

Opis choroby *

Definicja

Chronic hepatic porphyrias represent a sub-group of porphyrias (see this term). They are characterized by bullous photodermatitis caused by a deficiency of uroporphyrinogen decarboxylase (URO-D; the fifth enzyme in the heme biosynthesis pathway). Chronic hepatic porphyria encompasses two diseases: porphyria cutanea tarda and hepatoerythropoietic porphyria (extremely rare) (see these terms).

Dane

Klasyfikacja

Grupa fenomenów

Kod ORPHA

95161

Kod OMIM

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Kod ICD10

E80.2

Kod ICD11

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*Źródło

orphanet