Opis choroby *

Definicja

A rare central nervous system malformation characterized by a fluid-filled longitudinally oriented cavity (syrinx) within the spinal cord, which may or may not communicate with the central canal, does not have an ependymal lining, and is either idiopathic or seen as a familial malformation. Clinical manifestations in symptomatic patients include neuropathic pain, as well as sensory and motor disturbances. Typical presentations may be cape-like loss of pain and temperature sensation along the torso and arms, or disproportionately greater motor impairment in upper compared to lower extremities.

Dane

Klasyfikacja
Wada morfologiczna

Synonimy
Congenital syringomyelia
Wrodzona jamistość rdzenia

Kod ORPHA
99856

Kod OMIM
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Kod ICD10
Q06.4

Kod ICD11
8D66.0

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*Źródło

orphanet