Opis choroby *

Definicja

A rare, congenital, non-syndromic malformation of neurenteric canal, spinal cord and column, characterized by intraspinal, predominantly intradural-extramedullary cystic mass located typically ventral to the spinal cord. Histopathology reveals columnar or cuboidal epithelium with or without cilia and mucus globules. Patients may be asymptomatic or present with signs and symptoms of compression of the spinal cord and associated nerve roots, such as focal weakness, progressive paresis, paresthesias, gait disturbance, or radicular pain. Concomitant congenital vertebral anomalies are frequently observed.

Dane

Klasyfikacja
Wada morfologiczna

Kod ORPHA
268865

Kod OMIM
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Kod ICD10
Q06.8

Kod ICD11
LA07.Y

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*Źródło

orphanet