OPEN ACCESS TEXTBOOK OF GENERAL SURGERY

THE SPLEEN

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INTRODUCTION

“a spleen is a strange thing — we technically don ’t need one, but maybe spleens are kept in our bodies in case we mutate or evolve and if we grow wings or tentacles we need to have the spleen in place in order for them to work.. ” Douglas Coupland, The Gum Thief, 2007

The spleen represents the largest single aggregation of lymphatic tissue in the human body and has many important haematological and immunological functions. The asplenic state is however well tolerated with few longterm complications. It thus follows that the spleen can be removed for a variety of conditions, especially haematological conditions, where it is central to the pathophysiology. Splenectomy is a safe procedure with very low rates of peri-operative morbidity and mortality. In recent years, laparoscopic splenectomy has become the standard of care where technically feasible.

ANATOMY

Gross Anatomy

The spleen is a dark red-purple, almost oval organ, situated in the left upper abdominal cavity. The adult spleen measures on average 12cm in its long axis, with a weight of 150 g. It is located deep to the ninth, tenth and eleventh ribs toward the posterior of the left upper quadrant. The long axis of the spleen follows the course of the tenth rib. Two surfaces are evident. The convex and smooth superolateral surface is in contact with the undersurface of the left hemidiaphragm. The inferomedial surface has an irregular concave appearance due to multiple impressions left upon its surface by neighboring viscera. These impressions, arranged around the splenic hilum, are made by the greater gastric curvature, splenic flexure of the colon, left kidney and often the tail of the pancreas.

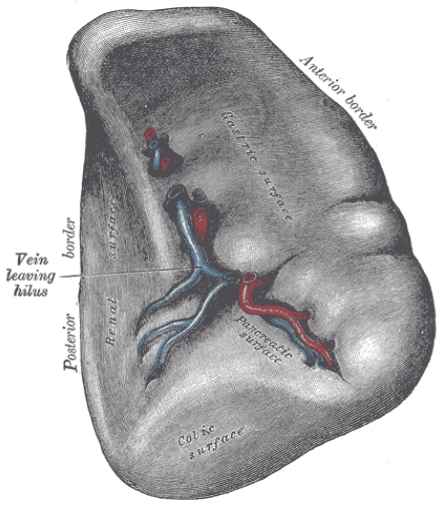

Fig 1 The visceral surface of the spleen.

Peritoneum covers most of the splenic surface, except at the hilum, where the peritoneal folds form three ligaments which anchor the spleen in position:

- The splenorenal ligament contains the splenic artery, splenic vein and tail of the pancreas
- The gastrosplenic ligament contains the short gastric and left gastroepiploic vessels
- The splenocolic ligament which is generally avascular

Arterial blood supply is from the spenic artery, a branch of the celiac axis. The artery courses behind the pancreas to the splenic hilum, via the splenorenal ligament, where it divides into segmental branches. Venous drainage is via the splenic vein to the portal venous system. Unlike lymph nodes, the spleen has no afferent lymphatic supply; efferent lymphatics drain to lymph nodes in the splenic hilum and along the coeliac axis. Innervation, which regulates splenic blood flow, is mainly sympathetic.

Accessory spleens (spleniculi) may be found in the splenic hilum, gastrosplenic and splenoral ligaments as well as in the greater omentum.

The spleen enlarges in an inferior and medial direction. A palpable spleen indicates splenomegaly and is considered an abnormal finding. Distinguishing features of splenomegaly are a mass in the left upper abdomen, of which the inferior border may be in the right lower abdomen, a palpable notch in the inferior border, dull percussion over the mass, inferomedial displacement during inspiration and an enlarged spleen is rarely ballottable.

Microscopic Anatomy

A connective tissue capsule of 1 to 2 mm thickness envelopes the spleen. Thin trabeculae extend from the capsule to the interior of the organ, forming a support structure for the splenic parenchyma. The splenic parenchyma consists of two different tissues:

- Red pulp: Composed of splenic cords which are supplied by sheathed capillaries and drained by venous sinuses. The cords are sponge-like cavities containing macrophages and other white blood cells and act as a blood filtration system.
- White pulp: Aggregates of lymphoid tissue arranged around central arteries. The lymphatic tissue is populate, mainly with B lymphocytes, with some T lymphocytes and plasma cells also present.

The relatively small amount of connective tissue within the spleen leads to a soft and friable organ, easily damaged by blunt trauma and traction on its ligamentous attachments.

PHYSIOLOGY

Haematological Functions

The unique structure of the spleen facilitates its action as a filter, which removes aged and abnormal red cells from the circulation by phagocytosis. To traverse the narrow splenic cords, red cells have to be pliable, an ability lost when red cells age past 120 days or are deformed, e.g. in hereditary spherocytosis. The spleen does not, however, represent the only site of red cell destruction and red cells do not appear to survive longer in circulation after splenectomy. A peripheral blood smear of the asplenic individual does nonetheless display increased numbers of abnormal red cells, containing Howell-Jolly bodies (nuclear remnants) or Papenheimer bodies (siderotic granules). Platelets and leukocytes are also removed from the circulation when abnormal or covered with antibodies.

The spleen functions as a site of extramedullary haematopoiesis in the fetus until the seventh month in utero and, in the adult, in the presence of myeloproliferative conditions. Regarding reservoir function, up to one third of all platelets may be present in the spleen, but red blood cells are not stored to any significant extent.

Immunological Functions

The peculiar structure of the splenic cords within the red pulp, lined by macrophages, provides an excellent framework for interaction between foreign elements which have gained access to the circulation, such as bacteria. After phagocytosis, antigens are presented to lymphocytes, leading to a specific and amplified immune response. The spleen also produces opsonins, such as tuftsin and properdin, which facilitate phagocytosis of bacteria. Within the white pulp, lymphocytes and antibodies (especially lgM) are produced by the resident T lymphocytes, B lymphocytes and plasma cells in response to activation by foreign antigens.

Table 1 Functions of the spleen	
Haematological	**Immunological**
Removal of aged and deformed red blood cells from circulation	Removal of bacteria from circulation
Removal of platelets and leukocytes from circulation if abnormal or coated with antibodies	Opsonin production
Extramedullary haematopoiesis in certain pathological states	Antibody production
Platelet reservoir	Lymphocyte proliferation

Indications for splenectomy

The indications for performing splenectomy may be classified into two broad groups:

- Trauma/disease localized to the spleen or its blood supply
- Haematological conditions which are potentially curable or ameliorated by splenectomy

SPLENIC DISORDERS

Splenic cysts

Cysts of the spleen, depending on the cause, are usually single and asymptomatic. These are found incidentally during imaging investigations. Larger cysts may present with splenomegaly or with left hypochondrial abdominal pain. Cysts may be classified as congenital, hydatid or pseudocysts.

- Congenital Cysts have an epithelial lining and may be either simple, epidermoid or dermoid cysts. Small and asymptomatic cysts are observed while large and/or symptomatic cysts are treated by partial or total splenectomy.

- Hydatid cysts occur as a result of Echinococcus Granulosis infestation and become symptomatic when large enough to cause local pressure effects or when complicated (rupture; secondary infection). The diagnosis is confirmed with serological testing after suggestive imaging.

- Pseudocysts have no epithelial lining and form after the resolution of a haematoma caused by splenic trauma. Other causes are splenic infarction with liquefactive necrosis, Tuberculosis or Syphilis.

Splenic Abscesses

Splenic abscesses occur as a result of haematogenous bacterial seeding, penetrating trauma or extension of sepsis from an adjacent structure. These are suspected in the setting of fever and left hypochondrium abdominal pain, tenderness or peritonism. Risk factors are sickle cell disease, splenic trauma, intravenous drug abuse and immunodeficiency. The diagnosis, when suspected, is confirmed with imaging (ultrasound or CT). Treatment consists of appropriate antibiotic therapy, drainage of the abscess percutaneously or surgically e.g. laparoscopic drainage, open drainage or by splenectomy. If haematogenous seeding is suspected, the source should be located and treated.

Splenic Artery Aneurysms

Aneurysms of the splenic artery are the most frequent visceral artery aneurysm and are twice as common in females as in males. Occurrence is mainly in three groups of patients: 1) elderly patients with atherosclerosis 2) young females as a congenital lesion and 3) as a complication of acute pancreatitis or pancreatic pseudocysts (Which are false aneurysms).

Splenic artery aneurysms are usually single lesions and asymptomatic. They may cause left upper abdominal or left flank pain but are rarely palpable. Rupture is characterized by sudden

onset of abdominal pain with circulatory collapse and may be free into the peritoneal cavity or into an adjacent hollow viscus. Asymptomatic aneurysms may be diagnosed incidentally on an abdominal XRay by noting a calcified lesion in the form of an “eggshell” in the left upper quadrant.

Observation is recommended for asymptomatic aneurysms less than 2 cm size. Surgery is indicated for aneurysms which are symptomatic and/or larger than 2 cm. Pregnancy is a serious risk factor for rupture and surgery is recommended for all young females who aim to become pregnant or are in their first two trimesters of pregnancy.

For aneurysms close to the splenic hilum, splenectomy is performed, while more proximal lesions may be treated by excision followed by splenic artery reconstruction or ligation.

SPLENIC TUMOURS

- Benign Tumours Haemangiomas and lymphangiomas occur rarely, are mostly asymptomatic and may be complicated by rupture or thrombocytopenia (Kasabach-Merritt Syndrome).

- Malignant Tumours

Table 2 Malignant tumours of the spleen	
Primary	**Secondary / Metastatic**
Non-Hodgkin lymphoma	Lung, breast, stomach, pancreas, colon, melanoma
Nonlymphoid malignancies e.g. langiosarcoma, plasmacytoma	

Splenic malignancy may present with epigastric of left hypochondrium abdominal discomfort, splenomegaly and constitutional symptoms in the setting of lymphoma. Complications are rupture and hypersplenism.

While non-lymphoid malignancies are rare, the spleen is involved in one third of patients with Non-Hodgkin lymphoma, which is the most common primary splenic malignancy. Splenectomy is not indicated in Non-Hodgkin lymphoma, unless diagnostic uncertainty exists or the spleen is affected in isolation (<1% of cases). Staging laparotomy for Non-Hodgkin lymphoma (splenectomy, lymph node sampling, hepatic wedge biopsy) is now rarely performed due to the accuracy of modern imaging modalities such as CT and positron emission tomography CT (PET-CT).

Left-sided Portal Hypertension

Splenic vein thrombosis due to acute or chronic pancreatitis may lead to a segmental form of portal hypertension along the greater curvature of the stomach. Gastric varices and splenomegaly are prominent, while oesophageal varices are uncommon. Endoscopic management of gastric varices is technically difficult with a low success rate. Splenectomy and devascularization of the greater curvature of the stomach is the treatment of choice.

SPLENIC TRAUMA

Rupture of the spleen can occur in four scenarios:

- Blunt abdominal trauma is the leading cause of splenic rupture. The spleen is the most frequently injured abdominal organ in patients with blunt abdominal trauma
- Penetrating abdominal trauma to the left lower chest, left flank and left hypochondrium.
- Iatrogenic trauma due to traction on its ligamentous attachments during surgical procedures in the

vicinity of the spleen e.g. left hemicolectomy
- "Spontaneous" rupture of a pathological spleen

The clinical presentation of splenic rupture secondary to trauma varies according to injury severity and associated injuries sustained. Clues to spenic injury are abdominal pain and tenderness worst in the left hypochondrium, left shouldertip pain, left rib fractures or an unexplained low haematocrit.

In the context of blunt abdominal trauma, splenic rupture presents in either haemodynamically stable or unstable patients. Stable patients, i.e. those that are normotensive without tachycardia or rapidly responsive to fluid resuscitation, are investigated with CT for suspected intra-abdominal injury. CT is accurate for the detection of splenic injury and the quantity of haemoperitoneum. CT is also used to classify the severity of injury according to the American Association for the Surgery of Trauma Spleen Injury Scale and reliably and consistently identifies certain other injuries such as renal injuries and vertebral fractures.

Spleen injury scale (1994 Revision) of the American Association for the Surgery of Trauma		
Grade	**Type of Injury**	**Injury Description**
I	Haematoma	Subcapsular, <10% splenic surface area
	Laceration	Capsular tear <1cm parenchymal depth
II	Haematoma	Subcapsular, 10-50% splenic surface area, intraparenchymal <5cm diameter
	Laceration	Capsular tear 1-3cm parenchymal depth not involving trabecular vessel
III	Haematoma	Subcapsular, > 50% splenic surface area, intraparenchymal > 5cm diameter
	Laceration	>3cm parenchymal depth or involving trabecular vessel
IV	Laceration	I Laceration involving segmental or hilar vessels with devascularization
V	Laceration	Entirely shattered spleen
	Vascular	Hilar vascular injury with splenic devascularization

Table 3 Spleen injury scale Adapted from Moore EE et al, J Trauma 1995;38:323 324

Historically, splenectomy was performed for all splenic injuries. Nonoperative management, first reported in 1971, has subsequently been shown to be a feasible alternative strategy. Evidence that stable patients with splenic injury can be managed nonoperatively, together with concerns regarding postsplenectomy infection, have established nonoperative management of stable patients as the current standard of care. These patients must, however, be evaluated critically by an experienced clinician and the following criteria adhered to before committing to a selective nonoperative strategy:

- no clinical or radiological indication for laparotomy
- no ongoing blood transfusion requirement attributable to splenic injury
- patients should be monitored in a high care environment
- constant availability of a surgeon in case of sudden deterioration as a result of bleeding

Bed rest, regular abdominal examination and serial haematocrit estimation is recommended during a

96 hour observation period. Failure of non-operative management, requiring splenectomy, is indicated by the following:

- development of peritoneal irritation suggestive of hollow visceral injury
- development of haemodynamic instability
- development of increasing intravenous fluid requirement to maintain haemodynamic stability
- drop in haematrocrit requiring transfusion of more than two units of packed red cells

Up to 60% of stable patients with splenic injury may be managed successfully. Failure rates increase for patients with more severe splenic injury (Grade IV and V injuries), large haemoperitoneum and age > 55 years, but these predictors do not serve as absolute contraindications to nonoperative management. Successfully managed patients are advised to avoid contact sport for 3 to 6 months.

Haemodynamically unstable patients with suspected blunt abdominal trauma are investigated with FAST (Focused Assessment by Sonography for Trauma) or DPL (Diagnostic Peritoneal Lavage). If these investigations indicate haemoperitoneum, emergency laparotomy is performed. At laparotomy the ruptured spleen declares itself as the culprit and is removed. Attempts at splenic salvage, such as partial splenectomy and splenorrhaphy, may be made within reason.

HAEMATOLOGICAL CONDITIONS TREATED OR AMELIORATED BY SPLENECTOMY

The second group of conditions where splenectomy may be indicated is represented by haematological disorders and includes haemolytic anaemias, purpuras, hypersplenism and myeloproliferative disorders.

Haemolytic Anaemias

Haemolytic anaemia is characterized by anaemia, jaundice, splenomegaly and reticulocytosis. These anaemias are caused by hereditary or acquired disorders. Depending on the specific disorder, jaundice, splenomegaly and symptoms occur to a varying degree. Mild forms may display compensated anaemia with high reticulocyte counts, while more severe forms develop symptomatic splenomegaly, hypersplenism and may require blood transfusion. Pigment gallstones frequently complicate haemolytic anaemia as a result of increased bilirubin load presented to the liver. Splenectomy, as a therapeutic measure, does not directly cure the underlying haematological disorder, but normalises red cell survival by removing the site of abnormal or antibody-coated red cell destruction. Splenectomy is indicated under the following circumstances:

- symptomatic splenomegaly
- symptomatic anaemia, persistent transfusion requirement
- hypersplenism and symptomatic cholelithiasis.

At splenectomy, the gallbladder is removed if cholelithiasis is present. Due to the risk of overwhelming postsplenectomy infection, splenectomy should be delayed in children until the age of four years. Hereditary causes frequently requiring splenectomy are hereditary spherocytosis, hereditary elliptocytosis and Thalassaemia major. Autoimmune haemolytic anaemia is the only acquired haemolytic disorder occasionally treated by splenectomy.

Purpuras

Idiopathic thrombocytopaenic purpura (ITP) constitutes the most common haematological splenectomy indication. The spleen produces lgG antiplatelet factors which destroy platelets and also acts as the major

site of platelet destruction. It is characterized by abnormal bleeding, a low platelet count and a normal bone marrow biopsy. ln children less than sixteen years, ITP is usually transient and rarely requires surgery, whereas in adult patients it is rarely (<15%) cured by medical therapy. Options for medical therapy are corticosteroids, plasmapheresis, intravenous immunoglobulins and eltrombopag. In practice, splenectomy is indicated when patients fail to respond to 6 weeks of steroid therapy, can not tolerate steroids or if the platelet count falls after tapering of steroid therapy. Splenectomy induces remission in 85% of patients, with platelet counts usually > 10 000/mm3 at day 7 postoperatively. ln ITP, the spleen is characteristically of normal size and ideally suited to laparoscopic removal.

Immune-mediated thrombocytopaenia may be associated with HIV infection, Systemic Lupus Erythematosus and Chronic Lymphocytic Leukemia. Indications for splenectomy are the same as for ITP.

Hypersplenism

Hypersplenism is characterized by splenomegaly, pancytopaenia and a normal bone marrow biopsy. Secondary hypersplenism may complicate a variety of disorders such as rheumatoid arthritis (Felty's Syndrome), Sarcoidosis, storage diseases, haematological malignancies, Malaria and portal hypertension (Banti's Syndrome). Primary hypersplenism without an apparent cause after careful investigation is rare. Patients may either be symptomatic due to the splenomegaly itself with abdominal discomfort or due to complications of pancytopaenia. The decision to perform splenectomy in these patients is individualized, with the patient's haematologist and surgeon taking into account alternative treatments, degree of symptoms, degree of pancytopaenia, complications of pancytopaenia and the procedure-related risk.

Secondary causes of hypersplenism	
Inflammatory conditions	e.g. Rheumatoid Arthritis, SLE, Sarcoidosis
Chronic infective conditions	e.g. Malaria, TB, Syphilis
Acute infective conditions	e.g. Infectious mononucleosis, SBE
Congestive Splenomegaly	Portal Hypertension
Storage Diseases	e.g. Gaucher's disease, amyloidosis
Chronic Haemolytic Anaemias	e.g. Hereditary spherocytosis, Thalassaemia major
Malignancy	Lymphoma, leukemia, metastatic carcinoma, myeloproliferative disorders

Myeloproliferative disorders

These disorders are characterized by the proliferation of precursors of myeloid elements which retain their ability to differentiate e.g. into red blood cells (Polycythaemia rubra vera) and platelets (Essential thrombocytosis). One of these disorders, myelofibrosis, is complicated by fibroblast and connective tissue proliferation in the bone marrow, liver and spleen. Extensive bone marrow fibrosis leads to bone marrow failure and extramedullary haematopoiesis in the liver and spleen. Massive splenomegaly is typical. Although splenectomy does not cure the disease, it is indicated to relieve the symptoms of splenomegaly and decrease transfusion requirements.

Table 5 Summary of potential splenectomy indications
Trauma / rupture
Splenic cysts
Splenic abscesses
Splenic artery aneurysms
Benign or malignant tumours limited to the spleen
Left sided portal hypertension
Certain haemolytic anaemias
Idiopathic thrombocytopaenic purpura
Hypersplenism / symptomatic splenomegaly
As part of another surgical procedure e.g. distal pancreatectomy or en-bloc tumour excision
Staging of Non-Hodgkin lymphoma (historic)

Splenectomy

Preoperative preparation

For elective splenectomy, the preoperative phase is influenced by the indication for splenectomy. In general, attention should be paid to preoperative haematocrit, coagulation studies and specimens sent for blood typing and cross matching. Patients must be vaccinated against Streptococcus pneumoniae, Haemophilus influenzae and Neisseria meningitidis at least 14 days prior to the procedure. After emergency splenectomy, vaccination is administered before hospital discharge. Venous thromboembolism prophylaxis is also provided routinely.

When ITP is the indication for surgery, platelets are ordered preoperatively and only transfused after division of the splenic artery. Perioperative cover with steroids is provided for patients currently or recently on corticosteroids where the risk of an Addisonian crisis exists.

Surgical technique

The spleen can be removed by open or laparoscopic approaches. Laparoscopic splenectomy is considered in most patients requiring splenectomy. In comparison to open splenectomy, less postoperative pain, shorter hospital stay, improved cosmesis and earlier return to full activity make laparoscopic splenectomy the procedure of choice. Open splenectomy may be favoured in the presence of a massive spleen, portal hypertension or extensive adhesions due to previous abdominal surgery.

A detailed description of the operative technique is beyond the scope of this text, but essentially the following manoeuvres are performed at splenectomy, whether by open or laparoscopic routes:

- division of the splenocolic an phrenocolic ligaments
- division of the attachments between spleen and diaphragm
- division of the gastrosplenic ligament with careful ligation of the short gastric vessels without injury to the greater curvature of the stomach
- separate identification and ligation of the splenic artery and vein after dissection of the splenorenal ligament, taking care not to injure the tail of the pancreas
- removal of the spleen from the abdominal cavity, which at laparoscopic splenectomy is performed by morselisation within a sterile bag and piecemeal extraction without spillage into the abdominal cavity
- careful search to identify and remove accessory splenic tissue

When removing a massively enlarged spleen, the splenic artery is ligated before mobilization begins to minimize intraoperative bleeding.

Postoperative complications

Early postoperative complications are haemorrhage, gastric perforation, injury to the tail of the pancreas with resultant pancreatic pseudocyst/fistula,

subphrenic abscess and atelectasis of the lower lobe of the left lung. Portal vein thrombosis is a rare but well recognized complication, especially after splenectomy for myeloproliferative disorders.

The platelet count is expected to rise after splenectomy and levels of 500 000/mm3 are often seen. Levels in excess of 1 000 000/mm3 are occasionally seen. This is thought to impart a small, yet unsubstantiated risk of thrombotic events and routine anticoagulation is not currently recommended.

The most feared long-term complication of splenectomy is overwhelming postsplenectomy infection (OPSI). This is a result of loss of the immunological function ofthe spleen as previously described, rendering the asplenic individual especially susceptible to infection with encapsulated bacteria e.g. Streptococcus pneumoniae, Haemophilus influenzae and Neisseria meningitidis. The early clinical course is characterized by mild non-specific influenza-like symptoms, followed by sepsis which may rapidly lead to septic shock and death from multi organ failure.

The risk of OPSI is greatest in young children, especially those younger than 2 years. The incidence is highest in the first 2 years after splenectomy, but patients remain at risk lifelong, with an annual incidence of approximately 0.5%. Although the cumulative lifelong risk of OPSI is fortunately less than 5%, mortality of the condition is around 50%.

The following are preventive strategies to decrease the incidence of OPSI:

- Patient education: Asplenic individuals must be made aware of the increased risk of serious infection. They should present as soon as possible to their nearest medical service with any acute febrile illness.
- Immunization: Splenectomized patients must be vaccinated against the relevant organisms, Streptococcus pneumoniae, Haemophilus influenzae and Neisseria meningitidis. Patients are revaccinated every 5 years.
- Antibiotic prophylaxis: Although evidence regarding antibiotic prophylaxis for children in the age of vaccination is lacking, most authorities recommend a daily dose of penicillin or amoxycillin in the first 2 years after splenectomy. For adults, standby antibiotics are recommended, to be taken at the first sign of infection.