Answer to Radiographic quiz

Radiographic findings
Fusiform dilatation of the entire common bile duct and both common hepatic bile ducts. No evidence of obstruction or filling defects with free passage of the contrast to the duodenum.

Diagnosis
Type I Choledochal cyst

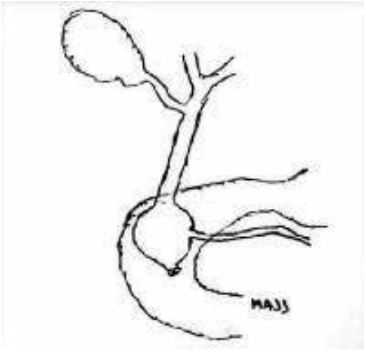
OVERVIEW OF THE DISEASE

Choledochal cysts
Choledochal cysts are unusual congenital anomalies of the bile ducts. They consist of cystic dilatations of the extrahepatic biliary tree, intrahepatic biliary radicles, or both.
Alonso-Lej et al provided the first systematic description of choledochal cysts in 1959 based on the clinical and anatomic findings in 96 cases. The resultant system classified choledochal cysts into 3 types and outlined therapeutic strategies

ANATOMY
The following discussion of the pertinent anatomy of choledochal cysts is based on the Todani classification.

• Type I choledochal cysts are most common and represent 80-90% of the lesions. Type I cysts are dilatations of the entire common hepatic and common bile ducts or segments of each. They can be saccular or fusiform in configuration. Type I cysts can be divided into 3 subclassifications, including type IA cysts, which are typically saccular and involve the entire extrahepatic bile duct (common hepatic duct plus common bile duct) or the major portion of the duct.

• Type II choledochal cysts (2%) are relatively isolated protrusions or diverticula that project from the common bile duct wall. They may be sessile or may be connected to the common bile duct by a narrow stalk.



• Type III choledochal cysts (1.5 – 5%) are found in the intraduodenal portion of the common bile duct. Another term used for these cysts is choledochocele.

for each. The classification system for choledochal cysts currently includes 5 major types (1). (See diagrams below.)

PATHOPHYSIOLOGY

The pathogenesis of choledochal cysts is most likely multifactorial. Some aspects of the disease are consistent with a congenital etiology, others with a congenital predisposition to acquiring the disease under the right conditions.

The vast majority of patients with choledochal cysts have an anomalous junction of the common bile duct with the pancreatic duct (anomalous pancreatobiliary junction (APBJ). An APBJ is characterized when the pancreatic duct enters the common bile duct 1 cm or more proximal to where the common bile duct reaches the ampulla of Vater. Miyano and Yamataka have demonstrated such APBJs in more than 90% of their patients with choledochal cysts.

The APBJ allows pancreatic secretions and enzymes to reflux into the common bile duct. In the relatively alkaline conditions found in the common bile duct, pancreatic proenzymes can become activated. This results in inflammation and weakening of the bile duct wall. Severe damages may result in complete denuding of the common bile duct mucosa. From a congenital standpoint, defects in epithelialization and recanalization of the developing bile ducts during organogenesis and congenital weakness of the duct wall have also been implicated. The result is the formation of a choledochal cyst.

CLINICAL MANIFESTATIONS

Infants commonly present with conjugated hyperbilirubinemia, failure to thrive or abdominal mass.

In patients older than 2 years, chronic intermittent abdominal pain is the presenting symptom 50-96 % of the time and intermittent jaundice 34-55% of the time.

They most commonly affect females (4:1) and Asians and can be detected at any point in life (80% in childhood).

They are 70 – 80% more frequent in women than men.

Typical presentation: recurrent right upper quadrant pain, jaundice, and/or palpable mass.

They may be complicated by stone formation, cholangitis, pancreatitis, or rupture.

Type I choledochal cysts are often associated with recurrent pancreatitis and eventually cholangiocarcinoma.

Rarely, they can show malignant degeneration .Cholangiocarcinoma develops in up to 7% of cases.

They can be associated with gallbladder aplasia, double gallbladder, annular pancreas, and biliary.

RADIOLOGICAL WORKUP AND TYPICAL FINDINGS:

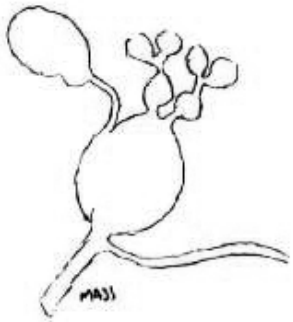
Conventional Radiography:

Findings: Plain abdominal radiographs are of little use in the diagnosis of choledochal cysts. They offer no specific information related to this diagnosis. In patients presenting with abdominal pain, radiographs are frequently ordered as part of the standard workup. At most, radiographs may suggest displacement of an adjacent hollow viscus, such as the duodenum, by a mass.

Computed Tomography:

Findings: Abdominal CT scanning is useful in the diagnostic algorithm for choledochal cysts. CT is highly accurate and offers a great deal of information that is helpful, not only in confirming the diagnosis but also in planning surgical approaches.

CT scans of a choledochal cyst demonstrate a dilated cystic mass with clearly defined walls, which is separate from the gallbladder. The fact that this mass arises from or actually is the extrahepatic bile duct usually is clear from its location and its relationships to surrounding structures. The cyst is typically filled with bile, which produces water-like attenuation. Depending on the patient's age and clinical history, the wall of the cyst can appear thickened, especially if multiple episodes of inflammation and cholan-



• Type IVA cysts (19%) are characterized by multiple dilatations of the intrahepatic and extrahepatic biliary tree. Most frequently, a large solitary cyst of the extrahepatic duct is accompanied by multiple cysts of the intrahepatic ducts. Type IVB choledochal cysts consist of multiple dilatations that involve only the extrahepatic bile duct.

• Type V choledochal cysts ("Caroli's disease") are defined by dilatation of the intrahepatic biliary radicles. Often, numerous cysts are present with interposed strictures that predispose the patient to intrahepatic stone formation, obstruction, and cholangitis. The cysts are typically found in both hepatic lobes. Occasionally, unilobar disease is found, which most frequently involves the left lobe.

Caroli's disease is associated with medullary sponge kidney and autosomal recessive polycystic kidney disease.

gitis have occurred. In addition, CT scanning is superior to US in defining the extent of the cyst in the extrahepatic biliary system and in detecting intrahepatic disease.

MRI:
Findings: These cysts appear as large fusiform or saccular masses that may be extrahepatic, intrahepatic, or both, depending on the type of cyst. They produce a particularly strong signal on T2-weighted images. Associated anomalies of the pancreatic duct, its junction with the common bile duct, and the long common channel formed by the 2 are usually well demonstrated on MRI/MRCP images.

Ultrasound:
Findings: US is the initial screening examination of choice in patients with choledochal cysts. Pertinent findings include a cystic extrahepatic mass. Depending on the skill of the operator, the specific type or class of choledochal cyst may be identified. Newer high-resolution US machines help clinicians make such diagnoses. Furthermore, advances in US technology have enabled ultrasonographers to make the diagnosis in the antenatal period.
US findings are diagnostic in many patients; however, in the preoperative period, complementary studies, such as ERCP, CT, or MRI/MRCP, may be helpful in delineating details of the surrounding anatomy, the location of an APBJ, and the length of the common pancreatobiliary channel.
Abdominal US findings can help in detecting associated conditions and complications of choledochal cysts, such as choledocholithiasis, intrahepatic biliary dilatation, portal vein thrombosis, gallbladder or biliary neoplasms, pancreatitis, and hepatic abscesses.

Nuclear Medicine:
Findings: Hepatobiliary scintigraphic modalities are used commonly in the setting of acute cholecystitis and in the investigation of neonatal jaundice. In addition, these techniques are useful in the diagnosis of choledochal cysts. Hepatobiliary scintigraphy can help differentiate between choledochal cysts and other peri-hepatic cystic masses, but a large choledochal cyst may also compress the gallbladder causing non-visualization of the gallbladder.

THERAPY:
The treatment for choledochal cysts is surgical. The treatment of choice for a type I choledochal cyst is complete excision of the cyst with construction of a Roux-en-Y biliary-enteric anastomosis to restore biliary continuity with the gastrointestinal tract.
Type II choledochal cysts can usually be excised entirely, and the defect in the common bile duct can be closed primarily over a T-tube. This approach can be used because, typically, type II choledochal cysts are lateral diverticula of the bile duct.
Therapy for type III choledochal cysts, or choledochoceles, depends on the size of the lesion. Choledochoceles with a diameter of 3 cm or smaller may be approached endoscopically and effectively treated by means of sphincterotomy. Choledochoceles larger than 3 cm in diameter are often associated with some degree of duodenal obstruction. These cysts are excised surgically by using a transduodenal approach. If the pancreatic duct is found to be entering the choledochocele, it must be reimplanted into the duodenum after the cyst is excised.
For type IV choledochal cysts, the dilated extrahepatic duct is completely excised, and a Roux-en-Y biliary-enteric anastomosis procedure is performed. No therapy is specifically directed at the intrahepatic ductal disease, except if intrahepatic ductal strictures, hepatolithiasis, or hepatic abscesses are present. In these patients, interventional radiologic techniques can be performed. If the disease is limited to specific hepatic segments or a lobe, these may be resected.
A type V choledochal cyst, or Caroli disease, is defined only by the dilatation of the intrahepatic ducts. If dilatation is limited to a single hepatic lobe, usually the left, the affected lobe is resected. Patients who have bilobar disease and signs of biliary cirrhosis, portal hypertension, or liver failure may be candidates for liver transplantation.
Patients with choledochal cysts require lifelong follow-up monitoring. They remain at increased risk for development of cholangiocarcinoma, even after complete excision of the cyst.

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