



**European  
Reference  
Network**

for rare or low prevalence  
complex diseases

 **Network**  
Neurological Diseases  
(ERN-RND)

# Scales to measure Dystonia

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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 Parkinsonism and genetic 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 the following scale as best clinical practice for the assessment and rating of Dystonia.**



#### Disclaimer:

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## **METHODOLOGY**

The development of the Diagnostic Flowcharts for Dystonia was done by the Disease group for Dystonia, Paroxysmal Disorder and NBIA of ERN-RND. Scales used in the clinical practice of the disease group members were mapped, and the decision on which scale should be proposed was taken by anonymous majority voting.

### Disease group for Dystonia, Paroxysmal Disorder and NBIA:

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

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

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## Endorsement process:

- Mapping of used disease scales by disease group – 06 - 12/2018
- Survey to decide on scales by anonymous majority voting – 31.01. – 24.02.2019
- Consent on document by whole disease group – 26.03.2019



## Recommended scales:

Domain	Scale
Generalised dystonia	<p><a href="#">Fahn-Marsden Dystonia Rating Scale</a>, Burke RE, Fahn S, Marsden CD, Bressman SB, Moskowitz C, Friedman J. Validity and reliability of a rating scale for the primary torsion dystonias. <i>Neurology</i> 1985;35:73–77.</p>
Generalised dystonia in children	<p><a href="#">The Movement Disorder-Childhood Rating Scale</a>, Battini R, Sgandurra G, Petacchi E, Guzzetta A, Di Pietro R, Giannini MT, Leuzzi V, Mercuri E, Cioni G. Movement disorder-childhood rating scale: Reliability and validity. <i>Pediatr Neurol</i> 2008;39:259-265</p>
Blepharospasm	<p>Defazio G, Hallett, M, Jinnah HA, Stebbins GT, Gigante AF, Ferrazzano G, Conte A, Fabbrini G, Berardelli A. <a href="#">Development and Validation of a Clinical Scale for Rating the Severity of Blepharospasm</a>. <i>Mov Disord</i>. 2015 April; 30(4): 525–530.</p> <p>Battini R, Sgandurra G, Petacchi E, Guzzetta A, Di Pietro R, Giannini MT, Leuzzi V, Mercuri E, Cioni G. <a href="#">Movement disorder-childhood rating scale: Reliability and validity</a>. <i>Pediatr Neurol</i> 2008;39:259-265</p>
Cervical Dystonia	<p>Toronto Western Spasmodic Torticollis Rating Scale for cervical dystonia</p> <ul style="list-style-type: none"> <li>- Consky, E, Basinski, A, Belle, L, Ranawaya, R, and Lang, AE. <a href="#">The Toronto Western Spasmodic Torticollis Rating Scale (TWSTRS): assessment of validity and inter-rater reliability</a> (abstract). <i>Neurology</i>. 1990; 40: 445</li> <li>- Consky ES, Lang AE. Clinical assessments of patients with cervical dystonia. In: Jankovic J, Hallett M, eds. <i>Therapy with Botulinum Toxin</i>. New York, NY: Marcel Dekker, Inc.:1994;211-237</li> </ul>



Laryngeal Dystonia	<a href="#">Vocal Performance Questionnaire (VPQ)</a> , Carding PN, Horsley IA, Docherty GJ. A study of the effectiveness of voice therapy in the treatment of 45 patients with nonorganic dysphonia. J Voice. 1999; 13:72–104.
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