Published in final edited form as:
Mov Disord. 2009 September 15; 24(12): 1820–1828. doi:10.1002/mds.22681.

Development of a Brief Ataxia Rating Scale (BARS) Based on a Modified Form of the ICARS

Jeremy D. Schmahmann, MD[*], **Raquel Gardner, MD**, **Jason MacMore, BA**, and **Mark G. Vangel, PhD**
Department of Neurology, Massachusetts General Hospital and Harvard Medical School, Boston, Massachusetts, USA

Abstract

To develop a brief ataxia rating scale (BARS) for use by movement disorder specialists and general neurologists. Current ataxia rating scales are cumbersome and not designed for clinical practice. We first modified the International Cooperative Ataxia Rating Scale (ICARS) by adding seven ataxia tests (modified ICARS, or MICARS), and observed only minimally increased scores. We then used the statistics package R to find a five-test subset in MICARS that would correlate best with the total MICARS score. This was accomplished first without constraints and then with the clinical constraint requiring one test each of Gait, Kinetic Function-Arm, Kinetic Function-Leg, Speech, and Eye Movements. We validated these clinical constraints by factor analysis. We then validated the results in a second cohort of patients; evaluated inter-rater reliability in a third cohort; and used the same data set to compare BARS with the Scale for the Assessment and Rating of Ataxia (SARA). Correlation of ICARS with the seven additional tests that when added to ICARS form MICARS was 0.88. There were 31,481 five-test subtests (48% of possible combinations) that had a correlation with total MICARS score of ≥0.90. The strongest correlation of an unconstrained five-test subset was 0.963. The clinically constrained subtest validated by factor analysis, BARS, had a correlation with MICARS-minus-BARS of 0.952. Cronbach alpha for BARS and SARA was 0.90 and 0.92 respectively; and inter-rater reliability (intraclass correlation coefficient) was 0.91 and 0.93 respectively. BARS is valid, reliable, and sufficiently fast and accurate for clinical purposes. © 2009 Movement Disorder Society

Keywords

ataxia; dysmetria; rating scale; assessment; cerebellum

Quantitation of motor deficit in ataxic disorders is a prerequisite for measuring clinical severity of disease in neurology clinics and in research studies. Two scales currently in use for the quantitation of motor deficit in ataxic disorders are the International Cooperative Ataxia Rating Scale (ICARS)[1] and the Scale for the Assessment and Rating of Ataxia (SARA).[2] The ICARS, a 19-item, 100-point scale has been validated[3] and used in studies of

[*]Correspondence to: Dr. Jeremy D. Schmahmann, Department of Neurology, Massachusetts General Hospital, Suite 340, Charles River Plaza South, 55 Fruit Street, Boston, Massachusetts 02114. jschmahmann@partners.org.

Potential conflict of interest: Nothing to report.

Author Roles

Jeremy D, Schmahmann, M.D. contributed to conception and design, data acquisition and analysis, drafting, editing and revising the text. Raquel Gardner, M.D. contributed to data acquisition and anlysis, and editing and revising the text. Jason MacMore, B.A. contributed to data acquisition and analysis, technical support, and revising the text. Mark G. Vangel, Ph.D. contributed to concept and design, statistical analysis, drafting, editing and revising the text.

hereditary spinocerebellar ataxia and idiopathic late onset ataxia,[4] stroke and tumor[5] among others. It has been shown to be a useful tool for assessing and monitoring cerebellar motor function,[6] but it has been criticized for containing redundancies,[7] for being insensitive to noncerebellar aspects of the neurological examination in disorders such as Friedreich ataxia[8] and multiple system atrophy,[9] and its length limits its use in the routine clinical setting. The more recently introduced SARA is an 8-item, 40-point scale that has been validated in studies of spinocerebellar ataxias.[10, 11] This shorter scale can still take from 5 to 40 minutes to administer,[2] and it includes redundancies while excluding oculomotor assessment.

We therefore set about developing a Brief Ataxia Rating Scale (BARS) that could be administered in the clinical setting rapidly and without special appliances or equipment.

METHODS

General Approach

The approach we adopted was to determine, first, whether adding traditional tests of cerebellar function to ICARS would increase the likelihood of detecting the cerebellar motor syndrome (thus, the Modified ICARS [MICARS]). We then examined whether we could derive a 5-item subset of tests (BARS) from MICARS that would correlate strongly with the MICARS, and replace it in the clinical setting.

Patient Population

In the Ataxia Unit of the Massachusetts General Hospital we (JDS) examined 91 patients with cerebellar disorders using the MICARS over a period of 3 years. The MICARS scores in these patients (Group 1) constituted the Initial Data Set. We used the Initial Data Set to develop the BARS, as we describe below.

We next validated the BARS in the examination (JDS) of a second cohort of 32 patients, nine of whom were examined more than once separated by intervals of at least 6 months, for a total of 41 assessments (Group 2). A medical student research assistant (RCG) was trained to perform the rating scales, and she independently evaluated a third cohort of 29 patients (Group 3). Sixteen of these patients were examined by JDS within 3 months of the assessment by RCG, and were used to determine reliability of the different test instruments. Both clinicians (JDS and RCG) also performed ataxia evaluations on 35 healthy controls.

Test Procedures

The 120-point MICARS was developed by adding seven additional tests (for the extra 20 points) in the categories of Kinetic function, Speech disorders and Oculomotor function (see Table 1). These tests were administered as described in the Appendix.

Data Analysis

The degree to which the seven additional supposed tests of cerebellar motor function predicted the total ICARS score was tested by correlation analysis using the statistic program R.[12] Paired *t* tests were then used to compare MICARS score (as a percentage) with ICARS score for individual patients. Scores were further analyzed for the categories in which new tests were added (Kinetic, Speech, and Oculomotor functions), normalizing the scores for each category.

We then investigated, using R, whether there was a subset of five tests that would correlate most closely with the score of the total MICARS minus those five tests. On the first pass there were no constraints on which five tests could be selected. On the second pass, we constrained the analysis by clinical criteria such that each of the five categories were

represented by one test, that is, Gait and Posture; Kinetic Function— Arm; Kinetic Function —Leg; Speech; and Oculomotor function. To assess the validity of this clinically constrained analysis, we then used factor analysis to determine in an independent manner whether there was any grouping of five factors within the Initial Data Set that would match this group of five clinically constrained categories.

After we defined the five-test subset that would constitute the BARS, we prospectively validated the findings from the Initial Data Set by performing MICARS (with ICARS and BARS embedded within it) in a separate cohort of patients in Group 2. We examined the degree to which scores on the BARS correlate with those of the MICARS-minus-BARS. For each of the tests evaluated (ICARS, the seven additional tests added to ICARS to constitute MICARS, and the five-test subset that constitutes the BARS), 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 the tests by comparing scores on the same patients by both raters.

Finally, we applied Cronbach alpha and ICC analyses to SARA. This was possible because the eight tests that constitute SARA are all included in MICARS, with the exception that SARA does not consider eye movements. (SARA assesses Gait, equivalent to MICARS test number 1, Stance—MICARS test number 3; Sitting—MICARS 7; Finger chase—MICARS 17; Finger to Nose—MICARS 13; Rapid alternating movements—MICARS 15; Heel to Shin—MICARS 8; Speech—any one of MICARS 19, 20, or 21).

This study was approved by the Institutional Review Board of the Massachusetts General Hospital.

RESULTS

All 35 healthy subjects score between 0 and 4 on the MICARS. A wide variety of patients with cerebellar disorders was examined (Table 2). In the Initial Data Set, the correlation of the seven additional tests (that when added to ICARS constitutes MICARS) with ICARS was r = 0. 88 for Group 1; r = 0.85 for Group 2; and r = 0.84 for Group 3. The mean ± standard deviation of the total score for the MICARS (as percentage) in Group 1 (32.0 ± 18.8) was no different than that for ICARS (28.8 ± 18.2), P = 0.25. Percentage subscores of the tests added to make MICARS were compared to the subscore tests in ICARS by normalizing for each subtest: Percentage subscores differed for Kinetic Function—Leg, MICARS 34.7 ± 22.2, ICARS 27.0 ± 22.0 ($P < 0.0001$); Kinetic Function—Arm, MICARS 29.0 ± 17.4, ICARS 25.2 ± 16.8 ($P < 0.0001$); and Speech, MICARS 31.7 ± 20.9, ICARS 27.0 ± 19.1 ($P < 0.0001$). Oculomotor scores were similar, MICARS 46.4 ± 27.2, ICARS 46.3 ± 26.0 ($P = 0.63$). Gait and equilibrium tests were not amended in MICARS.

There are 26 tests in the MICARS, so the number of possible five-test subtests is equal to the number of combinations of 26 objects taken five at a time without regard to order, which equals 65,780. In the Initial Data Set, there were 31,481 subsets of five tests (48%) with a correlation coefficient ≥0.90 between the sum of scores of the five-test subset combinations and the total MICARS score minus that five-test subset. The strongest five-test subset correlation with MICARS minus the five-test subset was 0.963; the 1000th strongest was 0.945.

When the data were constrained by requiring that the five-test subset combination include one test from each of the major categories assessed, the sum of the scores for tests 1, 8, 12, 20, and 24 produced a correlation with MICARS minus that subset of 0.952. This five-test subset tested walking capacity, heel-to-shin test for decomposition of movement, finger to nose test for decomposition and dysmetria, dysarthria assessed by clarity of speech, and

abnormalities of the ocular pursuit system. These five tests formed the basis of the BARS (Table 3). In the validation cohorts the correlation of BARS with MICARS-minus-BARS was 0.94 (Group 2) and 0.92 (Group 3).

To provide an independent test of the validity of our selection of the five-test subset based on clinical criteria, we applied factor analysis to MICARS. The Initial Data Set resolved into five factors; they conformed closely to the five groupings identified by clinical criteria (Table 4). Factor 1 included tests 1 through 6 that assess gait. Tests 12, 13, and 14 measure upper extremity dysmetria and formed Factor 2. Speech impairment measured by tests 19, 20, and 21 were the basis for Factor 3. Tests 22, 23, and 24 evaluate eye movements and contributed to Factor 4. And Factor 5 included tests 8 and 9 that measure lower extremity incoordination.

The internal consistency (Cronbach's alpha) of the evaluation instruments for Groups 1, 2, and 3 was ICARS: 0.96, 0.95, and 0.96, the seven additional tests that when added to ICARS constitute the MICARS: 0.86, 0.80, and 0.80; these additional tests analyzed but with oculomotor findings excluded: 0.91, 0.91, and 0.86; MICARS: 0.95, 0.93, and 0.96; and BARS 0.90, 0.89, and 0.86.

The reliability of the evaluation instruments assessed by the ICC on the 16 patients examined independently by both clinicians was ICARS: 0.92; the seven additional tests that when added to ICARS form the MICARS: 0.93; and BARS: 0.91.

SARA was subjected to the same statistical challenges. This was possible because all the components of SARA are contained within MICARS. Cronbach's alpha of internal consistency for SARA for Groups 1, 2, and 3 was 0.92, 0.90, and 0.89. The ICC of interrater reliability was 0.93.

DISCUSSION

Objective, observer-independent, and quantifiable tests of ataxia are sorely needed, but until such tests are developed and validated, clinical rating scales of ataxia remain the principal measure of the severity of cerebellar motor incapacity. Earlier clinicians[13–16] introduced the bedside tests for ataxia, dysmetria, dysarthria and oculomotor abnormalities. ICARS does not include some of these tests that are routinely used in the clinical setting (decomposition of movement and rapid tapping of the legs, overshoot and rebound in the upper extremities, impaired rapid alternating syllables, and oculomotor abnormalities at rest, with saccade, and with the vestibulo-ocular reflex cancellation test; see Table 2 and Appendix). The results of our study show that these additional tests correlate well with ICARS total score, indicating that they indeed test cerebellar motor function. Further, MICARS scores (as percent) for the amended categories are higher than ICARS scores, perhaps because these additional tests provide further opportunity for detecting cerebellar motor abnormalities, and while partially redundant, they probe these cerebellar features in different ways that appear to be clinically meaningful. The internal consistency of ICARS and MICARS are extremely high (Cronbach's alpha 0.96 and 0.95 respectively in Group 1). This speaks to the great redundancy in the tests. The seven additional tests have lower values, however, (Cronbach's alpha 0.86 to 0.80), and we show that this is because they include oculomotor assessment.

ICARS and MICARS are lengthy, cumbersome and impractical in the routine clinical setting. Here we show for the first time the astonishing result that there are literally thousands (≥31,000) of possible combinations of five-test subtests within MICARS that correlate extremely well (≥0.90) with the total MICARS score. This establishes with certainty that it is possible to derive a test of ataxia that is short, accurate, and easy to administer, and second, that there are a multitude of possible correct solutions when seeking

a five-test subset that would reflect the total score. So which to choose? We elected to constrain the five-test subset by ensuring that it includes aspects of the cerebellar motor exam known for over a century to be important: Gait, Kinetic function—Arms, Kinetic function—Legs, Speech, and Oculomotor function. We validated this selection independently using factor analysis. The five factors matched the clinically determined groupings precisely. To our knowledge, this is the first time that clinical determinations of the early neurologists have been put to the test and affirmed in this independent manner. Further, the high ICC for the BARS attests to its reliability in clinical practice, although we may have over-estimated inter-rater reliability because the second rater was personally instructed by the first. This will need to be re-evaluated in future studies when the learning of the BARS scoring system is made more generally available through instruction video or in written descriptions.

Our observation that quantifying oculomotor impairments reduces internal consistency in the seven additional tests of the MICARS is consistent with the conclusions of earlier investigators.[7, 10] In contrast to the determination by these investigators that oculomotor assessments be excluded from ataxia rating scales, however, we view this result in a positive light, and believe the data speak to the exact opposite conclusion. The lower internal consistency with inclusion of the eye movement evaluation indicates that the scores of extremity coordination, gait, and articulation cannot accurately predict the oculomotor score. Moreover, our data show that even with the eye movement score included, BARS correlation with MICARS is still extremely tight. Further, we believe that the clinical justification for inclusion of eye movement evaluation in an ataxia rating scale outweighs any putative statistical rationale for its exclusion. The BARS (Table 3) reflects this with eye movements scored for abnormalities of pursuit and saccade, as well as for nystagmus. The syndrome of isolated downbeat nystagmus underscores the inadequacy of ignoring eye movements in a cerebellar movement disorder rating scale, and the slowing of eye movements seen in some spinocerebellar ataxias mandates inclusion of this parameter as well. The factor analysis provides further independent support for this assertion.

The derivation of the BARS and its component tests as described here has relevance for other scoring systems, including SARA. The eight motor functions evaluated by SARA are all contained within MICARS, except that eye movements are excluded, and the assessment of sitting, which is included in SARA has low utility.[11] SARA thus contains tests that are redundant or have low utility, while excluding potentially important findings. It has more tests than BARS and takes longer to perform, but it is not appreciably better than BARS in its correlation with ICARS or its inter-rater reliability. SARA may be insufficiently discriminative for comprehensive observations in clinical trials when compared to MICARS, and unnecessarily redundant as well as unjustifiably restrictive for routine clinical use when compared to BARS.

The assessment of each motor domain is necessary because there is topography of motor function in the cerebrocerebellar system.[17–27] BARS focuses on each of the five major motor domains (coordination of gait, arm, leg, speech, and eye movements), and thus the resulting score provides an estimate of overall cerebellar motor function. BARS provides a quantitative measure of the neurological examination that is performed routinely in every cerebellar patient, and it takes no extra time to perform. It can be readily learned and applied to the clinical assessment of ataxia. BARS affords the movement disorder specialist, and indeed the general neurologist, the opportunity to quantitate ataxia in the clinic and at essentially no time cost—an advance over the current state of “mild, moderate, and severe”. It is not clear whether BARS is sufficiently reliable for the fine-grained analysis required in research settings, but since it is embedded within ICARS, it is possible to analyze this

retrospectively and in a prospective manner. Future studies of intraand inter-rater reliability of BARS will be important to establish its widespread utility.

Acknowledgments

Supported in part by RO1 MH067980, and the Birmingham Foundation. Presented in part to the 59th annual meeting of the American Academy of Neurology, Boston, March 2007. The statistical analysis was performed principally by Mark Vangel, PhD.

APPENDIX

Tests Added to ICARS to Constitute the MICARS

1. Decomposition of leg movement is assessed with the draw-a-circle test. The patient lies on the examination table and elevates each leg in turn, the leg held straight out from the hip at an angle of ~30°, tracing a circle in the air of ~2 feet in diameter. If the traced circle is degraded by edges or corners (resembling a hexagon) the effort is graded 1. Marked decomposition of the circle consisting of jagged lines or chaotic motion is scored 2.
2. Integrity of leg tapping is assessed with the patient standing on one leg while holding onto a firm structure such as the examination table, and tapping the heel of the other foot on the ground in as regular and rapid a motion as possible. Mild irregularity in rate, rhythm and force of tapping, or mild slowing, is graded 1. Attempted foot tapping that is markedly slow, with poor regularity and variable force is graded 2.
3. Overshoot is measured with the patient's arm extended towards the examiner's finger, and is scored according to the degree to which the patient overshoots the end-point after the examiner has rapidly moved the finger in the horizontal or vertical planes. Overshoot of less than 10 cm is scored 1, greater than 10 cm scored 2.
4. Rebound is measured with the patient's arms extended straight in front with palms facing up. The patient is requested to maintain the arms in that position. A quick downward displacement of the limb by the examiner followed by upward rebound above the starting position scores 1 when the rebound is less than 10 cm, and 2 when it is greater than 10 cm.
5. Dysarthria is assessed by the ability to produce conversational speech as well as lingual, palatal and buccal consonants rapidly and smoothly. Examples include buttercup, pa-ta-ka, and la-la-la, me-me-me, and go-go-go. Speech and/or consonants that are mildly slow and abnormal score 1, whereas more clearly abnormal and severely slow responses score 2.
6. The eyes should be quiet at rest, and the presence of abnormalities is scored as either present or absent. These findings may include, for example, square wave jerks, nystagmus in primary position, and opsoclonus.
7. The presence of saccadic intrusions into the vestibulo-ocular reflex (VOR) may be detected at the bedside by asking the patient to hold the arms outstretched in front with the thumbs pointing upwards. The patient is asked to focus on the thumbs, while the torso and arms are rotated passively by the examiner holding the patient at the shoulders and rotating about the axis, while observing the patient's eye movements. There should be essentially no motion of the eyes during this test. If saccadic eye movements are noted, then the cancellation of the VOR has failed, and the test is positive.

References

1. Trouillas P, Takayanagi T, Hallett M, et al. 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. 1997; 145:205–211.
2. Schmitz-Hubsch T, du Montcel ST, Baliko L, et al. Scale for the assessment and rating of ataxia: development of a new clinical scale. Neurology. 2006; 66:1717–1720. [PubMed: 16769946]
3. Storey E, Tuck K, Hester R, Hughes A, Churchyard A. Inter—rater reliability of the International Cooperative Ataxia Rating Scale (ICARS). Mov Disord. 2004; 19:190–192. [PubMed: 14978674]
4. Richter S, Dimitrova A, Maschke M, et al. Degree of cerebellar ataxia correlates with three-dimensional mri-based cerebellar volume in pure cerebellar degeneration. Eur Neurol. 2005; 54:23–27. [PubMed: 16088175]
5. Schoch B, Dimitrova A, Gizewski ER, Timmann D. Functional localization in the human cerebellum based on voxelwise statistical analysis: a study of 90 patients. Neuroimage. 2006; 30:36–51. [PubMed: 16253526]
6. Schoch B, Regel JP, Frings M, et al. Reliability and validity of ICARS in focal cerebellar lesions. Mov Disord. 2007; 22:2162–2169. [PubMed: 17712842]
7. Schmitz-Hubsch T, Tezenas du Montcel S, Baliko L, et al. Reliability and validity of the International Cooperative Ataxia Rating Scale: a study in 156 spinocerebellar ataxia patients. Mov Disord. 2006; 21:699–704. [PubMed: 16450347]
8. Ribaÿ P, Pousset F, Tanguy ML, et al. Neurological, cardiological, and oculomotor progression in 104 patients with Friedreich ataxia during long-term follow-up. Arch Neurol. 2007; 64:558–564. [PubMed: 17420319]
9. Tison F, Yekhlef F, Balestre E, et al. Application of the International Cooperative Ataxia Scale rating in multiple system atrophy. Mov Disord. 2002; 17:1248–1254. [PubMed: 12465064]
10. Subramony SH. SARA—a new clinical scale for the assessment and rating of ataxia. Nat Clin Pract Neurol. 2007; 3:136–137. [PubMed: 17290240]
11. Weyer A, Abele M, Schmitz-Hübsch T, et al. Reliability and validity of the scale for the assessment and rating of ataxia: a study in 64 ataxia patients. Mov Disord. 2007; 22:1633–1637. [PubMed: 17516493]
12. Development Core Team R: a language and environment for statistical computing . Vienna, Austria: R foundation for statistical computing; 2007. Available at www.r–project.org.
13. Brown S. On hereditary ataxia, with a series of twenty-one cases. Brain. 1892; 15:250–282.
14. Marie P. Sur l'hérédo-ataxie cérébelleuse. Semaine méd. 1893; 13:444–447.
15. Babinski, JFF. De l'asynergie cérébelleuse. Rev Neurol. 1899; 7:806–816.
16. Holmes G. The symptoms of acute cerebellar injuries due to gunshot wounds. Brain. 1917; 40:461–535.
17. Bolk, L. Das cerebellum der säugetiere. Haarlem: De Erven F. Bohn; 1906.
18. Snider RS. Recent contributions to the anatomy and physiology of the cerebellum. Arch Neurol Psychiatr. 1950; 64:196–219.
19. Chambers WW, Sprague JM. Functional localization in the cerebellum. I. Organization in longitudinal corticonuclear zones and their contribution to the control of posture, both extrapyramidal and pyramidal. J Comp Neurol. 1955; 103:105–130. [PubMed: 13263445]
20. Chambers WW, Sprague JM. Functional localization in the cerebellum. II. Somatotopic organization in cortex and nuclei. Arch Neurol Psychiatr. 1955; 74:653–780. [PubMed: 13268132]
21. Oscarsson O. Functional organzation of the spino- and cuneocerebellar tracts. Physiol Rev. 1965; 45:495–522. [PubMed: 14337566]
22. Nitschke MF, Kleinschmidt A, Wessel K, Frahm J. Somatotopic motor representation in the human anterior cerebellum. A high resolution functional MRI study Brain. 1996; 119:1023–1029.
23. Schmahmann, JD.; Pandya, DN. The cerebrocerebellar system. In: Schmahmann, JD., editor. Int Rev Neurobiol. Vol. 41. San Diego: Academic Press; 1997. p. 31-60.The Cerebellum and Cognition

24. Grodd W, Hülsmann E, Lotze M, Wildgruber D, Erb M. Sensorimotor mapping of the human cerebellum: fMRI evidence of somatotopic organization. Hum Brain Mapp. 2001; 13:55–73. [PubMed: 11346886]

25. Schmahmann JD, MacMore J, Ko R. The human basis pontis. Clinical syndromes and topographic organization Brain. 2004; 127:1269–1291.

26. Schmahmann JD, Rosene DL, Pandya DN. The motor corticopontine projection in rhesus monkey. J Comp Neurol. 2004; 478:248–268. [PubMed: 15368534]

27. Stoodley CJ, Schmahmann JD. Functional topography in the human cerebellum: a meta-analysis of neuroimaging studies. Neuroimage. 2009; 44:489–501. [PubMed: 18835452]

TABLE 1

Modified International Cooperative Ataxia Rating Scale (MICARS)

I. Posture and gait disturbances

1.Walking capacities

0:Normal

1:Almost normal naturally, but unable to walk with feet in tandem position

2: Walking without support, but clearly abnormal and irregular

3: Walking without support but with considerable staggering; difficulties in half turn

4: Walking with autonomous support no longer possible; the patient uses the episodic support of the wall for 10-meter test

5: Walking only possible with one stick

6: Walking only possible with two special sticks or with a Stroller

7: Walking only with accompanying person

8: Walking impossible, even with accompanying person (wheelchair)

2. Gait speed

0: Normal

1: Slightly reduced

2: Markedly reduced

3: Extremely Slow

4: Walking with autonomous support no longer possible

3. Standing capacities, eyes open

0: Normal: able to stand on one foot more than 10 seconds

1: Able to stand with feet together, but no longer able to stand on one foot more than 10 seconds

2: Able to stand with feet together, but no longer able to stand with feet in tandem position

3: No longer able to stand with feet together, but able to stand in natural position without support, with no or moderate sway

4: Standing in natural position without support, with considerable sway and considerable corrections

5: Unable to stand in natural position without strong support of one arm

6: Unable to stand at all, even with strong support of two arms

4. Spread of feet in natural position without support, Eyes open

0: Normal (<10 cm)

1: Slightly enlarged (>10 cm)

2: Clearly enlarged (25 cm < spread < 35 cm)

3: Severely enlarged (>35 cm)

4: Standing in natural position impossible

5. Body sway with feet together, eyes open

0: Normal (<10 cm)

1: Oscillations

2: Moderate oscillations (<10 cm at the level of head)

3: Severe oscillations (>10 cm at the level of head), threatening the upright position

4: Immediate falling

6. Body sway with feet together, eyes closed

0: Normal (<10 cm)

1: Slight oscillations

2: Moderate oscillations (<10 cm at the level of head)

3: Severe oscillations (>10 cm at the level of head), threatening the upright position

4: Immediate falling

7. Quality of sitting position

0: Normal

1: With slight oscillations of the trunk

2: With moderate oscillations of the trunk and legs

3: With severe dysequilibirium

4: Impossible

II. Kinetic functions

8. Knee-tibia test (decomposition of movement and intention tremor) (Left and Right scored)

0: Normal

1: Lowering of heel in continuous axis, but the movement is decomposed in several phases, without real jerks, or abnormally slow

2: Lowering jerkily in the axis

3: Lowering jerkily with lateral movements

4: Lowering jerkily with extremely long lateral movements or test impossible

9. Action tremor in the heel-to-knee test (Left and Right scored)

0: Normal

1: Tremor stopping immediately when the heel reaches the knee

2: Tremor stopping <10 seconds after reaching the knee

3: Tremor continuing >10 seconds after reaching knee

4: Uninterrupted tremor or test impossible

10. Decomposition of leg movement (Left and Right scored)

0: Normal

1: Corners or edges on the circle

2: Markedly decomposed attempts at circle

11. Decomposition of leg tapping (Left and Right scored)

0: Normal

1: Slightly slow and irregular

2: Clearly slow and irregular

12. Finger-to-nose test: decomposition and dysmetria (Left and Right scored)

0: Normal

1: Oscillating movement without decomposition of the movement

2: Segmented movement in 2 phases and/or moderate dysmetria in reaching nose

3: Segmented movement in more than 2 phases and/or considerable dysmetria in reaching nose

4: Dysmetria preventing the patient from reaching nose.

13. Finger-to-nose test: intention tremor of the finger (Left and Right scored)

0: Normal

1: Simple swerve of the movement

2: Moderate tremor with estimated amplitude <10 cm

3: Tremor with estimated amplitude between 10 cm and 40 cm.

4: Severe tremor with estimated amplitude >40 cm

14. Finger-finger test (action, tremor and/or instability) (Left and Right scored)

0: Normal

1: Mild instability

2: Moderate oscillations of finger with estimated amplitude <10 cm

3: Considerable oscillations of finger with estimated amplitude between 10 and 40 cm

4: Jerky movements >40 cm of amplitude

15. Pronation-supination alternating movements (Left and Right scored)

0: Normal

1: Slightly irregular and slowed

2: Clearly irregular, and slowed movement, but without elbow sway

3: Extremely irregular, and slowed, but with sway of the elbow

4: Movement completely disorganized or impossible

16. Rebound of the arms (Left and Right scored)

0: None

1: Less than 10 cm

2: Greater than 10 cm

17. Overshoot of the arms (Left and Right scored)

0: None

1: Less than 10 cm

2: Greater than 10 cm

18. Drawing of Archimedes' spiral on a predrawn pattern

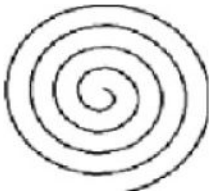
0: Normal

1: Impairment and decomposition, the line quitting the pattern slightly, but without hypermetric swerve

2: Line completely out of the pattern with recrossings and/or hypermetric swerves

3: Major disturbances due to hypermetria and decomposition

4: Drawing completely disorganized or impossible



III. Speech disorders

19. Dysarthria: fluency of speech

0: Normal

1: Mild modification of fluency

2: Moderate modification of fluency

3: Considerable slow and dysarthric speech

4: No speech

20. Dysarthria: Clarity of speech

0: Normal

1: Suggestion of slurring

2: Definite slurring, most words understandable

3: Severe slurring, speech not understandable

4: No speech

21. Dysarthria: Alternating syllables

0: Normal

1: Slightly irregular

2: Clearly irregular, dysrhythmic and slurred

IV. Oculomotor disorders

22. Abnormal eye movements at rest

0: Absent

1: Present

23. Gaze-evoked nystagmus

0: Normal

1: Transient

2: Persistent but moderate

3: Persistent and severe

24. Abnormalities of the ocular pursuit

0: Normal

1: Slightly saccadic

2: Clearly saccadic

25. Dysmetria of the saccade

0: Absent

1: Bilateral clear overshoot or undershoot of the saccade

26. Saccadic intrusions into vestibulo-ocular reflex cancellation

0: Absent

1: Present

International Cooperative Ataxia Rating Scale (ICARS) tests and measures of severity are shown in regular font. The seven additional tests which when added form the Modified ICARS (MICARS) are shown in bold font.

TABLE 2

Diagnoses of patients examined in each group

Group 1	
ILOCA	22
Cerebellar stroke	12
MSAc	8
Friedreich's ataxia	6
Post infectious cerebellitis	6
ADCA	6
Nonprogressive cerebellar ataxia	6
SCA 6	4
SCA 7	4
SCA 3	3
SCA 8	3
Adrenoleukodystrophy	2
Alcohol related cerebellar degeneration, cerebellar agenesis, gliadin associated ataxia, histiocytosis of cerebellum, Lhermitte-Duclos, post-tumor resection, SCA 1, SCA 2, superficial siderosis	1 each
Group 2	
Cerebellar stroke	13
ILOCA	6
MSAc	2
ADCA	2
Cerebellar atrophy with hypogonadotrophichypogonadism, Gerstmann-Straussler-Scheinker disease, gliadin associated ataxia, measles cerebellitis, nonprogressive cerebellar ataxia, SCA 2, SCA 3, SCA 6, superficial siderosis	1 each
Group 3	
MSAc	13
ILOCA	10
Behcet's disease, Friedreich's ataxia, familial recessive ataxia, gliadin associated ataxia, SCA 1, SCA 7	1 each

ADCA, autosomal dominant cerebellar ataxia; ILOCA, idiopathic late onset cerebellar ataxia; MSAc, multiple system atrophy, cerebellar type; SCA, spinocerebellar ataxia.

TABLE 3

The Brief Ataxia Rating Scale (BARS)

Gait
0: Normal
1: Almost normal naturally, but unable to walk with feet in tandem position
2: Walking without support, but clearly abnormal and irregular
3: Walking without support but with considerable staggering; difficulties in half turn
4: Walking without support not possible; uses support of the wall for 10-meter test.
5: Walking possible only with one cane
6: Walking possible only with two canes or with a stroller
7: Walking possible only with one accompanying person
8: Walking impossible with one accompanying person (2-person assist; wheelchair)
Knee-tibia test (decomposition of movement and intention tremor) (Left and Right scored)
0: Normal
1: Lowering of heel in continuous axis, but movement is decomposed in several phases, without real jerks, or abnormally slow
2: Lowering jerkily in the axis
3: Lowering jerkily with lateral movements
4: Lowering jerkily with extremely long lateral movements, or test impossible
Finger-to-nose test (decomposition and dysmetria of arm and hand) (Left and Right scored)
0: Normal
1: Oscillating movement of arm and/or hand without decomposition of the movement
2: Segmented movement in 2 phases and / or moderate dysmetria in reaching nose
3: Segmented movement in more than 2 phases and / or considerable dysmetria in reaching nose
4: Dysmetria preventing the patient from reaching nose
Dysarthria
0: Normal
1: Mild impairment of rate/rhythm/clarity
2: Moderate impairment of rate/rhythm/clarity
3: Severely slow and dysarthric speech
4: Speech absent or unintelligible
Oculomotor abnormalities
0: Normal
1: Slightly slowed pursuit, saccadic intrusions, hypo/hypermetric saccade, nystagmus
2: Prominently slowed pursuit, saccadic intrusions, hypo/hypermetric saccade, nystagmus
TOTAL (out of 30)

TABLE 4

Principal component analysis of factor loading using the method of Varimax rotation for the 26 MICARS tests in the 91 patients examined in Group 1

Question	Factor 1	Factor 2	Factor 3	Factor 4	Factor 5
1	**0.785**		0.239	0.421	0.199
2	**0.745**	0.14	0.178	0.4	0.182
3	**0.741**	0.202	0.26	0.283	0.266
4	**0.647**		0.263	0.269	0.207
5	**0.763**	0.191	0.187	0.313	0.223
6	**0.671**	0.214		0.362	0.231
7	0.361	0.321	0.553	0.133	
8a	0.334	**0.488**		0.554	0.123
8b	0.31	**0.543**	0.108	0.529	
9a	0.115	**0.836**	0.227	0.148	0.125
9b	0.127	**0.829**	0.256	0.175	
10a	0.376	0.333	0.101	0.593	
10b	0.307	0.269		0.61	
11a	0.364	0.218		0.687	0.105
11b	0.447	0.262		0.593	
12a	0.343	0.142	**0.316**	0.597	0.3
12b	0.427	0.163	**0.338**	0.555	0.2
13a	0.326	0.124	**0.412**	0.535	0.267
13b	0.358	0.227	**0.38**	0.545	0.177
14a	0.164	0.247	**0.736**	0.291	0.134
14b	0.152	0.191	**0.745**	0.291	
15a	0.243	0.131	0.331	0.699	0.178
15b	0.284	0.17	0.303	0.598	
16a	0.189	0.29	0.145	0.441	0.164
16b	0.227	0.325	0.195	0.403	
17a	0.144		0.189	0.631	0.229
17b	0.293	0.133	0.132	0.599	0.107
18	0.306	0.136	0.44	0.541	0.13
19	0.231		0.317	**0.68**	0.235

Question	Factor 1	Factor 2	Factor 3	Factor 4	Factor 5
20	0.107		0.256	**0.77**	0.227
21	0.219		0.207	**0.741**	0.275
22	0.212		0.129	0.151	**0.55**
23	0.109				**0.697**
24	0.175	0.114		0.393	**0.754**
25	0.152		0.17	0.293	0.279
26	0.105	0.129	0.109	0.176	0.636

The letters a and b represent left and right sides respectively. Values in bold represent the MICARS tests that cluster together as a group.