Management of Choledocal Cysts

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ACGME Core Competencies

- Patient Care
- Medical Knowledge
- Practice Based Learning/Improvement
- Interpersonal Communication Skills
- Professionalism
- Systems-based Practice
1) Clinical presentation child vs. adult?
2) Todani classification?
3) Etiology—what is Long common channel theory?
4) What is the reason for surgical intervention?
5) What are complications related to surgical management?
Case Presentation

- 18 F without significant PMHx
- Presented to ER in 7/2007 with c/o of acute onset of epigastric pain, associated with nausea and vomiting.
- Symptoms similar to two previous episodes in the past that resolved spontaneously
- CT scan performed demonstrating a diagnosis of Type I choledochal cyst (CC)
- No specific episodes of pancreatitis/ cholangitis
- PE: afebrile, VSS, no evidence of jaundice
  - Abd: soft non tender, non distended, +bowel sounds
Case Presentation

Labs (admission):
- CBC/SMA7/Liver Function Tests: WNL
- Amylase-26 Lipase-84

Imaging:
- CT scan (7/07): CHD/CBD dilatation from Rt PV into pancreatic head
- MRCP (9/07): Dilated extrahepatic ducts (CHD/CBD) through HOP to ampulla, no filling defects
Operative Procedure

- Excision of Choledocal cyst with Roux-en-Y hepaticojejunostomy
Operative Procedure

1) Evaluate extent of cyst
2) Lysis of pericystic adhesions
3) Perform cholecystectomy and choledochocystectomy
4) Perform internal drainage by Roux-en-Y jejunal loop
5) Place Jackson-Pratt drain and close abd wall
Choledochal cyst
(8x6.5cm)

Mild chronic cholecystitis
Uncomplicated post operative course

Return of bowel function on POD#4

Diet advanced

Discharged home on POD#6
Management of Choledochal Cysts
Choledochal cysts are congenital anomalies of the bile ducts.
Cystic dilatations of the extrahepatic biliary tree, intrahepatic biliary radicles, or both.
First described by Vater and Ezler in 1723; Douglas published the first complete clinical description of the anomaly in 1853.
Frequency rates range from 1 case per 100,000-150,000 to 1 case per 2 million live births.
3 or 4 times more prevalent in females than males.
- The female-to-male ratio is between 3:1 and 4:1.
Highest in Asian countries, especially Japan (1/1000)
60% of patients are diagnosed during first decade of life, about 20% go undiagnosed until adulthood.
Alonso-Lej et al classified (CC) into 3 types, which was later modified by Todani.

- **Type I** - Cystic dilatation of the common bile duct; most common, comprising 50–85% of all biliary cysts.
- **Type II** - Simple diverticulum of the extrahepatic biliary tree, comprising less than 5% of all cysts.
- **Type III** - Cystic dilatation of the intraduodenal portion of the extrahepatic CBD (choledochocoele) comprise approximately 5%.
- **Type IV** - Multiple cysts of the intrahepatic and extrahepatic biliary tree; IVA (both intrahepatic and extrahepatic cysts) and IVB (multiple extrahepatic cysts only); type IVA (30-40%).
- **Type V** - Isolated intrahepatic biliary cystic disease (Caroli's disease).
Classification


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• The exact cause of choledochal cysts remains obscure; Several theories:
  ◆ Congenital
  ◆ Weakness of the wall of the bile duct
  ◆ Obstruction of the distal choledochus
  ◆ Combination of obstruction and weakness
  ◆ Reflux of pancreatic enzymes into the CBD secondary to an anomaly of the pancreaticobiliary junction (APBJ)
• Long Common Channel (LCC) theory, first described by Babbitt
  - APBJ: entry of the pancreatic duct into the CBD 1 cm or more proximal to where the CBD reaches the ampulla of Vater, this allows for reflux of pancreatic juices into biliary system, possibly leading to increased intraductal pressure and inflammation
  - Not always responsible for CC, accounts for about 60-80% of cases
Etiology

• Pancreaticobiliary maljunction
• Classic triad for choledochal cysts is pain, jaundice, and abdominal mass.
  ◆ It is found in only a minority of children at the time of presentation.

• Infants commonly present with elevated conjugated bilirubin (80%), failure to thrive, or an abdominal mass (30%).

• In patients older than 2 years of age, abdominal pain is the most common presenting symptom.

• Intermittent jaundice and recurrent cholangitis are also common, as is pancreatitis.
Choledochal cysts in adults are usually suspected or diagnosed by hepatobiliary imaging studies, often initiated for evaluation of an upper abdominal complaint.

May be visualized non-invasively by ultrasonography, computed tomography (CT), magnetic resonance cholangiopancreaticography (MRCP), or by direct ductal imaging using ERCP.

“Gold standard” for visualizing a biliary duct cyst is MRCP, with sensitivity similar to that, but without the risk of complications, of ERCP.
Imaging-CT Scan

QuickTime™ and a TIFF (LZW) decompressor are needed to see this picture.
Why is it important to operate?

• Cholangiocarcinoma: such patients have a 20- to 30-fold higher risk than general population and this risk remains high even after surgical treatment.

• The risk of cholangiocarcinoma in the first decade of life is only 0.7%; however, this increases to 14% at 20 years and is postulated to increase even further throughout life.

• Cholangiocarcinoma may develop in all kinds of cysts but type I and IV cysts are associated with a higher incidence, even after cyst excision.

• Complete excision of these lesions is recommended as soon as possible, preferably before puberty, in order to decrease the chance of developing cancer.
Choledochal cysts were historically managed with biliary-enteric drainage via cyst-enterostomy (increased risk of bile duct and gallbladder cancer)- complete cyst excision recommended.

Main goal of management is to prevent malignant degeneration of the cyst via surgical excision.

Initial step should be to rule out metastatic disease.

Complete cyst excision may be difficult; the back wall of the cyst may be left intact to prevent injury to the portal vein or hepatic artery.

After cyst excision and cholecystectomy, the bile duct is reconstructed: hepaticojejunostomy, hepaticoduodenostomy or most commonly used technique Roux-en-Y hepaticojejunostomy
• After the cyst has been exposed, the gallbladder, which usually arises from the mid-portion of the choledochal cyst, should be dissected away from the hepatic bed.

• The goal is then to excise the intrapancreatic portion of the cyst without injuring the pancreatic duct or the long common channel. The distal-most portion of the choledochal cyst is encircled and transected as it enters the pancreas.

• If the cyst extends distally into the pancreas, the mucosa of the intrapancreatic portion of the cyst should be stripped away prior to closure at the point of distal transection.
Operative Management-type I

- Care must be taken to recognize atypical biliary anatomy, which may be encountered at the most proximal portion of dissection near the hilum.
- Cyst is transected at the bifurcation or more proximally if it extends into the individual hepatic ducts.
- Excised cyst should be examined grossly for malignancy and should be sent for frozen section.
- Reconstruction of the biliary tree is typically performed with a Roux-en-Y hepaticojejunostomy.
Operative Management

- **Type II**: treated with simple cyst excision.
  - CBD should be closed transversely (avoid stricturing)

- **Type III**: excision uncommon (low malignancy rate)
  - Endoscopic sphincterotomy (symptomatic)
  - Transverse duodenotomy for resection
Operative Management

- **Type IV**: require complete resection of the extrahepatic biliary tree when possible.
  - Regarding intrahepatic ducts, surgery should be individualized depending on: lobes affected, strictures or stones present, cirrhosis or malignancy
  - If intrahepatic cysts are localized into one lobe, hepatic lobectomy is the preferred approach.
  - For diffuse intrahepatic disease, liver transplantation would be considered.
Operative management

- **Type V** (Carolí’s disease):
  - If unilateral or segmental with cirrhosis: resection of the involved parenchyma.
  - Ursodeoxycholic acid may improve bile flow, reducing the incidence of biliary sludge, stones, and cholangitis.
  - In the absence of cirrhosis or malignancy, Roux-en-Y hepaticojejunostomy with bilateral transhepatic Silastic stents may be indicated to improve biliary drainage (stents left for 6-12 months).
  - Patients with Carolí's disease and liver failure may warrant liver transplantation.
Operative Management

Type I
EXCISION, ROUX-EN-Y
HEPATOJEJUNOSTOMY
EXCISION, HEPATICODUODENOSTOMY
Roux-en-Y choledochocystosto-
jejunitomy
Choledochocystoduodenostomy

Type IVA
Extrahepatic component
EXCISION, ROUX-EN-Y HEPATICO-
JEJUNOSTOMY
EXCISION, HEPATICODUODENOSTOMY

Type IVB
Intrahepatic component
Hepatic resection ±
Roux-en-Y hepatojejunostomy
Transhepatic intubation

Type II
EXCISION

Type III
TRANSODUDENAL EXCISION
Transduodenal sphincteroplasty
Endoscopic sphincterotomy

Type IV
Type V
(Caroli’s disease)

HEPATIC RESECTION
Roux-en-Y intrahepatic
cholangiojejunostomy
Transhepatic intubation

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Choledochal Cyst Disease in Children and Adults: A 30-Year Single-Institution Experience

- 92 pts with CC disease seen between 1976-2006 at Johns Hopkins Hospital; Differences between children and adults evaluated

- Findings:
  - Demographic: 90% of pts were female, 79% were adults, 91% non-Asian
  - Presentation: RUQ pain (91%), followed by n/v(47%)
  - Imaging: U/S (76%), followed by CT scan (66%)
  - Adults vs children: 71% children presented with jaundice and 97% adults with RUQ pain
Choledochal Cyst Disease in Children and Adults: A 30-Year Single-Institution Experience

- Excision of CC with Roux-en-Y hepaticojejunostomy - most common
- 34% overall complication: adults > children
- Post op length of stay: adult vs child 10:8
- Five pts with malignancy at time of resection (4) type I and (1) type IV
  - (2) type I with cholangiocarcinoma w/in cyst
  - (1) type I with GB cancer (1) type I embryonal rhabdomyosarcoma
  - Type IV-died at 7 months
  - (1) type I with neg margins, but + periportal LN-died
Choledochal Cyst Disease in Children and Adults: A 30-Year Single-Institution Experience

- Five pts without malignancy at time of resection: (1) type V, (1) type IV, (3) type I
  - Both type V/IV died of cholangiocarcinoma
  - All type I-died unrelated causes
- Conclusion:
  - Adults outnumber children
  - Difference in clinical presentation
  - Both intrahepatic/extrahepatic CC if untreated: increase Ca risk
  - For extrahepatic dz: complete excision eliminates risk for cholangiocarcinoma
  - Type IV and V- liver transplantation can only potentially eliminate Ca risk
Complications after surgery have been mainly observed with types I, IV, and V choledochal cysts.

The overall morbidity rate is less than 10%.

Postsurgical complications include:

- Cholangitis
- Biliary stone formation
- Anastomotic stricture
- Residual debris
  - Biliary stone formation
  - Pancreatitis
  - Cholangitis
- Intrahepatic bile duct dilatation
Summary

- Choledochal cysts require proper diagnosis and treatment to address associated symptoms, risk of malignancy, and disease progression.

- The majority of cases of biliary cysts (type I and IVA) can be treated effectively with cyst resection, cholecystectomy, and biliary reconstruction.

- While operative therapy decreases the risk of subsequent cancer, patients continue to require long-term surveillance for recurrent cholangitis, intrahepatic stones, pancreatitis, postoperative biliary strictures, and malignancy.