

[Open Peer Review on Qeios](#)

Progressive polyneuropathy with bilateral striatal necrosis

INSERM

Source

INSERM. (1999). Orphanet: an online rare disease and orphan drug data base.

Progressive polyneuropathy with bilateral striatal necrosis. ORPHA:217396

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.