Choledochal Cyst

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History

- 1723: First described by Vater and Ezler
- 1924: First choledochal cyst excision described by MacWorter
  - original procedures involved external or internal drainage along with cholecystectomy
- 1959: Alonso-Lej and colleagues proposed first classification system
- 1969: Babbitt described anomalous pancreaticobiliary duct union (APBDU)
- 1977: Todani et al described classification system based on site of cystic change
- 1995: Farello first described laparoscopic resection of a choledochal cyst
Epidemiology

- ~80% diagnosed in infants and young children within first decade of life
- Incidence
  - 1 in 100,000 to 1 in 150,000 in western countries
  - Much higher in Asian populations, as high as 1 in 1,000 to 1 in 13,000 in Japan
- Four times more common in females
Pathophysiology

- Anomalous pancreaticobiliary duct union (APBDU)
  - common bile duct and pancreatic duct junction occurs outside the duodenum
  - reflux of pancreatic fluid into the biliary tree
  - believed to be secondary to arrest in migration of the choledochopancreatic junction into the duodenal wall
- Long common channel
  - defined as insertion of the CBD farther than 15 mm from the ampulla of vater
APBDU

Source: Soares KC, et al.
Pathophysiology

- 80-96% of pediatric choledochal cysts are associated with APBDU
- Nagi et al:
  - series of 2,885 patients undergoing ERCP
    - ~90% of patients diagnosed with APBDU had a choledochal cyst
- Other mechanistic hypotheses for choledochal cysts include a weak bile duct wall, sustained increased intrabiliary pressure, inadequate autonomic innervations, sphincter of Oddi dysfunction, and distal obstruction of the CBD
Classification

- Multiple classification systems
- Todani (1977): most widely accepted
  - based on site of cystic change
  - five types
    - Type I (80-90% of all choledochal cysts)
    - Type IV (15-20%)
<table>
<thead>
<tr>
<th>Type</th>
<th>Description</th>
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</thead>
<tbody>
<tr>
<td>I</td>
<td>Cyst confined to the extrahepatic bile duct</td>
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<tr>
<td>IA</td>
<td>Diffuse dilatation</td>
</tr>
<tr>
<td>IB</td>
<td>Focal dilatation</td>
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<tr>
<td>IC</td>
<td>Fusiform dilatation</td>
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<tr>
<td>II</td>
<td>True diverticula of the extrahepatic duct</td>
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<tr>
<td>III</td>
<td>Choledochocoele</td>
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<tr>
<td>IV</td>
<td>Multiple cystic dilatation</td>
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<tr>
<td>IVA</td>
<td>Both intrahepatic and extrahepatic duct involvement</td>
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<tr>
<td>IVB</td>
<td>Only extrahepatic duct</td>
</tr>
<tr>
<td>V</td>
<td>Caroli disease</td>
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</tbody>
</table>
Classification

Type Ia

Type Ib

Type Ic

Type II

Type III

Type IV

Type V

Source: Gonzales KD, Lee H.
Type I

- Most common (at least 90% of cases)
- Cystic/saccular or fusiform dilation of common bile duct
- Three subtypes
  - Ia - cystic dilation of the entire CBD (gallbladder arises from the choledochal cyst)
  - Ib - cystic dilation of a segment of the CBD
  - Ic - Fusiform dilation of the CBD
Type I

- Typically appear as anechoic cystic lesions which communicate with the biliary tract
- May be associated with mild enlargement of the intrahepatic bile ducts secondary to bile stasis
Type I (CT, MRCP, ERCP)

Source: Soares et al.
Type II

- True diverticula of the CBD
- No dilation of the common bile, extrahepatic, or intrahepatic ducts
- Represent ~2% of reported cases
- Appear as anechoic cysts juxtaposed to the CBD with a normal appearing gallbladder and CHD
- Can resemble gallbladder duplication
Type II (CT, MRCP, MRCP)
Source: Soares KC, et al.
Type III

- Also known as choledochoceles
- Comprise 1-4% of cases
- Characterized by intraduodenal location at the pancreaticobiliary junction
- More evenly distributed between sexes (as opposed to other CC)
- Much lower incidence of malignant transformation
- Biliary tract symptoms less common; pancreatitis commonly seen
- APBDU much less commonly seen with choledochoceles
Type III

Source: Gonzales KD, Lee H.
Type III (MRCP, ERCP)
Source: Cha SW, Park MS, Kim KW, et al.
Type IV

- Multiple cystic dilations of biliary tree
- Can include both intra- and extrahepatic duct involvement
- Two subtypes:
  - IVA - extends from the CBD and CHD into the intrahepatic biliary tree
    - primary ductal stricture around hepatic hilum commonly seen
  - IVB - multiple dilations of the extrahepatic biliary tree (classically described as ‘string of beads’ appearance) without intrahepatic involvement
Type IVA - CT

Case courtesy of Dr Mohammad Taghi Niknejad, Radiopaedia.org, rID: 20611
Type IVA - CT (cont’d)

Case courtesy of Dr Mohammad Taghi Niknejad, Radiopaedia.org, rID: 20611
Type V

- Also known as Caroli’s disease
- Intrahepatic saccular or fusiform dilation with no underlying obstruction or extrahepatic biliary tree involvement
- Thought to arise from ductal plate malformation
- Can be associated with polycystic kidney disease
- When associated with congenital hepatic fibrosis - Caroli’s syndrome
- Enhancement of portal vein surrounded by dilated intrahepatic bile ducts (“central dot sign”)
Type V (MRCP)
Case courtesy of Dr Frank Gaillard, Radiopaedia.org, rID: 8362
Type V (US, MRCP, MRCP)
Source: Soares KC, et al.
Carolí disease, “central dot sign”
Source: Levy, AD
Clinical Presentation

- Usually diagnosed in childhood (in utero and adult diagnosis also)

- Classic triad (only seen in ~20%)
  - abdominal pain
  - jaundice
  - RUQ mass

- Rare to see complete triad, but more commonly seen in pediatric patients as opposed to adults

- May present with cholangitis, pancreatitis, portal hypertension, LFT abnormalities
Clinical Presentation

- Patients can manifest symptoms at any point in life
- 80% of patients are symptomatic before age 10
- Classification based on age at presentation:
  - ‘Infantile’ (<12 mo) – obstructive jaundice, acholic stools, hepatomegaly (all similar to biliary atresia)
  - ‘Adult’ (>12 mo) – more symptoms including fever, nausea/vomiting, jaundice
Adult vs Pediatric

- **ADULTS**
  - more likely to present with biliary or pancreatic symptoms and abdominal pain

- **CHILDREN**
  - more likely to present with abdominal mass and jaundice

- Cyst rupture - rare, typically only seen in neonates and infants
Diagnosis

- More choledochal cysts are being diagnosed as incidental findings secondary to increased use of imaging modalities.

- Diagnosis usually established using multimodality approach (US, CT, MRI):
  - Ultrasound most frequently used.
  - Cholangiography (ERCP, PTC) is the most sensitive technique to define anatomy - may be difficult to perform in pediatric population.
  - MRCP gaining popularity:
    - Highly sensitive (70-100%) and specific (90-100%) in choledochal cyst diagnosis and classification.
Pathology

- Fibrosis of cyst wall lined with columnar epithelium and lymphocytic infiltration typical of pediatric CC

- Adult CC includes evidence of inflammation and hyperplasia

- Most show some degree of change in liver including portal fibrosis, central venous distention, parenchymal inflammation, and bile duct proliferation
Pathology

- Type I (and sometimes Type IV) lack biliary mucosa
- Type II closely resembles gallbladder duplication
- Type III lined by duodenal mucosa
- Type V can have extensive hepatic fibrosis
- Increased risk of malignant transformation with age
  - half of patients >50 years old have invasive biliary neoplasm
  - less than 1% have invasive biliary neoplasm before the age of 10
Pathology

- Malignancy
  - most commonly associated with Type I and IV cysts
  - II, III, and V have minimal neoplastic risk
  - believed to occur through multistep genetic events (early K-ras and p53, late DPC-4)
  - most cases of malignant transformation are cholangiocarcinoma (gallbladder carcinoma reported in 10-25% of CC-related malignancies)
Management

- Risk of malignant transformation warrants complete and total excision whenever possible

- Fetal and newborn diagnosis is associated with early progression to liver fibrosis (particularly type IV)
  - RCT has shown that early excision in prenatally diagnosed asymptomatic cysts resulted in significantly less hepatic fibrosis

- Early excision recommended
Type I & IV

Management:

- complete extrahepatic bile duct excision down to level of communication with the pancreatic duct
- cholecystectomy
- restoration of bilioenteric continuity
- hepatectomy warranted in type IVA cysts with a significant intrahepatic component likely to result in complications if not removed
Bilioenteric reconstruction

- Roux-en-Y hepaticojejunostomy preferred
- Hepaticoduodenostomy has been reported but no longer reconstruction of choice
  - associated with increased rates of gastric cancer (due to bile reflux) and biliary cancer
  - significantly more cases of postoperative reflux and gastritis
- wide anastomosis imperative - prevent stricture
Type II & III

- Extremely low risk of malignant transformation
- Type II
  - diverticulectomy
  - primary CBD closure at the diverticulum
- Small choledochoceles
  - endoscopic sphincterotomy
- Large choledochoceles
  - trans-duodenal excision, especially if associated with complications
Type V

- Liver resection or orthotopic liver transplant (OLT)
- Localized or unilobar cystic disease - hepatic resection
  - incomplete resection leads to poor long-term outcomes
- Asymptomatic bilobar disease - non-operative with aggressive surveillance
- Complicated bilobar disease (cholangitis, portal hypertension, suspicion of malignancy) - OLT
Laparoscopic Approach

- Laparoscopic resection with roux-en-Y hepaticojejunostomy shown to be safe with comparable outcomes to open approach in retrospective studies

- Reported advantages:
  - improved intra-operative visualization of deeper structures
  - decreased post-operative pain
  - shorter hospital stay
  - improved cosmetic result
  - decreased post-operative ileus
- Tang et al. (2011, China)
  - Retrospective analysis of 62 children undergoing laparoscopic resection
  - 8.2% morbidity (bile leak, adhesive SBO, intestinal necrosis, cholangitis, anastomotic narrowing)

- Ono et al. (2010, Japan)
  - Retrospective review of 56 patients undergoing open resection and reconstruction
  - 16/56 late complications within 10 years (biochemical liver dysfunction, persistent dilation of intrahepatic ducts, recurrent abdominal pain, recurrence of CBD adenocarcinoma)
Outcomes

- Post operative morbidity and mortality typically lower in children compared to adults
  - Late complications occur in up to 40% of adult patients (stricture, cancer, cholangitis, cirrhosis)
- Type IVA cysts most commonly associated with post-operative complications
Prognosis

- Overall, resection has good prognosis
  - 89% event free rate
  - 5 year OS rates >90%
- Risk of malignancy
  - Remains elevated, even after excision
  - Choledochal cyst associated biliary malignancy associated with extremely unfavorable outcomes (median survival 6-21 months)
Surveillance

- Long term surveillance indicated, especially given risk of malignancy
- Particularly important in cases with persistent intrahepatic biliary dilatation
- Regular biochemical evaluation and abdominal US or cross-sectional imaging
Sources


- Levy AD. Biliary Tract - Pathology. [www.radiologyassistant.nl](http://www.radiologyassistant.nl)

sources


