Opis choroby *

Definicja

A rare sporadic human prion disease characterized by adult onset of progredient neurodegeneration presenting as a combination of psychiatric, sleep, and oculomotor disturbances, with development of progressive cognitive impairment (the predominantly affected cognitive domains being memory, temporal and/or spatial orientation, language, executive functions, and attention), postural instability, and sometimes additional motor abnormalities and autonomic hyperactivity, in the course of the disease. Bilateral thalamic hypometabolism on FDG-PET imaging and positive prion seeding activity in the cerebrospinal fluid are present in many cases. The disease is fatal within typically two to three years.

Dane

Klasyfikacja
Choroba

Kod ORPHA
586130

Kod OMIM
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Kod ICD10
A81.9

Kod ICD11
6D85.5

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*Źródło

orphanet