

Opis choroby *

Definicja

A rare genetic skin disease characterized by the triad of congenital scleroatrophy predominantly of the hands with sclerodactyly, palmoplantar keratoderma, and nail changes (consisting of hypoplasia, ridging, clubbing, and white discoloration). Additional features include palmar hypohidrosis and a high susceptibility to early-onset squamous cell carcinoma of affected skin areas.

Dane

Klasyfikacja

Choroba

Synonimy

Palmoplantar hyperkeratosis-sclerodactyly syndrome
Scleroatrophic syndrome
Zespół hiperkeratoza dłoniowo-podeszwowa - sklerodaktylia
Zespół Hurieza
Palmoplantar keratoderma-sclerodactyly syndrome
Scleroatrophic syndrome
Sclerotylosis

Kod ORPHA

384

Kod OMIM

181600

Kod ICD10

Q82.8

Kod ICD11

EC20.30

*Źródło

orphanet