

Opis choroby *

Definicja

A rare genetic autoinflammatory syndrome characterized by early-onset of repeated episodes of fever, nodular neutrophil-rich panniculitis, arthralgia, and lipodystrophy. Additional reported features include diarrhea, failure to thrive, lymphadenopathy, and vasculitis. Laboratory examination may reveal elevated serum C-reactive protein and leukocytosis with neutrophilia in the absence of infection.

Dane

Klasyfikacja

Choroba

Synonimy

ORAS

Otulipenia

Niedobór OTULIN

Zespół autoinflammatoryjny związany z OTULIN

OTULIN deficiency

OTULIN-related autoinflammatory syndrome

Otulipenia

Kod ORPHA

500062

Kod OMIM

617099

Kod ICD10

D89.8

Kod ICD11

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[*Źródło](#)

orphanet