

## Opis choroby \*

### Definicja

A rare, genetic, autoinflammatory syndrome with immune deficiency disease characterized by recurrent and severe flares of generalized pustular psoriasis associated with high fever, asthenia, and systemic inflammation, due to IL36R antagonist deficiency. Psoriatic nail changes (e.g. pitting and onychomadesis) and ichthyosis may occasionally be associated.

### Dane

#### Klasyfikacja

Choroba

#### Synonimy

Deficiency of IL-36R antagonist

Niedobór antagonisty IL-36R

Niedobór IL-36Ra

Deficiency of IL-36Ra

#### Kod ORPHA

404546

#### Kod OMIM

614204

#### Kod ICD10

L40.1

#### Kod ICD11

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#### \*Źródło

orphanet