Jaundice & Hepatosplenomegally

Professor Dr. Md. Aminul Islam
Professor, Dept. Of Medicine
CBMCB

Definition

- Yellow appearance of the skin, sclera and mucous membranes
- Resulting from an increased bilirubin concentration in the body

When Jaundice is clinically detectable

- **When serum bilirubin is > 3mg/dl (>50mmol/L)**
- **Best seen in daylight**

Why jaundice is best seen in sclerae

- **Because sclerae have a particular affinity for bilirubin due to their high elastin content**

Other sites to look for jaundice

- **Underneath the tongue**
- **Skin**

D/D of yellow coloration of skin

- Carotenaemia/ carototenoderma
- Use of the drug qunacrine
- Excessive exposure to phenol

Billirubin production and metabolism

- **Sources of unconjugated billirubin**
  - **Haemoglobin breakdown**
  - **Catabolism of other haem containing protein e.g. myoglobin and cytochrome enzymes**
  - **Ineffective erythropoiesis**

Measurement of serum billirubin

- **Types: Based Vanden Bergh reaction**
  - **Direct**
  - **Indirect**
- **Direct fraction provides as approximate estimation of conjugated bilirubin in serum**
- **Indirect fraction provides an estimate of unconjugated bilirubin in serum**

Measurement of urine bilirubin …

- **Unconjugated bilirubin**
  - **bound to albumin in the serum**
  - **not found in the urine**
- **Conjugated bilirubin**
  - **Always filtered at the glomerulus**
  - **Majority is reabsorbed from the tubules**
  - **Small fraction is excreted in the urine**

Mechanism of jaundice production

- Increased production of bilirubin
  - Haemolysis
- Impaired excretion of bilirubin
  - Congenital non-haemolytic hyperbilirubinaemia
    - Gilbert's syndrom
    - Najjar syndrome type I, type II
    - Dubin – Jhonson Syndrome
  - Hepatocellular jaundice
  - Cholestasis

The evaluation of jaundice

- **Whether the hyperbilirubinaemia is conjugated or unconjugated**
- **Whether other LFT are abnormal**

If a patient has isolated elevation of serum bilirubin Think of the following -

- **Unconjugated hyperbilirubinaemia**
  - **Haemolytic disorder**
  - **Ineffective erythropoiesis**
    - **In iron, folate, B_{12} deficiencies**
  - **Inherited conditions**
    - **Gilbert's syndrome**
    - **Crigler-Najjar syndrome type-I,II**
- **Conjugated hyperbilirubinaemia**
  - **Inherited conditions**
    - **Dubin-Johnson syndrome**
    - **Rotor syndrome**

If a patient has raised serum bilirubin with other LFT abnormalities consider:-

- **Hepatocellular disease**
- **Intra or extrahepatic cholestasis**

Haemolytic jaundice

- Results from increased destruction of RBC or their precursors in the marrow
- Jaundice due to haemolysis is mild
- Causes
  - Haemolytic aneamia due to any cause

Haemolytic jaundice…

- **Clinical features**
  - **Anaemia**
  - **Jaundice (mild)**
  - **Splenomegaly**
  - **Normal colored stool and urine**

Triad of haemolytic anaemia

Haemolytic anaemia

- **Investigation**
  - **Plasma bilirubin usually < 6 mg/dl**
  - **Indirect Vanden Bergh reaction**
  - **Liver enzymes normal**
  - **No bilirubin, ⇧ urinary uroblinogen**
  - **Serum haptoglobin low**
  - **PBF: features of haemolytic anaemia**

Congenital non-haemolytic hyperbilirubinaemia

- **Gilbert's syndrome**
- **Other forms are very rare**

Gilbert's syndrome

Causes

- ⇩ Glucoronyl transferase

  ⇩

- ⇩ Conjugation of bilirubin
- Asymptomatic & is usually detected incidentally
- Bilirubin level increase during fasting and fail during treatment with phenobarbital
- Excellent prognosis, needs no treatment

Hepatocellular jaundice

Causes:

- **Inability of the liver to transport billirubin into the bile**

Most common causes

- **Acute viral hepatitis (A,B,C,D,E viruses)**
- **Drug induced hepatotoxicity**
- **Alcoholic hepatitis**

Hepatocellular jaundice …

Investigation

- **⇧ conjugated and unconjugated bilirubin**
- **⇧ AST, ALT**

Cholestasis jaundice

- **Pathogenesis**
  - **Failure of conjugated bilirubin to enter bile canaculi**
  - **Obstruction of bile flow in the extrahepatic bile ducts**
- **Types**
  - **Intra-hepatic cholestasis**
  - **Extra-hepatic cholestasis**

Cholestatic jaundice: Causes

- **Intra-hepatic cholestasis**
  - **Primary biliary cirrhosis**
  - **Viral hepatitis**
  - **Alcohol**
- **Extra- hepatic cholestasis**
  - **Choledocholithiasis**
  - **Carcinoma of the head of the pancreas**
  - **Stricture of bile duct, etc**

Cholestatic Jaundice: CF

- Jaundice
- Dark (yellow) urine
- Pale stool
- Palpable GB in Ca-head of the pancreas

Cholestatic jaundice…

- **Investigations**
  - **Serum billirubin : raised**
  - **Alkaline phosphatase: Markedly raised**
  - **AST & ALT : slightly raised**
  - **USG, CT abdomen, ERCP etc according to clinical suspicion**
- **Treatment**
  - **According to cause**

Hepatosplenomegaly: Common causes

- Chronic kala-azar
- Chronic malaria
- Hereditary haemolytic anaemia
- Lymphoma
- Leukaemia
- CLD with portal hypertension
- Infectious mononucleosis
- Disseminated tuberculosis

Hepatosplenomegaly

- Things to search for in a patient
  - Lymph node (lymphoma, CLL)
  - Stigmata of chronic liver disease
  - Pigmentation (Kala-azar)
  - Bony tenderness (leukaemia)
  - In young/child -
    - Anaemia, jaundice, haemolytic facies (hereditary haemolytic anaemia)

Hepatosplenomegaly: How to investigate

- **Hb, TC, DC, ESR, PBF**
  - **Kala-azar (leukopenia with relative lymphocytosis and monocytosis)**
  - **Leukaemia**
  - **Hereditary haemolytic anaemia (microcytic hypochromic blood picture)**
  - **Myelofibrosis (leukoerythroblastic blood picture, tear drop cells)**
- **MP**
- **X-ray chest (lymphoma ⇨bilateral hilar lymphadenopathy)**

Hepatosplenomegaly: How to investigate …

- Bone marrow (LD bodies, leukaemia myelofibrosis)
- If lymph node is palpable ⇨ FNAC/ Biopsy
- Hb-electrophoresis for hereditary haemolytic anaemia
- Other investigations according to suspicion

Thank you all