

## Opis choroby \*

### Definicja

A group of rare neurodegenerative diseases characterized by the accumulation of prions, abnormal variants of the cellular prion protein, primarily in brain tissue of affected individuals, as well as massive, rapid neuronal death, and an invariably fatal course. Human prion diseases most often occur sporadically but may also be of genetic origin or infectiously acquired. Irrespective of etiology, they are transmissible to other individuals.

### Dane

#### Klasyfikacja

##### Kategoria

#### Synonimy

TSE

Choroba prionowa

Transmissible spongiform encephalopathy

#### Kod ORPHA

56970

#### Kod OMIM

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#### Kod ICD10

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#### Kod ICD11

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#### \*Źródło

orphanet