

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	Synonimy
Choroba	IAD
	IAD
	Idiopatyczna duktopenia dorosłych
	Idiopathic adult ductopenia

Kod ORPHA	Kod OMIM	Kod ICD10
480512	-	K83.8

Kod ICD11

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*Źródło

orphanet