

## Opis choroby \*

### Definicja

A rare acquired human prion disease characterized by a progressive, invariably fatal neuropsychiatric disorder resulting from transmission via consumption of products from prion-diseased cows or via blood transfusion from an affected individual. Patients typically present early psychiatric symptoms (such as depression, anxiety, apathy, withdrawal, and delusions), as well as persistent painful sensory symptoms, ataxia, myoclonus, chorea, or dystonia, and dementia. Brain MRI often shows bilateral FLAIR hyperintensities involving the pulvinar thalamic nuclei. Neuropathological examination reveals spongiform change and extensive deposition of abnormal prion protein with florid plaques throughout the cerebrum and cerebellum.

### Dane

#### Klasyfikacja

Choroba

#### Synonimy

Variant MCJ

vCJD

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vCJD

#### Kod ORPHA

576370

#### Kod OMIM

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#### Kod ICD10

A81.0

#### Kod ICD11

8E01.2

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#### [\\*Źródło](#)

orphanet