Opis choroby *

Definicja

A rare genetic disease characterized by thickening of the skin on palms and soles restricted to areas of weight bearing and/or friction (focal, non-epidermolytic palmoplantar keratoderma) and oral and esophageal leukokeratosis, associated with a very high lifetime risk of developing squamous cell carcinoma of the esophagus. The skin lesions appear in childhood and can be complicated by fissuring and infection.

Dane

Klasyfikacja
Choroba

Synonimy
Bennion-Patterson syndrome
Hiperkeratoza dłoni i stóp - rak przełyku
Keratosis palmoplantaris-esophageal carcinoma syndrome
Zespół Benniona i Pattersona
Zespół hiperkeratozy dłoni i stóp oraz nowotworu przełyku
Zespół Howella i Evansa
Howell-Evans syndrome
Keratosis palmoplantaris-esophageal carcinoma syndrome
Palmoplantar hyperkeratosis-esophageal carcinoma syndrome
Tylosis-oesophageal carcinoma syndrome

Kod ORPHA
2198

Kod OMIM
148500

Kod ICD10
Q82.8

Kod ICD11
EC20.31

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*Źródło

orphanet