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TABRIZ UNIVERSITY OF MEDICAL SCIENCES

Biliary Stones and Polyps

CASE HISTORY 1

- 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.

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CASE HISTORY 1

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

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DIFFERENTIAL DIAGNOSIS

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

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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

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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)

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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)

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PROTECTIVE FACTORS

- Ascorbic acid (Vitamin C)
  - %13, female
- Coffee
  - %40, 2-3
  - Decaffeinated coffee
- Vegetable protein
  - %20
- Poly- and monounsaturated fats

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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

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Characteristics of gallstone pain

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Characteristics of gallstone pain

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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.

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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.

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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

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CASE HISTORY CONTINUE

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

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‘Just in case’

- *What is your advise ?*
- *What are the indications for prophylactic surgery?*

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GALLSTONE TREATMENT

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

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GALLSTONE TREATMENT

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

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CONTINUED

- *What are the indications for nonsurgical treatment ?*

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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

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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

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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

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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)

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CONTINUED

- *What are the indications for prophylactic surgery?*

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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

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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

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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

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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

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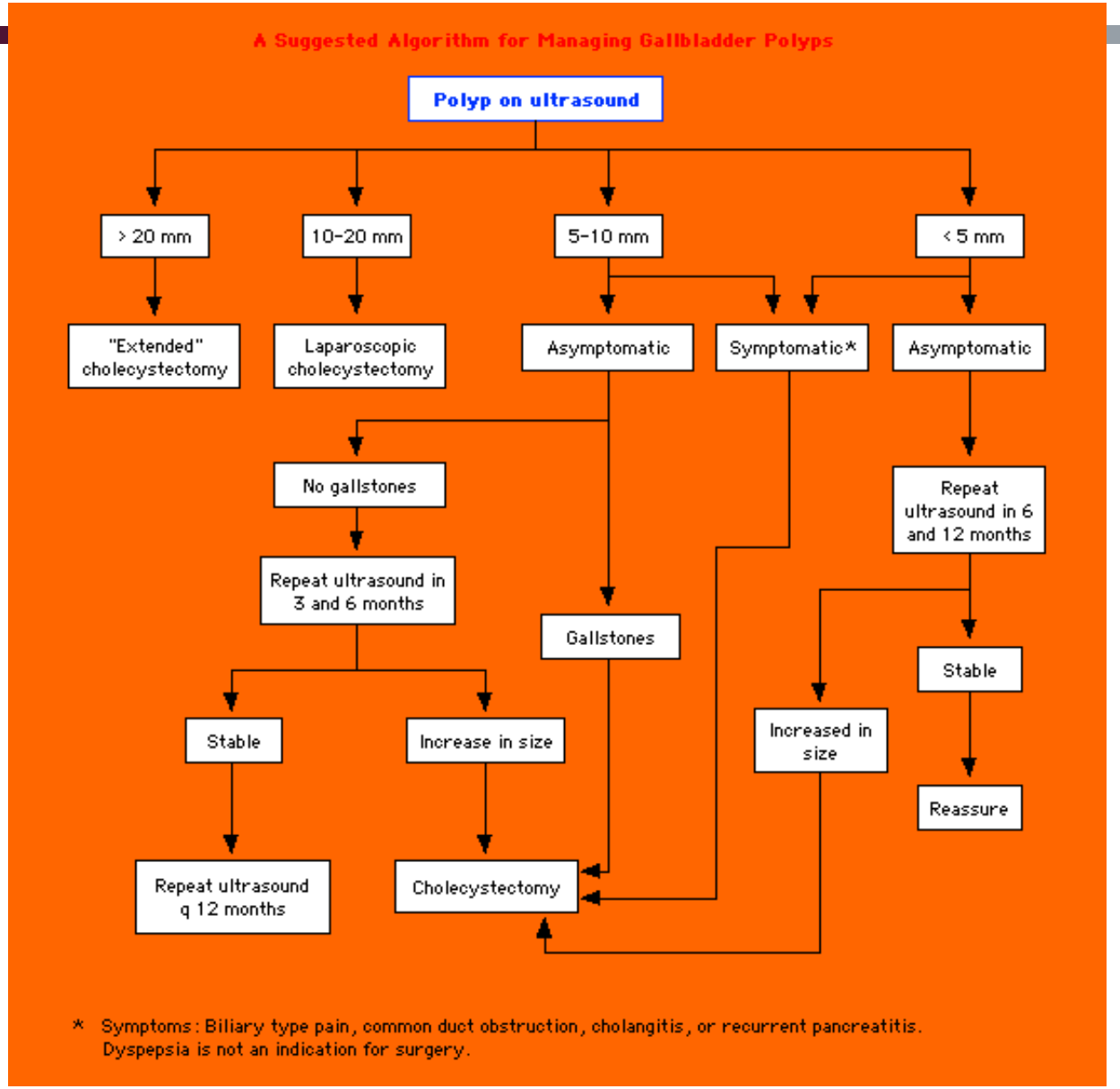

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ROLE OF PROPHYLACTIC CHOLECYSTECTOMY

- 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.

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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.

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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

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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

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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

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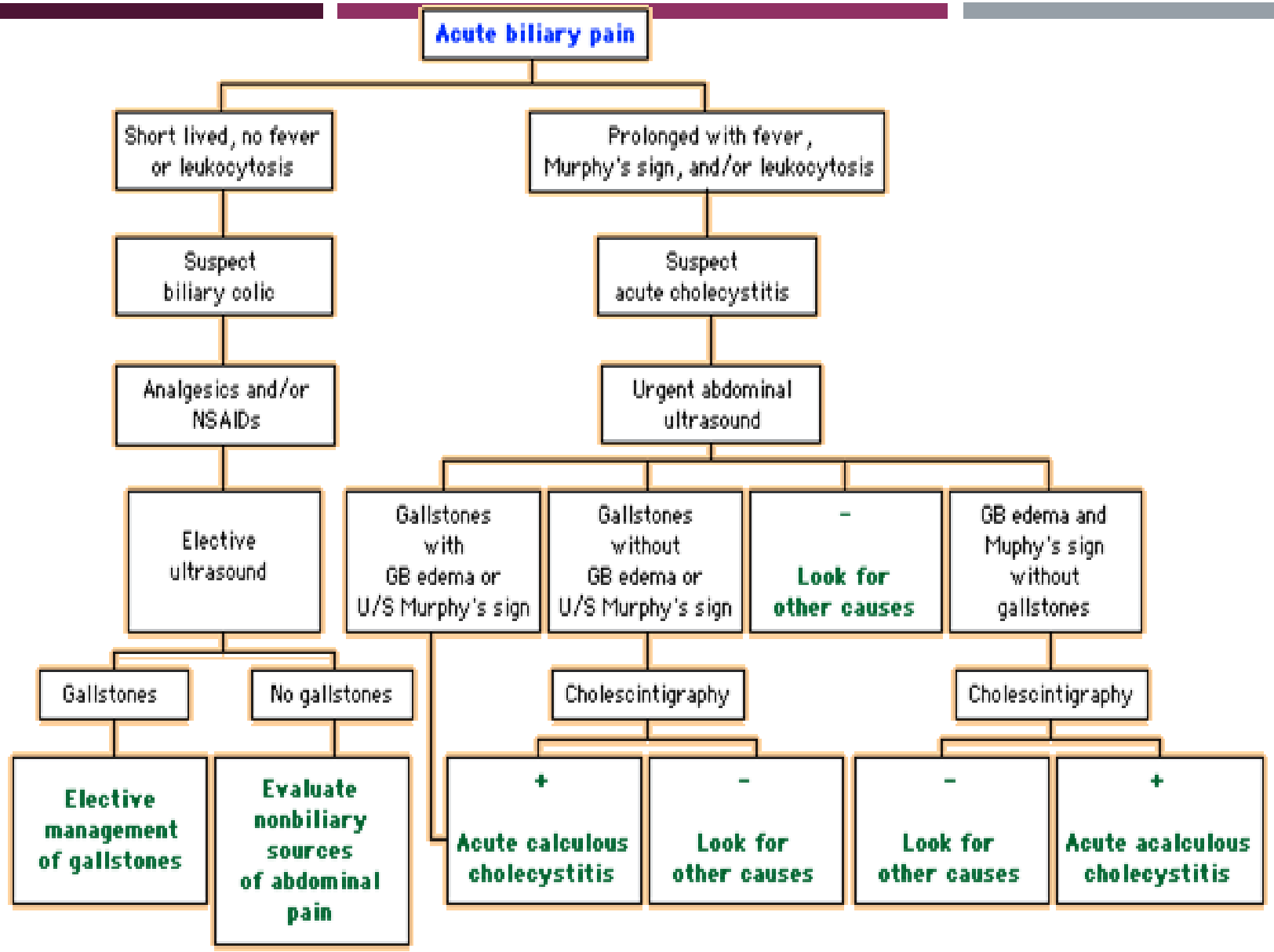

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ULTRASOUND

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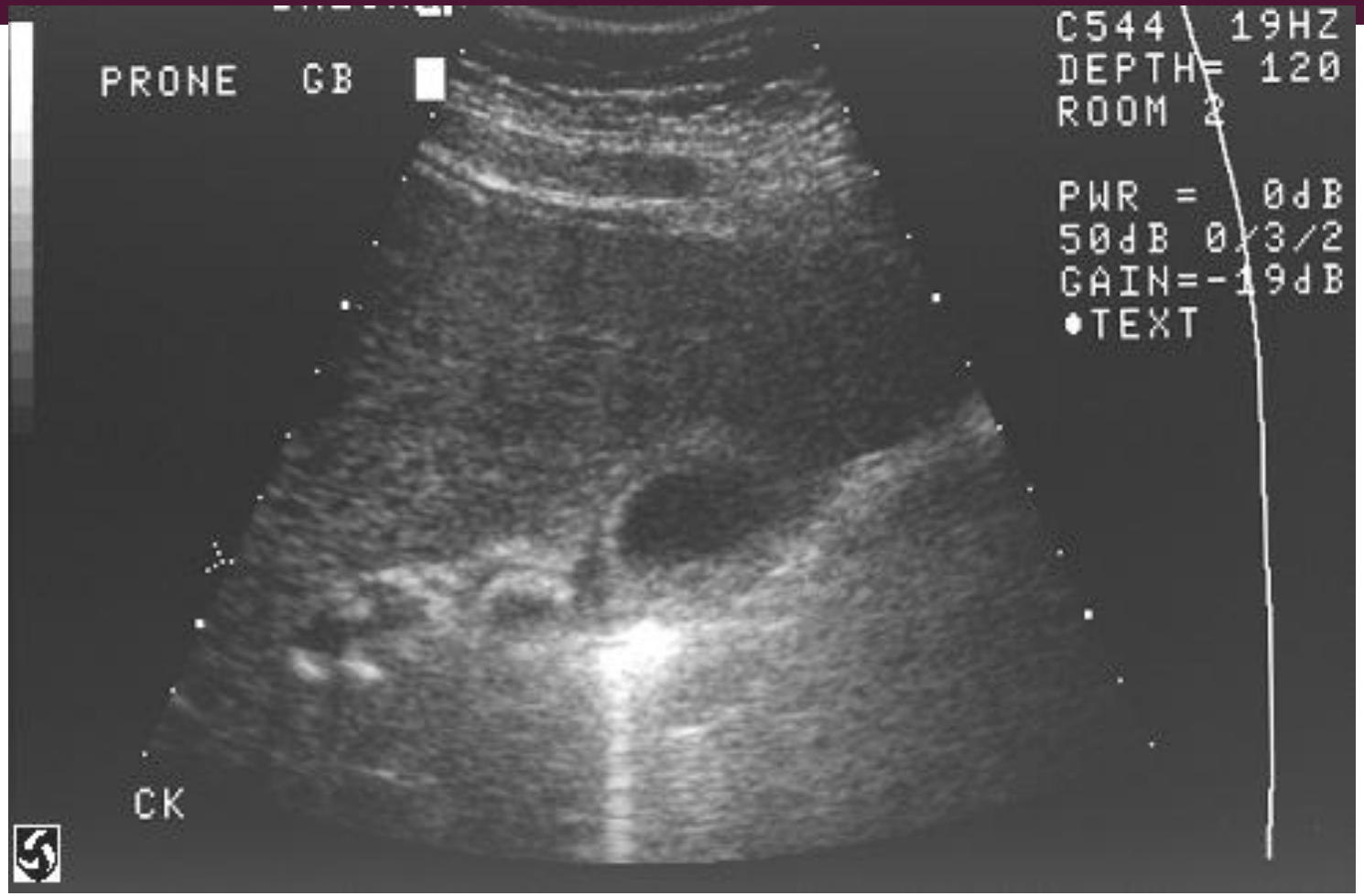

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ULTRASOUND

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ABDOMINAL ULTRASOUND

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

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ABDOMINAL ULTRASOUND

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

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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.

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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

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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

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ACUTE CHOLECYSTITIS

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

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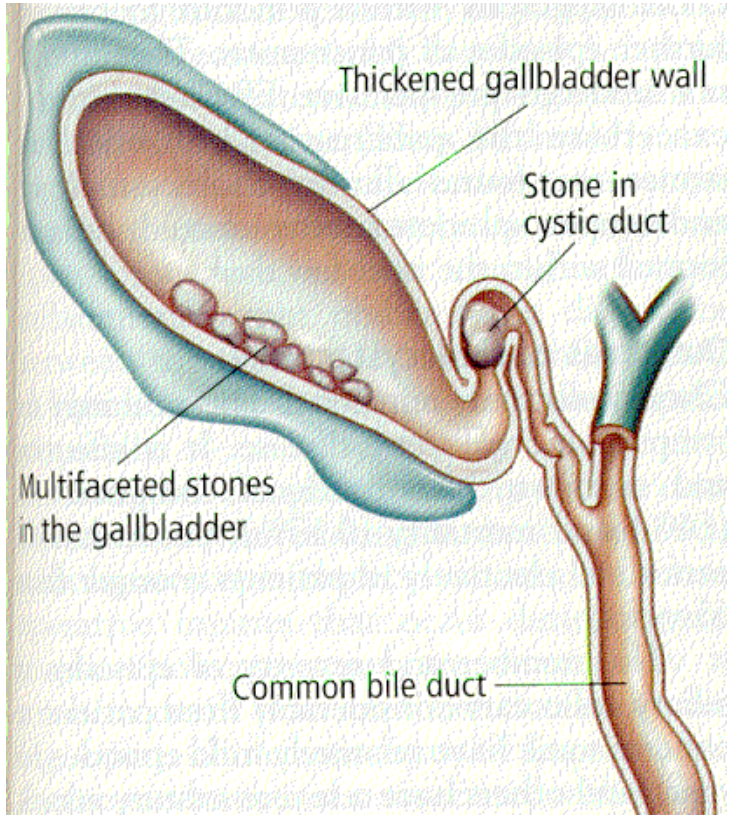
ACUTE CHOLECYSTITIS

- Distention and inflammation of the gallbladder

- Persistent obstruction of cystic duct ⇒ chemical irritants in the bile
  - Lysolecithin
  - Prostaglandins



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CHOLECYSTITIS

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

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DIAGNOSIS

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

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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

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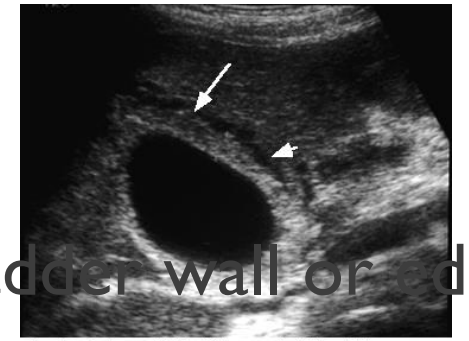
- Thickened gallbladder wall or edema
- Pericholecystic fluid
- Sonographic Murphy's sign



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

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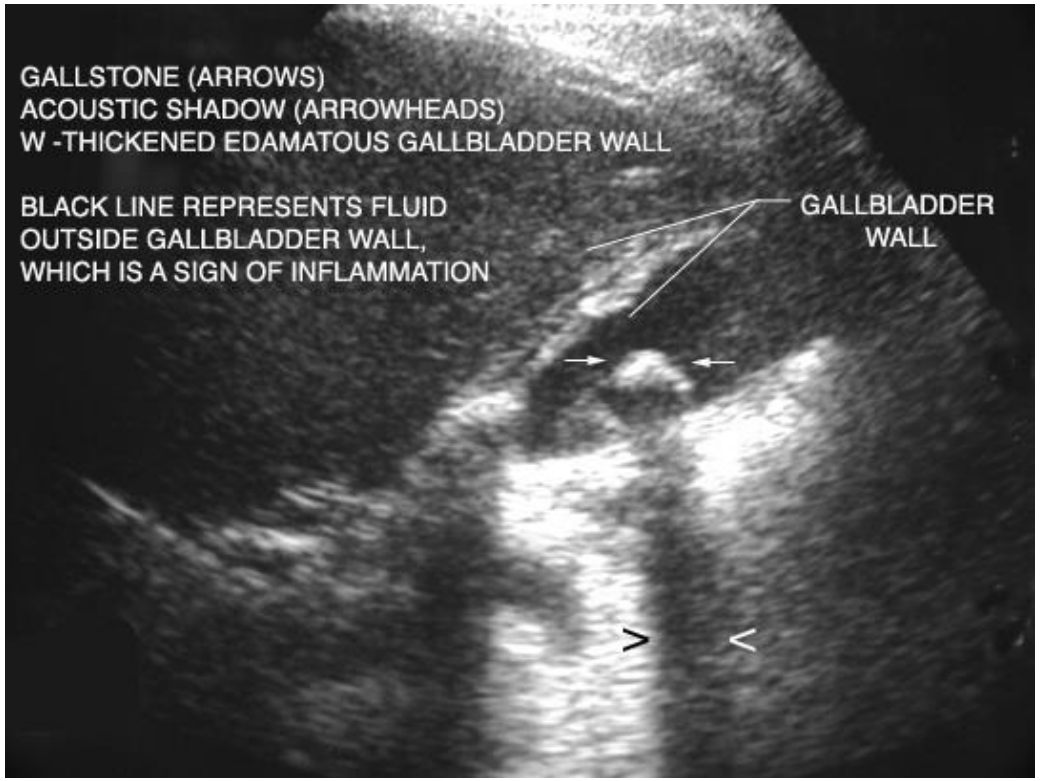
ACUTE CHOLECYSTITIS



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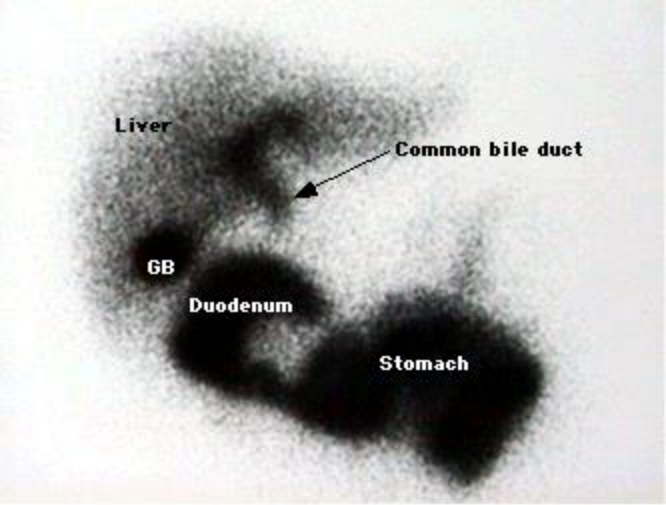
CHOLESCINTIGRAPHY

- 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.

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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

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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

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CHOLESCINTIGRAPHY

- False negative results are uncommon
  - Incomplete cystic duct obstruction

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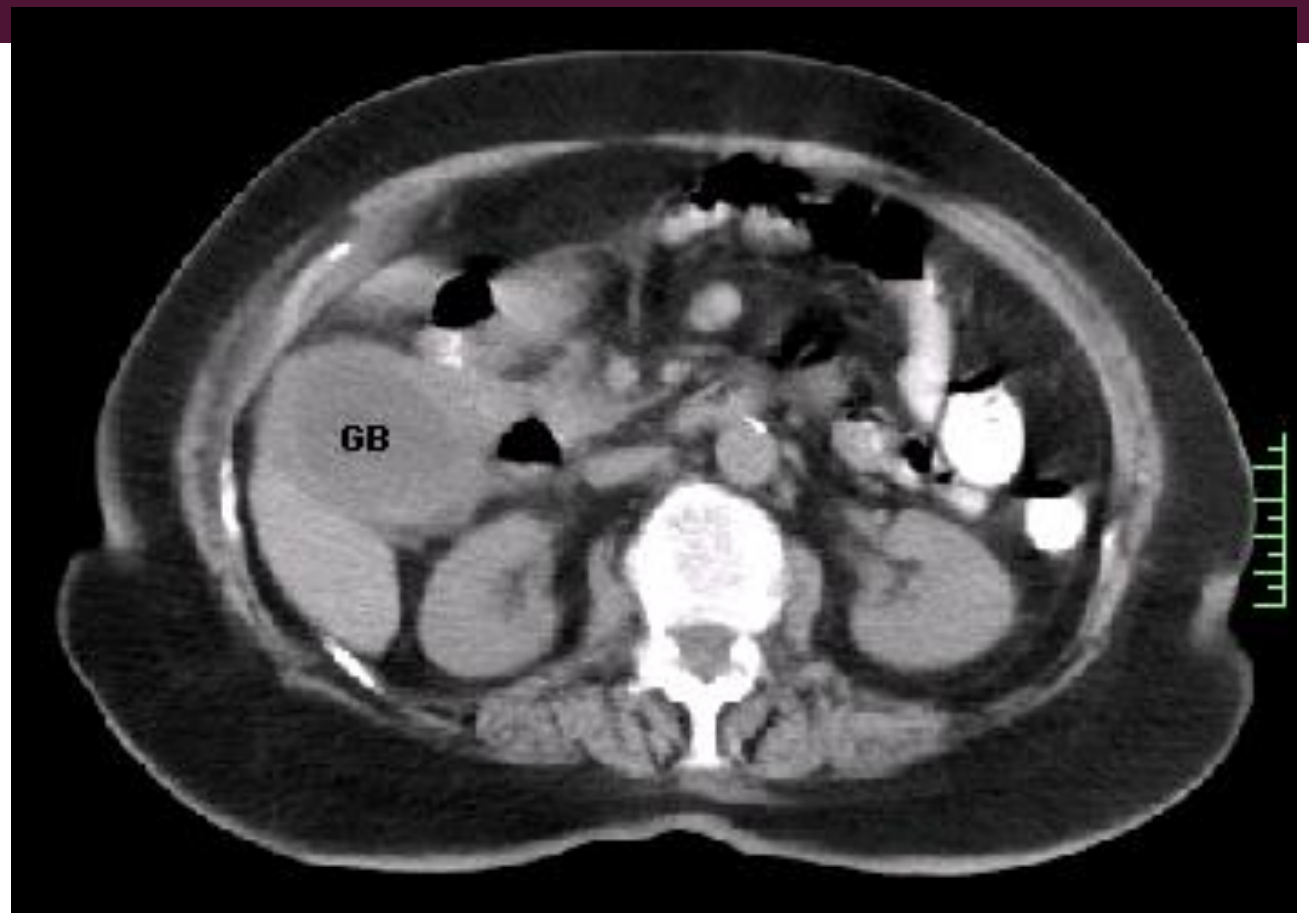
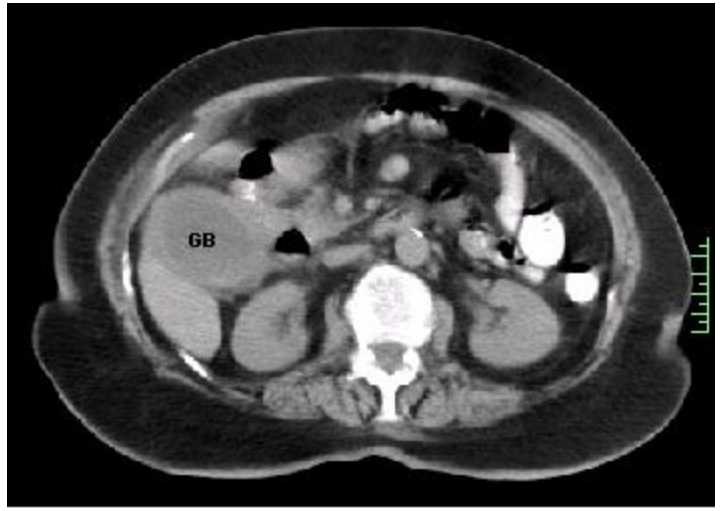
CT SCAN



CT SCAN

- CT scan
  - Usually unnecessary, although it is 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.

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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

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QUESTION

- What are you doing for management of this patient ?

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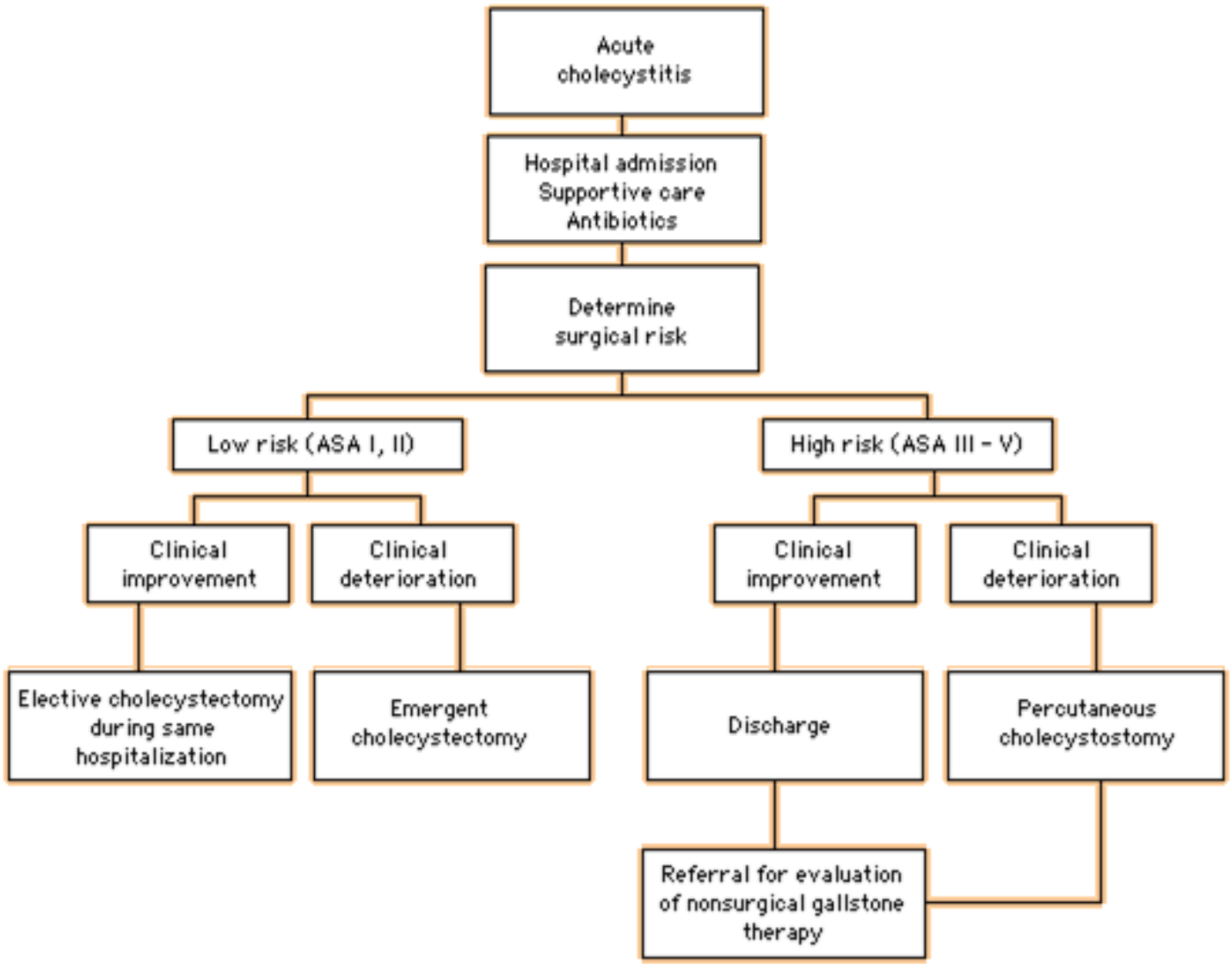
Treatment of Acute Cholecystitis[1]



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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

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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)

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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

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MANAGEMENT CONT.

- SURGERY is the only definitive treatment

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MANAGEMENT CONT.

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

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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
- Comp[illegible]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.

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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

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- 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.

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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

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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.

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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.

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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

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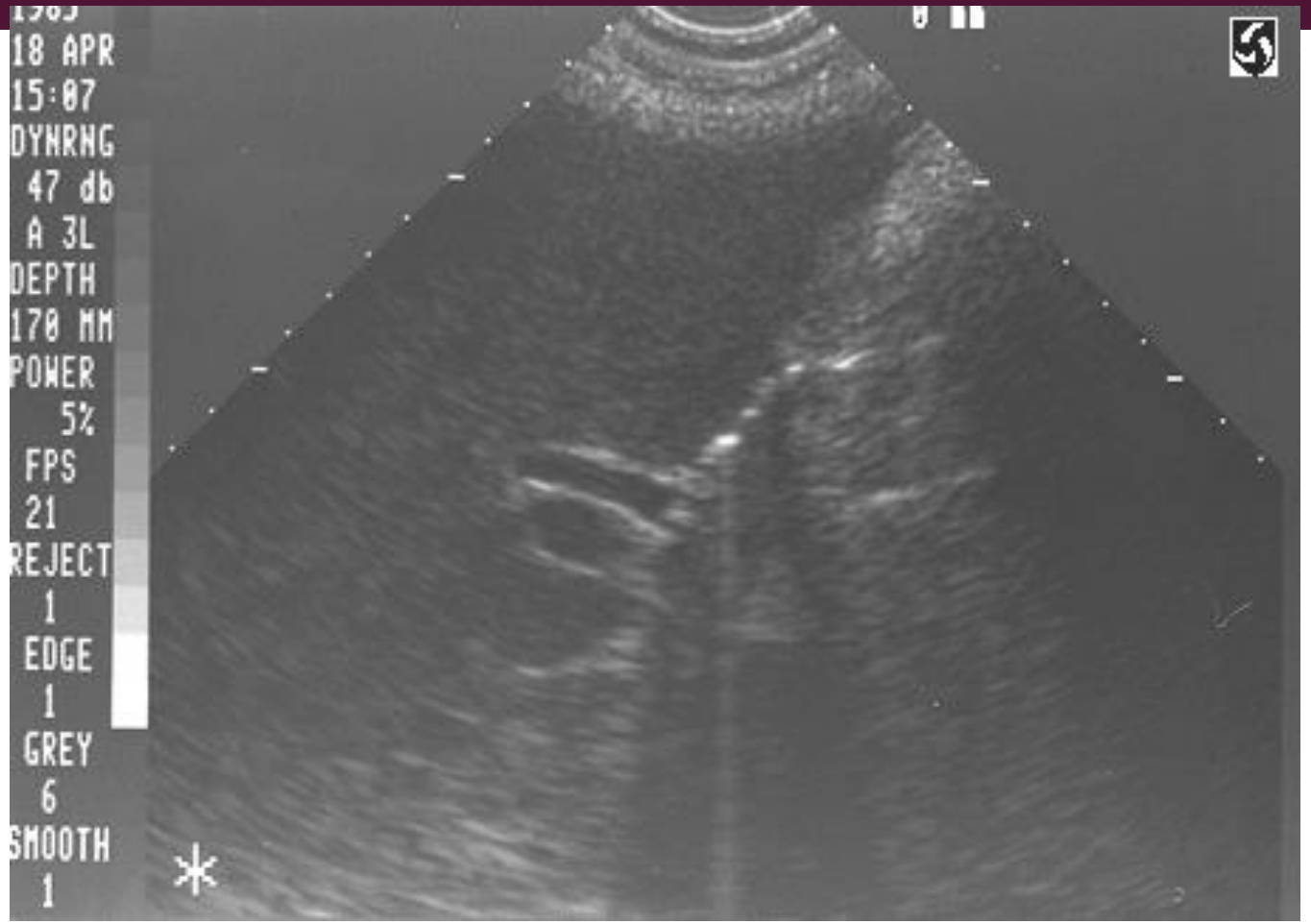
ULTRASOUND



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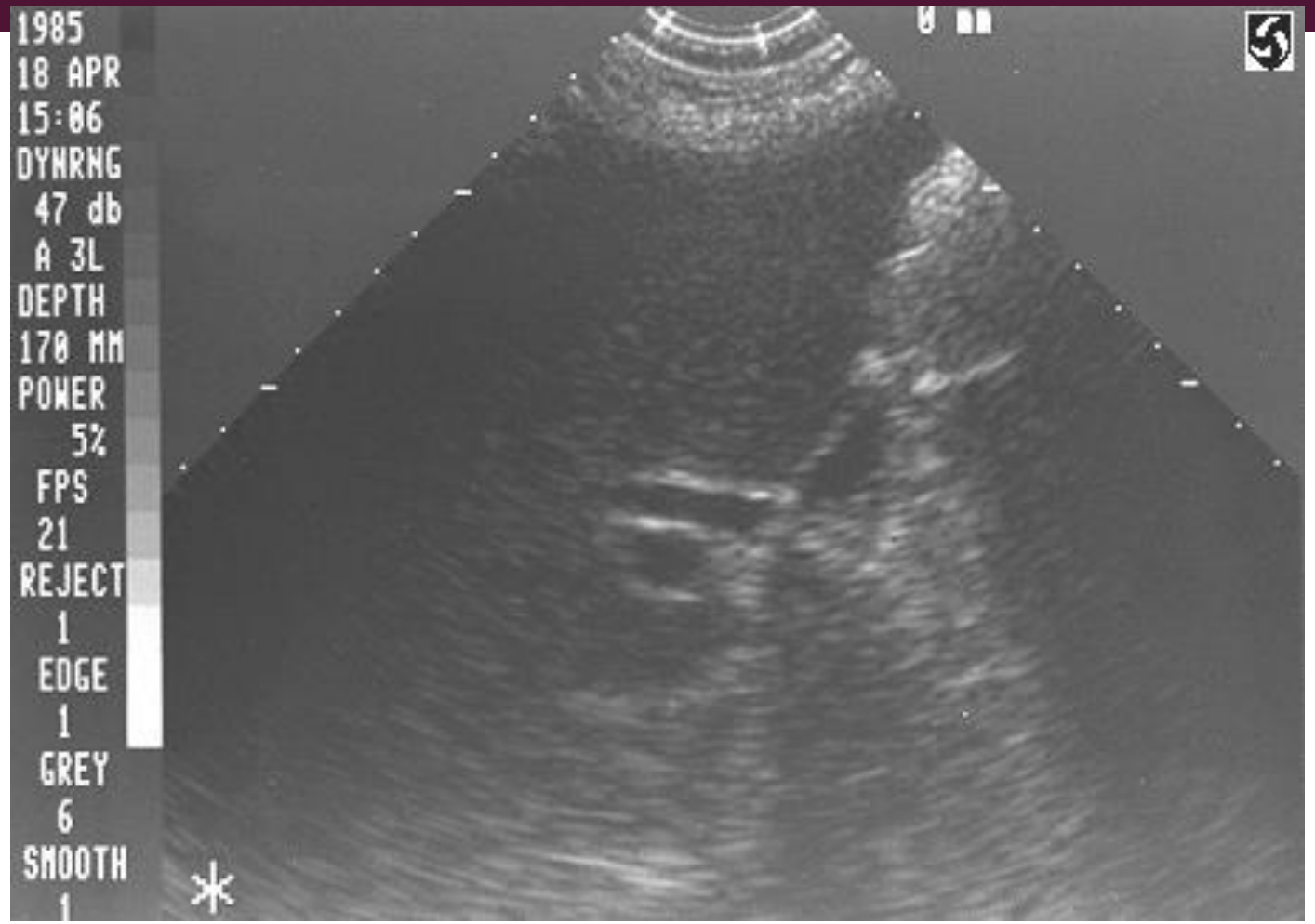
ULTRASOUND



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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

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ABDOMINAL ULTRASOUND

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

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QUESTION

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

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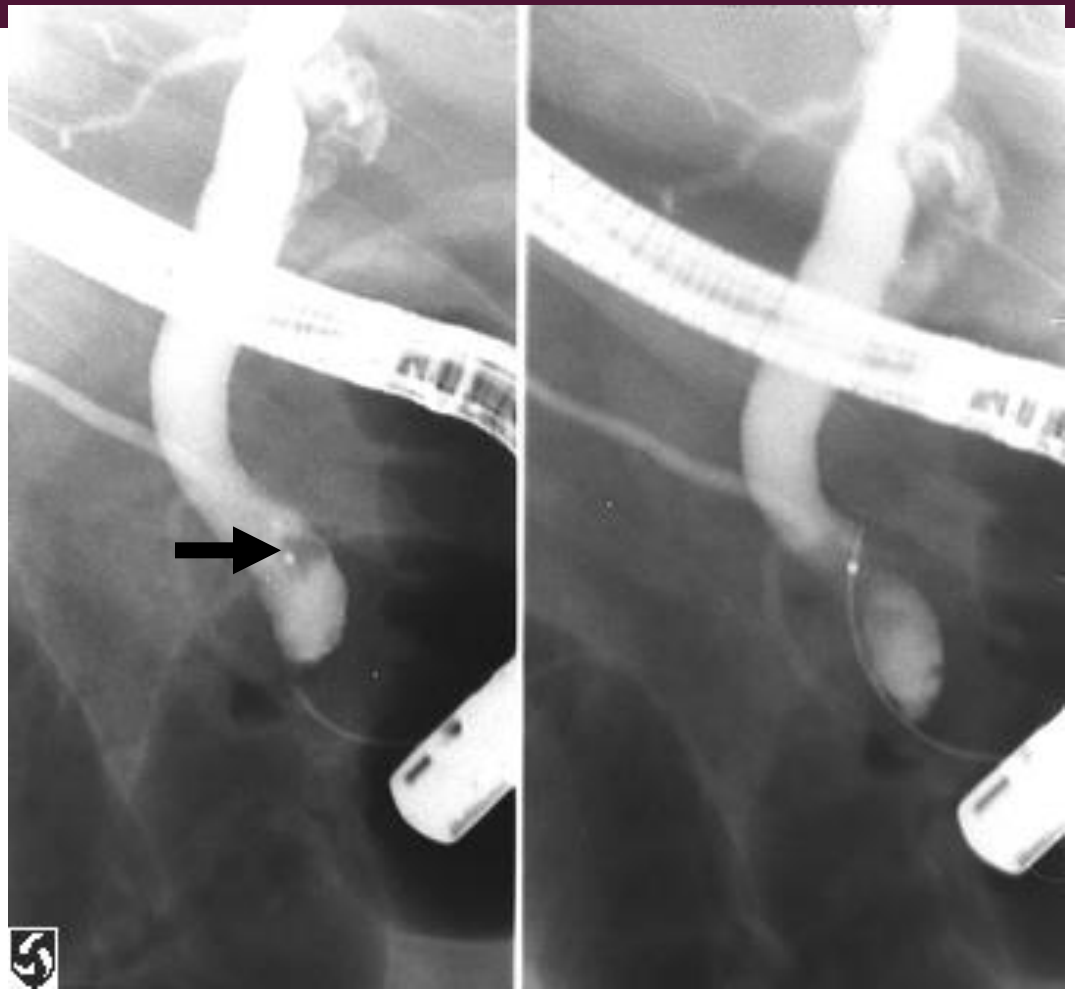
ENDOSCOPIC RETROGRADE CHOLANGIOPANCREATOGRAPHY WITH STONE EXTRACTION



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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.

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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.

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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.

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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

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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

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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

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SPECTRUM OF BILE DUCT STONE
CHOLEDOCHOLITHIASIS

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

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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

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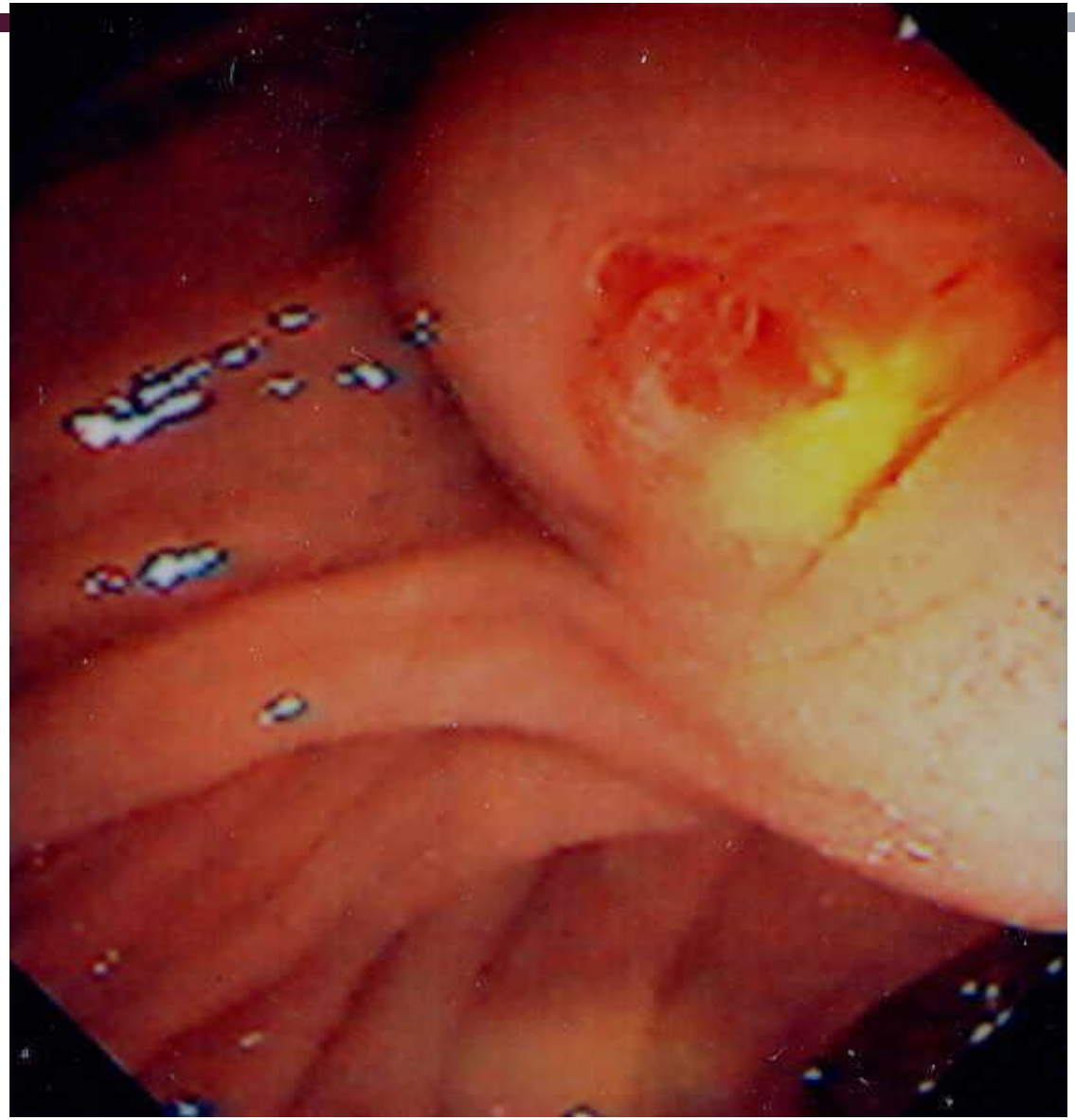
- فیلم
- ERCP و سنگ



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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.

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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.

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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.

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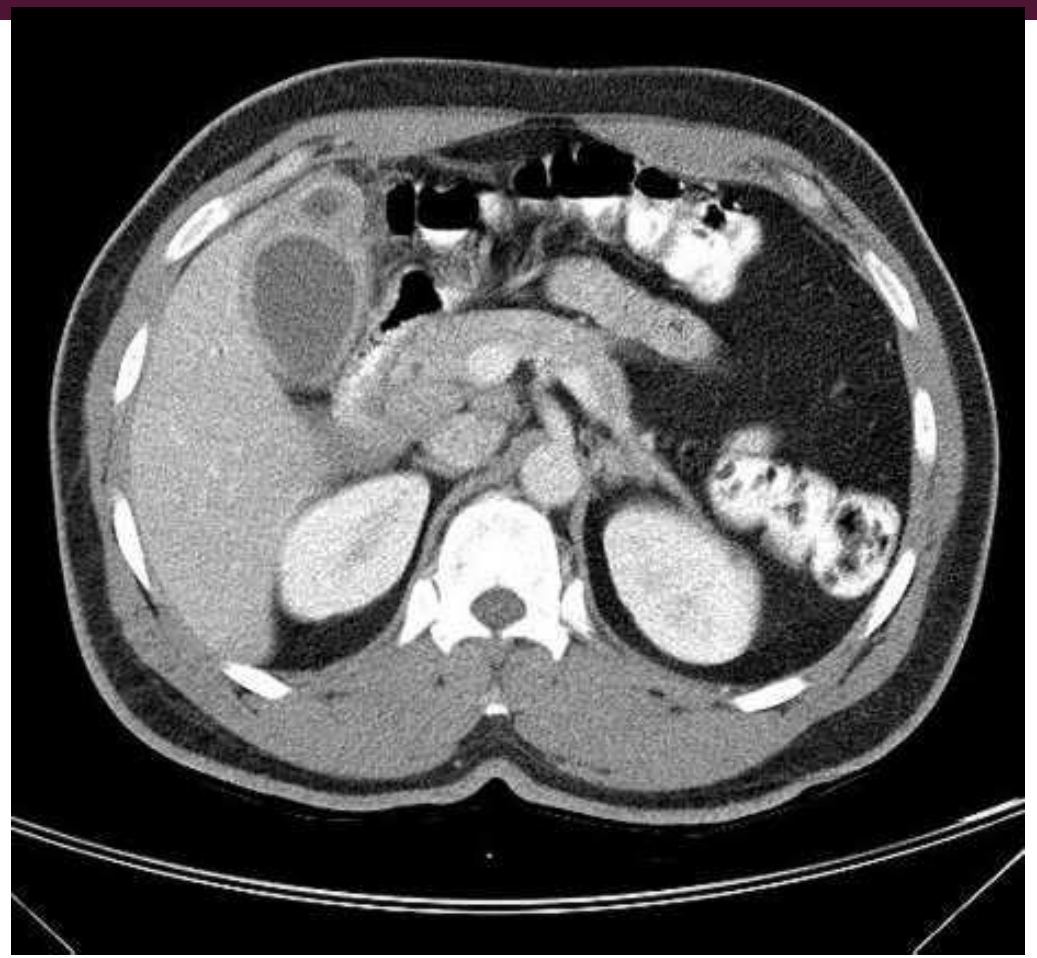
CT SCAN



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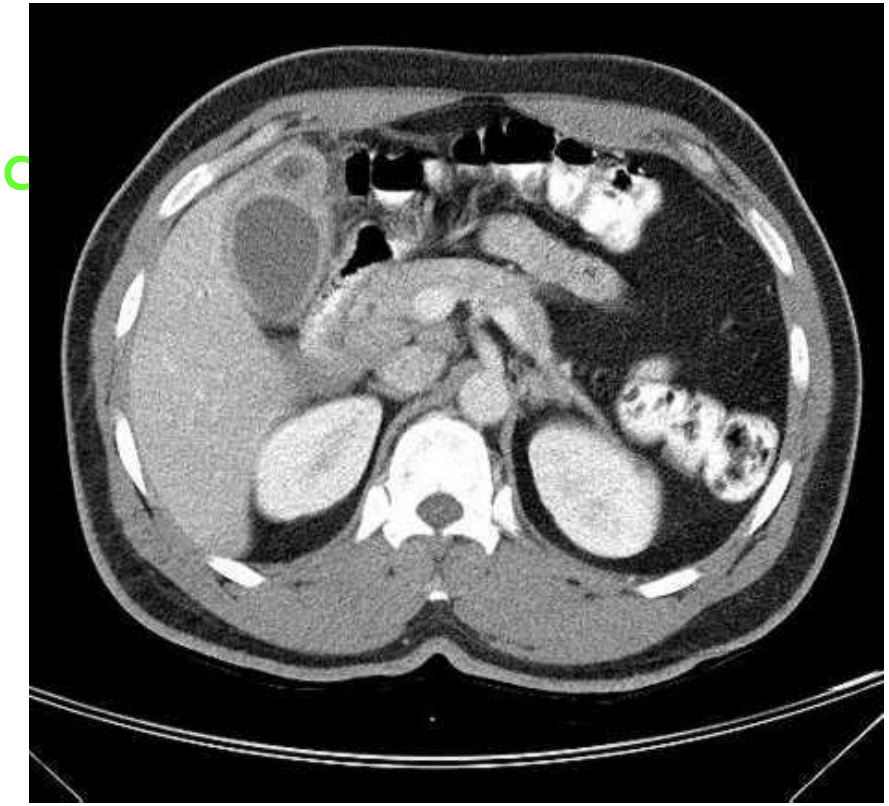
CT SCAN

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



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ERCP

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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.

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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.

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QUESTION

- What is your diagnosis ?

DIAGNOSIS

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

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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

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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

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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

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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

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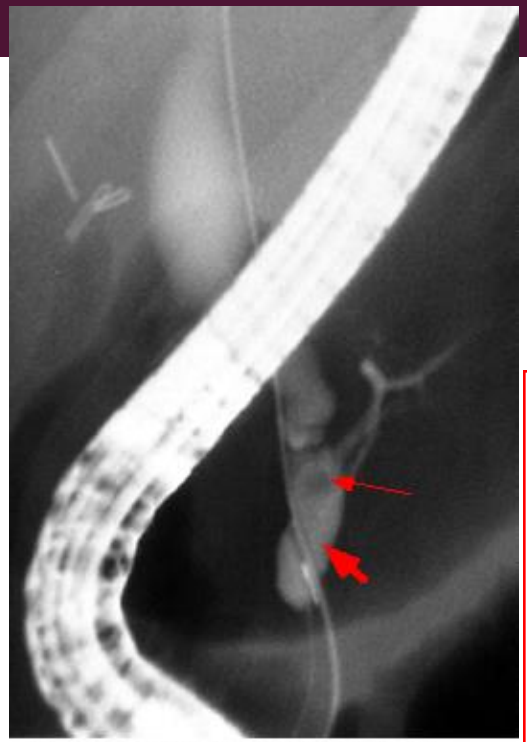
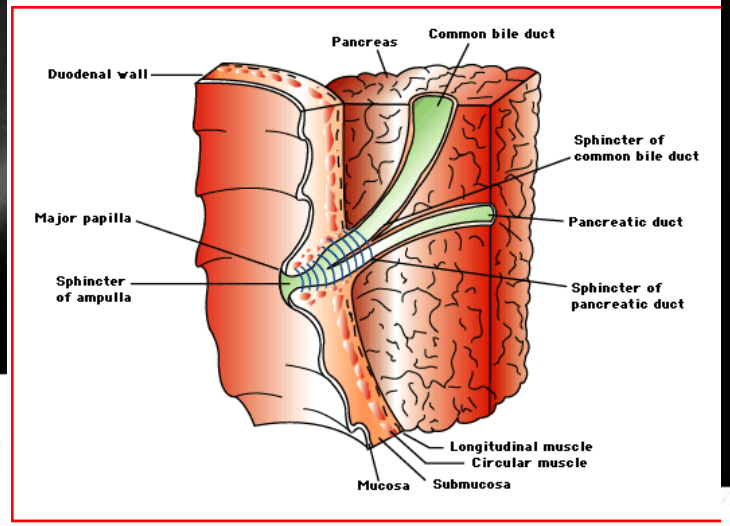


Anomalous pancreaticobiliary junction ERCP demonstrating an anomalous pancreaticobiliary junction in a child with recurrent abdominal pain and pancreatitis. Note the long, dilated common channel (thick arrow) containing a stone (thin arrow). The patient also has pancreas divisum. Courtesy of Mark Topazian,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

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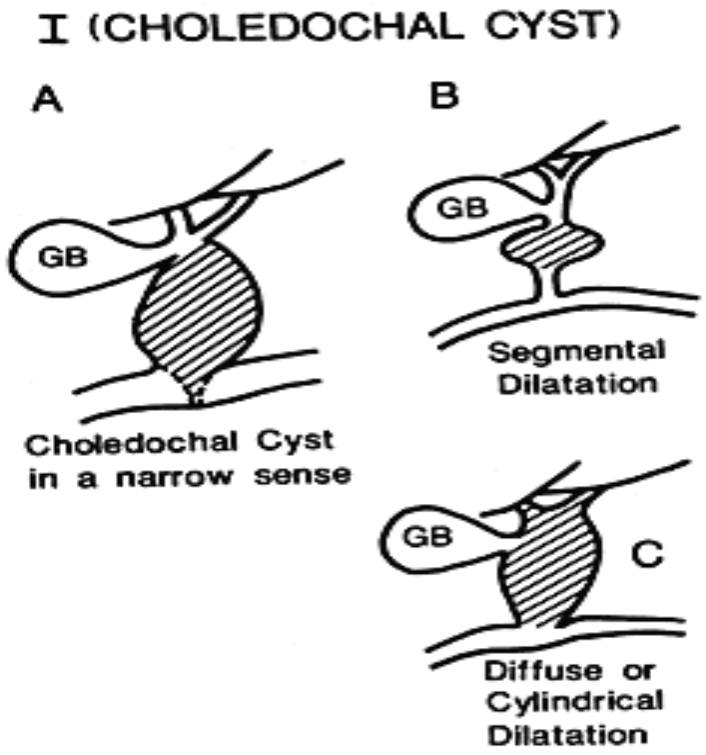
A classification scheme for cysts of the extrahepatic bile ducts (choledochocal cysts) was proposed initially in 1959

- 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



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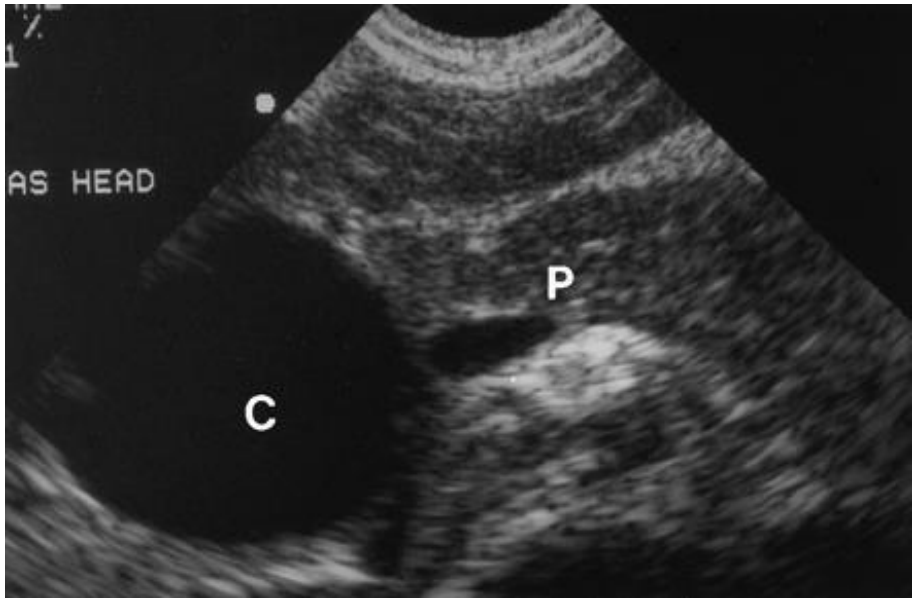
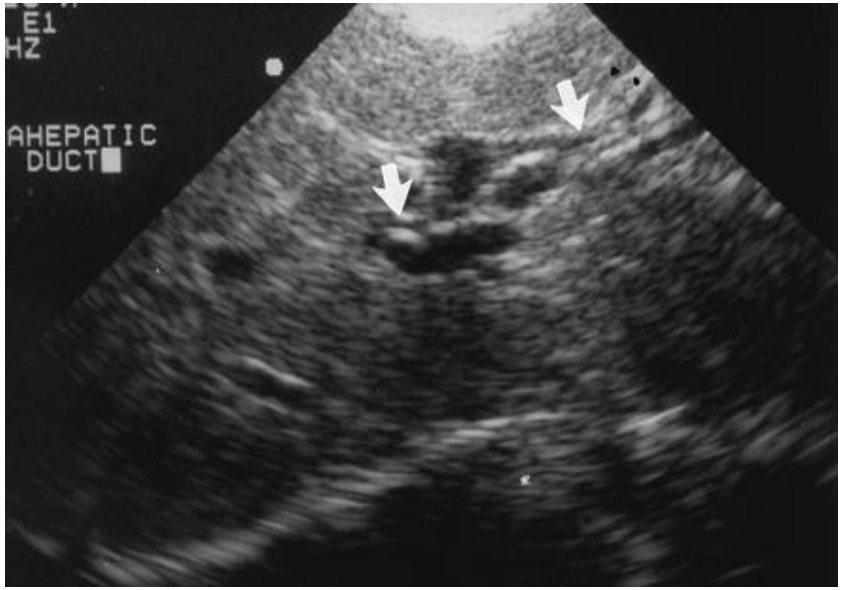
CHOLEDOCHAL CYST (TYPE IA)





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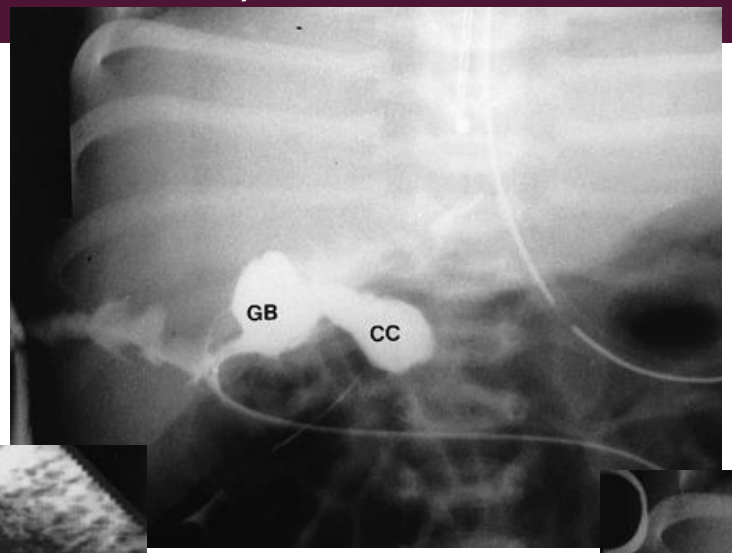
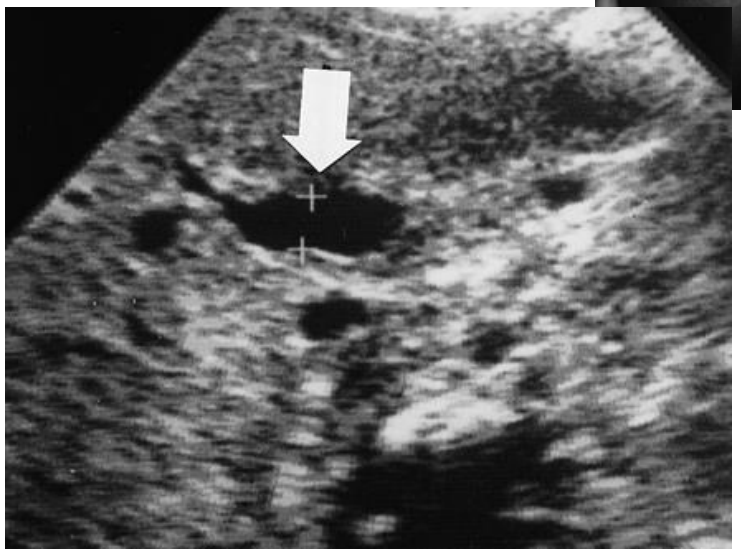
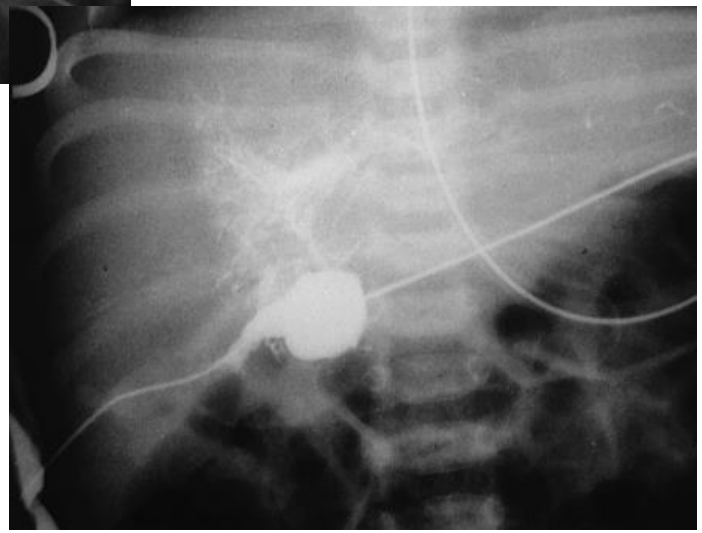
CHOLEDOCHAL CYST (TYPE IC)







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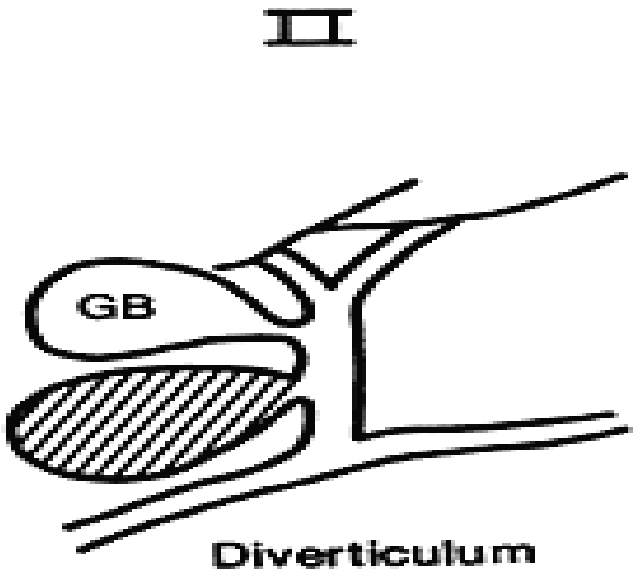
- Type II, the rarest biliary cyst, is a true diverticulum of the extrahepatic bile duct located proximal to the duodenum.



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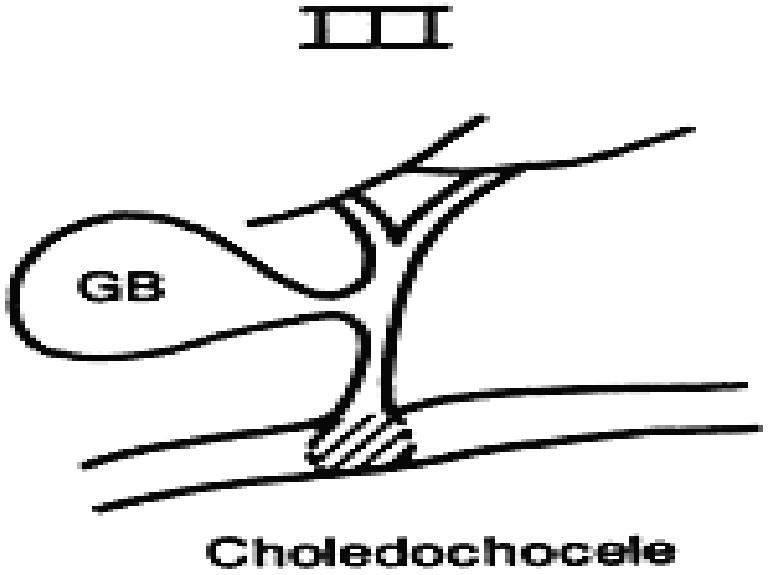
- Type III is a cystic dilatation limited to the intraduodenal portion of the distal common bile duct, also known as a choledochocele.



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TYPE III

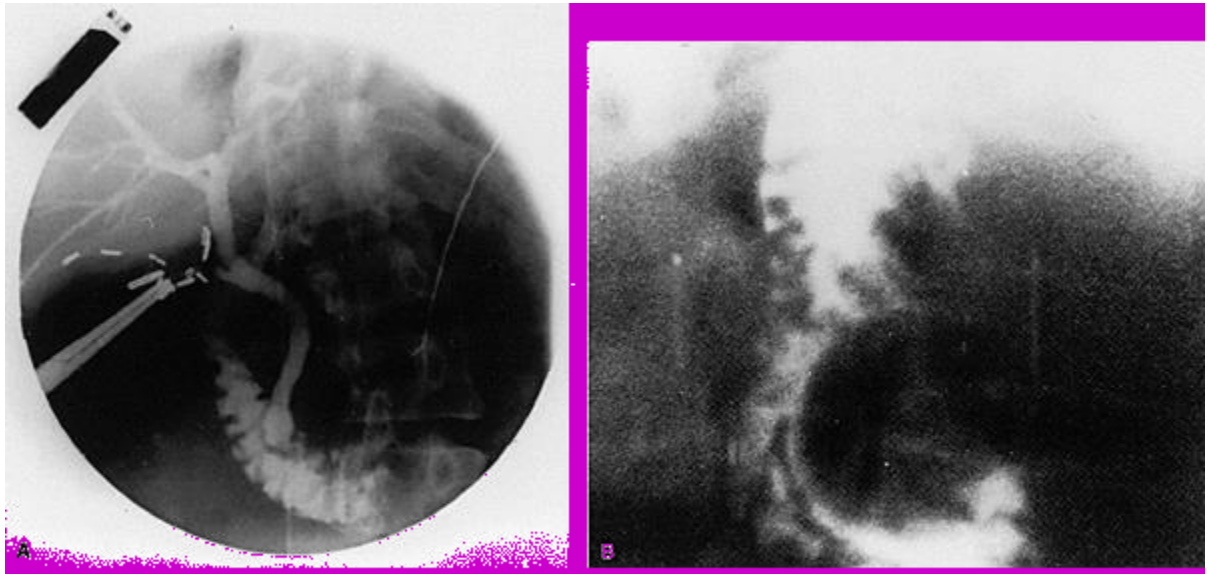

FIGURE 100-11. Choledochocele cyst—type III. A: Cholangiography reveals a club-shaped enlargement of the distal common bile duct bulging into the duodenum. B: Upper gastrointestinal series reveals a smoothly rounded filling defect in the second portion of the duodenum.

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CLASSIFICATION

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

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- 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 th trahepatic cysts without intrahepatic involvem
  - Type IVA is the sec on type of biliary cyst

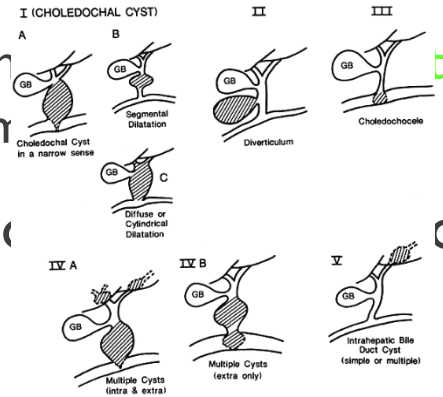

FIGURE 100-7. Todani's classification of biliary cysts based on location. Hatched areas represent cystic dilations. See text for details.

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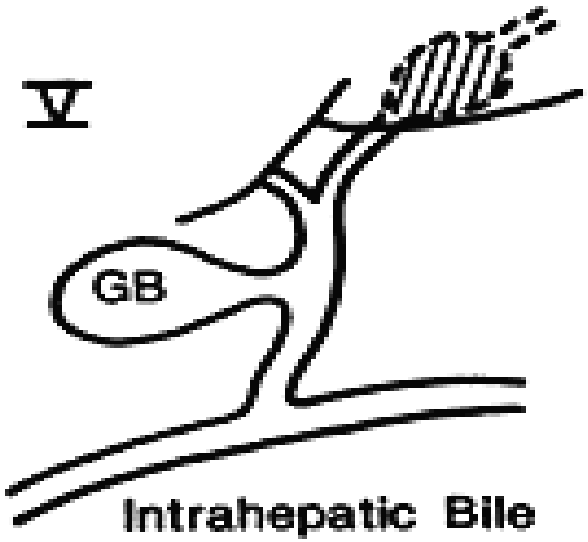
- Type V includes isolated or multiple cystic dilatations of the intrahepatic ducts without extrahepatic duct disease, also known as Caroli's disease.



Intrahepatic Bile Duct Cyst (simple or multiple)

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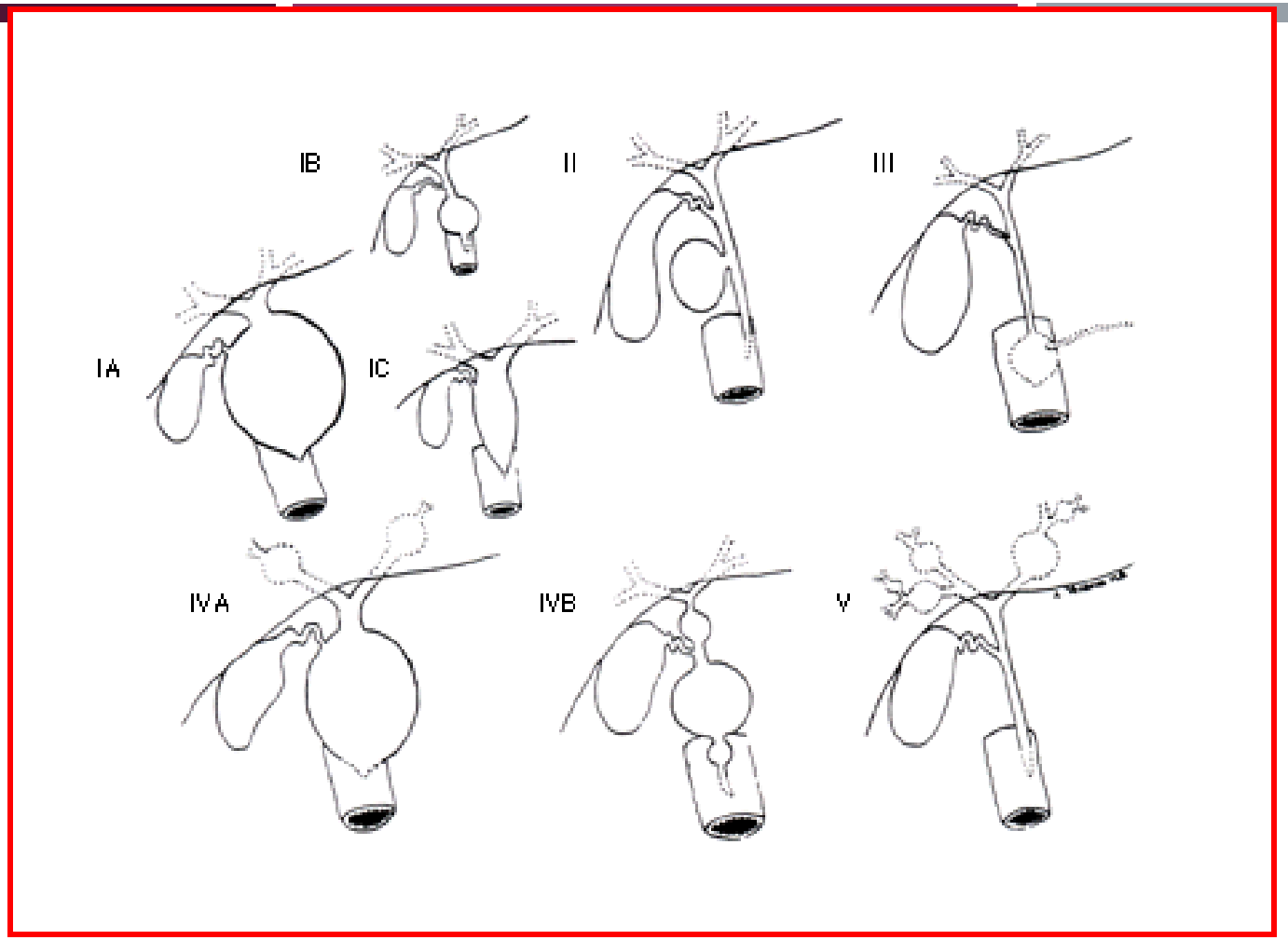
- Film



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 dilatations of the intrahepatic ducts. Reproduced with

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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

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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).

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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

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DIAGNOSIS

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

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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

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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.

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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

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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

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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

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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

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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

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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

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COMPLICATIONS

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

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DISCUSSION

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

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DISCUSSION

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

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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).

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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

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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

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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

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DISCUSSION

- Treatment is usually surgical

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

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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.

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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

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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

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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

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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

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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.

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PENTAX
THE END
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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.

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CASE HISTORY 2

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

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PRESENT ILNNESS(3)

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

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PHYSICAL EXAMINATION

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

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LAB DATA

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

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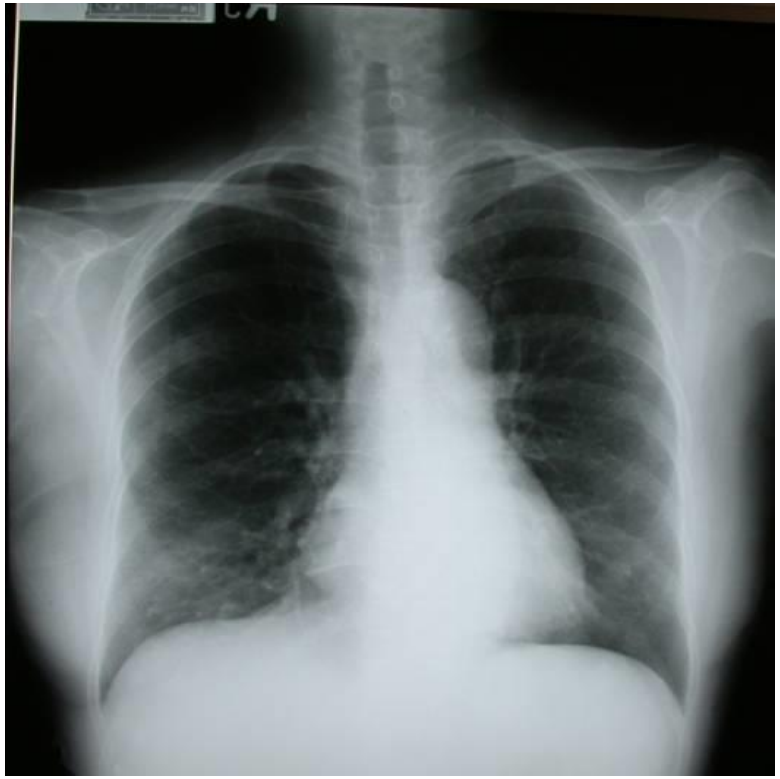
CXR: NORMAL



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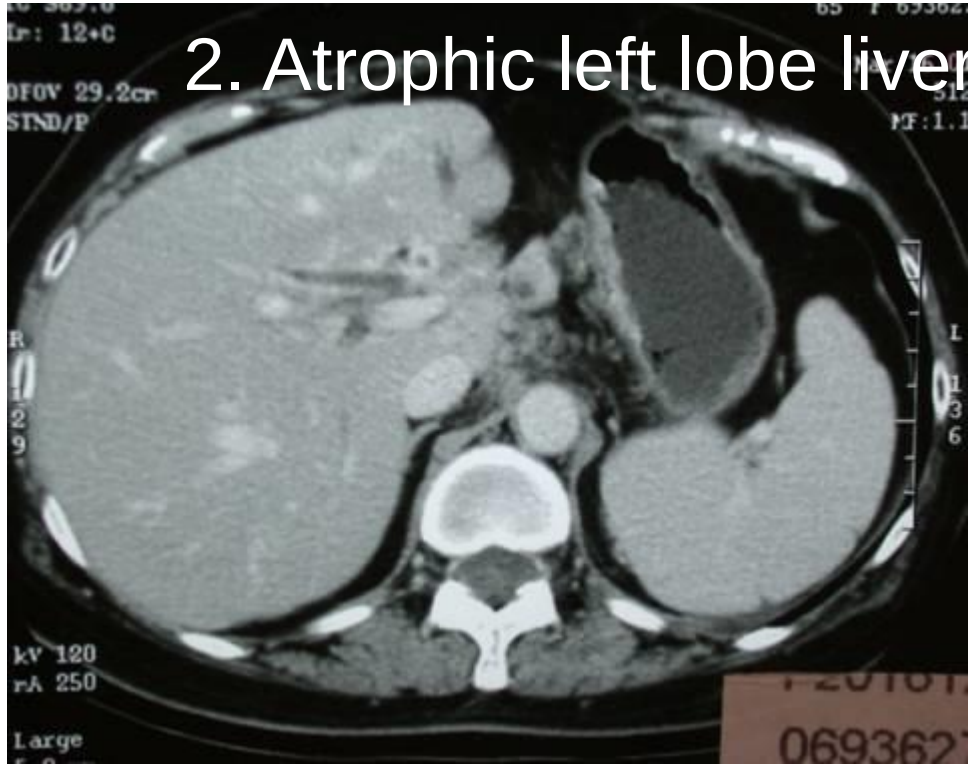
KUB: GALLSTONES NOTED

ABDOMINAL CT



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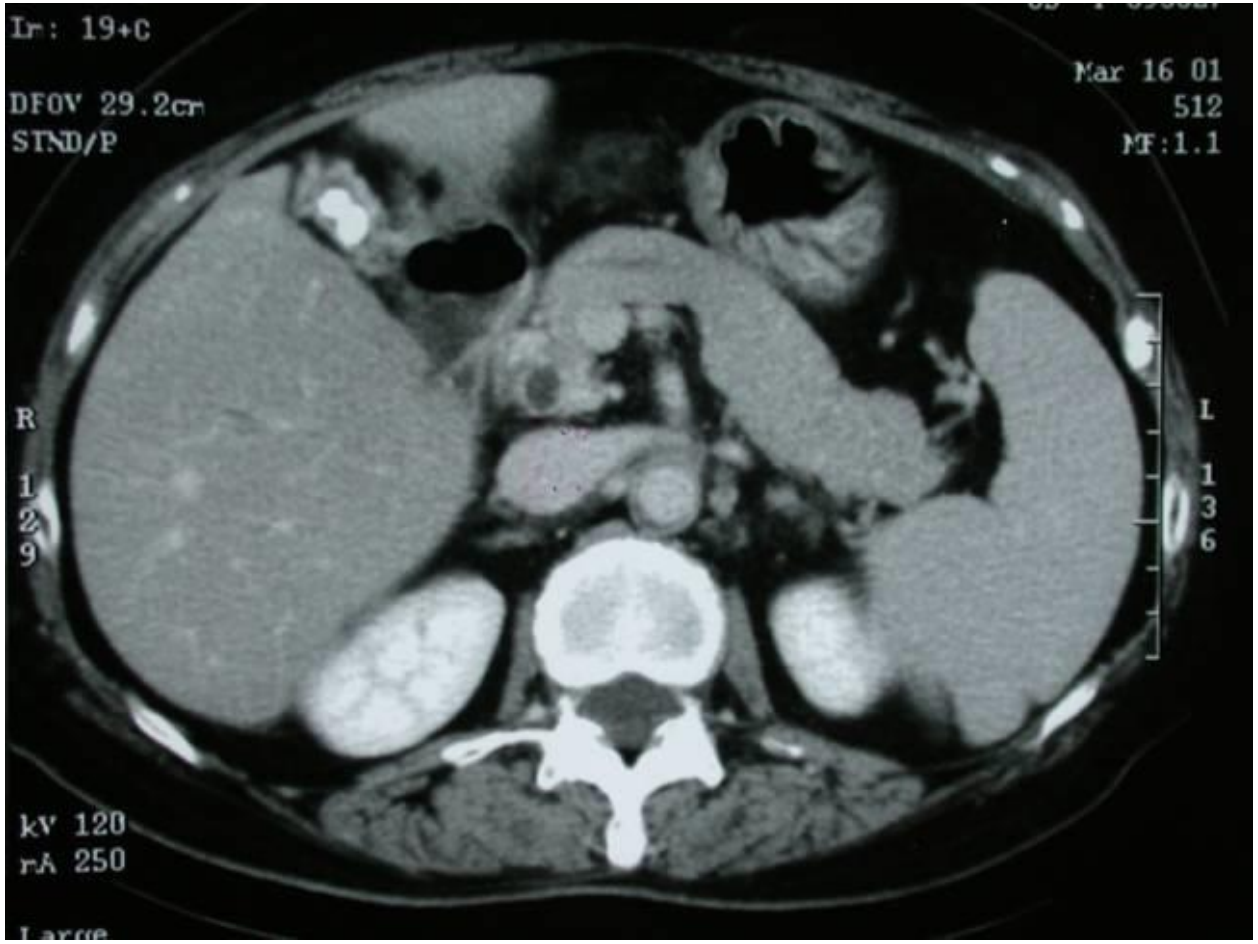
MULTIPLE GALLSTONES IN GALLBLADDER DILATED CBD NORMAL PANCREAS



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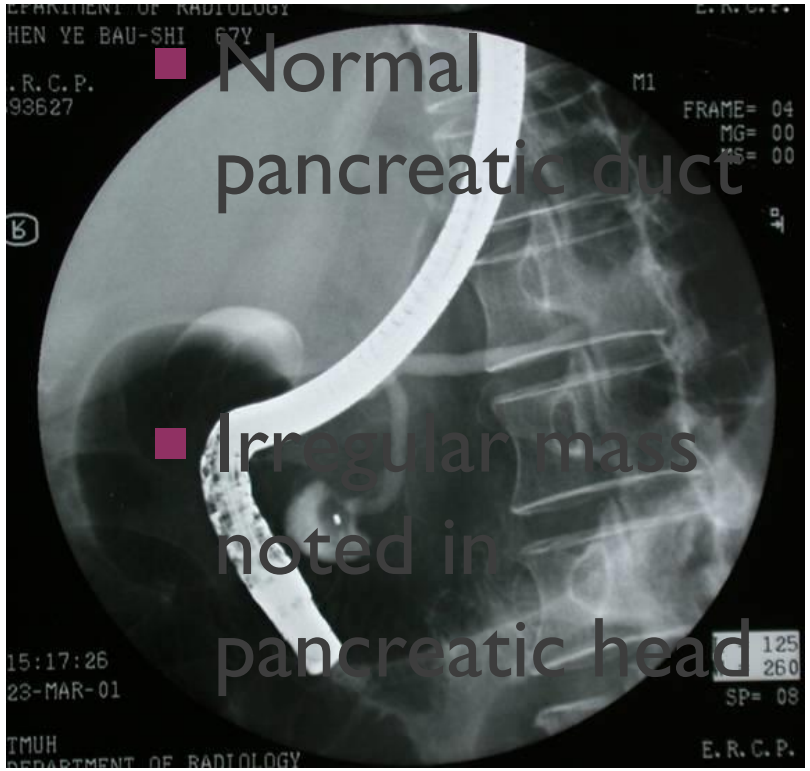
ERCP



- Normal pancreatic duct
- Irregular mass noted in pancreatic head

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MRCP

- Dilated right IHDs, CBD and pancreatic duct
- Stricture of the CHD

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BARIUM STUDY

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

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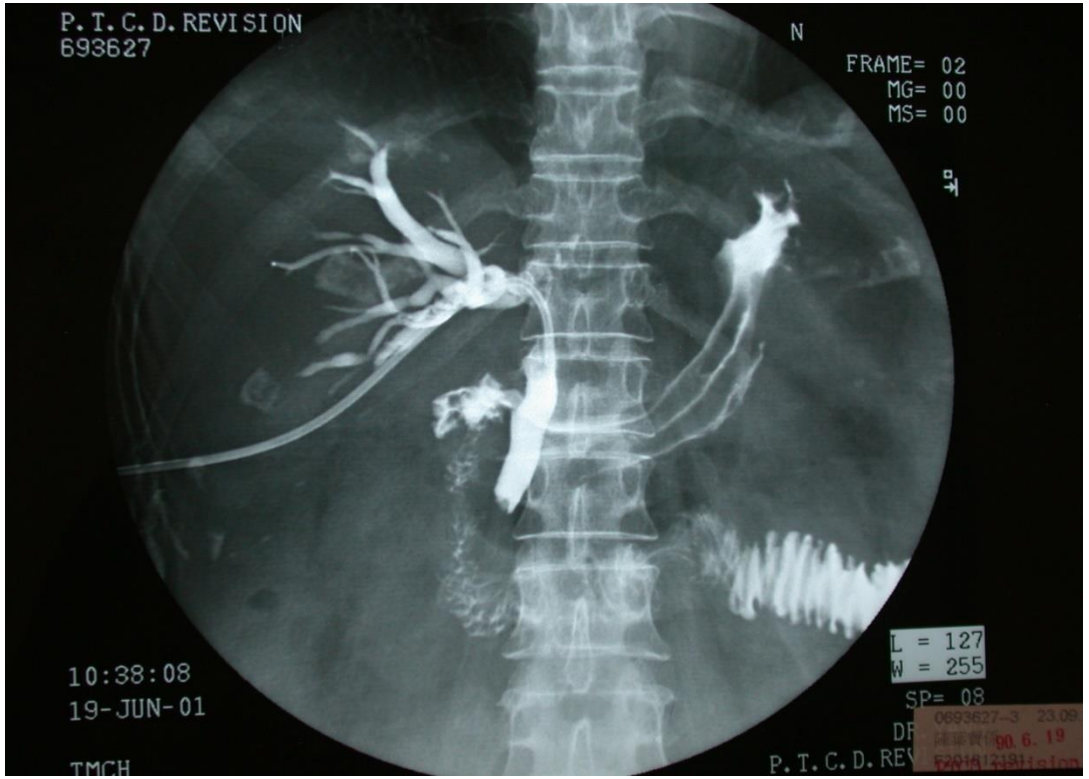
PTCD

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



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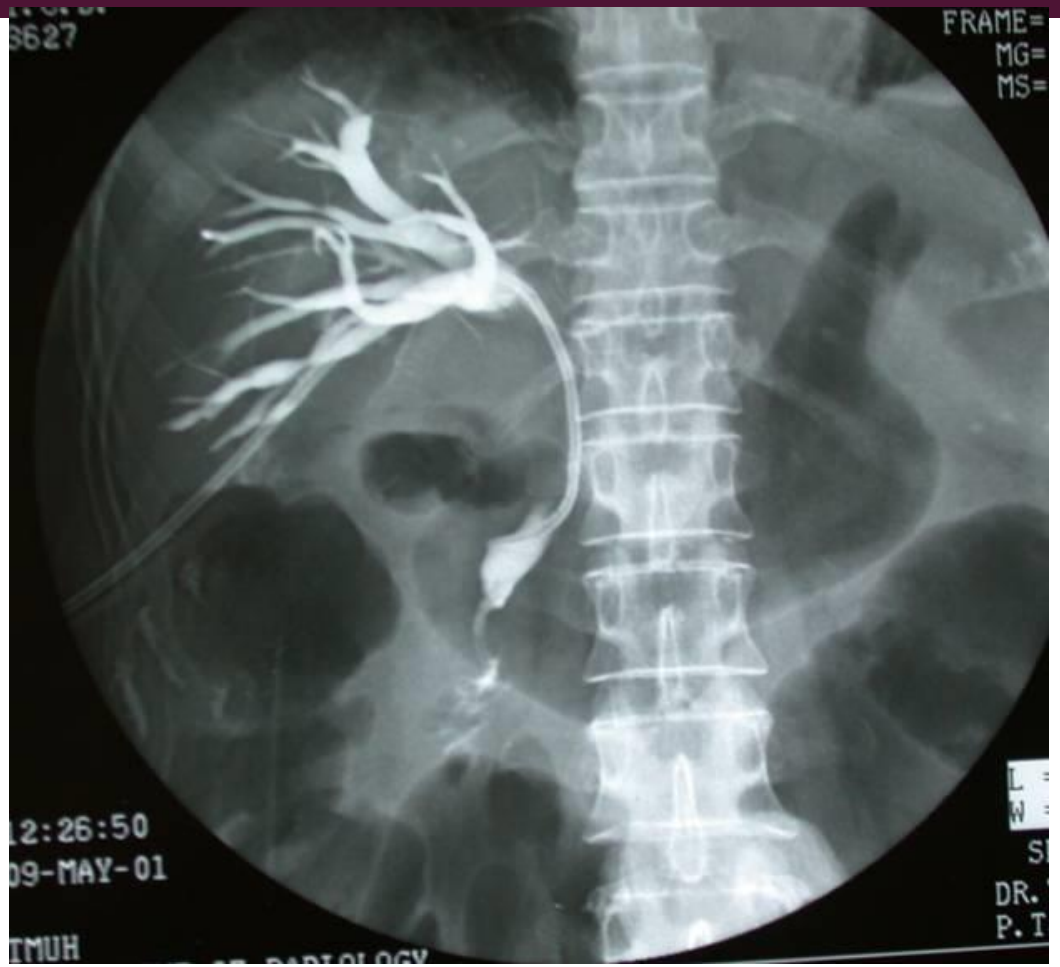
DILATATION OF THE RIGHT INTRAHEPATIC DUCTS STRICTURE OF THE COMMON BILE DUCT.



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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

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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

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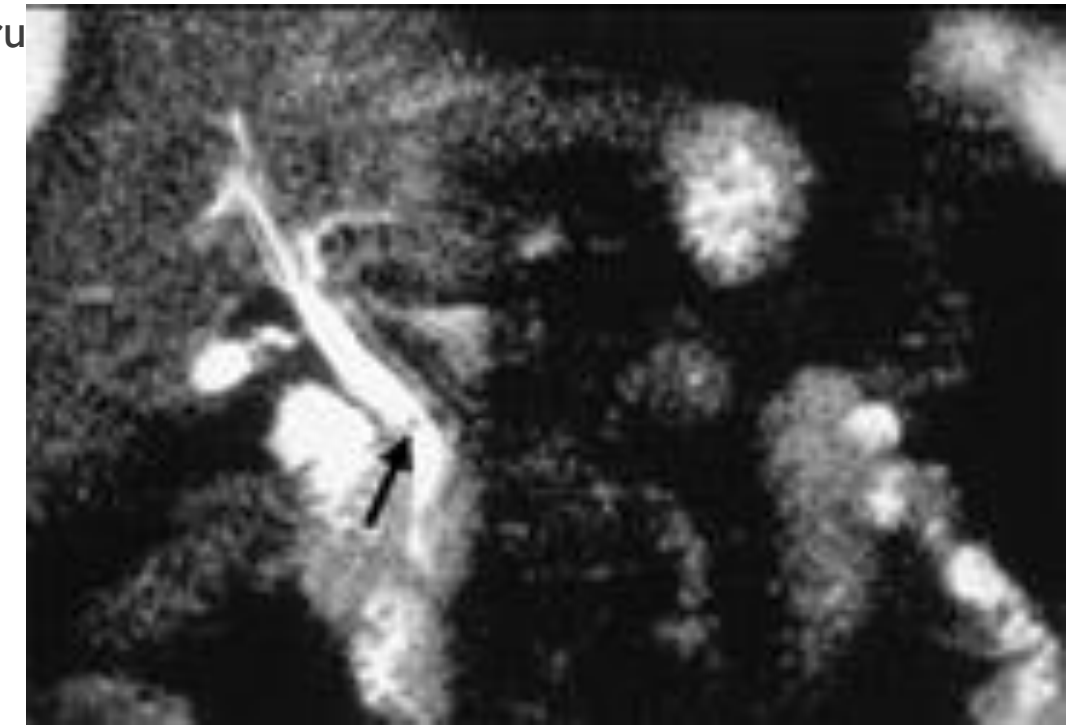
CHOLEDOCHOLITHIASIS

- Choledocholithiasis—20% of obstru



- PTC and ERCP are the most efficacious examination
- Most patients have gallstones in gallbladder

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CHOLEDOCHAL CYST (UNCOMMON CONGENITAL DISORDER)

- Slight
  proxir
  of obs
- Narro
- Abrup

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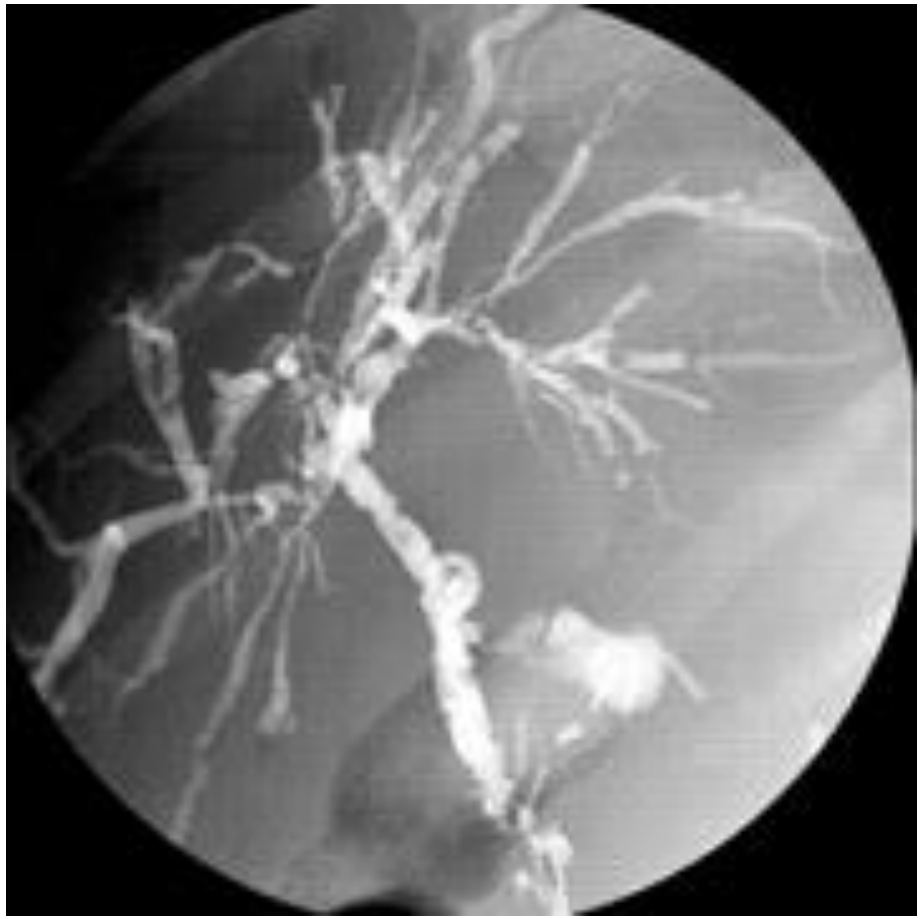
SCLEROSING CHOLANGITIS

- 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



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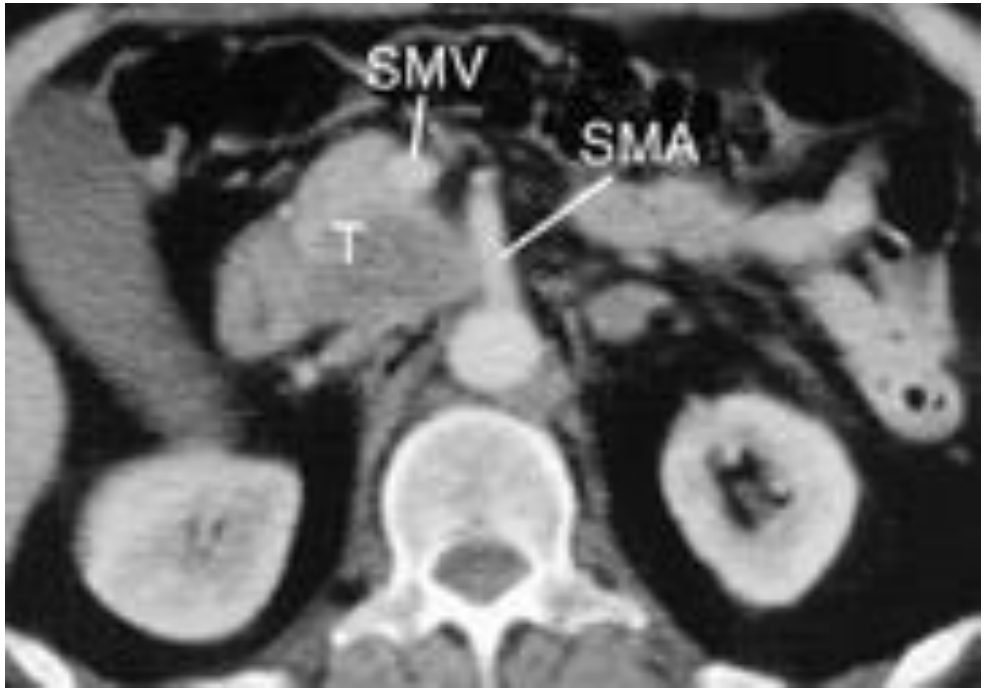
AMPULLARY AND PANCREATIC TUMOR

- 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)

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CHOLANGIOCARCINOMA

- *Peripheral cholangiocarcinoma*
  - Present as an intrahepatic hypodence 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

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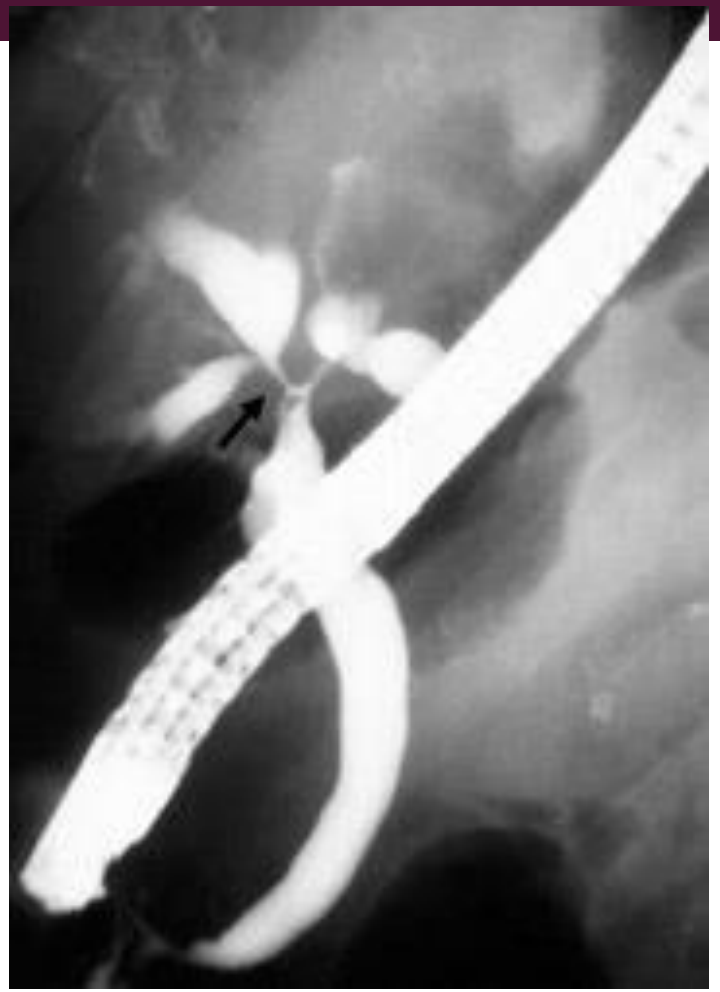
CHOLANGIOCARCINOMA



- 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.

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SURGICAL INTERVENTION

- Cholecystectomy + tumor biopsy
- Pathology: adenocarcinoma, hilum

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DISCUSSION

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

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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

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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

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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

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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.

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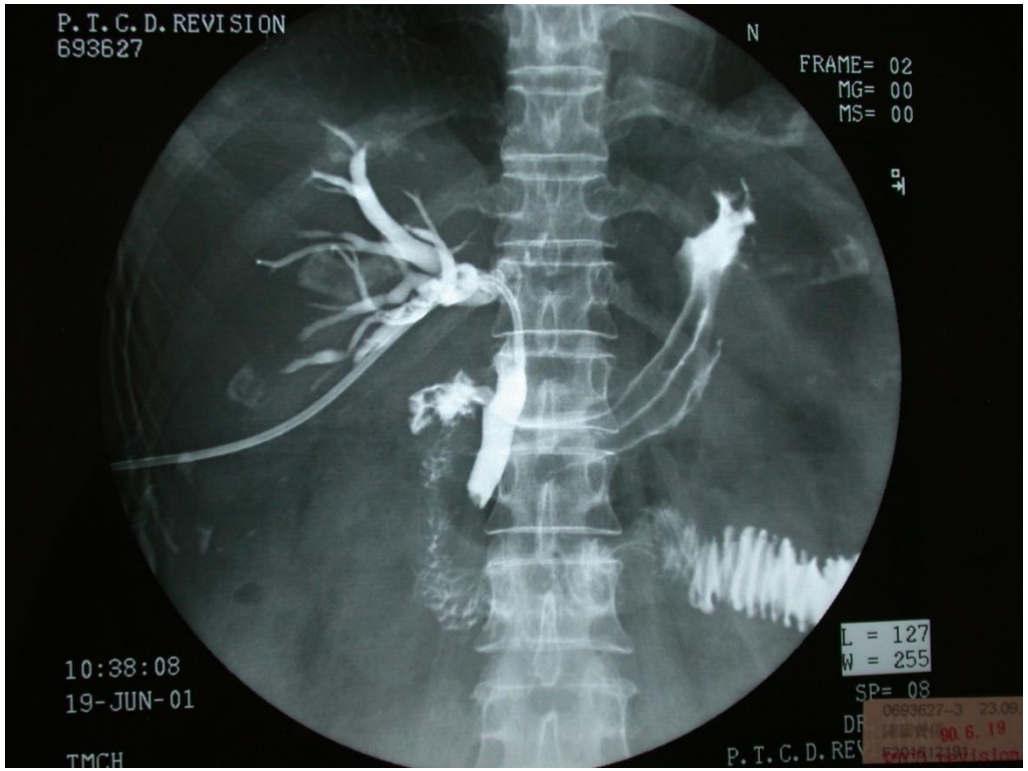
- Until recently cholangiography either by ERCP or PTCD available to display correctly the full extent of CCC with an accuracy varying between 89 and 96%.



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- 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.

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