Clinical and genetic characteristics of late-onset Huntington's disease

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Résumé en anglais	BACKGROUND: The frequency of late-onset Huntington's disease (>59 years) is assumed to be low and the clinical course milder. However, previous literature on late-onset disease is scarce and inconclusive. OBJECTIVE: Our aim is to study clinical characteristics of late-onset compared to common-onset HD patients in a large cohort of HD patients from the Registry database. METHODS: Participants with late- and common-onset (30-50 years)were compared for first clinical symptoms, disease progression, CAG repeat size and family history. Participants with a missing CAG repeat size, a repeat size of ≤35 or a UHDRS motor score of ≤5 were excluded. RESULTS: Of 6007 eligible participants, 687 had late-onset (11.4%) and 3216 (53.5%) common-onset HD. Late-onset (n = 577) had significantly more gait and balance problems as first symptom compared to common-onset (n = 2408) (P < .001). Overall motor and cognitive performance (P < .001) were worse, however only disease motor progression was slower (coefficient, -0.58; SE 0.16; P < .001) compared to the common-onset group. Repeat size was significantly lower in the late-onset (n = 40.8; SD 1.6) compared to common-onset (n = 44.4; SD 2.8) (P < .001). Fewer late-onset patients (n = 451) had a positive family history compared to common-onset (n = 2940) (P < .001). CONCLUSIONS: Late-onset patients present more frequently with gait and balance problems as first symptom, and disease progression is not milder compared to common-onset HD patients apart from motor progression. The family history is likely to be negative, which might make diagnosing HD more difficult in this population. However, the balance and gait problems might be helpful in diagnosing HD in elderly patients.
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Liens

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