

Opis choroby *

Definicja

A rare genetic skin disease characterized by generalized poikiloderma with marked accentuation in flexural regions and on extensor surfaces, sclerosis of palms and soles, and linear and reticulated hyperkeratotic and sclerotic bands in the axilla and the antecubital and popliteal fossae. Subcutaneous calcification, finger clubbing, Raynaud phenomenon, and cardiac abnormalities (such as severe aortic stenosis) have also been reported.

Dane

Klasyfikacja

Choroba

Kod ORPHA

221039

Kod OMIM

173700

Kod ICD10

Q82.8

Kod ICD11

EC10

*Źródło

orphanet