The incidence of choledochal cysts has increased due to an increase in the detection rate by means of because of advancements in technology. The incidence in Asia is somewhat higher than that in western countries. The reason for this geographical difference is still remains unclear. Further, there is also an unexplained difference in the female preponderance with the female: male ratio being commonly reported as 4:1. The most widely accepted hypothesis regarding the etiology is an anomalous arrangement of the pancreaticobiliary ductal junction. The triad of jaundice, right upper quadrant pain, and a palpable subcostal mass is diagnostic but is not seen present in all cases.

Choledochal cysts can be associated with biliary atresia, congenital hepatic fibrosis, and cystic disease of the kidney, especially renal tubular ectasia, sometimes in combination with cortical and medullary cysts. Reported complications of choledochal cysts include have been reported to cause complications such as secondary calculus formation, pancreatitis, biliary cirrhosis, cyst rupture with bile peritonitis, cholangitis, intrahepatic abscess, portal vein thrombosis, and malignant transformation into cholangiocarcinoma.

Ultrasonography is preferred for initial evaluation. It reveals an anechoic cystic structure separate from the gall bladder that communicates with the hepatic ducts. The differential diagnosis, which is based on ultrasound findings, includes other fluid-filled structures in this region, namely pancreatic pseudocysts, large right renal cysts, enteric duplication cysts, and hepatic artery aneurysms. Hepatobiliary scintigraphy can also complement support the diagnosis by showing revealing late accumulation of radioisotopes in the cystic structure.