that the ICARS-T sub-scales were determined by five factors with eigenvalues greater than 1 that explained 74% of the total variance. The Kaiser–Meyer–Olkin measure of sampling adequacy was 0.848 and the Bartlett test was significant ($p < 0.001$), indicating that both were favorable for the use of factor analysis.

According to the factor analyses, except item 7, the posture and gait disturbance sub-scale items formed factor 2. Item 7 shared the same factor as the kinetic function items (factor 1; items 10, 11, 12, 13). Kinetic functions related with the lower extremities (items 8 and 9) formed factor 3. Oculomotor disturbances and speech disturbances formed factor 4, but item 17 alone formed factor 5 (Table 6).

4.3. Hypothesis testing

BBS exhibited no significant correlation with SARA and ICARS-T (rho: –0.048 and –0.008, respectively). The EDSS total score had

Table 3
Floor- ceiling effect of SARA.

SARA		Mean	Std. Deviation	p value
sara item 1	Floor (n = 22)	1,18	0,66	< 0.001
	Ceiling (n = 22)	4,05	1,29	
sara item 2	Floor (n = 22)	0,91	0,61	< 0.001
	Ceiling (n = 22)	2,95	0,95	
sara item 3	Floor (n = 22)	0,00	0,00	< 0.001
	Ceiling (n = 22)	0,82	0,73	
sara item 4	Floor (n = 22)	0,00	0,00	< 0.001
	Ceiling (n = 22)	2,09	1,15	
sara item 5 (right)	Floor (n = 22)	0,73	0,46	< 0.001
	Ceiling (n = 22)	1,59	0,67	
sara item 5 (left)	Floor (n = 22)	0,68	0,48	< 0.001
	Ceiling (n = 22)	2,14	0,99	
sara item 6 (right)	Floor (n = 22)	0,91	0,53	< 0.001
	Ceiling (n = 22)	1,86	0,77	
sara item 6 (left)	Floor (n = 22)	0,91	0,43	< 0.001
	Ceiling (n = 22)	2,14	1,04	
sara item 7 (right)	Floor (n = 22)	0,59	0,50	< 0.001
	Ceiling (n = 22)	1,55	0,67	
sara item 7 (left)	Floor (n = 22)	0,64	0,49	< 0.001
	Ceiling (n = 22)	1,95	0,95	
sara item 8(right)	Floor (n = 22)	0,64	0,49	< 0.001
	Ceiling (n = 22)	1,73	0,63	
sara item 8 (left)	Floor (n = 22)	0,64	0,58	< 0.001
	Ceiling (n = 22)	2,00	0,82	

Table 4
Inter rater and test-retest reliability of ICARS.

	İnter-rater reliability	**Test-re test reliability**
Posture and gait disturbances	1.00**	1.00**
icars item 1	1.00**	1.00**
icars item 2	1.00**	1.00**
icars item 3	1.00**	1.00**
icars item 4	1.00**	1.00**
icars item 5	1.00**	1.00**
icars item 6	1.00**	1.00**
icars item 7	1.00**	1.00**
Kinetic functions	1.00**	1.00**
icars item 8 (right)	1.00**	1.00**
icars item 8 (left)	1.00**	1.00**
icars item 9 (right)	1.00**	1.00**
icars item 9 (left)	1.00**	1.00**
icars item 10 (right)	1.00**	1.00**
icars item 10 (left)	0.962**	1.00**
icars item 11 (right)	1.00**	1.00**
icars item 11 (left)	1.00**	1.00**
icars item 12 (right)	1.00**	1.00**
icars item 12 (left)	1.00**	1.00**
icars item 13 (right)	1.00**	1.00**
icars item 13 (left)	1.00**	1.00**
icars item 14	1.00**	1.00**
Speech disorders	1.00**	1.00**
icars item 15	1.00**	1.00**
icars item 16	1.00**	1.00**
Oculomotor disorders	1.00**	1.00**
icars item 17	1.00**	1.00**
icars item 18	1.00**	1.00**
icars item 19	1.00**	1.00**
icars total score	1.00**	1.00**

** Intraclass correlation coefficient (ICC) is significant (p < 0.001).

significant correlations with both SARA and ICARS-T (rho: 0.557 and 0.707, respectively). C-FSS had moderate correlation with SARA and a high correlation with ICARS-T (rho: 0.469 and 0.653, respectively). In addition, SARA and ICARS had high correlation (rho: 0.807) (Table 7).

4.4. Predictive validity

According to the AUC value, ICARS-T is the best scale to discriminate mild and moderate ataxia (AUC: 0.875). Although lower than ICARS-T, SARA-T demonstrates good discriminant ability for difference in ataxia severity with the AUC value of 0,758. However, BBS had lower AUC values than ICARS-T and SARA-T (AUC:0.510) (Table 8). The lower AUC values of BBS, concurrent with the correlations and it means that BBS is obviously inadequate to describe the

Table 5
inter-rater reliability- test re-test reliability of SARA.

	Inter-rater reliability	**Test- retest reliability**
sara item 1	1.00**	1.00**
sara item 2	1.00**	1.00**
sara item 3	1.00**	1.00**
sara item 4	1.00**	1.00**
sara item 5 (right)	0.964**	0.945**
sara item 5 (left)	0.990**	1.00**
sara item 6 (right)	1.00**	0.977**
sara item 6 (left)	0.982**	0.983**
sara item 7 (right)	0.985**	0.985**
sara item 7 (left)	0.990**	0.990**
sara item 8 (right)	1.00**	1.00**
sara item 8 (left)	1.00**	1.00**
sara total score	1.00**	1.00**

Intraclass correlation coefficient (ICC) is significant (p < 0.001).

Table 6
Factor analyses of the ICARS scale with varimax rotation.

	Factor 1	Factor 2	Factor 3	Factor 4	Factor 5
Eigenvalue	12.091	2.566	1652	1.388	1.022
Percent of variance	48.364	10.263	6.608	5.554	4.087
Cumulative of variance	48.364	58.627	65.235	70.789	74.876
icars item 1	,385	**,779**	,298	,120	−,013
icars item 2	,358	**,726**	,281	−,005	,042
icars item 3	,363	**,742**	,165	,162	,143
icars item 4	,358	**,759**	,129	,224	−,156
icars item 5	,423	**,812**	,169	,072	,153
icars item 6	,199	**,825**	,127	,063	,163
icars item 7	**,748**	,393	,012	,015	−,195
icars item 8 (right)	,208	,217	**,735**	,117	,280
icars item 8 (left)	,247	,289	**,722**	,209	,036
icars item 9 (right)	,149	,139	**,833**	,074	,097
icars item 9 (left)	,203	,192	**,838**	,130	−,123
icars item 10 (right)	**,711**	,175	,301	−,114	,318
icars item 10 (left)	**,702**	,391	,260	,158	,118
icars item 11 (right)	**,721**	,237	,187	,076	,079
icars item 11 (left)	**,832**	,252	,075	,162	−,033
icars item 12 (right)	**,844**	,262	,126	−,063	−,034
icars item 12 (left)	**,816**	,302	,197	,042	−,021
icars item 13 (right)	**,538**	,090	,274	,320	,193
icars item 13 (left)	**,643**	,345	,262	,199	−,032
icars item 14	,275	**,505**	**,410**	,257	,002
icars item 15	,293	**,513**	**,401**	**,515**	−,172
icars item 16	,129	**,536**	**,429**	**,518**	−,164
icars item 17	,037	,091	,099	,254	**,810**
icars item 18	,013	,195	,025	**,849**	,214
icars item 19	,081	,034	,252	**,844**	,132

Table 7
Spearman correlations of ataxia rating scales with BBS, EDSS and C-FSS.

	SARA	**ICARS**	**ICARS PG**	**ICARS KF**	**ICARS SD**	**ICARS OD**
C-FSS	0.469**	0.653**	0.598**	0.549**	0.481**	0203
BBS	−0.048	−0.008	−0.128	0.114	−0.024	−0.010
EDDS	0.557**	0.707**	0.760**	0.471**	0.590**	0.275*
SARA		0.807**	0.730**	0.678**	0.742**	0.430**

ICARS KF: ICARS kinetic functions, **ICARS PG:** ICARS posture and gait disturbances **ICARS SD:** ICARS Speech Disorders **ICARS OD:** ICARS Oculomotor Disorders.
* Spearman correlation is significant (p < 0.005).
** Spearman correlation is significant (p < 0.001).

Table 8
Predictive validity of balance and ataxia scales.

	Group 1 (n = 22)	Group 2 (n = 58)	p value	AUC
BBS total score	45.18 ± 12.15	44.98 ± 11.52	0.946	0.510
SARA total score	7.47 ± 5.18	11.07 ± 5.31	0.008	0.758**
ICARS total score	14.63 ± 5.87	28.81 ± 14.29	< 0.001	**0.875****

** Area under the ROC curve is significant (p < 0.001).

cerebellar symptoms.

5. Discussion

To our knowledge, this is the first study to investigate the validity and reliability of ICARS and SARA in MS patient and also the first application of both scales in a Turkish version. The present cross-sectional study results showed that the SARA-T and ICARS-T are reliable in MS patients with different severities of ataxia in clinical examination by different physiotherapists. Another important finding was that although the ICARS-T scale had some sub-scale structural problems, it is basically a valid scale for MS patients with ataxia. Both ICARS-T and SARA-T have convergent validity and good internal consistency. In addition to these findings, both scales are close in their performance to discriminate the severity of ataxia in MS.

The only existing sub-scale that briefly assesses the severity of ataxia in MS is C-FSS. Although the importance of detailed ataxia assessment is known, a novel scale has not been developed, nor have the existing scales been studied. Winser and colleagues published a study protocol and emphasized the need for comprehensive, optimal measures of ataxia in MS, but the results of the study following this protocol have not yet been published (Winser et al., 2014). Considering the need for a standardized scale in MS patients, SARA and ICARS appear to be an appropriate option that is frequently used in degenerative ataxias. The original validation of these scales were done in subjects with spinocerebellar ataxia of which the majority had non cerebellar symptoms to different extends, similar as in MS. This supports the use of a generic ataxia scale in the context of concomitant affection of other neurological systems. However, moderate and severe pyramidal symptoms interfere the kinetic function assessment. So it is difficult to obtain accurate results in patients who have predominantly pyramidal symptoms.

Moreover, the ICARS scale has not been translated into another language until now, although the SARA scale has been translated into many languages (Sato et al., 2009; Kim et al., 2014; Tan et al., 2013; Braga-Neto et al., 2010). Neto and colleagues developed a Portuguese version of SARA for hereditary ataxias and found no significant correlation between ICARS and SARA (Braga-Neto et al., 2010), while Song and colleagues found significant correlation in Chinese version of SARA and ICARS (Tan et al., 2013). In the present study, we found a significant correlation between the Turkish version of the scales but not a significant correlation with the BBS. Likewise, the C-FSS had a significant correlation with these scales, while did not have any correlation with the BBS. This indicates that the BBS may not be sufficiently sensitive to ataxic symptoms This insufficiency might be attributed to the floor effect with BSS.

We found significant correlation with the kinetic functions of ICARS-T and C-FSS. ICARS-T evaluates kinetic functions with seven different items, while C-FSS evaluates functions broadly with only three items. The major deficiency of the C-FSS relates to assessing tremor and dysmetry in the same items. Although we found significant correlation, according to our clinical experience, the use of ICARS-T will be more reliable, especially in research in which it is important to reveal subtle differences in kinetic functions. On the other hand, oculomotor disorders part of ICARS showed lowest correlation with C-FSS. This together with ceiling effects observed for ICARS item 17, may question oculomotor function for the assessment of ataxia in MS. Interestingly this coincidence with findings reported in the SARA original publication, where similar findings led to exclusion of oculomotor testing from the final version of the scale (Schmitz-Hubsch et al., 2006a).

Another important finding is that our factor analysis results did not coincide with the ICARS sub-scales. Kinetic functions were disrupted over three functions. These results are similar to the findings of Schmitz-Hübsch et al.'s study on spinocerebellar ataxia patients (Schmitz-Hubsch et al., 2006b). However, they identified four factors, unlike in our results. Similarly, Tison and colleagues found four factors in a study with multiple system atrophy patients (Tison et al., 2002). In our research, item 17 loaded differently and formed a single factor, so we found five factors. This structural problem was taken into consideration by researchers. Interestingly, item 17 showed a floor and ceiling effect. We attributed this to the fact that very few of our MS patients had severe nystagmus.

This study has some potential limitations. We found excellent correlation between raters for both ICARS-T and SARA-T assessments. This may be the consequences of the raters' competence in ataxia rehabilitation. They both hold PhD degrees in neurological rehabilitation, and they have been working together for 10 years. Also raters were acquainted the English version of the scale. If the proficiency of the researchers were different, the correlation could possibly change. In future studies, the results should be strengthened by comparing different health professionals' scores (neurologists/occupational therapists) and various competencies (junior/senior). Another limitation is that we selected ataxic patients according to their C-FSS assessment. It would be better if the patients were divided into the groups according to their neurological involvement such as ataxic, spastic, sensory. But spastic patients could not perform the kinetic functions tests. For this reason, we had to exclude high pyramidal involvement.

The results of this study will be useful for both clinicians and researchers. It is very difficult to evaluate ataxic symptoms in MS patients with various neurological symptoms in addition to ataxia. The use of BBS alone leads to insufficient examination of MS patients with ataxia. Despite some deficiencies, it is appropriate to use of ICARS and SARA until new, optimized MS-specific ataxia rating scales have been developed. In future studies, the performance of MS –specific ataxia rating scales should be improved by considering carefully spasticity and strength loss. It is very important to assess ataxia despite these pyramidal symptoms.

Declaration of interest

The authors report no conflicts of interest. The authors certify that no funding has been received for the conduct of this study and/ or preparation of this manuscript

Appendix A. Supplementary material

Supplementary data associated with this article can be found in the online version at http://dx.doi.org/10.1016/j.msard.2017.09.032.

References

Altman, D.G., 1990. Practical Statistics for Medical Research. Taylor & Francis.
Ashizawa, T., Xia, G., 2016. Ataxia. Continuum (Minneap Minn). **22**(4Movement Disorders): p. 1208–1226.
Bastian, A.J., 1997. Mechanisms of ataxia. Phys. Ther. 77 (6), 672–675.
Braga-Neto, P., et al., 2010. Translation and validation into Brazilian version of the Scale of the Assessment and Rating of Ataxia (SARA). Arq. Neuropsiquiatr. 68 (2), 228–230.
Cattaneo, D., Regola, A., Meotti, M., 2006. Validity of six balance disorders scales in persons with multiple sclerosis. Disabil. Rehabil. 28 (12), 789–795.
Cattaneo, D., Jonsdottir, J., Repetti, S., 2007. Reliability of four scales on balance disorders in persons with multiple sclerosis. Disabil. Rehabil. 29 (24), 1920–1925.

Ghez, C, T.W., The cerebellum in Principles of Neural Science, Fourth Edition, Kandel, E., Schwartz, J., Jessell, T., Editors, 2000, McGraw-Hill Companies, Incorporated.
Kesselring, J., Beer, S., 2005. Symptomatic therapy and neurorehabilitation in multiple sclerosis. Lancet Neurol. 4 (10), 643–652.
Kim, B.R., et al., 2014. Korean version of the scale for the assessment and rating of ataxia in ataxic stroke patients. Ann. Rehabil. Med. 38 (6), 742–751.
Klockgether, T., 2010. Sporadic ataxia adult onset: classification and diagnostic criteria. Lancet Neurol. 9 (1), 94–104.
Kurtzke, J.F., 1983. Rating neurologic impairment in multiple sclerosis: an expanded disability status scale (EDSS). Neurology 33 (11), 1444–1452.
Lynch, D.R., et al., 2006. Measuring Friedreich ataxia: complementary features of examination and performance measures. Neurology 66 (11), 1711–1716.
Mariotti, C., Fancellu, R., Di Donato, S., 2005. An overview of the patient with ataxia. J. Neurol. 252 (5), 511–518.
Mokkink, L.B., et al., 2010. The COSMIN checklist for assessing the methodological quality of studies on measurement properties of health status measurement instruments: an international Delphi study. Qual. Life Res. 19 (4), 539–549.
du Montcel, S.T., et al., 2008. Composite cerebellar functional severity score: validation of a quantitative score of cerebellar impairment. Brain 131 (5), 1352–1361.
Nachbauer, W., Eigentler, A., Boesch, S., 2015. Acquired ataxias: the clinical spectrum, diagnosis and management. J. Neurol. 262 (5), 1385–1393.
Polman, C.H., et al., 2005. Diagnostic criteria for multiple sclerosis: 2005 revisions to the "McDonald Criteria". Ann. Neurol. 58 (6), 840–846.
Sato, K., et al., 2009. Reliability of the Japanese version of the scale for the Assessment and Rating of Ataxia (SARA). Brain Nerve 61 (5), 591–595.
Schmahmann, J.D., et al., 2009. Development of a Brief Ataxia Rating Scale (BARS) Based on a Modified Form of the ICARS. Mov. Disord. 24 (12), 1820–1828.
Schmitz-Hubsch, T., et al., 2006a. Scale for the assessment and rating of ataxia: development of a new clinical scale. Neurology 66 (11), 1717–1720.
Schmitz-Hubsch, T., et al., 2006b. Reliability and validity of the International Cooperative Ataxia Rating Scale: a study in 156 spinocerebellar ataxia patients. Mov. Disord. 21 (5), 699–704.
Siva, A., et al., 1999. Frontiers in Multiple Sclerosis. Martin Dunitz.
Swingler, R.J., Compston, D.A., 1992. The morbidity of multiple sclerosis. Q. J. Med. 83 (300), 325–337.
Tallaksen, C.M., 2008. Hereditary ataxias. Tidsskr. Nor. Laege. 128 (17), 1977–1980.
Tan, S., et al., 2013. Reliability and validity of the Chinese version of the Scale for Assessment and Rating of Ataxia. Chin. Med. 126 (11), 2045–2048.
Tison, F., et al., 2002. Application of the International Cooperative Ataxia Scale rating in multiple system atrophy. Mov. Disord. 17 (6), 1248–1254.
Todd, P.K., Taylor, J.P., The Aetiologic Spectrum of Cerebellar Ataxia: Acquired Causes of Ataxia, in eLS. 2001, John Wiley & Sons, Ltd.
Trouillas, P., et al., 1997. International Cooperative Ataxia Rating Scale for pharmacological assessment of the cerebellar syndrome. The Ataxia Neuropharmacology Committee of the World Federation of Neurology. J. Neurol. Sci. 145 (2), 205–211.
Weinshenker, B.G., Issa, M., Baskerville, J., 1996. Long-term and short-term outcome of multiple sclerosis: a 3-year follow-up study. Arch. Neurol. 53 (4), 353–358.
Winser, S., et al., 2014. Balance assessment in multiple sclerosis and cerebellar ataxia: rationale, protocol and demographic data. Phys. Med. Rehabil. Int. 1 (5), 6.