

Sporadyczna śmiertelna bezsenność

Kod Orpha: 586130 Kod OMIM:

Opis choroby *

Definicja

A rare sporadic human prion disease characterized by adult onset of progressive 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

[* Źródło](#)

orphanet

Rozszerzony opis choroby

Brak opisu rozszerzonego dla tej choroby. Opracowanie w toku.

Dostępna na stronie www.orphanet.pl