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**HUNTINGTON'S DISEASE: CLINICAL ASPECTS IN 81 PATIENTS (Abstract)\*.**  
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Huntington's disease is a genetically inherited degenerative neurological disorder, characterized by motor alterations, including involuntary movements such as chorea, dementia and psychiatric disturbances. The objective of this study is a retrospective analysis of the clinical features of a patient group with this diagnosis. The results of brain magnetic resonance performed in some of the patients were also analysed.

Charts from 81 patients seen at the Hospital das Clínicas da Faculdade de Medicina da Universidade de São Paulo between 1957 and 1995 were evaluated. There were 42 women and 39 men. There were 62 white, 10 black, 8 mulatto and 1 asian patients. The age of onset ranged from 5 to 65 years with a mean of 39.23 years.

The disease was maternally inherited in 23 cases, paternally inherited in 48 and in 10 cases this could not be established. There was no statistical correlation between the age of onset and sex or inheritance.

The disease was clinically manifested by choreic form in 78 cases and primary rigid form in 3 cases. In the latter 3 cases, the disease onset was significantly earlier than mean for the remainder. The initial clinical manifestation was chorea in 67 patients (82.7%), mental disorders in 21 (25.9%), and others in 6 (7.4%). Associated neurological features included chorea (97.5%), dysarthria (85.2%), alterations in muscle tone (69.1%), pyramidal signs (69.1%), dysphagia (50.6%), oculomotor disorders (39.4%), urinary incontinence (32.9%), dystonia (24.7%), bradykinesia (13.6%), other involuntary movements (9.7%), seizures (3.7%) and cerebellar syndrome (1.2%). There was a statistically significant association between the finding of pyramidal signs and paternal inheritance.

Associated mental disorders included dementia (62.5%), depression (42.3%), mania (1.3%) and psychosis (10.1%). The age of onset in patients with depressive symptoms was significantly higher than in those patients not presenting such symptoms. There was no significant difference in the age of onset in patients with or without psychosis.

In 17 patients, the brain magnetic resonance study showed variable degrees of caudate nucleus atrophy. However, the degree of atrophy was not directly related to the duration of the disease. In 14 patients, there was cortical atrophy which was predominant in the posterior regions in 10 of them. Signal intensity abnormalities in T2-weighted images in the putaminal region were observed in 5 patients. In 4 of them, there were putaminal hypointense signals and in 1 case (with primary rigid form of the disease) there was a central hypointense signal with peripheral hyperintense rim in this structure.

**KEY WORDS:** Huntington's disease, chorea, clinical aspects, magnetic resonance image.

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