Zespół Caroliego
Kod Orpha: 480520 Kod OMIM:

Opis choroby *

Definicja
A rare genetic hepatic disease characterized by multiple segmental cystic dilatations of both central and smaller peripheral bile ducts associated with congenital hepatic fibrosis. Age of symptom onset is variable, as is disease progression. Patients present recurrent cholangitis, hepatolithiasis, and cholecystolithiasis. Portal hypertension may appear later in the disease course, and the risk of developing cholangiocarcinoma is increased significantly. The syndrome is often associated with autosomal recessive polycystic kidney disease.
Dane

Klasyfikacja
Zespół wad wrodzonych

Kod ORPHA
480520

Kod OMIM
-

Kod ICD10
Q44.6

Kod ICD11
-

---

*Źródło

orphanet

Rozszerzony opis choroby

Brak opisu rozszerzonego dla tej choroby. Opracowanie w toku.