## **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

<b>Kod ORPHA</b> 586130	Kod OMIM -	<b>Kod ICD10</b> A81.9
<b>Kod ICD11</b> 6D85.5		
<u>*Źródło</u>		
orphanet		