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Psychiatric symptoms do not correlate with cognitive decline, motor symptoms, or CAG repeat length in Huntington's disease

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Abstract

Objectives: To investigate the hypothesis that psychiatric disturbances in Huntington's disease are related to degree of cognitive or motor compromise and to determine correlations between CAG repeat length within the gene for Huntington's disease and disease severity.

Design: Consecutive series of patients with Huntington's disease.

Setting: Neurological specialty hospital.

Patients: Seventeen men and 12 women from 24 families.

Main outcome measures: The Hamilton Psychiatric and Anxiety Rating Scales and Brief Psychiatric Rating Scale were used to assess psychiatric disturbances; Folstein's Quantified Neurological Examination to evaluate motor status; and the Mini-Mental State Examination, Raven Progressive Matrices), Phonemic Verbal Fluency Test, Short Tale Test, Visual Search Test, and Benton's Visual Orientation Line Test to evaluate cognitive function. The length of the CAG repeat sequence in the Huntington's gene was determined by quantitative polymerase chain reaction.

Results: Cognitive test scores correlated significantly with each other; of these, results of the Visual Search and Short Tale tests correlated significantly with the Folstein's Quantified Neurological Examination score ($P = .05$ and $P = .03$, respectively). Results of the Folstein's Quantified Neurological Examination also correlated with the illness duration and the length of the CAG repeat. Although psychiatric scores correlated significantly among themselves ($P < .01$), neither cognitive compromise, motor deterioration, nor CAG length were related to the extent of psychiatric compromise. Patients who were depressed when they were examined tended to have a history of psychiatric disorders.

Conclusions: The lack of correlation between disease severity and psychiatric disturbances indicates that psychiatric disorders progress nonlinearly, possibly because of differential degeneration of the striatal-cortical circuits; the possibility that psychiatric disorders are prevalent in certain families with a member who has Huntington's disease is being further investigated. The lack of correlation between CAG length and cognitive and psychiatric variables needs further investigation.

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