

# Erythrodermia złuszcząca

## Kod Orpha: 314 Kod OMIM: 609536

### 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

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#### [\\*Źródło](#)

orphanet

### Rozszerzony opis choroby

Brak opisu rozszerzonego dla tej choroby. Opracowanie w toku.