

## Opis choroby \*

### Definicja

Peeling skin syndrome (PSS) refers to a group of rare autosomal recessive forms of ichthyosis (see this term) that is characterized clinically by superficial, asymptomatic, spontaneous peeling of the skin and histologically by a shedding of the outer layers of the epidermis. PSS presents with either an acral (acral PSS) or a generalized distribution (generalized PSS type A (non inflammatory) or B (inflammatory)) (see these terms). Some cases remain difficult to classify, suggesting that there could be additional subtypes of PSS.

### Dane

#### Klasyfikacja

Grupa fenomenów

#### Synonimy

Deciduous skin

Skóra liściasta

Familial continuous skin peeling syndrome

Idiopathic deciduous skin

Keratosis exfoliativa congenita

PSS

Peeling skin disease

#### Kod ORPHA

817

#### Kod OMIM

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#### Kod ICD10

Q80.8

#### Kod ICD11

EC20.1

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#### \*Źródło

orphanet