

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