The spleen

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Anatomy

- The weight of the normal adult spleen is 75–250 g.
- It lies in the left hypochondrium between the gastric fundus and the left hemidiaphragm, with its long axis lying along the tenth rib.
- The hilum sits in the angle between the stomach and the kidney and is in contact with the tail of the pancreas.
- The tortuous splenic artery arises from the coeliac axis and runs along the upper border of the body and tail of the pancreas, to which it gives small branches.
- The short gastric and left gastroepiploic branches pass between the layers of the gastrosplenic ligament.

- The splenic vein is formed from several tributaries that drain the hilum.
- *The vein runs behind the pancreas, receiving several small tributaries from the pancreas before joining the superior mesenteric vein at the neck of the pancreas to form the portal vein.*
- The nodes and lymphatics drain via retropancreatic nodes to the coeliac nodes.

FUNCTIONS OF THE SPLEEN

- **Immune function** The spleen processes foreign antigens and is the major site of specific immunoglobulin M (IgM) production.
- **Filter function**. Macrophages in the reticulum capture cellular and non-cellular material from the blood and plasma. This will include the removal of effete platelets and red blood cells.
- **Pitting**. Particulate inclusions from red cells are removed, and the repaired red cells are returned to the circulation. These include Howell–Jolly and Heinz bodies, which represent nuclear remnants and precipitated haemoglobin or globin subunits, respectively.
- **Reservoir function**. This function in humans is less marked than in other species, but the spleen does contain approximately 8 per cent of the red cell mass. An enlarged spleen may contain a much larger proportion of the blood volume.
- **Cytopoiesis**. From the fourth month of intrauterine life, some degree of haemopoiesis occurs in the fetal spleen.

INVESTIGATION OF THE SPLEEN

- Conditions that result in splenomegaly can be diagnosed on the basis of the history and examination findings and from laboratory examination.
- In haemolytic anaemia, a full blood count, reticulocyte count and tests for haemolysis will determine the cause of the anaemia.
- Splenomegaly associated with portal hypertension caused by cirrhosis is diagnosed on the history, physical signs of liver dysfunction, abnormal tests of liver function and endoscopic evidence of oesophageal varices.

Radiological imaging

- Plain radiology is rarely used in investigation, but the incidental finding of calcification of the splenic artery or spleen may raise the possible diagnosis of a splenic artery aneurysm, an old infarct, a benign cyst or hydatid disease. Multiple areas of calcification may suggest splenic tuberculosis.
- Ultrasonography can determine the size and consistency of the spleen, and whether a cyst is present.

- computed tomography (CT) with contrast enhancement is more commonly undertaken to better characterize the nature of the suspected splenic pathology and to exclude other intra-abdominal pathology.
- Magnetic resonance imaging (MRI) may be similarly useful.
- Radioisotope scanning is used occasionally to provide information about the spleen. The use of technetium-99m (99mTc)-labelled colloid is normally restricted *to determining whether the spleen is a significant site of destruction of red blood cells.*

CONGENITAL ABNORMALITIES OF THE SPLEEN

Splenic agenesis is rare, but is present in 10 per cent of children with congenital heart disease. Polysplenia is a rare condition resulting from failure of splenic fusion.

Splenunculi are single or multiple accessory spleens that are found in approximately10 –30 per cent of the population. They are located near the hilum of the spleen in 50 per cent of cases and are related to the splenic vessels or behind the tail of the pancreas in 30 per cent. The remainder are located in the mesocolon or the splenic ligaments. *Their significance lies in the fact that failure to identify and remove these at the time of splenectomy may give rise to persistent disease*.

Hamartomas are rarely found in life and vary in size from 1 cm in diameter to masses large enough to produce an abdominal swelling.

Non-parasitic **splenic cysts** are rare. Splenic cysts are classified as primary cysts (true) or pseudocysts (secondary) on the basis of the presence or absence of lining epithelium.

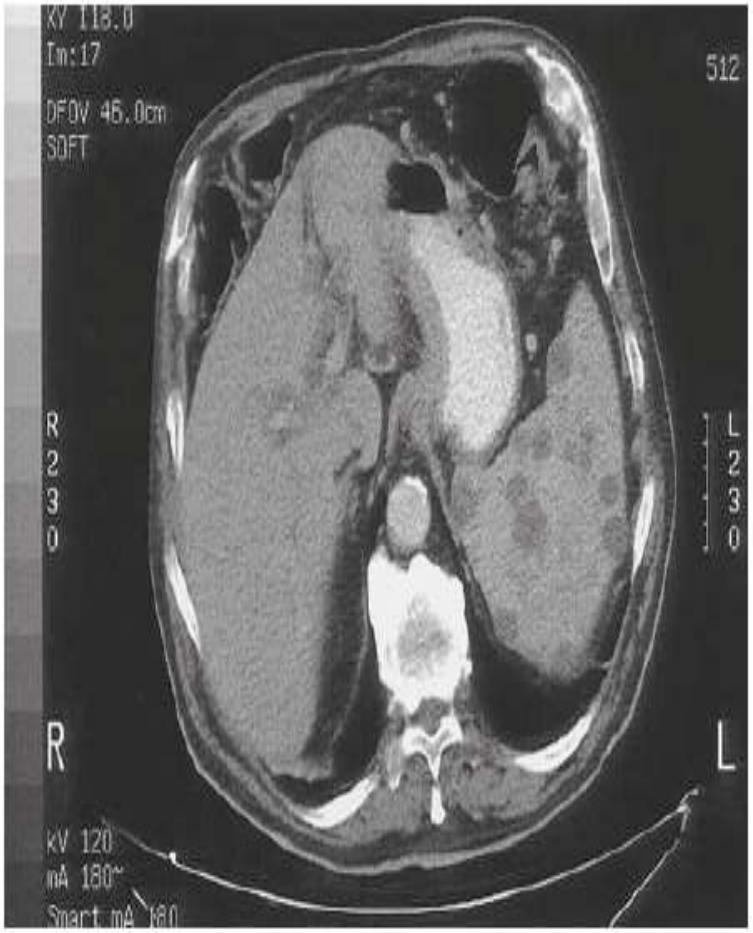

Figure 66.2 Computed tomography scan showing multiple low-density areas in the spleen consistent with multiple benign splenic cysts.

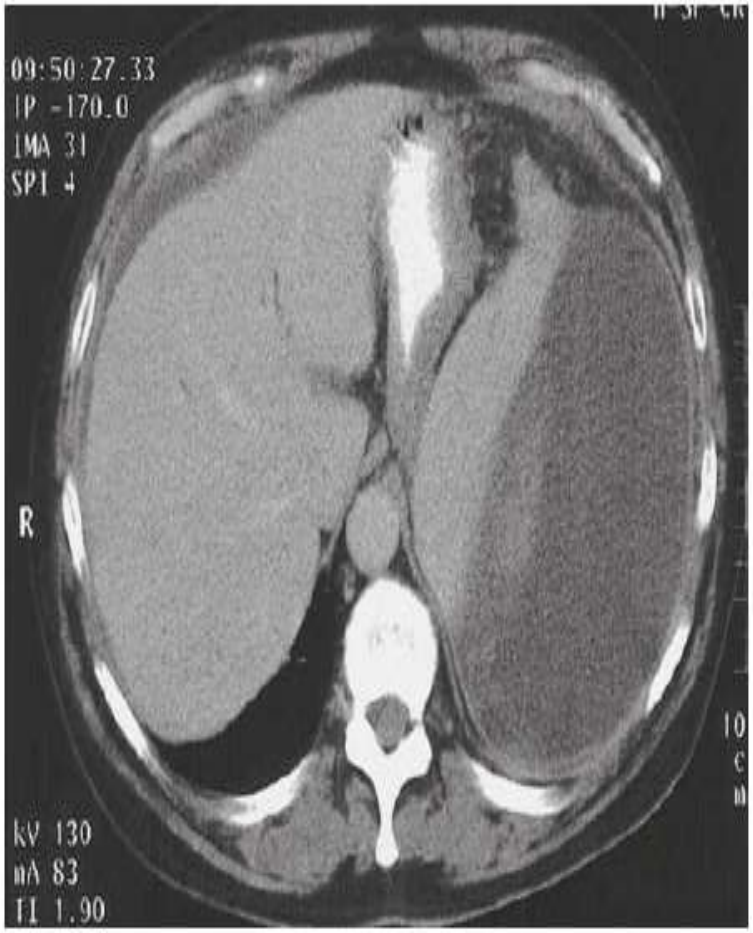

Figure 66.3 Computed tomographic scan showing a large pseudocyst involving the spleen. There is displacement of the stomach medially, and a trace of ascitic fluid is present above the liver.

Hypersplenism

is a clinical syndrome that is characterised by splenic enlargement, any combination of anaemia, leukopenia or thrombocytopenia, compensatory bone marrow hyperplasia and improvement after splenectomy.

Table 66.1 Causes of splenic enlargement.

Infective	Bacterial	Typhoid and paratyphoid Typhus Tuberculosis[a] Septicaemia Splenic abscess[b]
	Spirochaetal	Weil's disease Syphilis
	Viral	Infectious mononucleosis HIV-related thrombocytopenia[b] Psittacosis
	Protozoal and parasitic	Malaria Schistosomiasis[a] Trypanosomiasis Kala-azar Hydatid cyst[c] Tropical splenomegaly[a]
Blood disease	Acute leukaemia Chronic leukaemia Pernicious anaemia Polycythaemia vera Erythroblastosis fetalis	Idiopathic thrombocytopenic purpura[c] Hereditary spherocytosis[a] Autoimmune haemolytic anaemia[a] Thalassaemia[a] Sickle cell disease[a]
Metabolic	Rickets Amyloid Porphyria Gaucher's disease[b]	
Circulatory	Infarct Portal hypertension Segmental portal hypertension[b]	(Pancreatic carcinoma, splenic vein thrombosis)

Collagen disease	Still's disease
	Felty syndrome[a]
Non-parasitic cysts	Congenital
	Acquired
Neoplastic	Angioma
	Primary fibrosarcoma
	Hodgkin's lymphoma[b]
	Other lymphomas
	Myelofibrosis[b]

Tropical splenomegaly

Massive splenic enlargement frequently occurs in the tropics from malaria, kala-azar and schistosomiasis.

It may result from occult infection or be related to malnutrition.

The massive splenomegaly observed in this condition may require removal in those patients disabled by anaemia or local symptoms.

Life-long anti-malarial therapy is indicated in malaria endemic areas.

Schistosomiasis

This condition is prevalent in Africa, Asia and South America.
It is caused by infection with *Schistosoma mansoni* in 75 per cent of cases and by *Schistosoma haematobium* in the remainder. The splenic enlargement may result from portal hypertension associated with hepatic fibrosis. Splenomegaly can occur at any age.

- The diagnosis is based on examination of the urine and faeces for ova, abnormal liver function tests and the presence of hypochromic anaemia.
- Successful medical treatment of established cases does not result in regression of splenomegaly, and removal of the painful and bulky spleen is indicated where there is no evidence of hepatic or renal insufficiency. Splenectomy may be required as part of a devascularisation procedure in patients with portal hypertension associated with schistosomiasis.

Leukaemia

Leukaemia should be considered in the differential diagnosis of splenomegaly and the diagnosis is made by examining a blood or marrow film.

Splenectomy is reserved for hypersplenism that occurs during the chronic phase of chronic granulocytic leukaemia.

Idiopathic thrombocytopenic purpura

In most cases of idiopathic thrombocytopenic purpura (ITP), the low platelet count results from the development of antibodies to specific platelet membrane glycoproteins that damage the patient's own platelets.

Two distinct clinical types are evident:

the acute condition in children and a chronic condition in adults.

Acute ITP often follows an acute infection and has a spontaneous resolution within two months.

Chronic ITP persists longer than six months without a specific cause being identified.

Clinical features

•The adult form

• normally affects females between the ages of 15 and 50 years

• it can be associated with other conditions, including systemic lupus erythematosus, chronic lymphatic leukaemia and Hodgkin's disease.

•The childhood form

• is distributed equally between males and females

• commonly presents before the age of five years.

•Purpuric patches (ecchymoses) occur on the skin and mucous membranes.

• Following trauma or pressure, examination often reveals numbers of petechial haemorrhages in the skin

• There is a tendency to spontaneous bleeding from mucous membranes (e.g. epistaxis); in women, menorrhagia and the prolonged bleeding of minor wounds are common.
• Haemorrhage from the urinary and gastrointestinal tracts and haemarthrosis are rare.
• *Although intracranial haemorrhage is also uncommon, it is the most frequent cause of death*.
• *The diagnosis is made based upon the presence of cutaneous ecchymoses and a positive tourniquet test*.
• The spleen is palpable in fewer than 10 per cent of patients, and the presence of gross splenic enlargement should raise the suspicion of an alternative diagnosis.

Investigations

- Coagulation studies are normal, and a bleeding time is not helpful in diagnosis.
- Platelet count in the peripheral blood film is reduced (usually <60 × 109/L).
- Bone marrow aspiration reveals a plentiful supply of platelet-producing megakaryocytes.

Treatment

•The course of the disease differs in children and adults.

• The disease regresses spontaneously in 75 per cent of pediatric cases following the initial attack.

• Short courses of corticosteroids in both adult and child are usually followed by recovery.

•*Splenectomy is* usually recommended if a patient has

• two relapses on steroid therapy or

• if the platelet count remains low.

•Generally, this is indicated where the ITP has persisted for more than 6–9 months.

•Up to two-thirds of patients will be cured by surgical intervention, and 15 per cent will be improved, but no benefit will be derived in the remainder. ***The response to steroids predicts a good response to splenectomy.***

Haemolytic anaemias

Hereditary spherocytosis

Hereditary spherocytosis is an autosomal dominant hereditary disorder characterised by the presence of spherocytic red cells, caused by various molecular defects in the genes that code for alpha- and beta-spectrin, ankyrin, band 3 protein, protein 4.2 and other erythrocyte membrane proteins. **These proteins are necessary to maintain the normal biconcave shape of the erythrocyte**. **Spherocytosis arises essentially from an increase in permeability of the red cell membranes to sodium**. As this ion leaks into the cell, the osmotic pressure rises, resulting in swelling and increased fragility of the spherocyte. A large number of red cells are destroyed in the spleen, where there is a relative deficiency of both glucose and oxygen.

The clinical presentation is generally in childhood, but may be delayed until later life.

- Mild intermittent jaundice is associated with mild anaemia, splenomegaly and gallstones.
- Examination reveals splenomegaly, and the liver may also be palpable.
- Chronic leg ulcers may arise in adults with the disease.
- Haematological investigations include the fragility test.

Immature red blood cells (reticulocytes), which differ from adult cells by possessing a reticulum, are discharged into the circulation by the bone marrow to compensate for the loss of erythrocytes by haemolysis.

- Faecal urobilinogen is increased as this route excretes most of the urinobilinogen

All patients with hereditary spherocytosis should be treated by splenectomy but, in juvenile cases, this is generally delayed until six years of age to minimise the risk of post splenectomy infection, but before gallstones have had time to form.
Ultrasonography should be performed preoperatively to determine the presence or absence of gallstones.

Acquired autoimmune haemolytic anaemia

This condition is divided into immune and non-immunemediated forms. It may arise following exposure to agents such as chemicals, infection or drugs, e.g. alpha-methyldopa, or be associated with another disease (e.g. systemic lupus erythematosus).

In most instances, the cause is unknown, and red cell survival is reduced because of an immune reaction triggered by immunoglobulin or complement on the red cell surface.

This condition is more common in women after the age of 50 years. In half the patients, the spleen is enlarged and, in 20 per cent of cases, pigment gallstones are present. Anaemia is invariably present and may be associated with spherocytosis because of red cell membrane damage

In the immune type, antibody, which coats the red cells, can be detected by agglutination when anti-human globulin is added to a suspension of the patient's erythrocytes (Coombs' test positive). The disease runs an acute self-limiting course, and no treatment is necessary.

Splenectomy should, however, be considered

- if corticosteroids are ineffective,
- when the patient is developing complications from long-term steroid treatment or
- if corticosteroids are contraindicated. Eighty per cent of patients respond to splenectomy.

Thalassaemia (synonyms: Cooley's anaemia, Mediterranean anaemia)

Thalassaemia (Greek, *Thalassa* meaning sea (because the disease occurs in people of Mediterranean origin)) results from a defect in haemoglobin peptide chain synthesis and is transmitted most commonly as a recessive trait. The disease is really a group of related diseases, alpha, beta and gamma, depending upon which haemoglobin peptide chain's rate of synthesis is reduced. Most patients suffer from beta thalassaemia, in which a reduction in the rate of beta-chain synthesis results in a decrease in haemoglobin A. Intracellular precipitates (Heinz bodies) contribute to premature red cell destruction.

Graduations of the disease range from heterozygous thalassaemia minor to homozygous thalassaemia major, which is associated with chronic anaemia, jaundice and splenomegaly.

Patients with homozygous thalassaemia major frequently develop clinical signs in the first year of life, and these include retarded growth, enlarged head with slanting eyes and depressed nose, leg ulcers, jaundice and abdominal distension secondary to splenomegaly. Red cells are small, thin and misshapen and have a characteristic resistance to osmotic lysis.

In the more severe forms, nucleated red cells and other immature blood cells are seen. The final diagnosis is by haemoglobin electrophoresis. Blood transfusion may be required to correct profound anaemia, but the patient may become transfusion dependent because of the development of hypersplenism.

Splenectomy is therefore of benefit in patients who require frequent blood transfusion and if haemolytic antibodies have developed as a result.

Sickle cell disease

Sickle cell disease is a hereditary, autosomal recessive haemolytic anaemia occurring mainly among those of African origin in whom the normal haemoglobin A is replaced by haemoglobin S (HbS). The HbS molecule crystallises when the blood oxygen tension is reduced, thus distorting and elongating the red cell.

The resulting increased blood viscosity may obstruct the flow of blood in the spleen. Splenic microinfarcts are therefore common. Depending upon the vessels affected by vascular occlusion, patients may complain of bone or joint pain, priapism, neurological abnormalities, skin ulcers or abdominal pain due to visceral blood stasis.

The diagnosis is made by the finding of characteristic sickle-shaped cells on blood film, although this investigation has largely been replaced by haemoglobin electrophoresis.

Hypoxia that provokes a sickling crisis should be avoided and is particularly relevant in patients undergoing general anaesthesia.

Adequate hydration and partial exchange transfusion may help in a crisis. Splenectomy is of benefit in a few patients in whom excessive splenic sequestration of red cells aggravates the anaemia. Chronic hypersplenism usually occurs in late childhood or adolescence, although *Streptococcus pneumoniae* infection may precipitate an acute form in the first five years of life.

Felty's syndrome

Patients with rheumatoid arthritis may develop leukopenia. This is referred to as Felty syndrome if it is extreme and associated with splenomegaly.

- There is no definite relationship between the severity of the arthritic changes and the leukopenia and splenomegaly.
- Splenectomy produces only a transient improvement in the blood picture, but rheumatoid arthritis may respond to steroid therapy to which it had previously become resistant

SPLENECTOMY

The common indications for splenectomy are:

- trauma resulting from an accident or during a surgical procedure, as for example during mobilisation of the oesophagus, stomach, distal pancreas or splenic flexure of the colon;
- removal en bloc with the stomach as part of a radical gastrectomy or with the pancreas as part of a distal or total pancreatectomy;
- to reduce anaemia or thrombocytopenia in spherocytosis, idiopathic thrombocytopenic purpura or hypersplenism;
- in association with shunt or variceal surgery for portal Hypertension

Preoperative preparation

- In the presence of a bleeding tendency, transfusion of blood, fresh-frozen plasma, cryoprecipitate or platelets may be required.
- Coagulation profiles should be as near normal as possible at operation, and platelets should be available for patients with thrombocytopenia at operation and in the early postoperative period.
- Antibiotic prophylaxis appropriate to the operative procedure should be given and consideration should be given to the risk of post-splenectomy sepsis
- *vaccinating against pneumococcus , Meningococcus and H. influenza type B*

Postoperative complications

• Immediate complications specific to splenectomy include haemorrhage resulting from a slipped ligature.
• Haematemesis from gastric mucosal damage and gastric dilatation is uncommon.
• Left basal atelectasis is common, and a pleural effusion may be present.
• Adjacent structures at risk during the procedure include the stomach and pancreas.
• A fistula may result from damage to the greater curvature of the stomach during ligation of the short gastric vessels.
• Damage to the tail of the pancreas may result in pancreatitis, a localised abscess or a pancreatic fistula.
• Postoperative thrombocytosis may arise and, if the blood platelet count exceeds 1 × 106/mL, prophylactic aspirin is recommended to prevent axillary or other venous thrombosis.

•Post-splenectomy septicaemia may result from *Streptococcus pneumoniae*, *Neisseria meningitides*, *Haemophilus influenzae* and *Escherichia coli*. The risk is greater in the young patient, in splenectomised patients treated with chemoradiotherapy and in patients who have undergone splenectomy for thalassaemia, sickle cell disease and autoimmune anaemia or thrombocytopenia.

• Opportunist post-splenectomy infection (OPSI) is a major concern. It is thought that children who have undergone splenectomy before the age of five years should be treated with a daily dose of penicillin until the age of ten years. Prophylaxis in older children should be continued at least until the age of 16 years, but its use is less well defined in adults.

Furthermore, compliance is problematic in the long term but, as the risk of overwhelming sepsis is greatest within the first 2–3 years after splenectomy, it seems reasonable to give prophylaxis during this time.

However, all patients with compromised immune function should receive prophylaxis. Satisfactory oral prophylaxis can be obtained with penicillin, erythromycin or amoxicillin, or co-amoxiclav. Suspected infection can be treated intravenously with these same antibiotics and cefotaxime, ceftriaxone or chloramphenicol in patients allergic to penicillin and cephalosporins. **If elective splenectomy is planned, consideration should be given to vaccinating against pneumococcus, meningococcus C (both repeated every five years) and H. influenza type B (Hib) (repeated every ten years).**

The latter two vaccines are commonly delivered as a combined preparation. Yearly influenza vaccination has been recommended as there is some evidence that it may reduce the risk of secondary bacterial infection. Such vaccinations should be administered at least 2 weeks before elective surgery or as soon as possible after recovery from surgery but before discharge from hospital. Pneumococcal vaccination is recommended in those patients aged over two years.

Haemophilus influenzae type b vaccination is recommended irrespective of age.

• Asplenic patients should carry a medical alert and an up-to-date vaccination card.
• They require specific advice regarding travel and animal handling.
•Patients who have undergone splenectomy and are travelling to countries where malaria is present are strongly advised to use all physical anti-mosquito barriers, as well as anti-malarial therapy, since they are at increased risk of severe malaria.
•Overwhelming post-splectomy sepsis due to *Capnocytophaga canimorsus* may result from dog, cat or other animal bites. In the trauma victim, vaccination can be given in the postoperative period, and the resulting antibody levels will be protective in the majority of cases.