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Biliary Stones and Polyps

CASE HISTORY I

- 42 year old male with known gallstones presents with severe epigastric pain x 12 hrs.
- He's had the pain almost every day for last 2 years.
- He has some bloating and indigestion.
- His friend who is a surgeon has advised him to undergo a cholecystectomy.
- Exam and labs are normal.

CASE HISTORY I

- *What is the differential diagnosis?*
- *What would you advise?*
- *What is definition of asymptomatic gallstone?*

DIFFERENTIAL DIAGNOSIS

- Gastroesophageal reflux
- Peptic ulcer disease
- Pancreatitis
- Irritable bowel syndrome
- *Any disorder that can produce abdominal pain*



GALLSTONE DISEASE

- Prevalence (10-20%) is dependent on age, gender, ethnicity, and other factors
- 20 million persons with gallstones in US
- 70-80% are cholesterol stones
- Most stones are silent

RISK FACTORS FOR GALLSTONES

- Age
 - <40 years: 1-3% of men, 5-10% of women
 - >60 years: 15-20% of men, 30-40% of women
- Gender
 - Incidence in pre-menopausal women is x10 men, but decreases to x2 post-menopausal
- Obesity or rapid weight loss (?Increased cholesterol secretion, gallbladder stasis due to fasting)

Major Risk Factors for the Development of Gallstones

- Age
- Female sex
- Genetic
 - Pima Indians and certain other Native Americans
 - Chileans
- Pregnancy
- Obesity
- Rapid weight loss
 - Very low calorie diet
 - Surgical therapy of morbid obesity
- Cirrhosis
- Hemolytic anemias
- Hypertriglyceridemia
- Medications
 - Estrogen and oral contraceptives
 - Clofibrate
 - Ceftriaxone
 - Octreotide
- Terminal ileal resection
- Gallbladder stasis
 - Diabetes mellitus
 - Total parenteral nutrition
 - Postvagotomy
 - Octreotide or somatostatinoma
 - Spinal cord injury
- Reduced physical activity (at least in men)

PROTECTIVE FACTORS

❖ Ascorbic acid (Vitamin C)

- %13, female

❖ Coffee

- %40, 2-3
- Decaffeinated coffee

❖ Vegetable protein

- %20

❖ Poly- and monounsaturated fats

GALLSTONE SYMPTOMS

- The cardinal symptom of gallstones is biliary colic
 - Moderately severe **crescendo** type pain in the right upper quadrant
 - Radiating to the **back and right shoulder**
 - May be accompanied by **nausea**
 - Usually steady and **not colicky** (despite its name)
 - Pain may be brought on after ingestion of **fatty foods**



Characteristics of gallstone pain

Characteristics of gallstone pain

GALLSTONE SYMPTOMS

- The pain of biliary colic usually lasts for **less than four hours**.
- **Fever** and **systemic symptoms** are unusual; pain lasting longer than four hours or associated with systemic symptoms may indicate the development of **acute cholecystitis**.

GALLSTONE SYMPTOMS

- Gallstones are sometimes implicated as the **source** of symptoms in patients with **dyspepsia**.
- However, such an association should be made carefully, since gallstones may silently coexist in patients with dyspepsia, and other causes of dyspepsia are more common.

DIAGNOSIS

- Abdominal ultrasound
 - >95% accuracy, inexpensive, non invasive
 - The best method for diagnosis
- Oral cholecystogram
 - Also very sensitive, but false negative in 25-40%
- CT
 - Expensive, not very sensitive

CASE HISTORY CONTINUE

- Treatment for dyspepsia improves his symptoms
- Despite his improvement, he **still wants** to undergo **cholecystectomy**

‘Just in case’

➤ *What is your advise ?*

➤ *What are the indications for prophylactic surgery?*

GALLSTONE TREATMENT

- *Treat only patients with proven gallstone disease*
- *Treat only patients with typical symptoms*
 - Recurrent pain
 - Cholecystitis
 - Complications of cholecystitis

GALLSTONE TREATMENT

- *Treatment is almost always surgical*
- Prophylactic surgery is not recommended routinely

CONTINUED

- *What are the indications for nonsurgical treatment ?*

NONSURGICAL TREATMENT

- The role for the medical management of gallstone disease has decreased in recent years with the advent of laparoscopic cholecystectomy
- Cholecystectomy remains the preferred method because of its
 - Reduced cost
 - Definitive nature
 - Safety

NONSURGICAL TREATMENT

- Three methods used alone or in combination are available for the nonsurgical management of patients with gallstone disease:
 - Oral bile salt therapy, primarily ursodeoxycholic acid
 - Contact dissolution
 - Extracorporeal shock-wave lithotripsy

NONSURGICAL TREATMENT

- Patients selected for oral bile acid therapy should have the following favorable characteristics:
 - Small stone size (< 1 cm)
 - Mild symptoms
 - Good gallbladder emptying (function)
 - Buoyant stones, suggesting a high cholesterol content
 - Minimal calcification on CT imaging
 - No evidence of acute cholecystitis
 - A patent cystic duct

- The primary aim of therapy should be to achieve complete gallstone dissolution

NONSURGICAL TREATMENT

PATIENT SELECTION

Recommendations

- Patients with **severe medical problems** who are **at high risk for or refuse surgery** and who have mild to moderately symptomatic gallstone disease should be considered for **medical therapy**
- Patients with complicated gallstone disease who cannot undergo surgery are better treated by
 - Percutaneous lithotomy
 - Gallbladder drainage
 - Endoscopic retrograde cholangiopancreatography (ERCP)

CONTINUED

- *What are the indications for prophylactic surgery?*

ROLE OF PROPHYLACTIC CHOLECYSTECTOMY

- There are **no** prospective **trials** of therapy, either surgical or medical, for asymptomatic gallstones.
- However, **decision analysis models** have shown **no benefit of a prophylactic cholecystectomy**.
- Thus, prophylactic cholecystectomy is **not indicated** in most patients with asymptomatic gallstones

ROLE OF PROPHYLACTIC CHOLECYSTECTOMY

- Possible exceptions include
 - Patients who are at increased risk for gallbladder carcinoma
 - Patients who are at increased risk for gallstone complications
 - In whom prophylactic cholecystectomy or incidental cholecystectomy at the time of another abdominal operation can be considered

ROLE OF PROPHYLACTIC CHOLECYSTECTOMY

- Patients at increased risk of complications
 - Diabetes mellitus
 - The magnitude of the risk and the risks and costs of cholecystectomy do not warrant prophylactic cholecystectomy in diabetics with asymptomatic gallstones

ROLE OF PROPHYLACTIC CHOLECYSTECTOMY

- Patients at increased risk for biliary cancer – an increased risk of **cholangiocarcinoma** and **gallbladder carcinoma** has been associated in some ethnic groups (such as native Americans)
 - Choledochal cysts
 - Caroli's disease
 - Anomalous pancreatic ductal drainage (in which the pancreatic duct drains into the common bile duct)
 - Gallbladder adenomas
 - Porcelain gallbladder

A Suggested Algorithm for Managing Gallbladder Polyps



* Symptoms: Biliary type pain, common duct obstruction, cholangitis, or recurrent pancreatitis. Dyspepsia is not an indication for surgery.

ROLE OF PROPHYLACTIC CHOLECYSTECTOMY

- Sickle cell disease
 - Pigment gallstones are common and often asymptomatic in patients with sickle cell disease.
 - Prophylactic cholecystectomy is not recommended, but an incidental cholecystectomy should be considered if abdominal surgery is being performed for other reasons.

ROLE OF PROPHYLACTIC CHOLECYSTECTOMY

- Hereditary spherocytosis
 - Some authorities recommend combined prophylactic splenectomy and cholecystectomy in young asymptomatic patients with gallstones
- Gastric bypass surgery
 - Morbidly obese patients who have undergone gastric bypass surgery, a form of bariatric surgery, have a high incidence of developing gallstones (greater than 30 percent)
 - An incidental cholecystectomy is recommended at the time of surgery.



CASE HISTORY CONTINUE

- After 3 year, he presents to the emergency room with:
 - Right upper quadrant (RUQ) and back pain from last night
 - Nausea and vomiting after eating fried chicken for dinner
- On physical examination
 - He has a temperature of 38 degrees Celsius
 - Moderate RUQ tenderness
 - Mild jaundice
 - Positive Murphy's sign

CASE HISTORY CONTINUE

- Lab tests:
 - White blood cell count (WBC) is 15 K/ml
 - T bili = 2
 - ALKP = 88
 - AST = 34
 - ALT = 66
- Plain radiographs of the abdomen are unremarkable

CASE HISTORY CONTINUE

- What study do you request next?

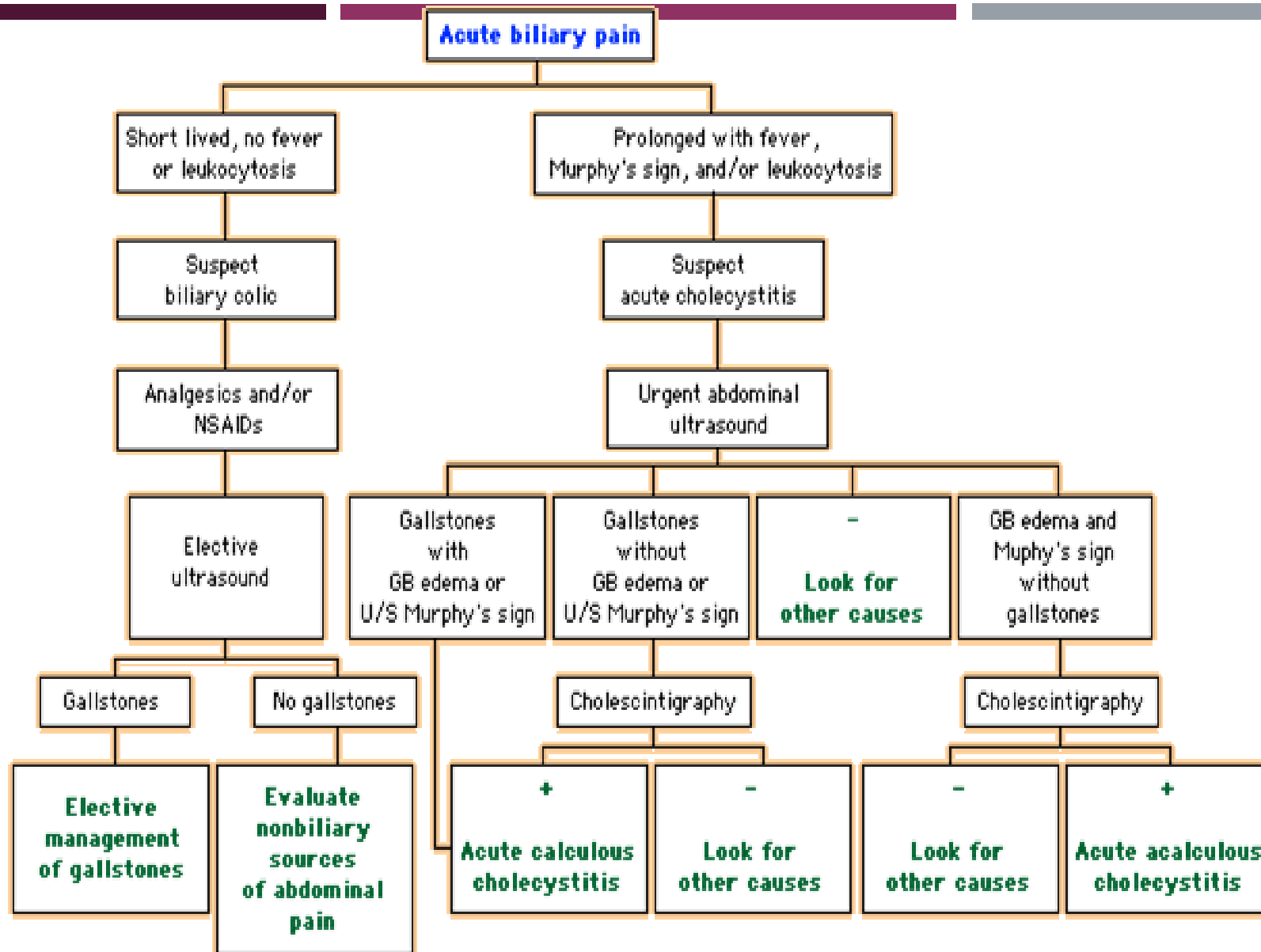
CLICK Endoscopic retrograde cholangiopancreatogram (ERCP)

CLICK Cholescintigraphy

CLICK Abdominal computed tomogram

CLICK Abdominal ultrasound

CLICK Abdominal MRI



ULTRASOUND



ULTRASOUND



ULTRASOUND



ULTRASOUND



ABDOMINAL ULTRASOUND

- Abdominal ultrasound demonstrates a gallstone lodged within the cystic duct
- There is thickening of the gallbladder wall, measuring 5.3 mm

ABDOMINAL ULTRASOUND

- A small amount of pericholecystic fluid is present
- The common bile duct is not dilated
- These findings are consistent with acute cholecystitis

ACUTE CHOLECYSTITIS

- Acute cholecystitis occurs in approximately **one third** of patients with gallstones and is caused by **obstruction** of the **cystic duct** by an impacted calculus.
- This results in gallbladder wall **inflammation** which may lead to **infection** and **necrosis**.

ACUTE CHOLECYSTITIS

- 90% of acute cholecystitis cases due to **gallstones**
- **Aging** is the most significant factor $\Rightarrow$ higher incidence of acute cholecystitis
- Acute cholecystitis is the **initial** presentation of symptomatic gallstones in **15% - 20%** of patients

ACUTE CHOLECYSTITIS

■ Symptoms:

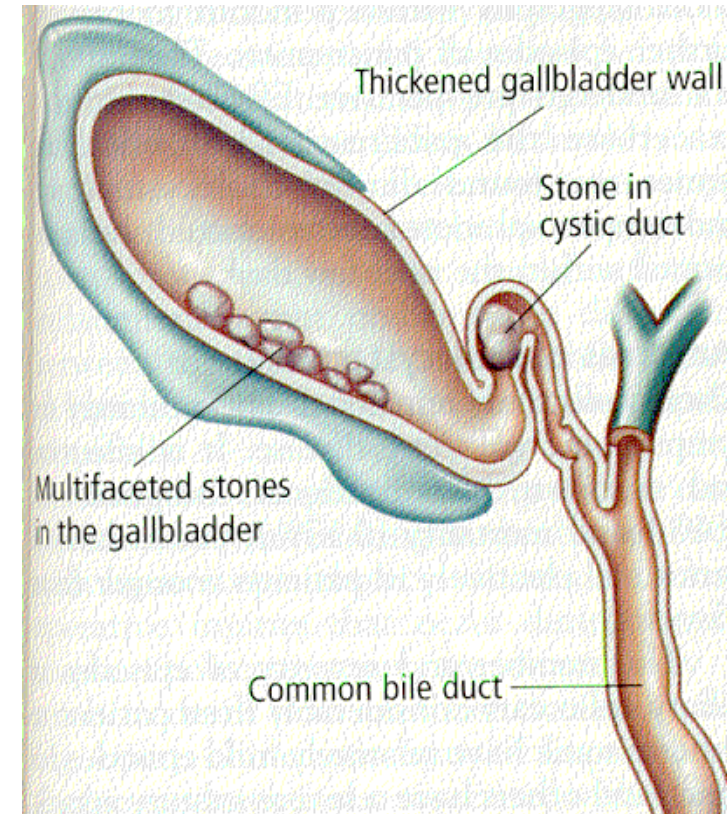
- Abdominal pain
 - Most commonly in the right upper quadrant or epigastrium
 - May radiate to the right shoulder or back
 - Lasts > 6-8 hrs
- Nausea, vomiting, and anorexia
- Usually fatty food ingestion - 1 hr before pain

ACUTE CHOLECYSTITIS

- Signs:
 - Tachycardia
 - *Low grade* fever
 - *Mild* leukocytosis
 - Biliary colic
 - Murphy's sign
 - Palpable gallbladder
 - Usually does not cause jaundice

ACUTE CHOLECYSTITIS

- Distention and inflammation of the gallbladder
- Persistent obstruction of cystic duct $\Rightarrow$ chemical irritants in the bile
 - Lysolecithin
 - Prostaglandins



CHOLECYSTITIS

- Diagnosis:
 - History and physical examination
 - Lab tests
 - Ultrasound
 - Obvious in 30%
 - Questionable in 60% - perform radionuclide scanning

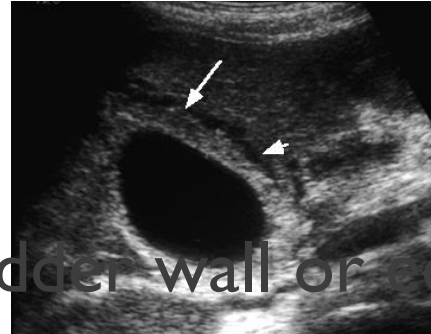
DIAGNOSIS

- Sonography is a sensitive %88 (85-95%) and specific % 80 (64-100%) modality for diagnosis of acute cholecystitis.

DIAGNOSIS

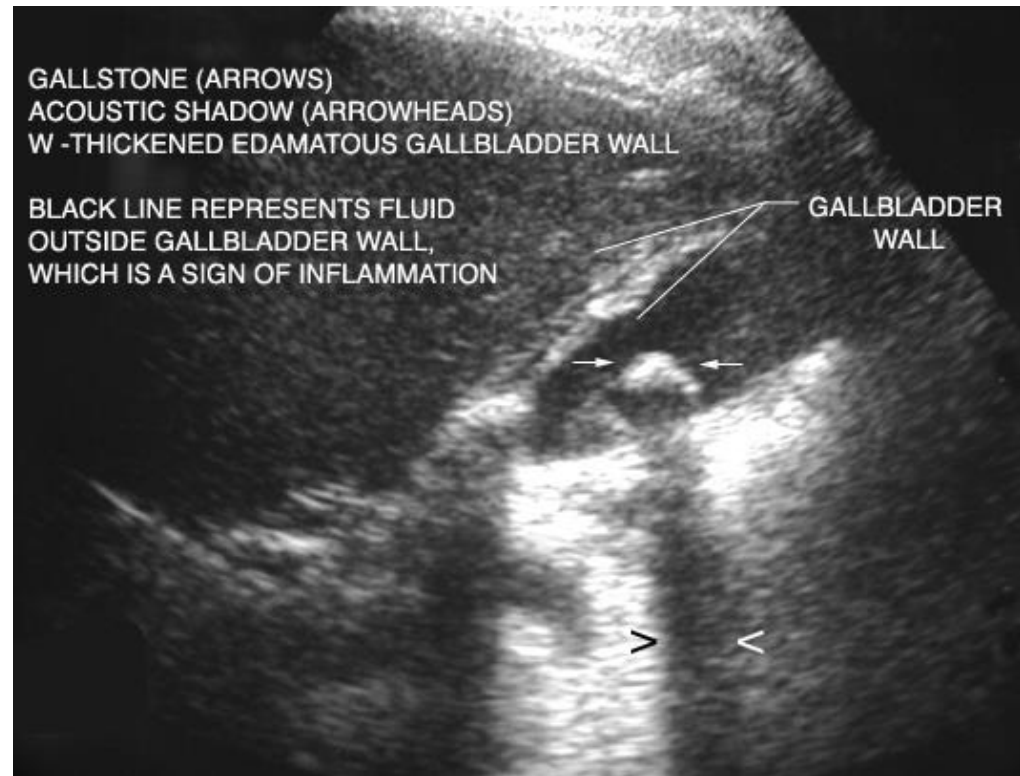
- The diagnostic criteria include cholelithiasis
 - **Sonographic Murphy's sign** with maximal tenderness over the sonographically localized gallbladder
 - (More accurate than hand palpation)
 - **Gallbladder wall**
 - Thickening (greater than 4 to 5 mm) (3mm) or
 - Edema (double wall sign)
 - **Pericholecystic fluid**
 - **Gallbladder dilatation**

- Thickened gallbladder wall or edema
- Pericholecystic fluid
- Sonographic Murphy's sign



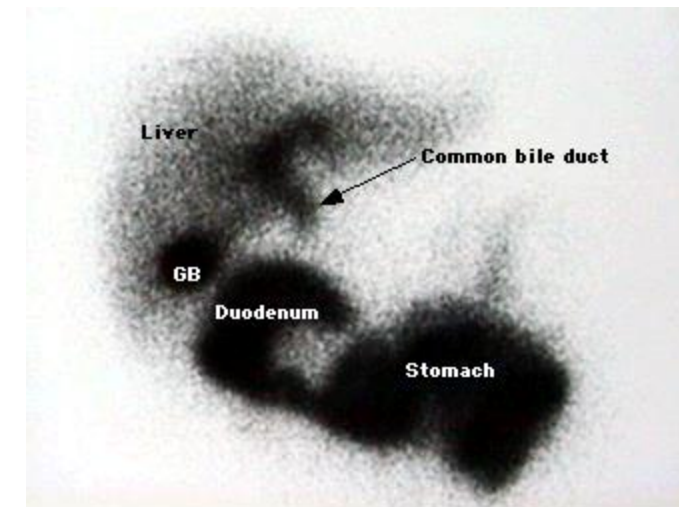
Acute cholecystitis Ultrasound of the right upper quadrant in a patient with acute cholecystitis reveals marked thickening of the gallbladder wall (arrow) with pericholecystic fluid surrounding the distended gallbladder (arrowhead). Courtesy of Jonathan Kruskal, MD.

ACUTE CHOLECYSTITIS



CHOLESCINTIGRAPHY

- Cholescintigraphy (generically referred to as a **HIDA** scan) is indicated if the diagnosis remains uncertain following ultrasonography
- Technetium labeled **hepatic iminodiacetic acid** (HIDA)
- Useful for demonstrating **patency** of the common bile duct and ampulla
- 30 to 60 minutes
- Positive if the gallbladder does **not visualize**
- Sensitivity 97 and specificity 90



Normal HIDA scan showing the visualized gallbladder, common duct and filling of the duodenum. Courtesy of Salam F Zakko, MD, FACP.

CHOLESCINTIGRAPHY

■ False positive test

- **Cystic duct obstruction** with a stone or tumor in the absence of acute cholecystitis
- **Severe liver disease**, which may lead to abnormal uptake and excretion of the tracer
- **Fasting** patients receiving total parenteral nutrition, in whom the gallbladder is already maximally full due to prolonged lack of stimulation

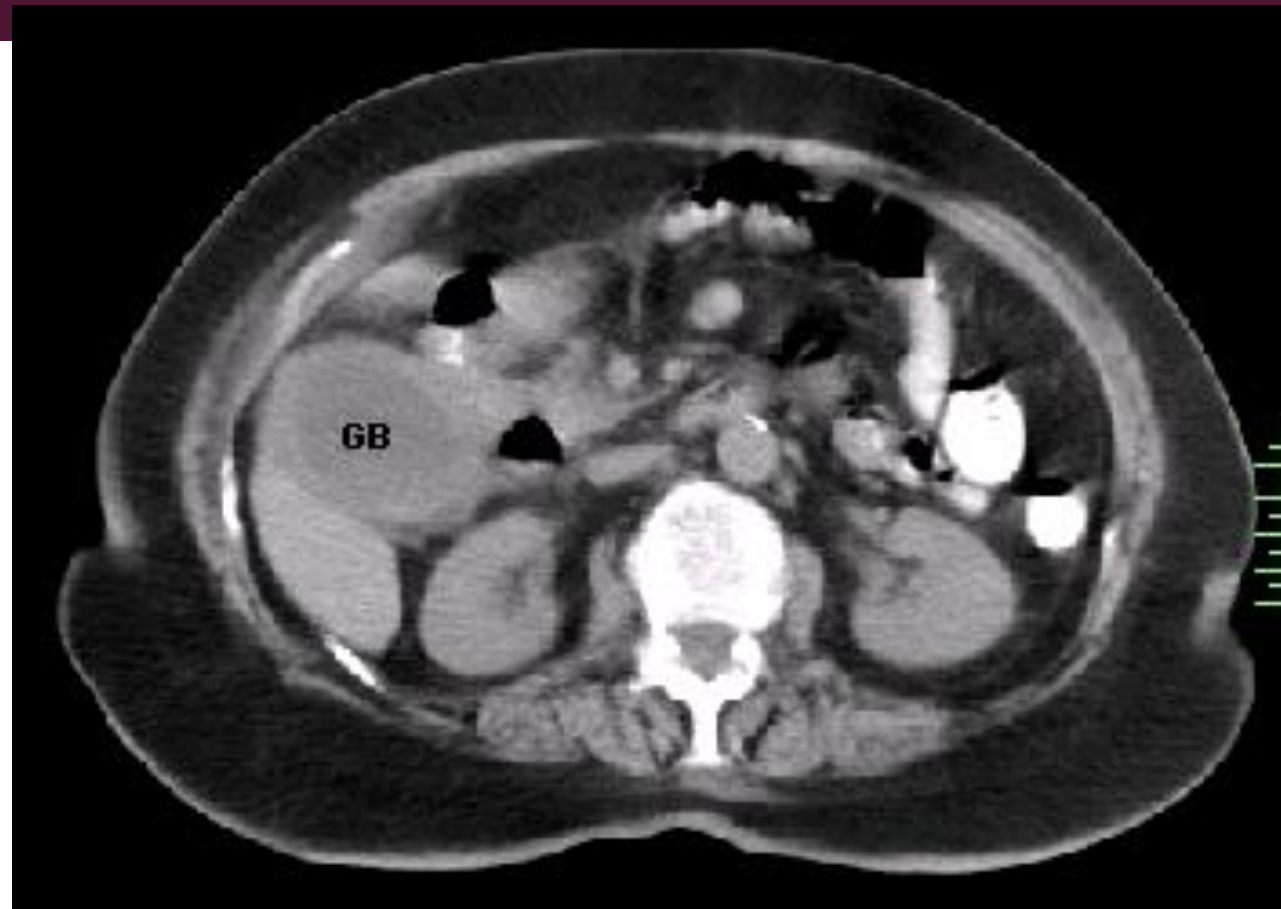
CHOLESCINTIGRAPHY

- False positive test
 - **Biliary sphincterotomy**, which may result in low resistance to bile flow, leading to preferential excretion of the tracer into the duodenum without filling of the gallbladder
 - **Hyperbilirubinemia**, which may be associated with impaired hepatic clearance of iminodiacetic acid compounds
 - Newer agents commonly used in cholescintigraphy (diisopropyl and m-bromothymethyl iminodiacetic acid) have generally overcome this limitation

CHOLESCINTIGRAPHY

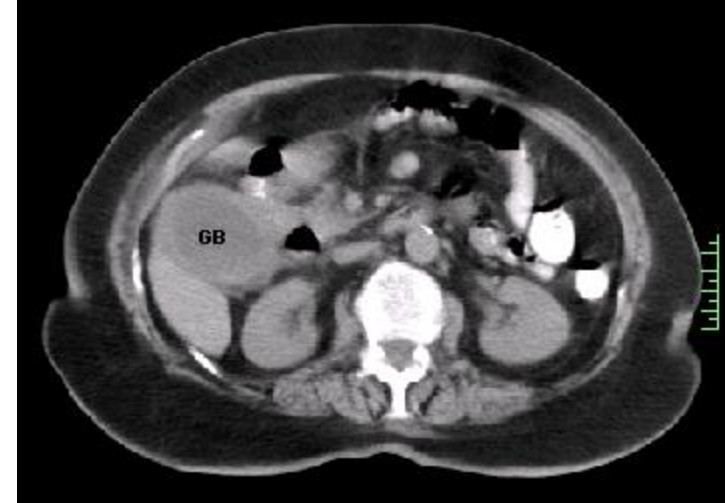
- False negative results are uncommon
 - Incomplete cystic duct obstruction

CT SCAN



CT SCAN

- CT scan
 - **Usually unnecessary**, although it can easily demonstrate gallbladder wall edema associated with acute cholecystitis
 - Other CT findings include
 - Pericholecystic stranding and fluid
 - High-attenuation bile
 - **Useful for complications**
 - Emphysematous cholecystitis
 - Gallbladder perforation



A CT scan showing a dilated gallbladder with a markedly thickened wall due to edema of acute cholecystitis. Courtesy of Salam F Zakko, MD, FACP.

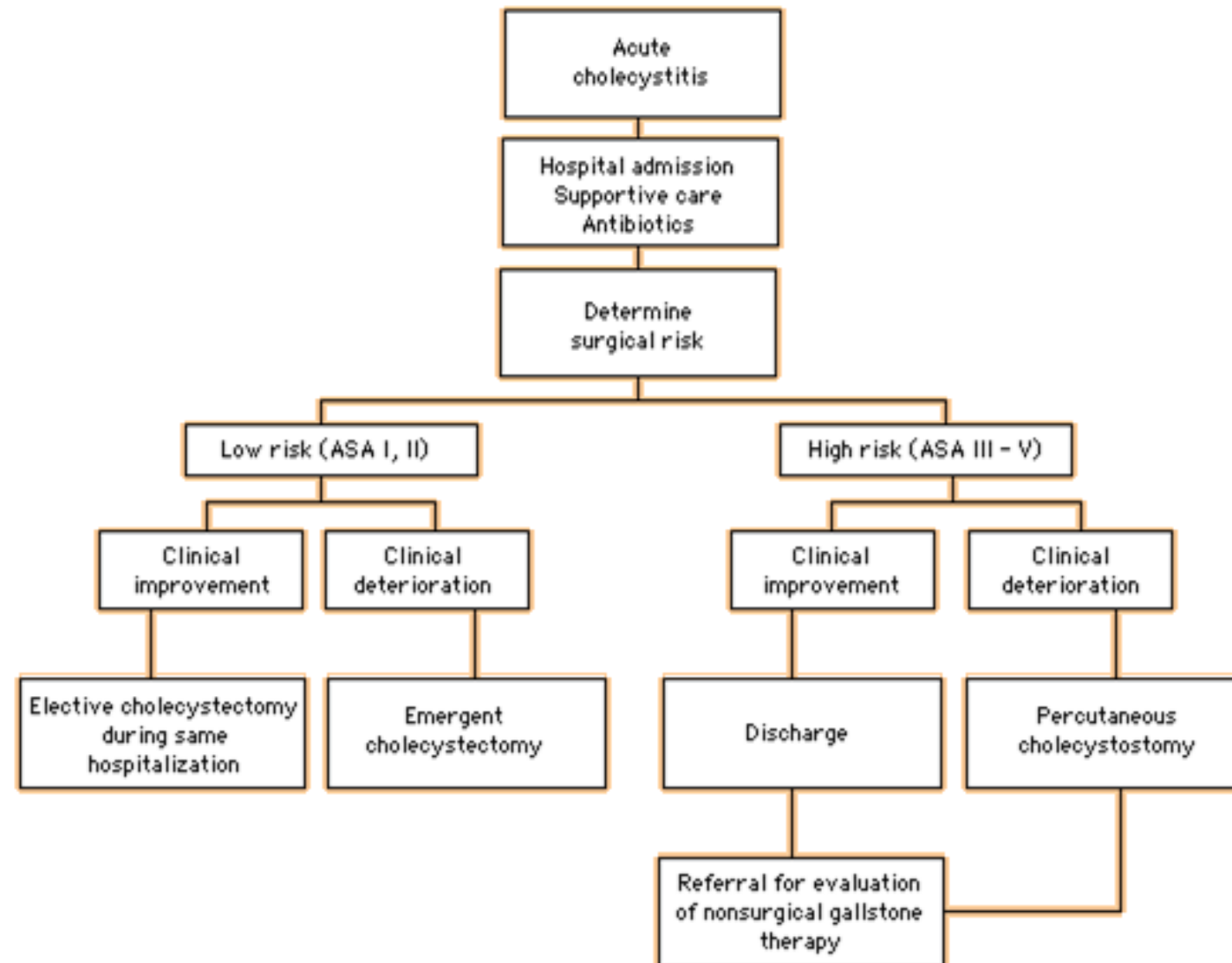
OTHERS

- Magnetic resonance cholangiography
 - Superior to ultrasound for **detecting stones in the cystic duct** (sensitivity 100 versus 14 percent)
 - But less sensitive than ultrasound for detecting **gallbladder wall thickening** (sensitivity 69 versus 96 percent)
- Oral cholecystography
 - No role
 - Cannot show gallbladder wall edema
 - Requires days to complete

QUESTION

- What are you doing for management of this patient ?

Treatment of Acute Cholecystitis*



MANAGEMENT OF ACUTE CHOLECYSTITIS

- Admitted to the hospital & supportive care with
 - IVFs
 - Bowel rest
 - Narcotic analgesia may be required
 - NSAIDs
 - May improve course of acute cholecystitis
 - Prevent the progression of biliary colic to acute cholecystitis ?
 - Injection of ketorolac (30 to 60 mg adjusted for age and renal function), which may also favorably alter the natural history of the disease

MANAGEMENT OF ACUTE CHOLECYSTITIS

- Almost **half** of patients have **positive** bile cultures
 - E. Coli is most common organism
 - Escherichia coli (41 percent)
 - Enterococcus (12 percent)
 - Klebsiella (11 percent)
 - Enterobacter (9 percent)

QUESTION

- What and when antibiotic are your choice?

MANAGEMENT OF ACUTE CHOLECYSTITIS

- No evidence exists showing a definite benefit with use of antibiotics
 - Although clear evidence of benefit is lacking, most patients who are hospitalized for an episode of acute cholecystitis are **given antibiotics**
 - We recommend the combination of **ampicillin** (2 gm IV every 4 h) and **gentamicin** (dosed according to weight and renal function) as empiric treatment
 - **3rd generation cephalosporin**
 - Cephalosporins do not treat **Enterococcus**

MANAGEMENT CONT.

- SURGERY is the only definitive treatment

MANAGEMENT CONT.

- ☀ 1st open cholecystectomy: 1886 by Justus Ohage
- ☀ 1st half of 20th century: supportive care $\Rightarrow$ delayed open cholecystectomy
- ☀ In 1970's – mid-1980's: open cholecystectomy early in the treatment course
- ☀ “Golden 72 hours” rule

MANAGEMENT OF ACUTE CHOLECYSTITIS

- Studies in early 1980's ⇒ **early surgery was better** than delayed surgery (using standard open approach)
- Laparoscopic surgery developed in late 1980's
- Comparison of laparoscopic cholecystectomy vs open cholecystectomy on

Major Complications of Laparoscopic Cholecystectomy in 8856 Cases^{*}

Complication	Frequency, percent
Major bleeding	1.38
Wound infection	0.6
Bile leak	0.4
Biliary injury	0.2
Bowel injury	0.16
TOTAL	2.58

^{*}Data from Strasberg, SM, Hertl, M, Soper, NJ. J Am Coll Surg 1995; 180:101.

TIMING OF SURGERY

- Early surgery = Within **72 hours** of admission or onset of symptoms
- Delayed surgery = Supportive care only followed by discharge and readmission in **6-12 weeks** for surgery

- Based on patient's **overall risk of surgery**
- American Society of Anesthesiologists (ASA) Scale is a guide for decisions on surgery

**American Society of Anesthesiologists
Physical Status Scale***

Class 1:	A normally healthy individual
Class 2:	A patient with mild systemic disease
Class 3:	A patient with severe systemic disease that is not incapacitating
Class 4:	A patient with incapacitating systemic disease that is a constant threat to life
Class 5:	A moribund patient who is not expected to survive 24 hours with or without operation

*Adapted from Feigal, DW, Med Clin North Am 1979; 63:1131.

COMPLICATIONS OF CHOLECYSTITIS

- Suspect when symptoms **don't resolve in 2-3 days** or if symptoms are severe or atypical
 - **Empyema** (pus filled gallbladder)
 - **Emphysematous cholecystitis** (infection by gas forming organisms; possibly in diabetics)
 - **Perforation**
- Treat with antibiotics and surgery



CASE HISTORY CONT.

- After 2 year from cholecystectomy, he presents with a 2-week history of intermittent right upper quadrant pain and a 2-day history of malaise, nausea and vomiting.
- Physical examination reveals a slightly jaundiced patient with tachycardia and mild RUQ tenderness without peritoneal signs.

CASE HISTORY CONT.

- Laboratory evaluation is remarkable for a bilirubin of 4.8 and slightly elevated alkaline phosphatase.
- Supine and upright plain radiographs of the abdomen are unremarkable.

QUESTION

- *What is the differential diagnosis?*

QUESTION

- What study do you request next?
 - Cholangiogram
 - Abdominal ultrasound
 - MR cholangiogram
 - Abdominal computed tomogram
 - Hepatobiliary (Tc-99m-IDA) scintigram

ULTRASOUND



ULTRASOUND



ABDOMINAL ULTRASOUND

- Abdominal ultrasound scans show a mildly dilated common bile duct measuring 8 mm in diameter
- An echogenic calculus with acoustic shadowing is visible in the distal portion of the common bile duct
- There is no intrahepatic biliary dilatation.
- The gallbladder absent

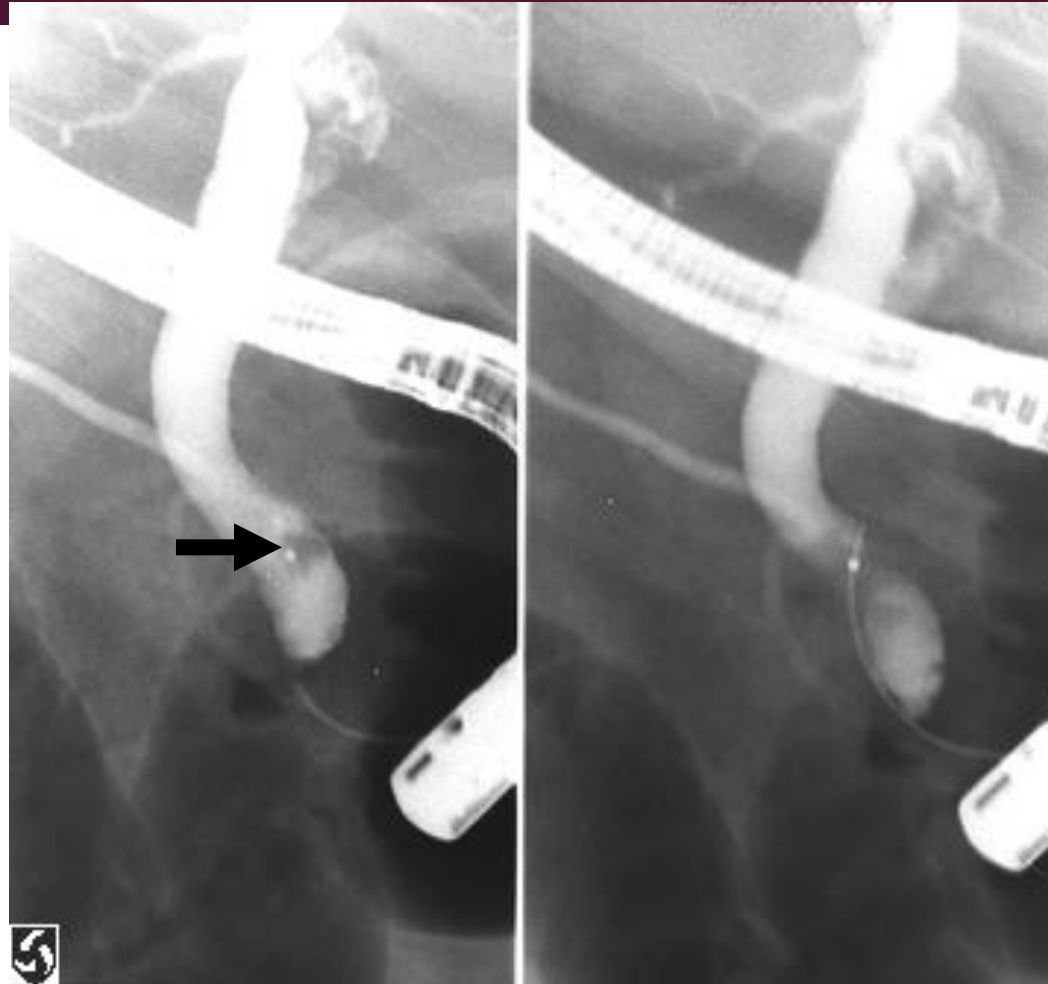
ABDOMINAL ULTRASOUND

- These findings are consistent with **choledocholithiasis** resulting in early or incomplete biliary obstruction.

QUESTION

- Which study to you request next?
 - MR cholangiogram
 - Abdominal CT
 - Endoscopic retrograde cholangiopancreatography with stone extraction
 - Oral cholangiogram
 - Percutaneous cholangiogram

ENDOSCOPIC RETROGRADE CHOLANGIOPANCREATOGRAPHY WITH STONE EXTRACTION



CASE HISTORY CONT.

- Patient underwent endoscopic retrograde **stone extraction** with **sphincterotomy**.
- Injection of the common bile duct during the procedure showed a filling defect (arrow) corresponding to the calculus in the distal common bile duct.

CHOLEDOCHOLITHIASIS

- Choledocholithiasis results from a gallstone that enters the common bile duct from the cystic duct or by erosion.
- Primary common duct stone arising in the intrahepatic or extrahepatic biliary tree is unusual.
- Choledocholithiasis in patients who have undergone cholecystectomy usually represents retained stones that were not identified at the time of surgery.

CHOLEDOCHOLITHIASIS

- About 75% of CBD stones can be delineated by ultrasound although detection will be limited in absence of bile duct dilatation.
- When ultrasonographic evaluation of distal common bile duct is limited by gas or particulate material in the adjacent duodenum, a CT scan or a cholangiogram can aid in establishing the definitive diagnosis.

ALGORITHM FOR CHOLEDOCHOLITHIASIS

- Supine and upright abdominal plain radiographs
 - to exclude bowel obstruction
- Abdominal ultrasound
 - to evaluate the gallbladder and the biliary tree
- Abdominal CT or ERCP
 - when choledocholithiasis is suspected and the distal common bile duct is not visible on ultrasound

BILE DUCT STONES

- **Secondary** bile duct stones
 - **Migrated** from the gallbladder
- **Primary** bile duct stones and intrahepatic stones
 - Common in **Southeast Asia**
 - Often associated with **chronic biliary infections**
 - Often a chronic **recurrent disease**

SPECTRUM OF BILE DUCT STONE CHOLEDOCHOLITHIASIS

- Biliary **pain** only: transient **obstruction**
- Obstructive **jaundice**: continued **obstruction**
 - Diagnosis is suggested by **typical biliary pain + jaundice with bilirubin < 10**
 - Ultrasound is 30% sensitive for showing bile duct stone

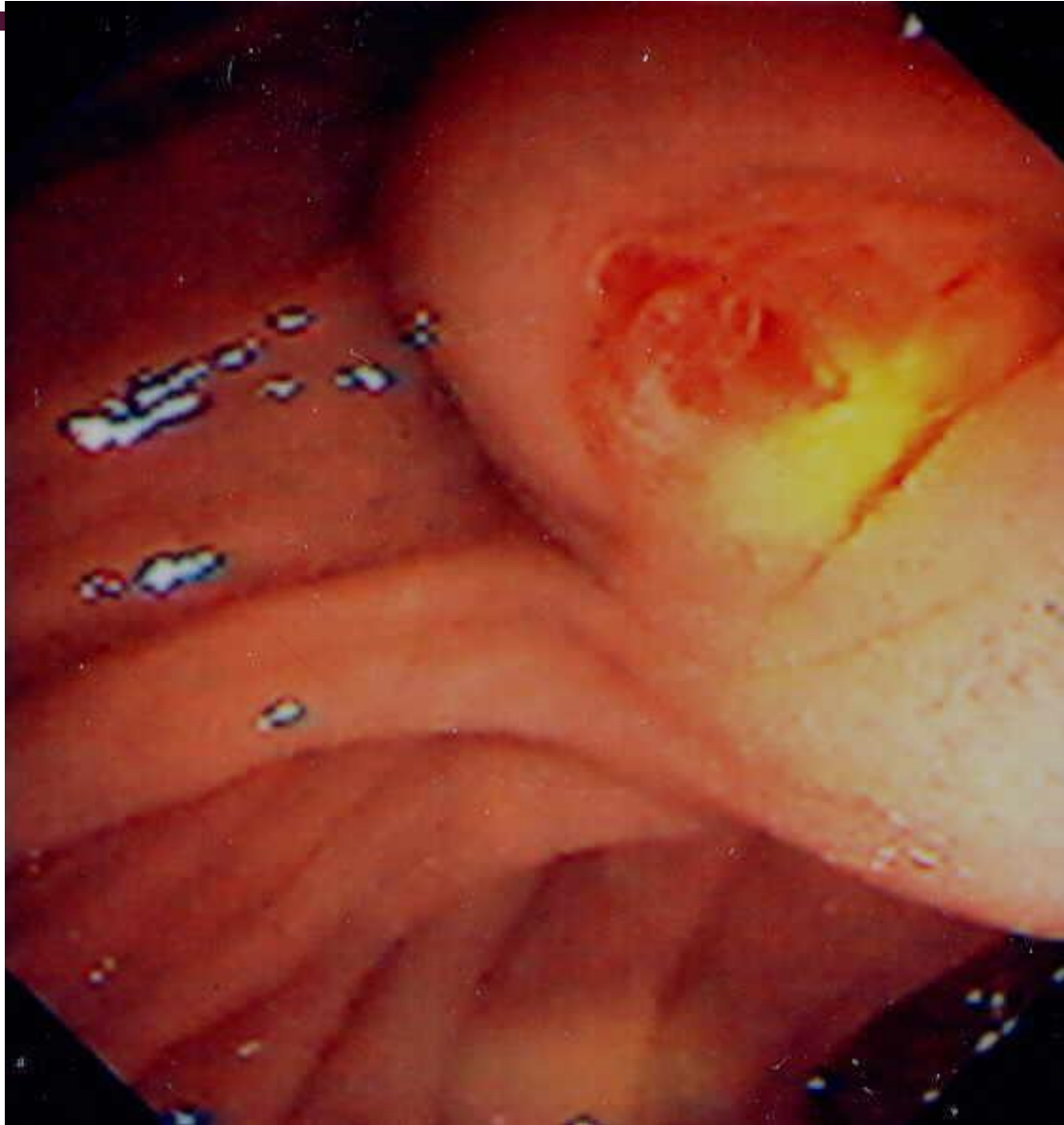
SPECTRUM OF BILE DUCT STONE CHOLEDOCHOLITHIASIS

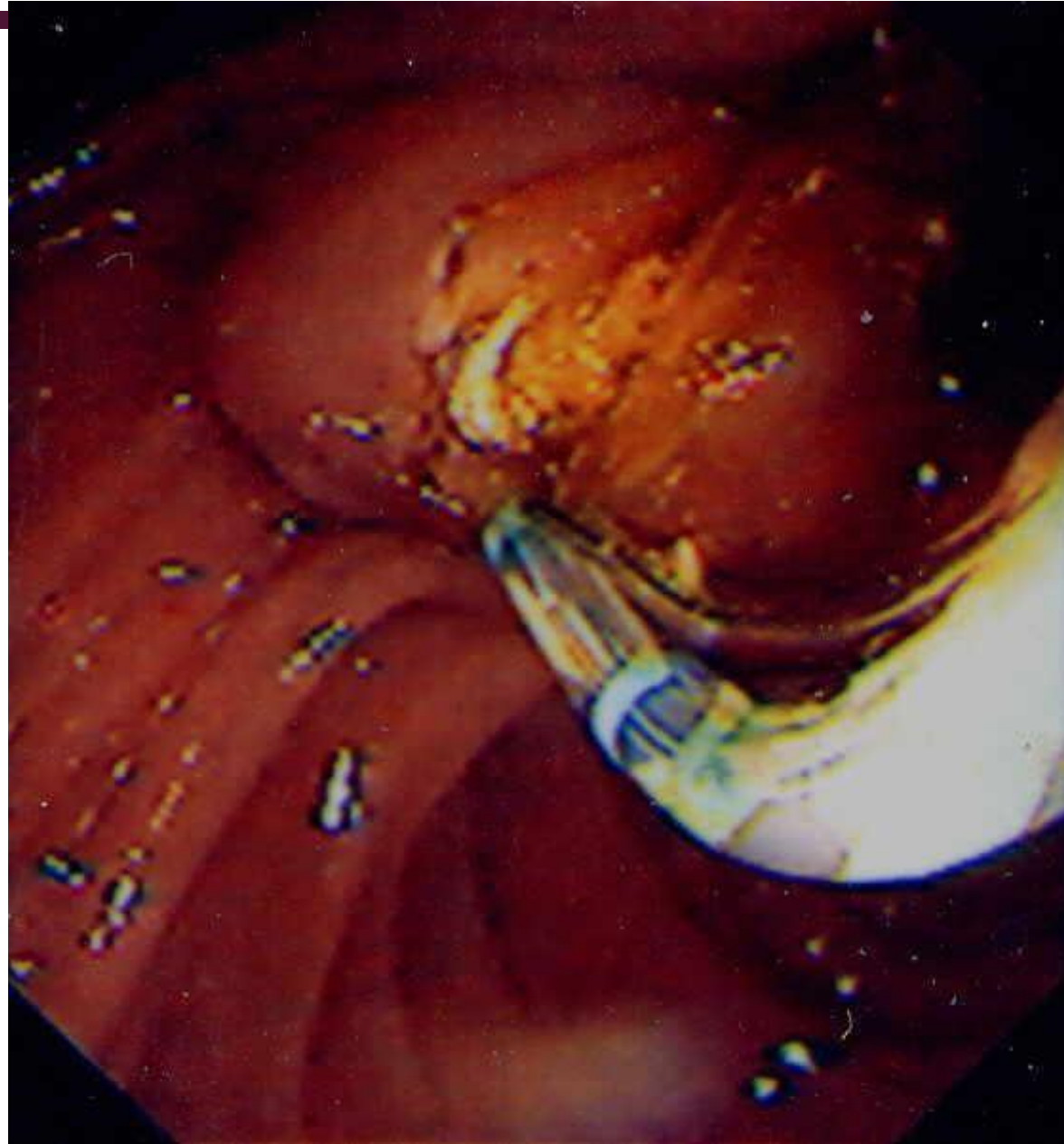
- Cholangitis: obstruction+infection
 - Charcot's triad of pain, fever, jaundice

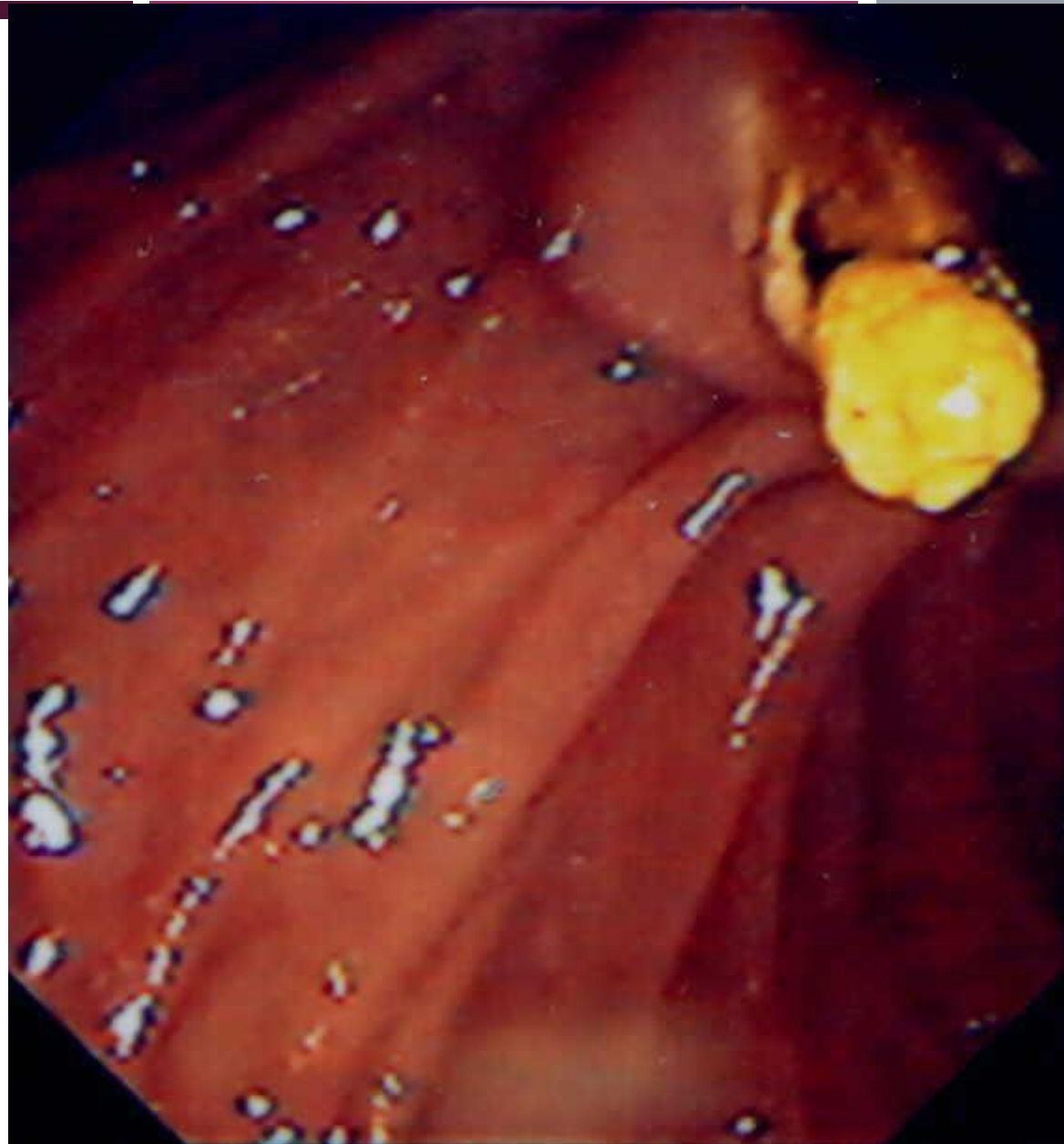
TREATMENT OF CHOLEDOCHOLITHIASIS

- Treatment is *almost always* indicated
- Supportive therapy for pain, jaundice, or cholangitis
- **Treatment is usually endoscopic** (as opposed to gallbladder stones); alternatives of surgery
- **Cholecystectomy** in patients with intact gallbladder

- فیلم
- و سنگ ERCP











CASE HISTORY2

- A 37-year-old man presents with 3 weeks of RUQ pain (colic type), fevers and weight loss.
- Examination reveals marked tenderness in the RUQ without a palpable mass.
- He is not jaundiced and there are no stigmata of chronic liver disease.

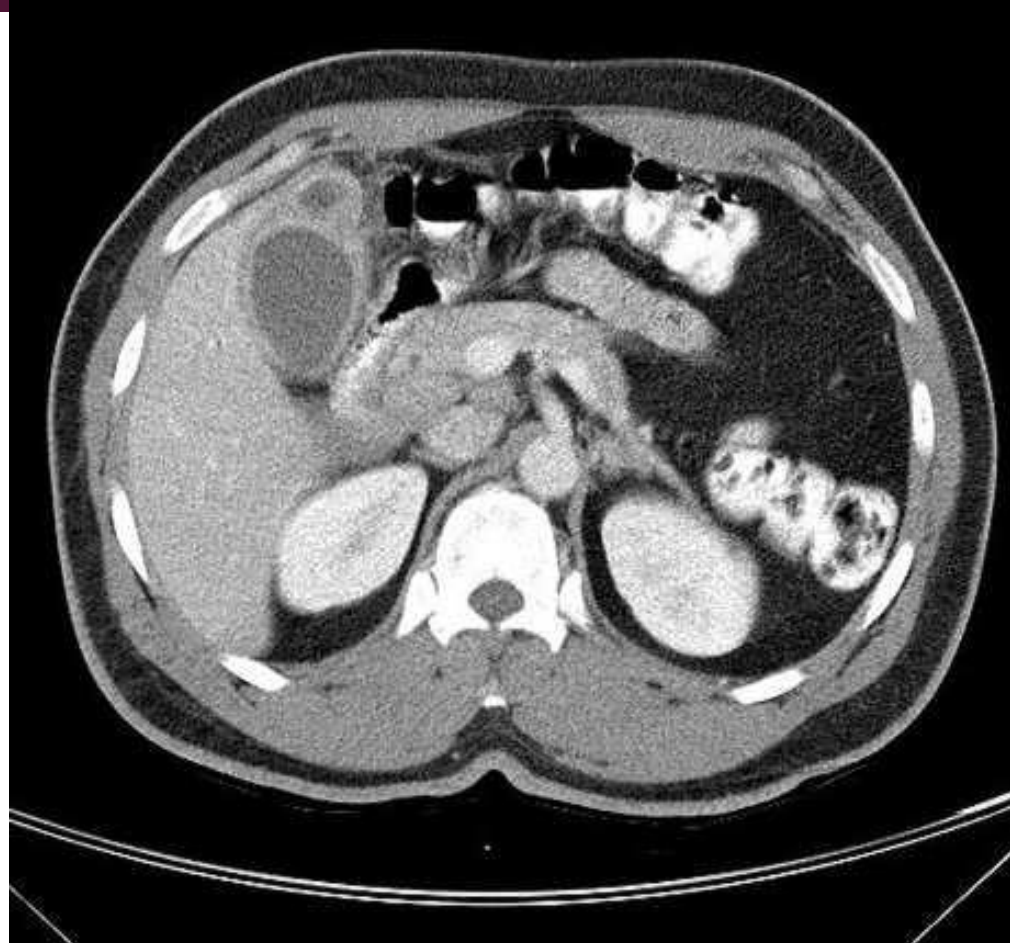
CASE HISTORY2

- An abdominal ultrasound suggests **dilatation** of the **extrahepatic ducts** to **17mm**.
- The gallbladder wall is **thickened** and contains **some sludge** but **no stones**.

CASE HISTORY2

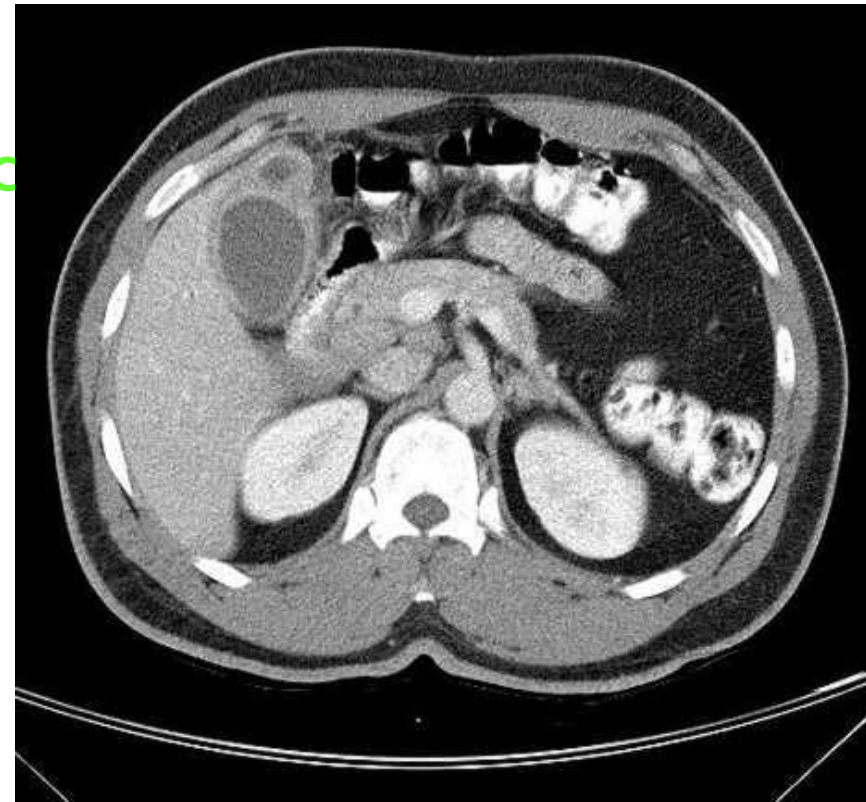
- Liver function tests are abnormal
 - AST = 191 (17-59)
 - Alt = 411 (21-72)
 - Alp = 1025 (38-126)
 - Bili = 0.7 (0.2-1.3)
 - Ca 19.9 = was 33 IU (0-37)
- CT scan and ERCP are performed.

CT SCAN



CT SCAN

- CT Scan confirms **dilated and thickened extrahepatic ducts** without significant intrahepatic dilatation.
- There is also **gallbladder wall thickening**, but no mass



ERCP



ERCP

- Cholangiography showing a **long stricture of the CBD** and **cystic dilatation** of the common hepatic and distal intrahepatic ducts.
- There is also **irregularity** and **occlusion of the cystic duct** and a choledochoduodenal fistula.



ERCP

- A nasobiliary catheter is placed for temporary drainage whilst surgical excision or bypass of the cyst is considered.
- Bile aspirates grew **E.coli and Klebsiella.**



QUESTION

- What is your diagnosis ?

DIAGNOSIS

- ❑ Probable **Cholangiocarcinoma** or Gallbladder carcinoma arising in **choledochal cyst** (Type I-C) with **cholangitis**.

DISCUSSION

- Choledochal cysts were **first** described by Vatero and Ezler in **1723**
- However, Douglas published the first detailed case in **1852**
- **Todani** et al published a classification scheme with **five types** and several subtypes

BILIARY CYSTS

EPIDEMIOLOGY

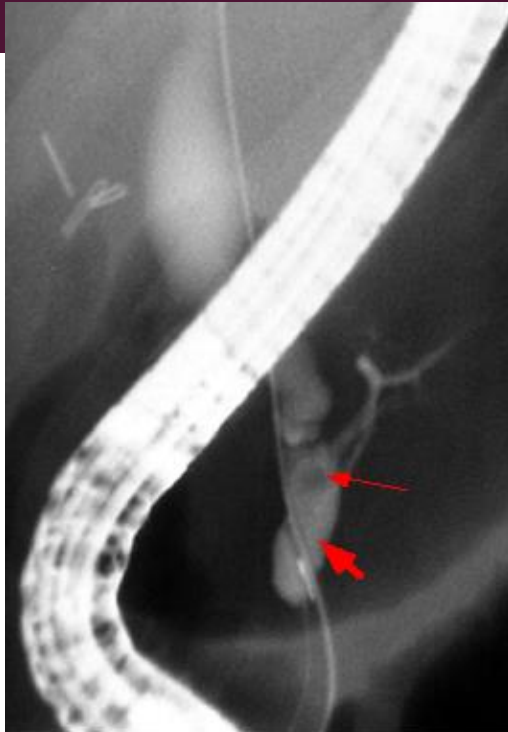
- The **incidence** of biliary cysts has been estimated to be **1:100,000** to **150,000** with even wider ranges reported
- The incidence is **higher** in some **Asian** countries; more than **half** of the reported cases have occurred in **Japan**
- The **female to male** ratio is about **3:1**
- In the past, the majority of cases were reported in children, although more recent series **report equal numbers in adults and children**
- Between **40% and 60%** of patients were diagnosed **before 10** years of age, **52% to 76% before 20** years of age, and 83% to 90% before 30 years of age

PATHOLOGY

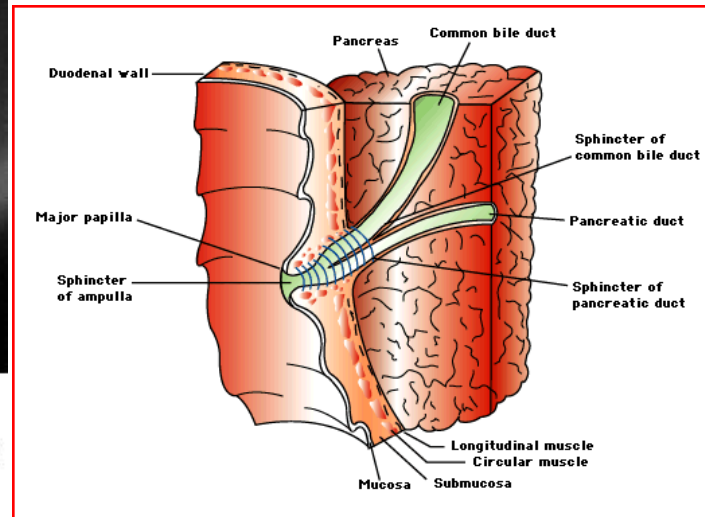
- Pathologic features of choledochal cysts are variable
 - Ranging from normal bile duct mucosa to carcinoma
- Malignancy, when present, is most commonly found in the posterior cyst wall

ABNORMAL PANCREATICOBILIARY JUNCTION

- An abnormal pancreaticobiliary junction (APBJ) is present in about 70 percent of patients with biliary cysts and may be a significant risk factor for the development of malignancy in the biliary cyst
- APBJ is characterized by a
 - long common channel (usually over 2 cm in length)
 - Ampulla of Vater is diminutive or flat in patients with APBJ



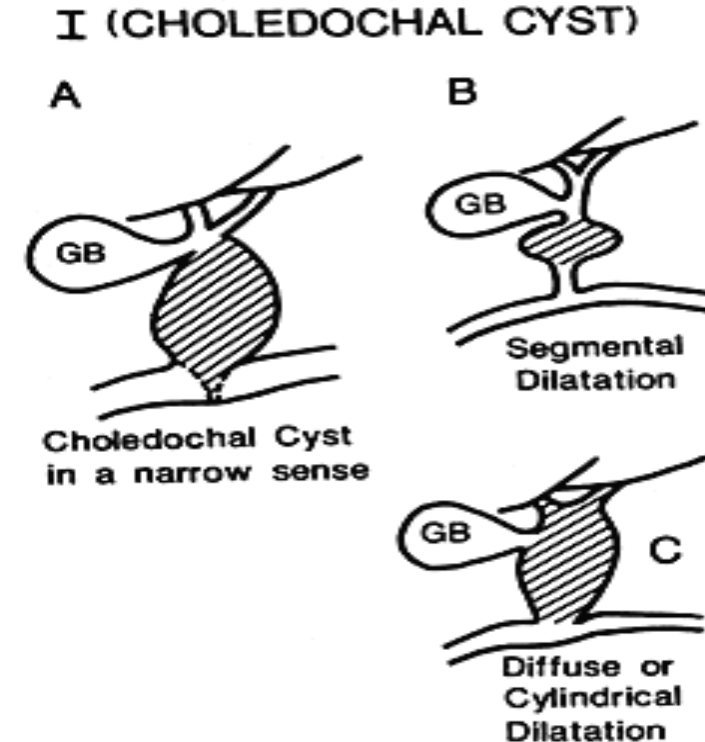
Anomalous pancreaticobiliary junction ERCP demonstrating an anomalous pancreaticobiliary junction in a child with recurrent abdominal pain and pancreatitis. Note the long, dilated common channel (thick arrow) containing a stone (thin arrow). The patient also has pancreas divisum. Courtesy of Mark Topazian, MD.



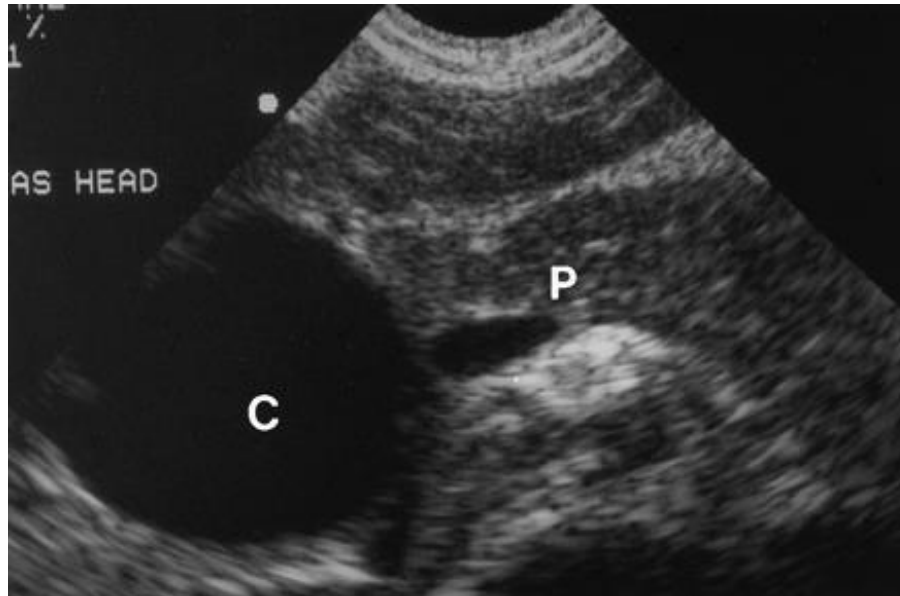
Type I biliary cyst Image obtained after ERCP showing a Type I biliary cyst associated with an anomalous pancreaticobiliary junction. Courtesy of Dr. Morton Burrell.

A classification scheme for cysts of the extrahepatic bile ducts (choledochoal cysts) was proposed initially in 1959

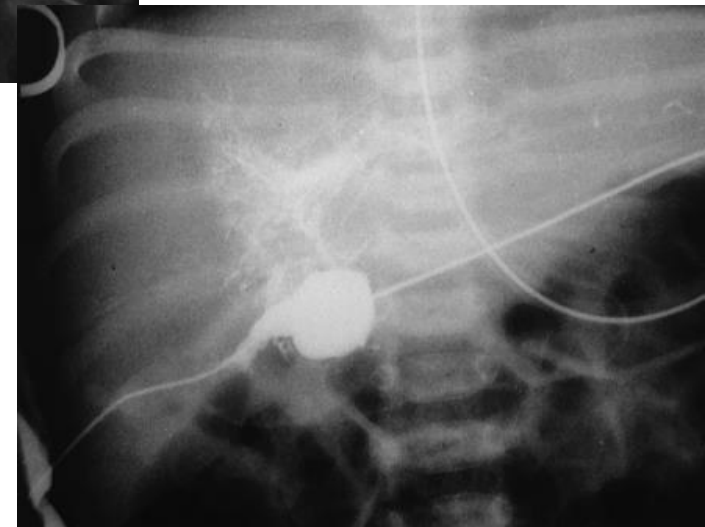
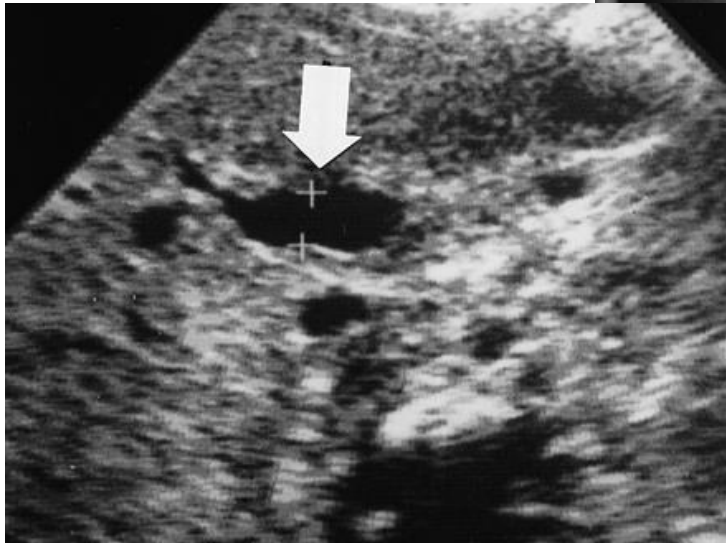
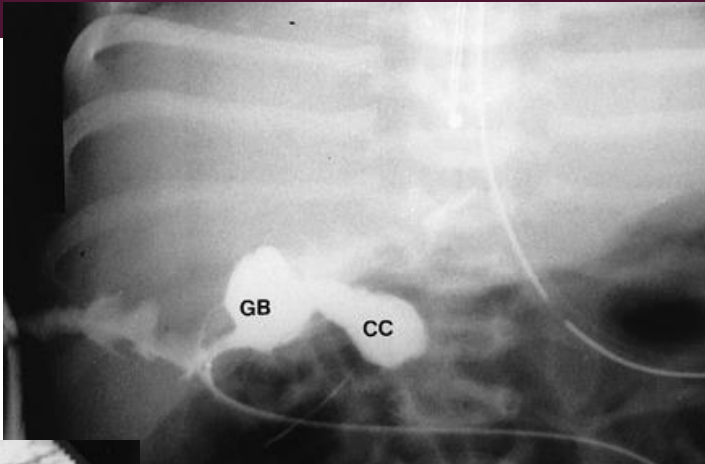
- **Type I** is a cystic dilatation of the common bile duct (CBD) and is the **most common**, comprising **50 to 85** percent of all biliary cysts



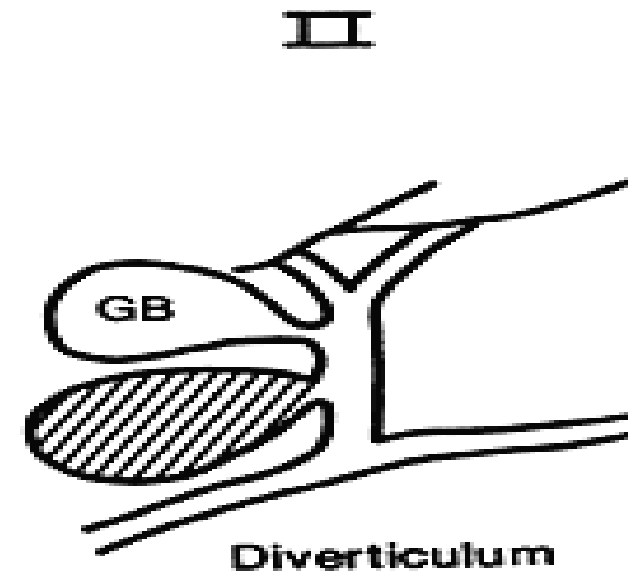
CHOLEDOCHAL CYST (TYPE IA)



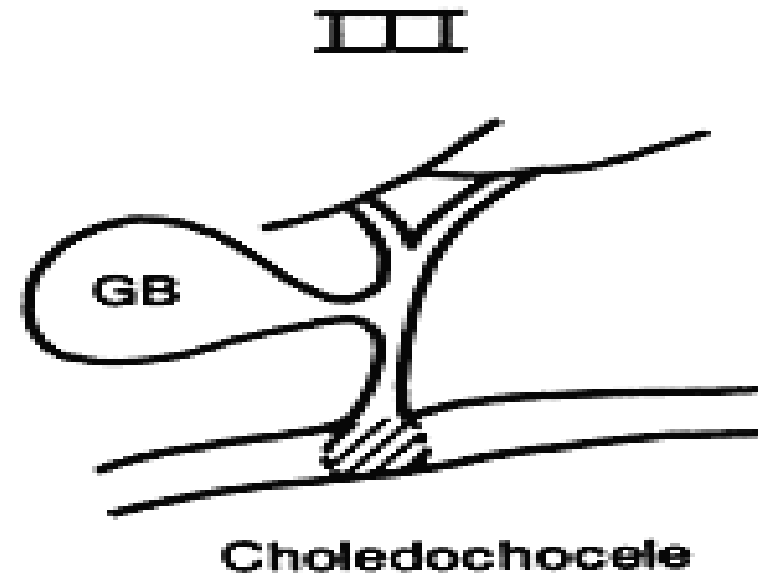
CHOLEDOCHAL CYST (TYPE IC)



- Type II, the rarest biliary cyst, is a true **diverticulum** of the **extrahepatic** bile duct located proximal to the duodenum.



- Type III is a cystic dilatation limited to the **intraduodenal portion** of the distal common bile duct, also known as a choledochoceles.



TYPE III



FIGURE 100-11. Choledochocyst—type III. A: Cholangiography reveals a club-shaped enlargement of the distal common bile duct bulging into the duodenum. B: Upper gastrointestinal series reveals a smoothly rounded filling defect in the second portion of the duodenum.

CLASSIFICATION

- They are subdivided into
 - Type IIIA (in which the bile duct and pancreatic duct enter the choledochoceles, which then drains into the duodenum at a **separate orifice**)
 - Type IIIB (a **diverticulum** of the intraduodenal bile duct or common channel)

- This classification was extended in 1977 to include intrahepatic cystic dilation
- Type IV includes cases of multiple cysts, including
 - Type IVA, in which there are both intrahepatic and extrahepatic cystic dilations

- Type IVB, in which the intrahepatic involvement

- Type IVA is the second most common type of biliary cyst

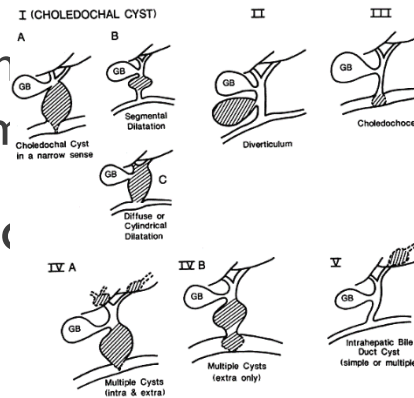
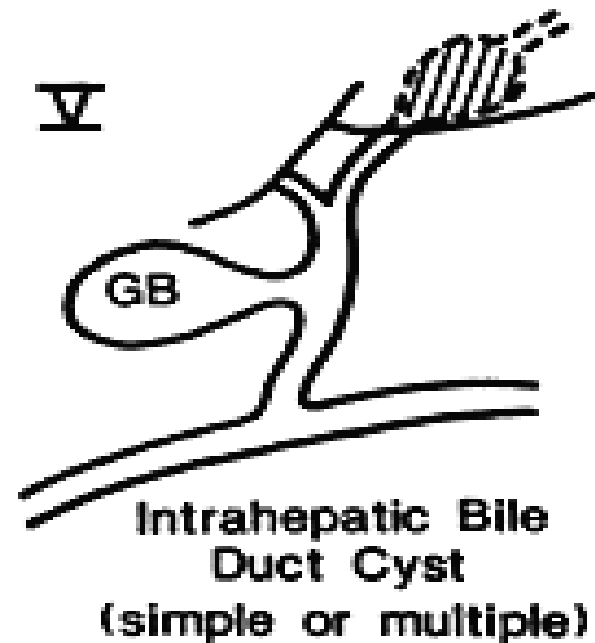
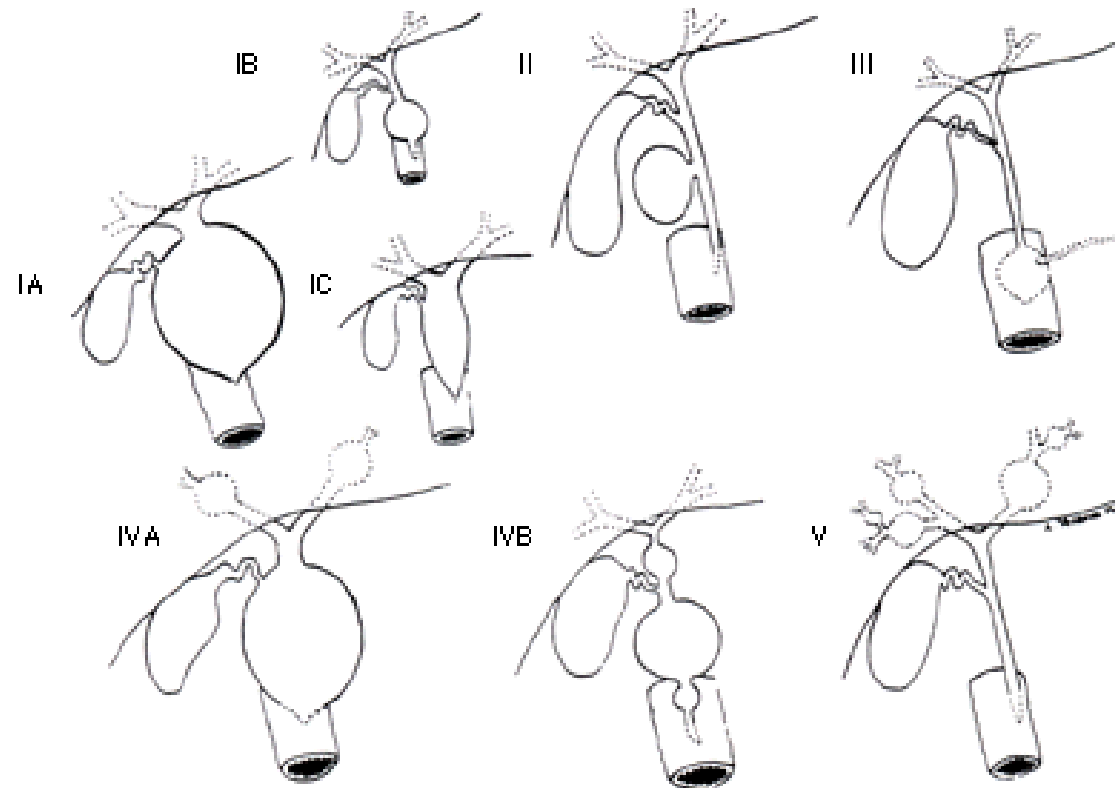


FIGURE 100-7. Todani's classification of biliary cysts based on location. Hatched areas represent cystic dilations. See text for details.

- **Type V** includes isolated or multiple cystic dilations of the **intrahepatic** ducts **without extrahepatic** duct disease, also known as **Caroli's** disease.



- Film



Classification of choledochal cysts according to Todani and colleagues (IA) common type; (IB) segmental dilatation; (IC) diffuse dilatation; (II) diverticulum; (III) choledochocoele; (IVA) multiple cysts (intra- and extrahepatic); (IVB) multiple cysts (extrahepatic); (V) single or multiple dilations of the intrahepatic ducts. Reproduced with

CLINICAL MANIFESTATIONS

- Infants with biliary cysts commonly present with
 - Conjugated hyperbilirubinemia (80 percent)
 - Failure to thrive
 - An abdominal mass (30 to 60 percent)
- The triad of **pain, jaundice**, and abdominal **mass** is found in **11** to **63 percent**

CLINICAL MANIFESTATIONS

- In contrast, **chronic** and **intermittent** abdominal pain appears to be the most **common** presenting symptom (50 to 96 percent) in patients **older than two**.
- **Intermittent jaundice** and **recurrent cholangitis** are also common (34 to 55 percent).
- An abdominal **mass** is **less common** (10 to 20 percent).

CLINICAL MANIFESTATIONS

- **Pancreatitis** has been described in approximately **20** percent of patients at diagnosis while **biliary lithiasis** has been described in only about **8** percent of patients
- Pancreatitis and ductal **stones** are more commonly encountered with **choledochoceles**

DIAGNOSIS

- Rarely, biliary cysts present with
 - Intraperitoneal **rupture**
 - **Bleeding** due to erosion into adjacent vessels
 - **Portal hypertension** and **cirrhosis**



DIAGNOSIS

- A diagnosis of biliary cyst should be **considered** in adults when a **dilated portion** of the bile ducts or ampulla is identified, especially in the **absence** of overt **obstruction**

DIAGNOSIS

- A **high** level of **suspicion** is required for diagnosis, particularly for **type I** cysts, which may go undiagnosed unless considered in the differential diagnosis of patients found to have ductal dilation.
- On the other hand, acute or chronic biliary obstruction may cause marked biliary dilation that mimics a type I cyst.

DIAGNOSIS

- Such patients usually present with jaundice or markedly elevated serum alkaline phosphatase, have a readily identifiable obstructing lesion such as a stone or stricture, and their biliary **dilation** often **improves after** appropriate **treatment**
- Careful evaluation for **APBJ** may help with diagnosis in **indeterminate** cases

DIAGNOSIS

- Cross-sectional imaging with **ultrasound** or **CT** may suggest the presence of a biliary cyst
- Direct cholangiography (whether intraoperative, percutaneous, or endoscopic) has long been considered the best test for diagnosis and evaluation

DIAGNOSIS

- Cholangiography
 - Demonstrates areas of cystic dilation
 - Excludes overt obstruction of the bile duct
 - Described the presence of an APBJ
 - Demonstrate stones or malignancy in the cyst

DIAGNOSIS

- MRCP also appears to be **useful** for diagnosis
 - Accurately demonstrates cystically dilated segments of the biliary tree
 - Identifies APBJ in over 75 percent of cases
- However, MR is **less sensitive** than direct cholangiography at **excluding obstruction**

DIAGNOSIS

- Endoscopic ultrasound (EUS) can also
 - Demonstrate extrahepatic biliary cysts
 - Provide detailed images of the cyst wall and pancreaticobiliary junction
- Intraductal ultrasound (IDUS) has been used for diagnosis of
 - Early malignant changes in a biliary cyst
 - More sensitive than direct cholangiography for detection of early malignancy in the cyst wall

DIAGNOSIS

- Choledochoceles, or type III cysts, are often first suspected during ERCP when a **dilated intramural** portion of the bile duct is seen **endoscopically**
- The dilated segment may become
 - Much more apparent during contrast injection
 - Ballooning in shape as it fills with contrast
- **Large** type III cysts may be apparent on **CT** or **upper GI series**
 - As a filling defect in the duodenal lumen

COMPLICATIONS

- largely due to obstruction and include
 - Jaundice
 - Stone formation
 - Recurrent cholangitis
 - Hepatic abscess
 - Portal vein thrombosis
 - pancreatitis, cyst rupture
 - Secondary biliary cirrhosis
 - Carcinoma

DISCUSSION

- Malignant change is estimated to occur in 3-20%, although carcinoma associated with type II cysts (choledochoceles) is more rare

DISCUSSION

- Most tumors are found synchronously when the cyst is diagnosed, although a significant proportion arise metachronously, even after 20 years following diagnosis (9)

DISCUSSION

- The cyst is often the site of malignant change, but there is also an increased risk of malignancy in any part of the bile ducts, gallbladder and pancreas.
- Most tumors are adenocarcinomas, although squamous tumors and cholangiocarcinoma occur.
- Prognosis of tumors is dismal, with only 5% patients surviving two years (10).

CANCER RISK

- Biliary cysts are associated with an increased risk of cancer, particularly **cholangiocarcinoma**
 - The incidence of malignancy varies with age
 - In a 1983 review of all published series of biliary cysts, the incidence of cancer was
 - 0.7 percent in patients under 10 years of age
 - 6.8 percent in patients 11 to 20 years of age
 - 14.3 percent in patients over 20 years of age
 - As high as 50 percent has been reported in older patients

CANCER RISK

- The risk of malignancy is best studied in type I and IV cysts
 - Type V cysts (**Caroli's disease**) have also been associated with a **7 percent** risk of malignancy
 - Cancer is an **uncommon** complication of **type III cysts**, and may be limited to those choledochoceles lined by biliary rather than duodenal epithelium

CANCER RISK

- Patients previously treated with **biliary bypass** rather than cyst resection have a **risk** of subsequent malignancy that seems **higher than** the risk in **unoperated** patients
- Patients who have previously undergone cyst **resection** continue to have an **increased risk of carcinoma**
- Malignancy may develop in portions of cysts that were
 - Residual at surgery
 - At the anastomotic site
 - Or in the pancreas

DISCUSSION

- Treatment is usually surgical
- Complete cyst resection and biliary bypass is the accepted treatment, although anatomic considerations often make complete resection difficult

DISCUSSION

- The development of carcinoma after biliary cyst excision is rare but has been reported (11).
- Hepatic resection and even liver transplantation has been advocated for intrahepatic cysts.

MANAGEMENT

- Because of the risk of malignant degeneration of the cyst, the current standard is surgical excision, with the goal of removing all of the cyst tissue when possible
- Excision of cysts can reduce complications such as
 - Recurrent cholangitis
 - Choledocholithiasis
 - Pancreatitis

Which were frequently seen following the older surgical strategy of cyst drainage by choledochojejunostomy, without resection

MANAGEMENT

- In the case of **extrahepatic cysts**, resection is usually followed by **hepaticojejunostomy** for reconstruction
- A **partial hepatectomy** may be indicated if **intrahepatic cysts** are present and resectable

MANAGEMENT

- Surgical resection
 - Effectively treats symptoms and appears to decrease
 - Although not eliminate, cancer risk
- The most frequent **long-term complication** of hepaticojejunostomy is
 - **Stenosis** of the biliary-enteric **anastomosis** leading to
 - Cholangitis
 - Jaundice
 - Cirrhosis
 - This complication occurs in up to **25 percent** of patients over time

MANAGEMENT

- Patients with **Caroli's** disease may eventually require liver **transplantation**
- Type **IIIA** cysts (choledochoceles) are often amenable to endoscopic **sphincterotomy**
- Type **IIIB** cysts (diverticular choledochoceles) may be **resected** surgically or endoscopically

MANAGEMENT

- There is no proven, effective method of **screening** biliary cysts for **dysplasia** or intra-mucosal **cancer**.
- If screening is attempted, intraductal ultrasound is probably the **most sensitive** available test for early malignancy in the **cyst wall**.



**THE
END**



**THE
END**



6 12 2004

CASE HISTORY 3

- This 67 y/o female patient suffered from dull abdominal pain about 3 months ago.
- The abdominal pain was dullness in nature without relationship to eating.
- No associated symptoms such as nausea, vomiting, diarrhea, or tarry stool.

CASE HISTORY 2

- Abdominal sonography showed
 - Diffuse liver parenchymal disease
 - Gallbladder stones with cholecystitis
 - CBD and IHD dilatation
 - Splenomegaly
 - Hepatic cyst were also noted

PRESENT ILLNESS(3)

- ERCP was performed
 - PD: normal size and shape
 - CBD: poor visualization due to poor filling
 - GB and cystic duct and IHD: not visualization

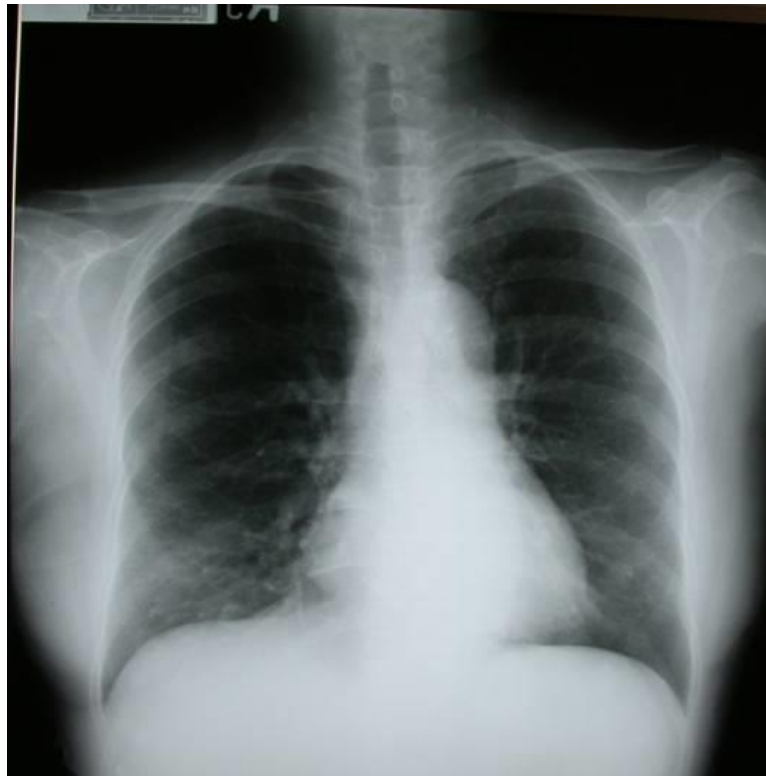
PHYSICAL EXAMINATION

- Icteric sclera
- Abdomen
 - Soft and mild distention
 - Mild RUQ tenderness
 - Murphy's sign(-)

LAB DATA

- WBC/DC:WNL
- Hb: 13.0
- Platelet: 119000
- AST/ALT: 34/34
- CEA: 8.14

CXR: NORMAL



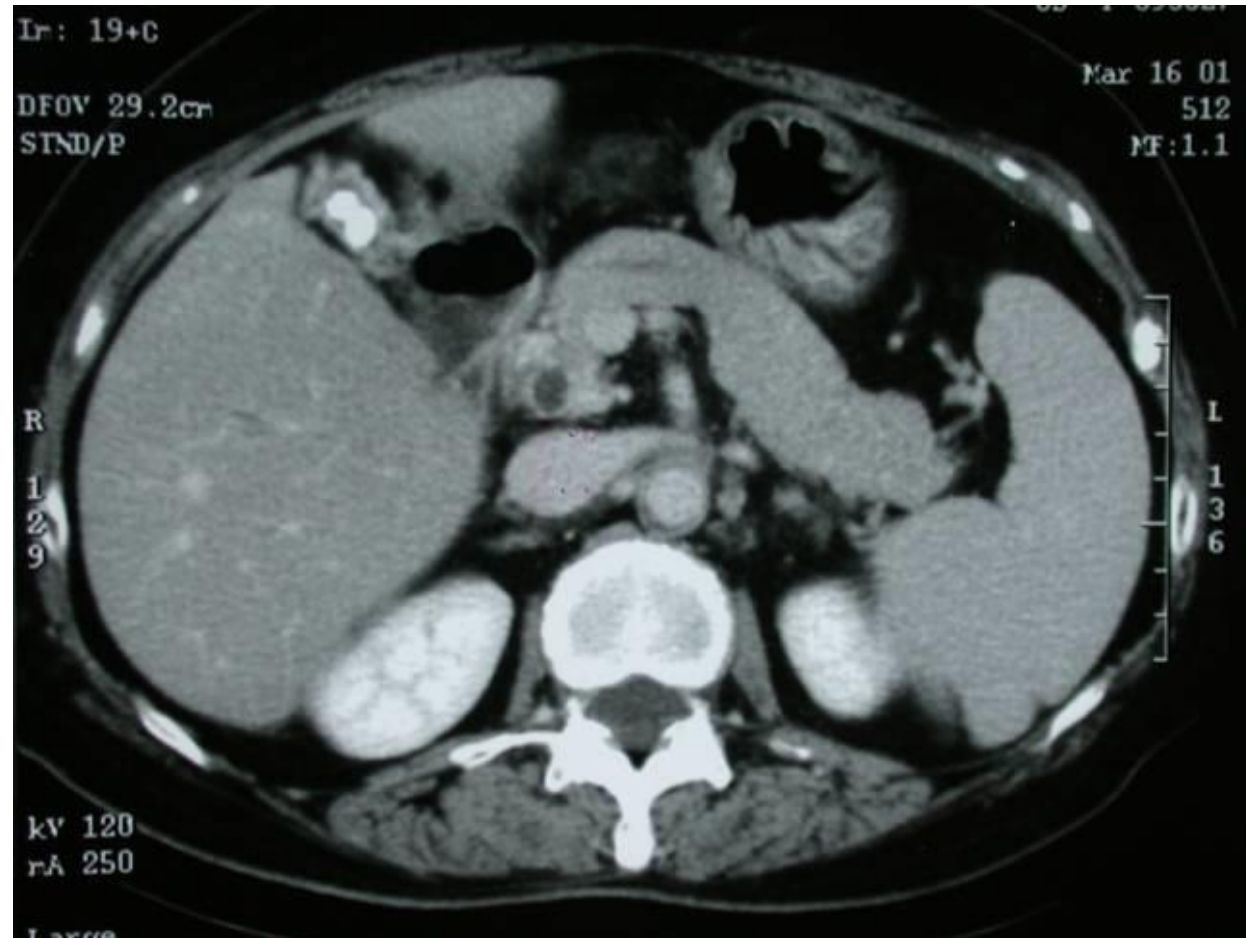
KUB: GALLSTONES NOTED



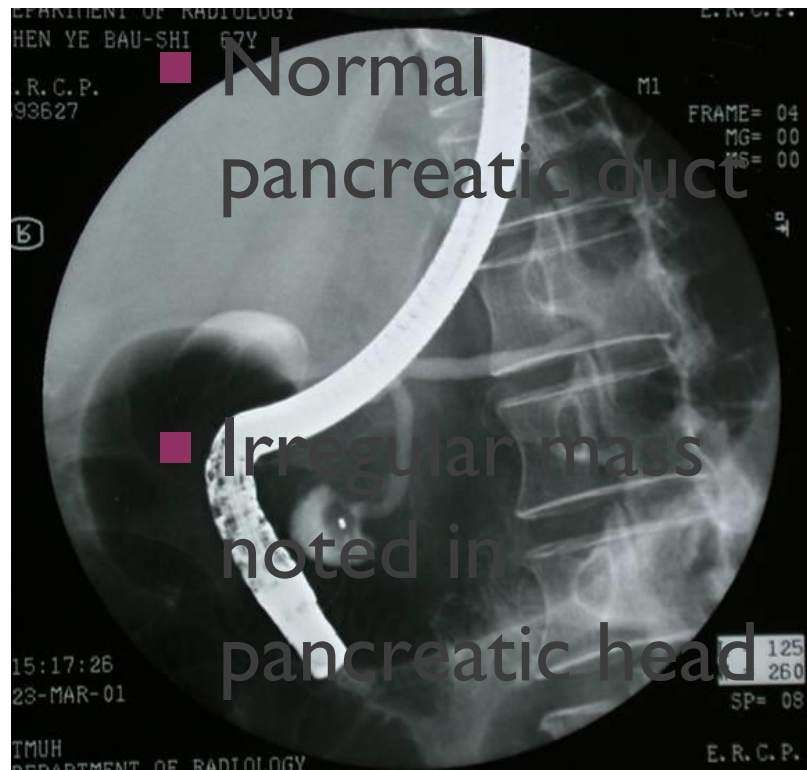
ABDOMINAL CT



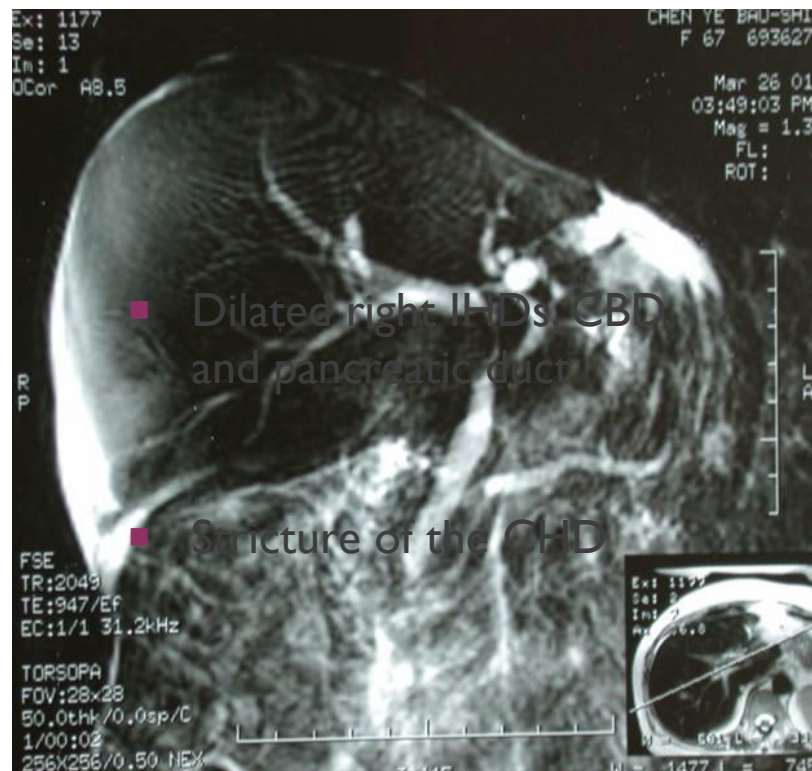
MULTIPLE GALLSTONES IN GALLBLADDER DILATED CBD NORMAL PANCREAS



ERCP



MRCP



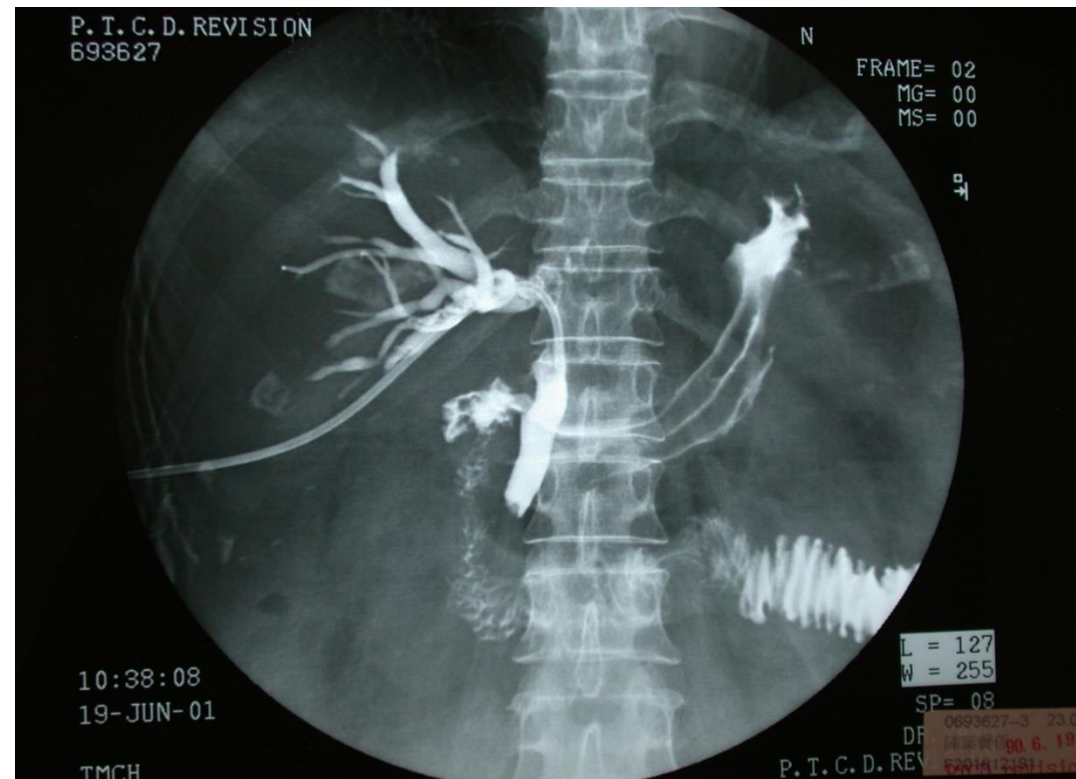
BARIUM STUDY

- mucosal tethering noted at the transverse and ascending colon
- Tumor seeding should be considered



PTCD

- Dilatation of the right intrahepatic ducts with stricture of the common bile duct.



DILATATION OF THE RIGHT INTRAHEPATIC DUCTS STRICTURE OF THE COMMON BILE DUCT.



SUMMARY OF IMAGE FINDINGS

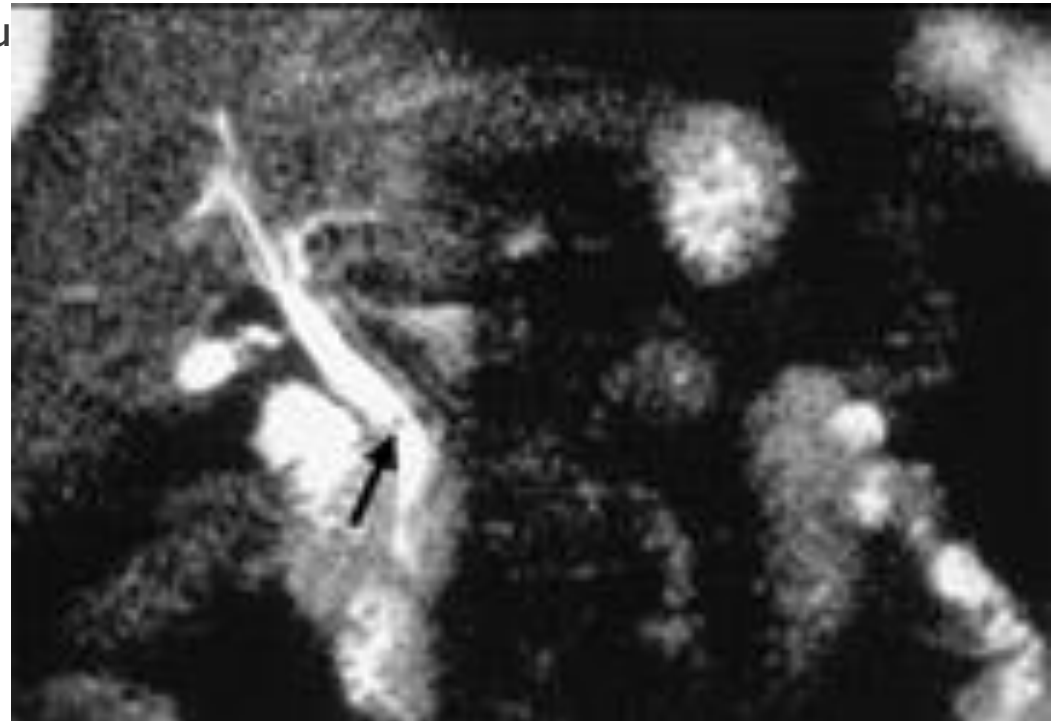
- Multiple gallstones in gallbladder
- Dilated CHD, IHDs and stricture of CBD
- Segmental narrowing with irregularity at the common hepatic artery and proximal portion of gastroduodenal artery
- mucosal tethering noted at the transverse and ascending colon
- Splenomegaly

DIFFERENTIAL DIAGNOSIS OF BILIARY TRACT OBSTRUCTION

- Benign—75%
- Benign stricture
 - Surgery/instrumentation
 - Trauma
 - Stone passage
 - Cholangitis
 - Choledochal cyst
- Stone impacted in duct
- Parasite (ascariasis)
- Liver cyst
- Malignant—25%
- Pancreatic carcinoma
- Ampullary/duodenal carcinoma
- Cholangiocarcinoma
- Metastasis

CHOLEDOCHOLITHIASIS

- Choledocholithiasis—20% of obstru
- PTC and ERCP are the most efficacious examination
- Most patients have gallstones in gallbladder



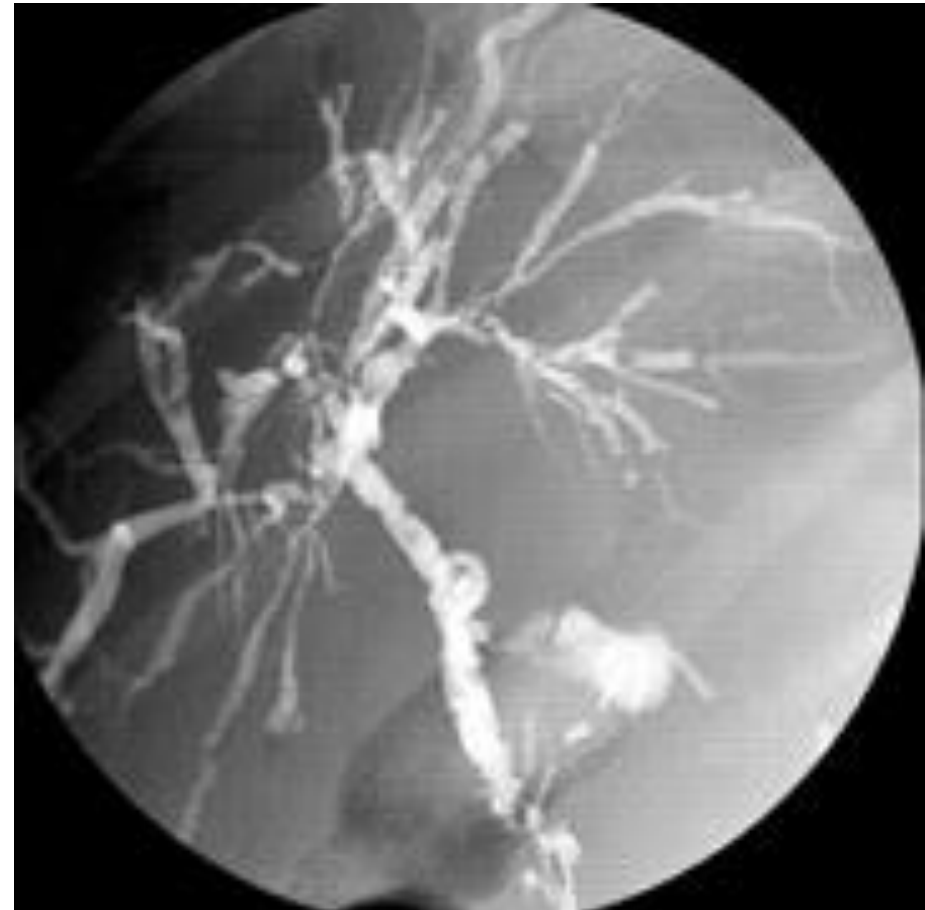
CHOLEDOCHAL CYST (UNCOMMON CONGENITAL DISORDER)

- Slight proximal dilatation of the common bile duct
- Narrowing of the distal common bile duct
- Abrupt transition between the two



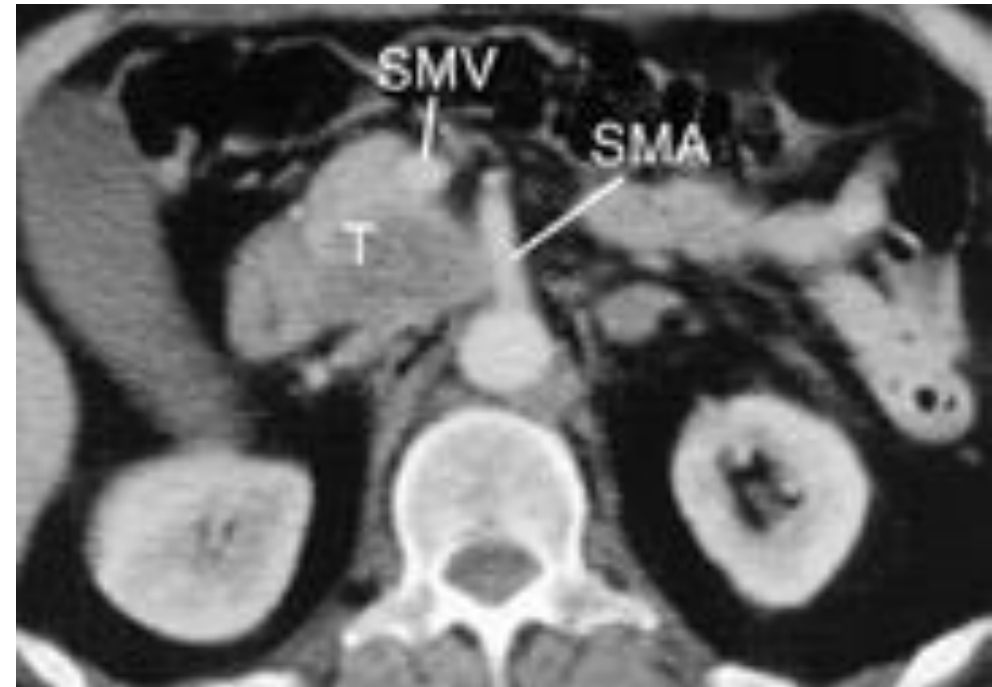
SCLEROSING CHOLANGITIS

- A history of ulcerative colitis in 50 % cases
- Elevated serum alkaline phosphatase
- Liver biopsy multiple short strictures and saccular dilations involving the intrahepatic and extrahepatic bile ducts give the biliary tree an irregular beaded appearance



AMPULLARY AND PANCREATIC TUMOR

- Most common cause of malignant bile duct stricture
- CT has become the gold standard for the diagnosis of pancreatic carcinoma

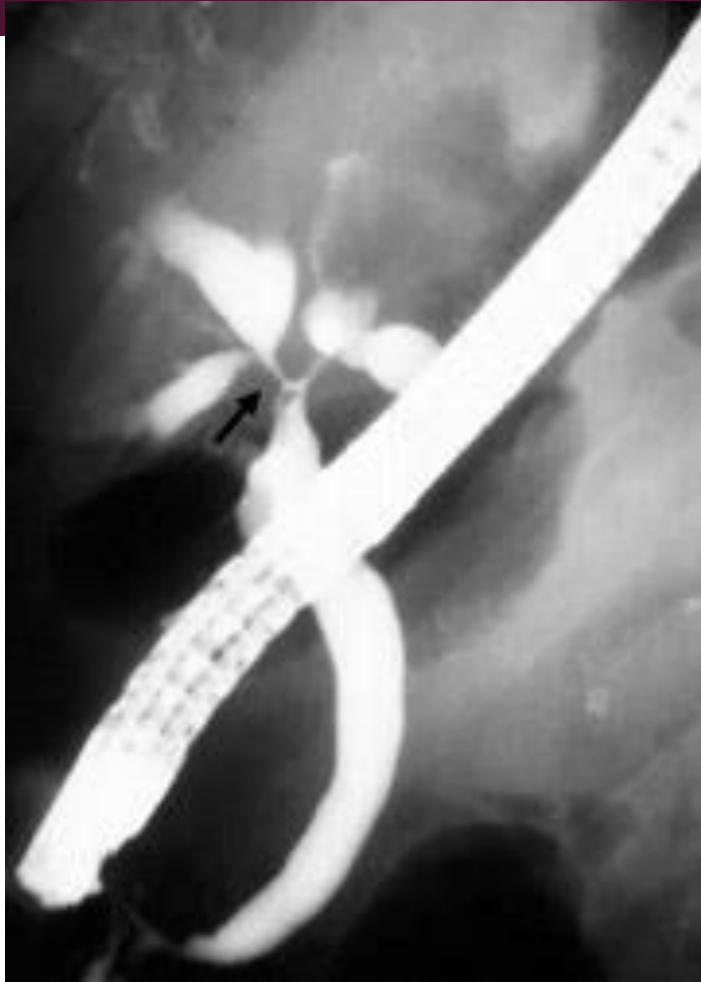


- *Character CT findings:* obstruction with uniform dilatation of the distal pancreatic duct in the absence of duct calculi (compared with the irregular chain of lakes of chronic pancreatitis)

CHOLANGIOCARCINOMA

- *Peripheral cholangiocarcinoma*
 - Present as an intrahepatic hypodense mass with adjacent biliary dilatation
- *Hilar cholangiocarcinoma*
 - **Klaskin's tumor** is usually small, poorly differentiated, aggressive, and cause obstruction of both ductal system
- *Extrahepatic cholangiocarcinoma*
 - Cause stenosis or obstruction of the CBD

CHOLANGIOCARCINOMA



- ERCP, demonstrating extreme stenosis of the confluence of the left and right hepatic duct (arrow), extending into the proximal portion of the common hepatic duct due to infiltrative form of CCC.

SURGICAL INTERVENTION

- Cholecystectomy + tumor biopsy
- Pathology: adenocarcinoma, hilum

DISCUSSION

- Cholangiocarcinomas (CCC) are malignancies of the biliary duct system, originating in the liver and terminating at the ampulla of Vater.

DISCUSSION

- The etiology of most bile duct cancers remains undetermined
 - Long-standing inflammation, as with primary sclerosing cholangitis (PSC)
 - Chronic parasitic infection, has been suggested as playing a role

DISCUSSION

- Symptoms may include
 - Jaundice
 - Clay-colored stools
 - Dark urine, pruritus
 - Weight loss
 - Abdominal pain
- The patient may have a palpable gallbladder, which commonly is known as Courvoisier sign

DISCUSSION

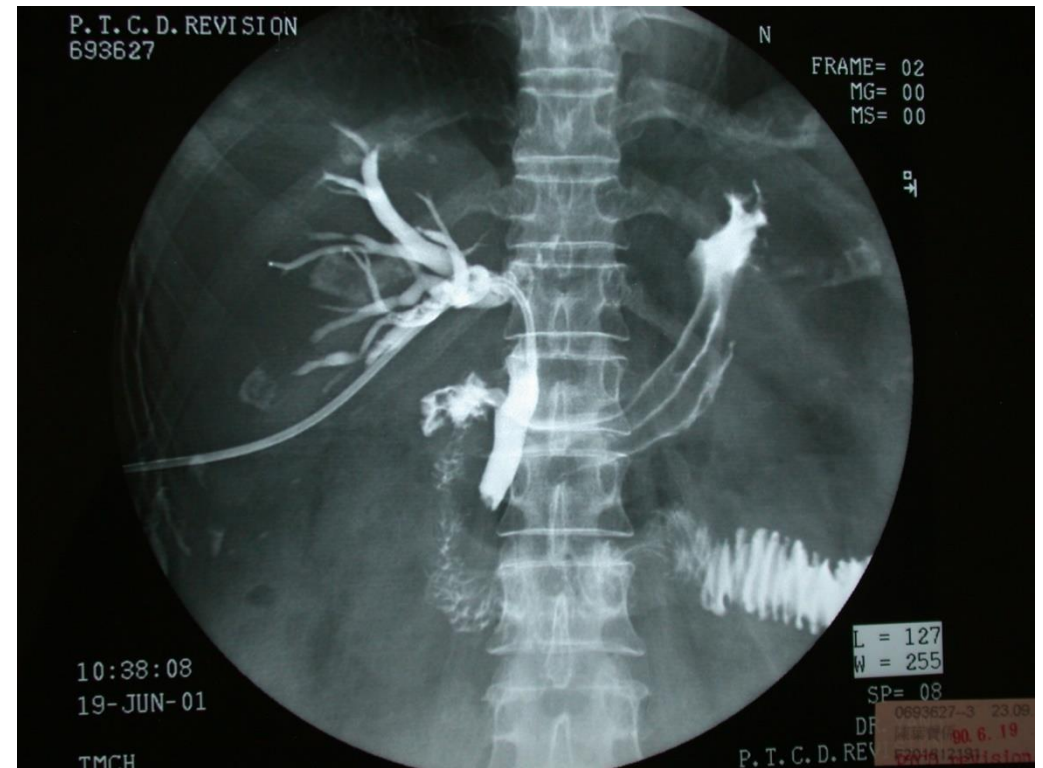
- Lab:
 - Elevated
 - Conjugated (i.e., Direct) bilirubin
 - Alkaline phosphatase
 - Gamma-glutamyltransferase (GGT)
 - AST/ALT may be normal or minimally elevated
 - With prolonged obstruction, PT can become elevated from vitamin K malabsorption

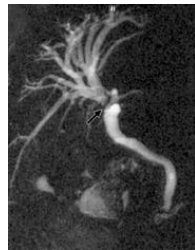
DISCUSSION

- In general, ultrasound or computed tomography (CT) scan is performed initially, followed by a type of cholangiography.
- CT scan resembles ultrasound in that it may demonstrate ductal dilatation and large mass lesions.



- Until recently cholangiography either by ERCP or PTCD available to display correctly the full extent of CCC with an accuracy varying between 89 and 96%.





- MRCP, revealing a marked dilatation of intrahepatic bile ducts. Extreme narrowing (arrow) of the confluence of the left and right hepatic duct.

DISCUSSION

- Complete surgical resection is the only therapy to afford a chance of cure.
- Unfortunately, only 10% of patients present with early stage disease and are considered for curative resection.
- Intrahepatic and Klaskin tumors require liver resection, which may not be an option for older patients with co-morbid conditions.

