LECTURE SEVEN OVERVIEW
(Lecture 7b)

Gastrointestinal Tract Development and Malformations

❖ Hypertrophic Pyloric Stenosis: *

- Occurs when the muscularis externa in the pyloric region hypertrophies and forms a **small palpable mass** (**"olive"**), causing a **narrow pyloric lumen** that **obstructs food passage**.

- *The barium contrast radiograph in Figure opposite shows the long, narrow, double channel of the pylorus (arrows) in a patient with hypertrophic pyloric stenosis.*

Hypertrophic Pyloric Stenosis
Clinical Presentation - 7b

- **Vomiting**, which is:
  - ***Projectile,***
  - ***Non-bilious***
  - ***Post-prandial*** (occurs after feeding)
  - **Infants treated with erythromycin** have increased incidence
- O/E:
  - ***Palpable small mass*** at the right costal margin

Development of G.I.T

Clinical : ***Foregut*** **Anomalies** – 8a

Developmental anomalies of the gall bladder anatomy:

- Are fairly common in which are found:
- **Bi-lobed gall bladder**
- **Diverticula** and
- **Septated gall bladder**

(the latter likely due to incomplete recanalization of the gall bladder lumen)

Development of G.I.T

Clinical: **Foregut Anomalies** – 8b

a. Intrahepatic gall bladder :

- Occurs when the gallbladder rudiment advances beyond the hepatic diverticulum and becomes buried within the substance of the liver.

***b. Floating gall bladder*:**

- Occurs when the gallbladder rudiment lags behind the hepatic diverticulum and thereby becomes suspended from the liver by a mesentery.
- A floating gall bladder is at risk for **torsion** (i.e., a twisting or rotation around the axis of the mesentery).

Development of G.I.T
Clinical : ***Foregut*** **Anomalies** - 9a

c. Developmental anomalies of the cystic duct *anatomy*:

- Are fairly common.
- ***Biliary atresia*:**
- Defined as the obliteration of extra-hepatic and/or intrahepatic ducts.
- Ducts are replaced by fibrotic tissue due to acute and chronic inflammation

Clinical Features (Biliary atresia):

- **Progressive neonatal jaundice (**onset soon after birth)

Blockage of bile pigments (*refer to haem metabolism by-products*) results in:

- **white clay–colored stool** *(from missing stercobilin)*
- **dark-colored urine** *(from missing urobilin)*
- Average survival time is 12–19 months with a 100% mortality rate*

Pathophysiology of Jaundice (in biliary atresia) - 9b

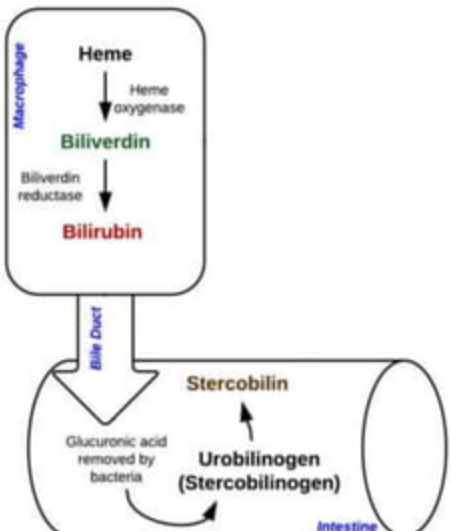

Figure 2 – **Bilirubin is produced as a byproduct of haem metabolism**

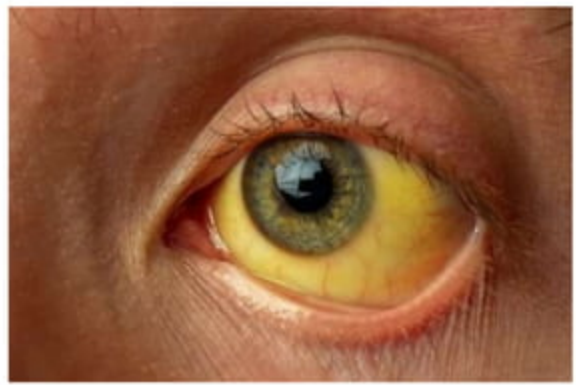

Figure 1: **Jaundice**: (resulting from bilirubin accumulation)

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Types of Jaundice - 9c

Types of Jaundice:

1. **Pre-hepatic Jaundice***
2. **Hepatic (Hepatocellular) Jaundice***
3. **Post-hepatic Jaundice***

❖ **Pre-hepatic jaundice:**

- Excessive RBC breakdown causing
- **Hyperbilirubinaemia** (because of overwhelmed liver's ability to conjugate bilirubin)

Types of Jaundice - 9d

❖ **Hepatocellular Jaundice:**

- Due to **hepatic cell dysfunction** (from disease)
- Liver loses ability to conjugate bilirubin
- In liver cirrhosis, liver cells also compress, causing: **biliary obstruction;** mixing of conjugated & conjugated bilirubin in blood

❖ **Post-hepatic Jaundice**:

- Due to **obstruction of biliary drainage**
- Non-excreted bilirubin will have been conjugated by the liver, resulting in **conjugated hyperbilirubinaemia**

Common Causes of Jaundice

❖ Pre-hepatic Causes:

- **Haemolytic anaemia**
- **Gilbert's syndrome**
- **Criggler-Najjar syndrome**

❖ Hepatocellular Causes:

- **Alcoholic liver disease**
- **Viral hepatitis**
- **Iatrogenic, e.g. medication**
- **Hereditary haemochromatosis**
- **Autoimmune hepatitis**
- **Primary biliary cirrhosis or primary sclerosing cholangitis**
- **Hepatocellular carcinoma**

Causes (Continued)

- ❖ Post-hepatic Causes:
- Intra-luminal causes:
  - ➢ **Gallstones**
- Mural causes:
  - ➢ **Cholangiocarcinoma,**
  - ➢ **Biliary strictures**,
  - ➢ **Biliary atresia**
  - ➢ D**rug-induced cholestasis**
- Extra-mural causes,
  - ➢ **Pancreatic cancer** or
  - ➢ **Abdominal masses** (e.g. lymphomas)

- **Bilirubinuria**: bilirubin in urine
- Bilirubinuria can give clues of type of jaundice
- **Conjugated bilirubin can be excreted via urine** (hence darkening of urine) but
- **Unconjugated bilirubin is insoluble in water** (hence normal urine observed)
- **Pale stools**: due to reduced stercobilin entering GIT

Investigations

(1)Laboratory Tests:

- **Liver Function Tests**
- **Coagulation Studies** (PT marker of liver synthesis function)
- **FBC** (Anaemia, raised MCV and thrombocytopenia seen in liver disease),
- **U&E's**

(2) Imaging:

- Abdominal Ultrasound
- Magnetic Resonance CholangioPancreatography (MRCP)

(3) Liver Biopsy

Liver Function Tests

Blood Marker	Significance
Bilirubin	Quantify degree of any suspected jaundice
Albumin	Marker of liver synthesising function
AST and ALT	Markers of hepatocellular injury*
Alkaline Phosphatase	Raised in biliary obstruction (as well as bone disease, during pregnancy, and certain malignancies)
Gamma-GT	More specific for biliary obstruction than ALP (however not routinely performed)

	Viral Serology	Non-Infective Markers
Acute Liver Injury	Hepatitis A, Hepatitis B, Hepatitis C, and Hepatitis E, CMV and EBV	Paracetamol level Caeruloplasmin Antinuclear antibody and IgG subtypes
Chronic Liver Injury	Hepatitis B Hepatitis C	Caeruloplasmin Ferritin and transferrin saturation Tissue Transglutaminase antibody Alpha-1 antitrypsin Autoantibodies*

Management of Jaundice

- **Definitive Management**:
  - Treat cause (e.g. surgery to remove obstruction)
- **Symptomatic Management**:
  - e.g. Anti-histamines for itching; treat hypoglycaemia, monitor coagulopathy (Vitamin K, FFP prn);

- **Preventive Management:**
  - Health Education to:
  - Avoid alcoholic cirrhosis
  - Avoid hepatitis
  - etc