Differential Diagnosis Of Splenomegaly

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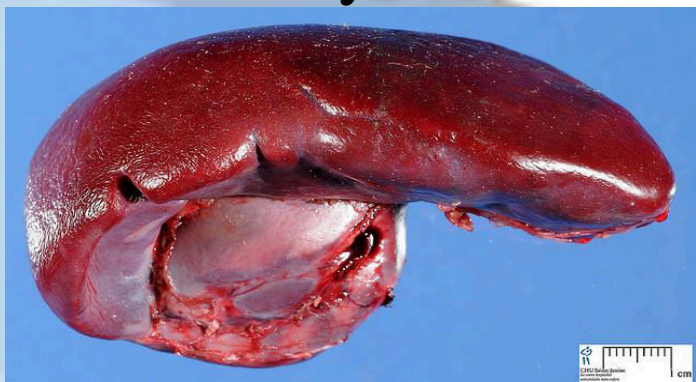
The normal spleen

- The spleen is a reticulo-endothelial organ that arises from the dorsal mesogastrium in the 5th week of intra-uterine life.
- **Normal size and location:**
  - Weight - <250g, and it decreases in size with advancing age
  - Location- It lies within the left upper quadrant (LUQ) of the peritoneal cavity in relation to 9-12 ribs, with maximum cephalocaudal diameter of 13 cm by USG



Funtions of spleen

1. Phagocytosis of red blood cells and particulate matter

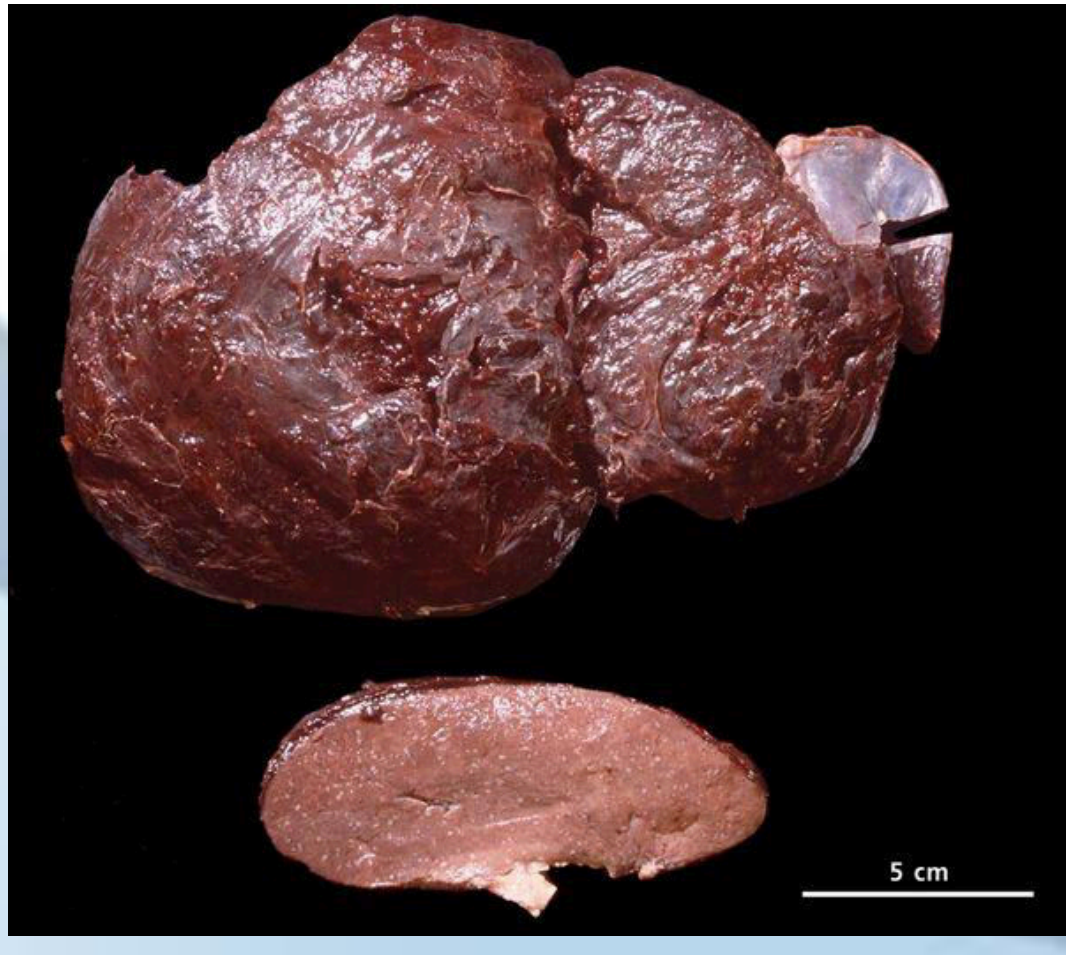
2. Hematopoiesis

3. Antibody production

4. Sequestration of formed blood element

Splenomegaly

- A result of increase in the functions of spleen in the red and white pulps
- Size: 400-500g , and >1000g is considered as massive splenomegaly



HACKETT'S CLASSIFICATION

0	Spleen not palapable
1	Palpable <halfway between LCM on deep inspiration
2	Palpable <halfway between LCM and umbilicus
3	>halfway to umbilicus but not beyond
4	Palpable below umbilicus but not below horizontal line midway between umbilicus and pubic symphysis
5	Beyond horizontal line

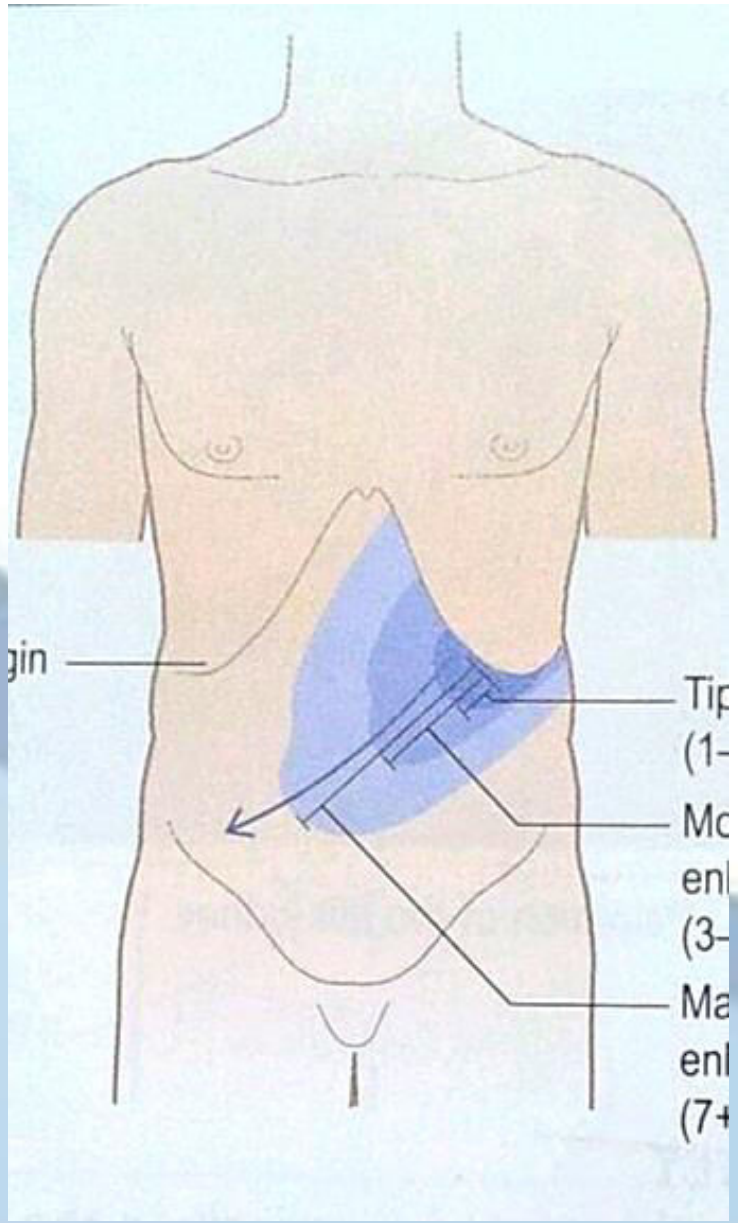
Splenomegaly

- Also classified as:

❖ **Mild splenomegaly-** spleen <5cm below the left costal margin (LCM)

❖ **Moderate splenomegaly-** 5-8 cm below LCM and maximum cephalocaudal diameter being 11-20cm

❖ **Massive splenomegaly-** >8cm below LCM and maximum cephalocaudal diameter being >20cm



Etiology of splenomegaly

- **Hypersplenism**
- **Infectious causes**

Non-specific splenitis	Infectious mononucleosis
Tuberculosis	Enteric fever
Syphilis	Malaria
Kala-azar	Trypanosomiasis
Echinococcosis	Schistosomiasis

ENLARGEMENT DUE TO INCREASED DEMAND FOR SPLENIC FUNCTION

Lymphohematogenous disorders

Hodgkin's disease	Non-hodgkin's lymphoma/leukemia
Multiple myeloma	Myeloproliferative disorders
Hemolytic anaemias	Thrombocytopenic purpura
Chronic myeloid leukemia (CML)	Chronic lymphocytic leukemia (CLL)
Polycythemia vera	Autoimmune hemolytic anaemia
Myelofibrosis with myeloid metaplasia	Hereditary spherocytosis

- **Immunologic-inflammatory conditions**

Rheumatoid arthritis (Felty's syndrome)	SLE

- **Storage diseases**

Gaucher's disease	Niemann-Pick's disease
Mucopolysaccharidoses	Amyloidosis

- **Enlargement due to abnormal splenic or portal blood flow- congestive states related to portal hypertension**

Liver cirrhosis	**Splenic vein thrombosis**
Hepatic vein obstruction	Portal vein thrombosis- intra/extrahepatic
Banti's Disease	Congestive heart failure

- **Splenic cystic lesions and neoplasms**

Hydatid cystic disease	**Epidermoid cysts**
Pseudocysts	Splenic abscess
Hematomas(large solitary abscesses)	Primary neoplasms: hemangio-sarcoma, lymphoma, hamartoma

Hypersplenism

- **Etiology:**

1. Neoplastic infiltrations-lymphomas and hemagiosarcomas
2. Diseases of bone marrow in which the spleen becomes site of extramedullary hematopoiesis
3. Metabolic/genetic disorders- Gaucher's disease.

- **Clinical features**:- present due to underlying disorder or are secondary to the depletion of circulating blood cells

❖ h/o LUQ fullness, discomfort (may be severe), early satiety

❖ h/o hematemesis due to gastroesophageal varices

❖ h/o recurrent infections in severe leukopenia

❖ h/o fatigue

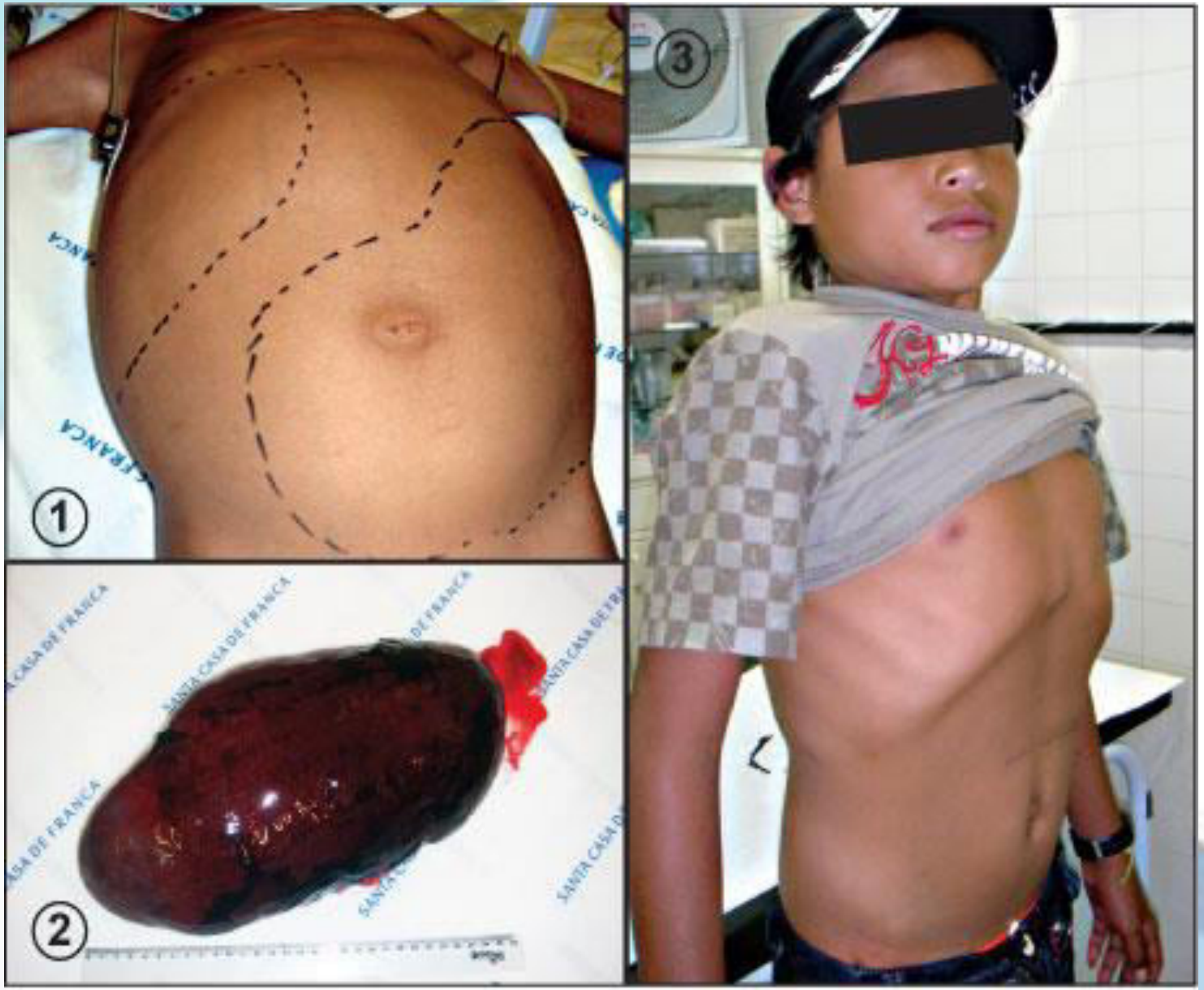
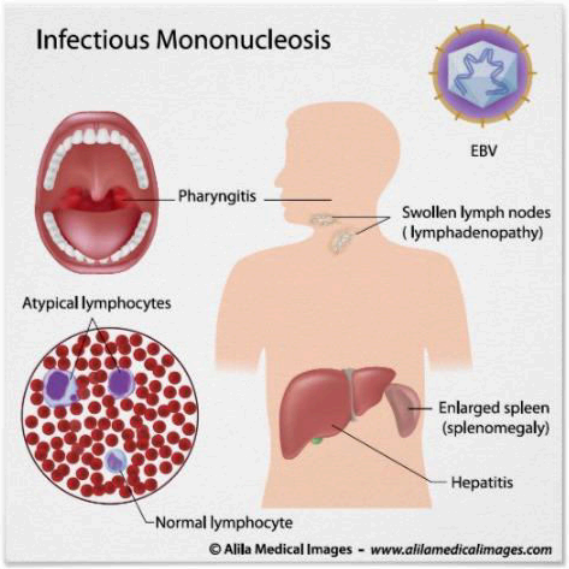


FIGURE 1 - Malnutrition and hepatosplenomegaly.

FIGURE 2 - Spleen: 2.700g, 28cm long, 18cm wide and 10cm thick.

FIGURE 3 -Two months after splenectomy.

Infectious mononucleosis



- **Etiology:**

❖ Is a viral infection caused by EBV

❖ Transmission : intimate contact with body secretion, mostly oropharyngeal, and in the individuals with congenital immunodeficiency.

❖ Virus infects the B lymphocytes leading to humoral and cellular immune response to virus.

❖ The humoral response is diagnostic

❖ T lymphocyte response determines the clinical presentation. However, ineffective T lymphocyte response leads to Blymphocyte proliferation and B cell lymphomas

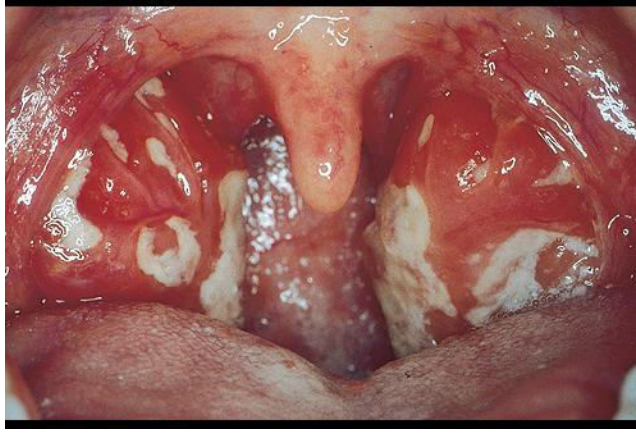
- **Clinical features:**

❖ Most assymptomatic

❖ Fatigue, prolonged malaise, nausea and anorexia without vomitings

❖ Early signs include- low grade fever, lymphadenopathy, pharyngitis, maculo-papular rash

❖ Late signs- splenomegaly, hepatomegaly, jaundice, palatal petechiae and uvular oedema. Splenic tenderness is present

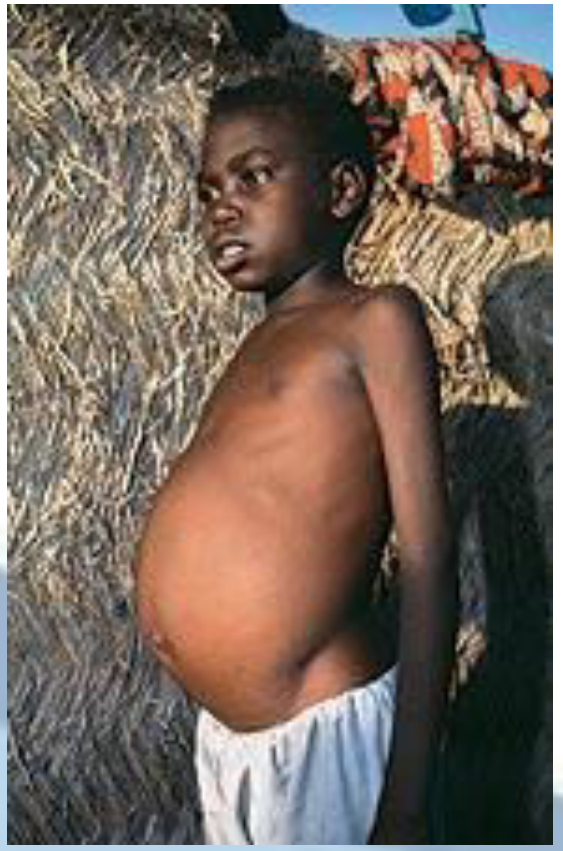


Malaria-hyperactive malarial splenomegaly

- Formerly known as tropical splenomegaly syndrome, HMS is the most common cause of massive splenomegaly in malaria endemic areas
- **Etiopathogenesis:**

❖ There are increased levels of antibodies for P.falciparum, P.vivax, and P.ovale due to chronic antigenic stimulation

❖ Chronic exposure to malaria leading to exaggerated stimulation of polyclonal B lymphocytes that leads to excess IgM production (polyclonal)



- **Clinical features:**

❖h/o chronic abdominal swelling- 64% cases

❖h/o weight loss

❖h/o weakness, headache and malaise- indicate severe anaemia

❖Chronic splenic enlargement

❖Rarely intermittent fever

Schistosomiasis

- **Etiopathogenesis:**

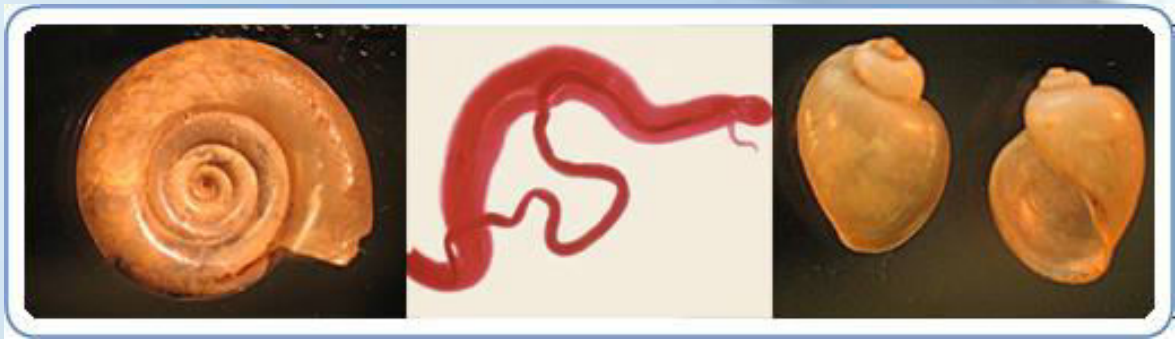
❖ Mostly prevalent in Africa , Asia and South America

❖ Caused by the infection with S.mansoni-75% cases and S.haematobium-25% cases

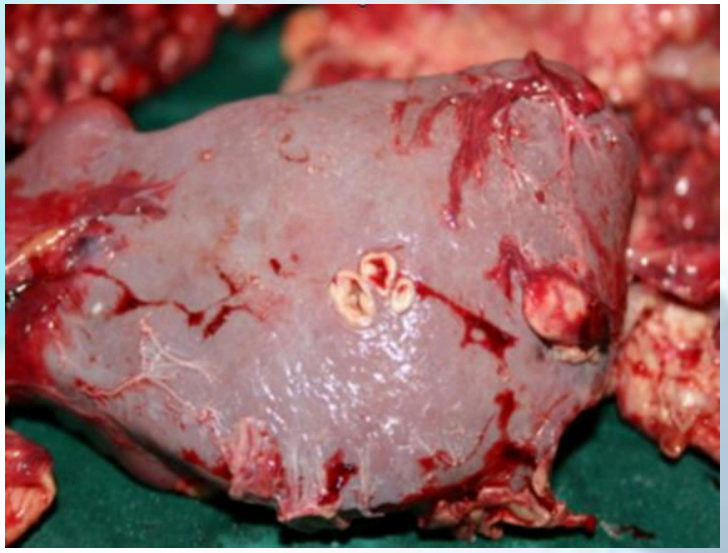
❖ Splenic enlargment is due to hyperplasia which is induced by phagocytosis of disintegrated worms, ova and toxins, and also by portal hypertension which is the result of hepatic fibrosis

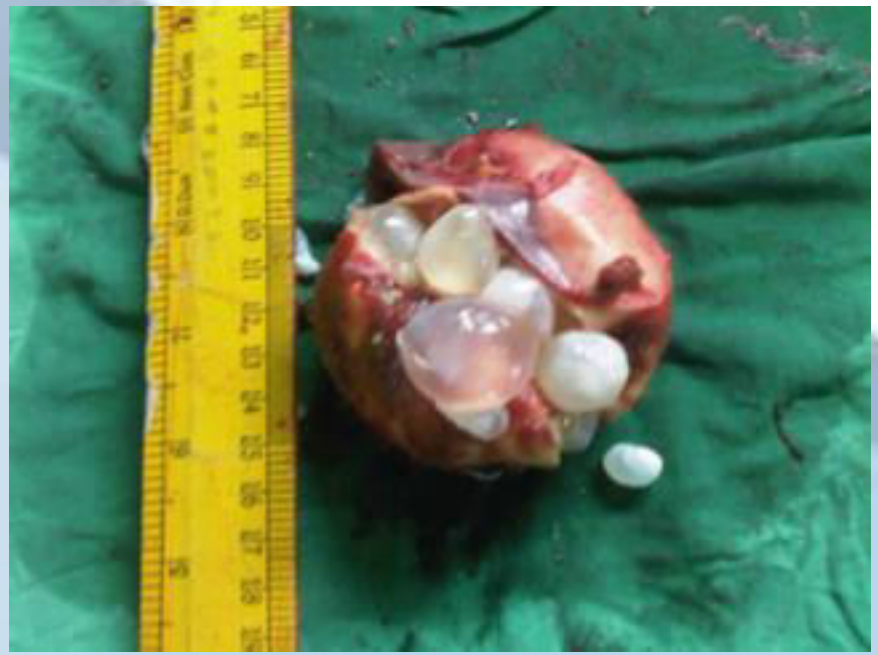
- **Clinical features**

❖ Splenomegaly- degree reflects the extent of hepatic fibrosis



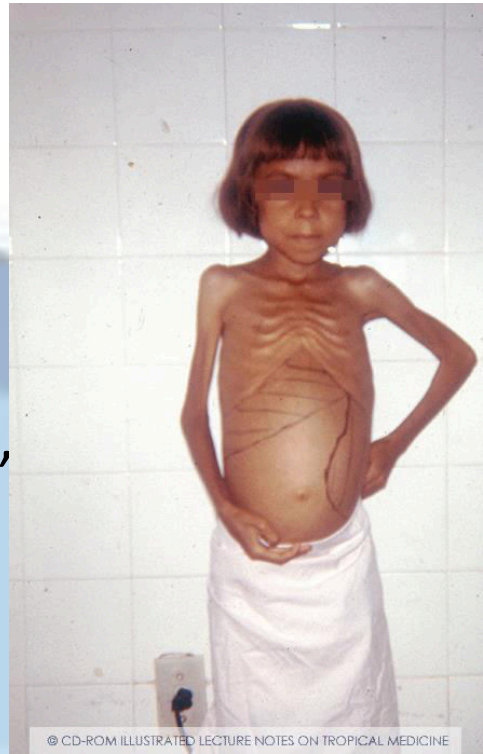
Echinococcosus Hydatid Cyst





Visceral leishmaniasis

- **Etiology:**
- It is a protozoal disease that is caused by L.donovani.
- Visceral leishmaniasis is the most fatal form, also known as kala-azar or Black water fever.
- Systemic infections of liver, spleen and bone marrow
- **Clinical features:**
- Pentad- fever, weight loss, night sweats, weakness, anorexia, wasting, skin hyperpigmentation
- **O/E:** massive hepatosplenomegaly
- **Complications**: amyloidosis, glomerulonephritis, cirrhosis
- HIV- associated Leishmaniasis- GIT ulcerations, pleural effusion, odynophagia

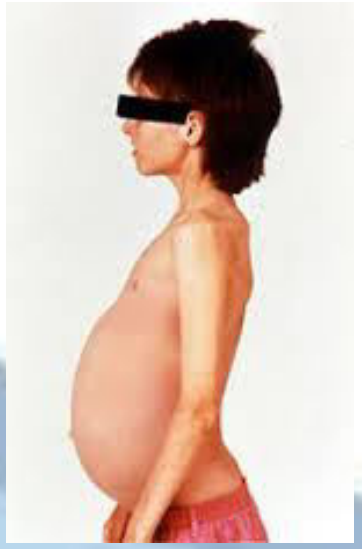


Gaucher's disease

- **Etiopathogenesis:**

❖ It is an autosomal recessive disorder due to deficiency of beta-glucosidase (enzyme needed in degeneration of sphingolipid glucocerebroside.

❖ Lipid accumulation within the white pulp of the spleen, liver or bone marrow



❖ **Clinical features:**

❖ massive splenomegaly- 3.6-4.1kgs. Splenic enlargement begins <12years.

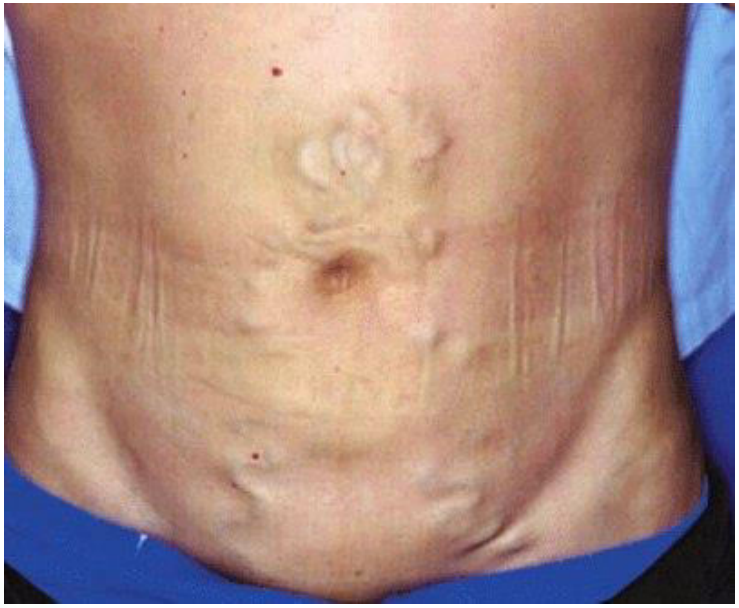
❖ Anemia

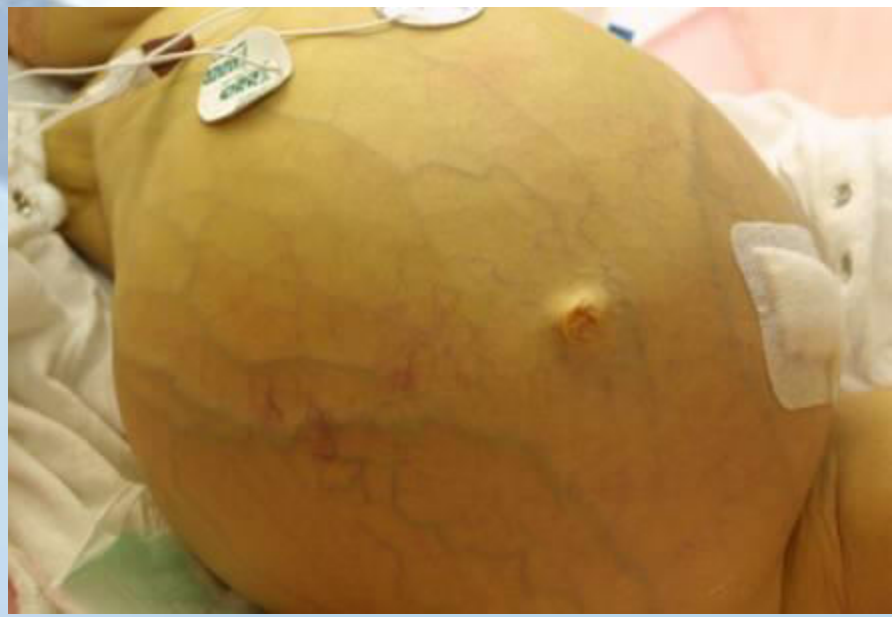
❖ Yellowish-brown discolouration of skin of hands and face

❖ Pingecula (conjunctival thickening)

Portal hypertension

- This is an elevation in portal pressure >12mmHg (normal- 5-10mmHg) and is found in liver cirrhosis, extrahepatic portal vein occlusion, intrahepatic veno-occlusive disease or occlusion of the main hepatic veins (Budd-Chiari Syndrome – BCS)
- Assymptomatic- usually diagnosed following chronic liver disease and encephalopathy, ascites or oesophageal variceal bleeding.





Portal hypertension

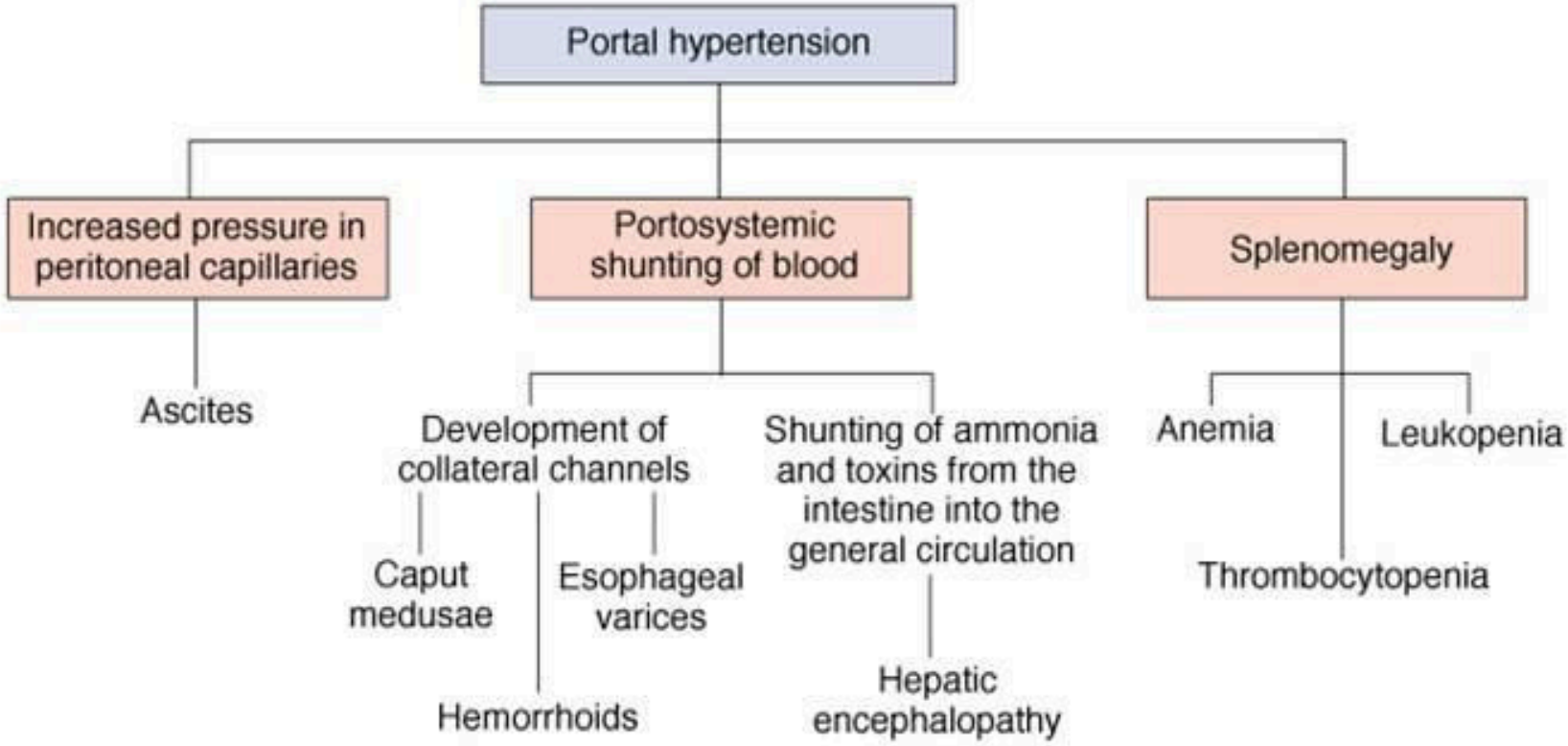
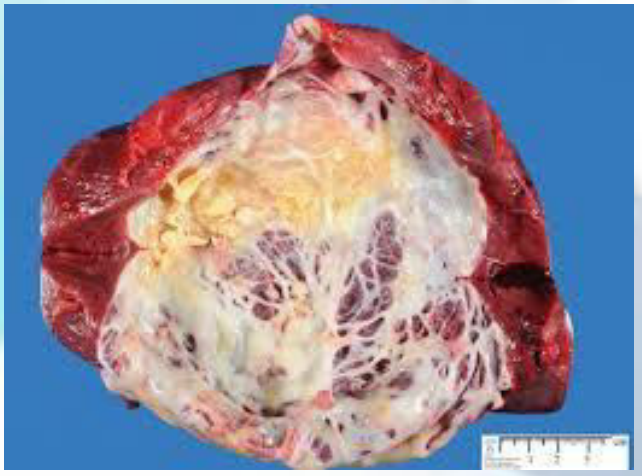


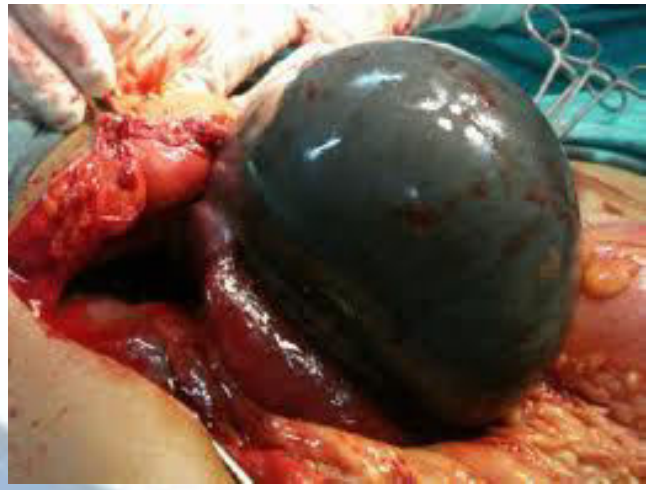
Figure 40-13 Mechanisms of disturbed liver function related to portal hypertension.

Other splenic cysts

Epidermoid cysts,

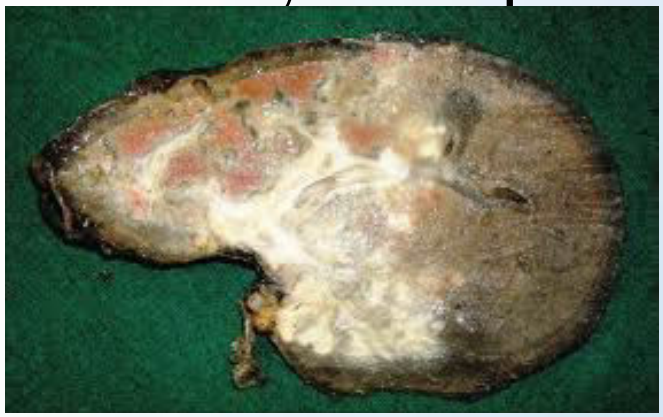


Pseudocysts



Splenic abscess-

Actinomycotic splenic abscess



Uncommon causes of splenomegaly

Disease	Clinical features
Chronic myeloid leukemia (CML)	Fatigue, weight loss, LUQ abdominal pain, fever, petechiae, ecchymosis, splenomegaly->5cm below LCM
Myelofibrosis(myeloid metaplasia)	Fever, night sweats, abdominal bloating, splenomegaly, portal hypertension
Beta-thalassemia Major	Chr anaemia, splenomegaly with abd distension, jaundice

Disease	Clinical features
Myelodysplastic syndrome	Weakness, dyspnoea, spontaneous bleeding, splenomegaly, hepatomegaly
ITP	Spontaneous bleeding, menorrhagia, splenomegaly-25% cases, positive Tourniquet test
AIHA	Splenomegaly- 50% cases Pigment gall stones-20% cases

Disease	Clincial features
Sickle cell disease	Joint pains, skin ulcers, priapism, abdominal pain, neurological abnormalities
Hereditary spherocytosis	Fever, easy fatiguability, splenomegaly, pigment gall stones-85% cases, jaundice

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THANK YOU