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

Brockmann K¹, Srulijes K, Pflederer S, Hauser AK, Schulte C, Maetzler W, Gasser T, Berg D.

Author information

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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