Late Onset Huntington Disease: Phenotypic and Genotypic Characteristics of 10 Cases in Argentina.

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Abstract

BACKGROUND:

Huntington's disease (HD) is a neurodegenerative disorder that includes motor, psychiatric and cognitive manifestations with typical onset of symptoms is in the forties. A percentage of patients (4.4% - 11.5%) may be exceptions to this and manifest symptoms later (>60 years old). Diagnosis of Late onset HD (LoHD) can be a challenge, due to the low suspicion of the disease at this age.

OBJECTIVE:

To review the genotype and phenotype of LoHD in an Argentinian cohort.

METHODS:

We reviewed the medical records and genetic testing of a total of 95 individuals with clinical and molecular diagnosis of Huntington's disease, based on 2 institution's registry.

RESULTS:

Among our HD cohort, 10 patients (10.52%) had LoHD, with variable results regarding family history. The average of repetitions of the expanded allele was 40 (range 38-44). All cases had mild motor symptoms at onset.

CONCLUSIONS:

Late onset HD can be a diagnostic challenge, due to its slow progression, unawareness of manifestations among patients and in many cases, mild symptomatology that does not warrant medical attention.

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