



**HAL**  
open science

## Temporal dynamics of the Scale for the Assessment and Rating of Ataxia in Spinocerebellar ataxias.

Paul Moulaire, Pierre-Emmanuel Poulet, Emilien Petit, Thomas Klockgether, Alexandra Dürr, Tetsuo Ashizawa, Sophie Tézenas Du Montcel

► **To cite this version:**

Paul Moulaire, Pierre-Emmanuel Poulet, Emilien Petit, Thomas Klockgether, Alexandra Dürr, et al.. Temporal dynamics of the Scale for the Assessment and Rating of Ataxia in Spinocerebellar ataxias.. Ataxia Global Conference 2022, Nov 2022, Dallas, United States. hal-03831598v1

**HAL Id: hal-03831598**

**<https://hal.science/hal-03831598v1>**

Submitted on 24 Nov 2022 (v1), last revised 28 Oct 2022 (v2)

**HAL** is a multi-disciplinary open access archive for the deposit and dissemination of scientific research documents, whether they are published or not. The documents may come from teaching and research institutions in France or abroad, or from public or private research centers.

L'archive ouverte pluridisciplinaire **HAL**, est destinée au dépôt et à la diffusion de documents scientifiques de niveau recherche, publiés ou non, émanant des établissements d'enseignement et de recherche français ou étrangers, des laboratoires publics ou privés.



Distributed under a Creative Commons Attribution 4.0 International License

## **Temporal dynamics of the Scale for the Assessment and Rating of Ataxia in Spinocerebellar ataxias.**

### Background

SARA scale is the reference clinical scale to assess the severity of cerebellar ataxia. In a context of upcoming therapeutics trials, a reliable clinical outcome is needed to assess the efficiency of the treatments.

### Objectives

To precisely assess the temporal dynamics of SARA scale.

### Methods

We analyzed data from four cohorts (EUROSCA, RISCA, CRC-SCA, SPATAX) including 1210 participants and 4092 visits. The linearity of the progression and the variability was assessed with an ordinal Bayesian mixed effect model (Leaspy). We did sample size calculations for therapeutics trials with different scenarios to improve the responsiveness of the scale.

### Results

Seven of the eight different items had a non-linear progression. The speed of progression was different between most of the items with an average time for a one-point increase from 3.5 years [3.4;3.6] (median, 95% credible interval) for the fastest item to 11.4 [10.9;12.0] years. The total SARA score had a linear progression with an average time for a one-point increase of 0.95 [0.92;0.98] years. After removing the four last items and re-scaling all items from 0 to 4, variability increased and progression was slower, and thus would require larger sample size in a future therapeutic trial.

### Conclusion

Despite a heterogeneous temporal dynamic at the item level, the global progression of the SARA scale was linear. Changing the initial scale deteriorates the responsiveness. This new information about the temporal dynamic of the scale should help to design outcome of future clinical trials.

Authors:

Paul Moulairé<sup>1</sup>, MSc, Pierre Emmanuel Poulet<sup>2</sup>, MSc, Emilien Petit<sup>2</sup>, MSc, Thomas Klockgether<sup>3</sup>, MD, Alexandra Durr<sup>2</sup>, MD, PhD, Tetsudo Ashisawa<sup>4</sup>, MD, Sophie Tezenas du Montcel<sup>2</sup>, MD, PhD, for the READISCA consortium

Author affiliations :

<sup>1</sup>. Sorbonne Université, Paris Brain Institute, INSERM, Institut Pierre Louis d'Épidémiologie et de Santé Publique, INRIA, CNRS, APHP, 75013 Paris, France.

<sup>2</sup>. Sorbonne Université, Paris Brain Institute, INSERM, INRIA, CNRS, APHP, 75013 Paris, France.

<sup>3</sup>. German Center for Neurodegenerative Diseases (DZNE), 53127 Bonn, Germany.

<sup>4</sup>. Weill Cornell Medicine at The Houston Methodist Research Institute, Houston, TX 77030, USA.