Congenital Dilations of the Biliary Tract

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Facts

- Choledochal cysts are abnormal dilations of the bile ducts, congenital in origin.
- Usually a surgical problem of infancy or childhood, but ~20% present in adults.
- Associated with anomaly involving the anatomy of the pancreatobiliary duct junction (APBDJ).
- Clinical presentation and treatment: Children and Adults:
  - Children are more likely to present with jaundice and/or a mass.
  - Adults present incidentally or on evaluation of symptoms related to the long-term consequences/complications.

DIAGNOSIS -Classification-

Alonso-Lej’s classification
- Applies only to cyst of the common choledocho.
- Two cases and analysis of n = 94

Todani’s Modification
  - N= 17 = Type I
  - N= 0 , Type II or III
  - N= 20, Didn’t fit Alonso-Lej’s classification


Alonso-Lej F, Reiner WB Jr., Passaglia D. Congenital Bile Duct Cysts—New After Surg; 1959; 208, 1
**DIAGNOSIS -Etiology-**

- **Hypothesis:** Anomalous arrangement of the pancreatobiliary ductal junction.
- **Long common channel –Pancreatic juice**
  - Inflammation, ectasia, dilation.
- **Pancreatobiliary duct:** leads to cystic dilation of the extrahepatic BD.
- **Not all patients with BD cysts have APBDJ and not all pts with APBDJ have BD cyst.**
  - The prevalence of APBDJ ranges from ~20-90%


**DIAGNOSIS -Demographics-**

- **Biliary Cysts are uncommon (~ 1% of benign biliary Dz)**
- **Geographic population Prevalence: Asia**
  - > 1/3 of cases have been reported from Japan.
  - Frequency of 1 in 100,000 to 1 in 2 million live births in western countries.
- **More common in females: F to M ratio of 3:1 to 8:1**
- **Prevalence: Based on selected review of the world’s literature (35 reports, 1959-2004).**

<table>
<thead>
<tr>
<th>Todani Cyst Type</th>
<th>I</th>
<th>II</th>
<th>III</th>
<th>IV</th>
<th>V</th>
</tr>
</thead>
<tbody>
<tr>
<td><em>Total # Cases</em></td>
<td>2365</td>
<td>83</td>
<td>102</td>
<td>457</td>
<td>28</td>
</tr>
<tr>
<td>Percentage</td>
<td>78</td>
<td>3</td>
<td>3</td>
<td>15</td>
<td>1</td>
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</tbody>
</table>


**Clinical Presentation in Adults**

- **May remain Asymptomatic indefinitely.**
- **<20 % of patients with BD cysts present as Adults (> 16 y/o).**
- **When Symptomatic--- intermittent.**
  - Similar to Biliary Stone dz regardless of cyst type.
  - RUQ pain, Fever, & mild jaundice.
- **Cholangitis—most common Sx.**
- **~15% adults present with evidence of cirrhosis or Portal HTN.**
  - Signs of chronic Liver Dz. (ascites, muscle wasting, etc.)
- **Other Presentations:**
  - Pancreatitits- (30-70%), N, V and intense pain.
  - Weight loss; unusual, but might indicate malignancy.

**Imaging**

- **Imaging studies should clearly define the relationship of the cyst to the other portal structures.**
- **Studies:**
  - Ultrasonography
  - Hydroxymidoacetic acid scan (HIDA)-limited value.
  - Computed Tomography (CT) +/- cholecystography
  - Magnetic resonance cholangiopancreatography (MRCP)
  - Endoscopic retrograde cholangiopancreatography (ERCP).
  - Percutaneous transhepatic cholangiography (PTC)
Ultrasonography

- Non-invasive, low cost.
- Permits evaluation of liver and pancreas.
- Shows hypoechoic dilation of BD.
- High Resolution US:
  - Focal duct wall thickening or nodularity.
  - Intracystic stones: echogenic features and acoustic shadowing.
- Limited use in detecting Type III cyst because of bowel gas.

Cholangiography

- Some form of Cholangiography (MRI or CT) is necessary in all patients.
- Defines cyst anatomy: Location, shape and extent of duct involvement.
- Note: all cyst communicate with the ductal system.
- Direct Cholangiography: ERCP and/or PTC.
  - Stones, strictures, papillary projections.
  - Typically, the BD joins the PD ~ 2-4 cm proximal to the duodenum to the long common channel (APBDJ)

Complications of Biliary Cysts

- Cystolithiasis - most frequent condition (2-72%).
  - Uncommon in children.
- Hepatolithiasis - seen with long-term F/U, may occur with or without associated stricture.
  - Usually occurs in Type IV bile duct cysts.
  - Can lead to intrahepatic abscess and/or lobar atrophy.
- Calculous Cholecystitis: Acute or chronic.
- Pancreatitis.
- Cirrhosis with portal Hypertension.
- Malignant Degeneration-Cholangiocarcinoma (2.5-28%)
  - 20-30 X than general population.
  - Age related: 0.7% first decade; > 14% after age of 20 year of age.
  - Only 57% are intracystic.

Malignancy After Incomplete Excision

30 year old woman with history of choledochal cyst who presented with postprandial pain.

- Upper GI shows a filling defect (arrows) in the choledochal cyst
- Axial CT with intravenous contrast confirms a large heterogeneously enhancing mass (arrows) in the common bile duct.
Malignancy in Unresected Cyst

29 year old woman with history of type I cyst treated with a Roux-en Y hepatojejunostomy bypass without cyst resection.

PTC-shows extrinsic compression in the medial aspect of the Roux limb by a large mass (arrows).

Axial CT with contrast demonstrates an 8 cm heterogeneous mass (arrows) centered at the head of the pancreas.

Management

-Surgical-

- Preoperative Planning:
  - Clear delineation of biliary and pancreatic duct anatomy by direct or indirect cholangiography is critical.
  - If extensive unilateral hepatic involvement (usually the left side) is found, complete cyst excision with concomitant partial hepatectomy should be planned and discussed with the patient in the preoperative setting.

- Always anticipate coexisting malignancy in adults:
  - weight loss, jaundice, elevated tumor markers, mass or intracystic mural nodules.

Surgical Technique

Pertinent Anatomy:

Surgical Technique
-OPEN-

- **Choledochal Cyst Excision**
  - Cyst is obvious bulging from the lateral edge of the HD ligament.
  - Cholecystectomy is started by incising the peritoneum overlying the dilated and distorted gallbladder neck. A top-down dissection is used to mobilize the gallbladder from the liver bed.
  - A Kocher maneuver.
  - Peritoneum is incised on the superior border along the first portion of the duodenum and it is reflected downward and rolled inferiomedially to expose the anterior cyst wall.
  - The lower end of the cyst is encircled with a vessel-loop and used for traction.
  - Dissection progresses inferiorly along the distal CBD duct to achieve a "complete" cyst excision.
  - Note: care must be taken not to injure the underlying pancreatic duct.

Surgical Technique
-Cyst Dissection-

- Normal-sized distal CBD is divided and over sewn with a 3-0 or 4-0 absorbable suture depending on the duct size.
- The proximal divided end of the cyst is also over sewn to avoid bile spillage.
- The cyst is then reflected upward to allow dissection away from the portal vein and the hepatic artery.

Surgical Technique
-Alternative Approach-

- **Note:** rarely, intense inflammation of the cyst wall is found as a result of subclinical cholangitis or pancreatitis.
- An internal cyst dissection approach should be done.
• If preoperative imaging and intraoperative findings demonstrate a transition to a normal caliber common hepatic duct, the specimen is divided at this point.
• For cyst with intrahepatic extension, the hilar plate is lowered, and the hepatic bifurcation is dissected to expose both the takeoff of the right and left ducts.
• Under these circumstances, the hepatic confluence is included in the specimen.

Surgical Technique
-Cyst Excision-

Surgical Technique
-Biliary-enteric Reconstruction-

Surgical Technique
-Minimally Invasive Approach-
LAPAROSCOPIC
ROBOT-ASSISTED

• Pertinent steps for the minimally invasive surgical management approach of bile duct cyst include:
  – Jejuno-jejunostomy:
  – Alternatively, this step can be omitted completely by choosing to instead reconstruct by hepatico-duodenostomy.
  – MIS hepaticojejunostomy:
Surgical Technique
-Minimally Invasive Approach-
LAPAROSCOPIC
ROBOT-ASSISTED

POSTOPERATIVE MANAGEMENT
• Emphasis is placed on controlling post-operative incisional pain.
• Observation for signs of common early complications such as bleeding, ileus, wound infection and/or biliary leakage.
• Abdominal drainage management.
• For long-term surveillance, patients are seen in follow-up at 6 and 12 months, and annually thereafter.
• Liver function tests and a post-operative radionucleotide scan are obtained to document the patency of the anastomosis and are useful as baseline studies.

RESULTS
• Long-term follow-up data (ranging from 17 - 25 years) from several studies has shown that primary complete cyst excision with a WIDE bilioenteric anastomosis is the treatment choice for bile duct cysts.
• Choice for biliary reconstruction remains controversial.
• Nevertheless, good long-term outcomes can be expected regardless of the biliary reconstruction technique chosen, as long at a complete cyst excision can be done.

CONCLUSIONS
• The term Choledochal cysts, should be abandoned and exchanged for Congenital Bile Duct Cysts.
• Bile duct cysts carry a substantial risk for malignant transformation and should be completely excised.
• The preferred operative management of these cysts has begun to shift toward a minimally invasive surgical approach.
• While the laparoscopic technique has not been universally embraced, data supporting its feasibility, safety and efficacy continues to mount.
CONCLUSIONS

• Bile duct cysts affect young females most commonly and they have the most to gain from the cosmetic benefits of this approach.

• While the laparoscopic approach is gradually gaining acceptance, the role of robotic-assisted resection is even more unclear, especially given its high cost and the current financial constraints of our healthcare system.