GBA-associated Parkinson's disease: reduced survival and more rapid progression in a prospective longitudinal study.

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Abstract

BACKGROUND: Parkinson's disease (PD) patients with GBA mutations show an earlier age at onset and more severe non-motor symptoms compared with PD patients without GBA mutations.

OBJECTIVE: This study was undertaken to evaluate progression of motor and non-motor symptoms in sporadic PD patients depending on the mutational GBA status.

METHODS: We used regression analysis to evaluate independent effects of the mutational GBA status, age at onset, age at examination, and disease duration on motor (Unified Parkinson's Disease Rating Scale [UPDRS]-III, Hoehn and Yahr [H&Y] stage, Levodopa [L-dopa]-equivalent-dosage) and non-motor characteristics (cognition and mood). Disease progression was assessed prospectively over 3 years.

RESULTS: The GBA-associated PD patients compared with non-mutation PD patients, although younger and with an earlier age at onset, show (1) a more rapid disease progression of motor impairment and cognitive decline and (2) reduced survival rates.

CONCLUSIONS: The mutational GBA status, rather than older age and age at onset, presents an important predictor for disease progression in this specific subgroup of PD patients.

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KEYWORDS: GBA; Parkinson; dementia; progression; survival

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