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
Variant MCJ
vCJD

Kod ORPHA
576370

Kod OMIM
-

Kod ICD10
A81.0

Kod ICD11
8E01.2

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*Źródło

orphanet