Choledochal cysts

What are choledochal cysts?

Choledochal cyst is a congenital dilatation of part or whole of the bile duct. The cystic dilatations of the biliary tree can involve the extrahepatic and/or the intrahepatic biliary tract. There are five major classes of choledochal cysts:

[1]

- Type I cysts consist of saccular or fusiform dilatations of part or all of the extrahepatic bile duct:
    - Type IA is saccular and involves all or most of the entire extrahepatic bile duct.
    - Type IB is saccular and involves a limited segment of the bile duct.
    - Type IC is more fusiform and involves all or most of the extrahepatic bile duct.
- Type II choledochal cyst is an isolated diverticulum protruding from the wall of the common bile duct or joined to the common bile duct by a narrow stalk.
- Type III choledochal cyst (choledochocele) arises from the intraduodenal portion of the common bile duct.

- Type IV:
    - Type IVA cysts consist of multiple dilatations of the intrahepatic and extrahepatic bile ducts.
    - Type IVB choledochal cysts are multiple dilatations involving only the extrahepatic bile ducts.
- Type V (Caroli's disease) consists of multiple dilatations limited to the intrahepatic bile ducts.[2]

50-80% of all choledochal cysts are type I, 2% type II, 1.4-4.5% type III, 15-35% type IV and 20% type V.[3]

How common are choledochal cysts? (Epidemiology)

- Choledochal cysts are relatively rare in Western countries. Reported frequency rates range from 1 case per 100,000-150,000 live births.[3]
- Choledochal cysts are much more common in Asian populations with a reported incidence of 1 in 1,000 and about two thirds of cases occur in Japan.[3]
- Choledochal cysts are three or four times more prevalent in females than in males.[3]
- Most patients with choledochal cysts are diagnosed during infancy or childhood, although the condition may be discovered at any age. Approximately 67% of patients present before the age of 10 years.

Choledochal cyst symptoms[4]

- Choledochal cysts are often asymptomatic.
- 80% of patients present before the age of 10 years but presentation can be at any age.
- The classic triad of intermittent abdominal pain, jaundice and a palpable right upper quadrant abdominal mass occurs in fewer than 20% of patients, although almost two thirds of patients present with two of the three symptoms.

- Infants generally present with obstructive jaundice and abdominal masses, whereas older patients present most commonly with pain, fever, nausea, vomiting and jaundice.
- Symptoms are often due to the associated complications of ascending cholangitis and pancreatitis.
- Complications associated with all types of choledochal cysts result from bile stasis, stone formation, recurrent cholangitis and inflammation.
- Adults with choledochal cysts can present with one or more severe complications - eg, liver abscesses, cirrhosis, portal hypertension, recurrent pancreatitis, gallstones, cholangiocarcinoma.
- About 1-12% of patients present with spontaneous rupture and symptoms and signs of abdominal pain, sepsis and peritonitis.

Differential diagnosis[5]

The differential diagnosis includes biliary lithiasis, primary sclerosing cholangitis, pancreatic pseudocyst, biliary papillomatosis, and biliary hamartoma.

Biliary atresia is commonly associated with choledochal cysts and must, therefore, be ruled out in neonatal obstructive jaundice. Cystic biliary atresia very closely resembles choledochal cyst.

Investigations[5]

- Raised white blood cell count, (increased immature neutrophils in patients with cholangitis).
- Abnormal LFTs - cholestasis.
- Serum amylase and lipase concentrations may be increased in the presence of pancreatitis. Serum amylase concentrations also may be elevated in biliary obstruction and cholangitis.
- Abdominal ultrasonography is the test of choice for the diagnosis.[6] Antenatal ultrasound can demonstrate a choledochal cyst as early as 12 weeks of gestation. Endoscopic ultrasonography allows a good view of the intrapancreatic portion of the common bile duct.[4]

- Technetium ^{99m} Tc hepatobiliary iminodiacetic acid (HIDA) scan is often used and is particularly useful for showing continuity with bile ducts and diagnosis of cyst rupture.[4]
- Abdominal CT scan and MRI help to delineate the anatomy of the lesion and of the surrounding structures. They can also help to identify the presence and extent of intrahepatic duct involvement. Magnetic resonance cholangiopancreatography (MRCP) is now considered to be the gold standard.[4]
- Percutaneous transhepatic cholangiography (PTC) or endoscopic retrograde cholangiopancreatography (ERCP) are helpful in demonstrating the presence of an anomalous pancreatobiliary junction and in identifying associated extrahepatic or intrahepatic strictures and stones.[7]

Associated diseases

There is a common association with anomalous pancreatobiliary duct junction.[8]

Choledochal cyst treatment and management[5]

Choledochal cysts should be treated by complete surgical excision because of the risk of bile duct malignancy. A biliary-enteric anastomosis restores continuity with the gastrointestinal tract. Partial cyst resection and internal drainage are less satisfactory because of occasional pancreatitis, cholangitis and cholangiocarcinoma.[9]

Patients who present at a late stage, after the development of advanced cirrhosis and portal hypertension, may not be good candidates for surgery because of the associated high morbidity and mortality rates.

Patients who present with cholangitis should be treated with broad-spectrum antibiotic therapy directed against common biliary pathogens - eg, *E. coli* and *Klebsiella* spp.[10]

- **Type I and IV**: management consists of complete extrahepatic bile duct cyst excision down to the level of communication with the pancreatic duct, cholecystectomy, and restoration of bilioenteric continuity.

- **Type II and III**: are associated with an extremely low risk of malignant transformation. Diverticulectomy of type II followed by primary closure of the common bile duct at the diverticulum neck is usually sufficient. Appropriate management of small choledochoceles consists of endoscopic sphincterotomy. Transduodenal excision may be considered for large choledochoceles with associated complications such as gastric outlet obstruction or pancreatitis.
- **Type V (Caroli's disease)**: management consists of liver resection or orthotopic liver transplant. Localised or unilobar cystic disease is best managed with hepatic resection.

Complications

- With increasing age at presentation, the risks of intrahepatic strictures and stones, segmented hepatic atrophy/hypertrophy, secondary biliary cirrhosis, portal hypertension, liver failure and biliary malignancy all increase significantly.[11]
- The reported incidence of cholangiocarcinoma in patients with choledochal cyst ranges from 9-28%.[12] Cancer also presents 10-15 years earlier.
- Postoperatively, patients are at risk of developing pancreatitis and ascending cholangitis.

Prognosis

- The prognosis is generally excellent following surgical excision but depends on the age of presentation (worse prognosis with older age at presentation) and presence of complications.
- Following excision, there is an incidence of 0.7-6% of malignant disease, which is thought to be due to residual cyst tissue or subclinical malignant disease not detected before surgery.[13]

Further reading

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