

Opis choroby *

Definicja

A rare motor neuron disease characterized by isolated lower motor neuron features, including progressive flaccid weakness, muscle atrophy, fasciculations, and reduced or absent tendon reflexes. Onset is in late adulthood, with men being affected more often than women. Upper motor neuron signs may develop later in some cases. Occurrence of respiratory insufficiency determines the prognosis. Neuropathological analysis shows intraneuronal Bunina bodies and ubiquitin-positive inclusions.

Dane

Klasyfikacja

Choroba

Synonimy

PMA

PMA

Kod ORPHA

454706

Kod OMIM

-

Kod ICD10

G12.2

Kod ICD11

8B60.3

*Źródło

orphanet