

Unified Huntington's disease rating scale: a follow up.

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An objective assessment of the clinical findings in patients with Huntington's disease (HD) is necessary for an evaluation of the longitudinal progression of the disease features. The Unified Huntington's Disease Rating Scale (UHDRS) is a scale to assess clinical performance and functional capacity. The authors examined the 1_year change in UHDRS scores in 78 patients with HD examined either in Leiden, the Netherlands (24 men, 25 women), or in Rochester, New York, United States (12 men, 17 women). A significant decline was seen in motor function, measured with the total motor scale. The total dystonia score increased significantly; the total chorea score did not. The frequency of behavioral disorders tended to increase. The scores on independence scale, functional assessment, total functional capacity, and symbol digit decreased significantly. No relation was observed between the UHDRS items and the age at onset or duration of illness. Thirteen patients with 2_year follow up showed a clear increase in score on the total motor scale and a decline on the independence scale and in total functional capacity. The UHDRS may also be used as a tool for determining therapeutic intervention. Annual evaluation of the total motor scale in every patient gives a clear description of the motor progression of the disease. The authors suggest performing a total UHDRS evaluation every second year for every HD patient as part of the routine longitudinal evaluation.