

## Opis choroby \*

### Definicja

Primary intraosseous venous malformation is a rare, genetic vascular anomaly characterized by severe blood vessel expansion (most frequently within the craniofacial bones) with painless bone enlargement (usually of mandibule, maxilla and/or orbital, nasal, and frontal bones), typically resulting in facial asymmetry and contour deformation. Midline abnormalities, such as diastasis recti, supraumbilical raphe, and hiatus hernia, are commonly associated. Additional features reported include gingival bleeding, ectopic tooth eruption, exophthalmos, loss of vision, nausea, and vomiting.

### Dane

#### Klasyfikacja

Choroba

#### Synonimy

Intraosseous hemangioma  
Malformacja naczyniowa kości  
Naczyniak wewnątrzkościowy  
Osseous venous malformation

#### Kod ORPHA

140436

#### Kod OMIM

606893

#### Kod ICD10

D18.0

#### Kod ICD11

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#### \*Źródło

orphanet