Opis choroby *

Definicja

A rare acquired human prion disease characterized by rapidly progressive, fatal neurodegeneration, caused by the consumption of prion-containing tissue in endocannibalistic funeral rituals in Papua New Guinea until the late 1950s. After a decades-long asymptomatic period and a non-specific prodromal phase with headaches and arthralgia, the most prominent neurological feature is ataxia, in addition to other symptoms involving the cerebellum, brain stem, mid-brain, hypothalamus, and cerebral cortex, and emotional changes including inappropriate euphoria and compulsive laughter, or depression and apprehension. The last reported patient died in 2005 with an incubation period extending over four decades.

Dane

Klasyfikacja
Choroba

Kod ORPHA
454745

Kod OMIM
245300

Kod ICD10
A81.8

Kod ICD11
8E01.1

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*Źródło

orphanet