

## Opis choroby \*

### Definicja

Progressive polyneuropathy with bilateral striatal necrosis is a rare, genetic disorder of thiamine metabolism and transport characterized by the childhood-onset of recurrent episodes of flaccid paralysis and encephalopathy, associated with bilateral striatal necrosis and chronic progressive axonal polyneuropathy with proximal and distal muscle weakness, areflexia, contractures and foot deformities.

### Dane

### Klasyfikacja

Choroba

#### Kod ORPHA

217396

#### Kod OMIM

613710

#### Kod ICD10

E88.8

#### Kod ICD11

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### \*Źródło

orphanet