

## ORGANIZATION NEWS

### Highlights From the Rehabilitation Measures Database

*This content is provided as a service by the American Congress of Rehabilitation Medicine and is not peer reviewed by the Archives.*

# Measurement Characteristics and Clinical Utility of the International Cooperative Ataxia Rating Scale in Individuals With Hereditary Ataxias

Maryleen K. Jones, PT, MPT, CLT, NCS, Stephanie A. Combs-Miller, PT, PhD, NCS

The International Cooperative Ataxia Rating Scale (ICARS), developed by the Ataxia Neuropharmacology Committee of the World Federation of Neurology,<sup>1</sup> can be used to quantify the level of impairment related to hereditary ataxias, monitor response to pharmacological intervention, and plan clinical trials. The test can be completed in 15 to 30 minutes and is typically administered by a physician or physical therapist. Total scores range from 0 to 100, with higher scores representing greater impairment. A score of 0 represents normal, whereas a score of 100 represents maximum impairment. The scale consists of 19 items with 4 subscales including posture and gait disturbances (34 points), kinetic function (52 points), speech disorder (8 points), and oculomotor disorders (6 points). The ICARS has demonstrated reliability and validity in individuals with spinocerebellar disease,<sup>2</sup> Friedreich ataxia,<sup>3</sup> multisystem atrophy,<sup>4</sup> and chronic alcoholism.<sup>5</sup>

This Rehabilitation Measures Database summary provides a review of the psychometric properties of the ICARS with spinocerebellar disease, Friedreich ataxia, multisystem atrophy, and chronic alcoholism. A full review of the ICARS as well as reviews of more than 100 other instruments can be found at [www.rehabmeasures.org](http://www.rehabmeasures.org).


Please address correspondence to [rehabmeasures@ric.org](mailto:rehabmeasures@ric.org).

#### BIBLIOGRAPHY

1. Trouillas P, Takayanagi T, Hallett M, et al. International Cooperative Ataxia Rating Scale for pharmacological assessment of the cerebellar syndrome. *J Neurol Sci* 1997;145:205-211.
2. Schmitz-Hubsch T, Tezenas duMontcel 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.
3. Metz G, Coppard N, Cooper JM, et al. Rating disease progression of Friedreich's ataxia by the International Cooperative Ataxia Rating Scale: analysis of a 603-patient database. *Brain* 2013;136:259-268.
4. 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.
5. Fitzpatrick LE, Jackson M, Crowe SF. Characterization of cerebellar ataxia in chronic alcoholics using the International Cooperative Ataxia Rating Scale (ICARS). *Alcohol Clin Exp Res* 2012;36: 1942-1951.
6. Fahey MC, Corben L, Collins V, et al. How is disease progression in Friedreich's ataxia best measured? A study of four rating scales. *J Neurol Neurosurg Psychiatry* 2007;78:411-413.
7. Burk K, Malzig U, Wolf S, et al. Comparison of three clinical rating scales in Friedreich ataxia (FRDA). *Mov Disord* 2009;24:1779-1784.
8. Morales JA, Donis KC, Serrano-Munuera C, et al; Iberoamerican Multidisciplinary Network for the Study of Movement Disorders (RIBERMOV) Study Group. Ataxia rating scales—psychometric profiles, natural history and their application to clinical trials. *Cerebellum* 2012;11:488-504.

This instrument summary is designed to facilitate the selection of outcome measures by trained clinicians. The information contained in this summary represents a sample of the peer-reviewed research available at the time of this summary's publication. The information contained in this summary does not constitute an endorsement of this instrument for clinical practice. The views expressed are those of the summary authors and do not represent those of authors' employers, instrument owner(s), the *Archives of Physical Medicine and Rehabilitation*, the Rehabilitation Measures Database, the United States Department of Education or the Retirement Research Foundation. The information contained in this summary has not been reviewed externally.

The Rehabilitation Measures Database and Instrument Summary Tear-sheets are funded by the National Institute on Disability and Rehabilitation Research, United States Department of Education through the Rehabilitation Research and Training Center on Improving Measurement of Medical Rehabilitation Outcomes (H133B090024) and the Retirement Research Foundation (2011-027).

	<b>Measure Name:</b>	<b>Acronym:</b>	<b>Summary Author:</b>												
	<b>International Cooperative Ataxia Rating Scale</b>	ICARS	Jones, MK; Combs-Miller, SA												
<b>Population Reviewed:</b>	<b>Admin Time:</b>	<b>Items:</b>	<b>Score:</b>												
Spinocerebellar Disease; Friedreich's Ataxia; Multisystem Atrophy; Chronic Alcoholism	15 to 30 minutes	19	0/100 (min / max)												
<b>Purpose and Administration Instructions:</b>															
<ul style="list-style-type: none"> <li>Evaluates the level of ataxia-related impairment due to hereditary ataxias and chronic alcoholism.</li> <li>The patient is evaluated and a score quantified on an ordinal scale consisting of 19 items with four subscales for posture and gait disturbances (34 points), kinetic function, (52 points), speech disorder (8 points), and oculomotor disorders (6 points).</li> </ul>															
<b>Required Equipment:</b>															
Score sheet and pen; Pre-drawn pattern of Archimede's Spiral															
<b>Training:</b>															
Specialized training not required, but knowledge of movement impairments would be helpful for scoring accuracy															
<b>Validity:</b>		<b>Reliability:</b>													
<ul style="list-style-type: none"> <li><b>Predictive Validity:</b> <i>Adequate</i> in Friedreich's Ataxia.<sup>4</sup></li> <li><b>Concurrent Validity in Spinocerebellar Ataxia:</b> <ul style="list-style-type: none"> <li><i>Excellent</i> correlation to Barthel Index<sup>2</sup></li> <li><i>Adequate</i> correlation to disease duration<sup>2</sup></li> </ul> </li> <li><b>Construct Validity</b> <ul style="list-style-type: none"> <li><i>Adequate</i> correlation of ICARS score with Spinocerebellar Ataxia to disease staging by Klockgether &amp; Colleagues<sup>2</sup></li> <li><i>Adequate</i> correlation of number of years of heavy ETOH consumption to ICARS score<sup>5</sup></li> <li><i>Adequate</i> effect size for Friedreich's &amp; Spinocerebellar Ataxia.<sup>6,7</sup></li> </ul> </li> </ul>		<ul style="list-style-type: none"> <li><b>Test-retest Reliability:</b> <ul style="list-style-type: none"> <li><i>Excellent</i> in Spinocerebellar Ataxia<sup>2</sup></li> </ul> </li> <li><b>Inter/Intra-rater Reliability:</b> <ul style="list-style-type: none"> <li><i>Excellent</i> in Spinocerebellar Ataxia and Friedreich's Ataxia.<sup>2,3</sup></li> </ul> </li> <li><b>Internal Consistency:</b> <ul style="list-style-type: none"> <li><i>Adequate</i> in Spinocerebellar Ataxia<sup>2</sup></li> <li><i>Excellent</i> in Multiple System Atrophy Cerebellar Type<sup>4</sup></li> <li><i>Adequate</i> in Multiple System Atrophy Parkinson's Type.<sup>4</sup></li> <li><i>Poor</i> in Friedreich's Ataxia<sup>7</sup></li> </ul> </li> </ul>													
<b>Floor/Ceiling Effects for Friedreich's Ataxia:</b>		<b>Minimal Detectable Change (MDC):</b>													
<ul style="list-style-type: none"> <li>Adequate for ICARS total score: Floor effect 1% and ceiling effects 2%<sup>3</sup></li> <li>Adequate for subscales: Posture and Gait: 6% Ceiling Effect, Oculomotor: 10% Floor Effect<sup>3</sup></li> </ul>		Spinocerebellar Ataxia: 1.96 points <sup>2</sup>													
		<b>Standard Error of Measurement (SEM):</b>													
		Spinocerebellar Ataxia: 4.18 points <sup>2</sup>													
<b>Scoring:</b>															
All items are summed to calculate a total score; A higher score indicates decreased independence with ADL's and functional mobility. <sup>8</sup>															
<b>Considerations:</b>		<b>Abbreviations:</b>													
<ul style="list-style-type: none"> <li>Item ratings on subscales are not "qualified" with ratings being termed "slightly reduced", "markedly reduced" and "extremely slow." For consistency, users may want to operationally define these terms.</li> <li>Further research is needed to determine the sensitivity of the ICARS over a respective assessment period.</li> <li>Further research is needed to validate the use of the ICARS in other neurological populations that experience ataxia, such as Multiple Sclerosis, Stroke, Traumatic Brain Injury, Neoplastic Brain Injury and Spinal Cord Injury.</li> </ul>		ADL's : Activities of Daily Living													
		<b>Cut-off Criteria:</b>													
		<table border="1"> <thead> <tr> <th></th> <th><i>r</i></th> <th>ICC</th> </tr> </thead> <tbody> <tr> <td><b>Excellent</b></td> <td>≥ .6</td> <td>≥ .75</td> </tr> <tr> <td><b>Adequate</b></td> <td>.31-.59</td> <td>.40 -.74</td> </tr> <tr> <td><b>Poor</b></td> <td>≤ .3</td> <td>&lt; .4</td> </tr> </tbody> </table>			<i>r</i>	ICC	<b>Excellent</b>	≥ .6	≥ .75	<b>Adequate</b>	.31-.59	.40 -.74	<b>Poor</b>	≤ .3	< .4
	<i>r</i>	ICC													
<b>Excellent</b>	≥ .6	≥ .75													
<b>Adequate</b>	.31-.59	.40 -.74													
<b>Poor</b>	≤ .3	< .4													