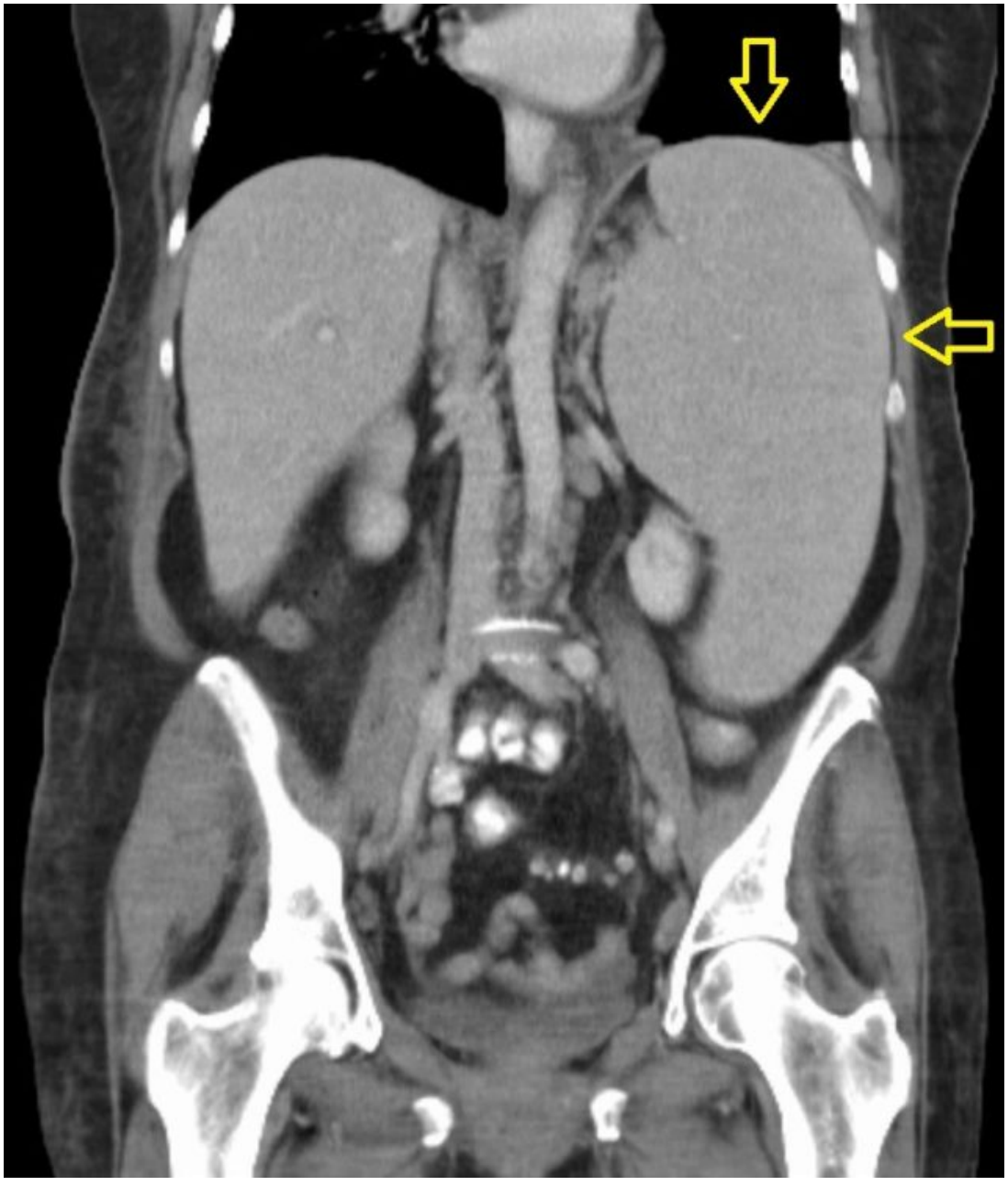
Splenomegaly

See online here

Splenomegaly is the enlargement of the spleen. The spleen is an integral part of the immune system that provides efficient immunosurveillance (production of WBCs defending the body from various infections) and hematopoiesis (“cemetery” for damaged and worn off erythrocytes). The spleen is allocated in the left upper quadrant of the abdomen behind the ribs 9-11, on the left from the stomach, and the tail of the pancreas, above the left kidney, the splenic flexure of the colon.

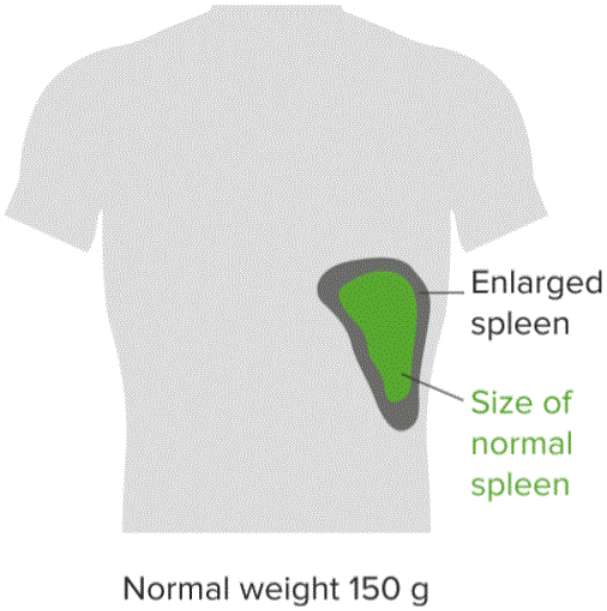


Definition and Background

Spleen: The spleen is a firm abdominal organ, dull red in color, that lies under the diaphragm on the left side. It measures 1 X 3 X 5 inches, weighs approximately 7 oz and lies between the 9th and 11th ribs, hence summarized by the odd numbers 1, 3, 5, 7, 9, and 11.

The spleen consists of the two formations: the red pulp and the white pulp that are covered by a dense fibrous covering called the splenic capsule. The **red pulp** filters damaged red blood cells, hosts macrophages and platelets, and release them into the bloodstream when needed. The **white pulp** is the place where white cells grow and mature (extramedullary hemopoiesis).

The spleen is not a vital organ. Though spleen is the largest lymphoid organ and has a variety of functions, including immunosurveillance and hematopoiesis, the body can manage without the spleen when it is removed due to particular reasons (injury, blood diseases).



"Splenomegaly. Enlarged spleen"

Splenomegaly: Splenomegaly is the enlargement of the spleen. The normal spleen is usually not palpable on abdominal examination, except in adolescents and thin slender build individuals. The splenomegaly is classically a clinical sign, but with technological advancements, even mild splenomegaly is detected on the imaging studies. Usually, when the spleen is palpable, it is often enlarged two-to-three times than the normal size.

Hypersplenism: Hypersplenism means increased functioning of the spleen, while splenomegaly simply denotes an increased size of the spleen. It is not necessary that all the enlarged spleens are hyperfunctioning. The **four cardinal features of hypersplenism** are:

1. Splenomegaly
2. Anemia, leukopenia, thrombocytopenia and/or pancytopenia
3. Bone marrow hyperplasia (compensatory)
4. Improvement after splenectomy

Congestion of blood in the spleen cause:

- Sequestration of blood elements
- Activation of reticuloendothelial cells

Both processes lead to anemia and thrombocytopenia. In some cases there is increased the risk of traumatic splenic rupture, causing potentially fatal hemoperitoneum.

Etiology of Splenomegaly

The common causes of splenomegaly are:

- **Hematological**: The hematological disorders are one of the common causes of splenomegaly that include chronic myeloid leukemia (CML), chronic lymphocytic leukemia (CLL), acute leukemias, lymphomas, primary myelofibrosis, polycythemia vera, hairy cell leukemia, thalassemia, and different anemias.
- **Liver disease and portal hypertension:** Hepatic cirrhosis, portal or splenic vein thrombosis.
- **Storage diseases:** Gaucher's disease, Niemann–Pick disease.
- **Systemic diseases:** Sarcoidosis, amyloidosis, and collagen diseases – systemic lupus erythematosus, rheumatoid arthritis, Felty syndrome.
- **Infections:** Malaria, leishmaniasis, schistosomiasis, severe septicemia, bacterial endocarditis, typhoid, AIDS, hydatid disease, abscesses, and infectious mononucleosis.
- Splenic metastasis, cysts, and hemangiomas.
- Systemic mastocytosis
- Histiocytosis X

Epidemiology of Splenomegaly

Splenomegaly is a common finding and is reported in 2% to 5.6% of patients in the United States. There is no racial predilection but certain causes are more frequent in certain races such as sickle cell anemia in Blacks and certain lysosomal storage diseases in Ashkenazi Jews.

Some causes of splenomegaly have geographical predilection such as tropical splenomegaly syndrome (or hyperactive malarial syndrome), which is common in tropics where malaria is endemic. Tropical splenomegaly syndrome is twice more common in females. The acute leukemias and storage disorders are common in the children and adolescents, while the chronic leukemias are common in the older population.

Clinical Presentation of Splenomegaly

The clinical presentation of patients with splenomegaly depends upon the underlying etiology, for example, malaria presents with high-grade fever, chills, rigors, and sweating. The leukemias present with features of pancytopenia, and cirrhotic patients may present with abdominal distension, ascites, hematemesis and/or hepatic encephalopathy.

The splenomegaly itself is often asymptomatic and detected incidentally. When severely enlarged, it may cause abdominal distension, dull abdominal pain, and early satiety.

It is important to review the family history to identify relevant hereditary diseases, such as hemolytic anemias, and lysosomal storage diseases.

Diagnosis of Splenomegaly

Splenomegaly is a clinical sign rather than a full diagnosis. It is associated with a long list of causes (as described above). When splenomegaly is present, the patients should be properly evaluated to identify the etiologic cause, and in most cases, the causes are infections, portal hypertension, neoplasms, or autoimmune disorders.

Laboratory Studies

- Complete blood counts with peripheral thick and thin films (for malaria)
- Liver function tests and viral hepatitis profile
- Autoimmune screen and inflammatory markers
- Cytogenetic tests (BCR-ABL, tyrosine kinase)
- Karyotyping

Imaging Studies

- Ultrasound examination of the abdomen
- CT scan of the abdomen
- Bone marrow aspirate
- Lymph node biopsy
- Full liver screen

Grading of Splenomegaly

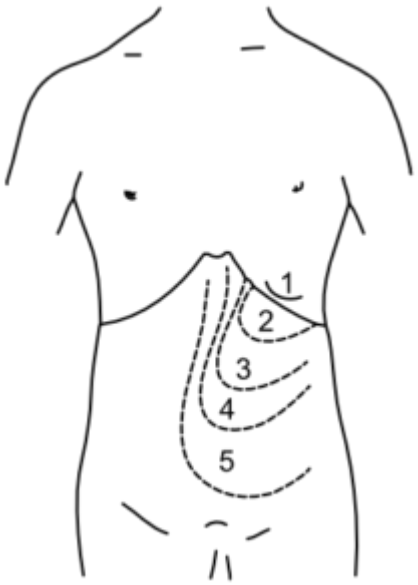


Image : “Splenomegaly” by Furfur. License: CC BY-SA 4.0

The splenomegaly has been graded from grade 0 to 5 according to the Hackett ’ s grading system, which is as follows:

Grade 0: Normal impalpable spleen.

Grade 1: Spleen palpable only on deep inspiration

Grade 2: Spleen palpable on mid-clavicular line, half way between umbilicus and costal

margin

Grade 3: The spleen expands towards the umbilicus

Grade 4: The spleen goes past the umbilicus

Grade 5: The spleen expands towards the pubis symphysis.

Differential Diagnosis of Splenomegaly

The differential diagnosis of splenomegaly is huge. It often can be shortlisted depending upon the splenic size and other clinical features.

The **massive splenomegaly** is when the spleen is palpable >8cm below the left costal margin with the maximum cephalocaudal diameter of spleen being >20cm. The common causes associated with massive splenomegaly are:

- Chronic Myeloid lymphoma (CML)
- Lymphoma
- Primary myelofibrosis
- Thalassaemia major or intermedia
- Gaucher’s disease
- Malaria
- Leishmaniasis
- Schistosomiasis

Moderate splenomegaly is when the spleen is palpable 5-8 cm below left costal margin with the maximum cephalocaudal splenic diameter being 11-20cm.

Mild splenomegaly is when the spleen is palpable <5cm below the left costal margin and maximum cephalocaudal splenic diameter is $\leq$10 cm.

The causes of mild and moderate splenomegaly are as follows:

- Congestive heart failure
- Liver cirrhosis and portal hypertension
- Severe bacterial sepsis
- Infective endocarditis
- Sickle cell anemia
- Acute infectious illnesses (typhoid, malaria, other tropical diseases)
- Acute viral infections (infectious mononucleosis)
- Systemic lupus erythematosus
- Tuberculosis (chronic forms)
- Idiopathic splenomegaly
- Immune hemolytic anemia
- Immune thrombocytopenic disorders
- Trypanosomiasis

Management of Splenomegaly

Conservative

The conservative management of splenomegaly depends upon the underlying disorder and patient’s clinical status. E,g, malaria is treated with antimalarials, infections with

antibiotics and leukemias with chemotherapeutic agents. The splenomegaly often resolves when the underlying cause is appropriately managed. The additional supportive measures may include blood transfusions/exchange transfusions and prophylactic antibiotics.

Low-dose radiotherapy has been used as palliative care in patients with advanced hematological problems (primary myelofibrosis).

Surgery (Splenectomy)

Splenectomy is the surgical removal of the spleen. It is required in certain cases when splenomegaly becomes a reason of the aggravation of the basic condition.

Indications for splenectomy:

- Hemolytic anemias (some cases), e.g. hereditary spherocytosis, autoimmune hemolytic anemia, thalassemia major or intermedia
- Chronic lymphocytic leukemia and lymphomas
- Chronic immune thrombocytopenia
- Primary myelofibrosis
- Tropical splenomegaly
- Splenic trauma and rupture

Recommendations for vaccination of patients before splenectomy

Pneumococcal polyvalent vaccine, *Hemophilus influenzae* type b and meningococcal conjugate vaccines are recommended **two weeks before elective** splenectomy. In cases of urgent splenectomy after trauma or splenic rupture, the vaccines should be given **two weeks after urgent** splenectomy.

Influenza vaccine is recommended annually.

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Notes