

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

[*Źródło](#)

orphanet