



**European
Reference
Network**

for rare or low prevalence
complex diseases

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

Scales to measure Huntingtons' Disease and Choreas

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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 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.

Recommendation for clinical use:

The European Reference Network for Rare Neurological Diseases The European Reference Network for Rare Neurological Diseases strongly recommends the use the following scales as best clinical practice for the assessment and rating of Huntingtons' Disease and other choreas.

Disclaimer:

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METHODOLOGY

The disease group HD and Chorea decided to endorse the scales used by Enroll-HD (www.enroll-hd.org) for ERN-RND.

Disease group for HD and Chorea:

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Disease group members:

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Flowchart development process:

- Consent on endorsement of the following scales for HD and Chorea during ERN-RND annual meeting 2019 – 18/06/2019

Recommended scales:

Domain	Scale
Motor Assessments	Unified Huntington’s Disease Rating Scale (UHDRS) ‘99 Motor ¹
	UHDRS Diagnostic Confidence Index ¹
Functional Assessments	UHDRS ‘99 Total Functional Capacity ¹
	UHDRS ‘99 Functional Assessment Scale ¹
	UHDRS ‘99 Independence Scale ¹
Behavioral Assessment	Problem Behaviors Assessment-Short (PBA-s) ²
	Hospital Anxiety/ Depression Rating Scale (HADS) ³ & Snaith Irritability Scale (SIS) ⁴ , combined
	Columbia Suicide Severity Rating Scale (CSSRS) ⁵
	Symbol Digit Modality Test ¹
Cognitive Assessments	

	Stroop Color Naming ¹
	Stroop Word Reading ¹
	Categorical Verbal Fluency ⁶
	Stroop Interference ¹
	Trail Making A & B ⁷
	Letter Verbal Fluency ⁶
	Mini Mental State Examination (MMSE) ⁸
Global	Global Clinical Impression ⁹
Physiotherapy Outcome Measures	Timed Up and Go (TUG) ¹⁰
	30-second Chair Stand Test ¹¹
Quality of Life	Short Form Health Survey-12 (SF-12) ¹²
	Caregivers Quality of Life Questionnaire ¹³
Health Economics	Client Services Receipt Inventory (CSRI) ¹⁴
	Work Productivity and Activity Impairment-Specific Health Problem Questionnaire (WPAI-SHP) ¹⁵

References

¹ Unified Huntington's Disease Rating Scale: reliability and consistency. Huntington Study Group.(1996) Movement Disorders, 11(2): 136-42.

² McNally, G, Rickards, H, Horton, M et al. (2015) Exploring the Validity of the Short Version of the Problem Behaviours Assessment (PBA-s) for Huntington's disease: A Rasch Analysis. Journal of Huntington's disease, 4 (4). pp. 347-369.

³ <https://www.svri.org/sites/default/files/attachments/2016-01-13/HADS.pdf>

⁴Snaith RP, Constantopoulos AA, Jardine MY, McGuffin P. (1978) A clinical scale for the self-assessment of irritability. *British Journal of Psychiatry*,132:164–171.

⁵ <https://depts.washington.edu/ebpa/sites/default/files/C-SSRS-LifetimeRecent-Clinical.pdf>

⁶ Rosser A, Hodges JR (1994) Initial letter and semantic category fluency in Alzheimer's disease, Huntington's disease, and progressive supranuclear palsy. *Journal of Neurology, Neurosurgery, and Psychiatry* 57: 1389-1394.

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¹⁰ Podsiadlo D, Richardson S (1991) The timed “Up & Go”: a test of basic functional mobility for frail elderly persons. *J Am Gertr Soc* , 39:142-8.

¹¹ <https://www.sralab.org/rehabilitation-measures/30-second-sit-stand-test>

¹² Ware J, Kosinski M, Keller S (1996) A 12-Item Short-Form Health Survey: Construction of Scales and Preliminary Tests of Reliability and Validity, *Medical Care*. 34(3):220-33.

¹³ Weitzner, M A, Jacobsen, P B, Wagner, H, Friedland, J, & Cox, C (1999). The Caregiver Quality of Life Index–Cancer (CQOLC) Scale: development and validation of an instrument to measure quality of life of the family caregiver of patients with cancer. *Quality of Life Research*, 8, 55–63.

¹⁴ Chisholm D, Knapp M, Knudsen HC, Amaddeo F, Gaité L, van Wijngaarden B, EPSILON Study Group (2000) Client Socio-Demographic and Service Receipt Inventory - European Version: development of an instrument for international research. *EPSILON Study 5*. *British Journal of Psychiatry* 177 Supplement 39 28-33.

¹⁵ http://www.reillyassociates.net/WPAI_SHP.html