Opis choroby *

Definicja

A rare systemic autoimmune disease characterized by cholestasis and diffuse cholangiographic abnormalities with circular and symmetrical bile duct wall thickening, and elevated serum IgG4 levels. Characteristic histopathological findings include dense infiltration of IgG4-positive plasma cells and extensive fibrosis in the bile duct wall. A marked response to steroid therapy is typical. Patients present with jaundice, cholangitis, pruritis, and sometimes associated findings of autoimmune pancreatitis, sialadenitis, and retroperitoneal fibrosis.

Dane

Klasyfikacja

Podtyp kliniczny

Kod ORPHA 447764

Kod OMIM

Kod ICD10

K83.0

Kod ICD11 4A43.0

*Źródło

orphanet