

Development and Validation of the Hereditary Spastic Paraplegia

Rating Scale (HSPRS)

R. Schüle¹, T. Holland-Letz², S. Klimpe³, J. Kassubek⁴, T. Klopstock⁵, V. Mall⁶, S. Otto⁷, B. Winner⁸, L. Schöls¹

¹ Hertie-Institute for Clinical Brain Research and Department of Neurology, Eberhard Karls-University Tübingen, ² Department of Medical Informatics, Biometry and Epidemiology, Ruhr-University Bochum, ³ Department of Neurology, Johannes Gutenberg University Mainz, ⁴ Department of Neurology, University of Ulm, ⁵ Department of Neurology, Ludwig-Maximilians University Munich, ⁶ Department of Neuropediatrics and Muscle Disorders, University of Freiburg, ⁷ Department of Neurology, Ruhr-University Bochum, ⁸ Department of Neurology, University of Regensburg

Background: Hereditary spastic paraplegias (HSP) comprise a group of neurodegenerative disorders affecting primarily the corticospinal tract. The natural history of HSP is unknown and a reliable instrument to measure disease severity and progression has not been available.

Methods: To measure disease severity in HSP we developed a clinical rating scale, HSPRS, consisting of 13 items designed to rate functional impairment occurring in pure forms of the disease. Additional symptoms constituting a complicated form of HSP are recorded in an inventory. Evaluation was performed in two independent patient cohorts in a two-step validation procedure.

Results: Application of HSPRS requires less than 15 minutes and does not depend on any special equipment. Thereby HSPRS is highly suitable for use in an outpatient setting. Interrater agreement of HSPRS was high (intraclass correlation coefficient = 0.99). Reliability was further supported by high internal consistency (Cronbach's alpha of 0.91). HSPRS values were almost normally distributed without apparent floor or ceiling effect. Construct validity was shown by high correlation of HSPRS to Barthel index and the International Cooperative Ataxia Rating Scale (convergent validity) and low correlation to Minimal Status Examination (discriminant validity).

Conclusion: HSPRS is the first validated score for HSP. Evaluation establishes HSPRS as a robust measure of disease severity in HSP and recommends HSPRS for forthcoming therapeutical trials.