

## Opis choroby \*

### Definicja

Woolly hair-palmoplantar keratoderma syndrome is a very rare, hereditary epidermal disorder characterized by hypotrichosis/woolly scalp hair, sparse body hair, eyelashes and eyebrows, leukonychia, and striate palmoplantar keratoderma (more severe on the soles than the palms), which progressively worsens with age. Pseudo ainhum of the fifth toes was also reported. Although woolly hair-palmoplantar keratoderma syndrome shares clinical similarities with both Naxos disease and Carvajal syndrome, cardiomyopathy is notably absent.

### Dane

#### Klasyfikacja

Choroba

#### Synonimy

KWWH type IV

KWWH typu IV

Rogowiec z włosami wełnistymi typu IV

Zespół włosów wełnistych i hiperkeratozy  
dłoniowo-podeszwowej

Keratoderma with woolly hair type IV

Woolly hair-palmoplantar hyperkeratosis  
syndrome

#### Kod ORPHA

420686

#### Kod OMIM

616099

#### Kod ICD10

Q82.8

#### Kod ICD11

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#### \*Źródło

orphanet