

## Opis choroby \*

### Definicja

A rare, neurodegenerative disease characterized by progressive cataracts, hearing loss, cerebellar ataxia, paranoid psychosis and dementia. Neuropathological features are diffuse atrophy of all parts of the brain, chronic diffuse encephalopathy and the presence of extremely thin and almost completely demyelinated cranial nerves.

### Dane

<b>Klasyfikacja</b>	<b>Synonimy</b>
Podtyp kliniczny	Familial dementia, Danish type Rodzinna demencja, typ duński

<b>Kod ORPHA</b>	<b>Kod OMIM</b>	<b>Kod ICD10</b>
97346	117300	I68.0*

**Kod ICD11**  
5D00.2Y

---

### \*Źródło

orphanet