Opis choroby *

Definicja

A group of rare central nervous system malformations characterized by varying degrees of absence or dysplasia of the derivatives of the prosencephalon (i. e. telencephalon and diencephalon), with an intact cranial vault. The spectrum comprises atelencephaly, the less severe form, in which only the telencephalon is affected, and aprosencephaly, where the diencephalon is also involved. The malformations may occur in an isolated form or in association with other anomalies.

Dane

Klasyfikacja
Wada morfologiczna

Synonimy
AP/AT spectum
AP/AT spectum

Kod ORPHA
566847

Kod OMIM
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Kod ICD10
Q04.3

Kod ICD11
LA05.Y

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*Źródło

orphanet