

Brief Ataxia Rating Scale: A Reliable Tool to Rate Ataxia in a Short Timeframe

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The worldwide prevalence of late onset ataxia is approximately 10.2 of 100,000 people.¹ Several scales have been developed to rate ataxias. The International Cerebellar Ataxia Rating Scale (ICARS) and the Scale for Assessment and Rating of Ataxia (SARA) are the two best-known, validated, and broadly used rating scales. There is apparently no clear advantage of one in relation to the other.² ICARS and SARA are long and time-consuming, and they become impractical in a wider clinical-practice context. Schmahmann et al.³ developed the Brief Ataxia Rating Scale (BARS), a shorter scale created from a MICARS, a modified ICARS that includes seven more tests in kinetic function, speech disorders, and oculomotor function. The objectives of this study were to translate MICARS and BARS in Brazilian Portuguese and to validate them by correlating with SARA in a heterogeneous group of patients with cerebellar ataxia.

Methods

The first step of this study was to translate both MICARS and BARS into Brazilian Portuguese. SARA has been previously validated in this language.⁴ Both MICARS and BARS were translated by two movement disorders specialists, who were native speakers of Brazilian Portuguese and were fluent in English. One consensus translation was performed based on the two previous independent translations (S.C., R.A.). The consensus translation was submitted to a back translation (R.M.). Consensus translation and back translation were compared with the original text and the necessary adjustments were made to create a comprehensive and verisimilar text (see Appendix S1). We then enrolled 35 consecutive ataxia patients from the UFMG Movement Disorders Clinic. All MICARS, SARA, and BARS tests were video recorded and two independent raters scored all patients. Statistical analyses were performed using IBM SPSS Statistics software package (version

23.0). Pearson correlation was used to measure the correlation between scales. As described by Schmahmann et al.,³ we also tested whether the five items of BARS correlated with MICARS minus those five items. Cronbach's alpha was used to test the internal consistency of the scores. Intraclass correlation coefficient (ICC) was used to evaluate the inter-rater reliability of MICARS and BARS. Bland-Altman plots were used to calculate the mean bias and the 95% limits of agreement between raters in both scales. The ethics committee of the institution approved the study, and all patients signed the informed consent.

Results

Of 35 patients, 18 were female (51%). Age at assessment varied from 16 to 73 years (44.8 ± 15.7 years) and age at onset was 5 to 72 years (35.9 ± 17.1 years). Disease duration ranged from 1 to 43 years (8.9 ± 8.6 years). A total of 65.7% were diagnosed with hereditary ataxia (SCA2 and 3, mitochondrial ataxia, Friedreich ataxia, and unknown recessive ataxia), 25.7% were diagnosed with autoimmune disorders, vasculopathies, multiple system atrophy, vitamin deficiency, and alcoholic cerebellar degeneration. Undetermined causes accounted for 8.6% of patients. Scores from [MICARS] correlated with [BARS] ($r = 0.93$; $P = 0.0001$) and [MICARS minus BARS] highly correlated with [BARS] ($r = 0.90$; $P < 0.0001$). SARA correlated with both BARS ($r = 0.91$) and MICARS ($r = 0.97$). Internal consistency (Cronbach's alpha) was 0.934 for MICARS and 0.972 for BARS. ICC was 0.98 for MICARS and 0.94 for BARS. The Bland-Altman plots of inter-rater agreement for MICARS and BARS are shown in Figures 1 and 2. Scores tended to distribute symmetrically around the mean difference of scores, and only two cases were outside the limits of agreement for each scale.

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Additional Supporting Information may be found online in the supporting information tab for this article.

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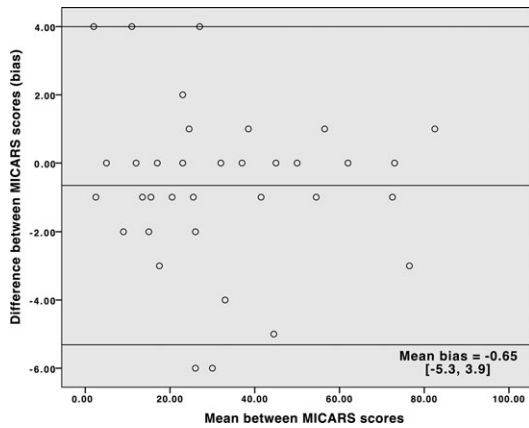


Figure 1 Bland-Altman Plot for MICARS. The lines represent the mean bias and the 95% limits of agreement.

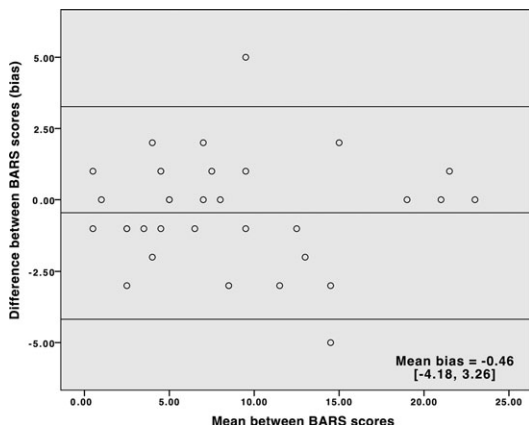


Figure 2 Bland-Altman Plot for BARS. The lines represent the mean bias and the 95% limits of agreement.

Discussion

Following scale translation into Brazilian Portuguese, we were able to demonstrate that the brief scale (BARS) highly correlates with MICARS and SARA with an elevated internal consistency and optimal reliability. BARS has some advantages: it is easier to learn and the score can estimate the overall cerebellar motor function quickly. We estimate that whereas it takes 15 to 20 minutes to perform MICARS or SARA, BARS can be performed in 3 to 5 minutes. One theoretical disadvantage of BARS, however, is that for research purposes, it can be less sensitive to detect minimal differences in scores, because it is less detailed than MICARS.

The relatively small sample is a limitation of this study. However, BARS inter-rater reliability and validity have been demonstrated in other studies with SARA or ICARS totaling 146 patients (50 with SCA7, 44 children with brain tumor, and 52 healthy children).^{5–7} BARS has also been used as the sole scale in few other studies so far, totaling 176 patients (17 with Niemann-Pick, 12 with SCA2, and 148 with multiple sclerosis).^{8–10}

We recommend BARS for use in clinical practice as a reliable tool to evaluate ataxia effectively and rapidly.

Author Roles

1. Research Project: A. Conception, B. Organization, C. Execution; 2. Statistical Analysis: A. Design, B. Execution, C. Review and Critique; 3. Manuscript Preparation: A. Writing of the First Draft, B. Review and Critique.

S.C.: 1A, 1B, 2B, 3A

F.C.: 1A, 2B, 3B

R.M.: 1B, 1C, 3B

L.H.: 1C, 2A

T.R.S.: 1C

V.G.C.: 1C

R.A.: 1C

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Appendix S1. Brief Ataxia Rating Scale Translation to Brazilian Portuguese.

Supporting Information

Supporting information may be found in the online version of this article.