

NEONATAL SURGICAL JAUNDICE

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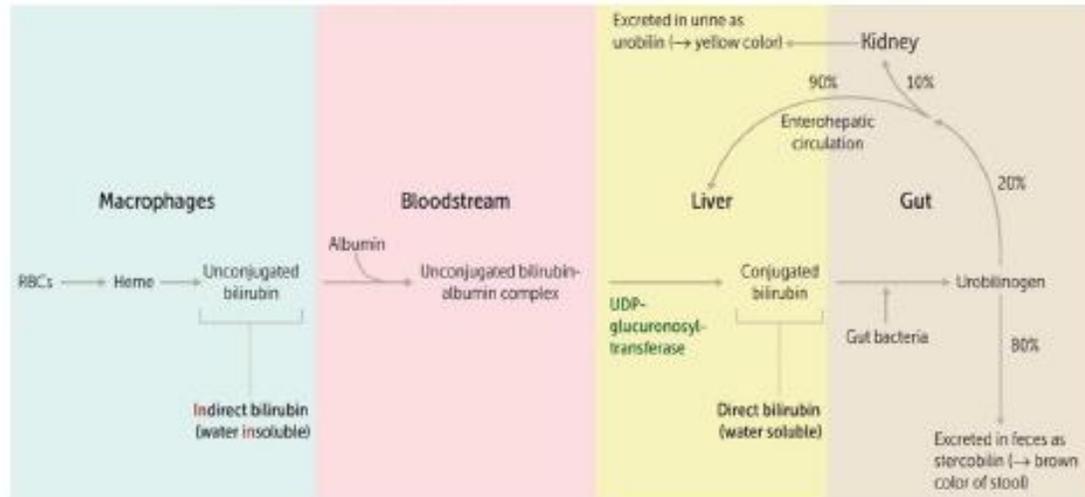
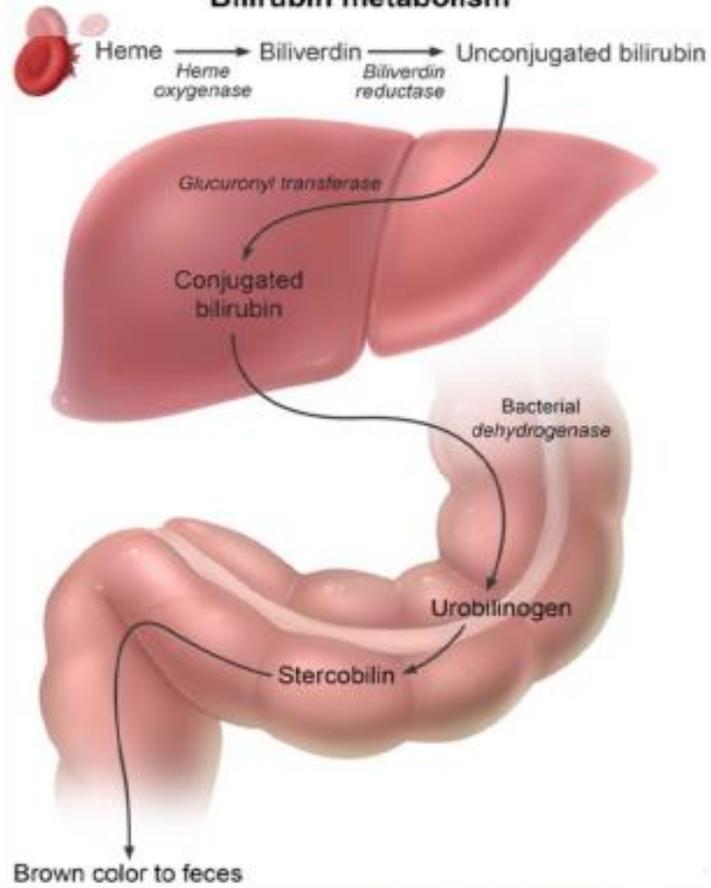
Dr. Abdallah Alraweh

INTRODUCTION

- Jaundice is the most common condition that require medical attention in newborns.
- It is a condition resulting from the accumulation of unconjugated hyperbilirubinemia
- In most infants, unconjugated hyperbilirubinemia is a normal transitional phenomenon in which it is benign and no intervention is required.
- Jaundice presents during the first week of life in **60% of term infants and 80% of preterm infants.**
- However, approximately 5-10% of neonates may have clinically significant jaundice in which the serum bilirubin may rise excessively, which can be cause for concern
- High bilirubin levels may be toxic to the developing central nervous system and may cause neurological impairment even in term newborns e.g. **kernicterus.**

BILIRUBIN METABOLISM

Bilirubin metabolism



TYPES OF NEONATAL JAUNDICE

Jaundice type	Onset/timing	Type of bilirubin	Causes
Physiological	After day 3 to day 7 approximately	Unconjugated	Normal physiological haemolysis of fetal erythrocytes (red blood cells). Haem breaks down to form unconjugated bilirubin. Hepatic immaturity, high red cell load at birth and short life span of red blood cells increase the tendency to develop this type of jaundice
Pathological	Within the first 24 hours	Unconjugated	Due to a disease process such as ABO haemolytic disease, Rhesus incompatibility, severe/congenital infection
Prolonged	Persists past the usual 7–10 days period	Conjugated or unconjugated	Exclusive breastfeeding although no intervention is necessary Infection Hypothyroidism Haemolytic diseases such as Glucose-6-phosphate dehydrogenase deficiency or Spherocytosis Hepatic disease or biliary conditions Total parenteral nutrition

BILIARY ATRESIA

INTRODUCTION TO BILIARY ATRESIA

- Biliary atresia (BA) is a **progressive, idiopathic, fibro-obliterative** disease of the extrahepatic biliary tree that presents with biliary obstruction exclusively in the neonatal period.
- Although the overall incidence is low (approximately 1 in 10,000 to 20,000 live births), BA is the **most common cause** of neonatal jaundice for which surgery is indicated and the **most common indication** for liver transplantation in children.
- Bile can't flow into the intestine, so it builds up in the liver and damages it; it could progress to hepatic fibrosis, cirrhosis, and liver failure.

PATHOGENESIS OF BA

- The causes of BA are **not well established** and are probably **multifactorial**; **genetic factors** (such as mutations in the CFC1 gene) may play a permissive role in some cases, but **infectious** (such as cytomegalovirus, reovirus, and group C rotavirus), or **immunologic mechanisms** are probably involved.

BA CLASSIFICATION

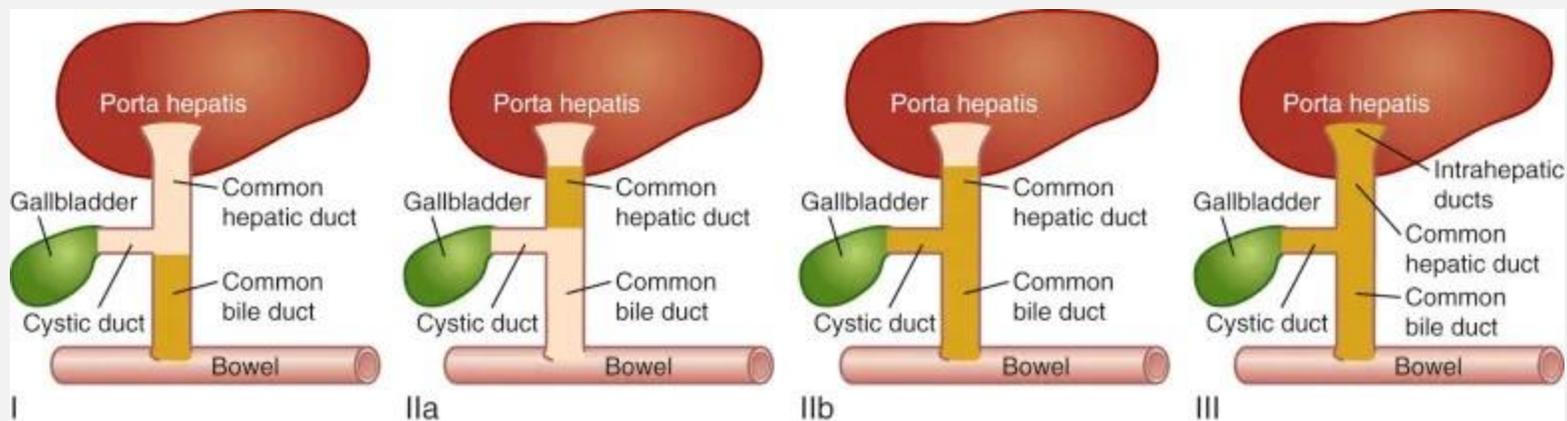
- **There are two classification**
- **1- according to association anomalies**
- **2- according to anatomical part**

ACCORDING TO THE ASSOCIATION ANOMALIES

- Infants with **BA** can be grouped into three categories :
- **BA without any other anomalies or malformations** – This pattern is sometimes referred to as **perinatal BA** and occurs in 70 to 85 percent of infants with **BA**. Typically, these children are born without visible jaundice, but within the first two months of life, jaundice develops and stools become progressively acholic.
- **BA associated with laterality malformations** – This pattern is also known as **biliary atresia splenic malformation (BASM)** or "embryonal" biliary atresia and occurs in 10 to 15 percent of infants with **BA**. The laterality malformations include situs inversus, asplenia or polysplenia, malrotation, interrupted inferior vena cava, and cardiac anomalies.
- **BA associated with other congenital malformations** – This occurs in the remaining 5 to 10 percent of **BA** cases; associated congenital malformations include intestinal atresia, imperforate anus, kidney anomalies, and/or cardiac malformations.

KASAI CLASSIFICATION OF BA

Type I	Atresia at the level of the common bile duct (patent cystic and CHD)
Type IIa	Atresia is at the level of the common hepatic duct (patent cystic and CBD)
Type IIb	Atresia in the cystic duct, CBD and CHD
Type III	Is the most frequent type, atresia occurs at the porta hepatis



CLINICAL PRESENTATION

- Most infants with BA are born at full term, have a normal birth weight, and initially thrive and seem healthy.
- **Scleral icterus** and/or **generalized jaundice** typically develop by eight weeks of age.
- Infants may also have **acholic** (very pale-colored) stools, **dark urine**, and/or a **firm liver and splenomegaly**.

INVESTIGATIONS

1) Blood test :

- Elevation in serum conjugated bilirubin (>2 mg/dL [34 micromol/L])
 - High Aminotransferase and Alkaline phosphatase *due to Bone remodeling*
 - High GGT (Gamma-glutamyl transferase).
-
- **Note: LIVER FUNCTION TEST;**
 - - Alkaline phosphatase elevation in infants due to the contribution of bone remodeling.
 - - GGT is more specific for hepatobiliary disease.

INVESTIGATIONS

2) Abdominal ultrasound:

- The main utility of the ultrasound is to exclude other anatomic causes of cholestasis (ie, choledochal cyst, intrahepatic bile duct dilatation, and gallstones)
- several findings (if present) support the diagnosis of BA:
 1. **Absence of the gallbladder.** (Note: The absence of a gallbladder is highly suggestive of the diagnosis of BA, but the presence of a gallbladder does not exclude the diagnosis of biliary atresia.)
 2. **Irregularly shaped gallbladder.**
 3. **Polysplenia.**
 4. **Triangular cord sign.**

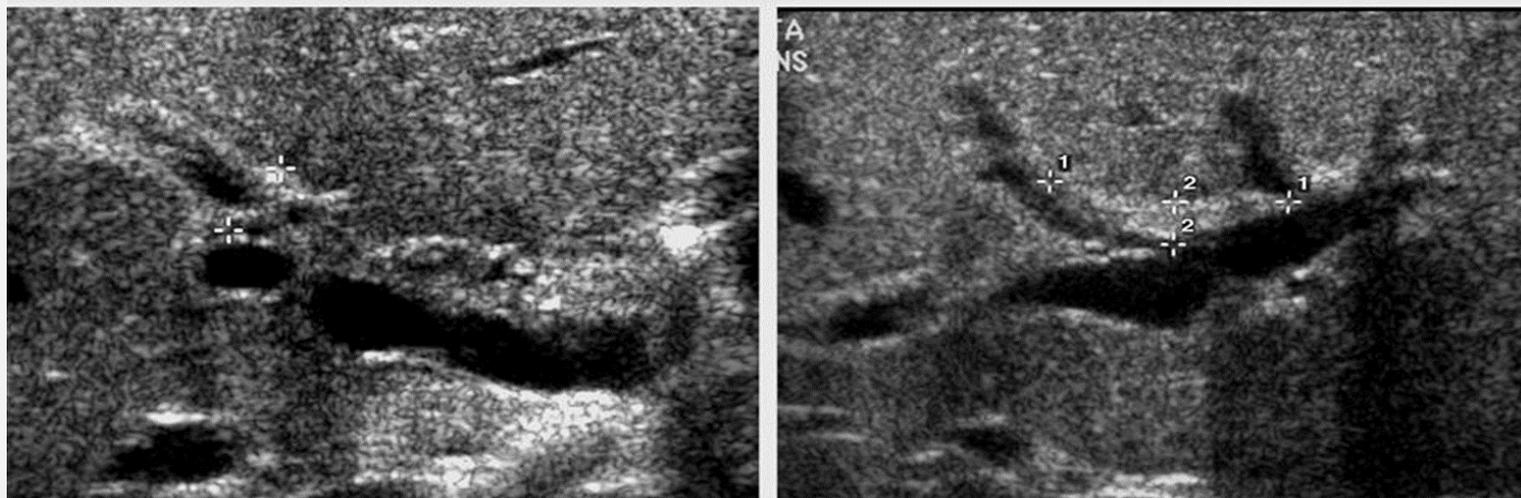
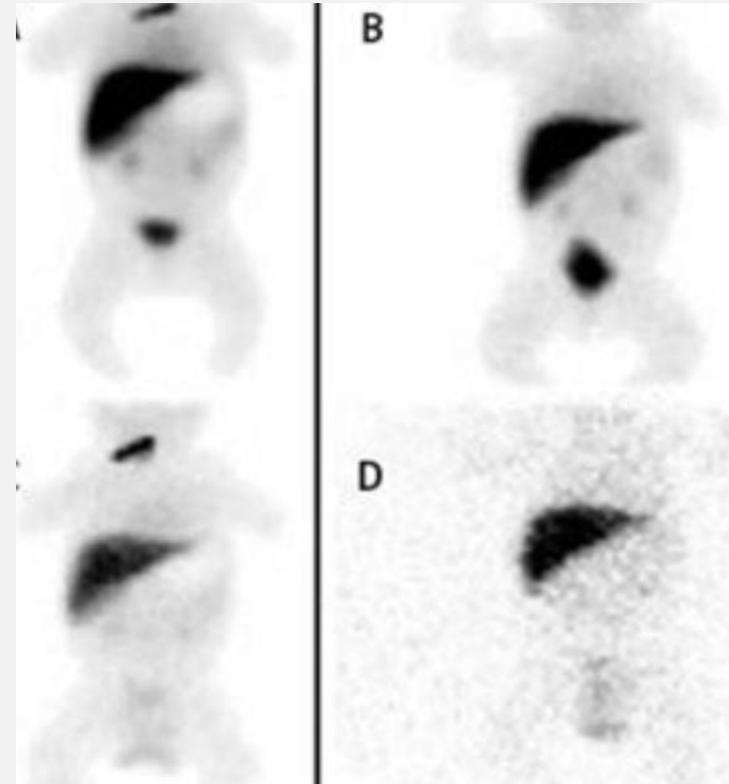


Figure 3. US images of a 35-day-old boy with biliary atresia. **(a)** Longitudinal scan shows TC sign (cursors) with a 5.1-mm-thick EARPV. **(b)** Transverse scan shows triangular echogenic area (cursors) in the anterior aspect of the portal vein in the porta hepatis.

3) Nuclear medicine scan and HIDA scan :

- The patient is given “hepatobiliary iminodiacetic acid”
If there is a good hepatic uptake with no evidence of excretion into the bowel during 24 hours (Excretion by alternate route the kidney and bladder) then it is biliary atresia.



4) Percutaneous Liver biopsy

- Active inflammation with bile duct degeneration and fibrosis
- Bile duct proliferation
- Portal stromal edema

6) Intraoperative cholangiography

- which is the gold standard in the diagnosis of BA. It is essential that patency be investigated both proximally into the liver and distally into the bowel to determine whether BA is present.
- If the intraoperative cholangiogram demonstrates biliary obstruction (ie, if the contrast does not fill the biliary tree or reach the intestine), the surgeon should perform a Kasai HPE at that time.
- In some cases, the cholangiogram cannot be performed, because the gallbladder and biliary tree are atretic; in this case, the surgeon makes the diagnostic decision based on visual inspection of the biliary tree.



DIAGNOSIS

- The possibility of BA is suggested by the clinical presentation of neonatal conjugated hyperbilirubinemia and/or acholic stools.
- The suspicion of BA is strengthened by the results of a variety of tests (typically ultrasound, hepatobiliary scan, and liver biopsy).
- The definitive diagnosis is made by an intraoperative cholangiogram.

DDX

- In **term infants**, BA and neonatal hepatitis account for a majority of cases of neonatal cholestasis.
- In **premature infants**, cholestasis more often results from total parenteral nutrition or sepsis.

TREATMENT

نربط الـ jejunum مع الـ liver

نصلها في حاله الـ intrahepatic
Before Liver Transplant

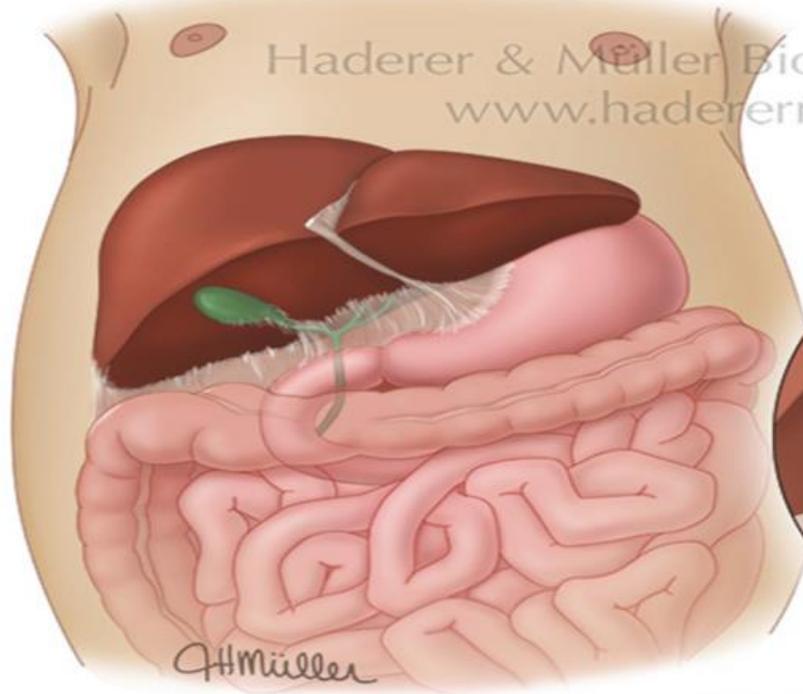
- If BA is confirmed by cholangiogram, a **Kasai procedure** (**hepatoportenterostomy [HPE]**) should be performed promptly. This operation is undertaken in an attempt to restore bile flow from the liver to the proximal small bowel.
- For this procedure, a **Roux-en-Y** loop of bowel is created by the surgeon and directly anastomosed to the hilum of the liver, following excision of the biliary remnant and portal fibrous plate.
- This surgery should be performed as soon as the diagnosis of BA can be made and preferably before **8 weeks** of age.
- Younger age at the time of the Kasai HPE is associated with better outcomes.

for extra Biliary atresia

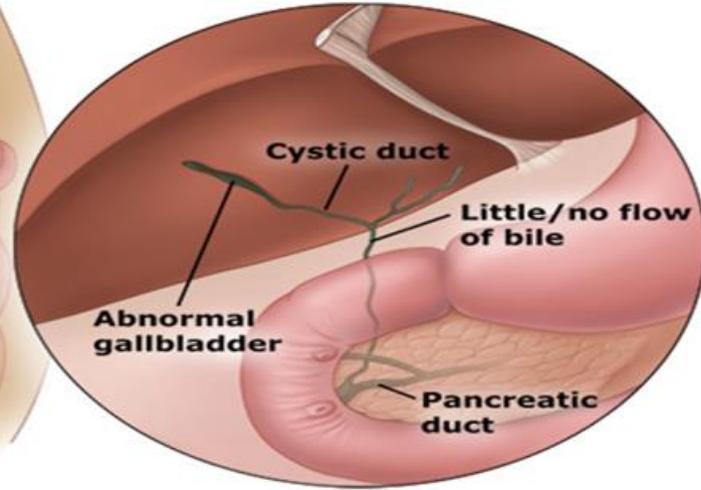
بس فيه طريقه
في المريء
anti reflux

نخلي الـ dringع ما يرجع
و بالتالي نقل الـ Cholangitis

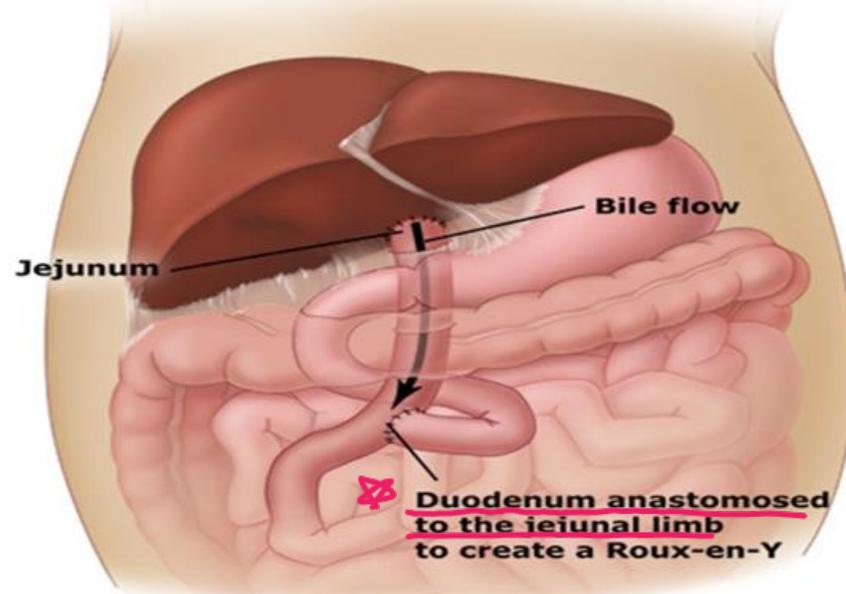
NORMAL



BILIARY ATRESIA



KASAI PROCEDURE



TREATMENT

- **Nutrition and monitoring :**
- Infants with BA who are jaundiced should be treated with supplements of fat-soluble vitamins and monitored for fat-soluble vitamin deficiencies. In addition, they should be given high-calorie formulas or other nutritional supplements as required to sustain normal rates of growth.
- **Medications:**
- It is suggested that infants and children be treated with ursodeoxycholic acid (UDCA) after Kasai HPE.
- It is also suggested to treat all patients with prophylactic antibiotics for at least the first year after Kasai HPE to reduce the risk of ascending cholangitis. Repeated episodes of cholangitis hasten the progression of liver disease.

TREATMENT

- **Liver transplantation:**
- Patients with persistent jaundice three months after Kasai HPE should be referred for liver transplant evaluation. Other indications for early liver transplantation include failure to thrive despite vigorous nutritional rehabilitation, complications of portal hypertension (recurrent variceal bleeding or intractable ascites), and progressive liver dysfunction.
- **Prognosis:**
- At least 60 to 80 percent of patients with BA will eventually require liver transplantation. A minority of patients with BA treated with Kasai HPE survive to 20 years or more without liver transplantation. However, many of these patients have chronic liver disease with cirrhosis and portal hypertension.

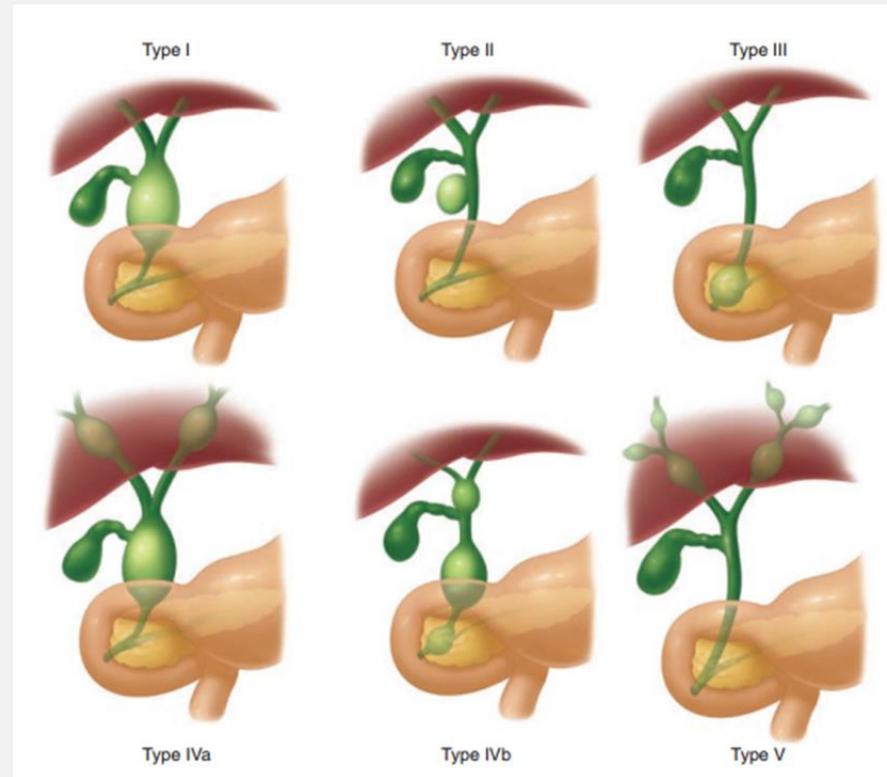
CHOLEDOCHAL CYSTS

EPIDEMIOLOGY & ETIOLOGY

- Is a rare congenital condition that causes abnormal dilatation of the bile ducts leading to obstruction and other complications.
- more common in Asian and females (F: M ratio 4:1).
- The majority (60%) were diagnosed in the first decade of life and others later. While (20%) of cases are diagnosed prenatally.
- Cause: the exact cause is unknown, but theories suggest that it may be due to **Pancreaticobiliary Malunion (PBMU)**.

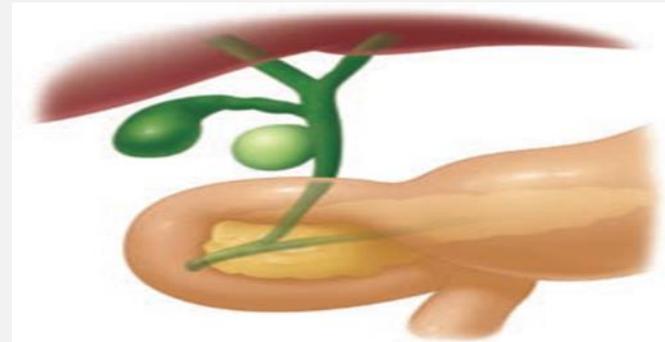
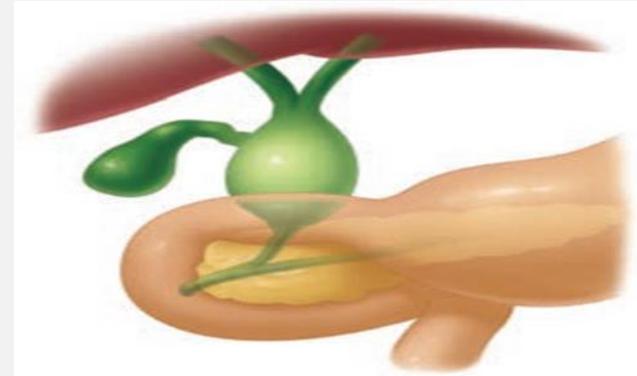
ANATOMIC CLASSIFICATION

- It's classified into 5 types :

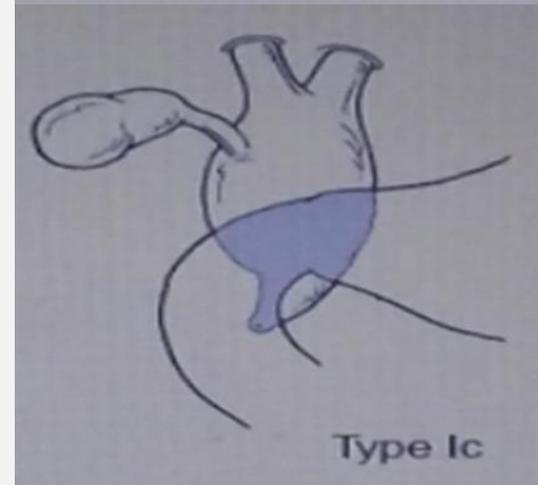
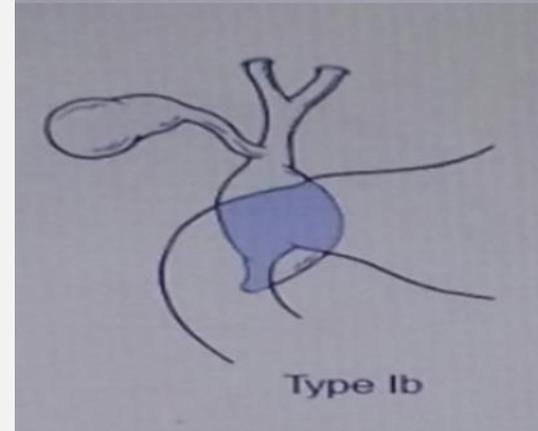
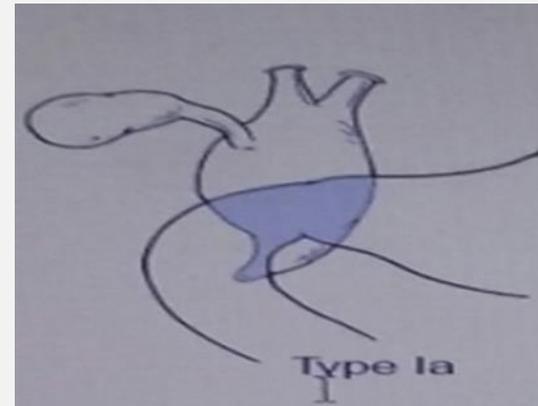


ANATOMIC CLASSIFICATION

- **Type 1 :**
 - It is the most common (>50% of cases).
 - **Cystic/saccular** or **fusiform** dilation
 - of the CBD (**extrahepatic**).
- **Type 2 :**
 - It is a true, single, **extrahepatic diverticulum**

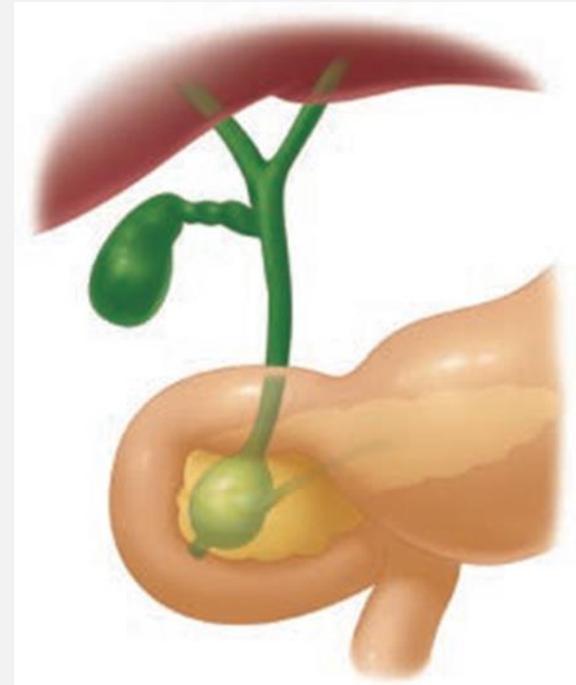


- **Type Ia** consists of cystic dilation of the **entire** common bile duct.
- **Type Ib** is cystic dilation of a **segment** of the common bile duct.
- **Type Ic** is **fusiform** dilation of the common bile duct.



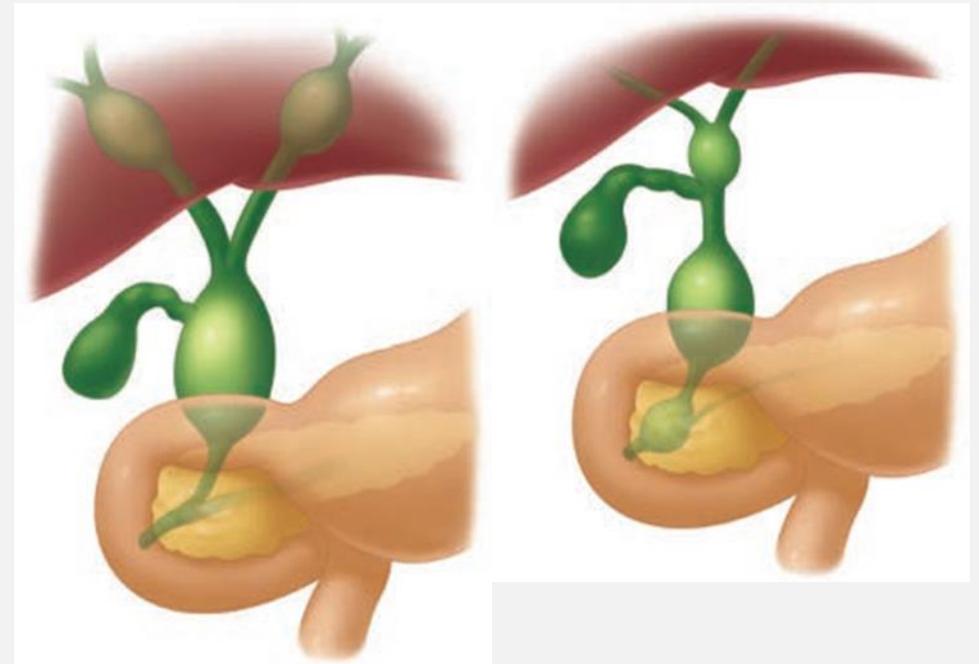
ANATOMIC CLASSIFICATION

- **Type 3 :**
- Also referred to as a **choledochocele** , usually has a normal CBD and main pancreatic duct with cystic dilation of the distal CBD that is either **intraduodenal** or **intrapancreatic** in location.
- The **choledochocele** is usually stenotic at their openings due to chronic inflammation.



ANATOMIC CLASSIFICATION

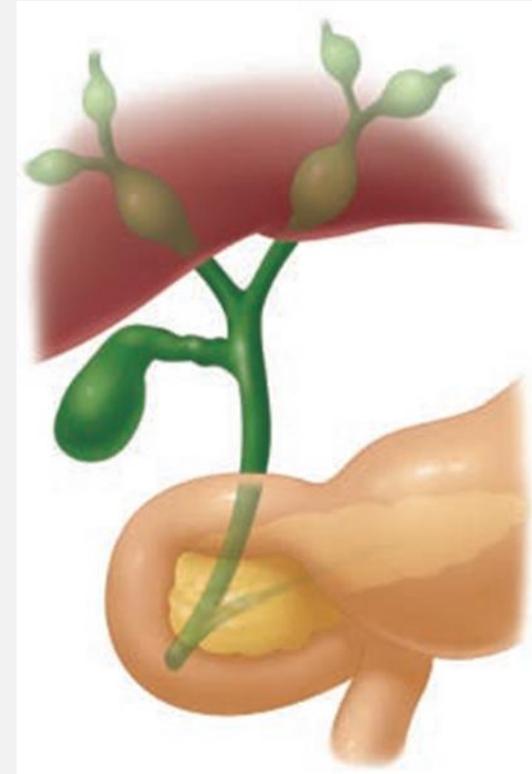
- **Type 4:** Multiple cysts.
- **Type 4A:** Is composed of **multiple** cysts located in both **intrahepatic** and **extrahepatic** bile ducts.
- **Type 4B:** Is composed of multiple cysts located **extrahepatic** bile duct only.



ANATOMIC CLASSIFICATION

- **Type 5 :**
- **Intrahepatic** single or multiple cysts only **without** extrahepatic dilation.
- Type V cysts in conjunction with hepatic fibrosis are commonly referred to as Caroli syndrom.

نمط ٥
liver transplant
لا عند
fibrosis



CLINICAL PRESENTATION

- **Prenatal:** Cystic mass in the abdomen.
- **In infant:** obstructive Jaundice, pale (acholic) stool with/without hepatomegaly

CLINICAL PRESENTATION

- **In children:** “ Classic triad “
jaundice, pain & palpable mass.
- The entire triad is present in fewer than 30% of patients
- **In adults:** recurrent ascending cholangitis, obstructive jaundice or recurrent pancreatitis.

CLINICAL PRESENTATION

- Undiagnosed choledochal cysts can lead to choledocholithiasis, cholangitis, obstructive jaundice, cirrhosis with portal hypertension, cyst rupture, or biliary carcinomas.

WORK UP

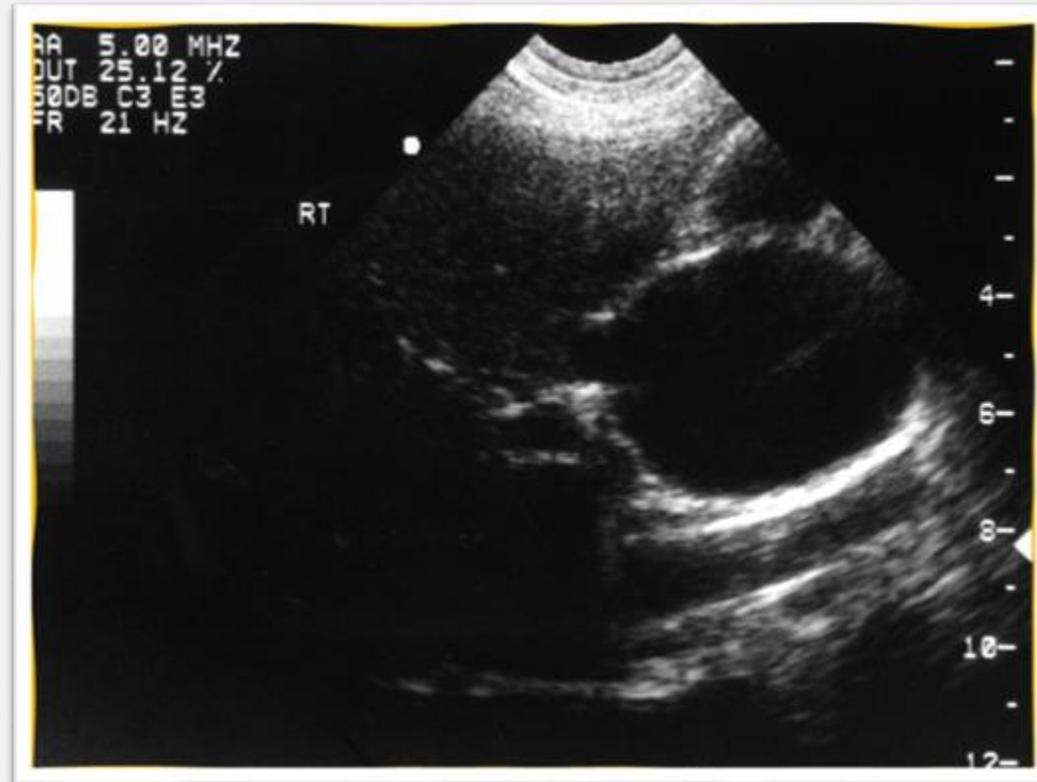
1) Laboratory studies:

- direct bilirubin = jaundice
- alkaline phosphatase = liver damage
- (AST) = liver damage
- (ALT) = liver damage
- (GGT) = liver damage \ damage of bile duct
- Coagulation profiles.
- (CBC) = anemia

WORK UP

2) Imaging Studies:

- **Ultrasound:** Best initial study.
- **HIDA scan:** if an anomaly was detected prenatally
- **MRCP:** Gold Standard diagnostic test.
- **CT:** first study in adults



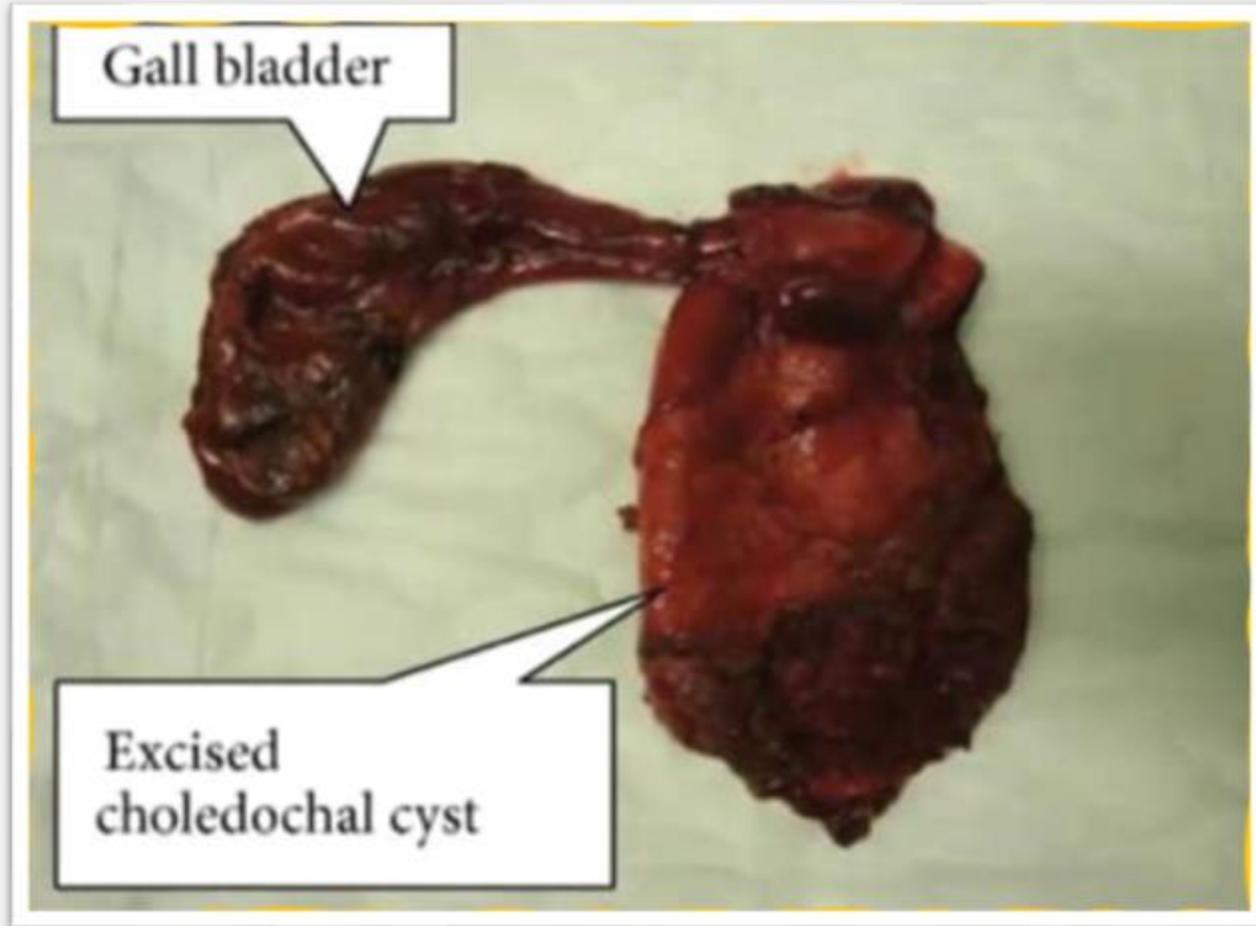
Ultrasound

Ultrasonogram shows a large unilocular cyst under the liver.

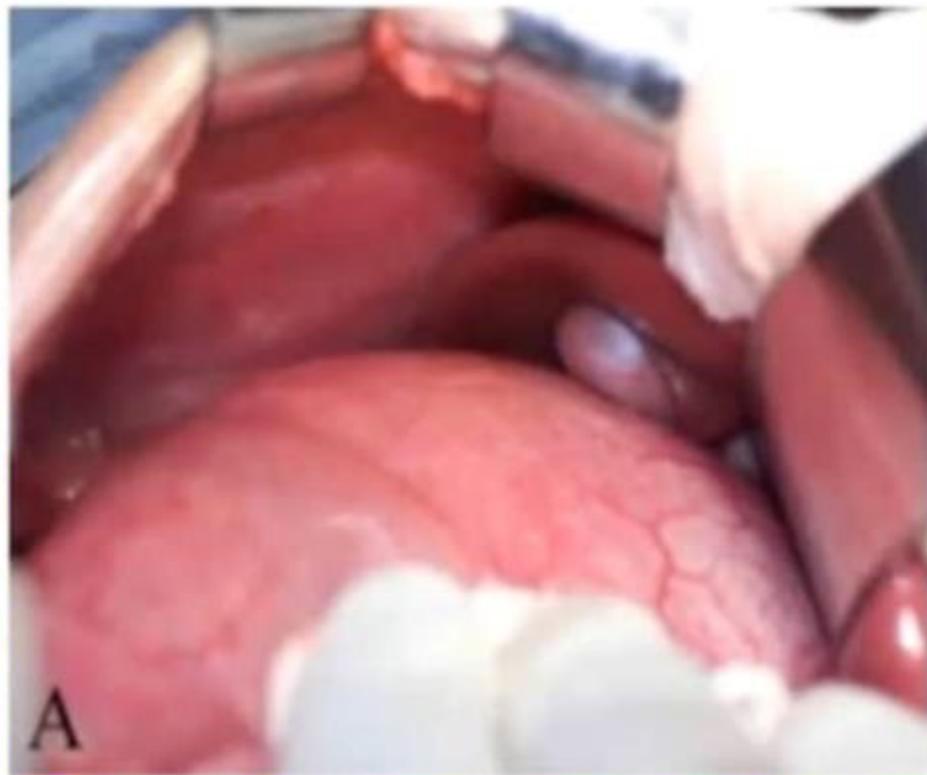


MRCP

shows dilated hepatic ducts and common bile duct (CBD) of a **type IV** cyst.

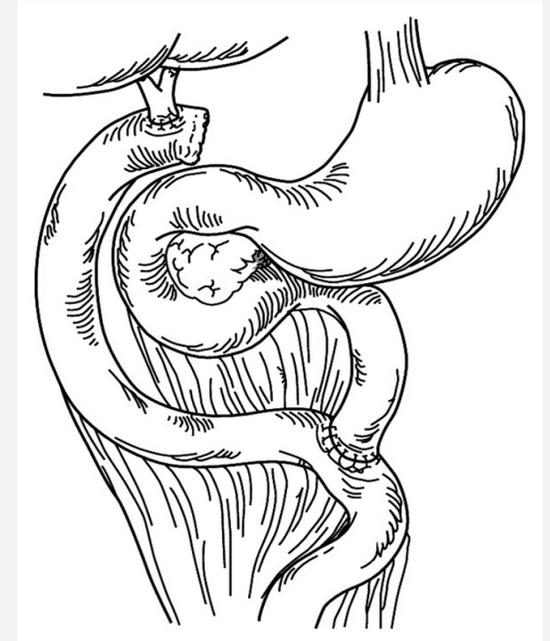


Complete excision with construction of biliary-enteric anastomosis



TREATMENT

- Treatment of choledochal cysts is **surgical**, except for **type V** multiple intrahepatic cysts, which may benefit from **conservative therapy** (including percutaneous drainage and medical management) for variable periods of time.
- **Total excision** of the cyst with adequate bile drainage is the standard treatment.
- **Total excision** of the cyst in **types I, II, and IV** followed by reconstruction of the biliary tree with **hepaticojejunostomy in a Roux-en-Y** fashion is the procedure of choice in treating choledochal cysts



TREATMENT

- **Intraoperative cholangiography** obtained via puncture of the cyst or via the gallbladder is always obtained. It outlines the exact anatomy of the choledochal cyst and its relationship with the pancreas. **Cholecystectomy is routinely performed at the same time.**

COMPLICATIONS

- Ascending Cholangitis
- pancreatitis
- Biliary stone formation
- Anastomotic stricture
- Bowel obstruction
- Malignancy: arise from the distal CBD

THANK YOU!