



Pediatric Neurology Part III: Chapter 196. Huntington's disease in children (Handbook of Clinical Neurology)

Derek Letort, Pedro Gonzalez-Alegre

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Huntington's disease (HD) is a dominantly inherited, fatal neurodegenerative disease. This incurable illness is characterized by a triad of a movement disorder, cognitive decline and psychiatric manifestations.

Although most patients with HD have disease onset in the adult years, a small but significant proportion present with pediatric HD. It has been long known that patients with early-onset HD commonly exhibit prominent parkinsonism, known as the Westphal variant of HD. However, even among patients with pediatric HD there are differential clinical features depending on the age of onset, with younger patients frequently presenting diagnostic challenges. In his chapter, the characteristics of patients with childhood- and adolescence-onset HD are discussed, focusing on the differential clinical features that can aid the clinical reach a correct diagnosis, the indications and rational use of genetic testing and the currently available options for symptomatic treatment.

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