A patient with an unusual congenital anomaly of the pancreaticobiliary tree

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Review of “Normal” Pancreaticobiliary Tract Anatomy

http://www.anabasisdev.com/cadeR/cmt_images/liver%20anatomy.JPG
(A) 30 days postfertilization: initiation of dorsal and ventral pancreatic buds on opposite sides of the gut tube. Ventral bud arises from a common outgrowth that will form the liver and gall bladder.

(B) Later, the ventral bud moves posteriorly to join the dorsal bud.

~ 6 weeks into development: fusion of the dorsal and ventral buds occurs in most people. The main duct is derived from the distal part of the dorsal bud and the proximal part of the ventral.

Review of “Normal” Pancreaticobiliary Tract Anatomy
Our Patient A.P.: History

A.P. is a 28 yo woman who presents to her PCP c/o intermittent, sharp epigastric pain radiating to the back for 10 years

- Lasts 2-6hrs, then resolves spontaneously
- ~once a week
- No improvement with antacids
- No diarrhea, constipation, appetite/weight/energy change, hematemesis, melena, hematochezia, fevers or chills.
Our Patient A.P.: History

PMH:
- Pancreatitis at age 16 and 22 of unknown etiology
- No prior operations

FH: NC

SH: No alcohol, nonsmoker, no drugs.

Meds: None, including OTCs.

PE: Comfortable. ABD soft, nontender, nondistended. No pain on palpation. Normal bowel sounds.

STUDIES:
- A recent upper endoscopy was wnl.
  - Biopsy urease test and culture were negative for H.Pylori
Clinical Differential of Epigastric Pain

1. Peptic ulcer disease
2. Gastroesophageal reflux disease
3. Gastritis
4. Pancreatitis
5. Myocardial infarction
6. Pericarditis
7. Ruptured aortic aneurysm

Fishman et al. UpToDate, 2007: Differential diagnosis of abdominal pain in adults
RUQ Ultrasound

- Because of A.P.’s h/o recurrent pancreatitis, an ultrasound was obtained to investigate pancreas and biliary tree

  - **U/S INDICATIONS:**
    - Acute RUQ pain
    - Suspected GB pathology
    - W/U jaundice and duct dilation
    - Guidance for biopsy/drainage
    - Liver evaluation
      - Typification of masses
      - Evaluation of parenchyma when CT/CT or IV contrast contraindicated
Our patient A.P.: RUQ Ultrasound

Patient A.P.
Abdominal Ultrasound

CBD TRV

1 L 0.69 cm
Our Patient A.P.: Abdominal Ultrasound Findings

- **Gallbladder:**
  - No stones or pericholecystic fluid. No edema in GB wall.

- **Biliary Tree:**
  - Intrahepatic ducts normal
  - Extrahepatic dilatation of the common bile (nl < 6mm) and cystic ducts
    - No stones are visualized in the common bile duct
    - No irregularity is noted along the length of the extrahepatic ducts.

- **Liver, pancreas and spleen are unremarkable**
DDx: Extrahepatic Bile Duct Obstruction

**Periportal**
- Cholangiocarcinoma
- GB Carcinoma
- HCC
- Metastases

**Mid-Duct**
- Pancreatic Cancer
- Bile Duct Cancer
- Iatrogenic Stricture
- Metastases
- Sclerosing cholangitis
- Pancreatitis
- CBD Stone

**Peri-Ampullary**
- Stone
- Pancreatic Cancer
- Periampullary Cancer
- Ampullary stenosis
- Choledochal Cyst
- Choledochocele
- Duodenal Diverticula

Gillianlieberman.com: The Gallbladder and Biliary Tract
http://www.anabasisdev.com/cadeR/cmt_images/liver%20anatomy.JPG
Our Patient A.P.: Further Studies

• Further Imaging was indicated to evaluate the biliary tract
  – ? ERCP vs MRCP
Endoscopic Retrograde Cholangiopancreatography (ERCP) vs Magnetic resonance cholangiopancreatography (MRCP)

**ERCP**
- Developed in 1970s for dx imaging of pancreatic and biliary system
- Current applications:
  - **Diagnostic**: cystic duct leaks, clarification of complex ductal anatomy/equivocal MRCP
  - **Therapeutic**: stone extraction, sphincterotomy, stricture dilation, stent placement, bx
- Advantage: Diagnostic and therapeutic
- Major Disadvantage: 5% complication rate (pancreatitis, hemorrhage, GI perforation)

**MRCP**
- Developed in early 1990s, it is now accepted as accurate technique to image pancreaticobiliary tract
  - Indications: Eval. of GB and extrahepatic bile ducts (ductal dilation/stenosis/obstxn, anamalous biliary anatomy
- Utilizes long T2 relaxation time of pancreatic and biliary secretions
- Advantages:
  - non-invasive
  - no ionizing radiation
  - no contrast required
- Disadvantage: solely diagnostic

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**Plan:** Will get MRCP due to risk of pancreatitis with ERCP.

Fulcher et al., Radiol Clin N Am 2002 (40): 1363-76
Ahmad, et al. Radiol Clin N Am 2002 (40): 1377-95
Pictures from:www.pennhealth.com/gi/prepare/ercp
FINDINGS:

1. Smooth stenosis of the distal CBD (short THICK arrow) with mild upstream dilation.

2. Normal pancreatic duct (arrowheads).

3. An accessory branch (thin arrow) of the pancreatic duct is noted.
MRCP Demonstrates 2 Abnormal Findings: Which one is responsible for her pain?

1. Stenosis of the distal CBD
   - May be due to operative injury, radiation, trauma, chronic pancreatitis, PSC or malignancy.
   - Pancreatitis: obstruction of the intrapancreatic segment of the CBD occurs in patients with chronic pancreatitis and accounts for ~10% of benign strictures
   - Presentation: Most asx. +/- Biliary colic, jaundice with severe dilation.

2. Abnormal “accessory” branch of PD
   - Unclear anatomy on MRCP
MRCP + Secretin

- **Secretin**
  - Hormone produced by S-cells of SI → stimulation of bicarbonate and fluid secretion by pancreas → distention of pancreatic ducts followed by dumping of fluid into duodenum
  - Helps to better detect anatomic variants and strictures
  - Thought to be better than MRCP alone

- MRCP + secretin was ordered to visualize the anatomy, flow characteristics and to try to reproduce the pain, which if associated with flow, would likely be diagnostic.

Remer et al., Radiol Clin N Am. 2002 (40): 1229-1242
Our Patient A.P.: MRCP + Secretin

Baseline

4 min

10 min

Gallbladder (arrow) is empty and no fluid is noted in the second portion of the duodenum (arrowheads).

• Unexpected filling of the gallbladder (arrow).

• Delay of duodenal filling (arrowheads).

There is increased distension of the gallbladder (arrow) and limited amounts of pancreatic secretions (arrowheads) in the duodenum.
MRCP + Secretin Results

- The MRI + secretin study demonstrated impaired flow of pancreatic secretions after a dose of secretin was given.

- Anatomy of “accessory branch” still unclear.
Step 2: Evaluation of unusual
“accessory branch” of pancreatic duct

- ERCP was ordered to further delineate anatomy.
Our Patient A.P.  ERCP-Cannulation via Major Papilla

Cannulation of major papilla with wire in the CBD (black arrows). Filling of aberrant duct (white arrow) that connects with pancreatic duct (black arrowheads) draining in the minor papilla (white arrowhead)
Repeated ERCP after cannulation of the minor papilla (white arrowhead) confirms the presence of a small aberrant branch (white arrow) of the pancreatic duct communicating with the CBD (black arrow). Retrograde filling of the CBD (black arrowheads) was observed.
Discussion

- Application of various imaging modalities, discovered a reason for A.P.'s recurrent pancreatitis and pain sx
  - Free flow between the pancreatic and bile ducts results in activation of pancreatic enzymes and inflammation $\rightarrow$ pancreatitis, pain, increased risk of malignancy
  - She was diagnosed with a congenital accessory bile duct that connected her Pancreatic Duct (of Santorini) to her main common bile duct

- A.P. underwent surgery and is now free of sx, 4 months later.