



**European  
Reference  
Network**

for rare or low prevalence  
complex diseases



**Network**

Neurological Diseases  
(ERN-RND)

# Scale for the assessment and rating of ataxia (SARA)

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# Introduction to the European Reference Network for Rare Neurological Diseases (ERN-RND):

ERN-RND is a European Reference Network established and approved by the European Union. ERN-RND is a healthcare infrastructure which focuses on rare neurological diseases (RND). The three main pillars of ERN-RND are (i) network of experts and expertise centres, (ii) generation, pooling and dissemination of RND knowledge, and (iii) implementation of e-health to allow the expertise to travel instead of patients and families.

ERN-RND unites 32 of Europe's leading expert centres in 13 Member States and includes highly active patient organizations. Centres are located in Belgium, Bulgaria, Czech Republic, France, Germany, Hungary, Italy, Lithuania, Netherlands, Poland, Slovenia, Spain and the UK.

The following disease groups are covered by ERN-RND:

- Ataxias and Hereditary Spastic Paraplegias
- Atypical Parkinsons' Disease
- Dystonia, Paroxysmal Disorder and Neurodegeneration with Brain Ion Accumulation
- Frontotemporal Dementia
- Huntingtons' Disease and other Chorea
- Leukodystrophies

Specific information about the network, the expert centres and the diseases covered can be found at the networks web site [www.ern-rnd.eu](http://www.ern-rnd.eu).

## Recommendation for clinical use:

**The European Reference Network for Rare Neurological Diseases strongly recommends the use of the Scale for the Assessment and Rating of Ataxia (SARA) as best clinical practice for the assessment and rating of Ataxia patients.**



#### Disclaimer:

Clinical practice guidelines, practice advisories, systematic reviews and other guidance published, endorsed or affirmed by ERN-RND are assessments of current scientific and clinical information provided as an educational service. The information (1) should not be considered inclusive of all proper treatments, methods of care, or as a statement of the standard of care; (2) is not continually updated and may not reflect the most recent evidence (new information may emerge between the time information is developed and when it is published or read); (3) addresses only the question(s) specifically identified; (4) does not mandate any particular course of medical care; and (5) is not intended to substitute for the independent professional judgement of the treating provider, as the information does account for individual variation among patients. In all cases, the selected course of action should be considered by the treating provider in the context of treating the individual patient. Use of the information is voluntary. ERN-RND provided this information on an “as is” basis, and makes no warranty, expressed or implied, regarding the information. ERN-RND specifically disclaims any warranties of merchantability or fitness for a particular use or purpose. ERN-RND assumes no responsibility for any injury or damage to persons or property arising out of or related to any use of this information or for any errors or omissions.

## **METHODOLOGY**

The endorsement process has been performed by the Disease group for Ataxia and Hereditary Spastic Paraplegias of ERN-RND.

### Disease group for Ataxia and Hereditary Spastic Paraplegias:

#### **Disease group coordinators:**

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#### **Disease group members:**

##### Healthcare professionals:

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Osredkar<sup>18</sup>; Massimo Pandolfo<sup>19</sup>; Joanna Pera<sup>20</sup>; Borut Peterlin<sup>18</sup>; Maria Salvadó<sup>5</sup>; Ludger Schöls<sup>14</sup>; Deborah Sival<sup>15</sup>; Matthis Synofzik<sup>14</sup>; Franco Taroni<sup>16</sup>; Sinem Tunc<sup>8</sup>; Bart van de Warrenburg<sup>21</sup>; Judith van Gaalen<sup>21</sup>; Martin Vyhnálek<sup>9</sup>; Michèl Willemsen<sup>21</sup>; Ginevra Zanni<sup>2</sup>; Judith Zima<sup>7</sup>; Alena Zumrová<sup>9</sup>

### Patient representatives:

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## Endorsement process:

- Mapping of used disease scales by disease group – June – December 2017
- Proposal for endorsement of rating scale by ERN-RND disease group coordinators – 15/05/2018
- Discussion in ERN-RND disease group during annual meeting – 08/06/2018
- Consent on endorsement of disease scale during ERN-RND annual meeting 2018 – 08/06/2018
- Consent on endorsement by whole disease group – 13/07/2018

## Scale



<p><b>1) Gait</b></p> <p>Proband is asked (1) to walk at a safe distance parallel to a wall including a half-turn (turn around to face the opposite direction of gait) and (2) to walk in tandem (heels to toes) without support.</p> <p><b>0 Normal, no difficulties in walking, turning and walking tandem (up to one misstep allowed)</b></p> <p><b>1 Slight difficulties, only visible when walking 10 consecutive steps in tandem</b></p> <p><b>2 Clearly abnormal, tandem walking &gt;10 steps not possible</b></p> <p><b>3 Considerable staggering, difficulties in half-turn, but without support</b></p> <p><b>4 Marked staggering, intermittent support of the wall required</b></p> <p><b>5 Severe staggering, permanent support of one stick or light support by one arm required</b></p> <p><b>6 Walking &gt; 10 m only with strong support (two special sticks or stroller or accompanying person)</b></p> <p><b>7 Walking &lt; 10 m only with strong support (two special sticks or stroller or accompanying person)</b></p> <p><b>8 Unable to walk, even supported</b></p>	<p><b>2) Stance</b></p> <p>Proband is asked to stand (1) in natural position, (2) with feet together in parallel (big toes touching each other) and (3) in tandem (both feet on one line, no space between heel and toe). Proband does not wear shoes, eyes are open. For each condition, three trials are allowed. Best trial is rated.</p> <p><b>0 Normal, able to stand in tandem for &gt; 10 s</b></p> <p><b>1 Able to stand with feet together without sway, but not in tandem for &gt; 10s</b></p> <p><b>2 Able to stand with feet together for &gt; 10 s, but only with sway</b></p> <p><b>3 Able to stand for &gt; 10 s without support in natural position, but not with feet together</b></p> <p><b>4 Able to stand for &gt;10 s in natural position only with intermittent support</b></p> <p><b>5 Able to stand &gt;10 s in natural position only with constant support of one arm</b></p> <p><b>6 Unable to stand for &gt;10 s even with constant support of one arm</b></p>
<p><b>Score</b></p>	<p><b>Score</b></p>
<p><b>3) Sitting</b></p> <p>Proband is asked to sit on an examination bed without support of feet, eyes open and arms outstretched to the front.</p> <p><b>0 Normal, no difficulties sitting &gt;10 sec</b></p> <p><b>1 Slight difficulties, intermittent sway</b></p> <p><b>2 Constant sway, but able to sit &gt; 10 s without support</b></p> <p><b>3 Able to sit for &gt; 10 s only with intermittent support</b></p> <p><b>4 Unable to sit for &gt;10 s without continuous support</b></p>	<p><b>4) Speech disturbance</b></p> <p>Speech is assessed during normal conversation.</p> <p><b>0 Normal</b></p> <p><b>1 Suggestion of speech disturbance</b></p> <p><b>2 Impaired speech, but easy to understand</b></p> <p><b>3 Occasional words difficult to understand</b></p> <p><b>4 Many words difficult to understand</b></p> <p><b>5 Only single words understandable</b></p> <p><b>6 Speech unintelligible / anarthria</b></p>



Score			Score		
<b>5) Finger chase</b> <b>Rated separately for each side</b> Proband sits comfortably. If necessary, support of feet and trunk is allowed. Examiner sits in front of proband and performs 5 consecutive sudden and fast pointing movements in unpredictable directions in a frontal plane, at about 50 % of proband's reach. Movements have an amplitude of 30 cm and a frequency of 1 movement every 2 s. Proband is asked to follow the movements with his index finger, as fast and precisely as possible. Average performance of last 3 movements is rated.			<b>6) Nose-finger test</b> <b>Rated separately for each side</b> Proband sits comfortably. If necessary, support of feet and trunk is allowed. Proband is asked to point repeatedly with his index finger from his nose to examiner's finger which is in front of the proband at about 90 % of proband's reach. Movements are performed at moderate speed. Average performance of movements is rated according to the amplitude of the kinetic tremor.		
<b>0 No dysmetria</b> <b>1 Dysmetria, under/ overshooting target &lt;5 cm</b> <b>2 Dysmetria, under/ overshooting target &lt; 15 cm</b> <b>3 Dysmetria, under/ overshooting target &gt; 15 cm</b> <b>4 Unable to perform 5 pointing movements</b>			<b>0 No tremor</b> <b>1 Tremor with an amplitude &lt; 2 cm</b> <b>2 Tremor with an amplitude &lt; 5 cm</b> <b>3 Tremor with an amplitude &gt; 5 cm</b> <b>4 Unable to perform 5 pointing movements</b>		
Score	Right	Left	Score	Right	Left
mean of both sides (R+L)/2			mean of both sides (R+L)/2		
<b>7) Fast alternating hand movements</b> <b>Rated separately for each side</b> Proband sits comfortably. If necessary, support of feet and trunk is allowed. Proband is asked to perform 10 cycles of repetitive alternation of pro- and supinations of the hand on his/her thigh as fast and as precise as possible. Movement is demonstrated by examiner at a speed of approx. 10 cycles within 7 s. Exact times for movement execution have to be taken.			<b>8) Heel-shin slide</b> <b>Rated separately for each side</b> Proband lies on examination bed, without sight of his legs. Proband is asked to lift one leg, point with the heel to the opposite knee, slide down along the shin to the ankle, and lay the leg back on the examination bed. The task is performed 3 times. Slide-down movements should be performed within 1 s. If proband slides down without contact to shin in all three trials, rate 4.		
<b>0 Normal, no irregularities (performs &lt;10s)</b> <b>1 Slightly irregular (performs &lt;10s)</b> <b>2 Clearly irregular, single movements difficult to distinguish or relevant interruptions, but performs &lt;10s</b> <b>3 Very irregular, single movements difficult to distinguish or relevant interruptions, performs &gt;10s</b> <b>4 Unable to complete 10 cycles</b>			<b>0 Normal</b> <b>1 Slightly abnormal, contact to shin maintained</b> <b>2 Clearly abnormal, goes off shin up to 3 times during 3 cycles</b> <b>3 Severely abnormal, goes off shin 4 or more times during 3 cycles</b> <b>4 Unable to perform the task</b>		
Score	Right	Left	Score	Right	Left



mean of both sides (R+L)/2		mean of both sides (R+L) / 2	
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## REFERENCE

T. Schmitz-Hübsch, S. Tezenas du Montcel, L. Baliko, et al.  
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# European Reference Networks

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Neurological Diseases  
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