

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