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
-

Kod ICD10
Q44.6

Kod ICD11
-

---

*Źródło

orphanet