

Biliary Tract Disease

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Overview

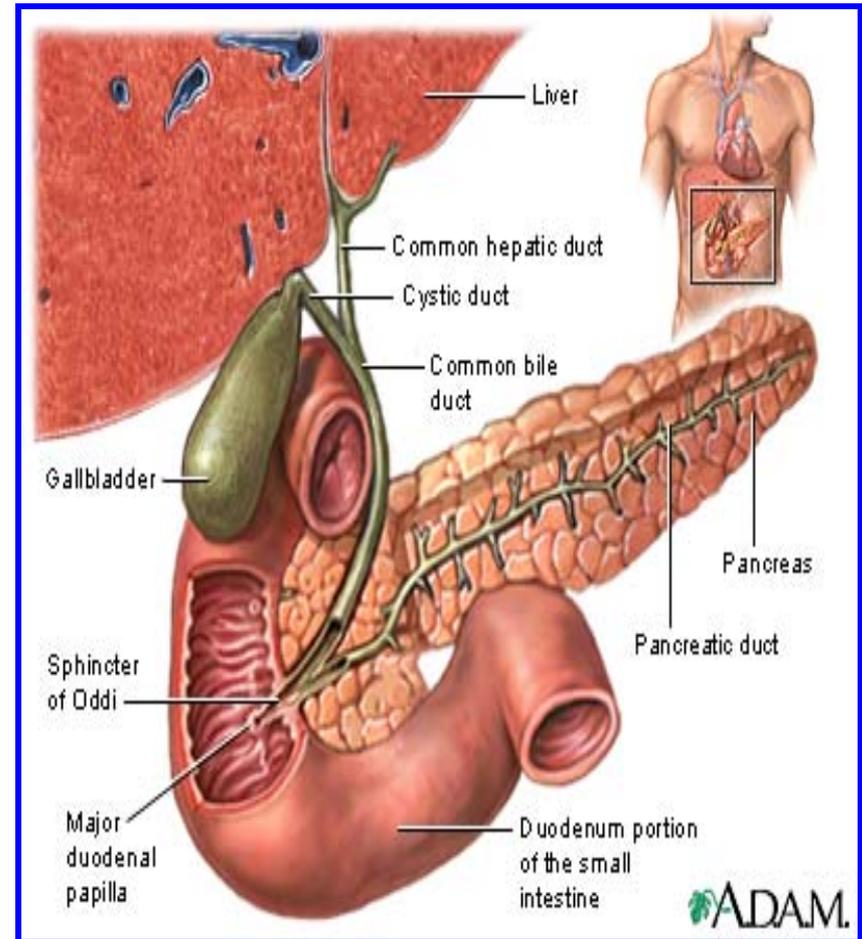
- Gallstones
- Biliary tract tumours
- Other conditions
 - Acute acalculous cholecystitis
 - Mirizzi's syndrome
 - Primary Biliary Cirrhosis
 - Primary Sclerosing Cholangitis
 - Biliary tract cysts
- Biliary strictures

Biliary Tract

Part of the digestive system.

Made up of:

- Intra hepatic ducts
- Extra hepatic ducts
- Gallbladder
- Common Bile Duct

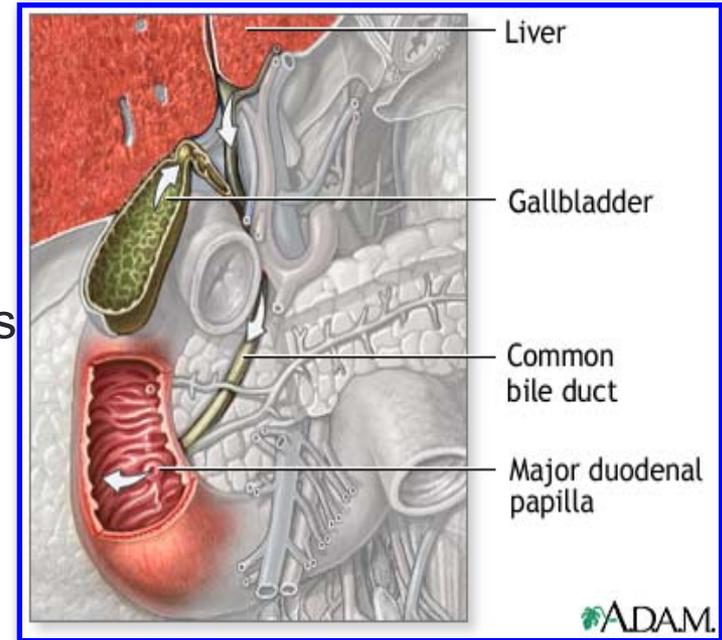


The Gallbladder

The gallbladder concentrates and stores bile.

Bile:

- Secreted by the liver
- Contains cholesterol, bile pigments and phospholipids
- Flows from the liver, through the hepatic ducts into the gallbladder
- Exits the gallbladder via the cystic duct
- Flows from the cystic duct into the common bile duct, into the small intestine
- In the small intestine, aids digestion by breaking down fatty foods and fat-soluble vitamins



Gallstones – Pathophysiology

- Cholesterol, ordinarily insoluble in water, comes into solution by forming vesicles with phospholipids
- If ratio of cholesterol, phospholipids, and bile salts altered, cholesterol crystals may form
- Gallstone formation involves a variety of factors:
 - Cholesterol supersaturation
 - Mucin hypersecretion by the gallbladder mucosa creates a viscoelastic gel that fosters nucleation.
 - Bile stasis
 - Occurs in diabetes, pregnancy, oral contraceptive use, and prolonged fasting in critically ill patients on total parenteral nutrition.

Gallstones – Frequency

- Gallstone disease is one of the most common and costly of all digestive diseases
- 9% of those > 60 years
- In USA, 6.3 million men and 14.2 million women aged 20-74 years have gallbladder disease
- Incidence of gallstones is 1 million new cases per year
- Prevalence is 20 million cases in USA

Gallstones



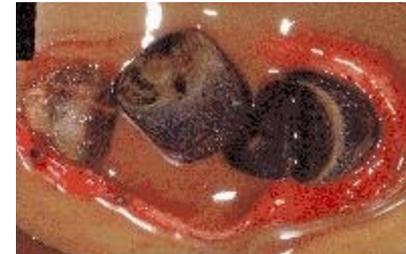
Sex

- Higher among females than males (lifetime risk of 35% vs 20%, respectively)
 - Due to endogenous sex hormones (enhance cholesterol secretion and increase bile cholesterol saturation)
 - Progesterone may contribute by relaxing smooth muscle and impairing gallbladder emptying.

Age

- Increased age is associated with lithogenic bile and increased rate of gallstones

Gallstones – Types



- Two main types:
 - Cholesterol stones (85%):
 - 2 subtypes—pure (90-100% cholesterol) or mixed (50-90% cholesterol).
 - Pure stones often are solitary, whitish, and larger than 2.5 cm in diameter.
 - Mixed stones usually are smaller, multiple in number, and occur in various shapes and colors.
 - Pigment stones (15%) occur in 2 subtypes—brown and black.
 - Brown stones are made up of calcium bilirubinate and calcium-soaps. Bacteria involved in formation via secretion of beta glucuronidase and phospholipase
 - Black stones result when excess bilirubin enters the bile and polymerizes into calcium bilirubinate (patients with chronic hemolysis)

Gallstones – Natural History



- 80% of patients, gallstones are clinically silent
- 20% of patients develop symptoms over 15-20 years
- About 1% per year
- Almost all become symptomatic before complications develop
- Biliary-type pain due to obstruction of the bile duct lumen
- Predictive value of other complaints (eg, intolerance to fatty food, indigestion) too low to be clinically helpful

Gallstones – Diverse symptoms

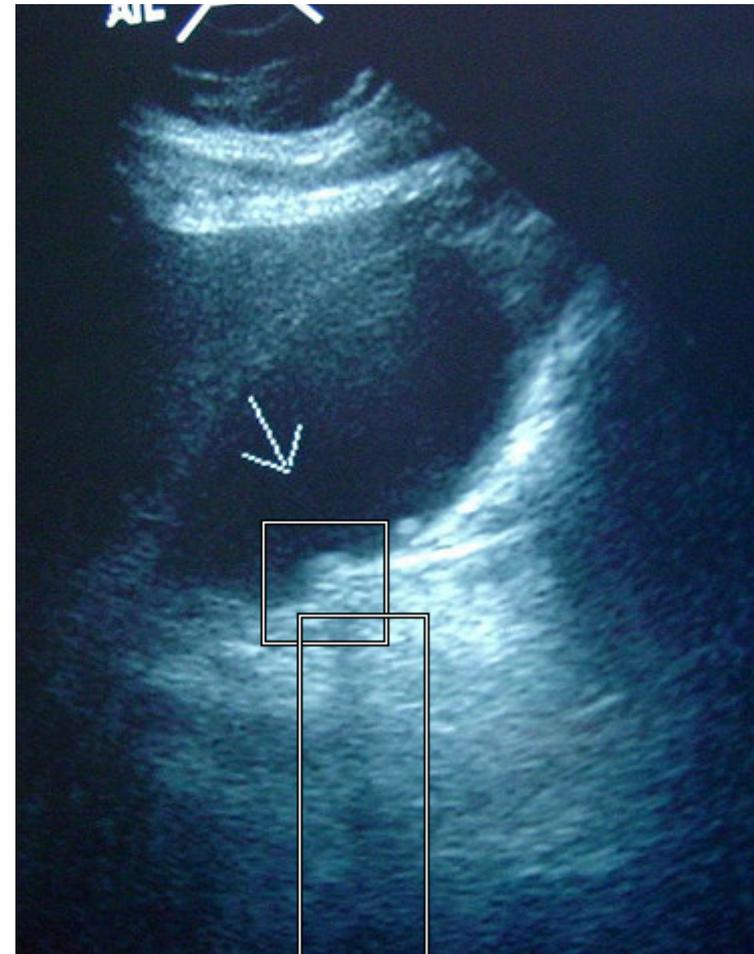
- Abdominal pain
 - Aching or tightness, typically severe and located in the epigastrium
 - May develop suddenly, last for 15 minutes to several hours, and then resolve suddenly
- Referred pain – posterior scapula or right shoulder area
- Nausea and vomiting
- Jaundice
- Pruritus:
 - Itching, typically worse at night.
- Fatigue
- Weight loss
- Miscellaneous:
 - Fatty food intolerance
 - Gas
 - Bloating
 - Dyspepsia

Complications of Gallstones

- In the gallbladder
 - Biliary colic
 - Acute and chronic cholecystitis
 - Empyema
 - Mucocoele
 - Carcinoma
- In the bile ducts
 - Obstructive jaundice
 - Pancreatitis
 - Cholangitis
- In the Gut
 - Gallstone ileus

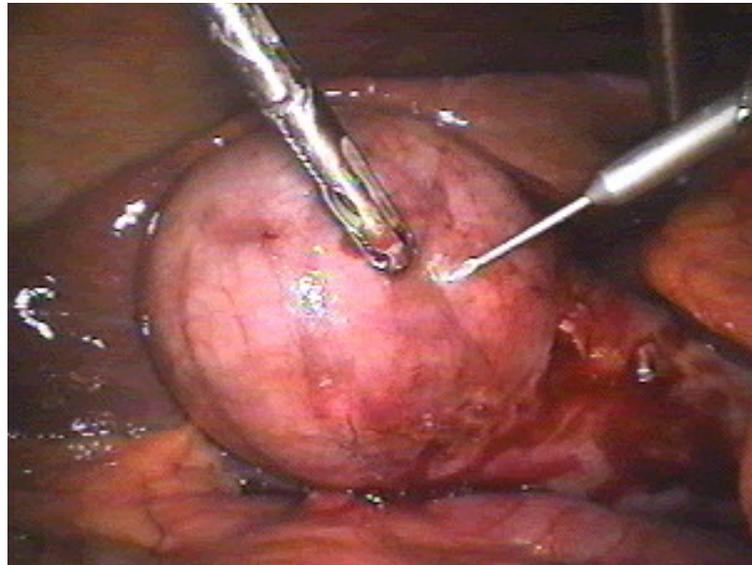
Biliary Colic

- Symptoms
 - Right upper quadrant pain
- Signs
 - Usually none
- Investigations
 - Bloods – U&E, FBC, LFT, Amylase, CRP
 - Ultrasound of abdomen
 - OGD (Oesophagogastroduodenoscopy)
- Treatment
 - Analgesia
 - Cholecystectomy



Acute Calculous Cholecystitis

- Inflammation of the gallbladder that develops in the setting of an obstructed cystic or bile duct
- Most patients have complete remission within 1-4 days.
- 25-30% of patients either require surgery or develop some complication
- Perforation occurs in 10-15% of cases.



Acute Calculous Cholecystitis

- Symptoms
 - Right upper quadrant pain – continuous, longer duration
- Signs
 - Fever, Local peritonism.
 - Murphy's sign
 - 2 fingers on RUQ, ask patient to breathe in. Positive if pain and arrest of inspiration
- Investigations
 - Bloods – U&E, FBC, LFT, Amylase, CRP
 - Ultrasound of abdomen
 - Thickened gallbladder wall, pericholecystic fluid and stones
 - OGD (Oesophagogastroduodenoscopy)
- Treatment
 - Nil by mouth
 - Analgesia
 - Intravenous antibiotics
 - Cholecystectomy

Empyema / Mucocoele

- Empyema refers to a gallbladder filled with pus due to acute cholecystitis
- Mucocoele refers to an overdistended gallbladder filled with mucoid or clear and watery content.



Empyema / Mucocoele

- Symptoms
 - Right upper quadrant pain – continuous, longer duration
- Signs
 - Fever, Local peritonism.
 - Murphy's sign
 - 2 fingers on RUQ, ask patient to breathe in. Positive if pain and arrest of inspiration
- Investigations
 - Bloods – U&E, FBC, LFT, Amylase, CRP
 - Ultrasound of abdomen
 - Thickened gallbladder wall, distended gallbladder, pericholecystic fluid, stones
- Treatment
 - Nil by mouth
 - Analgesia
 - Intravenous antibiotics
 - Cholecystectomy



Obstructive Jaundice

- Blockage of the biliary tree by gallstones
- Symptoms
 - Pain, Jaundice, dark urine, pale stools
- Signs
 - Jaundice.
- Investigations
 - Bloods – U&E, FBC, LFT, Amylase, CRP, Hepatitis screen, Coagulation screen
 - Ultrasound of abdomen
- Treatment
 - Endoscopic Retrograde CholangioPancreatogram



Ascending Cholangitis

- Obstruction of biliary tree with bile duct infection
- Symptoms
 - Unwell, pain, jaundice, dark urine, pale stools
 - Charcot triad (ie, fever, right upper quadrant pain, jaundice) occurs in only 20-70% of cases
- Signs
 - Sepsis (Fever, tachycardia, low BP), Jaundice.
- Investigations
 - Bloods – U&E, FBC, LFT, Amylase, CRP, Coagulation screen
 - Ultrasound of abdomen
- Treatment
 - Intravenous antibiotics
 - Endoscopic Retrograde CholangioPancreatogram

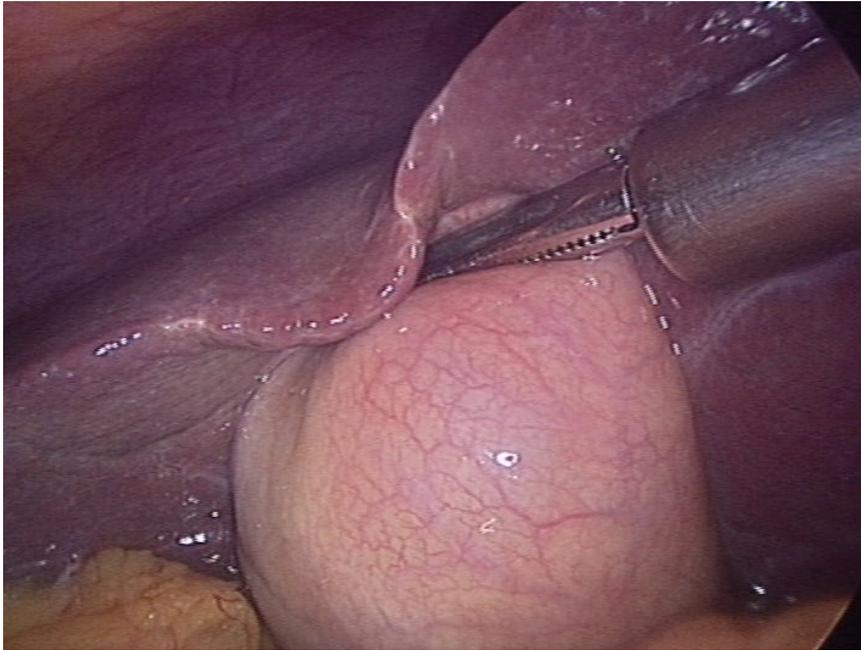
Acute Pancreatitis

- Acute inflammation of pancreas and other retroperitoneal tissues.
- Symptoms
 - Severe central abdominal pain radiating to back, vomiting
- Signs
 - Variable – None to Sepsis (Fever, tachycardia, low BP), Jaundice, acute abdomen
- Investigations
 - Bloods – U&E, FBC, LFT, Amylase, CRP
 - Ultrasound of abdomen
 - MRCP
 - CT Pancreas
- Treatment
 - Supportive
 - Endoscopic Retrograde CholangioPancreatogram

Gallstone ileus

- Obstruction of the small bowel by a large gallstone
 - A stone ulcerates through the gallbladder into the duodenum and causes obstruction at the terminal ileum
- Symptoms
 - Small bowel obstruction (vomiting, abdominal pain, distension, nil pr)
- Signs
 - Abdominal distension, obstructive bowel sounds.
- Investigations
 - Bloods – U&E, FBC, LFT, Amylase, CRP, Hepatitis screen, Coagulation screen
 - Plain film of abdomen – Air in CBD, small bowel fluid levels and stone
- Treatment
 - Laparotomy and removal of stone from small bowel.

Cholecystectomy



- Laparoscopic cholecystectomy standard of care
- Timing
 - Early vs interval operation
- Patient consent
 - Conversion to open procedure 10%
 - Bleeding
 - Bile duct injury
 - Damage to other organs

Mirizzi Syndrome

- Refers to common hepatic duct obstruction caused by an extrinsic compression from an impacted stone in the cystic duct
- Estimated to occur in 0.7-1.4% of all cholecystectomies
- Often not recognized preoperatively, which can lead to significant morbidity and biliary injury, particularly with laparoscopic surgery.

Acute Acalculous Cholecystitis

- Presence of an inflamed gallbladder in the absence of an obstructed cystic or common bile duct
- Typically occurs in the setting of a critically ill patient (eg, severe burns, multiple traumas, lengthy postoperative care, prolonged intensive care)
- Accounts for 5% of cholecystectomies
- Aetiology is thought to have ischemic basis, and gangrenous gallbladder may result
- Increased rate of complications and mortality
- An uncommon subtype known as acute emphysematous cholecystitis generally is caused by infection with clostridial organisms and occlusion of the cystic artery associated with atherosclerotic vascular disease and, often, diabetes.

Primary Sclerosing Cholangitis

- Chronic cholestatic biliary disease characterized by non-suppurative inflammation and fibrosis of the biliary ductal system
- Cause is unknown but is associated with autoimmune inflammatory diseases, such as chronic ulcerative colitis and Crohn colitis, and rare conditions, such as Riedel thyroiditis and retroperitoneal fibrosis
- Most patients present with fatigue and pruritus and, occasionally, jaundice
- Natural history is variable but involves progressive destruction of the bile ducts, leading to cirrhosis and liver failure
- Clinical features of cholangitis (ie, fever, right upper quadrant pain, jaundice) are uncommon unless the biliary system has been instrumented.

Primary Sclerosing Cholangitis

Medical Care

- Chronic progressive disease with no curative medical therapy
- Goals of medical management are to treat the symptoms and to prevent or treat the known complications
- Liver transplantation is the only effective therapy and is indicated in end-stage liver disease.

Surgical Care

- Indications for liver transplantation include variceal bleed or portal gastropathy, intractable ascites, recurrent cholangitis, progressive muscle wasting, and hepatic encephalopathy.
- Recurs in 15-20% of patients after transplantation.

Primary Biliary Cirrhosis

- Progressive cholestatic biliary disease that presents with fatigue and itching or asymptomatic elevation of the alkaline phosphatase.
- Jaundice develops with progressive destruction of bile ductules that eventually leads to liver cirrhosis and hepatic failure.
- Autoimmune illness has a familial predisposition
- Antimitochondrial antibodies (AMA) are present in 95% of patients
- Goals of treatment are to slow the progression rate of the disease and to alleviate the symptoms (eg, pruritus, osteoporosis, sicca syndrome)
- Liver transplantation appears to be the only life-saving procedure.

Biliary Tract Cysts

- Choledochal cysts
- Consist of cystic dilatations of the extra-hepatic biliary tree
- Uncommon abnormality
- 50% present with combination of jaundice, abdominal pain, and an abdominal mass.
- ? Due to anomalous union of the pancreatic and biliary ductal system.
- Classified into 5 types
- Treatment for choledochal cysts is surgical (excision of the cyst with construction).



Biliary Tract Tumours

Cholangiocarcinoma

Cancer of the Gall Bladder

Biliary Tree Neoplasms

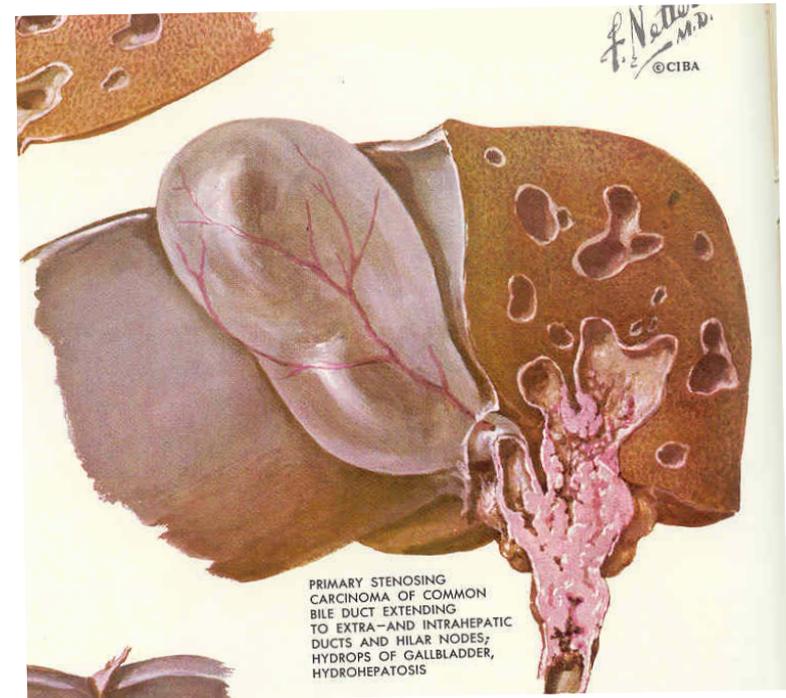
- Clinical symptoms:
 - Weight loss (77%)
 - Nausea (60%)
 - Anorexia (56%)
 - Abdominal pain (56%)
 - Fatigue (63%)
 - Pruritus (51%)
 - Fever (21%)
 - Malaise (19%)
 - Diarrhea (19%)
 - Constipation (16%)
 - Abdominal fullness (16%).
- Symptomatic patients usually have advanced disease, with spread to hilar lymph nodes before obstructive jaundice occurs
- Associated with a poor prognosis.

Cholangiocarcinoma

- Adenocarcinoma of the bile ducts
- May occur without associated risk factors
- Associated with chronic cholestatic liver disease such as:
 - Primary Sclerosing Cholangitis
 - Choledochal cysts
 - Asbestos.
- Accounts for 25% of biliary tract cancers
- Presentation:
 - Jaundice
 - Vague upper or right upper quadrant abdominal pain
 - Anorexia, weight loss
 - Pruritus.

Cholangiocarcinoma

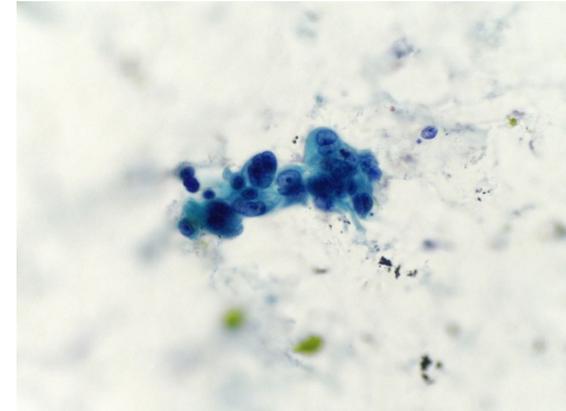
- Slow growing malignancy of biliary tract which tend to infiltrate locally and metastasize late.
- Gall Bladder cancer = 6,900/yr
- Bile duct cancer = 3,000/yr
- Hepatocellular Ca = 15,000/yr



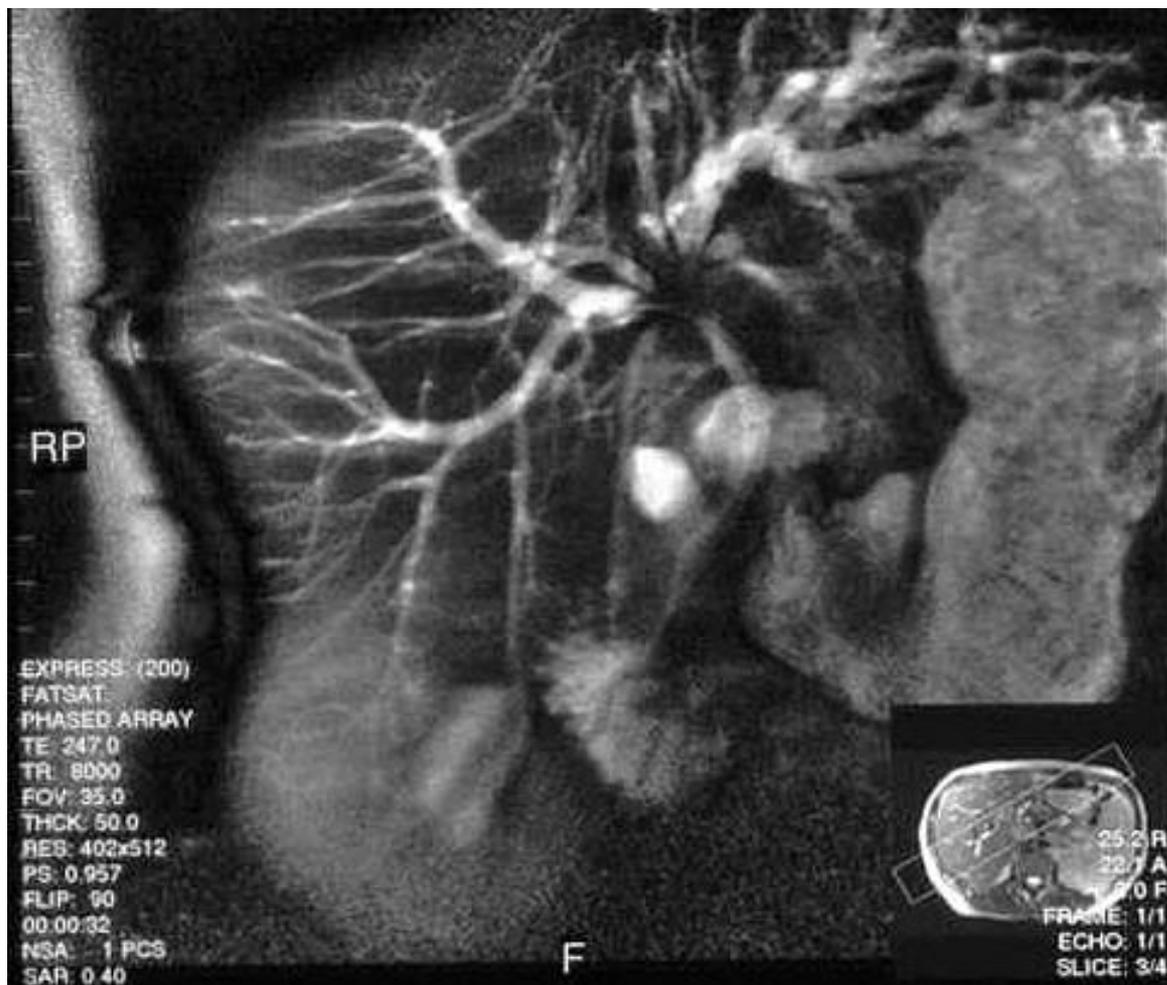
Cholangiocarcinoma

Diagnosis and Initial Workup

- Jaundice
- Weight loss, anorexia, abdominal pain, fever
- US – bile duct dilatation
- Quadruple phase CT
- MRCP/MRI
- ERCP with Stent and Brush Biopsy
- Percutaneous Cholangiogram with Internal Stent and Brush Biopsy

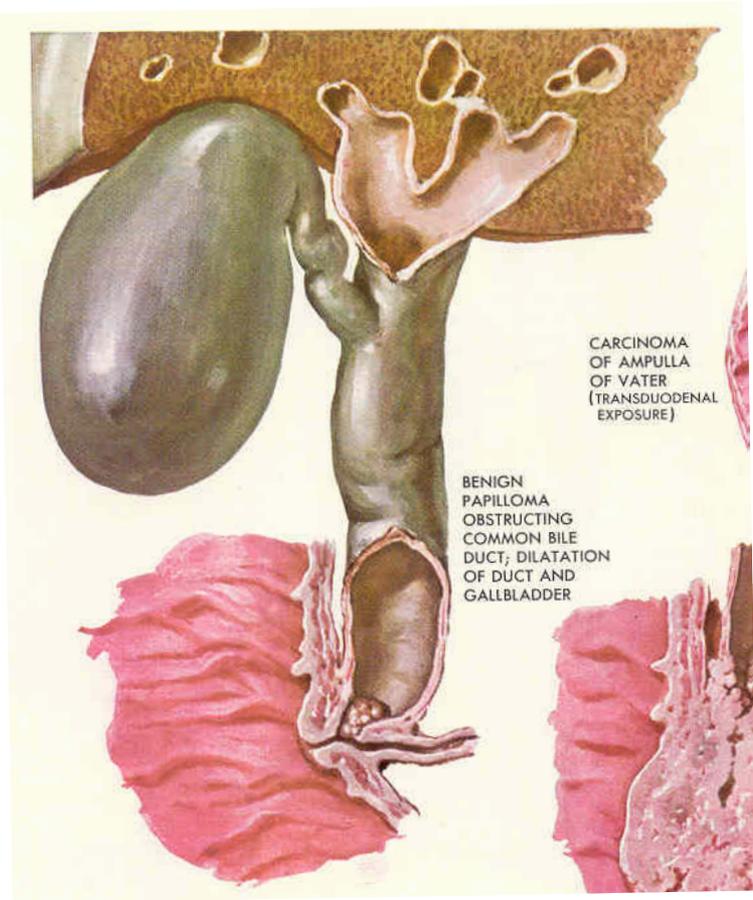


MRCP: Cholangiocarcinoma at the Bifurcation



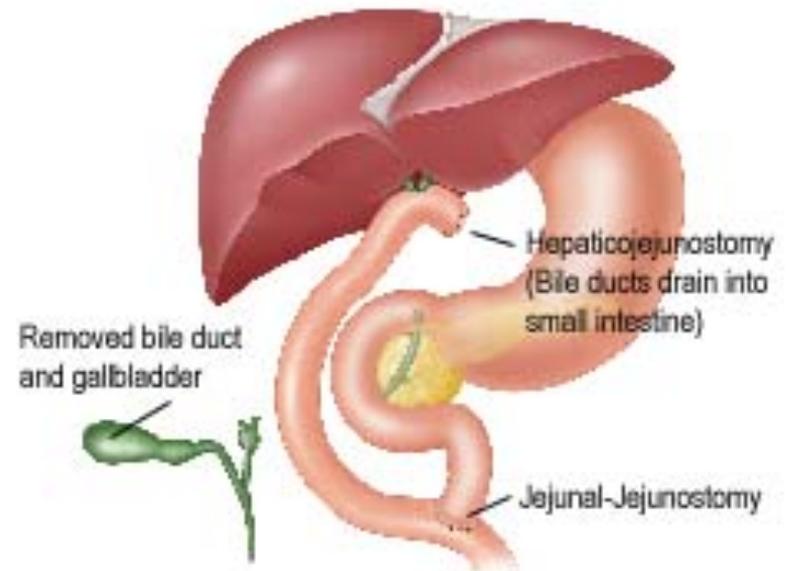
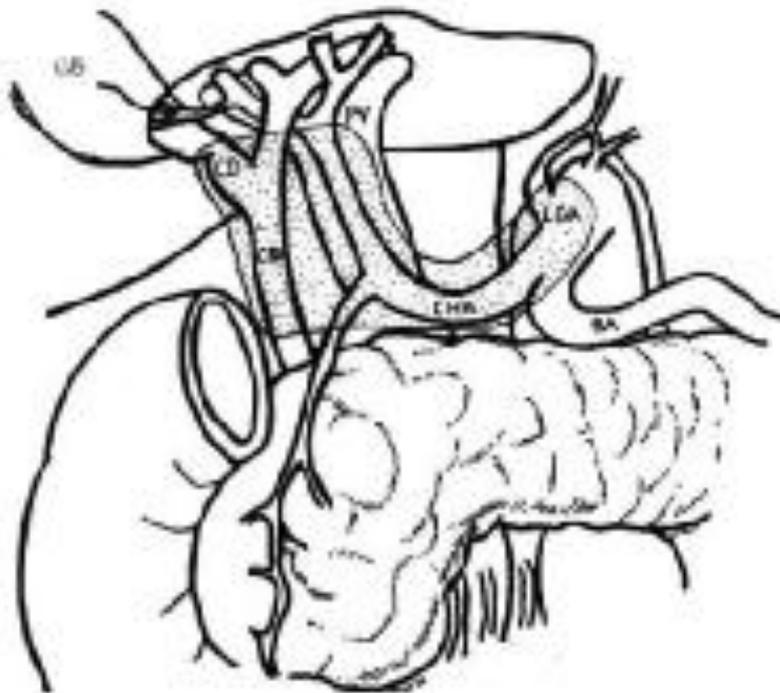
Klatskin tumour = Cholangiocarcinoma of junction of right & left hepatic ducts

ERCP: Distal CBD Cancer



Surgical Removal

- Node Dissection in Bile Duct Excision
- Roux-en-Y Hepaticojejunostomy



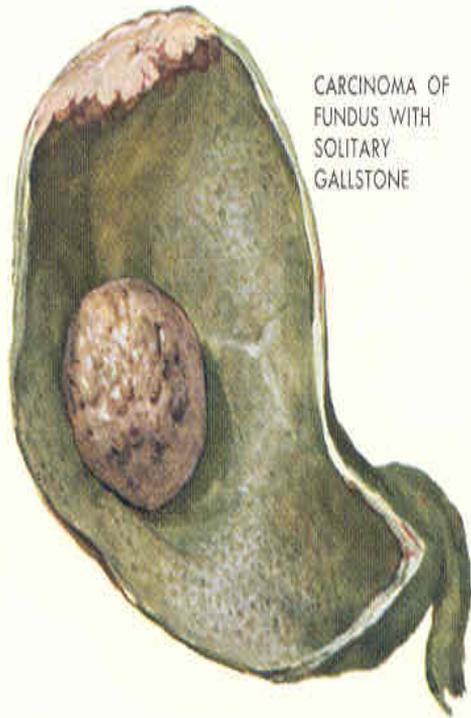
Roux-en-Y Hepaticojejunostomy Procedure performed for injuries and cancer of the bile duct.

Cholangiocarcinoma

If positive Margins or Unresectable:

- Stent
- Chemotherapy +/- Radiation Therapy
- Survival with surgery and chemo/radiation is 24 to 36 months
- With chemotherapy / radiation alone survival is 12 to 18 months

Gallbladder Cancer



CARCINOMA OF
FUNDUS WITH
SOLITARY
GALLSTONE

- 6th decade
- 1:3, Male:Female
- Highest prevalence in Israel, Mexico, Chile, Japan, and Native American women.
- Risk Factors: Gallstones, porcelain gallbladder, polyps, chronic typhoid and some drugs

Gallbladder Cancer

- Uncommon malignancy
- 2.5 per 100,000 population
- Represents 54% of biliary tract cancers.



Gall Bladder Cancer

Presentation (1)

- Discovered on pathology after a routine cholecystectomy. (T-1a/b - invades muscularis)
- CT/Chest and Abdomen, 4 phase CT of liver
- If negative for metastasis:
 - Radical cholecystectomy with nodal dissection, central hepatectomy, w or w/o bile duct excision
 - Excise port sites
 - Followed by Chemo/Radiation
- 5 year survival = 60%

Gall Bladder Cancer

Presentation 2

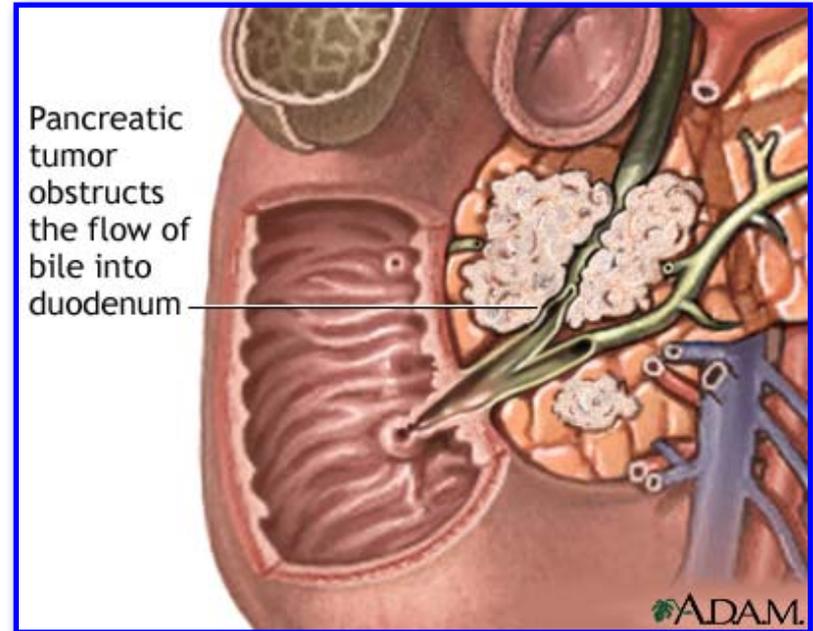
- RUQ pain, jaundice, weight loss: CT
- Biopsy yields adenocarcinoma consistent with GB primary
- Biliary Decompression
- Chemo/Radiation
- Median survival with chemoradiotherapy is 9 months.

Biliary Stricture

Biliary stricture is an abnormal narrowing of the bile duct.

Among biliary strictures:

- 90% are malignant
- Pancreatic cancer is the most common malignant cause, followed by cancers of the gallbladder, bile duct, liver, and large intestine.



Biliary Stricture – Non Cancerous Causes

Noncancerous causes of bile duct stricture include:

- Injury to the bile ducts during surgery for gallbladder removal (accounting for 80% of nonmalignant strictures)
- Pancreatitis (inflammation of the pancreas)
- Primary sclerosing cholangitis (an inflammation of the bile ducts that may cause pain, jaundice, itching, or other symptoms)
- Gallstones
- Radiation therapy
- Blunt trauma to the abdomen

Biliary Stricture – Patient Symptoms

Patients with biliary strictures may present with:

- Jaundice (yellow skin color)
- Abdominal pain
- Fever
- Vomiting

Biliary Stricture – Diagnostic Tests

Common diagnostics for biliary stricture are:

- Ultrasound
- CT
- MRI
- Biopsy
- Cholangiography

A cholangiogram is an X-ray of the bile ducts

- Can be performed:
 - Endoscopically
 - Percutaneously

Cholangiogram- Endoscopic Approach

Endoscopic retrograde
cholangiopancreatography (ERCP)

- Endoscopic tube is placed into the patient's mouth, through the stomach, and into the duodenal portion of the small intestine.
- Contrast is introduced into the biliary tract through the endoscope, in a retrograde manner.
- X-rays taken.



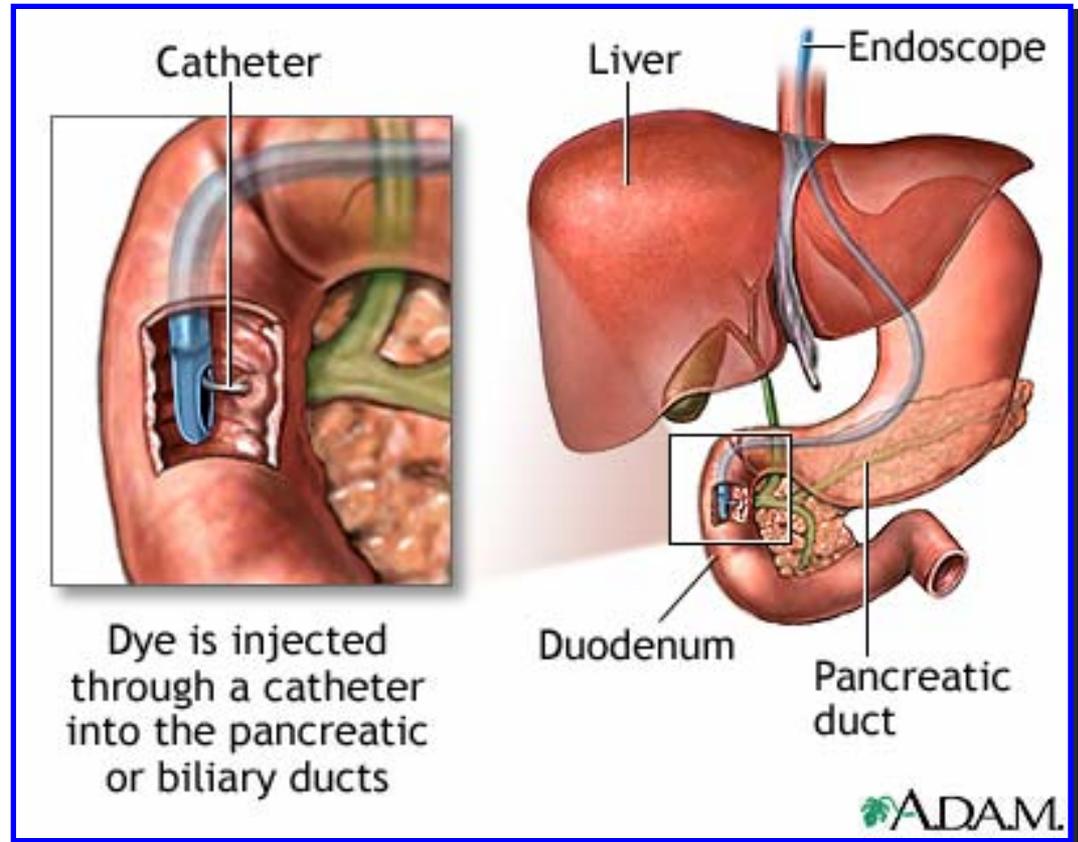
Indications For Biliary Stenting

Indications for stent insertion include:

- Ampullary Stenosis
- Management of patients with bile duct injury
- Management of benign or malignant biliary obstruction
- Prevention of obstruction where stone extraction is not possible at that time
- Management of selected pancreatic duct strictures, stones and sphincter of Oddi dysfunction

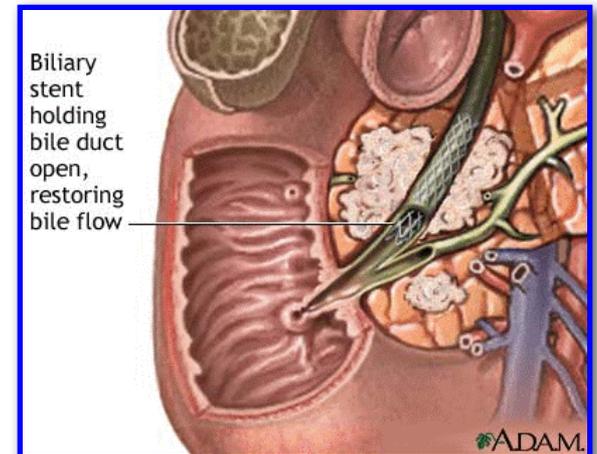
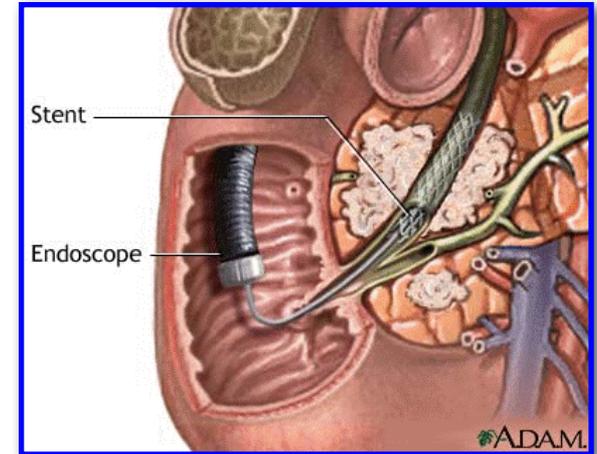
Stent Placement - Endoscopic Approach

The Endoscope is positioned in the duodenum at the opening of the bile duct.



Stent Placement -Endoscopic Approach

- A catheter is inserted through the endoscope into the ostium of the common bile duct.
- While maintaining the endoscope position in the duodenum, a wire is inserted through the catheter into the bile duct.
- The stent delivery system is then inserted over the wire to the site of obstruction, where the stent is deployed.



Stent Placement – Endoscopic Approach

Success rate of ERCP 90-95%

Complication rate of approximately 3-5%.

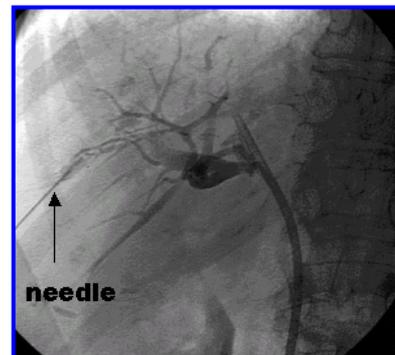
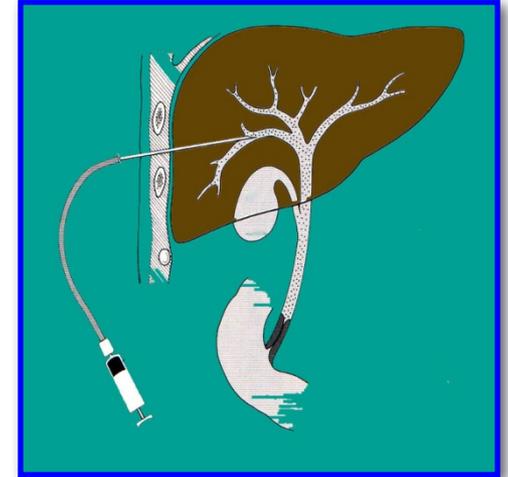
Complications:

- Pancreatitis
- Bleeding
- Perforation
- Infection
- Cardiopulmonary depression from conscious sedation.

Biliary Stent - Percutaneous Approach

For biliary stent placement using a percutaneous approach:

- A fine needle is inserted between the 4th and 5th rib on the patient's right side
- The puncture is through the liver
- The needle is inserted into an intrahepatic duct under image guidance.



Biliary Stent - Percutaneous Approach

Success rate of percutaneous transhepatic cholangiography approaches 95% when ducts are dilated.

Percutaneous approach associated with a 5-10% rate of major complications which include:

- Sepsis
- Bile leak
- Intraoperative haemorrhage
- Haemobilia
- Hepatic and perihepatic abscess
- Pneumothorax
- Skin infection and granuloma at the catheter entry site.

Percutaneous transhepatic cholangiography is contraindicated in patients with bleeding diatheses and significant ascites.

Summary

- Gallstones
 - In the gallbladder
 - Biliary colic
 - Acute and chronic cholecystitis
 - Empyema
 - Mucocoele
- Biliary tract tumours
- Other conditions
 - Acute acalculous cholecystitis
 - Mirizzi's syndrome
 - Primary Biliary Cirrhosis
 - Primary Sclerosing Cholangitis
 - Biliary tract cysts
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 - Cholangitis
 - In the Gut
 - Gallstone ileus