

## Opis choroby \*

### Definicja

A rare malformation characterized by missing scalp and flat bones over an area of the cranial vault. The size of the affected area is variable. In rare cases, acalvaria involves the whole of the dome-like superior portion of the cranium comprising the frontal, parietal, and occipital bones. Dura mater and associated muscles are absent in the affected area but the central nervous system is usually unaffected, although some neuropathological abnormality is often present (e.g. holoprosencephaly or gyration anomalies). Skull base and facial bones are normal.

### Dane

#### Klasyfikacja

Zespół wad wrodzonych  
Synonimy  
Primary acalvaria  
Bezczaszkwie

#### Kod ORPHA

945

#### Kod OMIM

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#### Kod ICD10

Q00.0

#### Kod ICD11

LA00.0Y

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#### \*Źródło

orphanet