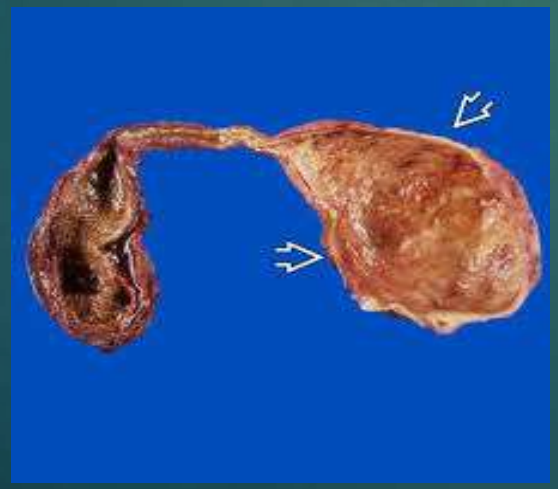
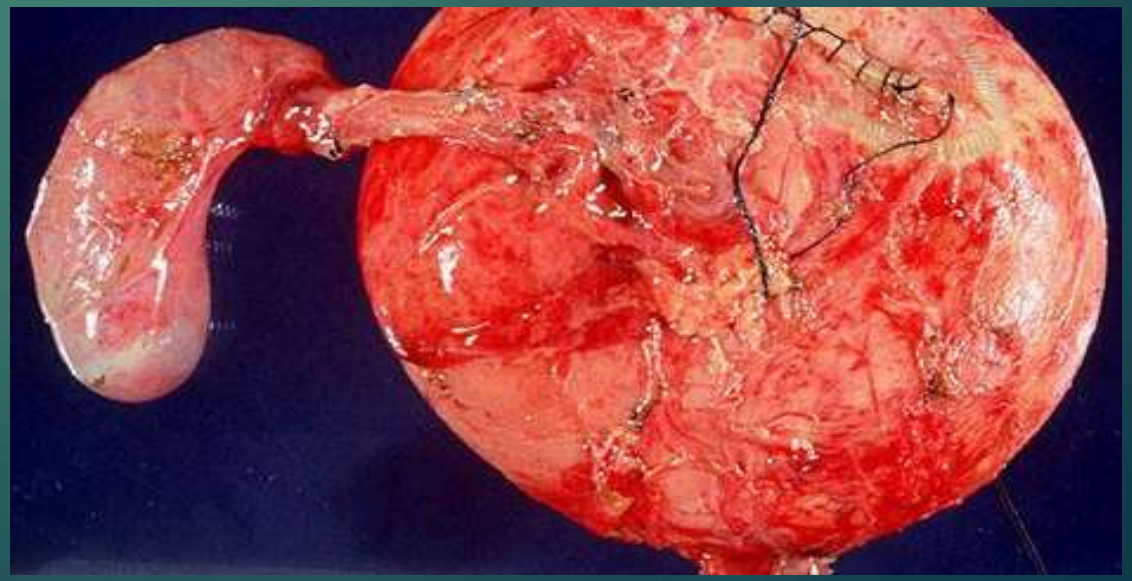
Choledochal Cysts

- They are **congenital bile duct anomalies.**
- These cystic dilatations of the biliary tree can involve the **extrahepatic** biliary radicles, the **intrahepatic** biliary radicles, or **both.**
- They may occur as a **single cyst** or in **multiples** within the biliary tree.
- Choledochal cysts are more prevalent in **females** than males with **3:1 ratio**





Pathophysiology

- **Abnormal union** between the **pancreatic duct** and the **common bile duct** allows pancreatic secretions to reflux into the common bile duct, where the **pancreatic proenzymes** become activated, damaging and weakening the bile duct wall.

- Defects in **epithelialization and recanalization** of the developing bile ducts and **congenital weakness of the ductal wall** also have been implicated.

- The result is the formation of a choledochal cyst.

Presentation

- **Infants:** frequently present with **obstructive jaundice** and **acholic stools. Palpable mass** in the right upper quadrant of the abdomen, accompanied with **hepatomegaly.**

- **Children:** diagnosed after infancy typically have a clinical picture of intermittent **biliary obstruction** or recurrent bouts of **pancreatitis**. Palpable right upper quadrant **mass** and **jaundice.**

- **Adults:** complain of vague epigastric or right upper quadrant pain and can develop **jaundice** or **cholangitis**. A **classic triad** of abdominal pain, jaundice, and a palpable right upper quadrant abdominal mass has been described in adults with choledochal cysts but is found in only **10-20%** of patients.

Diagnosis

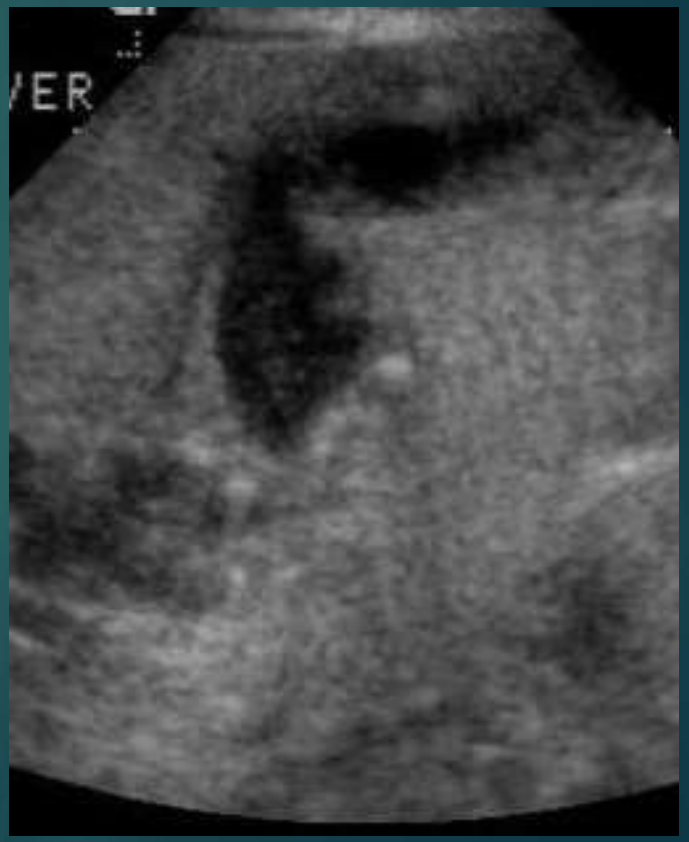
- **Lab studies** : No laboratory studies are specific for the diagnosis of a choledochal cyst. We do **(CBC, LFT and Chemistry panel),** to help us in diagnosis of associated cholangitis and pancreatitis.

- **Imaging Studies** : **(US, MRI, CT and MRCP).**

- **Invasive diagnostic studies** : **(ERCP and PTC)**

Imaging Studies

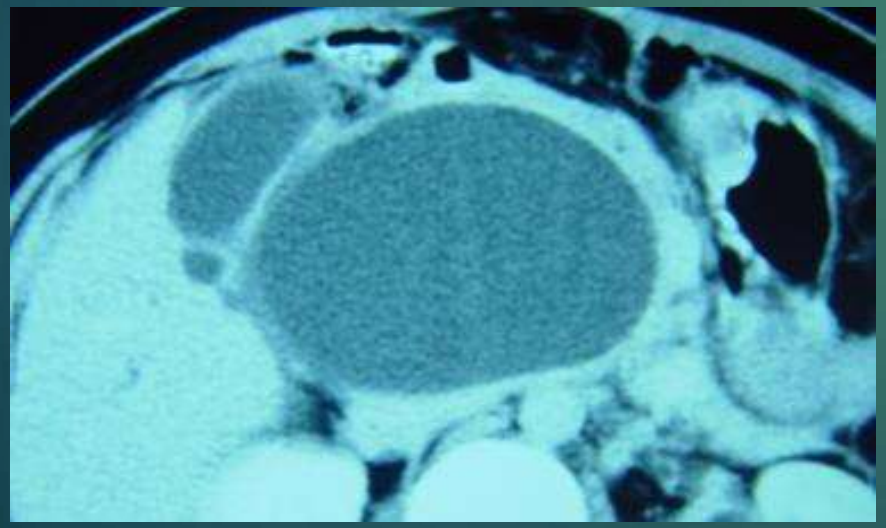
- **Ultrasound (US) :**
- Abdominal ultrasonography is the **test of choice** for the diagnosis of a **choledochal cyst.**
- Ultrasonography is useful in the **antenatal period** as well and can demonstrate a choledochal cyst in a fetus as early as the beginning of the **second trimester.**



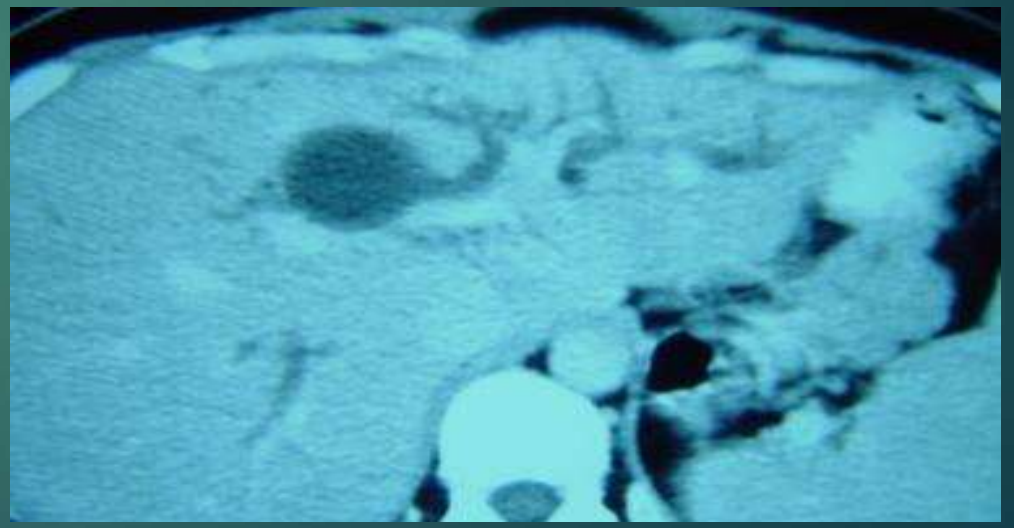
- **This picture shows a type 1 cyst in a 4 month child complaining of jaundice**

Imaging Studies

- **CT scan and MRI :**
- Abdominal CT scanning and MRI help to delineate the anatomy of the lesion and the surrounding structures. These tests also can assist in defining the presence and extent of intrahepatic ductal involvement.



CT scan demonstrating a large choledochal cyst and the adjacent gall bladder.



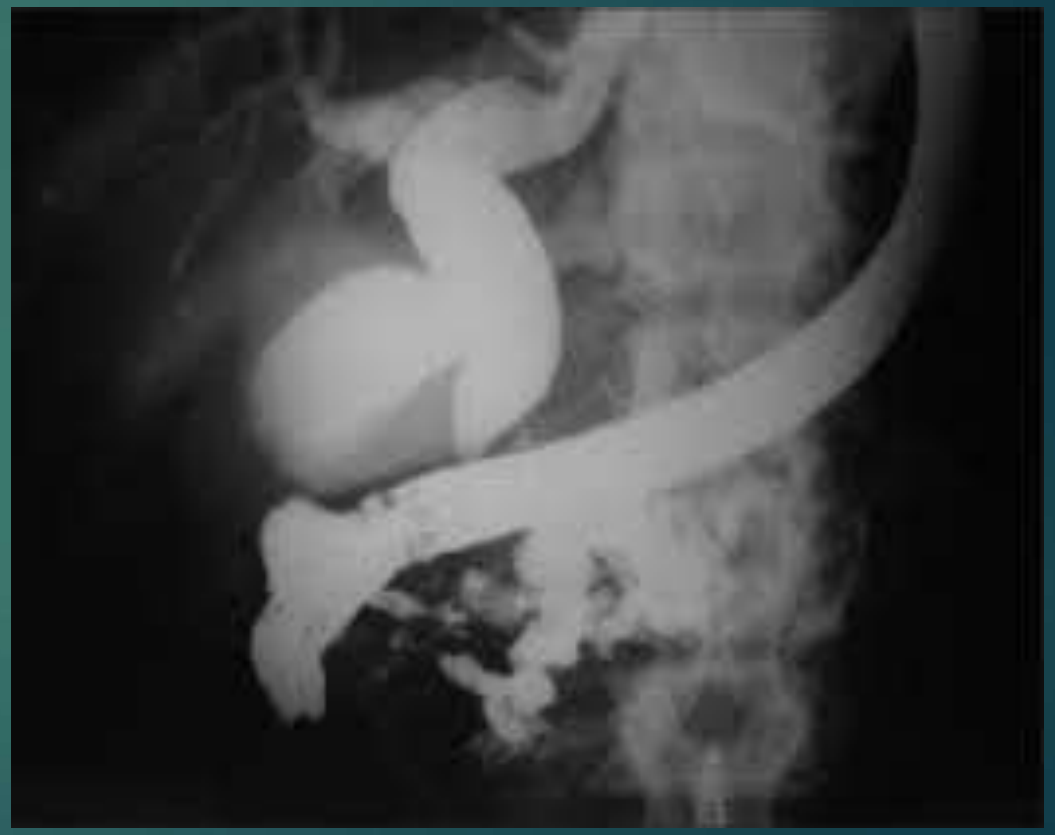
CT scan of a choledochal cyst demonstrating intrahepatic extension involving the main left hepatic duct.

Invasive Diagnostic Studies

- When **noninvasive** measures (Ultrasonography, CT scanning and MRI) **fail** to sufficiently detect the anatomy, they should be supplemented by the addition of :
- **Percutaneous Transhepatic Cholangiography (PTC)** or **Endoscopic Retrograde Cholangiopancreatography (ERCP)** which may have a sensitivity up to 100%.
- These studies are particularly helpful in demonstrating the presence of an **anomalous pancreatobiliary junction** and in showing associated extrahepatic or intrahepatic **strictures** and **stones.**

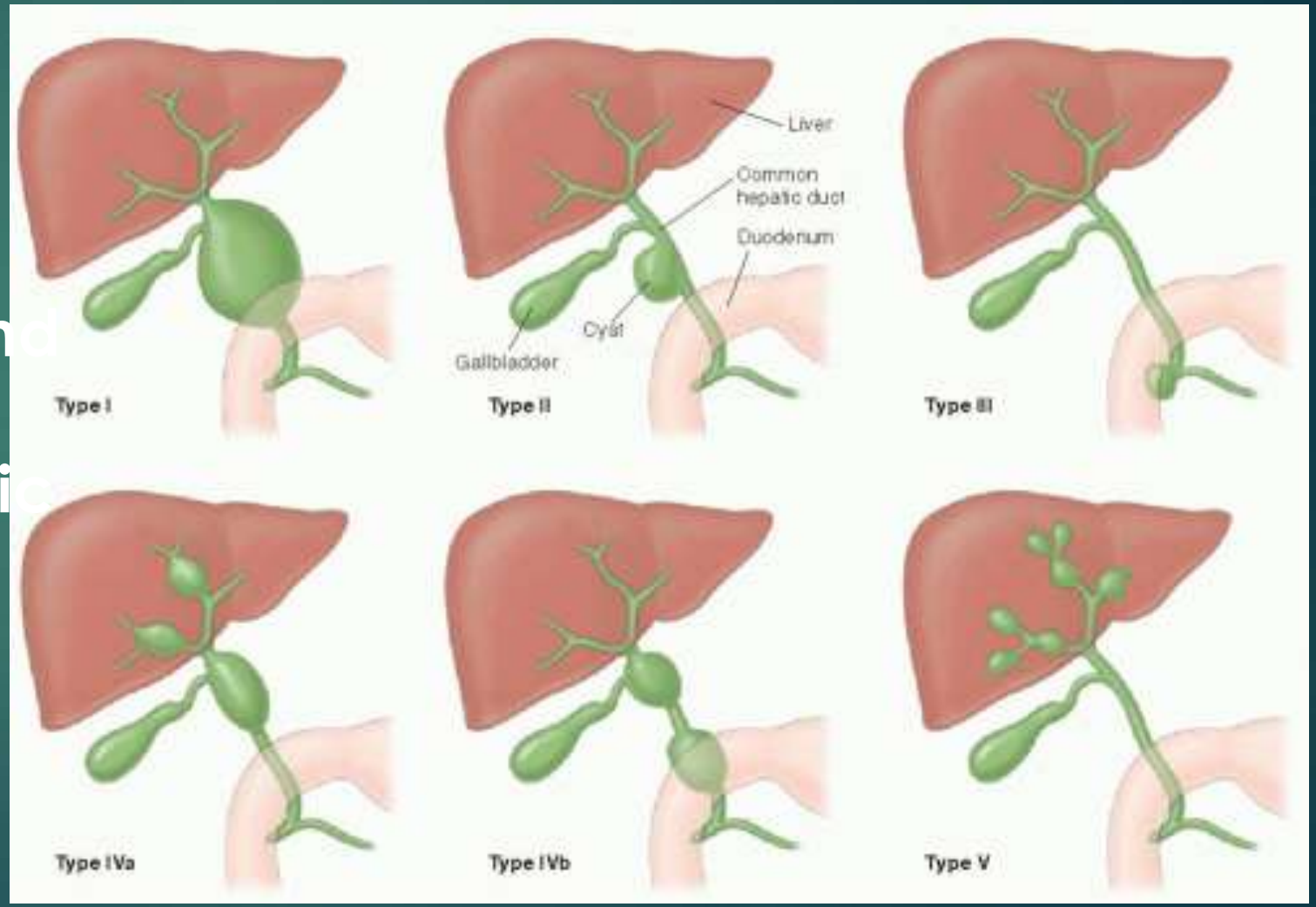
Percutaneous transhepatic cholangiogram(PTC)

Cholangiogram – Caroli's disease with characteristic stricture and segmental intrahepatic duct dilations.



Classification

* First type ,,, fusiform
* Second type,,, saccular
* Third type,,,,through duodenum
* 4a type ,,,, for all (inter and extra heptic)
* 4b ,,, be out (extra hepatic)
* 5 caroli,,,,intrahepatic



Classification

Type	Description
Type I	Fusiform dilation of the common bile duct (80-90% of the cases).
Type II	Bile duct diverticulum (0-2% of the cases).
Type III	Saccular dilation of the common bile duct inside the duodenum wall, or choledochocele.
Type IV	Multiple intra- and extrahepatic bile dilations (10-15%). Variant A: intra- and extrahepatic affectation. Variant B: involves only the extrahepatic bile duct.
Type V	Fusiform or saccular cystic dilation of the intrahepatic bile ducts, whether or not associated to hepatic fibrosis (Caroli disease).

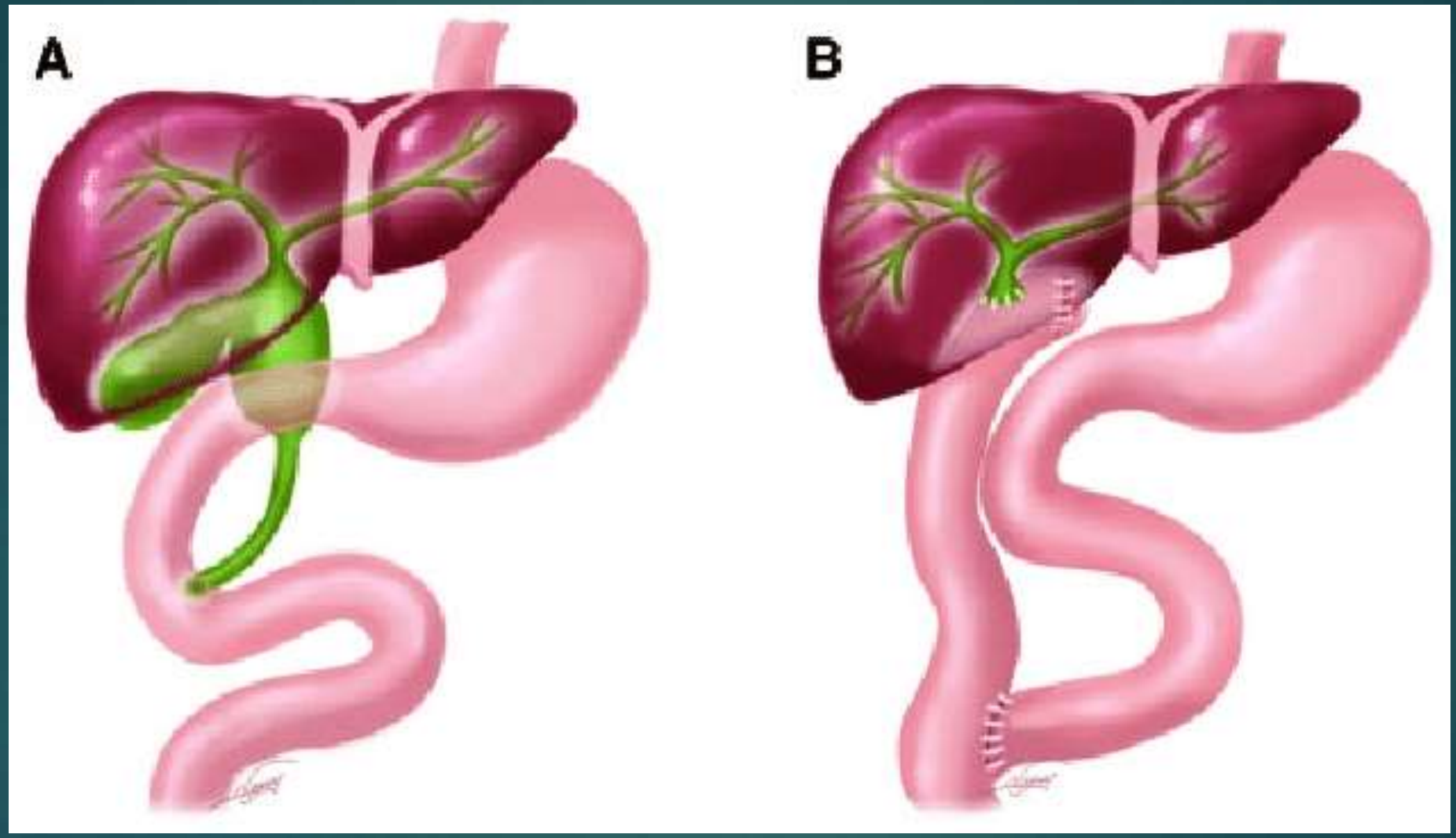
Based on the description handed by Kevin C. Soares, et al., 2014. [14].

Treatment and Management

- The **treatment of choice** for choledochal cysts is **complete excision** of the cyst with **construction** of a biliary-enteric anastomosis to restore continuity with the gastrointestinal tract.
- **Partial resection of the cyst** and **internal drainage procedures** expose patients to increased risks of **cholangitis, pancreatitis,** and **cholangiocarcinoma.**
- Appropriate **antibiotic therapy** and **supportive care** should be given to patients presenting with cholangitis. Broad-spectrum antibiotic therapy directed against common biliary pathogens, such as ***Escherichia coli*** and ***Klebsiella* species.**

Treatment according to type

Type	Procedure
I	The treatment of choice is complete excision of the involved portion of the extrahepatic bile duct. A Roux-en-Y hepaticojejunostomy is performed to restore biliary-enteric continuity.
II	Excision in its entirety. The resultant defect in the common bile duct is closed over a T-tube.
III	<3 cm → endoscopic sphincterotomy.
	>3 cm → lesions are excised surgically through a trans-duodenal approach.
IV	The dilated extrahepatic duct is completely excised and a Roux-en-Y hepaticojejunostomy is performed to restore continuity. In such instances, the affected segment or lobe of the liver is resected.
V (*Caroli disease*)	limited to one hepatic lobe → hepatic lobectomy. bilobar disease who begin to manifest signs of liver failure → liver transplantation

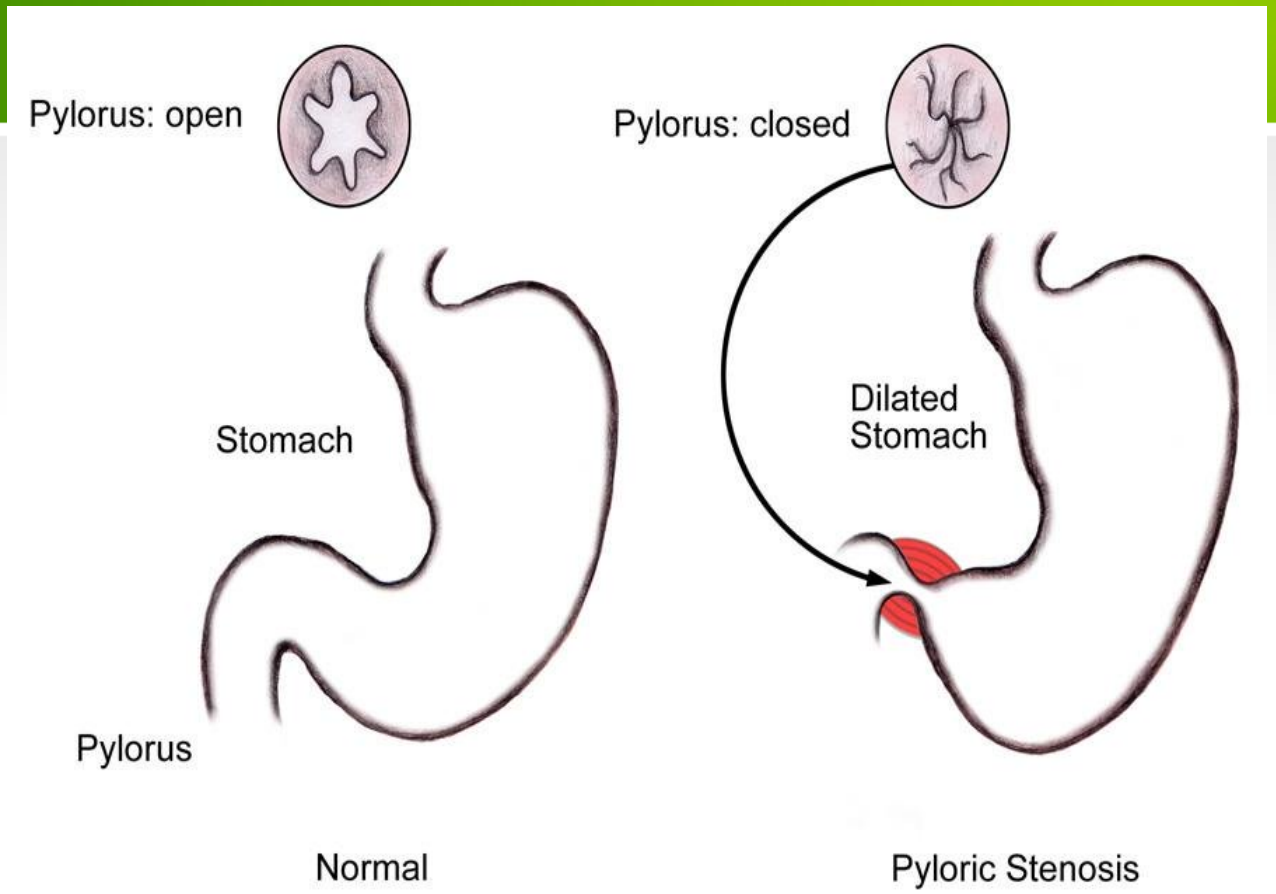

A
B

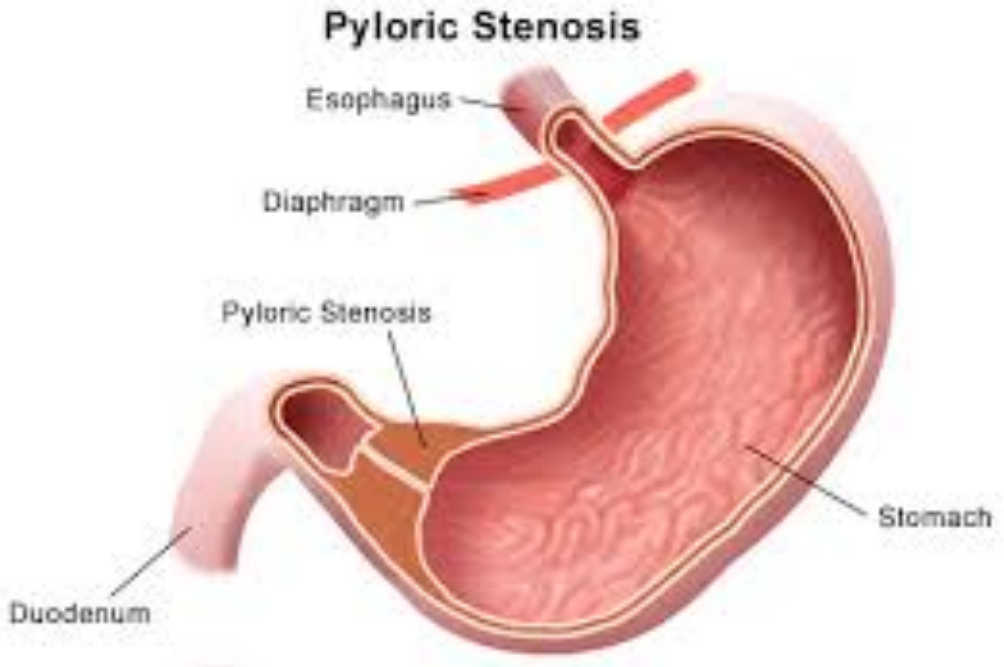
Pyloric Stenosis

- **Pyloric Stenosis** involves **narrowing and obstruction** of the pyloric channel because of **hypertrophy** of the **circular muscle** of the **pylorus.**

- It is the most common pediatric surgical disorder of infancy that necessitates surgery for associated emesis.

- Gastric outlet obstruction results in **emesis**, which is characteristically **nonbilious and projectile**. Protracted emesis, as well as failure of the stomach to empty into the duodenum, results in **progressive dehydration, electrolyte abnormalities, acid-base disorders, weight loss**, and, potentially, **shock.**


Pylorus: open
Pylorus: closed
Stomach
Dilated Stomach
Pylorus
Normal
Pyloric Stenosis


Pyloric Stenosis
Esophagus
Diaphragm
Pyloric Stenosis
Stomach
Duodenum

Presentation

Projectile vomiting is typically nonbilious but may have brown discoloration or a coffee-ground appearance due to associated gastritis, particularly if emesis has persisted for several days.

The vomiting occurs within 30-60 minutes after feeding.

The infant remains hungry and usually attempts to feed immediately after vomiting.

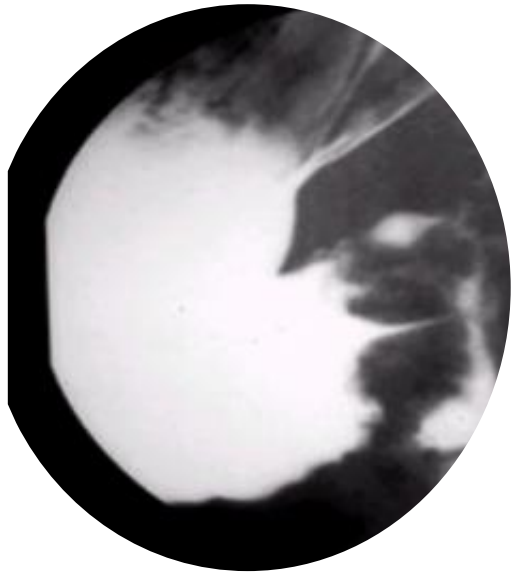
Diagnosis

Laboratory Studies:

- An **electrolyte panel** is essential for estimating the state of dehydration and acidosis/alkalosis in patients with pyloric stenosis. **Hypochloremic hypokalemic metabolic alkalosis** is the characteristic biochemical disturbance observed in pyloric stenosis.

Radiography:

- Upper GI (UGI) contrast studies have largely been supplanted by **Ultrasonography** as the **study of choice** for confirming pyloric stenosis. Although UGI studies have been reported as yielding an **accuracy** of **96%,** obvious disadvantages of such studies include **radiation exposure** and the risk of **aspiration** of contrast material.

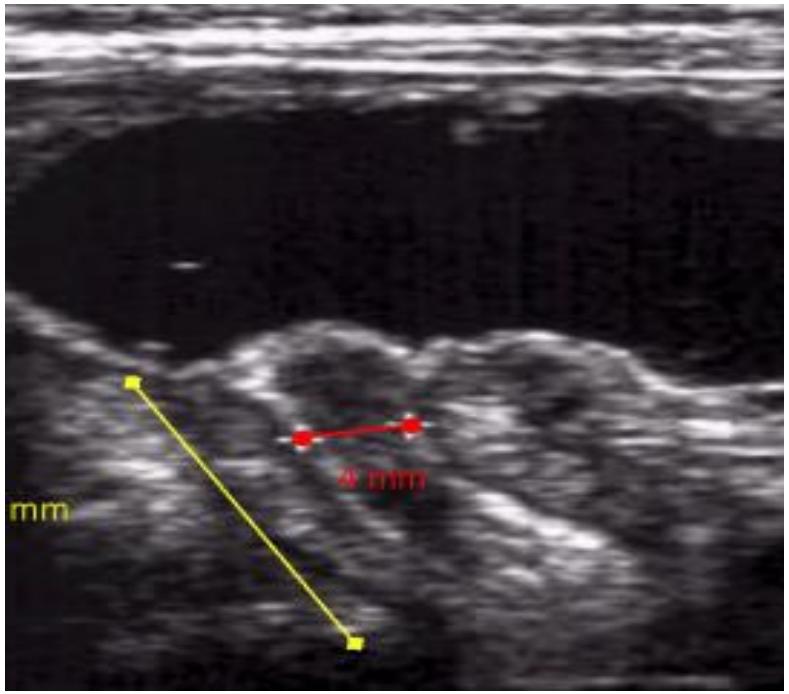


Diagnosis

- **Ultrasonography:**
- On US, a diagnosis of pyloric stenosis can be made through identification of an **elongated sausage-shaped mass** with the following characteristics:

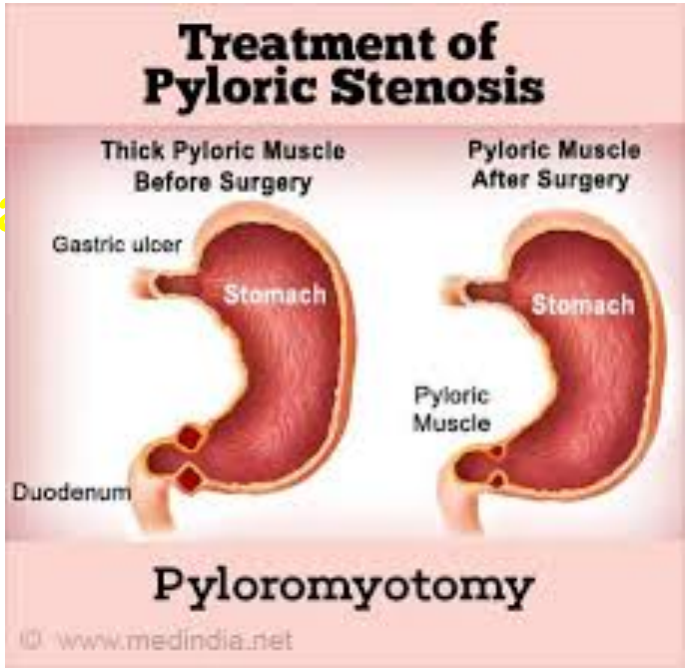
1. **Pyloric diameter >14 mm**
2. **Muscular thickness >4 mm**
3. **Length >16 mm**

- sensitivity of **91-100%** and a specificity of **100%** have been reported with these criteria.
  US is best performed with the stomach evacuated.



Treatment

- Pyloric stenosis is not a surgical emergency.
- Once the diagnosis of pyloric stenosis has been confirmed, adequate ongoing **preoperative fluid resuscitation** must be maintained by establishing adequate **urine output (1 mL/kg/hr)** and **correcting acid-base disorders and electrolyte abnormalities.**
- Pyloromyotomy may be performed either as an procedure, via a right-upper-quadrant (RUQ) incision or an **umbilical** incision operation), or as **laparoscopic** procedure.



Hirschsprung's disease

- It is aganglionic mega colon due to failure of caudal migration of neuroblasts derived from the neural crest.
- Incidence rate is 1:5000 births.
- 80% of cases are boys.
- 3-5% of patients have down's syndrome.
- >95% of cases are full term babies.
- Strong familial association in long segment and total colonic aganglionosis (15% and 25%).

- Aganglionosis begins at anorectal line
- 80% involve only rectosigmoid area
- 10% extend proximal to splenic flexure
- 10% involves the entire colon and part of small bowel
- Rarely involves entire gastrointestinal tract

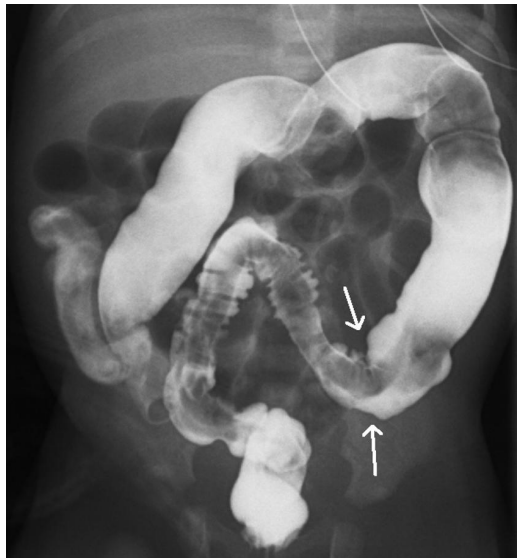
Presentation

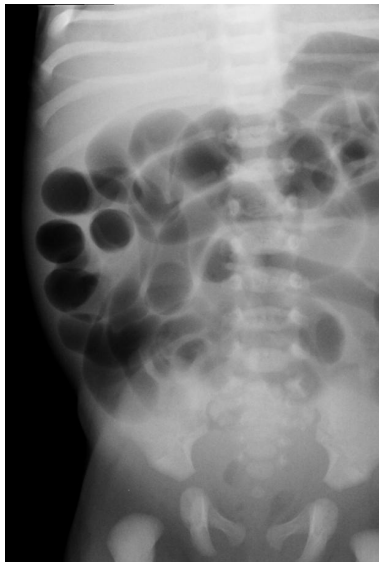
- Abdominal distention.
- Bilious vomiting.
- Failure to pass meconium in the first 24 hrs.
- May be associated with constipation and diarrhea.
- Failure to thrive.
- Rectal examination reveals a tight spastic rectum
- RULE: neonat with distended abdomin if term think about hirschsprung disease .IF preterm think about NEC .

Diagnosis

- Abdominal X-Ray.
- Barium enema.
- **Rectal biopsy (gold standard)**
- Anal manometry.

distal collapse trnsitional zone proximal dilated





Rectal biopsy types

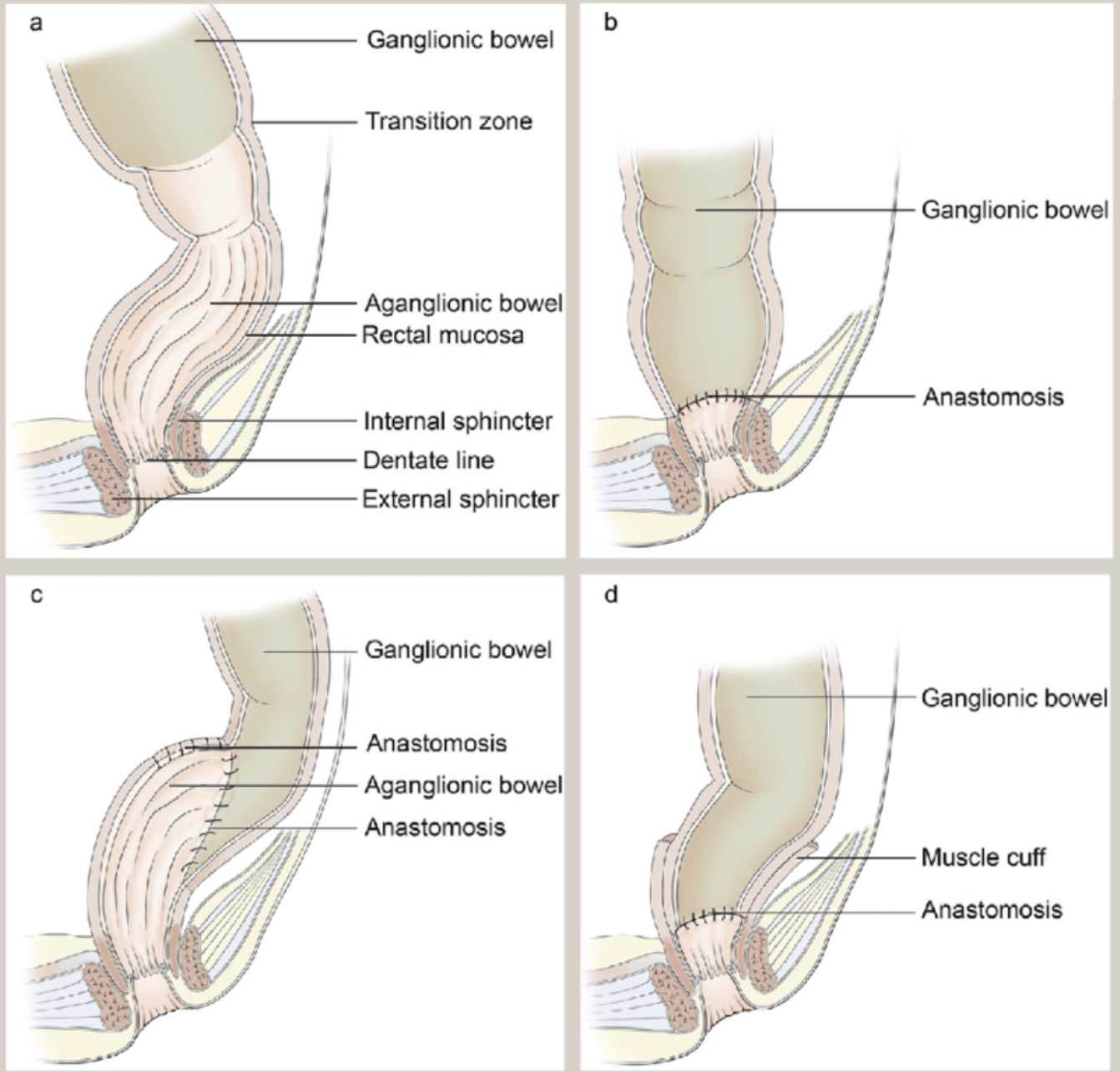
- Submucosal suction biopsy
  - Meissner's submucosal plexus
- Full thickness rectal biopsy
  - Auerbach's myenteric plexus
- Acetylcholinesterase staining
  - increased staining of neurofibrils

Treatment

- Laxatives and enemas.
- Colostomy in neonates.
- Surgical treatment : excision of the spastic segment and re-anastomosis to anal canal.

❖Surgical options are :

1. Swenson procedure.
2. Duhamel procedure.
3. Soave procedure.



B. Swenson procedure.

C. Duhamel procedure.

D. Soave procedure.

Post operative complications

- Leak at anastomosis (5-7%).
- Post-op enterocolitis (19-27%).
- Constipation.
- Strictures.
- Incontinence.

Biliary Atresia

Ahmad khader

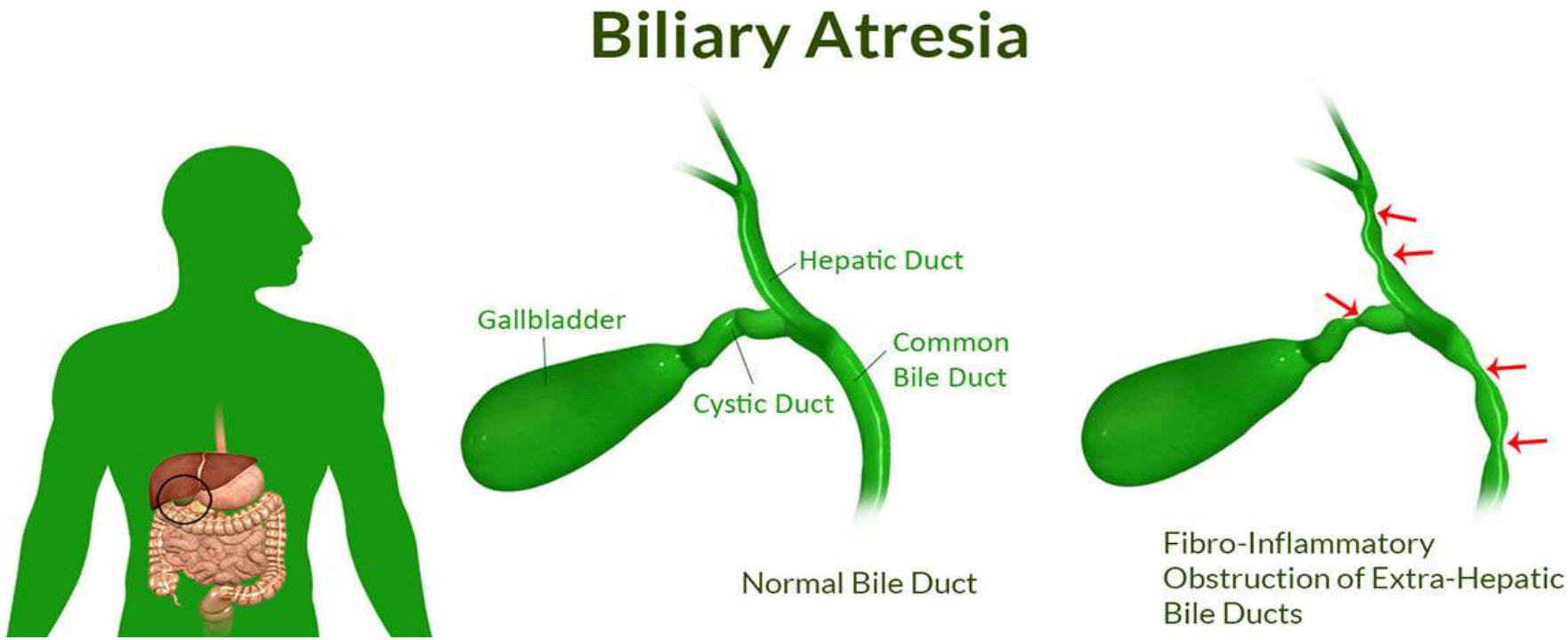
- It is a rare disease of the **liver** and **bile ducts** that occurs in **infants.**

- It is characterized by **obliteration or discontinuity** of the **extrahepatic** biliary system, resulting in **obstruction** to bile flow.

- **Extrahepatic** biliary atresia is slightly more common in **females** than in males.

- If not surgically corrected, **secondary biliary cirrhosis** invariably results.

- Patients with biliary atresia can be subdivided into 2 distinct groups: those with **isolated biliary atresia (neonatal form),** which accounts for **65-90%** of cases, and **(embryonic form)** patients with congenital anomalies such as situs inversus, polysplenia or asplenia comprising **10-35%** of cases.

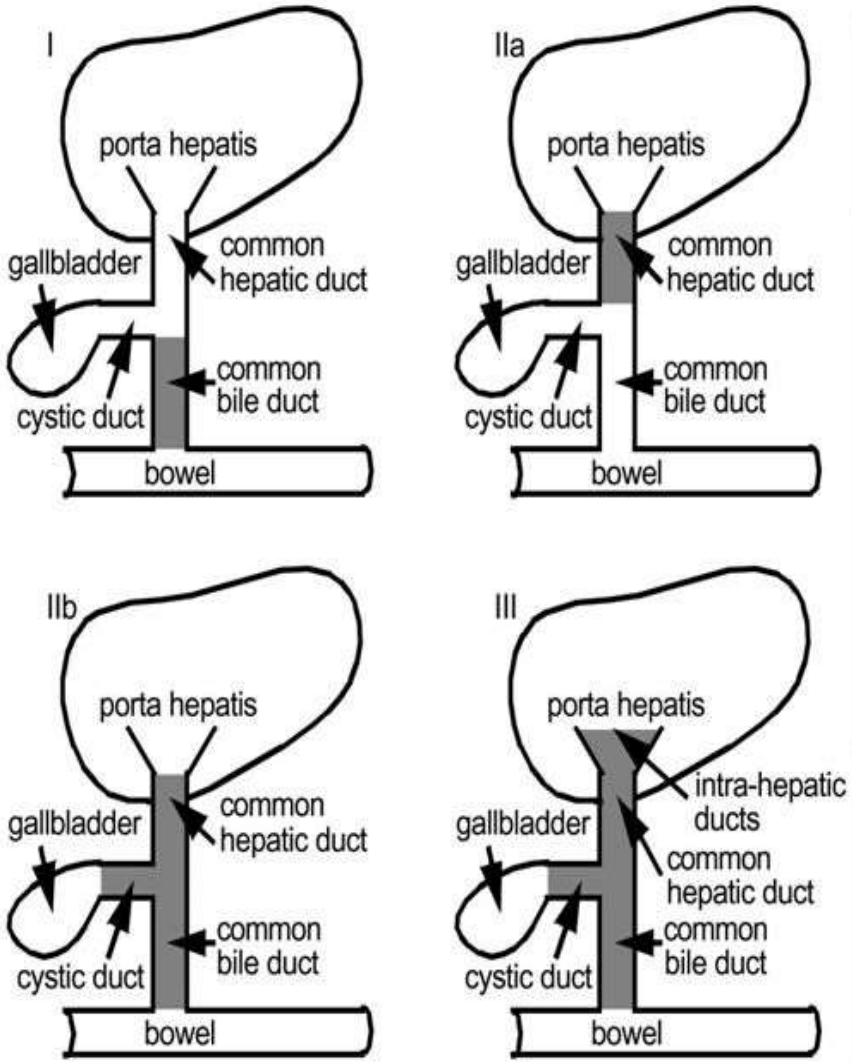


Types

- The following classification is based on the predominant site of atresia:

1. **Type I** involves obliteration of the **common duct**; the proximal ducts are patent
2. **Type IIA** is characterized by **atresia of the hepatic duct.**
3. **Type IIB** is obliteration of common hepatic duct, cystic and common bile duct
4. **Type III (>90% of patients)** involves **atresia** of the **right** and **left hepatic ducts** to the level of the **porta hepatis**

Kasai Classification



TYPE 1:

CBD is obliterated proximal bile ducts are patent

TYPE 2:

atresia of the hepatic duct is seen, cystic bile ducts at porta hepatis

TYPE 2A:

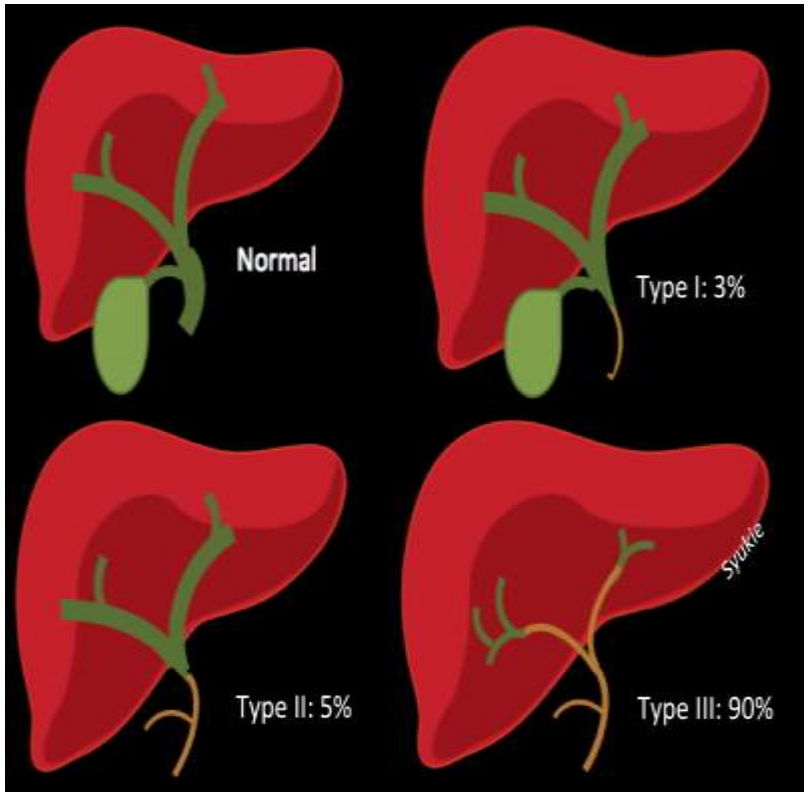
cystic and CBD are patent

TYPE 2B:

cystic, CBD and hepatic ducts are all obliterated

Type 3:

atresia refers to discontinuity of both R and L hepatic ducts to the level of the porta hepatis common, (>90% of cases)



Pathophysiology

- The pathogenesis of this disorder remains poorly understood.

- Early studies showed a **congenital** malformation of the **biliary ductular system.**

- **The more common neonatal type of the disease** is characterized by a progressive **inflammatory lesion.**
- which suggests a role for **infection** and/or **toxic agents** causing bile duct obliteration.

Presentation

- Typical symptoms include variable degrees of **jaundice**, **dark urine**, and **light stool.**
- Appetite, growth, and weight gain may be normal.
- On physical exam **hepatomegaly** may be present **early**, and the liver is often firm or hard to palpation.
- **Splenomegaly** is **common**, and an enlarging spleen suggests progressive cirrhosis with portal hypertension. (late)

Clinical presentation

- **Prolonged jaundice (ie. beyond 14 days age)**
- Plus:
  - **Pale stools**
  - **dark urine**
  - **Hepatomegaly**
  - **Failure to thrive**

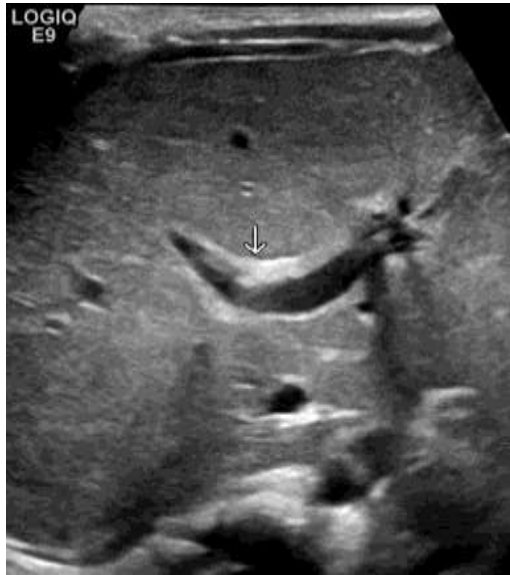
- **Other presentations:**
- Coagulopathy
- Abnormal antenatal scan ~5%
- Splenomegaly / portal hypertension (late signs)

Diagnosis

- **Labs :**
- **Conjugated hyperbilirubinemia (Direct),** defined as any level exceeding either 1 mg/dL **(total bilirubin < 5 mg/dL)** or 20% of total bilirubin **(total bilirubin >5 mg/dL),** is always **abnormal.**
- **Elevated Alkaline phosphatase (ALP), Gamma-Glutamyl transpeptidase (GGTP)**
- **Sweat chloride (Cl):** Biliary tract involvement is a well-recognized complication of Cystic Fibrosis (CF).

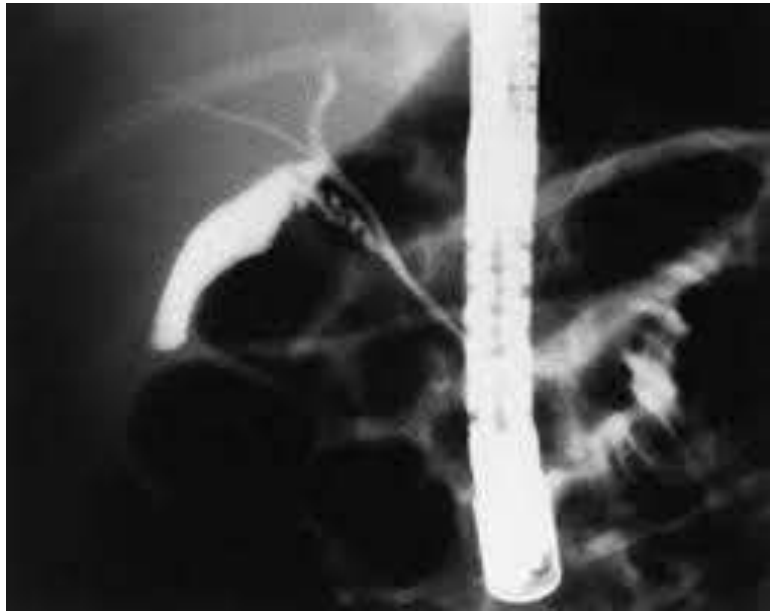
Diagnosis

- **Imaging :**
- **Ultrasonography (US) :** can exclude specific anomalies of the extrahepatic biliary system, particularly **choledochal cysts.**
- Ultrasonography may demonstrate absence of the gallbladder and no dilatation of the biliary tree.
- Unfortunately, the sensitivity and specificity of these findings, even in the most experienced centers, probably do **not exceed 80%.**
- **Technetium-99m iminodiacetic acid hepatobiliary imaging**



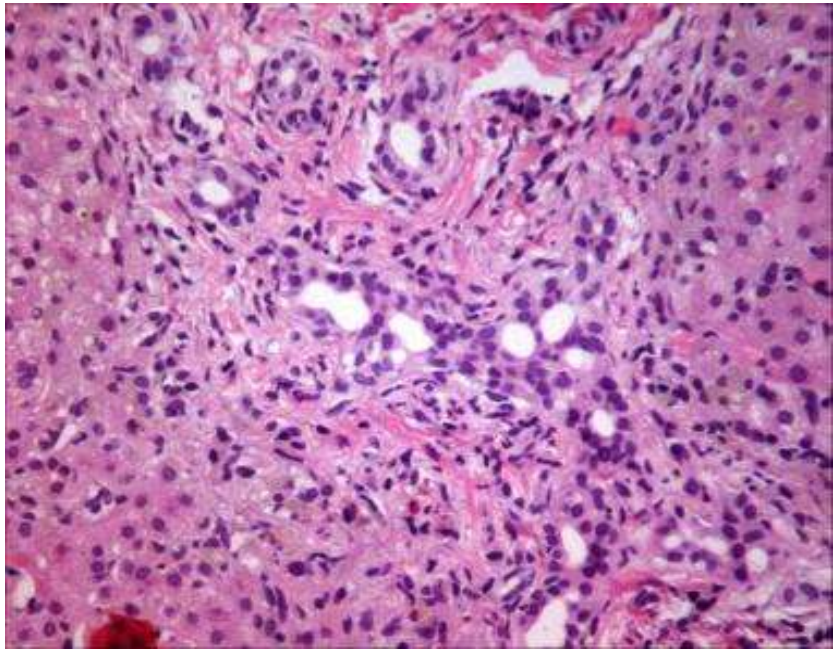
Diagnosis

- **ERCP :**
- Although not yet widely used, reports have demonstrated the use of ERCP in diagnosing biliary atresia
- **ERCP** may become part of the management algorithm in assessing **neonatal direct hyperbilirubinemia,** for which other studies have failed to confirm a diagnosis.



Diagnosis

- **Percutaneous Liver Biopsy :**
- Percutaneous liver biopsy is widely regarded as the most valuable study for evaluating neonatal cholestasis.
- **90%** sensitivity and specificity for biliary atresia.
- Bile ductular proliferation in liver biopsy specimen from patient with biliary atresia.



Treatment and Management

- **No primary medical treatment** is relevant in the management of extrahepatic biliary atresia.
- Once biliary atresia is suspected, **surgical intervention** is the only mechanism available for a **definitive diagnosis (intraoperative cholangiogram)** and **therapy (Kasai portoenterostomy).**
- In most cases of atresia, dissection into the porta hepatis and creation of **a Roux-en-Y anastomosis** with a 35-cm to 40-cm retro-colic jejunal segment is the procedure of choice.
- In order to prevent ascending cholangitis postoperatively, **prophylaxis** with **trimethoprim-sulfamethoxazole** has been used on a long-term basis.