



Scales to measure HSP in children before adolescence

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 strongly recommends the use the following scale as best clinical practice for the assessment and rating of pre-adolescent patients with HSP.

Disclaimer:

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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:

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Disease group members:

Healthcare professionals:

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Osredkar¹⁸; Massimo Pandolfo¹⁹; Joanna Pera²⁰; Borut Peterlin¹⁸; Maria Salvadó⁵; Ludger Schöls¹⁴; Deborah Sival¹⁵; Matthis Synofzik¹⁴; Franco Taroni¹⁶; Sinem Tunc⁸; Bart van de Warrenburg²¹; Judith van Gaalen²¹; Martin Vyhnálek⁹; Michèl Willemsen²¹; Ginevra Zanni²; Judith Zima⁷; Alena Zumrová⁹

Patient representatives:

Lori Renna Linton¹⁰, Cathalijne van Doorne¹⁰

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Endorsement process:

- Mapping of used disease scales by disease group – June – November 2018
- Proposal for endorsement of rating scale by ERN-RND disease group coordinators – November 2018
- Discussion in ERN-RND disease group – 11/02/2019
- Consent on endorsement of disease scale – 11/02/2019

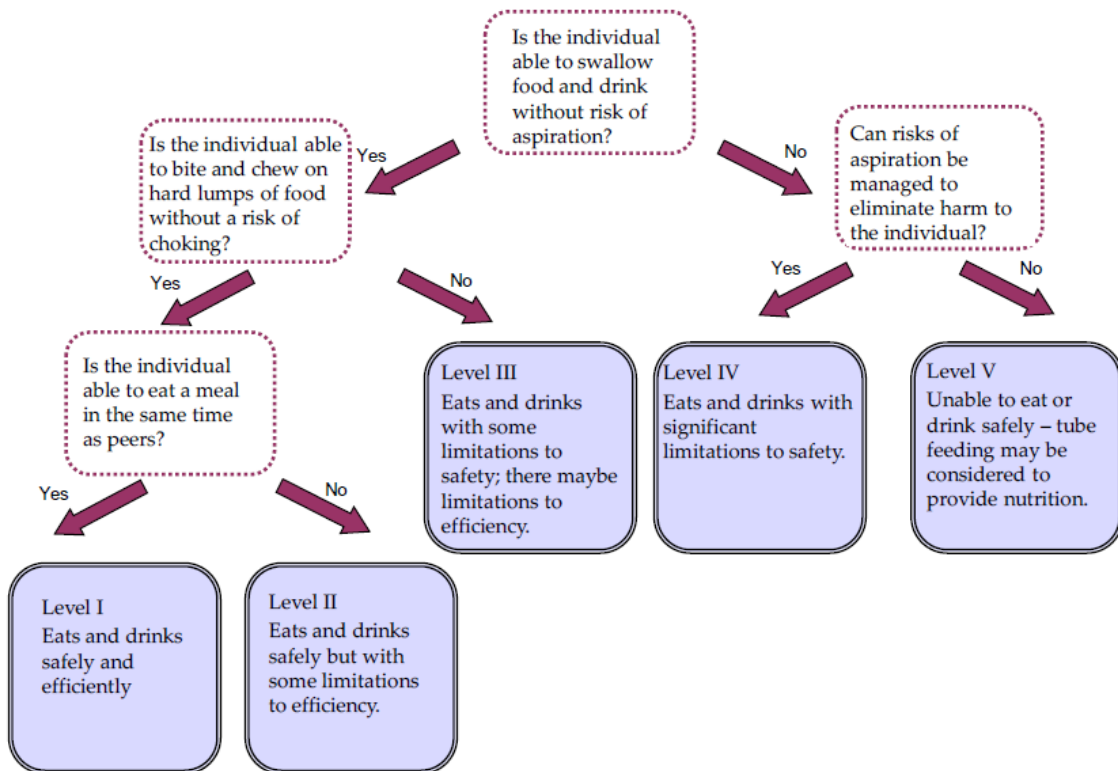
Recommended scales:

Domain	Scale
Gross motor abilities	GMFCS – Gross Motor Classification System Palisano et al. 1997, Dev Med Chil Neurol 39:214-23 CanChild: www.canchild.ca Illustrations Version 2 © Bill Reid, Kate Willoughby, Adrienne Harvey and Kerr Graham, The Royal Children’s Hospital Melbourne ERC151050
Fine motor abilities	MACS – Manual Ability Classification System For children <4 years: Mini MACS , Ann-Christin Eliasson, Lena Krumlinde-Sundholm, 2013 Contributors: Ann-Marie Öhrwall, Ulla Wahlström, Åsa Persson-Annerste
Language and communication	Viking Speech Scale , Lindsay Pennington, Tone Mjøen, Maria da Graça Andrada, Janice Murray, Viking Speech Scale, 2010©, Copyright © Newcastle University UK, Vestfold Hospital Trust Norway, Centro de Reabilitação de Paralisia Cerebral Calouste Gulbenkian-Lisbon and Manchester Metropolitan University UK ,2011.
Eating and drinking	EDACS - Eating and Drinking Ability Classification System , Sellers D, Mandy A, Pennington L, Hankins M, Morris C. Development and reliability of a system to classify the eating and drinking ability of people with cerebral palsy. Developmental medicine and child neurology 2014;56:245-51
Learning disability	<i>Apply scale used in daily routine, consider motor involvement.</i>
Urological issues	Dysfunctional voiding and incontinence symptoms score questionnaire , Akbal C, Genc Y, Burgu B, Ozden E, Tekgul S. Dysfunctional voiding and incontinence scoring system: quantitative evaluation of incontinence symptoms in pediatric population. The Journal of urology 2005;173:969-73.

EDACS - Eating and Drinking Ability Classification System

(<https://bit.ly/2Cy6E89>)

Eating and Drinking Ability Classification System - Algorithm



Other References

Wood E, Rosenbaum P. The gross motor function classification system for cerebral palsy: a study of reliability and stability over time. *Developmental medicine and child neurology* 2000;42:292-6.

Towns M, Rosenbaum P, Palisano R, Wright FV. Should the Gross Motor Function Classification System be used for children who do not have cerebral palsy? *Developmental medicine and child neurology* 2018;60:147-54.

Eliasson AC, Krumlinde-Sundholm L, Rosblad B, et al. The Manual Ability Classification System (MACS) for children with cerebral palsy: scale development and evidence of validity and reliability. *Developmental medicine and child neurology* 2006;48:549-54.

Eliasson AC, Ullenhag A, Wahlstrom U, Krumlinde-Sundholm L. Mini-MACS: development of the Manual Ability Classification System for children younger than 4 years of age with signs of cerebral palsy. *Developmental medicine and child neurology* 2017;59:72-8.

Pennington L, Virella D, Mjoen T, et al. Development of The Viking Speech Scale to classify the speech of children with cerebral palsy. *Research in developmental disabilities* 2013;34:3202-10.

Benfer KA, Weir KA, Bell KL, Ware RS, Davies PSW, Boyd RN. The Eating and Drinking Ability Classification System in a population-based sample of preschool children with cerebral palsy. *Developmental medicine and child neurology* 2017;59:647-54.