



# The Spastic Paraplegia Rating Scale (SPRS)

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Endorsed by ERN-RND: 26.07.2018



# 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 Parkinsonism and genetic Parkinson's 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 Spastic Paraplegia Rating Scale (SPRS) as best clinical practice for the assessment and rating of patients with Spastic Paraplegia.**



#### 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:**

Caterina Mariotti<sup>16</sup>; Rebecca Schuele-Freyer<sup>14</sup>

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

##### Healthcare professionals:

Segolene Ayme<sup>1</sup>; Enrico Bertini<sup>2</sup>; Kristl Claeys<sup>3</sup>; Maria Teresa Dotti<sup>4</sup>; Alexandra Durr<sup>1</sup>; Antonio Federico<sup>4</sup>; Josep Gámez<sup>5</sup>; Paola Giunti<sup>6</sup>; David Gómez-Andrés<sup>5</sup>; Kinga Hadziev<sup>7</sup>; York Hellenbroich<sup>8</sup>; Jaroslav Jerabek<sup>9</sup>; Mary Kearney<sup>10</sup>; Jiri Klempir<sup>11</sup>; Thomas Klockgether<sup>12</sup>; Thomas Klopstock<sup>13</sup>; Norbert Kovacs<sup>7</sup>; Ingeborg Krägeloh-Mann<sup>14</sup>; Berry Kremer<sup>15</sup>; Alfons Macaya<sup>5</sup>; Bela Melegh<sup>7</sup>; Maria Judit Molnar<sup>8</sup>; Isabella Moroni<sup>16</sup>; Alexander Münchau<sup>8</sup>; Esteban Muñoz<sup>17</sup>; Lorenzo Nanetti<sup>16</sup>; Andrés Nascimento<sup>17</sup>; Mar O'Callaghan<sup>17</sup>; Damjan



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:

Lori Renna Linton<sup>10</sup>, Cathalijne van Doorne<sup>10</sup>

<sup>1</sup> Assistance Publique-Hôpitaux de Paris, Hôpital Pitié-Salpêtrière, France: Reference Centre for Rare Diseases 'Neurogenetics'; <sup>2</sup> Pediatric hospital Bambino Gesù, Rome, Italy; <sup>3</sup> University Hospitals Leuven, Belgium; <sup>4</sup> AOU Siena, Italy; <sup>5</sup> Hospital Universitari Vall d'Hebron, Spain; <sup>6</sup> University College London Hospitals NHS Foundation Trust, United Kingdom; <sup>7</sup> University of Pécs, Hungary; <sup>8</sup> Semmelweis University, Hungary; <sup>8</sup> Universitätsklinikum Schleswig-Holstein, Germany; <sup>9</sup> Motol University Hospital, Czech Republic; <sup>10</sup> Patient representative; <sup>11</sup> General University Hospital in Prague, Czech Republic; <sup>12</sup> Universitätsklinikum Bonn, Germany; <sup>13</sup> Klinikum der Universität München, Germany; <sup>14</sup> Universitätsklinikum Tübingen, Germany; <sup>15</sup> University Medical Center Groningen, Netherlands; <sup>16</sup> Foundation IRCCS neurological institute Carlo Besta – Milan, Italy; <sup>17</sup> Hospital Clínic i Provincial de Barcelona y Hospital de Sant Joan de Déu, Spain; <sup>18</sup> University Medical Centre Ljubljana, Slovenia; <sup>19</sup> Université libre de Bruxelles, Belgium; <sup>20</sup> University Hospital in Krakow, Poland; <sup>21</sup> Stichting Katholieke Universiteit, doing business as Radboud University Medical Center Nijmegen, Netherlands;

## 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



## REFERENCE

**The Spastic Paraplegia Rating Scale (SPRS): a reliable and valid measure of disease severity. R. Schüle, T. Holland-Letz, S. Klimpe et al. Neurology 2006; Aug 8;67(3):430-4.**

