

## Opis choroby \*

### Definicja

A rare biliary tract disease characterized by loss of interlobular bile ducts resulting in chronic cholestasis, without any known cause. Loss of less than 50% of interlobular bile ducts is associated with a mild disease course, while loss of the majority of ducts results in a severe form, potentially leading to cirrhosis and liver failure. Patients typically present as young or middle-aged adults with episodic jaundice, pruritus, and elevated liver enzymes.

### Dane

#### Klasyfikacja

Choroba

#### Synonimy

IAD

IAD

Idiopatyczna duktopenia dorosłych

Idiopathic adult ductopenia

#### Kod ORPHA

480512

#### Kod OMIM

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#### Kod ICD10

K83.8

#### Kod ICD11

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#### \*Źródło

orphanet