

Splenomegaly

General Presentations:

The spleen is a hematopoietic organ supporting the megakaryotic, lymphoid, erythroid, myeloid, and reticuloendothelial systems. It is a reservoir for platelets and a site for the proliferation of B-lymphocytes. Other functions include filtering senescent and defective red blood cells, presenting antigens to the immune system, and destroying antibody coated microorganisms.

Splenomegaly is most often discovered under physical examination. One third of neonates and one tenth of children may have a normal, palpable spleen. However, when the tip of the spleen is felt beyond 2 cm below the left costal margin, it is considered abnormal.

An abnormally enlarged spleen signifies underlying pathological process and requires investigations. Mechanisms of splenomegaly can generally be classified as reactive, congestive, or infiltrative. Reactive mechanisms may include infections, autoimmunity, or hemolysis. Congestive mechanisms may be due to anatomical obstruction or backflow of blood. Infiltration of the spleen may be of a benign or a malignant origin.

Signs of Abnormal Splenomegaly
<ul style="list-style-type: none"> • >2 cm below left costal margin • Abnormally rough surface • Tender • Hard

Questions to ask:

- Is there any history of recent infection? (e.g. rash, pharyngitis, cough, SOB, fever, exposure, poor feeding, malaise etc.)
- Are there any constitutional symptoms such as fever, night sweats, or weight loss?
- Is there any abnormal bruising, bone pain, or history of frequent infections?
- Were there any complications during pregnancy, delivery, and after delivery? Are there any growth and developmental concerns?
- Was there persistent, unresolved jaundice following delivery?
- Does the child have any congenital heart diseases, storage diseases, bleeding disorders or liver diseases?
- Does the child have any history of surgeries or transfusions?
- Is there any family history of hematological diseases, autoimmune diseases, storage diseases, or malignancy? (See list under differential diagnosis) What is the family ethnicity?

- Is there any recent travel?
- Is there any history of trauma?

Differential Diagnoses:

S - P - L - E - E - N
<ul style="list-style-type: none"> • Sequestration of RBC: hereditary spherocytosis or other congenital or acquired hemolytic anemia • Proliferation: *viral, bacterial, fungal or parasitic infections; inflammation such as lupus or RA • Lipid deposition: Gaucher disease, Niemann-Pick disease • Endowment: congenital splenic hemangioma, hamartoma, or cysts • Engorgement: splenic trauma (intracapsular hematoma), *sequestration crisis in sickle cell disease, heart failure, portal hypertension • iNvasion: *malignancy, granulomatous, histiocytic, lymphoproliferation <p><i>*DO NOT MISS: sequestration crisis in sickle disease is a medical emergency with 10-15% mortality in the pediatric population!</i></p>

Investigations:

- A) General physical exam:
 - **DERM:** Petechiae and purpura (thrombocytopenia, autoimmune disorder, malignancy); jaundice (hemolytic anemia or liver disease); rashes (infection, lupus, RA, infective endocarditis)
 - **HEENT:** Icterus, cherry red retinal spots or cloudy cornea (lipid storage disease)
 - **CV/ RESP:** Murmur, SOB, fatigue (anemia or heart failure)
 - **GI:** tenderness, distension, ascites, hepatomegaly (liver disease, gallstones, trauma, and hemolytic anemia)
 - **MSK:** joint tenderness (RA, lupus, hepatitis), bone pain (malignancy)
 - **Neuro:** Poor vision (osteopetrosis), uveitis, iritis (sarcoidosis or rheumatoid arthritis), loss of developmental milestones (storage diseases, chronic infection, or immunodeficiency)
- B) Splenic Exam:
 - Bimanual Palpation:

- Patient should be supine and relaxed
- Relaxation is improved if legs and neck are slightly flexed
- Start palpating from **lower left quadrant in infants**, as the spleen tends to enlarge inferiorly towards the left iliac fossa. Palpation should be started from the **right lower quadrant in older children**.
- Percussion:
 - Castell's method: percuss the lowest intercostal space in the left anterior axillary line. Dullness to percussion indicates splenomegaly.
 - Traube's space: bound superiorly by the 6th rib; laterally by the mid-axillary line and inferiorly by the costal margin. Dullness to percussion indicates splenomegaly
- C) Laboratory Investigations:
 - CBC with differential
 - Peripheral blood smear
 - AST, ALT, GGT, alkaline phosphatase, bilirubin, prothrombin time, total protein and albumin
 - Additional investigations based on clinical suspicion:
 - Viral serology (EBV, CMV, Parvovirus B19, HIV etc.), acid beta-glucosidase (↓ in Gaucher disease), ANA (for SLE), bone marrow aspirate and biopsy
- D) Splenic Imaging (not routinely done):
 - Ultrasound (for identifying space occupying lesions such as cysts or abscess and differentiating between kidney vs splenic abnormalities)
 - CT scan or MRI: (for ruling out disseminated malignancy and liver diseases)
 - Radioactive (Tc-99m) sulfur colloid scintigraphy (the only radiological modality that provides functional information)

Conclusion

Splenomegaly is a symptom that can be caused by a variety of conditions that affect different organ systems. When evaluating a child with splenomegaly it is important to identify conditions in which immediate medical intervention is needed and provide appropriate treatment options.

References:

1) McClain K, Landaw S. Approach to the child with an enlarged spleen. (Last updated: July 22, 2008) In: UpToDate, Mahoney D, Hoppin A (Ed), UpToDate, Wellesley, MA, 2010.

- 2) Kainth M, Kanwar V, Sills R. Splenomegaly. eMedicine (Last updated September 4,2008). Available from: <http://emedicine.medscape.com/article/958739-overview> [Accessed on January 4, 2010]
- 3) Bickley L, Szilagyi P, Bates B. Bate's Guide to Physical Examination and History Taking, 10th ed. Philadelphia: Wolters Kluwer Health/ Lippincott Williams & Wilkins, 2009.
- 4) Kliegman R, Behrman, Jenson H, Stanton B. Nelson Textbook of Pediatrics, 18th ed. Philadelphia: Saunders, 2007.