Case Presentation

- BG G
- Born by CS on 9/11/2008 full term – 38 4/7 weeks
- Apgar scores were 8,9
- Birth weight 3335 gms
- Congenital endocushion heart defect diagnosed in utero
- Mother was 26 yrs old this was her 2nd pregnancy – the first was uneventful
Physical exam

- Pink, acyanotic, RA sat O2 was 87%, 96% on 0.1 l/min
- Systolic II/IV murmur at the left sternal border
- no dysmorphic features
- No palpable abdominal masses
- No acholic stools
- Feeding well
Initial Workup

- Echocardiogram – 2 chamber heart
  - Dextrocardia
  - Single atrium
  - Large RV, very small LV
  - Aorta originating from the RV,
  - Transposed great vessels
  - Pulmonic stenosis

- Abdominal US
  - Choledochal cyst

- Total bili 5.8 Direct bili 2.3 LFTs wnl
Dextrocardia
MRCP

High T2 signal structure just inferior to the intrahepatic ductal confluence with communication to the gallbladder – suspicious for choledochal cyst
HIDA – Tc99m

No clearance of radiotracer at 24 hours in the hepatic or common bile ducts suggestive of biliary atresia
Operative Intervention

- DOL #15
- Exploratory laparotomy, intraoperative cholangiogram, liver biopsy, broviac catheter placement
- Findings:
  - Preduodenal portal vein
  - Choledochal cyst
  - Polysplenia
  - Malrotation: ileo cecal junction on the left and the colon going to the right
Choledochal cyst
www.downstatesurgery.org
Intraoperative cholangiogram

- Bilobed choledochal cyst
- Flow into the small bowel
- Arborization of the hepatic ducts
Pathology

- Liver biopsy:
  - Intrahepatic duct paucity compatible with intrahepatic biliary atresia
• Contrast sweeps into the small bowel in the right upper quadrant.
• Compatible with mid gut malrotation
• No obstruction
MR of the abdomen

- Preduodenal portosplenic confluence, portal vein
- Choledochal cyst
- Multiple small soft tissue masses in the gastroplenic ligament consistent with polysplenia
- Aorta is anterior and to the right of the midline at the level of the diaphragm
- Sma is to the right of the smv
- No intrahepatic IVC
- Duplicated IVC
Heterotaxy syndrome/ Situs Ambiguous

- Abnormal arrangement of body organs or complete situs inversus
- Major cardiac anomalies
- GI anomalies
  - Midline liver
  - Malposition of stomach
  - Anomalies of intestinal rotation and fixation
    - Including malrotation, preduodenal portal vein
  - Intraperitoneal pancreas
- Asplenia or polysplenia
OR Take 2

- DOL #30
  - By now: T.B 6.5
  - Ast/alt 30/20
  - GGT 371

- Operative Procedure :
  - Exploratory laparotomy
  - Lysis of adhesions
  - Cholecystectomy
  - Resection of choledochal cyst and atretic common hepatic duct up to the porta hepatis
  - Hepatportoenterostomy with a Roux en-Y anastomosis
  - Modified Ladd’s procedure – appendectomy
  - Did not have to bypass the retro portal vein duodenum
Gall bladder

Choledochal cyst

Gall bladder
Pathology

- Choledochal cyst – type II
- Hepatic duct; dense fibroconnective tissue containing a minute atretic duct
Post-operative course

- POD #2 extubated, bilious drainage from the JP drain
- POD #5 greenish stools! T.Bili 2
- POD #6 tolerates PO diet
- POD #25 with green/yellow stools, tolerating diet and starting to gain weight – discharge planning in progress
Questions are guaranteed in life; Answers aren't.

www.downstatesurgery.org

Questions?
Surgical causes of neonatal jaundice
Neonatal jaundice

- Usual accumulation of unconjugated bilirubin
- Conjugated through glucuronyl transferase
- Conjugated → water soluble → then excreted into the biliary system → GI tract.
- Newborns
  - high Hgb, Rh, ABO and rare blood group abnormalities leading to hemolysis
  - immature conjugating system
  - infections
- So can have transient accumulation of bilirubin in the tissues – physiologic jaundice of the neonatal period
- Evident day 5-7
- Usually goes away by week 2
Persistent neonatal jaundice

- Biliary atresia (60%)
- Neonatal Hepatitis (35%)
  - Hepatic inflammation that can be secondary to several different causes
    - cmv, syphilis, herpes, toxo
    - Metabolic defects: Alpha 1-antitrypsin deficiency, galactosemia
- Choledochal cyst (5%)
  - Paucity and hypoplasia of interlobar bile ducts
Spontaneous perforation of extrahepatic bile ducts
- Ascites, mild jaundice, failure to thrive
- Usually occurs at the junction between the cystic and common bile ducts

Inspissated bile syndrome – bile plug syndrome
- Extrahepatic obstruction of the bile ducts by biliary sludge
- Associated with massive hemolysis, hemorrhage, TPN, cystic fibrosis and other intestinal diseases such as Hirschsprungs
Choledochal cyst

- Congenital biliary tract disorder
- 1: 2 million – 1: 13,000
- Idiopathic dilation of the common bile duct
Etiology:

- Can be associated with an anomalous jcn of the pancreatic and common bile ducts
- Possibly an abnormal pancreatic and biliary duct junction with the formation of a common channel by which the pancreatic enzymes are secreted.
- Reflux of pancreatic enzymes into the bile duct
  This weakens the bile duct wall by enzymatic wall destruction → dilation → inflammation → cyst formation
- Infectious agents
- Biliary autonomic dysfunction

www.downstatesurgery.org
Presentation

- More common in females than males 4:1
- More common in Asians – Japanese
- Can present in the neonatal period
- Usually present in toddlers
- Classically: Abdominal pain, mass, jaundice
- More commonly: Episodic abdominal pain, minimal jaundice
- Pts can develop cholangitis, infected cysts, portal hypertension and cirrhosis
- Rarely: Present with bile peritonitis from a ruptured cyst
Diagnosis

- Frequently diagnosed in the fetus during a screening prenatal US
- Postnatally – a cystic structure seen on US originating from the biliary tree
- CT and MRCP can be used in order to elucidate the cyst’s relationship with the biliary tree, porta hepatis and delineate its anatomy
Classes

- **Type I** 50%—sacular or fusiform dilatation of the common bile duct
- **Type II** 5%—isolated diverticulum off of the CBD
- **Type III** 5%—choledochoccele
- **Type IV** 5-10%—Dilatation of both intrahepatic and extrahepatic biliary ducts
- **Type V** or Carolli’s 1%—cystic dilatation of intra hepatic biliary ducts


Copyright © The McGraw-Hill Companies, Inc. All rights reserved.
Untreated can cause cholangitis and cholangiocarcinoma

Risk of ca in the 1st decade – 0.7%

Increases in the 2nd decade to 14%

In types I, II, IV: excision of the extrahepatic biliary tree, cholecystectomy and with a Roux-en-Y hepaticojejunostomy

IV may require additional segmental resection

Type III - sphincterotomy
Treatment

- Used to do a cyst-enterostomy however many of these pts went on to develop cholangiocarcinoma
  - cholecystectomy
  - Cyst excision
  - Biliary enteric reconstruction
Prognosis

- Generally excellent
- Complications:
  - Anastomotic stricture
  - Cholangitis
  - Intrahepatic stones
Biliary Atresia

- Incidence 1/20,000
- Obliterative process of the extrahepatic bile ducts
- Associated with hepatic fibrosis
- Ladd and Gross: arrest of development during the solid stage of bile duct formation.
- Etiology unclear
- Acquired – immune vs systemic viral infections
- Congenital – association with polysplenia
Atretic ducts – solid fibrous cords that may contain occasional islands of biliary epithelium

Three patterns

- Proximal extrahepatic bile ducts are patent and ducts distal to the cystic duct are obliterated
- The gallbladder, cystic duct and CBD are patent and proximal ducts occluded
- Entire extrahepatic system is occluded

Over time the failure to excrete bile results in progressive periportal fibrosis and obstruction of the intrahepatic portal veins → hepatic cirrhosis
Presentation

- Grey or acholic stools – secondary to obstructed bile flow
- Failure to thrive
- Liver failure and portal hypertension
- Bilirubin > 3 mg/dl
- Alk phos 500-1000
- GGT > 300
Diagnosis

- Technetium-99m iminodiacetate (HIDA) after pre-treatment with phenobarbital (promotes tracer uptake)
- If radionucleotide appears in the intestine then the biliary tree in presumed to be patent
- Ultrasound can exclude choledochal cyst
- 10% of pts - gallbladder is visualized and the distal ducts are patent.
- Percutaneous liver biopsy can r/o neonatal hepatitis
Intraoperative cholangiogram

- Can use the gallbladder as a conduit
- Defines the extrahepatic biliary anatomy
treatment

- Kasai
  - Excise scarred bile ducts and gall bladder
  - Portoenterostomy
    - Bile flow into the intestines
    - Roux-en-Y limb of the jejunum
    - Liver biopsy to evaluate the degree of fibrosis
    - Surgical success is increased if done before 8 weeks
  - Intussuscepted antireflux valve using appendix and jejunal limb – minimize postoperative cholangitis
    - Supposed to minimize postoperative cholangitis
      - Data suggests it actually does not impact outcome thus has fallen out of favor

- Complication
  - cholangitis
Outcome

-Usu successful if done before 2 months
-Two thirds of the require liver transplant secondary to progressive liver failure
  -1/3 of pts remain asymptomatic
  -1/3 never have bile flow and require early transplant
  -1/3 initially have good bile flow but subsequently develop cirrhosis
-Independent risk factors for failure
  -Bridging liver fibrosis
  -Post op cholangitic episodes
Without surgery or liver transplant life span – 19 months

Death
- Progressive liver failure
- Bleeding from esophageal varices
- Sepsis

Liver Transplant
A 54 yr male is found to have a type I choledochal cyst incidentally while being worked up for appendicitis. Treatment?

- Observe and do serial CTs Q 6 months
- Observe and do nothing unless symptomatic
- Cholecystectomy
- Cholecystectomy, choledochal cyst excision, hepatoenterostomy
A 54 yr male is found to have a type I choledochal cyst incidentally while being worked up for appendicitis. Treatment?

- Observe and do serial CTs Q 6 months
- Observe and do nothing unless symptomatic
- Cholecystectomy
- **Cholecystectomy, choledochal cyst excision, hepatoenterostomy**
A 3 week old infant is brought to the ER with lethargy and vomiting. The infant had been doing well at home until today, when she became lethargic and was vomiting greenish material. Which of the following would be most accurate in making the diagnosis?

- Abdominal US
- UGI
- CT
- XR
- Barium enema
A 3 week old infant is brought to the ER with lethargy and vomiting. The infant had been doing well at home until today, when she became lethargic and was vomiting greenish material. Which of the following would be most accurate in making the diagnosis?

- Abdominal US
- UGI
- CT
- XR
- Barium enema
Which of the following is included in the LADD’s procedure?

- Cholecystectomy
- Appendectomy
- Both
- neither
Which of the following is included in the LADD’s procedure?

- Cholecystectomy
- Appendectomy
- Both
- neither
The most common type of Choledochal cyst is

- I
- II
- III
- IV
- V
The most common type of Choledochal cyst is

- Type I
- Type II
- Type III
- Type IV
- Type V
Approximately 60% of pts with biliary atresia who undergo a Kasai procedure will eventually need a liver transplant.
TRUE OR FALSE

- Approximately 60% of pts with biliary atresia who undergo a Kasai procedure will eventually need a liver transplant.
References

- Maingot’s Abdominal Operations
- Siegel, M., Jaundice in Infants and Children, Ultrasound clin, 2006;1,431-446
- Schwartz’s Principles of Surgery