

## Opis choroby \*

### Definicja

A rare primary immunodeficiency disorder characterized by autosomal dominant inheritance, absolute neutrophil counts below  $0.5 \times 10^9/L$  in the peripheral blood (on three separate occasions over a six month period), granulopoiesis maturation arrest at the promyelocyte/myelocyte stage and early-onset, severe, recurrent bacterial infections.

### Dane

### Klasyfikacja

Choroba

#### Kod ORPHA

486

#### Kod OMIM

613107

#### Kod ICD10

D70

#### Kod ICD11

4B00.00

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### \*Źródło

orphanet