Splenomegaly

Introduction

Splenomegaly is a common finding on clinical examination, and frequently features in postgraduate assessments. The spleen does not normally descend below the left costal margin. The routine abdominal examination will identify whether the spleen is palpable, and if so splenomegaly is almost universally present. This is generally pathological and warrants investigation.

Clinical anatomy

The spleen is an ovoid organ, covered by a delicate fibroelastic capsule and surrounded by peritoneum, except at its hilum where its pedicles are the splenorenal and gastrosplenic ligaments (Moore and Daley, 2006). It sits in the left hypochondrium, posterior to the stomach, anterior to the diaphragm, superior to the left colic flexure and lateral to the left kidney (*Figure 1*). Its blood supply derives from the splenic artery, which subdivides into five branches that enter the hilum. As these do not anastomose, they generate vascular segments that allow for subtotal splenectomy.

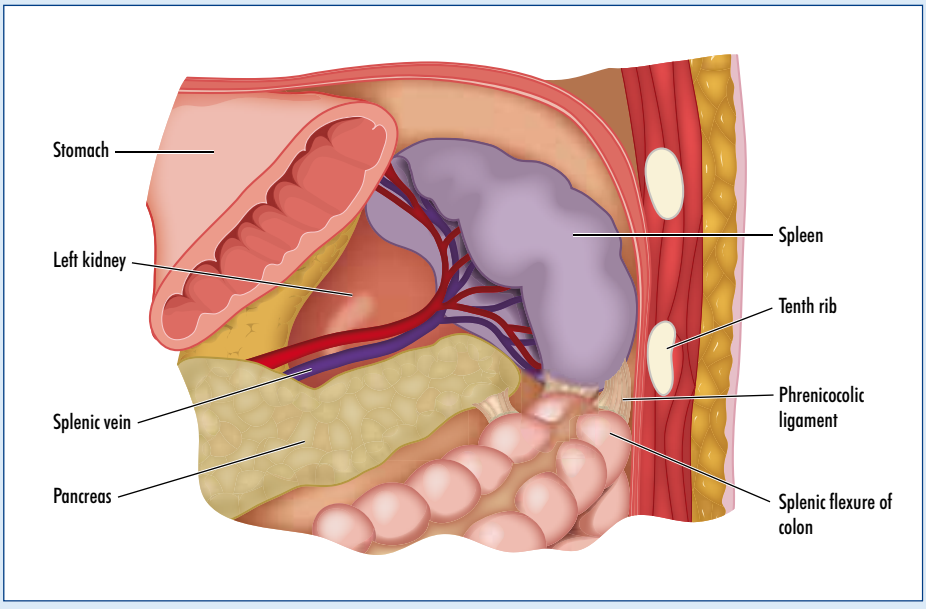
The surface anatomy of the spleen is such that it normally lies between the upper border of the ninth and the lower border of the eleventh ribs (*Figure 2*). When supine, the long axis runs parallel to the tenth rib. Its highest point is 4 cm lateral to the ninth thoracic spinous process, and the lowest is in the mid-axillary line at the level of the first lumbar spinous process. The superior surface curves convexly to fit the inferior surface of the diaphragm, the anterior border is sharp and notched, and the medial/inferior border rounded. When splenomegaly develops, it enlarges towards the right iliac fossa, with its notched anterior surface inferomedial to the left costal margin.

The physiological spleen varies in size and shape. The gold standard for determining splenomegaly is weight, typically defined as greater than 250 g (Pozo et al, 2009). However, this cannot be determined in vivo so size is a more practical measure. Its normal length is 8.5–11.5 cm, and width 5.5–7.5 cm (Larson et al, 1971).

Dr Nicholas F Brown *is FY1 in General Surgery,* **Dr Daniel JB Marks** *is Academic Clinical Fellow in Translational Medicine,* **Dr Philip J Smith** *is ST4 Academic Clinical Fellow in Gastroenterology and MRC Clinical Research Training Fellow and* **Dr Stuart L Bloom** *is Consultant in Gastroenterology, University College London Hospital, London NW1 2BU*

Correspondence to: Dr DJB Marks (d.marks@ucl.ac.uk)

Figure 1. Anatomy of the spleen and relations within the abdominal cavity.



Examining the spleen

Inspection

Splenomegaly is seldom visible on inspection, but may be apparent if the patient is particularly lean or the spleen is massively enlarged (*Figure 3*).

Palpation

Palpation of the spleen begins with the patient supine. Although textbooks classically state that it must at least double or triple in size before the tip is palpable below the costal margin, a 40% enlargement may in fact be detectable (Blackburn, 1953). Using the fingertips, palpate in the right iliac fossa as the patient inspires. If the spleen is not felt, move progressively in 2 cm steps towards the left hypochondri-

Figure 2. Surface anatomy of the normal spleen.

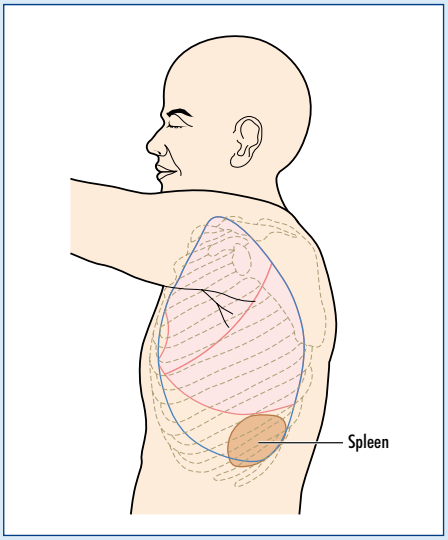
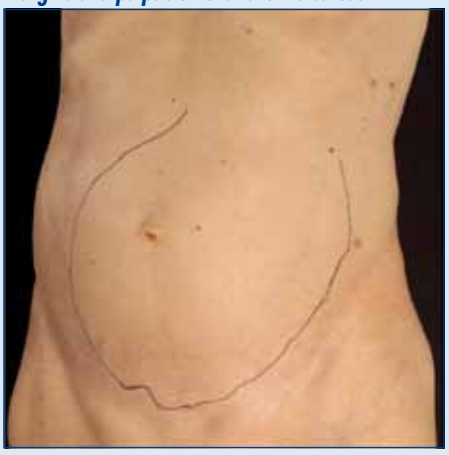


Figure 3. Massive splenomegaly in a patient with a haematological proliferative disorder. The margins and palpable notch are indicated.



um. If no mass is found, the Middleton method involves curling the fingers of both hands under the left costal margin (*Figure 4*), and asking the patient to inspire (Shaw and Dvorak, 1973). If this is still unrewarding, lie the patient in the right lateral decubitus position. Lift the lower ribcage anteriorly with the left hand, and palpate underneath for the spleen with the right (Grover et al, 1993).

Other potential sources of a mass in the left upper quadrant include the kidney (*Table 1* shows means of differentiation), stomach, pancreas, colon, adrenal gland, lymph nodes and liver.

Percussion

As the spleen enlarges into the abdomen, tympanic resonance is lost as air-containing bowel, stomach and lungs are displaced. Percussion begins as with palpation in the right iliac fossa, moving towards the left hypochondrium. In the event that an enlarged spleen has not yet advanced beyond the costal margin, there are a number of eponymous techniques to ensure it is not missed:

- Space of Traube. With the patient supine, percuss within a triangular area delineated by the sixth rib superiorly, mid-axillary line laterally and left costal margin inferiorly (*Figure 5*); this will be dull if splenomegaly is present (Barkun et al, 1991).
- Castell method. With the patient supine, percuss in the lowest intercostal space in the left anterior axillary line on both inspiration and expiration (*Figure 6*), looking for dullness with either (Castell, 1967).
- Nixon method. With the patient in the right lateral decubitus position, percuss along a line perpendicular to the midpoint of the left costal margin (*Figure 7*). Dullness extending more than 8 cm above the costal margin suggests splenomegaly (Sullivan and Williams, 1976).

Note that while resonance on percussion will help exclude splenomegaly, its absence could also result from other pathologies, particularly pleural effusion.

Reliability of these methods

Several studies have evaluated these techniques by comparing examination findings from multiple clinicians against spleen size on ultrasound or scintigraphy. Discriminant validity (the ability of a test to identify patients with splenomegaly from a group of otherwise normal individuals) has also been assessed (Holzbach et al, 1962; Halpern et al, 1974; Sullivan and Williams, 1976; Barkun et al, 1991; Grover et al, 1993).

All methods of palpation (supine, Middleton and right lateral decubitus) had similar discriminating abilities of 73–79%, with none overtly superior. Reliability varied according to body habitus, with validity rising to 83% in lean patients. The percussive methods (Traube, Castell and Nixon) had sensitivities of 60–80% and specificities of 70–80%, with no significant differences between the techniques. Importantly, if percussion was resonant, palpation added no supplemental clinical information. When com-

Figure 4. The Middleton method for splenic palpation. With the patient supine, curl the finger tips of both hands under the left costal margin, and apply pressure in a cranial direction.

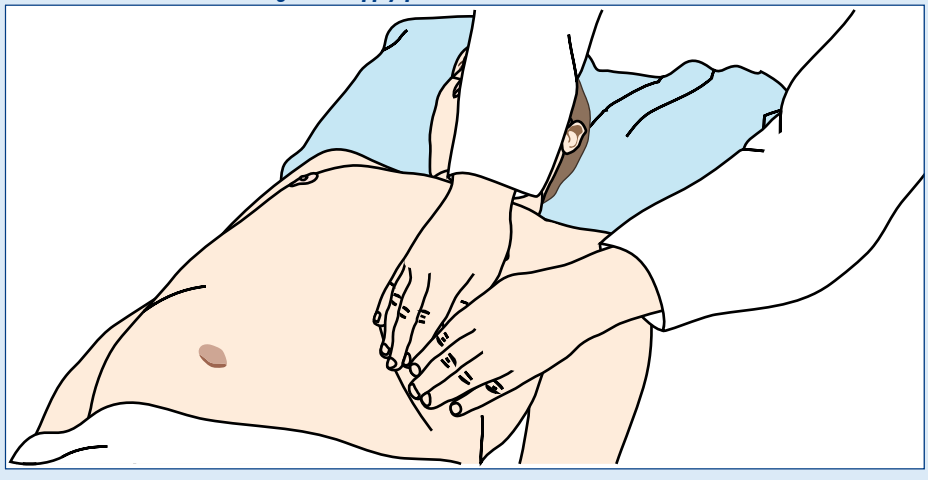


Table 1. Differentiating splenomegaly from an enlarged kidney

On inspiration, spleen moves towards right iliac fossa, kidney moves towards left iliac fossa
Spleen notched on the anterior border
Spleen dull to percussion; kidney retroperitoneal and therefore resonant as overlying bowel
The upper border of the kidney can be palpated; in the spleen it is hidden subchondrally
Kidney ballotable

Figure 5. The space of Traube. The anatomical boundaries are the left midaxillary line laterally, 6th rib superiorly and left costal margin inferiorly.

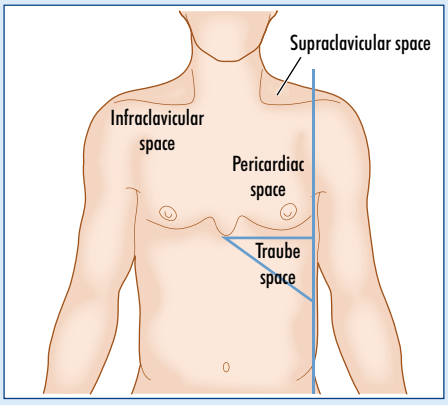
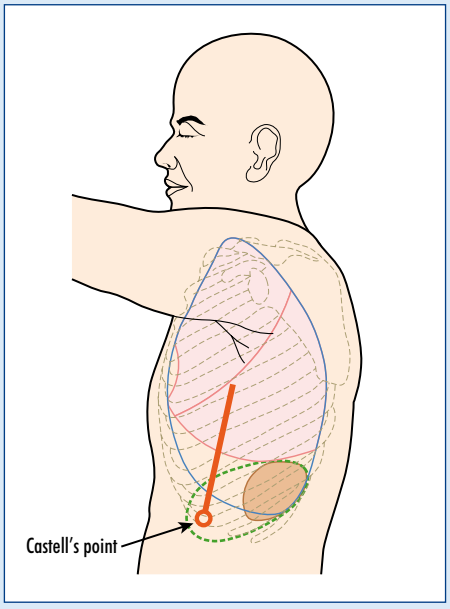


Figure 6. Castell test. Castell's point is the lowest intercostal space in the left anterior axillary line. Percuss on both inspiration and expiration.



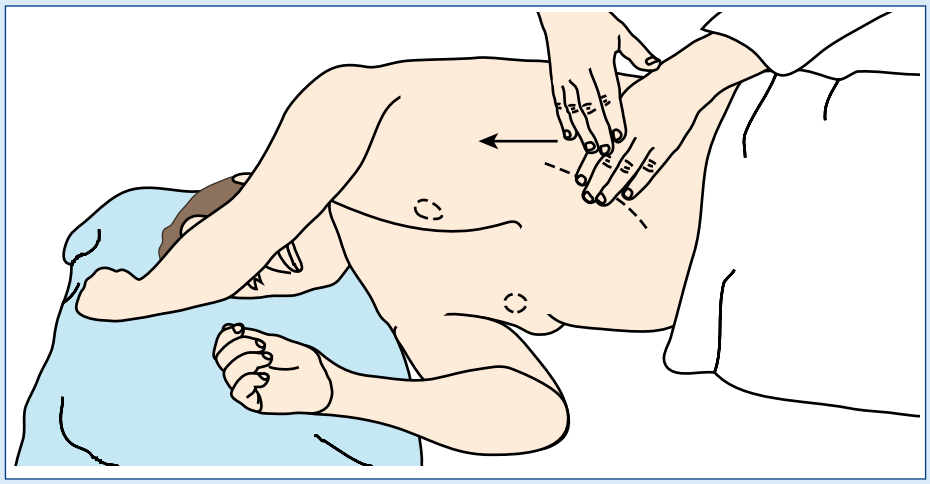


Figure 7. Nixon test. Identify the midpoint of the left costal margin. Percuss along a line perpendicular to this point, moving in a cranial direction. Loss of resonance extending for more than 8 cm above the costal margin is pathological.

bined, positive palpation and percussion still had sensitivity of 72% but specificity rose to 97%.

If both palpation and percussion suggest splenomegaly, the examiner can be confident in its diagnosis. If both are negative, massive splenic enlargement is improbable, but there will still be a 28% false negative rate compared to gold standard imaging. A useful tip, however, is that if the findings on attempts at splenic palpation are equivocal, the percussive manoeuvres help clarify the situation. If these are resonant, it is highly unlikely that any potential mass is an enlarged spleen.

Today the majority of cases of splenomegaly are probably first identified on imaging, usually ultrasonography (Mellstedt, 2007).

Completing the examination

To complete the examination, potential causes of secondary splenomegaly should be sought to provide clues as to the underlying aetiology. Examine for associated hepatomegaly, signs of chronic liver disease, lymphadenopathy and systemic autoimmune diseases such as rheumatoid arthritis or systemic lupus erythematosus. Also assess for sequelae of hypersplenism, including pallor, petechiae and bruising.

Causes of splenomegaly

When constructing the differential diagnosis, it is useful to divide aetiologies into those which can cause massive splenomegaly (>1000 g, or more practically where the spleen crosses the midline), which has a limited number of causes (*Table 2*), as opposed to non-massive splenomegaly (*Table 3*) (Swaroop and O'Reilly, 1999).

Investigation and management of splenomegaly

Splenomegaly almost always warrants investigation, initially with a combination of blood tests and imaging. In addition to routine laboratory investigations (including liver biochemistry) request a peripheral blood film, serum lactate dehydrogenase level, vitamin B_{12} and folate, Monospot test, serology for hepatitis B, hepatitis C, cytomegalovirus and human immunodeficiency virus, bacterial cultures and an autoimmune screen. Further specific microbiological testing (including malaria films) will be guided by a history of travel, occupational and recreational exposures. The full blood count will also identify haematological consequences of hypersplenism (the combination of splenomegaly and peripheral blood cytopaenias).

Ultrasound is the initial imaging modality of choice; as well as evaluating the spleen, it provides useful information regarding the liver, including portal and hepatic venous Doppler flows (Pozo et al, 2009). Computed tomography reconstruction and direct measurement is, however, more reproducible and reliable, and affords better detection of lymphadenopathy (not just restricted to the abdomen); it should be requested in patients in whom a neoplastic disorder is suspected. Echocardiography should be performed when diagnoses of endocarditis or pyogenic abscesses are possible. Patients with a possible or confirmed haematological malignancy may require bone marrow aspiration and trephine for further assessment.

In patients in whom an obvious cause is not identified, the management options are 'watchful waiting', splenic biopsy or diagnostic splenectomy. Watchful waiting can be adopted in young patients with mild splenomegaly and no systemic features (Pozo et al, 2009); one study identified that 3% of such individuals had persistent non-pathological palpable spleens (McIntyre and Ebaugh, 1967). Splenic biopsy can comprise fine needle aspiration and/or core biopsy. The procedure is safe with an overall adverse event rate of approximately 5%, with major complications in less than 1% (Civardi et al, 2001). Diagnostic splenectomy is rarely performed as the risks include mortality of 2.4% and postoperative complications in 41% (Pozo et al, 2009).

Specific management is dictated by the underlying cause. Cytopaenias resulting from hypersplenism are managed supportively (blood transfusions to correct anaemia, antibiotic prophylaxis or treatment of infections, and platelet transfusion as per routine guidelines). Indications for splenectomy are shown in *Table 4*.

Conclusions

Splenomegaly is frequently identified on clinical examination, and generally pathological. It is also a common sign to appear in postgraduate examinations. A number of techniques have been described to facili-

Table 2. Pathologies causing massive splenomegaly

Chronic myeloid leukaemia (most common)
Chronic lymphocytic leukaemia
Myelofibrosis
Polycythaemia rubra vera
Thalassaemia
Visceral leishmaniasis (kala-azar)
Malaria
Schistosomiasis
Gaucher's disease
Niemann–Pick disease

Table 3. Causes of splenomegaly by aetiological subgroup. Common causes in PACES are shown in bold

Haematological (49%)	Lymphoproliferative	**Lymphoma (30%)**
		Chronic lymphocytic leukaemia (15%)
		Acute lymphocytic leukaemia
		Multicentric Castleman disease
	Myeloproliferative	**Chronic myeloid leukaemia (21%)**
		Myelofibrosis (11%)
		Myelodysplastic syndrome (4%)
		Acute myeloid leukaemia
		Systemic mastocytosis
	Erythrocyte disorders	Thalassaemia
		Sickle cell disease*
		Spherocytosis or elliptocytosis
		Extramedullary haematopoeisis
		Polycythaemia rubra vera
Hepatic diseases (23%)	Chronic liver disease	
	Portal vein thrombosis	
	Budd–Chiari syndrome	
Infectious (13%)	Viral	Infectious mononucleosis
		Cytomegalovirus
	Bacterial	Subacute bacterial endocarditis
		Splenic abscess
		Typhoid fever (*Salmonella typhi*)
		Brucellosis
		Peliosis (*Bartonella* spp.)
		Tuberculosis
		Mycobacterium avium intracellulare
	Other	Malaria
		Visceral leishmaniasis (kala-azar)
		Schistosomiasis
		Histoplasmosis
Autoimmune	Felty's syndrome†	
	Systemic lupus erythematosus	
	Autoimmune haemolytic anaemias	
	Sjögren's syndrome	
Infiltrative	Metabolic diseases	Gaucher's disease
		Mucopolysaccharidoses
		Niemann–Pick disease
	Other infiltrative processes	Amyloidosis
		Sarcoidosis
		Splenic metastases‡

*In children. The spleen later auto-infarcts and patients become hyposplenic by adulthood. †In patients with rheumatoid arthritis. ‡Particularly melanoma

tate its detection. When discovered, the remainder of the examination and detailed review of the medical history should be directed towards elucidation of possible aetiologies. These can then be pursued with specific investigations. BJHM

Conflict of interest: none.

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Table 4. Indications for splenectomy

Rupture
Refractory immune thrombocytopaenic purpura
Severe or refractory hypersplenism
Severe discomfort in massive splenomegaly
Tumour debulking (lymphoma, metastases)
Diagnostic (if other modalities exhausted)

KEY POINTS

- Splenomegaly can often be detected on routine palpation of the abdomen.
- Use percussion to clarify equivocal findings on palpation: resonance effectively excludes splenomegaly.
- When palpation and percussion are both positive, splenomegaly can be diagnosed with confidence.
- A normal clinical examination has a false-negative rate of 28% when compared to imaging.
- Complete the examination by searching for clues as to the underlying aetiology and signs of hypersplenism.
- The differential diagnosis varies for massive and for non-massive splenomegaly.