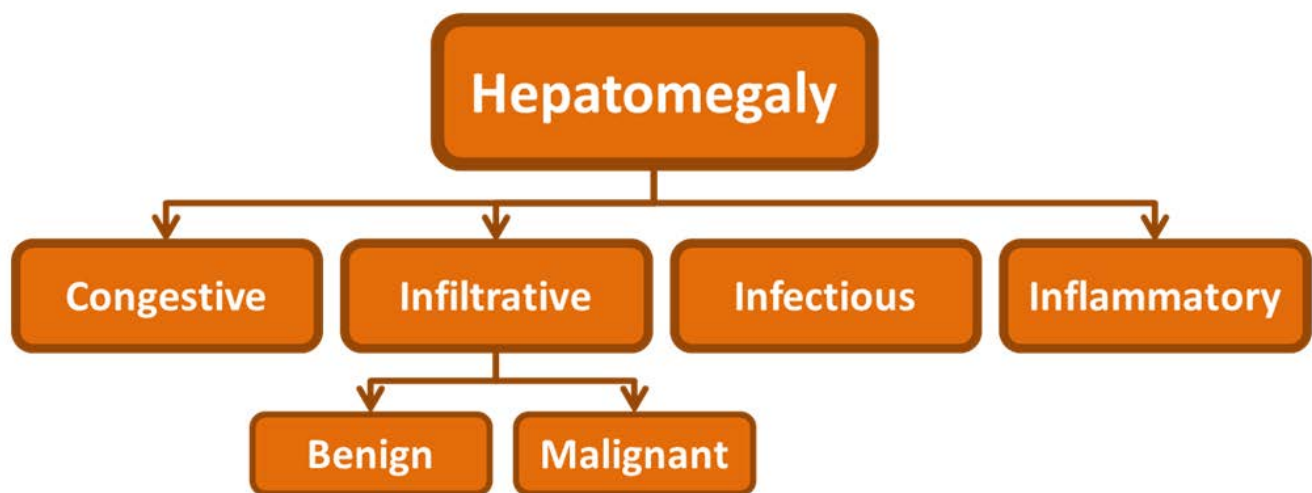


Chapter 11. Approach to Big Livers and Big Spleens

11.1 Hepatomegaly

- Hepatomegaly (big liver) is a liver >12 cm in the mid-clavicular line [see Chapter 2.4]
 - It may be noted on physical examination or on imaging (US, CT, MRI)
- Scheme for Hepatomegaly



- Clues in the history (Hx) and / or physical examination (PE) help identify the cause

Causes of Hepatomegaly

- **Congestive**
 - Congestive Heart Failure
 - *PE clues* = ↑jugular venous pressure (JVP), S3 or S4, murmur of tricuspid regurgitation, pulsatile liver, edema ± ascites
 - Constrictive Pericarditis
 - *PE clues* = ↑ JVP, pericardial knock, pulsus paradoxus, Kussmaul sign, pulsatile liver, edema ± ascites

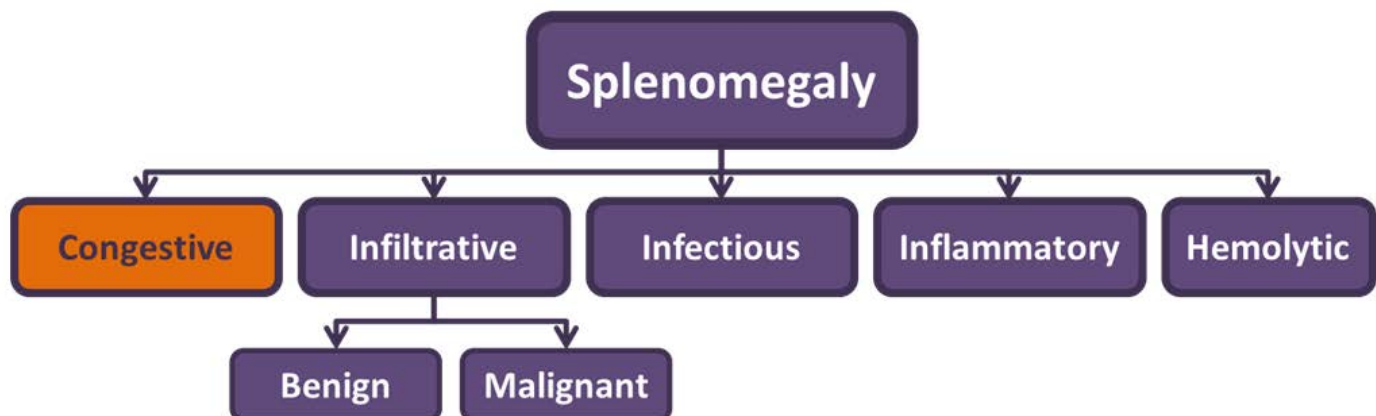
- Budd Chiari Syndrome
 - Obstruction of large hepatic veins by clot
 - *Hx clues* = hypercoagulable states such as antithrombin III deficiency, activated protein C resistance, protein C or S deficiencies, lupus anticoagulant, paroxysmal nocturnal hemoglobinuria (PNH), malignancy, connective tissue disease
 - *PE clues* = RUQ pain, weight gain, ascites \pm jaundice and can cause ALF
- Sinusoidal Obstruction Syndrome (SOS) or Veno-Occlusive Disease (VOD)
 - Obstruction or damage to the small central veins within the liver
 - *Hx clues* = can be seen with chemotherapy given for bone marrow transplantation (BMT)
 - *PE clues* = RUQ pain, weight gain, ascites \pm jaundice and can cause ALF
- **Infiltrative (Benign)**
 - Fatty \rightarrow Alcohol or NAFLD
 - *Hx clues* = alcohol abuse, diabetes, hyperlipidemia, obesity
 - *PE clues* = stigmata of chronic liver disease
 - Abnormal protein \rightarrow amyloidosis
 - Cysts \rightarrow Polycystic Liver Disease (PCLD)
 - Multiple cysts on imaging in liver \pm kidneys [see Chapter 12.1]
 - Red blood cells (extra-medullary hematopoiesis) \rightarrow myelofibrosis
- **Infiltrative (Malignant)**
 - Primary \rightarrow HCC or iCCA [see Chapter 13]
 - *Hx clues* = cirrhosis, hepatitis B virus, primary sclerosing cholangitis

- *PE clues* = stigmata of chronic liver disease
- Metastatic → breast, lung, GI, pancreas, etc.
 - *Hx clues* = older age, smoker, symptoms from primary cancer
- Hematologic → lymphoma, leukemia, MM
 - *Hx clues* = B symptoms (fever, night sweats, weight loss)
 - *PE clues* = lymphadenopathy ± splenomegaly
- **Infectious**
 - Viral Hepatitis → ABCs, EBV (mononucleosis)
 - Hx / PE clues (ABCs) = risk factors, stigmata of chronic liver disease
 - Hx / PE clues (Mono) = fever, sore throat, lymphadenopathy ± splenomegaly
 - Tuberculosis (TB)
 - *Hx clues* = sick contacts, cough, fever, night sweats, abnormal CXR
 - Liver abscess
 - *PE clues* = fever, RUQ pain
 - Schistosomiasis
 - *Hx clues* = travel to endemic countries and swimming in infected water
 - Life-cycle involves snails → humans infected through skin (rash) → travels to lung (cough) → swallowed taken up into portal circulation → adult worm lays eggs in mesenteric venules → circulate to liver and are shed in the stool
- **Inflammatory**
 - Alcoholic Hepatitis
 - *Hx clue* = ETOH binging

- *PE clue* = jaundice, RUQ, ascites, fever
- Chronic Liver Disease
 - *Hx clue* = risk factors
 - *PE clue* = stigmata of chronic liver disease
- Sarcoidosis
 - Inflammatory condition characterized by granulomas in lungs, lymph nodes, liver
- Histiocytosis X
 - Rare disorder with excess number of histiocytes (tissue macrophages) in the liver and other organs

11.2 Splenomegaly

- The spleen must enlarge 2-3 times to be felt on clinical examination [see Chapter 2.4]
- **Scheme for Splenomegaly**



Causes of Splenomegaly

- **Congestion – from cirrhotic or non-cirrhotic portal hypertension**
- Infiltrative – benign or malignant
- Infectious – e.g. mononucleosis
- Inflammatory
- Hemolytic conditions

- **Congestion from portal hypertension** is a common cause of splenomegaly
 - This is most commonly due to cirrhosis *[see Chapter 14]*
 - Rarely, it is due to **non-cirrhotic portal hypertension**, which can be:
 - **Pre-hepatic** = portal vein thrombosis, splenic vein thrombosis, splenomegaly
 - **Intra-hepatic**
 - **Pre-sinusoidal** = schistosomiasis, primary biliary cirrhosis (PBC), sarcoidosis, congenital hepatic fibrosis, idiopathic
 - **Sinusoidal** = Nodular Regenerative Hyperplasia (NRH), vitamin A or vinyl chloride toxicity
 - **Post-sinusoidal** = Budd Chiari Syndrome (BCS), sinusoidal obstruction syndrome (SOS)
 - **Post-hepatic** = cardiac disease, IVC obstruction
 - **Nodular regenerative hyperplasia (NRH)** can lead to non-cirrhotic portal hypertension

- Diagnosis is by liver biopsy (best seen on reticulin stain) which shows nodules, like in cirrhosis, but without fibrosis

- **Associations**
 - Autoimmune
 - RA = Felty's
 - SLE
 - PBC
 - Drugs
 - Sarcoidosis
 - Malignancy
- **Diagnosis on bx**
 - No fibrosis
 - Reticulin stain

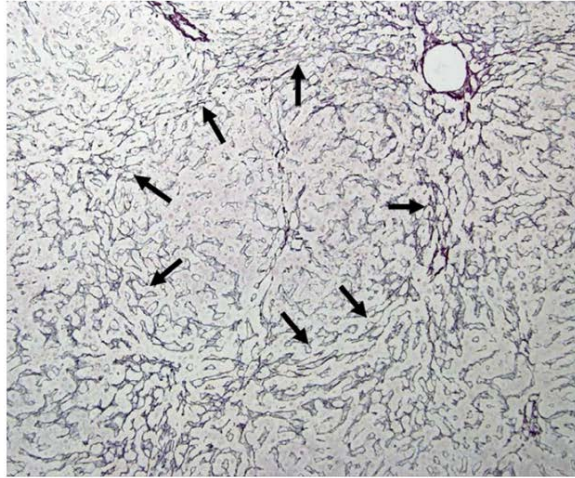
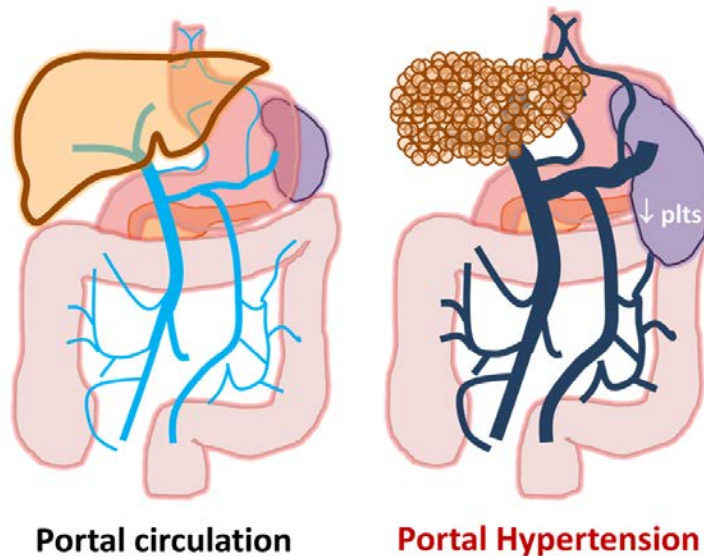


Image Source: http://livertox.nih.gov/Phenotypes_nodular.html

- Patients may have **hypersplenism** = overactive spleen with reduction in platelets, white blood cells or red blood cells

REMEMBER = low platelets (\downarrow plts) in a patient with chronic liver disease is a clue that the patient has developed cirrhosis



Abbreviations

BMT – bone marrow transplantation

CXR – chest x-ray

Hx – history

JVP – jugular venous pressure

PBC – primary biliary cholangitis

PCLD – polycystic liver disease

PE – physical examination

Plts – platelets

PNH – paroxysmal nocturnal hemoglobinuria

RA – rheumatoid arthritis

SLE – systemic lupus erythematosus

SOS – sinusoidal obstruction syndrome

TB – tuberculosis

VOD – veno-occlusive disease

Figure citations

Nodular regenerative hyperplasia. US National Library of Medicine. 2011. Retrieved on July 20, 2017 from https://livertox.nlm.nih.gov/Phenotypes_nodular.html