

## **Opis choroby \***

### Definicja

A rare, benign, primary bone dysplasia characterized by progressive replacement of normal bone and marrow with fibrous connective tissue in either one (monostotic) or multiple (polyostotic) bones. Clinical manifestations depend on the anatomic location of the replacement and may include bone pain, deformities, pathological fractures, and cranial nerve deficits.

### Dane

#### **Klasyfikacja**

Zespół wad wrodzonych

#### **Kod ORPHA**

249

#### **Kod OMIM**

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#### **Kod ICD10**

Q78.1

#### **Kod ICD11**

FB80.0

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\*Źródło

orphanet