

## Opis choroby \*

### Definicja

A rare association syndrome, reported in several members of two families to date, characterized by arterial dissection, occurring at an early age and presenting with a range of manifestations depending on the vascular territory involved (ex. headache, dysphasia, hemiparesis), in association with cystic medial necrosis and multiple lentigines (brown and black in color and mainly affecting the skin of the trunk and extremities).

### Dane

### Klasyfikacja

Zespół wad wrodzonych

#### Kod ORPHA

1682

#### Kod OMIM

600459

#### Kod ICD10

Q87.8

#### Kod ICD11

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### \*Źródło

orphanet