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Neurology

Historical Abstract: February 1, 1986

HUNTINGTON'S DISEASE IN VENEZUELA: NEUROLOGIC FEATURES AND FUNCTIONAL DECLINE

A B Young, I Shoulson, JB Penney, S Starosta-Rubinstein, F Gomez, H Travers, MA Ramos-Arroyo, SR Snodgrass, E Bonilla, H Moreno, NS Wexler

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We studied 65 Huntington's disease patients and 225 at-risk individuals over the past 4 years. The rate of decline of these untreated patients from Venezuela was similar to that seen in US patients who had received neuroleptic drugs. Chorea, oculomotor dysfunction, and dysdiadochokinesis were early symptoms; parkinsonian features and dystonia came later. Juvenile patients declined nearly twice as fast as adult-onset patients. No distinctive neurologic phenotypes were seen in children of two affected parents.

Comment from Ryan J. Uitti, MD, FAAN, Associate Editor: *This paper documented the course of Huntington disease from the world's region that made the greatest contributions to the understanding of the most common neurodegenerative genetic disorder.*

Huntington's disease in Venezuela: Neurologic features and functional decline

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