

Opis choroby *

Definicja

An atypical variant of progressive supranuclear palsy (PSP), a rare late-onset neurodegenerative disease, characterized by progressive freezing of gait, speech and writing early in the disease course. Later, axial rigidity, and facial immobility can occur, and supranuclear downgaze paresis may emerge after a decade. Neuropathological characteristics include tau pathology and neuronal loss in specific brain areas, especially in the globus pallidus, subthalamic nucleus, and substantia nigra. The tau pathology is less widespread compared to the other PSP sub-types.

Dane

Klasyfikacja

Podtyp kliniczny

Synonimy

PSP-PAGF

PSP-czysta akinezja z zamrożeniem chodu

PSP-PAGF

PSP-pure akinesia with gait freezing

Kod ORPHA

240094

Kod OMIM

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Kod ICD10

G23.1

Kod ICD11

8A00.10

*Źródło

orphanet