European Reference Networks

European Reference Network for rare or low prevalence complex diseases

Network: Neurological Diseases (ERN-RND)

Coordinator: Universitätsklinikum Tübingen — Deutschland

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 64 of Europe's leading expert centers as well as 4 affiliated partners in 24 member states and includes highly active patient organizations. Centers are located in Austria, Belgium, Bulgaria, Croatia, Cyprus, Czech Republic, Denmark, Estonia, Finland, France, Germany, Greece, Hungary, Ireland, Italy, Latvia, Lithuania, Luxembourg, Malta, Netherlands, Poland, Slovenia, Spain and Sweden.

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 Iron Accumulation
- Frontotemporal Dementia
- Huntington's Disease and other Chorea
- Leukodystrophies

Specific information about the network, the expert centers and the covered diseases can be found on the network's website ***www.ern-rnd.eu***.

Recommendation for clinical use:
The European Reference Network for Rare Neurological Diseases strongly recommends the use of the following scales as best clinical practice for the assessment and rating of Dystonia.

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 not 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 recommendations of clinical rating scales for Dystonia was developed by the Disease Group for Dystonia, Paroxysmal Disorders and NBIA of ERN-RND. Scales used in the clinical practice of the Disease Group members were mapped, and the decision on which scales should be proposed was taken by anonymous majority voting.

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Endorsement process:

- Mapping of used disease scales by disease group: June to December 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
- Endorsement on updated recommendations by whole disease group: 11.10.2024

RECOMMENDED SCALES

Domain	Scale
Generalised dystonia	**Fahn-Marsden Dystonia Rating Scale:** Burke RE, Fahn S, Marsden CD, Bressman SB, Moskowitz C, Friedman J. Validity and reliability of a rating scale for the primary torsion dystonias. Neurology 1985;35:73–77.
Generalised dystonia in children	**The Movement Disorder-Childhood Rating Scale:** 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. Pediatr Neurol 2008;39:259-265
Blepharospasm	Defazio G, Hallett, M, Jinnah HA, Stebbins GT, Gigante AF, Ferrazzano G, Conte A, Fabbrini G, Berardelli A. **Development and Validation of a Clinical Scale for Rating the Severity of Blepharospasm.** Mov Disord. 2015 April; 30(4): 525–530. 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**. Pediatr Neurol 2008;39:259-265
Cervical Dystonia *Please note:* *Regarding the specific rating scales for cervical dystonia, the Toronto Western Spasmodic Torticollis Rating Scale (TWSTRS) is the primary recommended scale for assessing cervical dystonia, especially for evaluating treatment outcomes such as botulinum toxin injections and deep brain stimulation in both clinical practice and trials, as it has been previously validated. Additionally, the Tsui scale is a reliable, brief, and easy-to-use tool that is also recommended for daily clinical routine.*	Toronto Western Spasmodic Torticollis Rating Scale for cervical dystonia - Consky, E, Basinski, A, Belle, L, Ranawaya, R, and Lang, AE. The **Toronto Western Spasmodic Torticollis Rating Scale (TWSTRS): assessment of validity and inter-rater reliability (abstract).** Neurology. 1990; 40: 445 - Consky ES, Lang AE. Clinical assessments of patients with cervical dystonia. In: Jankovic J, Hallett M, eds. **Therapy with Botulinum Toxin.** New York, NY: Marcel Dekker, Inc.:1994;211-237 - Jost WH, Hefter H, Stenner A, Reichel G. **Rating scales for cervical dystonia: a critical evaluation of tools for outcome assessment of botulinum toxin therapy.** J Neural Transm (Vienna). 2013 Mar;120(3):487-96. doi: 10.1007/s00702-012-0887-7. Tsui scale for cervical dystonia - Tsui JK, Eisen A, Stoessl AJ, Calne S, Calne DB. **Double-blind study of botulinum toxin in spasmodic torticollis**. Lancet. 1986 Aug 2;2(8501):245-7. doi: 10.1016/s0140-6736(86)92070-2. PMID: 2874278.

Laryngeal Dystonia	**Vocal Performance Questionnaire (VPQ):** 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.
Degree of Neurological Disability	**Modified Rankin Scale (mRS):** Rankin J. Cerebral Vascular Accidents in Patients over the Age of 60: II. Prognosis. Scottish Medical Journal. 1957;2(5):200-215.

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