Opis choroby *

Definicja

A rare, non-syndromic, hereditary palmoplantar keratoderma characterized by diffuse, yellowish, thick hyperkeratosis of the palms and soles with a sharp demarcation at the volar border and an erythematous margin, and the epidermolytic pattern of changes on the skin biopsy, including perinuclear vacuolization, granular degeneration of keratinocytes in the spinous and granular layer, and tonofilament aggregates. Painful fissures and hyperhidrosis are frequently associated.

Dane

Klasyfikacja
Choroba

Synonimy
Diffuse erythrodermic palmoplantar keratoderma, Voerner type
Epidermolityczny rogowiec dłoni i stóp Voerner
Epidermolityczny rogowiec dłoni i stóp Vörnera
EPPK
Rozlany erytrodermalny rogowiec dłoni i stóp, typu Voernera
Rozlany erytrodermalny rogowiec dłoni i stóp, typu Vörnera
Diffuse erythrodermic palmoplantar keratoderma, Vörner type
EPPK
Epidermolytic palmoplantar keratoderma of Voerner
Epidermolytic palmoplantar keratoderma of Vörner

Kod ORPHA
2199

Kod OMIM
144200

Kod ICD10
Q82.8

Kod ICD11
EC20.30

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*Źródło

orphanet