

# Izolowane wrodzone włóknienie wątroby

Kod Orpha: 485426 Kod OMIM:

## Opis choroby \*

### Definicja

A rare parenchymal liver disease characterized by progressive fibrosis of the portal tracts due to arrest of maturation of the ductal plate of the intrahepatic bile ducts. Clinically, it may manifest as a portal hypertensive, cholangitic, mixed, or latent form. Onset of symptoms is mostly in adolescence or young adulthood. Hepatocellular function is relatively well preserved.

### Dane

#### Klasyfikacja

Choroba

#### Synonimy

Isolated CHF

Izolowane CHF

#### Kod ORPHA

485426

#### Kod OMIM

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#### Kod ICD10

Q44.6

#### Kod ICD11

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[\\*Źródło](#)

orphanet

## Rozszerzony opis choroby

Brak opisu rozszerzonego dla tej choroby. Opracowanie w toku.