APPROACH TO A CHILD WITH JAUNDICE AND ASCITIS

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LEARNING OBJECTIVES

What is jaundice

Basic patho -physiology of bilirubin metabolism

Causes of jaundice

Approach to a child with jaundice

Ascites

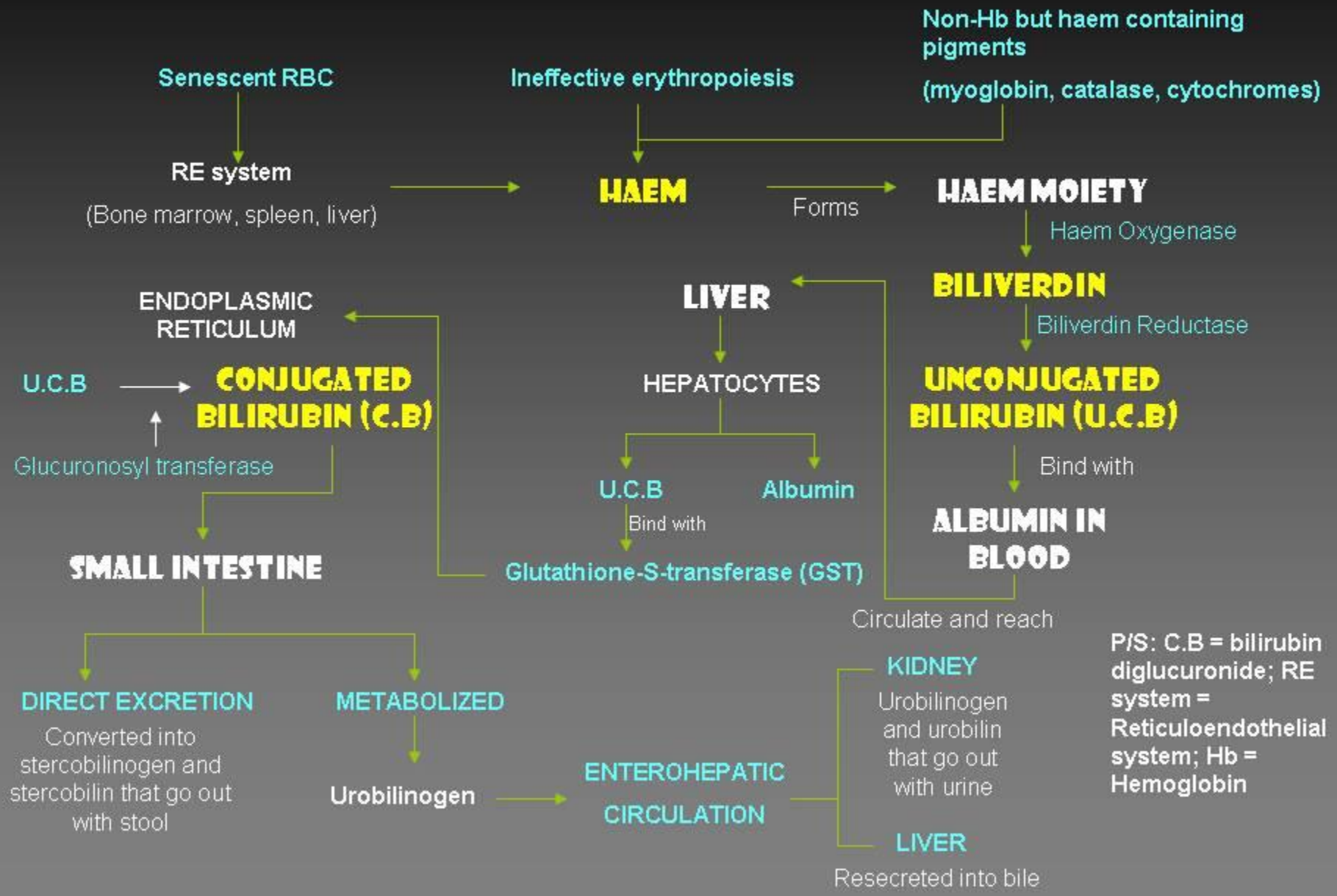
INTRODUCTION

- Symptom of disease rather than a disease
- In adults and older children sclera appears jaundiced when serum bilirubin is increased
- Gives yellowish hue to the skin, sclera, and mucous membranes
- Normal serum bilirubin <1mg%
- It is not a visible till s. bilirubin exceeds 2 mg/dl.
- In newborn->5 mg/dl
- However it is difficult to see sclera in newborn due to difficulty in opening eye

BILIRUBIN

- End product of hemoglobin metabolism that is excreted in bile.
- It comes from
  - from catabolism of circulating RBCs
  - from ineffective erythropoiesis (bone marrow)
  - from turnover of heme proteins

NORMAL BILIRUBIN METABOLISM



P/S: C.B = bilirubin diglucuronide; RE system = Reticuloendothelial system; Hb = Hemoglobin

What causes ↑ bilirubin?

1. Overproduction by reticuloendothelial system
2. Failure of hepatocyte uptake
3. Failure to conjugate or excrete
4. Obstruction of biliary excretion into intestine

Normal Range of Bilirubin

It is normal to have some bilirubin in your blood. Normal levels are:

- *Direct (also called conjugated) bilirubin: 0 to 0.3 mg/dL*
- *Total bilirubin: 0.3 to 1.9 mg/dL*

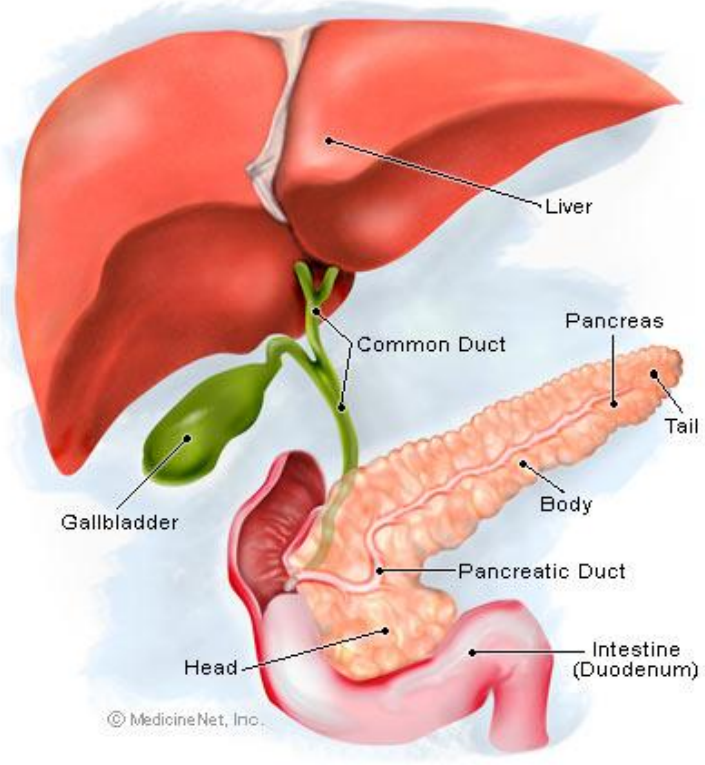
Causes of Jaundice

Jaundice occurs when there is:

- too much bilirubin being produced for the liver to remove from the blood (for example, patients with hemolytic anemia have an abnormally rapid rate of destruction of their red blood cells that releases large amounts of bilirubin into the blood)

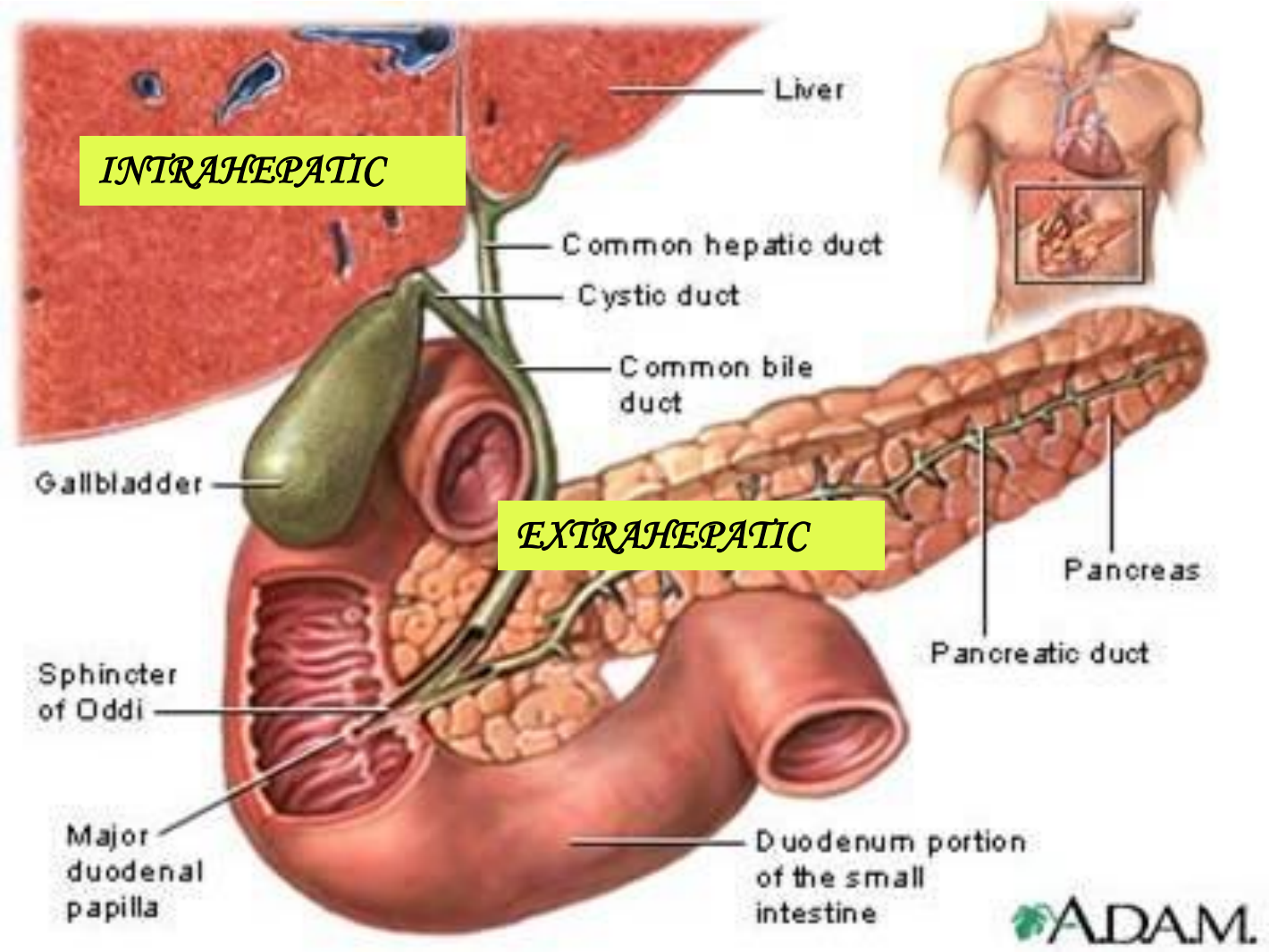
- a defect in the liver that prevents bilirubin from being removed from the blood, converted to bilirubin/glucuronic acid (conjugated) or secreted in bile; or

- blockage of the bile ducts that decreases the flow of bile and bilirubin from the liver into the intestines. For example, the bile ducts can be blocked by worms, cancer, gallstones, or inflammation of the bile ducts. The decreased conjugation, secretion, or flow of bile that can result in jaundice is referred to as cholestasis：however, cholestasis does not always result in jaundice.



❖ *Obstructive Jaundice*

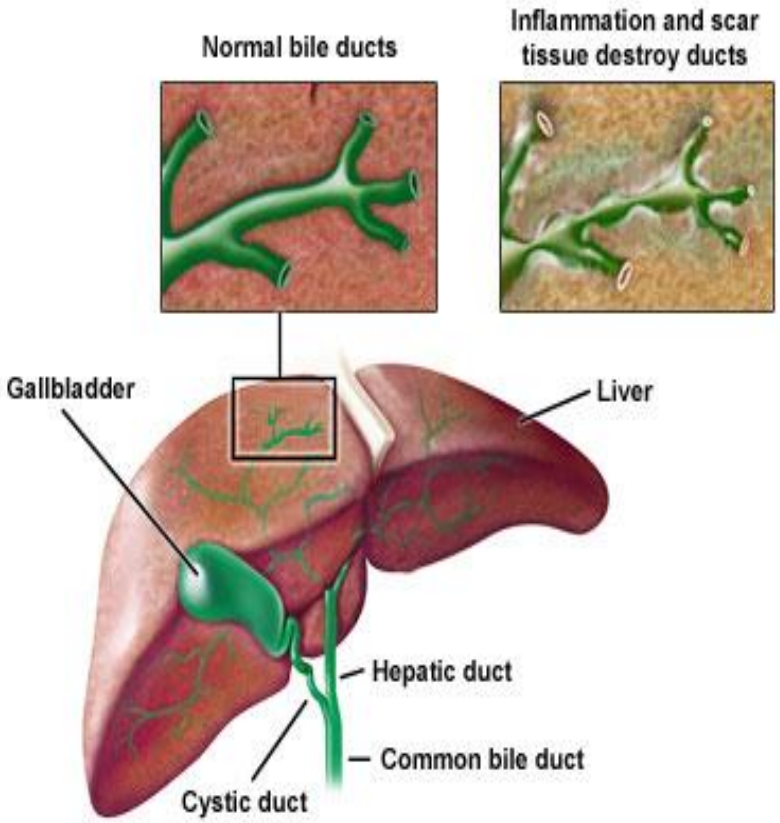
Obstructive jaundice is a condition in which there is blockage of the flow of bile out of the liver



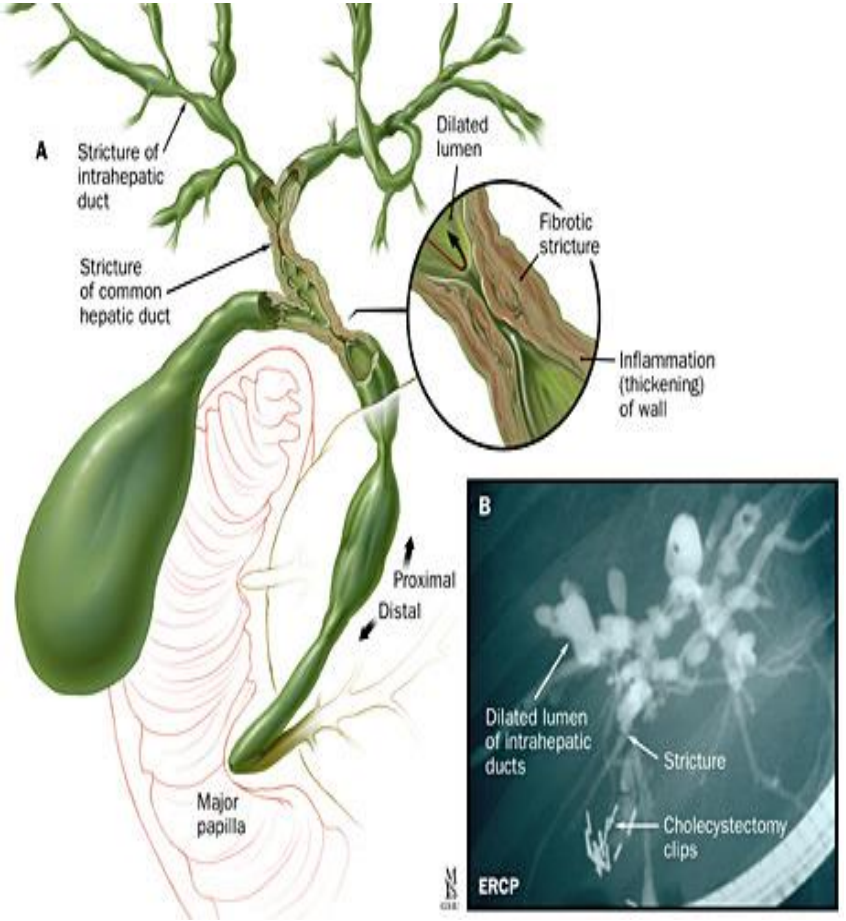
CAUSES OF OBSTRUCTIVE JAUNDICE: INTRAHEPATIC

Primary biliary cirrhosis

Sclerosing cholangitis (Inflammation/scarring)



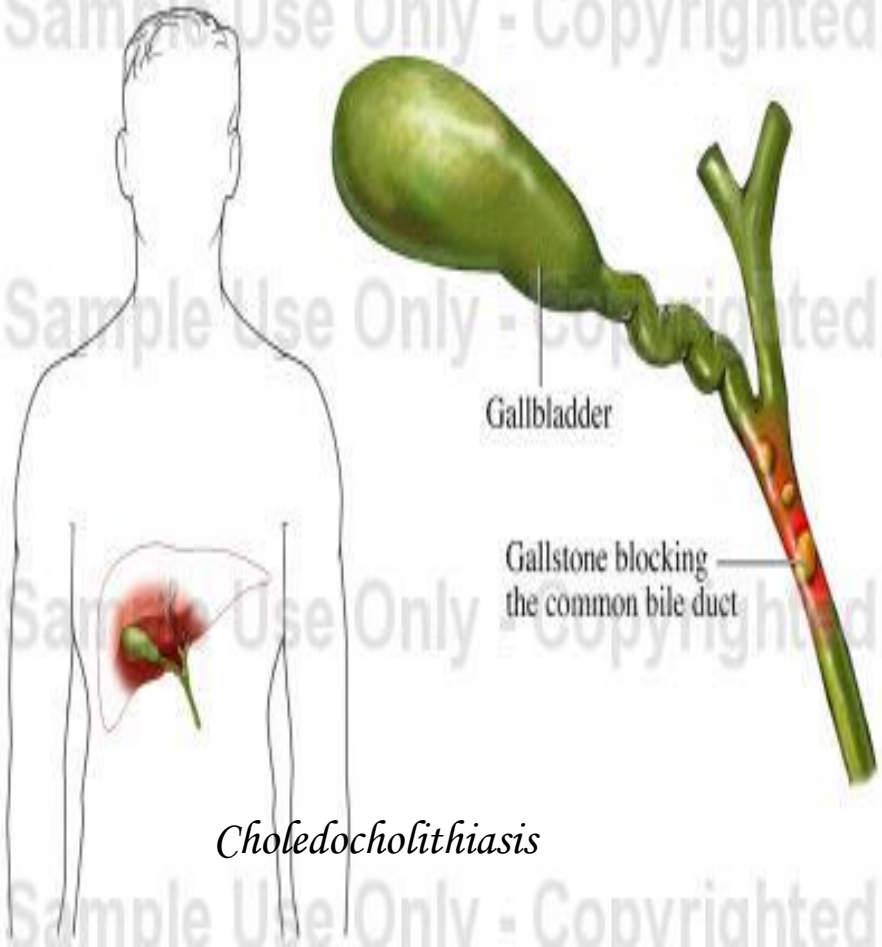
Primary biliary cirrhosis



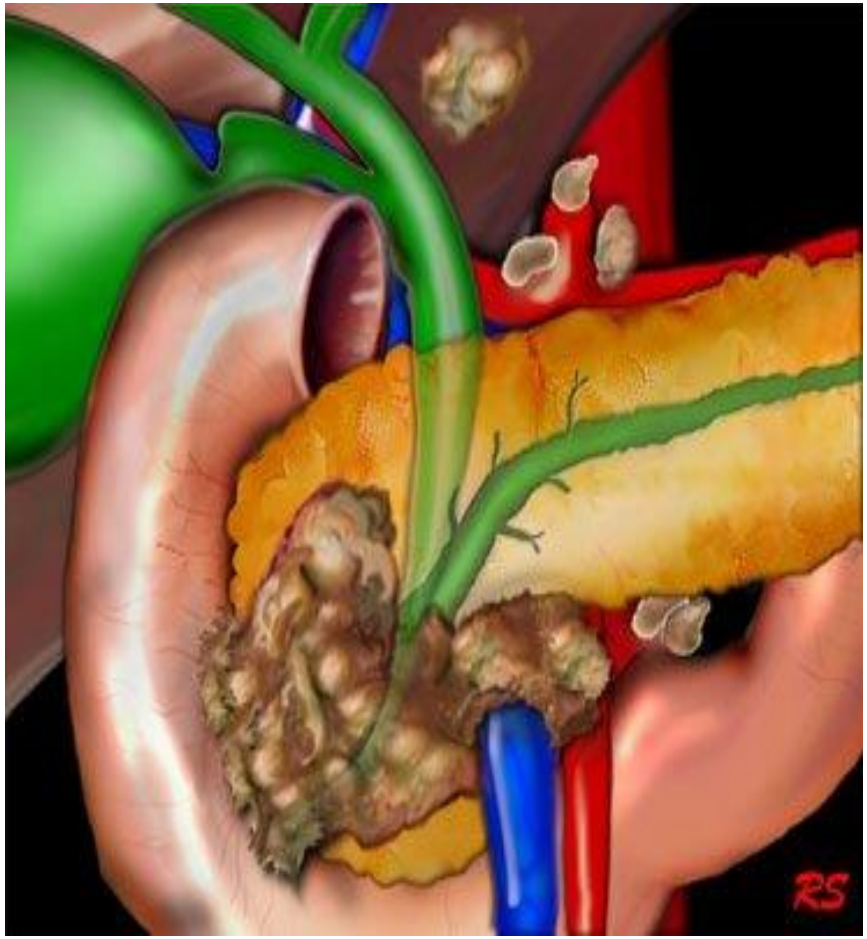
Sclerosing cholangitis (Inflammation/scarring)

CAUSES OF OBSTRUCTIVE JAUNDICE: EXTRAHEPATIC

- Choledocholithiasis
- Worms
- Malignancy : neoplasia, L.N.



Choledocholithiasis



Malignancy : Pancreatic (head of pancreas) carcinoma

Table 2. Differential Diagnosis Of Jaundice In Children

Unconjugated Hyperbilirubinemia

Increased bilirubin production

- Hemolysis: blood group incompatibility, G6PD, sickle cell disease

Decreased excretion/conjugation

- Crigler-Najjar syndrome (life-threatening) types I and II
- Congenital hypothyroidism
- Gilbert's syndrome
- Drugs: ketoconazole, ethinyl estradiol, amitriptyline, and HIV protease inhibitors

Impaired uptake

- Congestive heart failure
- Portosystemic shunts
- Drugs: rifampin, probenecid

Hyperbilirubinemia due to breastfeeding

- Breastfeeding jaundice
- Breast milk jaundice

Conjugated Hyperbilirubinemia

Extrahepatic cholestasis

- Choledocholithiasis, biliary sludge, inspissated bile syndrome
- Intrinsic and extrinsic tumors: cholangiocarcinoma
- Primary sclerosing cholangitis
- Acute and chronic pancreatitis
- Strictures after invasive procedure
- Parasitic infections: ascaris, lumbricoides, liver flukes
- Extrahepatic biliary atresia
- Choledochal cyst
- Alagille syndrome
- Cystic fibrosis
- Neonatal sclerosing cholangitis
- Congenital hepatic fibrosis/Caroli disease
- Spontaneous perforation of the bile ducts

Intrahepatic cholestasis

- Idiopathic neonatal hepatitis
- Sepsis and hypoperfusion states
- Bacterial infection: urinary tract infection, sepsis, syphilis
- Protozoal infection: toxoplasmosis
- Viral hepatitis infection: cytomegalovirus, HIV, HSV, rubella, parvovirus B19, ECHO virus, adenovirus
- Genetic/metabolic disorders: α1-antitrypsin deficiency, nonsyndromic paucity of bile ducts, disorders of bile acid synthesis, hypothyroidism, PFIC, cystic fibrosis, panhypopituitarism, neonatal hemochromatosis
- Carbohydrate disorders: tyrosinemia, galactosemia, fructosemia
- Lipid disorders: Wolman disease, Niemann-Pick disease, Gaucher disease
- Primary biliary cirrhosis
- Drugs/toxins:alkylated steroids, chlorpromazine, herbal medications (eg, Jamaican bush tea), arsenic
- Pregnancy
- Infiltrative diseases: lymphoma, tuberculosis
- Hepatic crisis in sickle cell disease

Toxic

- Total parenteral nutrition-associated cholestasis

Abbreviations: ECHO, enteric cytopathic human orphan; G6PD, glucose-6-phosphate dehydrogenase; HIV, human immunodeficiency virus; HSV, herpes simplex virus; Progressive Familial Intrahepatic Cholestasis, PFIC.

AN APPROACH TO A CHILD WITH JAUNDICE

CLASSIC APPROACH

- Proper detailed history
- Proper physical examination
- Appropriate investigations

IDENTIFY

Acute

Chronic (more than 6 months)

IDENTIFY

Hemolytic

Hepatocellular

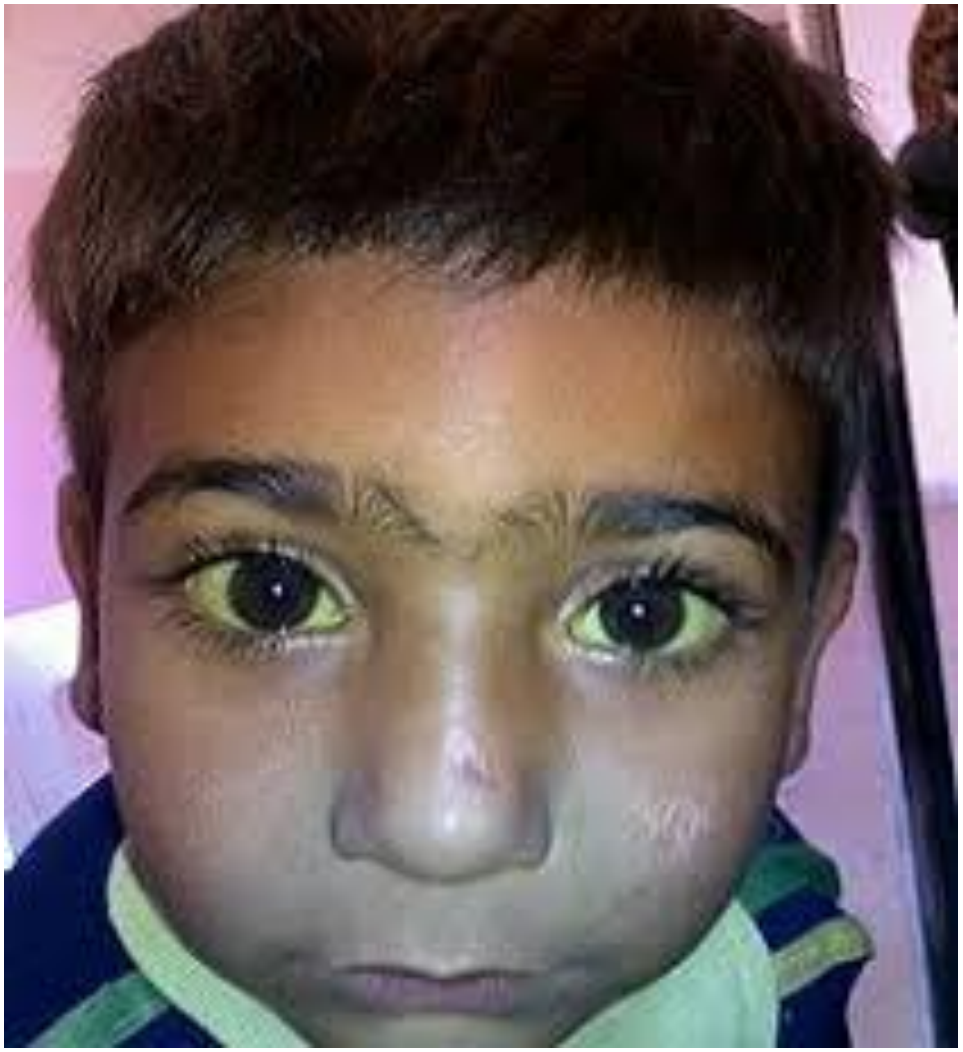
Cholestatic

TYPES OF JAUNDICE

TYPE	PRE HEPATIC	HEPATIC	POST HEPATIC
Urine color	normal	dark	dark
Stool color	normal	normal	acholic
Pruritis	no	No	yes

Signs and Symptoms of Jaundice

1. *yellow discoloration of the skin, mucous membranes, sclera of the eyes*
2. *light-colored stools*
3. *dark-colored urine*
4. *itching of the skin.*
5. *nausea and vomiting*
6. *abdominal pain*
7. *fever*
8. *weakness*
9. *loss of appetite*
10. *headache*
11. *confusion*
12. *swelling of the legs and abdomen*
13. *Skin stigmata*



Diagnosis of Jaundice

The health care provider will perform a physical exam. This may reveal liver swelling.

- *A bilirubin blood test will be done.*

Other tests vary, but may include:

- *Hepatitis virus panel to look for infection of the liver*
- *Liver function tests to determine how well the liver is working*
- *Complete blood count to check for low blood count or anemia*
- *Abdominal ultrasound*
- *Abdominal CT scan*
- *Endoscopic retrograde cholangiopancreatography (ERCP)*
- *Percutaneous transhepatic cholangiogram (PTCA)*
- *Liver biopsy*
- *Cholesterol level*
- *Prothrombin time*

IN CHILDREN

Hepatocellular (SGOT/SGPT more than twice of ALP)

Cholestatic (SGOT/SGPT less than twice of ALP)

Table of diagnostic tests

Function test	Pre-hepatic Jaundice	Hepatic Jaundice	Post-hepatic Jaundice
Total bilirubin	Normal / Increased	Increased	
Conjugated bilirubin	Normal	Increased	Increased
Unconjugated bilirubin	Normal / Increased	Increased	Normal
Urobilinogen	Normal / Increased	Increased	Decreased / Negative
Urine Color	Normal	Dark (urobilinogen + conjugated bilirubin)	Dark (conjugated bilirubin)
Stool Color	Normal	Normal/Pale	Pale
Alkaline phosphatase levels	Normal	Increased	
Alanine transferase and Aspartate transferase levels		Increased	
Conjugated Bilirubin in Urine	Not Present	Present	
Splenomegaly	Present	Present	Absent

REMEMBER

The prognostic value of

- Albumin
- Coagulation profile

NEONATAL JAUNDICE

- Jaundice is clinically detectable in the newborn when the serum bilirubin levels are greater than 5 mg/dl. This occurs in approximately 60% of term infants and 80% of preterm infants.

- Neonatal jaundice first becomes visible in the face and forehead. Blanching reveals the underlying color. Jaundice then gradually becomes visible on the trunk and extremities.

Signs and Symptoms of Neonatal Jaundice

Newborns, as the bilirubin level rises, jaundice will typically progress from the head to the trunk, and then to the hands and feet. Additional signs and symptoms that may be seen in the newborn include:

1. poor feeding
2. lethargy
3. changes in muscle tone
4. high-pitched crying
5. seizures.

ASCITES

DEFINITION

Accumulation of serous fluid in peritoneal cavity

- As a part of generalized edema- anasarca
- Isolated collection or disproportionate

CAUSES

1. **Isolated or disproportionate**

- **Hepatic**
  - Cirrhosis
  - Congenital hepatic fibrosis
  - Portal vein obstruction
  - Budd chiari syndrome
  - Neonatal cholestatis
- **Abdominal**
  - Peritoneal Tuberculosis
  - Acute pancreatitis
- **Renal**
  - Peritoneal dialysis
  - Obstructive uropathy
- **Cardiac**
  - Constrictive pericarditis
- **Neoplastic**
  - Lymphoma
  - Neuroblastoma
- **Urinary**
  - Perforation
  - Leakage from urinary tract
- **Chylous ascites**
- **Gynecological**
  - Ovarian tumor
  - Ovarian rupture

2. Ascitis with generalized edema

- **Renal-** Nephrotic syndrome, AGN, renal failure
- **Cardiac-** CHF, constrictive pericarditis
- **Polyserositis-** SLE, Dengue fever, sepsis
- **Severe Malabsorption**

CLINICAL FEATURE

Abdominal distension-hallmark

Five classical signs

- Bulging vein
- Flank dullness and fullness
- Shifting dullness
- Fluid thrill
- Puddle sign

Umbilical laughing/ herniation with tense ascites

D/D

- Gaseous distention
- Fecal retention
- Masses
- Obesity
- Pregnancy

EVALUATION OF CAUSE

- Generalized/ isolated or predominant
- Predominant- hepatic or intra-abdominal
- Age- Neonate- urinary, chylous

  Infancy- cholestatic
- Look for signs and symptoms of hepatic disease
- H/o contact for TB with pulmonary findings
- Presence of L. N.

ASCITIC FLUID ANALYSIS

- Transudative/ exudative (by SAAG –serum albumin-ascetic fluid albumin gradient)
- SAAG- ≥1.1g/dl-transudative

  <1.1g/dl- exudative
- Transudative- CLD and when ascites is a part of generalized edema
- Ascitic fluid- cytology, gram staining, culture

characteeristic	disease
Lymphocytic pleocytosis	tuberculosis
Polymorphic pleocytosis	Bacterial perotonitis
hemorrhagic	Malignancy, pancreatitis, tuberculosis
Milky white	Chylous ascitis

OTHER INVESTIGATIONS

- Ultrasonography– quantity, etiology, L.N., hepatic echotexture, size of portal vein
- Portal venous Doppler studies
- CT Abdomen-intra abdominal mass, malignancy etc.
- LFT
- UGI endoscopy- CLD and PH
- Chest X-ray
- Mantoux test

TREATMENT

- Low salt diet
- Diuretics
- IV albumin
- Repeated large volume paracentesis
- Depend on cause of ascites
  - ATT- TB
  - Antibiotics- bacterial infection
  - Interferons- Heptitis B and C
  - Steroids- autoimmune hepatitis
  - Surgery or propranolol- PH and Varices
  - Liver transplantation- decompensated liver, cirrhosis, portal hypertension

MCQ 1

- Regarding bilirubin metabolism, which of the following is true?

a) Normal elimination is through the urine and the stool

b) Serum bilirubin concentration is not influenced by medications

c) Bilirubin is primarily free in circulation

d) Heme protein is primarily broken down in circulation

MCQ 2

- Followings are alarming signs in a patient with jaundice, except:

a) Altered sensorium

b) Raised INR

c) Raised ALT

d) Persistent vomiting


Thank you