

Opis choroby *

Definicja

A rare immune deficiency with skin involvement characterized by early infantile onset of a clinical tetrad comprising generalized severe seborrheic-like erythroderma, recurrent secondary bacterial or fungal infections (most commonly *Staphylococcus aureus*, *Candida*, and gram-negative bacteria), persistent, profuse malabsorptive diarrhea, and failure to thrive or marked wasting. Associated systemic symptoms include fever, anemia, and weight loss. Further critical complications are impaired thermoregulation and severe fluid loss due to extensive exfoliation.

Dane

Klasyfikacja

Choroba

Synonimy

Leiner disease

Choroba Leinera

Kod ORPHA

314

Kod OMIM

609536

Kod ICD10

L21.1

Kod ICD11

EH40.2

*Źródło

orphanet