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

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The International Cooperative Ataxia Rating Scale (ICARS), developed by the Ataxia Neuropharmacology Committee of the World Federation of Neurology, 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, Friedreich ataxia, multisystem atrophy, and chronic alcoholism.

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.

Please address correspondence to rehabmeasures@ric.org.

BIBLIOGRAPHY

**Measure Name:** International Cooperative Ataxia Rating Scale  
**Acronym:** ICARS  
**Summary Author:** Jonas, MK; Combé-Miller, SA

<table>
<thead>
<tr>
<th>Population Reviewed:</th>
<th>Admin Time: 15 to 30 minutes</th>
<th>Items: 19</th>
<th>Score: 0/100 (min / max)</th>
</tr>
</thead>
</table>

**Purpose and Administration Instructions:**
- Evaluates the level of ataxia-related impairment due to hereditary ataxias and chronic alcoholism.
- 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).

**Required Equipment:**
Score sheet and pen; Pre-drawn pattern of Archimede’s Spiral

**Training:**
Specialized training not required, but knowledge of movement impairments would be helpful for scoring accuracy

**Validity:**
- **Predictive Validity:** Adequate in Friedreich’s Ataxia
- **Concurrent Validity in Spinocerebellar Ataxia:**
  - Excellent correlation to Barthel Index
  - Adequate correlation to disease duration
- **Construct Validity**
  - Adequate correlation of ICARS score with Spinocerebellar Ataxia to disease staging by Klockgether & Colleagues
  - Adequate correlation of number of years of heavy ETOH consumption to ICARS score
  - Adequate effect size for Friedreich’s & Spinocerebellar Ataxia

**Floor/Ceiling Effects for Friedreich’s Ataxia:**
- Adequate for ICARS total score: Floor effect 1% and ceiling effects 2%
- Adequate for subscales: Posture and Gait: 6% Ceiling Effect, Oculomotor: 10% Floor Effect

**Reliability:**
- **Test-retest Reliability:**
  - Excellent in Spinocerebellar Ataxia
- **Intra-rater Reliability:**
  - Excellent in Spinocerebellar Ataxia and Friedreich’s Ataxia
- **Internal Consistency:**
  - Adequate in Spinocerebellar Ataxia
  - Excellent in Multiple System Atrophy Cerebellar Type
  - Adequate in Multiple System Atrophy Parkinson’s Type
  - Poor in Friedreich’s Ataxia

**Minimal Detectable Change (MDC):**
Spinocerebellar Ataxia: 1.96 points

**Standard Error of Measurement (SEM):**
Spinocerebellar Ataxia: 4.18 points

**Scoring:**
All items are summed to calculate a total score; A higher score indicates decreased independence with ADL’s and functional mobility.

**Considerations:**
- 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.
- Further research is needed to determine the sensitivity of the ICARS over a respective assessment period.
- 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.

**Abbreviations:**
ADL’s : Activities of Daily Living

**Cut-off Criteria:**
<table>
<thead>
<tr>
<th>Cut-off</th>
<th>$r$</th>
<th>ICC</th>
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</thead>
<tbody>
<tr>
<td>Excellent</td>
<td>$\geq .6$</td>
<td>$\geq .75$</td>
</tr>
<tr>
<td>Adequate</td>
<td>$.31-.59$</td>
<td>$.40-.74$</td>
</tr>
<tr>
<td>Poor</td>
<td>$\leq .3$</td>
<td>$&lt; .4$</td>
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www.archives-pmr.org