Opis choroby *

Definicja

A rare primary cutaneous amyloidosis characterized by familial occurrence of lichen and/or macular amyloidosis due to fibrillary degeneration and apoptosis of basal keratinocytes, followed by conversion of filamentous masses into amyloid material in the papillary dermis. Patients typically present with a pruritic eruption of grouped hyperkeratotic papules, which may coalesce to form hyperkeratotic plaques, with a predilection for the lower limbs (lichen amyloidosis), or with hyperpigmented macules, sometimes with a reticulate pattern, most commonly arising on the back, chest or interscapular areas (macular amyloidosis).

Dane

Klasyfikacja
Choroba

Synonimy
FPLCA
FplCA

Kod ORPHA
353220

Kod OMIM
613955

Kod ICD10
L99.0*

Kod ICD11
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*Źródło

orphanet