

Introduction: Huntington disease (HD) is an autosomal dominant neurodegenerative disorder manifested by motor, behavioural and cognitive deficits with fatal consequences.

Aims: This study aims to validate the psychometric properties of a standard cognitive battery used in HD and establish language-specific normative values.

Methods: In the first study, cognitive performance was compared in 106 patients at different stages of HD and 100 healthy controls matched for age, sex, and education. The neuropsychological battery included the Symbol Digit Modalities Test, Stroop Word Reading Test, Stroop Colour Naming Test, Stroop Interference Test, Trail Making Test-A and B, Category and Letter Verbal Fluency. In addition, patients were comprehensively assessed for motor and functional status.

In the second study, the cognitive performance of 3,267 healthy subjects was assessed. The work focused on the stratification of cognitive performance concerning age, gender, language and level of education. Another aim was to establish the language-specific normative values and implement a web-based normative calculator to assess the degree of cognitive deficit in different languages (English, German, Spanish, Italian, Polish, French, Dutch, Danish).

Results: In the first study, analysis of variance showed that healthy controls performed significantly better than patients on all cognitive tests. Cognitive performance was correlated with motor and functional impairment ($p < 0.001$) independent of age and disease duration.

A normative study showed a significant non-linear decline in cognitive performance with advancing age in healthy subjects. Cognitive performance in all tests was significantly better in subjects who had attained higher education levels. In addition, language-related differences in cognitive performance were found in all tests. In terms of gender, there was a trend towards better performance on the SDMT for females.

Conclusion: The results of both studies provide normative data for a specific cognitive battery stratified by age, education, gender, and language to be used in the field of HD and other neurodegenerative diseases. The normative values obtained were used to create a freely available web-based calculator that allows for use in clinical and research practice.

Keywords: Huntington disease, Unified Huntington's Disease Rating Scale, neuropsychological test battery, cognitive performance, normative data, normative calculator.