Definition

• Congenital anomalies of the biliary tract that are manifested by cystic dilatation of the extrahepatic and/or intrahepatic bile ducts
• Single or multiple
• Also termed Biliary cysts

**Epidemiology**

- **Incidence**
  - Asia: 1:1000
  - North America is 1:150,000 live births
- **F:M distribution: 3:1 to 4:1**
- **Age distribution: Children >>> adults (20%)**

**Risk of Malignancy**

- 20-30 fold ↑ risk of Cholangiocarcinoma
- 10–30% of adults (mean age 32)
- Overall risk of cancer 10-15%
  - 0.7 % < 10 years of age
  - 7 % 11 - 20 years of age
  - 14 % >20 years of age
  - As high as 50 % in older patients
Non-Malignant Complications

- Stone formation: Cystolithiasis, cholelithiasis, choledocholithiasis, hepatolithiasis
- Cholangitis
- Secondary biliary cirrhosis
- Pancreatitis

Unusual Non-Malignant Complications

- Intraperitoneal cyst rupture
- Bleeding
- Gastric outlet obstruction
- Intussusception
Classification of Biliary Cysts According to Todani and Colleagues

I (IA) common type; (IB) segmental dilatation; (IC) diffuse dilatation
II Diverticulum
III Choledochocele
IV (IVA) multiple cysts (intra- and extrahepatic); (IVB) multiple cysts (extrahepatic)
V Single or multiple dilatations of the intrahepatic ducts.

Case

- 63-year-old male with history of lymphoma
- CT scan: lymphadenopathies and an infiltrating mass in the liver
- Liver biopsy: moderate to poorly differentiated adenocarcinoma, likely PB or GI primary
- Subsequently developed abnormal LFTs
Abnormal Pancreaticoiliary Junction (APBJ)

- Junction of the BD and PD outside the duodenal wall with a long common ductual channel (≥8 mm)
- Chronic reflux of pancreatic enzymes into the biliary tree, → inflammation, dilatation, and scarring
- Seen in 10-58% of Choledochal cysts
- Choledochal cysts are seen in 75% of APBJ

Komi Classification

- Type I A narrowed common bile duct joins the pancreatic duct at a right angle
- Type II A pancreatic duct joins the common bile duct at an acute angle
- Type III It is complicated by a patent accessory pancreatic duct

References:

Crittenden SL, McKinley MJ. Am J Gastroenterol 1985;80:643-647
Kimura K et al Gastroenterology. 1985 Dec;89(6):1258-65
Komi N et al Journal of Pediatric Surgery, 1992, 27, 728-731
APBJ and Cancer

- Significantly more K-ras mutations and p53 overexpression
- APBJ without biliary cysts: High incidence of GB cancer (73%); Strongly consider
  - Prophylactic cholecystectomy
  - Extrahepatic bile duct resection/cholecystectomy
- APBJ with biliary cysts: Increased incidence of Cholangiocarcinoma and GB cancer

Biliary Cyst Presentation

- Incidental: Imaging, prenatal ultrasound, or endoscopy
- Neonates: cholestatic jaundice, acholic stools, vomiting, irritability, failure to thrive, hepatomegally, abdominal mass
- Children and adults: triad of abdominal pain, jaundice, and a palpable mass (<20%)
- Abdominal pain > adults, Jaundice > children
- Older patients: >Pancreatitis and recurrent cholangitis
Work Up

• LFTs often normal unless complication e.g cholangitis, pancreatitis, stone, stricture, tumor, etc
• Cross-sectional imaging (MRI/MRCP preferred)
  – Confirm cyst/type
  – Assess cyst communicates with the biliary tree
  – Associated mass?
• High index of suspicion: Dilated bile duct or cystic liver lesion(s)

Case

• 12 year old female presents with abdominal pain
• Imaging: acute appendicitis and focal dilation of the bile duct (17 mm)
• MRCP
Case

- 23 year old female
- Intermittent RUQ, epigastric and lower quadrant pain, plus nausea of 4 months duration
- MRCP

Case

- 61 year old female presents to an OSH with AP
- Imaging showed distal biliary stricture with upstream dilation of 3.5 cm. LFTs elevated
- Working Dx distal CBD stricture
- ERCP performed with stent placement
- 1 week later worsening pain
- EUS: No pancreatic mass, normal distal cbd
Type 1

- 50-85% of all biliary cysts
- They only involve the extrahepatic duct
  - A: Cystic dilation involves the CBD, part or all of the CHD and extrahepatic portions of the L&R HD
    • Associated with an APBJ
  - B: Focal, segmental dilation of an extrahepatic bile duct (often the distal CBD)
    • Not associated with an APBJ
  - C: Smooth, fusiform dilation, usually from the PBJ to the extrahepatic portions of the L&R HD
    • Associated with an APBJ


Laith H. Jamil, MD
MRCP

ACG 2014
Copyright 2014 American College of Gastroenterology
ACG 2014 Annual Postgraduate Course
ACG 2014 Annual Postgraduate Course • October 18-19, 2014
Management

- Obstructive lesion \( \rightarrow \) EUS &/or ERCP
- Surgical excision and creating a Roux-en-Y hepaticojejunostomy
- Distal Margin:
  - Preop: MRCP, EUS, &/or ERCP
  - Intraop: Cholangiography, choledochoscopy
- If previously managed by a cystenterostomy \( \rightarrow \) surgery (30% post cystenterostomy risk of malignancy)

Timing in Neonates

- Unclear
- Early surgical intervention if
  - Progressive intrahepatic ductal dilation
  - Cyst enlargement
  - Deterioration of liver function
- Harbingers of obstruction and/or cholangitis
- External drainage procedures (as a bridge)
  - Nutritional compromise
  - Acute infection
Type II

• True choledochal diverticula arising from the extrahepatic duct
• Communicate with the bile duct through a narrow stalk
• 2% of biliary cysts

Management

• DDX: pancreatic, mesenteric, and hepatic cysts
• If in doubt about Dx → hepatobiliary scintigraphy or ERCP
• Treatment: Surgical excision
Case

- 82 year old male with RYGB presents with epigastric pain
- LFTs: TB 18 mg/dl, AP 258 U/L
- MRCP

DBE-ERCP
Type III (AKA Choledochoceles)

- Cystic dilations are limited to the intraduodenal portion of the distal CBD
- 1-5%
- Lining: duodenal epithelium or biliary epithelium (increased risk of malignancy)
- Several subtypes

Law R, Topazian M Gastroenterology 2014;12:196-203
Most Common Types

• Type A: The intramural bile duct opens into a cystically dilated segment, which communicates to the duodenal lumen via a separate orifice
• Type B: The bile duct that opens normally into the duodenal lumen, with the choledochoele arising as a diverticulum of the intra-ampullary common channel

Management

• Symptomatic: Treat
• Asymptomatic:
  – Type IIIA: Endoscopic sphincterotomy (followed by biopsy of the cyst epithelium), and/or endoscopic snare resection
  – Type IIIB: Surgical or endoscopic resection
• Follow up: Endoscopic biopsies of the cyst mucosa a year later (assess for dysplasia)
**Type IV**

- Multiple cysts
- 15-35%
- Type A: Both intrahepatic and extrahepatic cystic dilations
- Type B: Multiple extrahepatic cysts only

**Management**

- Surgical excision and creating a Roux-en-Y hepaticojejunostomy
- If previously managed by a cystenterostomy for symptomatic relief → surgery (30% post cystenterostomy risk of malignancy)
Symptomatic Type IV A

- If patient becomes symptomatic from residual intrahepatic cysts
  - Segmental hepatectomy
  - Surgical un-roofing
  - Liver transplantation in some cases

Case

- 84 year old male
- Presents with flank pain and hematuria
- Imaging showed kidney cysts and liver cyst
- MRI/MRCP
Case

- 30-year-old male with presumed cirrhosis secondary to congenital hepatic fibrosis
- Presents with abdominal pain, fever and mild abnormalities in LFTs (TB 1.7, AP 155)

Type V

- Single or multiple intrahepatic cystic dilations alone
- 20% of these cysts
- Caroli disease
  - Less common
  - Without other apparent hepatic abnormalities
- Caroli syndrome
  - More common
  - Associated with congenital hepatic fibrosis
**Carolí**

- Autosomal recessive
- Associated with autosomal recessive polycystic kidney disease (ARPKD)
- Saccular or fusiform dilatation of bile ducts → stagnation of bile → biliary sludge and intraductal lithiasis
- Bacterial cholangitis → septicemia and hepatic abscess formation
- Secondary biliary cirrhosis can occur due to biliary obstruction

---

**Diagnosis & Management**

- Imaging
- Rarely need liver biopsy
- Largely supportive (antibiotics, ERCP, cholangioscopy, EHL, ESWL)
- Hepatic resection: Disease limited to a single lobe
- Ursodeoxycholic acid: Dissolve intrahepatic stones
- Liver transplantation: Extensive disease and frequent complications

---

Okugawa T et al Gastrointest Endosc. 2002 Sep;56(3):366-71
Cancer Screening

- No proven effective method of screening for dysplasia or intramucosal cancer
- Case reports of EUS, IDUS, Cholangioscopy
- Periodic MRI/MRCP

Non Surgical Management

Type I, II, or IV

- Periodic imaging: Unproven value
- Can consider yearly imaging studies (MRI/MRCP, CT with contrast, or IDUS), particularly if findings will alter patient management
- Patient who refuse surgical resection/poor surgical candidates
  - Laparoscopic cholecystectomy
  - Endoscopic sphincterotomy
  - Endoscopic stent placement
Follow Up

- No clear guidelines
- Post-excisional malignant disease: 0.7%-6 %
- Type III cysts treated with endoscopic sphincterotomy: Endoscopic biopsies of the cyst mucosa a year later
- Post total cyst excision: Annual LFTs to screen for anastomotic biliary stenosis (25%)

Take Home Points

- High index of suspicious in unexplained cystic dilations of the biliary tree
- High risk for malignancy
- Patient with APBJ are at increase risk for GB cancer → strongly consider Cholecystectomy
- Cancer screening: Not proven
Take Home Points

• Type I → Surgery

• Type II → Surgery

• Type III → Endoscopic therapy

Take Home Points

• Type IV → Surgery

• Type V → Supportive/symptomatic treatment. May require surgery/liver transplantation

• Consider post surgical FU
Thank You