Splenomegaly

Definition / Supporting Information

The spleen tip is palpable in nearly one-third of healthy newborns and up to 10 percent of healthy children. A spleen which is enlarged more than 2 cm below the costal margin should be investigated.

Keywords / also known as: enlarged spleen

Essential History

Ask about:

- Birth history and the early neonatal period
  - A history of umbilical vein catheterisation or omphalitis in the neonatal period may suggest a diagnosis of portal vein thrombosis.
- Family history of:
  - Anaemia
  - Jaundice
  - Splenomegaly
  - Splenectomy
- Fever
- Pharyngitis
- Malaise
- Night sweats
- Weight loss
- Rash
- Gastrointestinal (GI) bleeding (see Haematemesis / Melaena)
- Jaundice
- Arthralgia (see Joint Pain)
- Bone pain
- Travel history
- History of trauma

‘Red Flag’ Symptoms and Signs

Ask about:

- Weight loss
- Bone pain
- Night sweats

- GI bleeding (see Haematemesis / Melaena)

Look for:

- Concomitant hepatomegaly, lymphadenopathy
- Signs of an underlying disease
  - Petechiae
  - Purpura
  - Pallor
- Firm to hard consistency
- Distended abdominal veins

Differential Diagnosis / Conditions

- Infection
  - Viral infection
    - The most common cause of splenomegaly in children
    - Splenic enlargement is usually transient and mild to moderate in severity.
    - Common viral infections include:
      - Epstein–Barr virus (infectious mononucleosis; splenomegaly occurs in 50–75% of cases of infectious mononucleosis)
      - Cytomegalovirus
      - HIV
  - Bacterial infection
    - Acute bacterial infections
    - Subacute bacterial endocarditis
    - Congenital syphilis
    - Tuberculosis
    - Other chronic bacterial infections
  - Parasitic infection
    - Toxoplasmosis
    - Malaria
    - Leishmaniasis
  - Fungal infection
    - Candidiasis
    - Histoplasmosis
    - Coccidioidomycosis
- Haematological disorders
  - Acute splenic sequestration crisis is a medical emergency that requires prompt recognition and treatment.
  - Haemolytic anaemias (congenital and acquired)

        - Red cell membrane defects (hereditary spherocytosis / hereditary elliptocytosis)
        - Red cell haemoglobin defects (sickle cell disease and related syndromes / thalassaemia)
        - Red cell enzyme defects (pyruvate kinase deficiency / glucose-6-phospate dehydrogenase deficiency / others)
        - Autoimmune haemolytic anaemia
    - Extramedullary haematopoiesis
        - Occurs in diseases associated with increased demand on the bone marrow for cell production (thalassaemia major / osteopetrosis / myelofibrosis)
- Infiltrative disorders
    - In malignant infiltration, the spleen is firm, massively enlarged, and crosses the midline of the body.
    - The spleen is commonly infiltrated in:
        - Leukaemias and lymphomas
        - Lipidoses
        - Mucopolysaccharidosis
        - Langerhans cell histiocytosis
    - Metastatic neoplasia of the spleen is rare and is usually caused by neuroblastoma.
- Congestive splenomegaly (Banti's syndrome)
    - Splenomegaly may occur from obstruction of the hepatic, portal, or splenic veins.
    - Common causes include:
        - Portal vein thrombosis
        - Cirrhosis
        - Congestive heart failure
    - Umbilical vein catheterisation or septic omphalitis in neonates may also result in obliteration of the portal, hepatic, or splenic veins.
- Inflammatory diseases
    - Systemic lupus erythematosus
    - Systemic onset juvenile idiopathic arthritis (Still's disease)
    - Serum sickness
    - Sarcoidosis
    - Immune thrombocytopenias and neutropenias
- Primary splenic disorders
    - Splenoptosis (wandering spleen)
        - A congenital fusion anomaly of the dorsal mesogastrium
        - Results in a spleen of normal size that moves freely within the peritoneal cavity

  - Cysts
    - Splenic cysts may mimic splenomegaly.
  - Haemangiomas and lymphangiomas
  - Subcapsular haemorrhage
    - Abdominal trauma may cause subcapsular haemorrhage of the spleen.
  - Accessory spleen may also mimic splenomegaly.
    - Found in 10–15% of individuals

Investigations

To be undertaken by non-specialist practitioners (eg, General Practitioner (GP) Team):

- Full blood count
- Leucocyte differential
- Reticulocyte count
- Examination of the peripheral blood smear
- Glandular fever screening
- C-reactive protein / erythrocyte sedimentation rate

To be undertaken by specialist practitioners (eg, Emergency Department / General Paediatric / Paediatric Gastroenterology Team(s)) if not already done:

- Ultrasonography
- Computed tomography
- Magnetic resonance imaging
  - Can further clarify abnormalities in size and shape
- Bone marrow aspiration

Treatment Approach

To be undertaken by non-specialist practitioners (eg, GP Team):

- Viral causes of splenomegaly require supportive care
- If needed seek telephone advice from the Paediatrician
- Treatment of splenomegaly should be aimed at the underlying disease entity.
  - Patients who have bacterial infections should receive appropriate antibiotic therapy.

To be undertaken by specialist practitioners (eg, Emergency Department / General Paediatric / Paediatric Gastroenterology Team(s)):

- Splenectomy – total or partial
  - May be indicated to help to control or stage some diseases that cause splenomegaly
    - Hereditary spherocytosis
    - Autoimmune thrombocytopenia or haemolysis
    - Lymphoma, Hodgkin's disease
  - Splenectomy may also be indicated to treat chronic, severe hypersplenism

When to Refer

Refer to a paediatric specialist if:

- Splenomegaly with concomitant hepatomegaly
- Palpation of a hard spleen
- Suspicion of cancer or other infiltrative disorders (see Suspected cancer: recognition and referral [NICE guideline 12, recommendations 1.10]
- Evidence of haemolytic anaemias
- Suspected splenic crisis (eg, in known sickle cell disease (SCD))
  - Young patients with SCD can develop splenic sequestration crisis which can cause hypovolaemic shock and lead to death
- Evidence of portal hypertension

'Safety Netting' Advice

- All children with splenomegaly should be counselled about the risks associated with contact sports.
- All children without spleens should:
  - Carry an HSC Public Health Agency splenectomy wallet card or necklet
  - Receive appropriate immunisations (given at least two weeks before splenectomy or at least two weeks afterwards):
    - *Pneumococcal* (see PHE Green Book Chapter 25)
    - *Haemophilus influenzae* type b (Hib) (see PHE Green Book Chapter 16)
    - *Meningococcal* ACWY conjugate (see PHE Green Book Chapter 22)
    - Yearly flu vaccine (see PHE Green Book Chapter 19)
  - Take daily antimicrobial prophylaxis against pneumococcal infections (in addition to immunisation):
    - Phenoxymethylpenicillin (Penicillin V).
    - Amoxicillin if cover is also needed for *H. influenzae*.
    - Erythromycin if penicillin-allergic.

- Antibacterial prophylaxis may be discontinued in children over 5 years of age with sickle-cell disease who have received pneumococcal immunisation and who do not have a history of severe pneumococcal infection.

Patient / Carer Information

Please note: whilst these resources have been developed to a high standard they may not be specific to children.

- Splenomegaly and Hypersplenism (Web page), Patient

Resources

National Clinical Guidance

Suspected cancer: recognition and referral (Web page), NICE clinical guideline NG12, National Institute for Health and Care Excellence

Medical Decision Support

Pneumococcal (Web page), Public Health England's Green Book

Haemophilus influenzae type b (Hib) (Web page), Public Health England's Green Book

Meningococcal (Web page), Public Health England's Green Book

Influenza (Web page), Public Health England's Green Book

Suggested Resources

Please note: these resources include links to external websites. These resources may not have national accreditation and therefore PCO UK cannot guarantee the accuracy of the content.

Kelly D, Bremner R, Hartley J, Flynn D (eds). The infant with splenomegaly. In: Practical Approach to Paediatric Gastroenterology, Hepatology and Nutrition. Oxford, UK: John Wiley & Sons; 2014

Bates B, Bickley LS, Hoekelman RA., A Guide to Physical Examination and History Taking. 6th ed. Philadelphia, PA: JB Lippincott; 1995

Buescher ES. Infections associated with pediatric sport participation. Pediatr Clin North Am. 2002;49(4):743-751 [PubMed]

Megremis SD, Vlachonikolis IG, Tsilimigaki AM. Spleen length in childhood with US: normal values based on age, sex, and somatometric parameters. Radiology. 2004;231(1):129-134 [PubMed]

Pearson HA. The spleen and disturbances of splenic function. In: Nathan DG, Orkin SH, Lampert R (eds). Nathan and Oski's Hematology of Infancy and Childhood. 5th ed. Philadelphia, PA: WB Saunders; 1997

Acknowledgements

Content Editor: Dr Srini Bandi

Clinical Expert Reviewer: Dr Patrick McKiernan

GP Reviewer: Dr Daniel Lang

AAP Reviewer: Jane Meschan Foy, MD, FAAP

Paediatric Trainee Reviewer: Dr Akhila Vasanth Hassan

Update reviewer: Dr Pemantah Sandheeah Ramdeny (trainee paediatrician)

Paediatric Specialty Group: British Society for Paediatric Gastroenterology, Hepatology and Nutrition

Update information

Created: 2015

Date last updated: 2018

Next review due: 2021