

# Splenomegaly and hepatomegaly in children

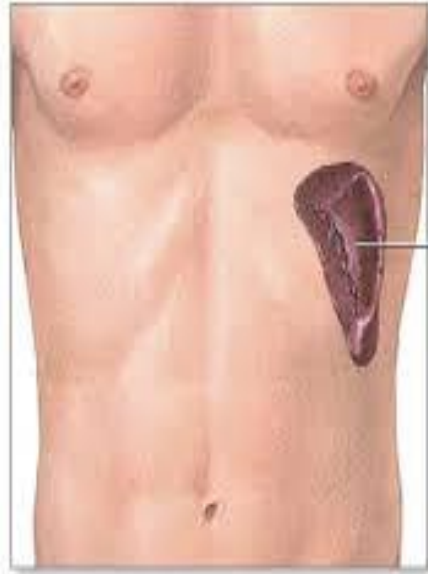
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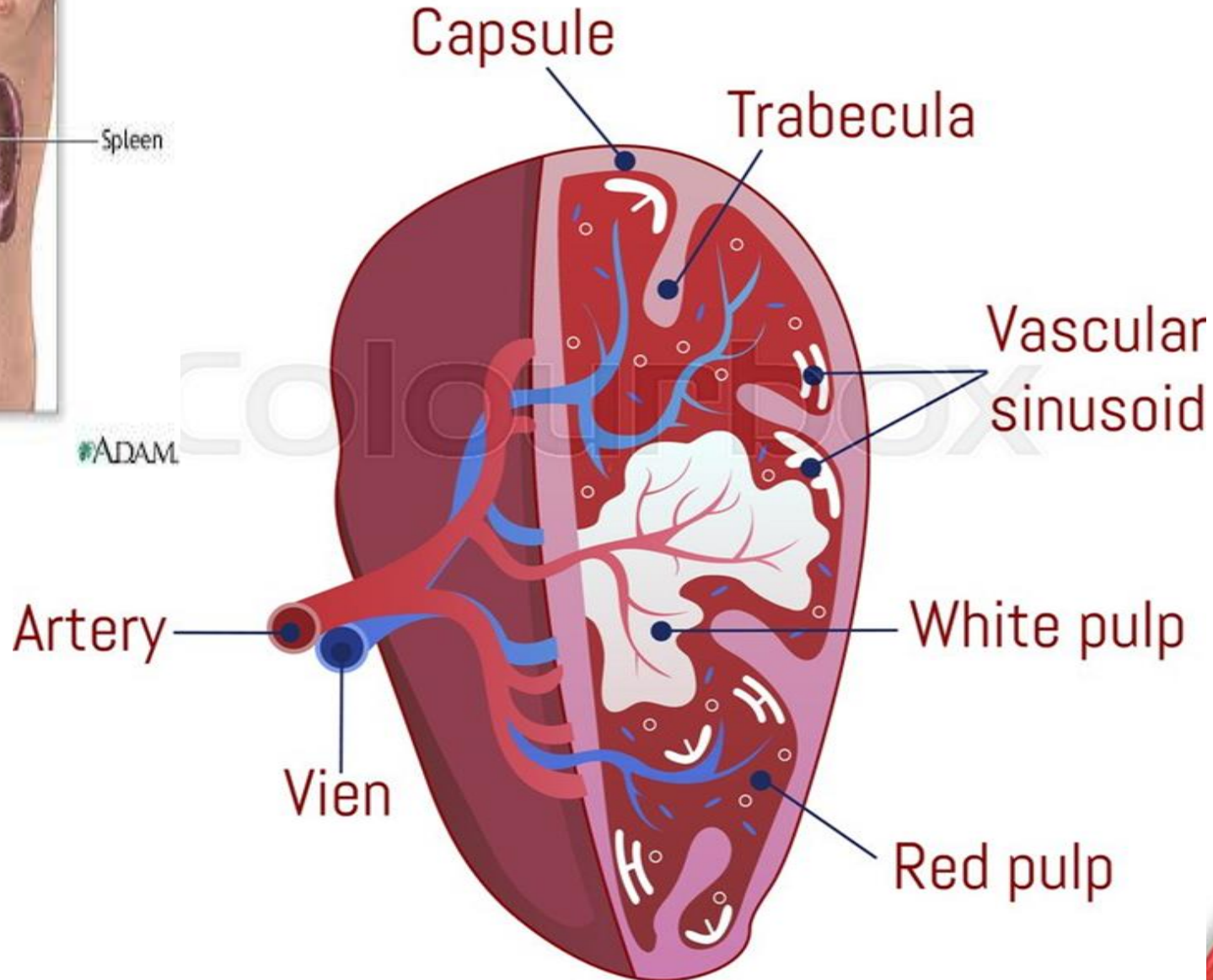


# SPLEEN ANATOMY



Spleen

ADAM



Capsule

Trabecula

Vascular  
sinusoid

White pulp

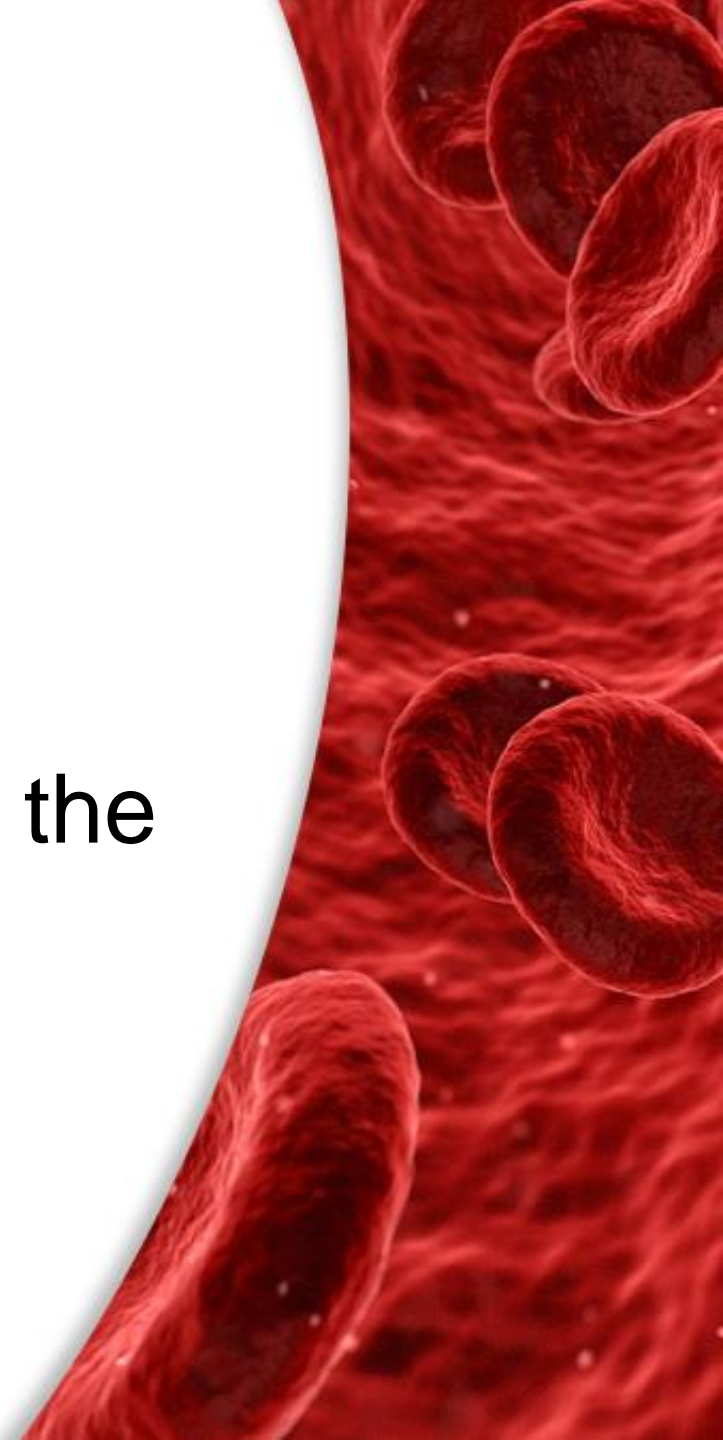
Red pulp

Artery

Vien

# Spleen function

- The spleen as a hematopoietic organ
- The white pulp of the spleen is a major part of the lymphoid tissue in the body.
- The largest component of the spleen is the red pulp.
- The spleen acts as a reservoir for platelets.

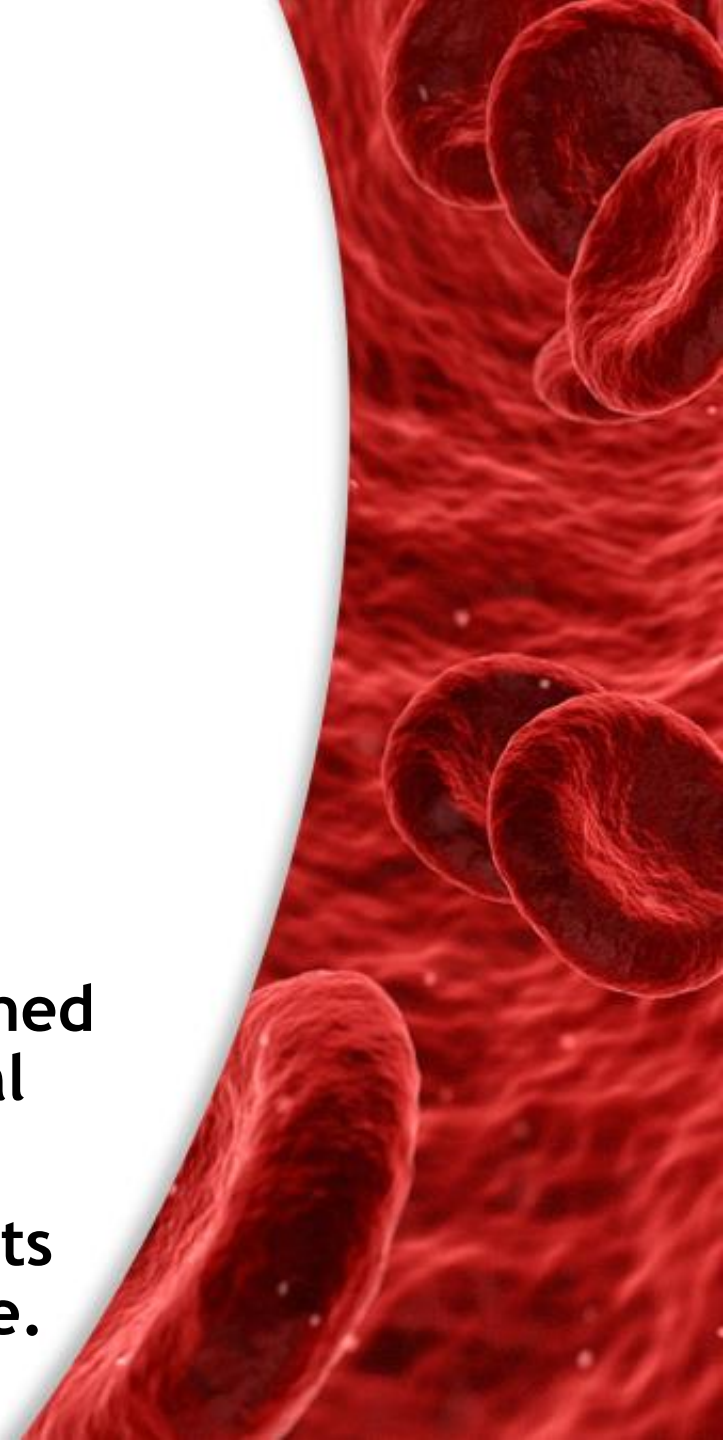




# Spleen size & splenomegaly

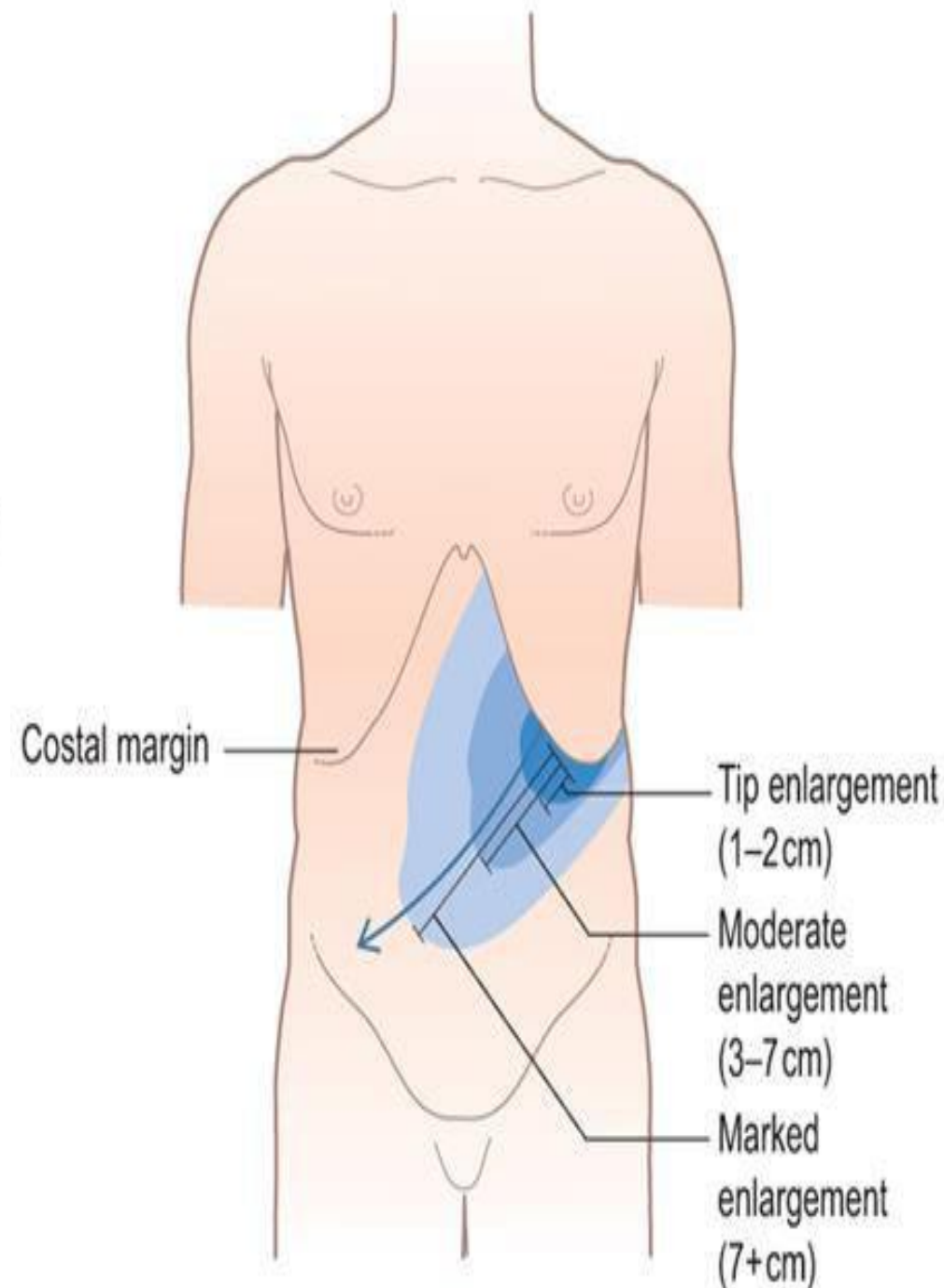
Normal values for splenic length (from the dome to the tip) measured by ultrasound

- Normal splenic length based upon age were as follows:
  - Age 3 months – 6 cm
  - Age 12 months – 7 cm
  - Age 6 years – 9.5 cm
  - Age 12 years – 11.5 cm
  - Age  $\geq 15$  years – 12 cm for girls and 13 cm for boys
- On physical examination, splenomegaly is generally defined as a palpable splenic edge felt  $>2$  cm below the left costal margin
- Massive splenomegaly: a spleen is massively enlarged if its lower pole is within the pelvic or if it crosses the midline.



## SPLENOMEGALY

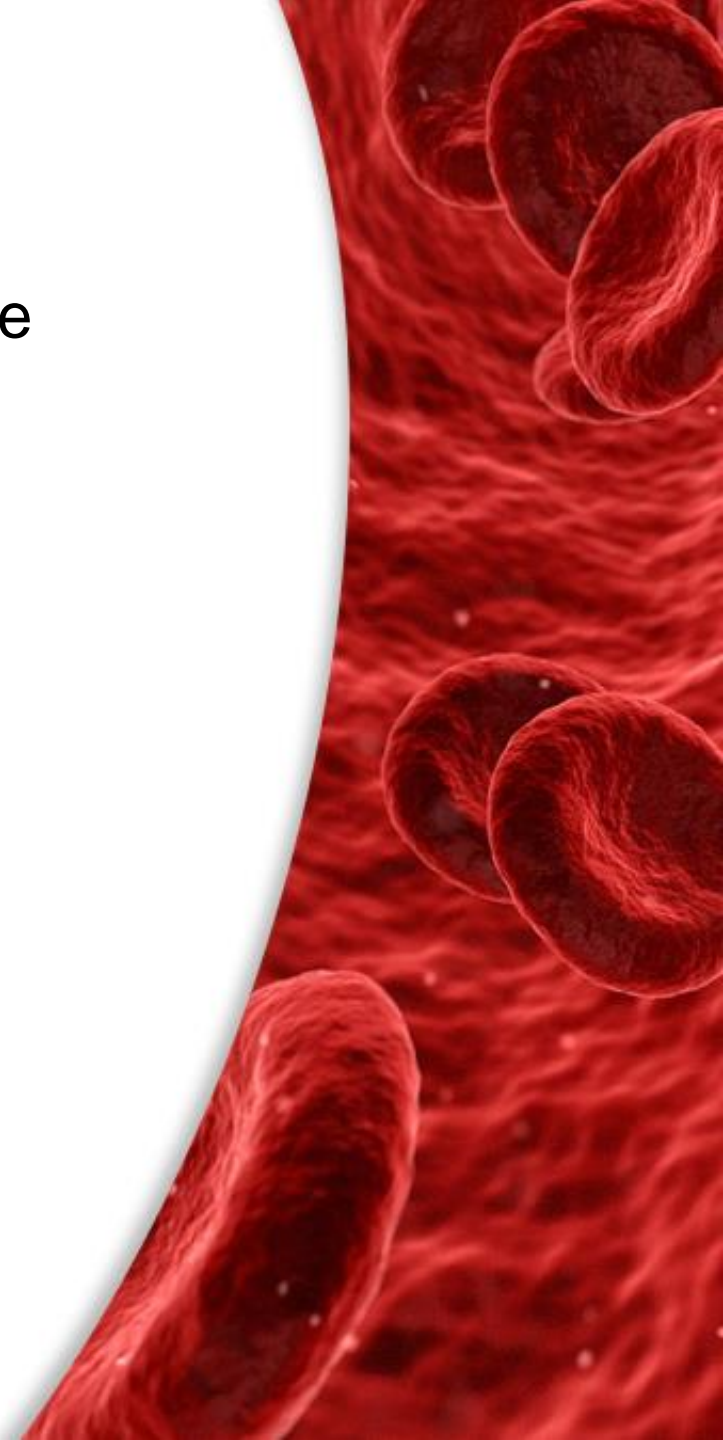
- Mild, moderate, massive
- **Massive** - beyond umblicus, crosses mid line into pelvis (>8cm)
- **Moderate** - b/w costal margin & umblicus (4-8cm)
- **Mild** - just palpable (1-3cm)



# Causes of splenomegaly

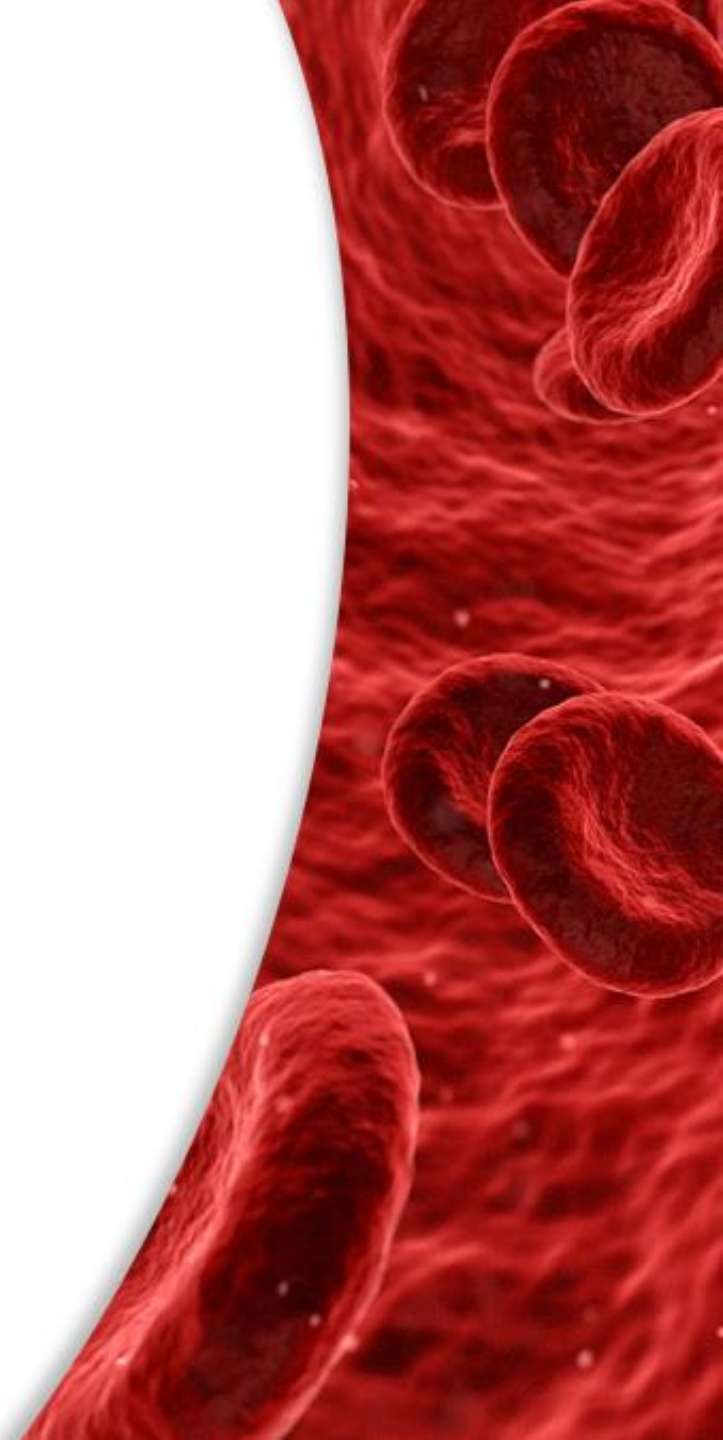
The most common causes of splenomegaly in children include

- **Infection**
- **Malignancy**
- **Disorders of immune regulation**
- Metabolic disorder
- **Liver disease**
- **storage diseases**
- **Hemolytic anemia.**
- Hemangiomata petechiae, purpura, icterus...
- Langerhans cell histiocytosis (seborrhea, or eczema)
- Signs of heart failure , murmurs, which suggest valvular or other structural heart disease or endocarditis,
- A hard or nodular spleen suggests malignancy or cystic



# CAUSES OF A MASSIVELY ENLARGED SPLEEN

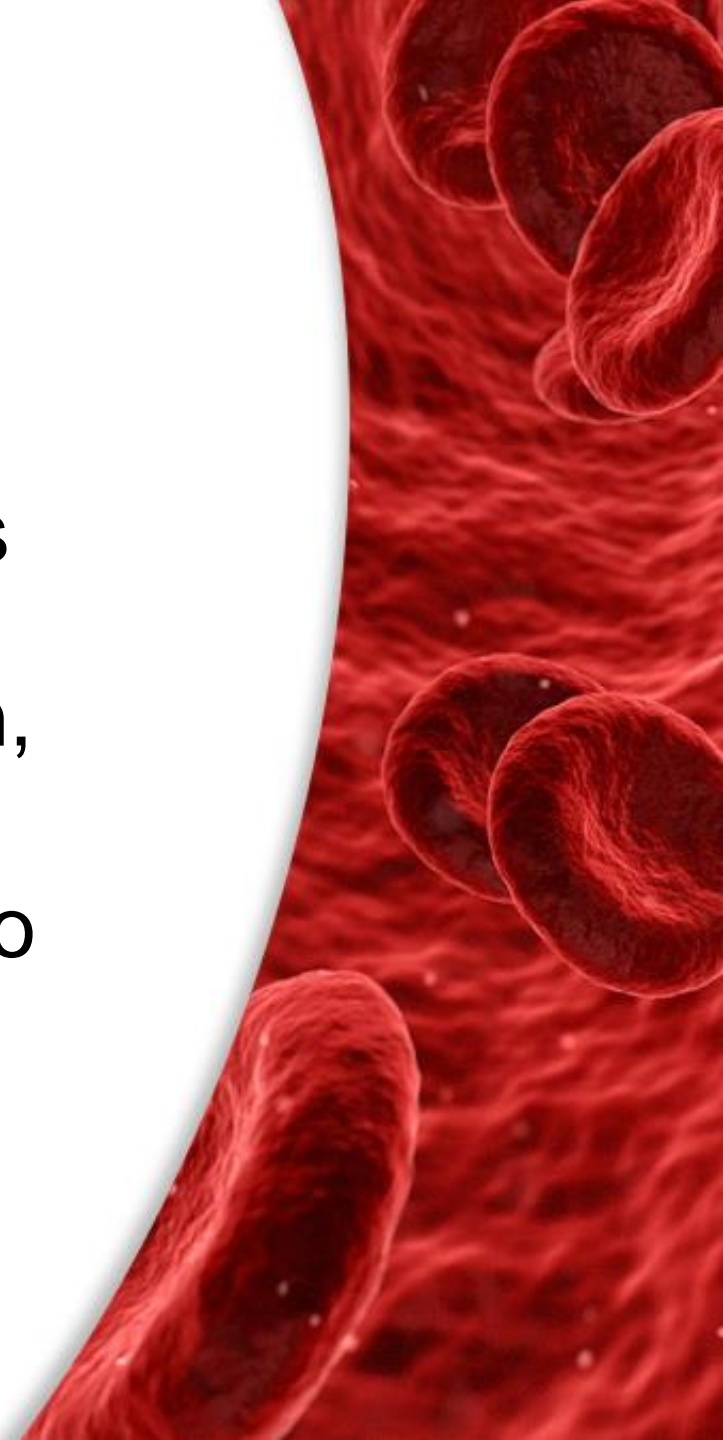
- Leukemia (lymphoid or myeloid)
- Lymphoma, usually the more indolent variants
- Thalassemia major
- Acute splenic sequestration in sickle cell disease
- Langerhans cell histiocytosis
- Hemophagocytic lymphohistiocytosis
- Autoimmune lymphoproliferative syndrome
- Malaria
- Gaucher disease
- HIV infection with *Mycobacterium avium* complex
- Kala-azar





# DIAGNOSTIC APPROACH

- For most children who present with unexplained splenomegaly, a likely diagnosis (or diagnostic category) can be arrived at based upon the history, physical examination, and initial imaging and laboratory findings.
- In most cases, additional testing is needed to establish the specific diagnosis.

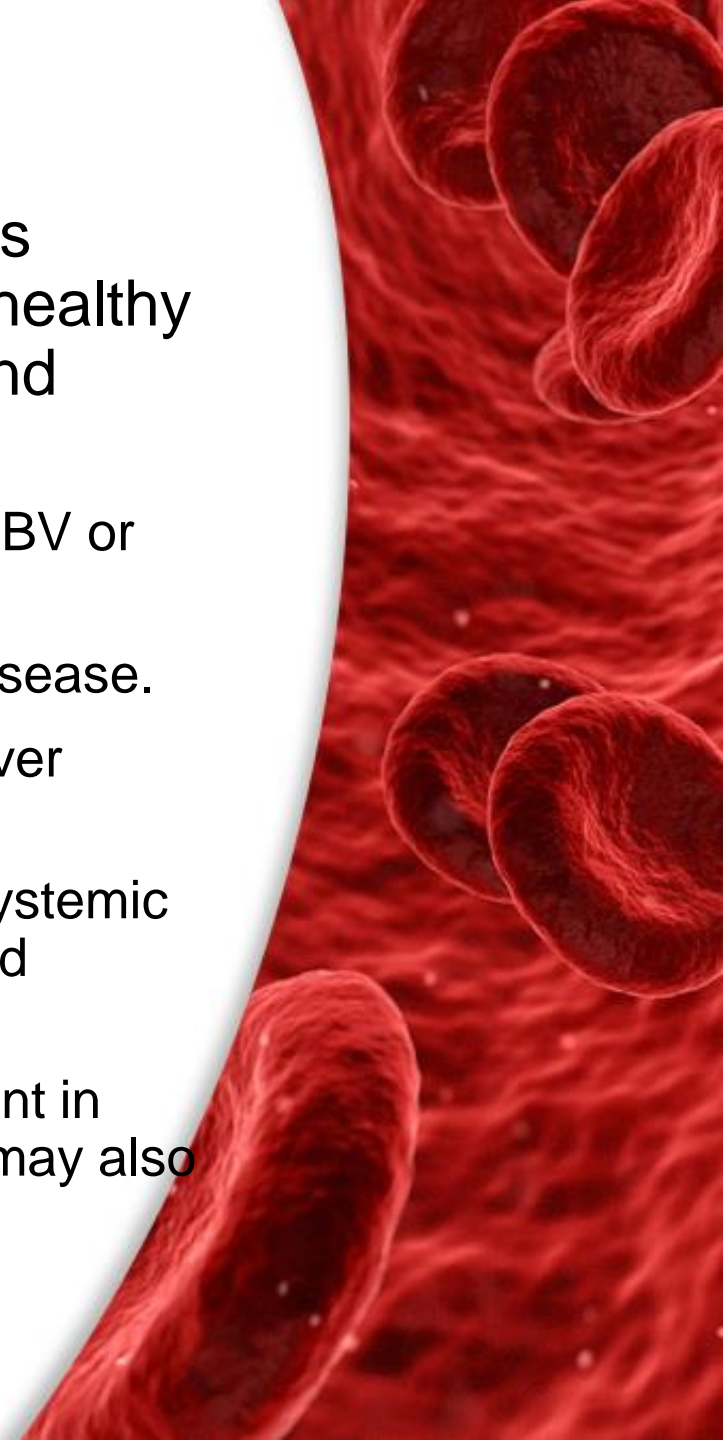




# Examination Findings

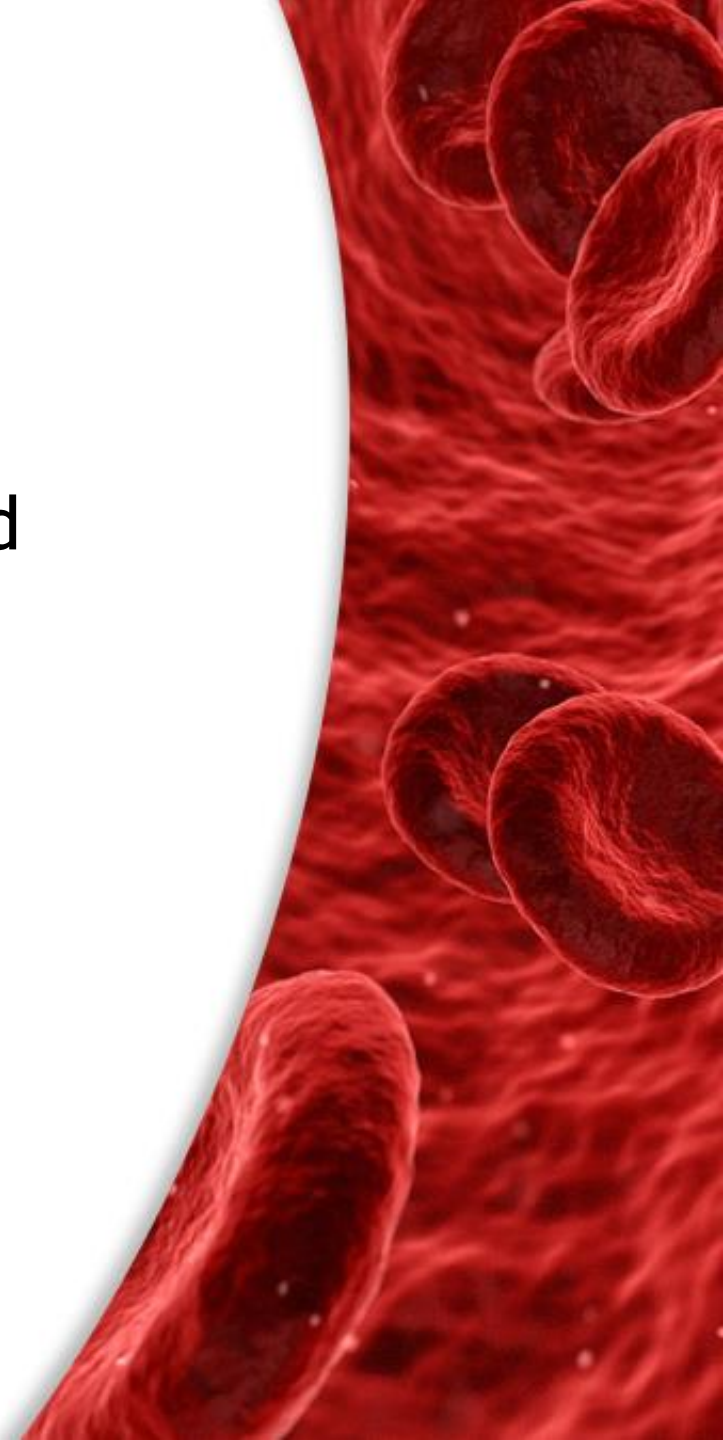
A palpable spleen may represent a normal finding, The spleen is palpable in nearly one-third of healthy neonates, 10 percent of healthy children, and approximately 3 percent of healthy adolescents and young adults

- **Lymphadenopathy** – Lymphadenopathy may be a sign of infection (eg, EBV or HIV-1) or malignancy
- **Jaundice** – Jaundice may indicate underlying hemolytic anemia or liver disease.
- **Hepatomegaly and other signs of liver disease** – Hepatomegaly, firm liver edge, ascites, and/or spider angiomas suggest underlying liver disease.
- **Rashes and/or joint swelling** – Rashes and/or joint swelling suggest a systemic autoimmune disorder such as systemic lupus erythematosus or rheumatoid arthritis.
- **Petechiae and ecchymoses** – Petechiae and ecchymoses may be present in patients with thrombocytopenia resulting from splenic enlargement. They may also be a sign of underlying malignant hematologic disease.
- **Fever**
- **Anemia**



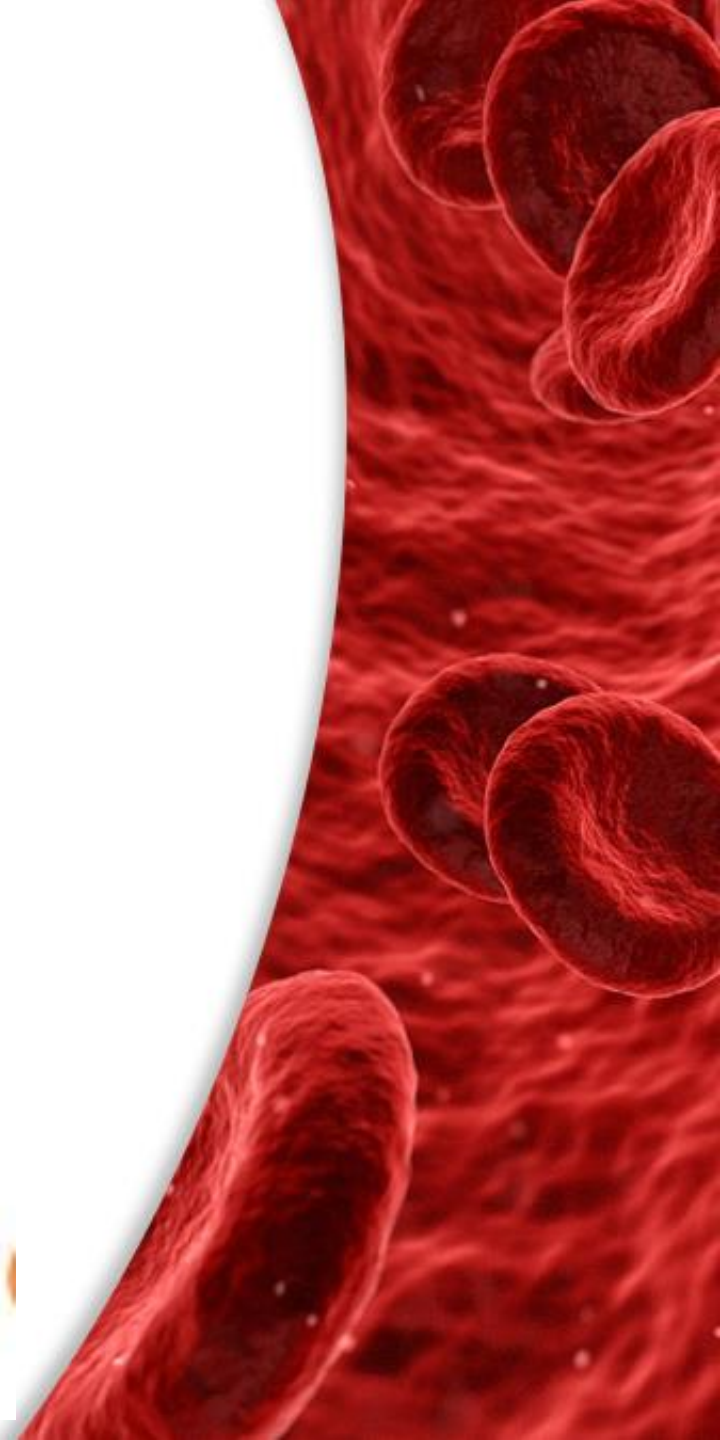
# Initial evaluation of the child with unexplained splenomegaly

- History and physical examination
- Complete blood count including platelet count and differential
- Reticulocyte count
- Review of the peripheral blood smear
- Liver function tests
- EBV and CMV serologies
- Chest radiograph
- Abdominal ultrasound



## IMAGING

- USG- sensitive & specific non-invasive
- CT scan – etiology of splenomegaly
  - liver size, heterogeneity
  - splenic mets, abscess, calcf., cysts
  - retro peritoneal LN
  - craniocaudal ln > 10 cm
- Liver- spleen colloid scan- (RBC –Cr51, Tc99)
  - hepatic steatosis, SOL, splenic functions
  - PHT, colloid shift +
- MRI/ Doppler usg- portal/splenic vein thrombosis
  - cavernomas





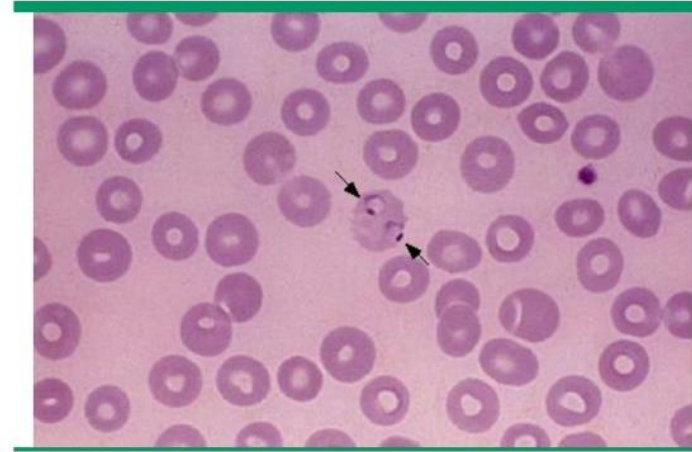
# Suspected infectious

**Viral infections, particularly EBV and CMV, are among the most common causes of splenomegaly in children.**

- Infectious mononucleosis - Fever, pharyngitis, fatigue, and lymphadenopathy - Lymphocytosis and atypical lymphocytes on peripheral blood smear-Diagnosis is made with EBV and CMV serologies and/or heterophile antibody testings

**Other infectious can cause splenomegaly, including:**

- Tuberculosis
- Infective endocarditis
- Malaria
- HIV
- Fungal infections
- Leishmaniasis
- Toxoplasmosis

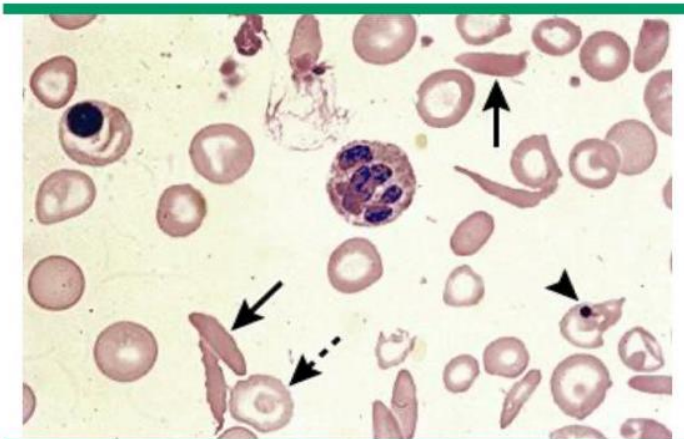


- Testing is guided by clinical findings and may include: TST to evaluate for TB - Serial blood cultures to evaluate for IE- Malaria blood smear (if relevant exposure) -HIV testing (if there are relevant risk factors)-Leishmaniasis to evaluate serology test or BMA

# Cause Of Hemolytic anemia

- **Congenital and acquired hemolytic anemias represent a major cause of splenomegaly .**
- **In these conditions, splenomegaly results from sequestration of destroyed red blood cells.**

**Peripheral blood smear in sickle cell anemia**



## Intrinsic red blood cell defects

Hemoglobinopathies (eg, sickle cell disease, thalassemias)

Membrane defects (eg, hereditary spherocytosis, elliptocytosis)

Enzyme deficiencies (eg, G6PD, pyruvate kinase deficiencies)

## Extrinsic hemolytic processes

Autoimmune hemolytic anemia (AIHA)

- Warm-reactive
- Cold agglutinin disease
- Paroxysmal cold hemoglobinuria

Hypersplenism

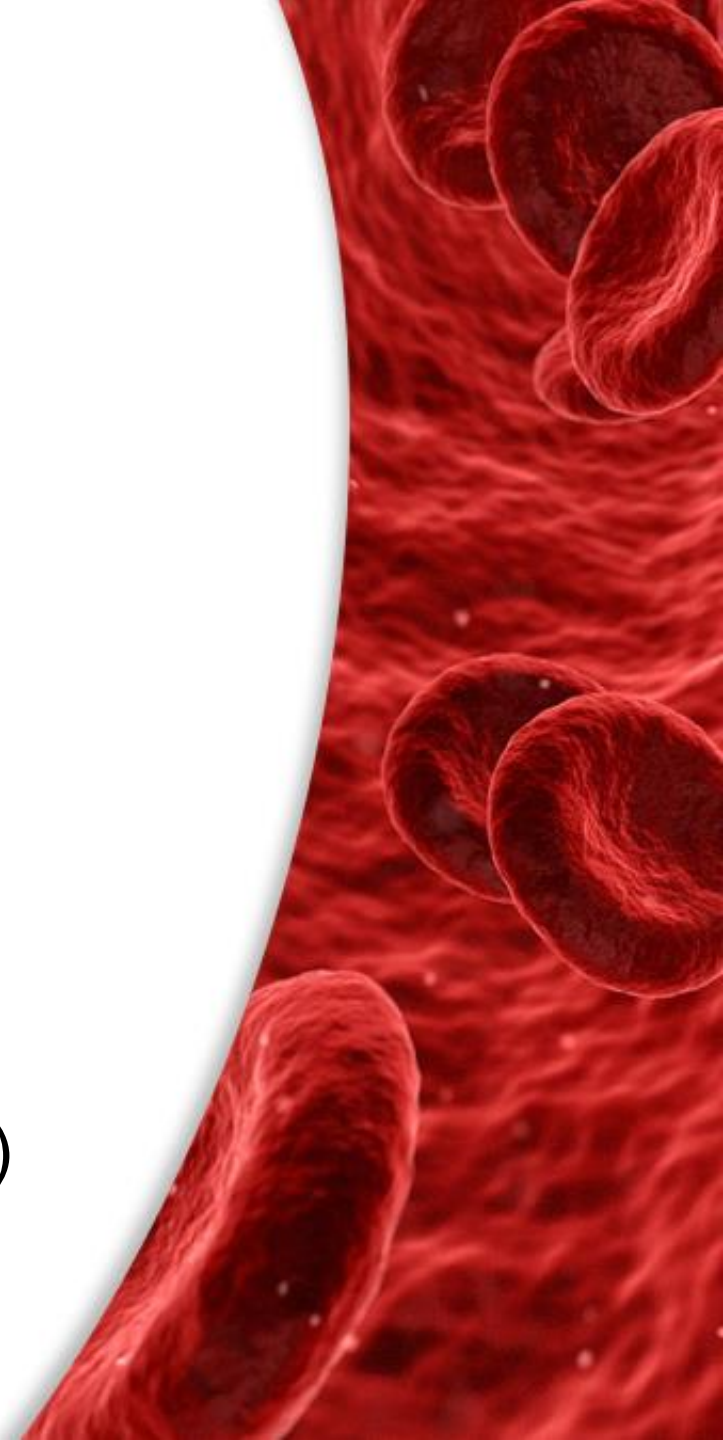
# Suspected hemolytic anemia

## Cause of Hemolytic anemias:

1. **Intrinsic** :RBC membrane defects - RBC enzyme defects - Hemoglobinopathies
2. **Estrinsic** : Autoimmune hemolytic anemia - Other acquired hemolytic anemias

## ➤ Testing for specific causes of hemolytic anemia

- Low hemoglobin
- Unconjugated hyperbilirubin
- Reticulocytosis
- Peripheral blood smear include :(polychromasia – spherocytes)
- Certain congenial RBC disorders may have specific findings on the blood smear (eg, elliptocytes, sickle cells)
- Serum markers of hemolysis (LDH, plasma free hemoglobin,haptoglobin)
- Coombs test





# Hematologic malignancy

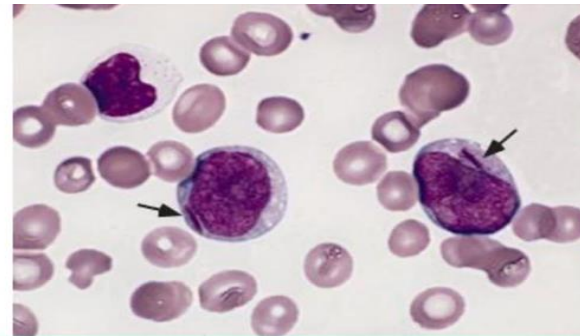
**Childhood leukemias and lymphomas are important causes of splenomegaly.** These include:

- Acute lymphoblastic leukemia (ALL), including precursor B cell and T cell
- lymphoma
- Acute myeloid leukemia JMLL
- Hodgkin lymphoma
- Non-Hodgkin lymphoma

## **Suggestive findings :**

- Persistent fevers, anorexia, weight loss, decreased activity level, bone pain, and/or easy bruising
- Cytopenias (neutropenia, anemia, and/or thrombocytopenia)
- Abnormal immature cells (blasts) in the peripheral Blood

**Diagnosis :** Bone marrow aspiration/biopsy



# Liver Disease/Portal Hypertension

**Splenomegaly is a feature of liver disease, particularly end-stage liver failure (cirrhosis).** Cirrhosis-related splenomegaly results from congestion due to portal hypertension.

## ➤ **Cause of liver disease :**

- Biliary atresia
- Viral hepatitis
- Wilson disease
- Other metabolic liver disease (eg, galactosemia)
- Alpha-1-antitrypsin deficiency
- Cystic fibrosis

## ➤ **Suggestive findings :**Jaundice, hepatomegaly, firm liver edge, ascites, spider angiomas

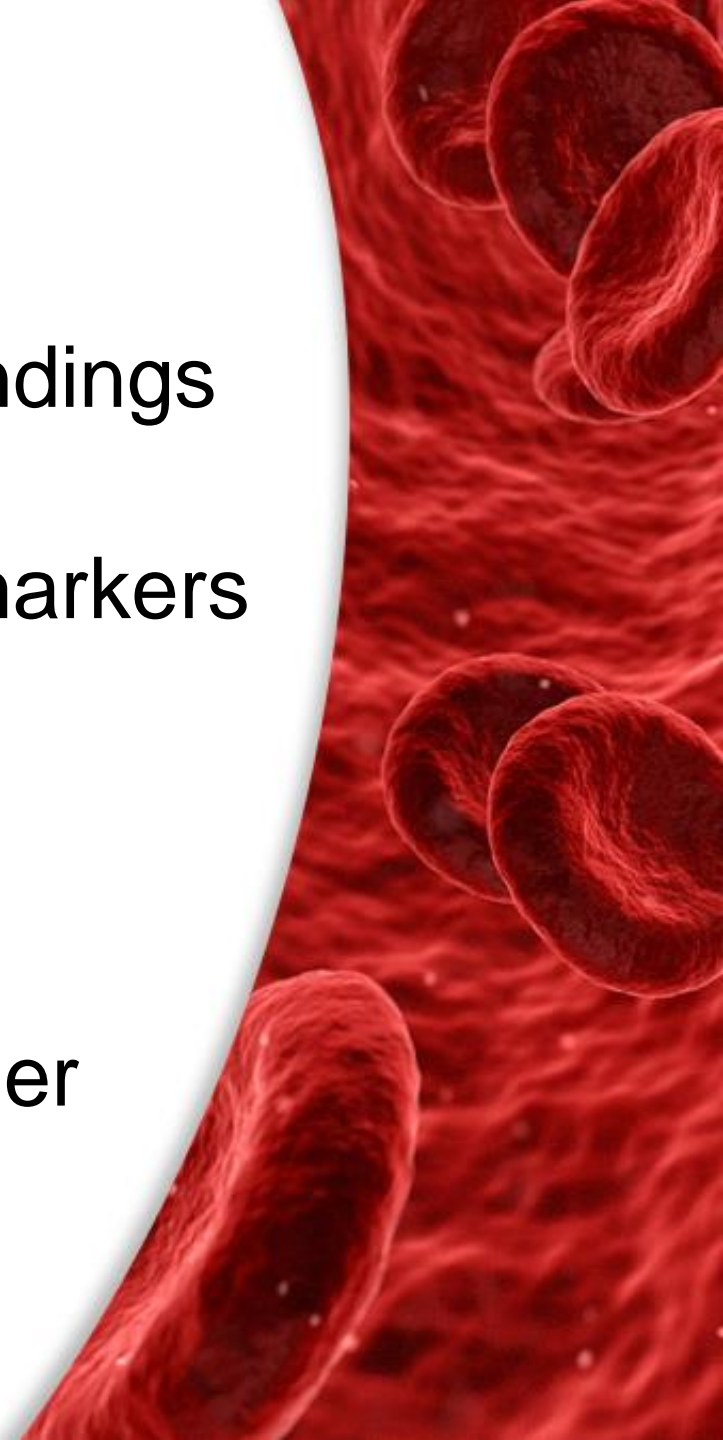
- Abnormal liver function tests
- Abnormal findings on abdominal imaging (slow or reversed portal blood flow suggestive of portal hypertension)
- Liver biopsy may be warranted in some cases



# Autoimmune Disease

Autoimmune disease may be suggested by findings of rashes and/or joint swelling. (**SLE, JRA** )

- **Laboratory findings include** : nonspecific markers of inflammation (eg, leukocytosis, elevated erythrocyte sedimentation rate, or C-reactive protein).
- Additional evaluation includes **antinuclear antibody titers** as a screening test and further evaluation based on the findings





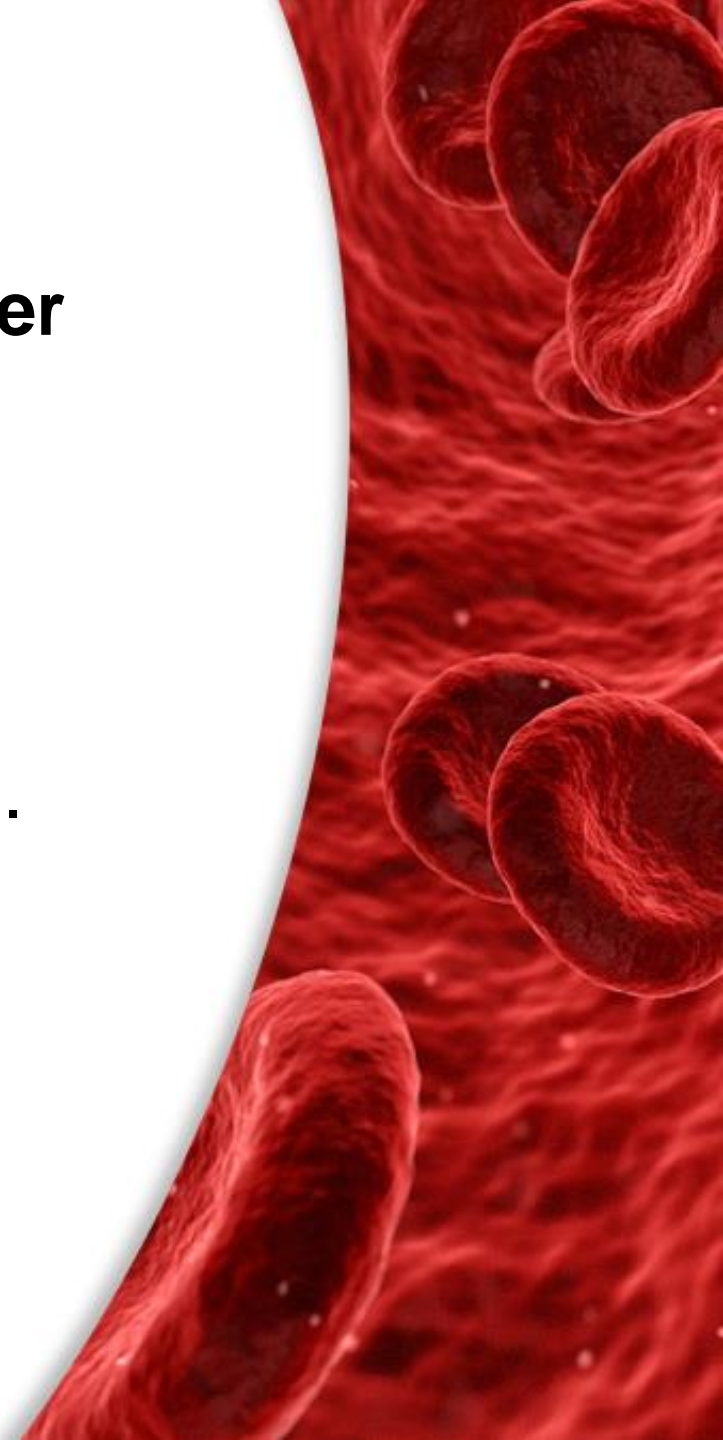
# Storage disorders

**Storage disease may be suspected based on other organ involvement** ( hepatomegaly, skeletal abnormalities, neurologic involvement).

- Plain radiographs may show bone ,focal splenic masses may be noted on ultrasound.
- The diagnosis is confirmed by demonstrating a specific enzyme deficiency and/or genetic mutation.

## **Storage disorders**

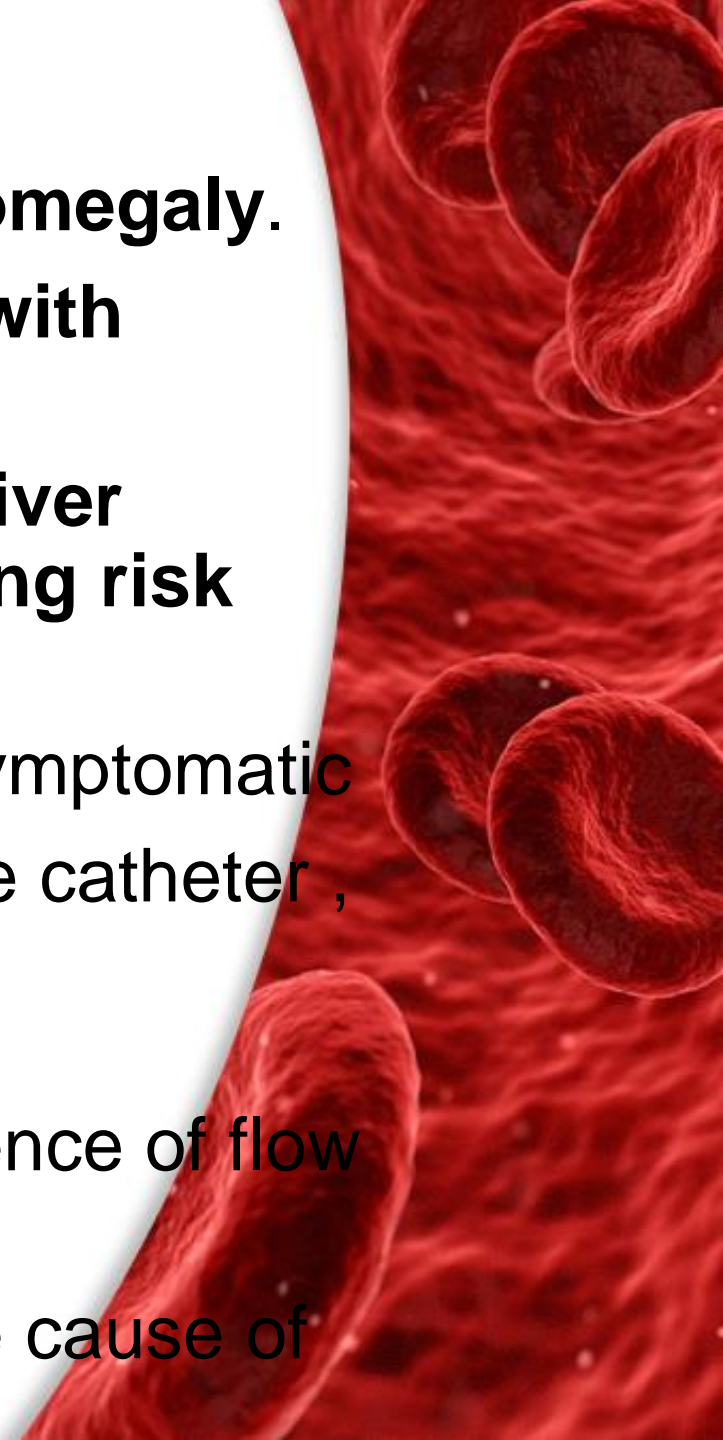
- Gaucher disease
- Niemann-Pick disease
- Mucopolysaccharidoses
- Other lysosomal storage disorders



# Portal vein thrombosis

**(PVT) is a cause of portal hypertension and splenomegaly.**

- **In neonates, PVT is most commonly associated with umbilical venous catheters.**
- **In older children, PVT may be a complication of liver transplant or may occur in children with underlying risk factors .**
- **Suggestive findings:** abdominal pain or may be asymptomatic
- **Underlying risk factor for thrombosis** (eg, invasive catheter , liver disease, malignancy, inherited thrombophilia, antiphospholipid antibodies)
- **Diagnosis:** Doppler ultrasound (filling defect or absence of flow Doppler images)
- Additional evaluation may be warranted to determine cause of thrombus



# Space-occupying lesions

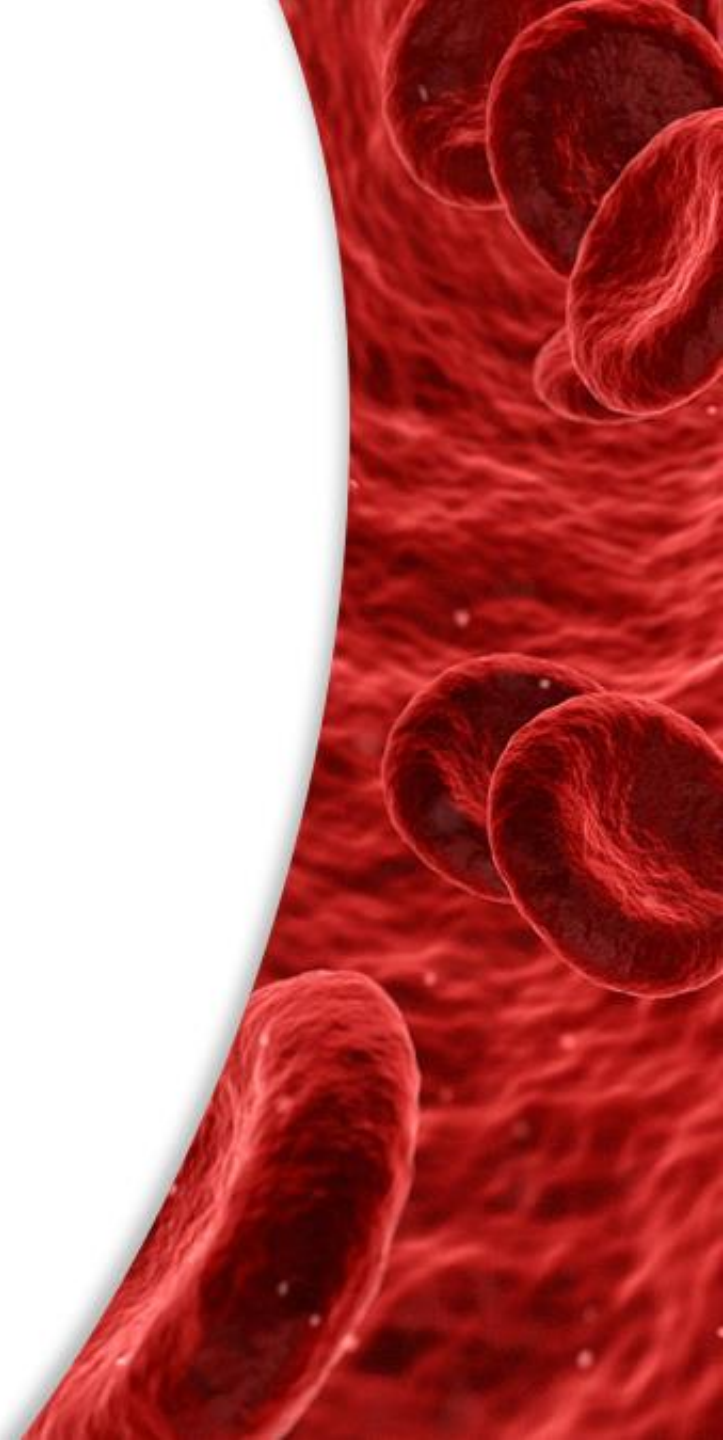
## Hemangioma, Hamartoma:

- Most are asymptomatic
- Additional imaging studies (eg, CT or MRI) may be warranted Diagnosis is based on imaging findings

## Intracapsular hematoma (trauma)

May be noted as incidental findings on abdominal imaging studies obtained for other reasons

- In most cases, there is a clear preceding history of substantial blunt abdominal trauma Relatively minor trauma can cause splenic rupture in children with underlying splenomegaly Hamartoma
- Cysts





# Hypersplenism

- Hypersplenism is a common disorder characterized by an **enlarged spleen** which **causes rapid and premature destruction of blood cells**.
- Hypersplenism is a **syndrome characterized by splenomegaly and any or all of the following cytopenias: anemia, leukopenia, or thrombocytopenia**.
- Implicit in the definition is that the cytopenias will correct after splenectomy.
- patients with splenomegaly do not have hypersplenism. Hypersplenism usually is the result of an identifiable pathologic process

