Chapter 43.1
Surgery of the spleen

P. Jane Clarke

Peter J. Morris

Introduction

The spleen was a source of intrigue to ancient physicians and philosophers alike. Galen described the spleen as an organ of mystery, and believed that it extracted 'melancholy' from the circulating blood and from the liver, and returned it to the stomach after a process of purification. Although it was noted by Aristotle that the asplenic state is compatible with life, the complexity of its function and anatomy are still a major topic of medical and scientific study. Thus the care of patients with splenic disorders often requires a multidisciplinary team of haematologists, pathologists, and surgeons.

Embryology

At 5 weeks of gestation, the part of the gut tube destined to become the stomach appears as a fusiform swelling in the foregut. It is attached to the dorsal body wall by a peritoneal fold, the dorsal mesogastrium. During the following weeks the stomach rotates in both its longitudinal and anteroposterior axes. As a result, the pyloric end moves to the right and the cardiac to the left; the left side faces anteriorly and the right posteriorly. These rotations result in the dorsal mesogastrium being pulled out and pouched to the left of the median plane. The pouch developed in this way lies behind the stomach and forms the omental bursa. At the same time, the fetal splenic tissue develops from condensations of mesoderm in the dorsal mesogastrium. This condensation has the effect of dividing the mesogastrium into two parts, that between the stomach and the fetal splenic tissue forming the gastrosplenic ligament and that between it and the kidney becoming the lienorenal ligament. The mesenchymal condensations then fuse to form the spleen.

Congenital abnormalities

Simple splenic agenesis is rare as an isolated abnormality but is found in 4.0 per cent of children with congenital cardiac disease. Because the spleen develops from independent collections of mesoderm that then fuse, both accessory spleens (splenunculi) and polysplenia can occur. Splenunculi are found in approximately 10 to 30 per cent of the population, and are therefore one of the most frequent congenital anomalies. They usually occur in the gastrosplenic ligament and the greater omentum. The spleen may retain its fetal, lobulated form or show deep notches on its diaphragmatic surface. In the rare condition of polysplenia, two to nine distinct parts are found, owing to failure of splenic

fusion.

Anatomy

Relations

The spleen is a lymphatic organ situated in the left hypochondrium between the gastric fundus and the left hemidiaphragm. Its long axis is in the line of the tenth rib; the hilum is in the angle between the stomach and the left kidney and makes contact with the tail of the pancreas. It is invested by visceral peritoneum. The diaphragmatic surface is moulded into a reciprocal convexity and the visceral surface has impressions from the stomach, left kidney, pancreas, and splenic flexure. Its size and weight vary depending on age and in different conditions, but a normal adult spleen weighs approximately 150 g (range 80–300 g), and measures 12 × 7 × 3 cm.

Surface anatomy

The long axis of the spleen lies along the tenth rib. The lower pole does not normally project beyond the midaxillary line. When the spleen enlarges the long axis extends down along the tenth rib and the anterior border approaches the costal margin. The spleen must at least treble in size before becoming palpable, which it does by passing in front of the splenic flexure of the colon. During clinical examination the spleen is palpated as a mass in the left upper quadrant that is ballotable, and is identified by the splenic notch and the lack of resonance to percussion over the mass (Fig. 1; see also Fig. 4).
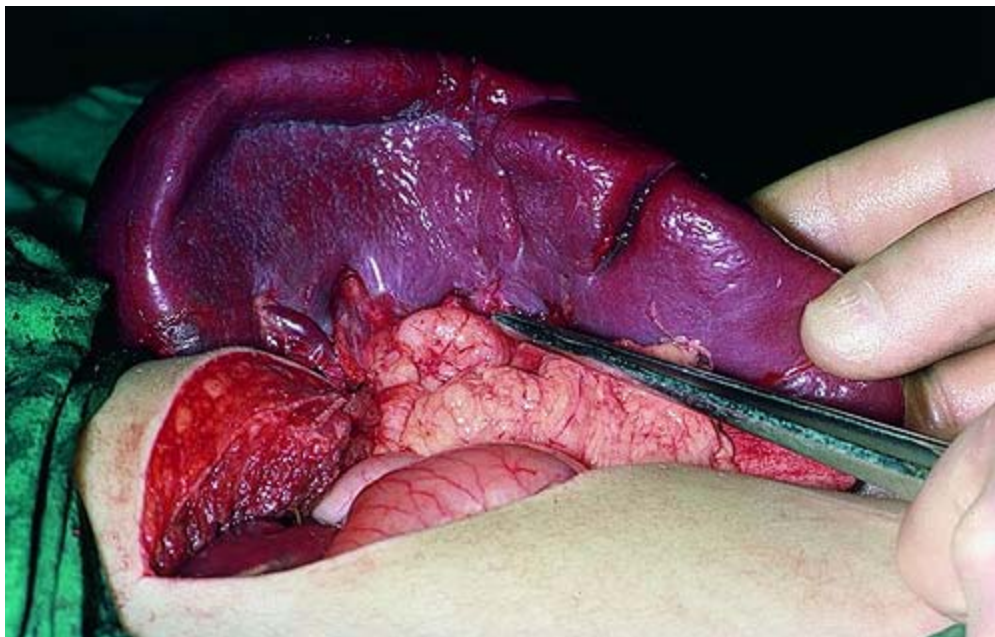

Fig. 1. An enlarged spleen delivered through a subcostal incision in a patient with Hodgkin's disease, showing the splenic notch.
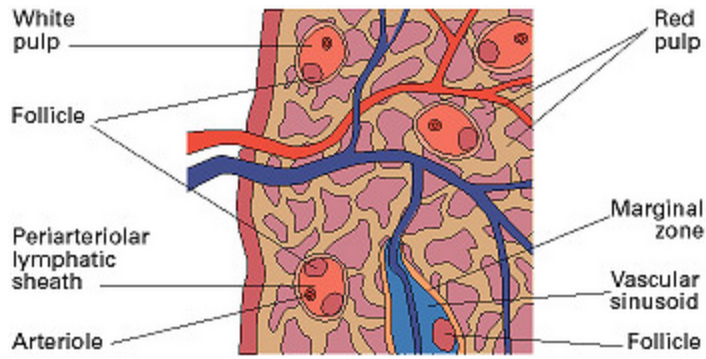
Fig. 2. A simple diagram of the anatomy of the spleen showing the white pulp, which comprises the arterioles, the periarteriolar lymphatic sheath, and the lymphoid follicles. The white pulp is embedded in the much larger mass of red pulp made up of the erythrocyte-filled sinusoids.
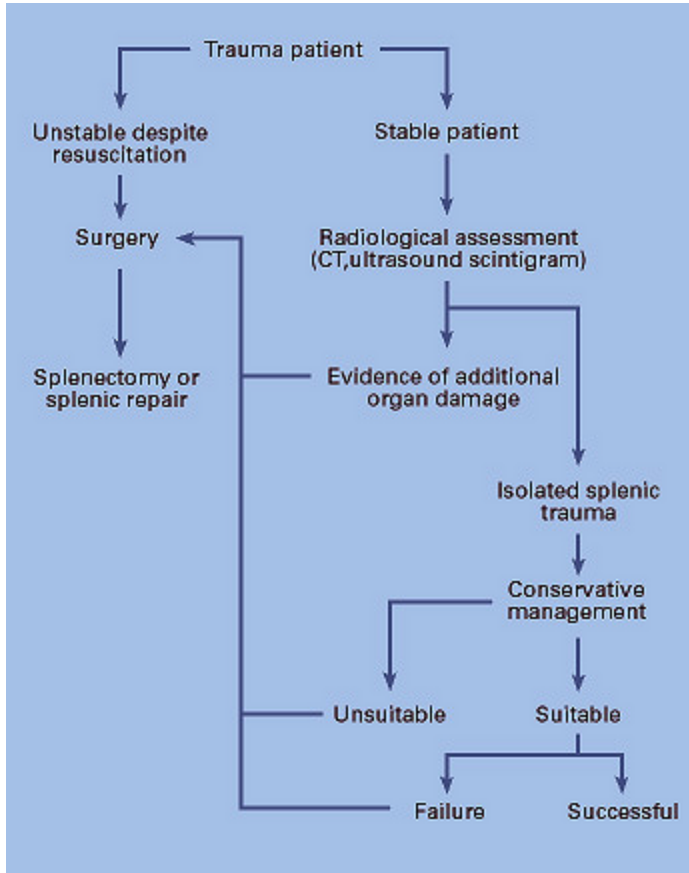
Fig. 3. An algorithm for the management of a patient with splenic trauma.
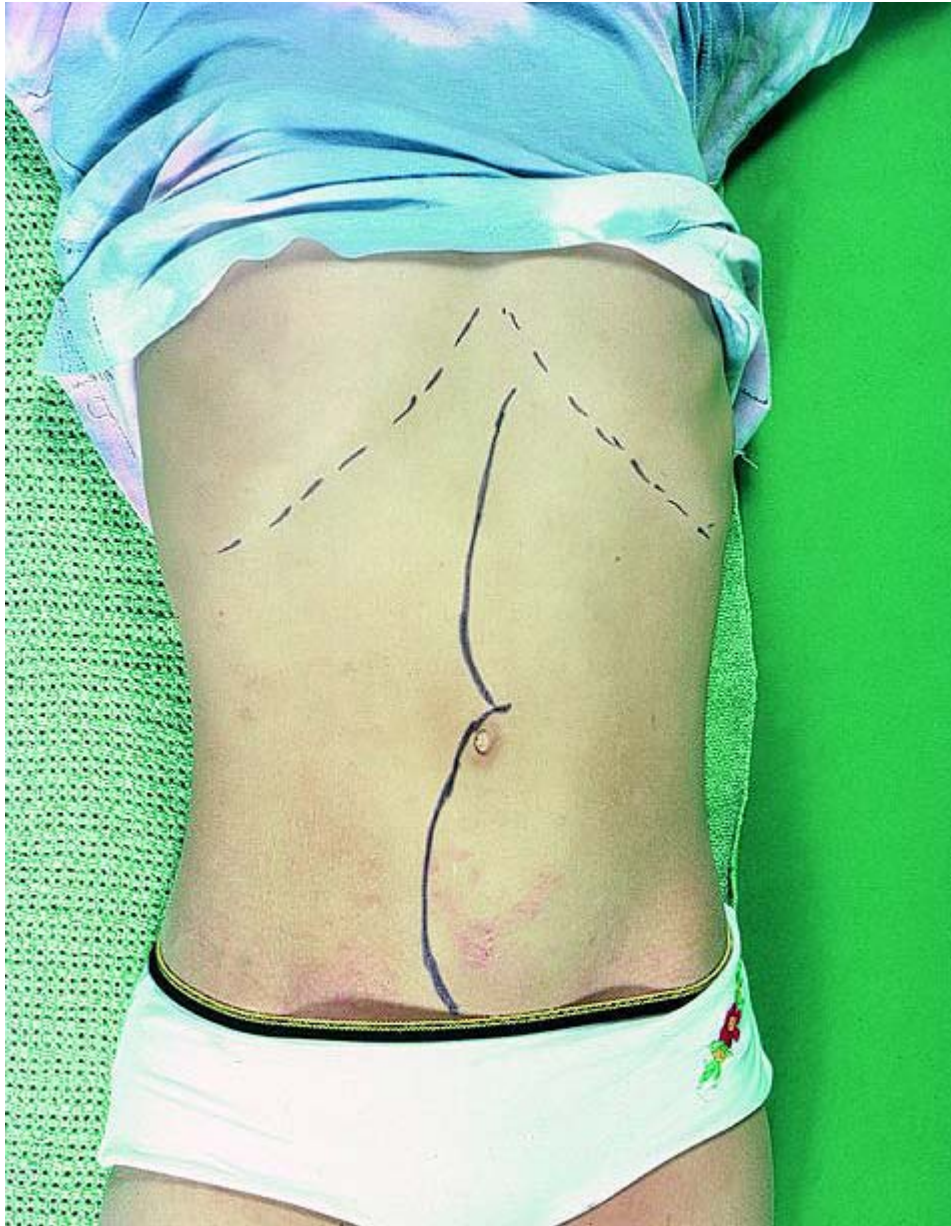
Fig. 4. An adolescent girl with β-thalassaemia who was transfusion dependent; the border of an enormous spleen is outlined, showing also the splenic notch.

Splenic vasculature

The arterial supply to the spleen passes through the splenic artery, a tortuous branch of the coeliac axis (usually from a common stem with a hepatic artery), running along the superior surface of the body and tail of the pancreas. The short gastric and left gastroepiploic branches of the splenic artery pass between the layers of the gastrosplenic ligament. The splenic artery divides at the splenic hilum into superior and inferior

branches, which then further divide into four or five segmental branches, each serving one segment of spleen. Radiopaque dye injection and corrosion casting techniques demonstrate each segment to be distinct, with a separate arterial supply and venous drainage. Such studies have clarified the existence of pyramidal

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segments in the polar regions and wedge-shaped segments in the central part of the spleen. Further subsegmentation has also been demonstrated, being similar in appearance and blood supply to accessory spleens or splenunculi. Vascular anastomoses between segments are uncommon. This knowledge of the arrangement of the segmental vasculature is important when considering splenic salvage surgery (see below).

The splenic vein forms from five or more tributaries leaving the hilum. It then runs behind the pancreas to join the superior mesenteric vein, thus forming the portal vein. Its tributaries correspond to the arterial branches. The lymphatic drainage of the spleen comprises extensive efferent vessels in the white pulp that run with the arterioles and emerge from nodes at the hilum. These nodes drain through the retropancreatic nodes to those at the coeliac axis. There are no afferent vessels. Sympathetic nerve fibres run from the coeliac plexus and mainly innervate branches of the splenic artery.

Microscopic anatomy

The spleen is surrounded by serosa and a collagenous capsule from which trabeculae penetrate the parenchyma. The trabeculae are dense connective-tissue fibres, rich in collagen and elastic tissue. In many mammals they contain non-striated myocytes but in man these are few in number. Between the trabeculae there is a network of reticular fibres supporting the splenic parenchyma, which consists of the red and white pulp separated by the marginal zone.

On macroscopic examination of the cut surface, the white pulp appears as discrete, grey–white nodules embedded in a red matrix, the red pulp. The white pulp consists of periarteriolar lymphatic sheaths and lymphoid follicles, whereas the red pulp (which comprises approximately 75 per cent of the splenic volume) consists of the venous sinusoids and the splenic cords (of Billroth).

The microscopic anatomy of the red and white pulp is based on a complex vascular arrangement (Fig. 2). Blood flow from the splenic artery enters the trabecular arteries from which central arteries supply the white pulp. The adventitia of the central arteries is then replaced by a periarteriolar lymphoid sheath (predominantly of T cells), and at various points these enlarge to form the splenic follicles. When stimulated by antigen the follicles form germinal centres, composed principally of B lymphocytes, that are similar in structure to the lymphoid follicles in lymph nodes. The central arterioles lie to one side of the germinal centres; they then lose their periarteriolar lymphoid sheath and continue through the transitional zone into the red pulp as several, very straight branches or penicillary arterioles (penicilli). The red pulp is composed of numerous, thin-walled venous sinusoids separated by the splenic cords, sponge-like structures consisting mainly of macrophages that are loosely connected via dendritic processes to form a filter through which blood can seep slowly. The blood entering the red pulp from the penicillary arterioles can take two routes to reach the splenic veins. The 'open' route is via the splenic cords, with blood

slowly filtering out to the sinusoids; the 'closed' route involves direct drainage from the penicillary arterioles to the veins without passing via the cords. It is thought that the majority of the circulation in health is via the closed, more rapid, circulatory pathway. The pulp venules collect the blood from both the sinuses and the cords and carry it to the trabecular veins, and thence to the hilum and the splenic veins.

Splenic function and physiology

Phagocytosis

A major function of the spleen is phagocytosis. Effete and damaged red cells are removed daily from the circulation, as well as particulate foreign matter, microbes, antigens, and cellular debris. This process occurs in the sinusoids and the splenic cords by the action of the endothelial macrophages. In addition, intact cells are held up and siderotic granules, as well as Howell–Jolly and Heinz bodies (nuclear remnants and precipitated haemoglobin or globin subunits, respectively), are removed before the red cells are returned to the circulation.

Immune response

The spleen comprises the largest single accumulation of lymphoid cells in the body, containing 25 per cent of the total T-lymphocyte population and 10 to 15 per cent of the B-lymphocyte population. Blood-borne cells and antigens are trapped in the periarteriolar lymphatic sheaths and presented to immunocompetent cells, leading

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to antibody production by plasma cells and an increase in size of the germinal centres in the lymphoid follicles. The spleen is particularly involved in the clearance of particulate antigens, immune complexes, and the antibody-coated cells generated in autoimmune responses. After splenectomy, patients have a decreased ability to generate an IgM response, a decreased capacity to respond to polysaccharide antigens, there is a decrease in the activity of their alternative complement pathway, and their production of tuftsin is defective. It is possible that these deficiencies in immune function explain the problem of overwhelming postsplenectomy infection that sometimes complicates the asplenic state (see below).

Erythrocyte storage

This function is less marked in man than other species but the spleen does contain a large volume of blood (approximately 8 per cent of the red cell mass) either in the venous sinuses or in the reticular meshes of the cords. During emergencies such as anoxia, large volumes of blood can be discharged into the circulation. Enlarged spleens may contain a much larger proportion of the blood volume (up to 40 per cent), including platelets and white cells.

Cytopoiesis

The red pulp contains groups of myelocytes, erythroblasts, and megakaryocytes. From the

fourth month of intrauterine life some degree of haemopoiesis occurs in the human fetus within the spleen, although this is a minor site compared with the liver. Although a large bulk of lymphoid tissue, the mature spleen is not a major site of lymphopoiesis. However, stimulation in the white pulp by antigenic challenge does result in the proliferation of germinal centres, which contributes to the circulating pool of competent T and B cells and macrophages. This may also occur in disease states, especially in myeloproliferative disorders, thalassaemias, and chronic haemolytic anaemias.

Indications for splenectomy (Table 1)

Splenic trauma

Diagnosis

Damage to the spleen may result from accidental or iatrogenic trauma, or may present as 'delayed' or 'spontaneous' rupture. It has been estimated that more than 20 per cent of all splenectomies are for damage to the spleen occurring during the course of another abdominal operation. The most common type of non-iatrogenic injury to result in splenic trauma is non-penetrating, blunt trauma to the upper abdomen, often associated with fractures of the left lower ribs. Delayed rupture is said to occur when the clinical signs develop after a delay of at least 48 h from the initial injury. It is generally believed to result from tearing of the capsule by the expansion of a subcapsular haematoma. The clinical management and presentation are the same as for immediate rupture. 'Spontaneous' rupture of a diseased spleen usually results from trivial trauma, often forgotten by the patient. It occurs most commonly as a complication of malaria, but is the most common cause of death in infectious mononucleosis.

Indication	Pathology
Bleeding	Trauma
	Iatrogenic
	Spontaneous
As part of a surgical resection	During total gastrectomy, distal pancreatectomy
Diagnostic	Splenomegaly of unknown cause
	Pyrexia of unknown origin
Staging	Hodgkin's disease
Therapeutic	Hypersplenism
	Idiopathic thrombocytopenic purpura
	Haemolytic anaemia
	Giant spleen with symptoms
Other	Splenic abscess
	Hydatid disease

Table 1 Indications for splenectomy

If the trauma is localized to the spleen, in the absence of other intraperitoneal damage, physical signs depend on the degree of blood loss; they vary from minimal tenderness in the left upper quadrant in an otherwise well patient to signs of peritonitis accompanied by evidence of shock. Peritoneal lavage can aid the diagnosis of intra-abdominal bleeding, especially in cases that are otherwise difficult to assess, such as an obtunded patient and those with multiple potential sites of blood loss. After clinical assessment of the degree of blood loss and appropriate resuscitation with colloid or blood, a management plan as suggested by the algorithm in Fig. 3 can be followed. The stable patient should be investigated with abdominal ultrasonography, computed tomography (**CT**), or splenic scintiscanning.

Operative management

Laparotomy is indicated for splenic trauma if there is obvious evidence of continuing blood loss despite adequate resuscitation, or if there is clinical suspicion of additional trauma to other organs such as the liver, pancreas, or bowel. In several series of adult patients with abdominal trauma, the incidence of major intra-abdominal injuries associated with blunt splenic trauma is approximately 30 to 60 per cent. If there is any doubt about the presence of an associated intra-abdominal injury, laparotomy should not be delayed. The surgical treatment of these injuries is dictated by the operative findings. At laparotomy the decision to perform splenectomy or splenic repair (splenorrhaphy), the techniques of which are discussed later, will depend largely on the degree of trauma and the expertise available. A simple grading system with suggested management is shown in Table 2.

Grade I:	Capsular injury not actively bleeding
Grade II:	Capsular or parenchymal injury requiring only topical haemostatic agents
Grade III:	Parenchymal injuries requiring suturing + haemostatic agents
Grade IV:	Parenchymal injuries requiring partial splenic resection
Grade V:	Injuries requiring splenectomy

Table 2 A grading system of splenic injuries and their management

The proportion of splenic injuries suitable for repair will vary with the type of trauma seen

in an individual centre and ranges from 30 to 90 per cent. The presence of extensive hilar injury makes it unlikely that the spleen will be suitable for repair, and avulsion or extensive fragmentation requires splenectomy, as does failure to achieve haemostasis following attempted splenorrhaphy. If repair is undertaken in conjunction with resection of splenic tissue, consideration should be given to the amount of spleen that will remain. The 'critical mass' required to confer substantial protection from bacterial infection in experimental models seems to be approximately one-quarter to one-third. Repair in the presence of significant peritoneal contamination is controversial, as is the repair of a diseased, ruptured spleen, but is probably inadvisable.

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After splenic salvage surgery, careful postoperative monitoring is necessary to alert the surgeon to rebleeding. If the clinical condition deteriorates and there is evidence of continuing blood loss, re-exploration of the abdomen should not be delayed. This assessment may be difficult, or indeed impossible in the presence of multiple other injuries, and such circumstances may make splenectomy preferable to attempted repair.

The complications of splenic repair are few, but reported series are small. True 'failures' seem rare and the majority represent inadequate assessment of the damage at the time of laparotomy, missed injuries, or overambitious attempts to repair severely damaged spleens.

Non-operative management

The suspicion of splenic trauma in a patient whose cardiovascular system is stable can be investigated with ultrasound, CT, or scintiscanning. If the radiological investigation confirms the presence of a splenic injury, a decision should be taken on whether it is appropriate to manage the patient conservatively. The proportion of cases judged to be suitable for non-operative management varies from 20 to 45 per cent. Patients are ideally cared for in a high-dependency or intensive care unit, where careful cardiovascular and haematological monitoring with repeated clinical examination is performed. Bed rest is imposed, and, assuming the patient remains stable, discharge to the ward can occur after 24 to 48 h of observation, after which activity can be increased up to discharge. It is prudent to restrict activity for 4 to 6 weeks and to avoid contact sports for up to 6 months. Regular rescanning (by either ultrasonography or isotope scinti-graphy) will monitor resolution of the trauma, which is complete at 3 months in 90 per cent of cases. Deterioration, or evidence of bleeding not controlled by transfusion, is an indication for surgery.

Up to nearly one-third of cases managed in this way will 'fail' and require surgery. Variations between series (failure rates of 0 to 30 per cent) reflect the differences in clinical judgement about which patients should be treated non-operatively. Most instances in which conservative management fails and a laparotomy is needed result in the patient losing their spleen. Indeed, the protagonists of splenic repair would argue that more spleens would be preserved overall if early laparotomy were to replace attempts at conservative management.

Conservative (non-operative) management of splenic trauma has been most widely

practised in children, because the increased incidence of overwhelming postsplenectomy infection in the child behoves the surgeon to attempt to conserve the spleen if possible. Fortunately, there are various reasons why conservative management of splenic injury in children is more likely to be successful than in adults. The type of injury suffered by children is less likely to be of the penetrating type (as associated with crimes of violence), and multiple injuries are also less common. Both criteria make non-operative management safer in the child. Furthermore, children have a better capacity for haemostasis, an increased resilience of the cardiovascular system to hypovolaemia, and an increased compliance of the splenic capsule and septa. All these factors improve the chances of a successful outcome for conservative management.

Splenectomy for haematological disease

Splenectomy may be required in the management of a variety of haematological disorders. Indications include the need to alleviate complications, to prevent repeated episodes of infarction, to prevent massive red-cell pooling and as part of the management of hypersplenism.

Hypersplenism is a clinical syndrome with many causes (Table 3). It is characterized by splenic enlargement, haematological cytopenia (reduction in one or more cellular components of the blood as a result of splenic pooling), maturation arrest in the marrow, and the premature release of immature cells into the circulation. Red cell and/or platelet survival may be decreased. Anaemia, neutropenia, and thrombocytopenia can all occur, either alone or in any combination, their relative severity depending on the nature of the underlying disease. Usually the decision whether the patient will benefit from splenectomy is made on the basis of knowledge about the particular disease process and the severity of the haematological cytopenia, but in difficult cases it may be necessary to use special tests of splenic function.

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These may include isotope studies of the splenic red-cell pool, splenic erythropoiesis and tests of red-cell survival, but despite their use the haematological outcome of splenectomy may be difficult to predict and the decision to operate is made by a multidisciplinary team that includes the haematologist, surgeon, immunologist, and microbiologist.

Lymphoma (Hodgkin's disease and non-Hodgkin's lymphoma)
Chronic lymphatic leukaemia
Hairy-cell leukaemia
Portal hypertension (Banti's syndrome)
Rheumatoid arthritis (Felty's syndrome)
Infection (e.g. malaria and kala-azar)
Infiltrative diseases (e.g. sarcoid)
Lipid storage disease (e.g. Niemann–Pick syndrome, Gaucher's disease)

Table 3 Causes of hypersplenism

Idiopathic thrombocytopenic purpura

Idiopathic thrombocytopenic purpura mainly affects females between the ages of 15 and 50 years. It presents with bruising, usually after trauma or pressure, and examination reveals variable numbers of petechial haemorrhages in the skin. The clinical course is often intermittent and chronic. The platelets of affected patients become sensitized by antiplatelet IgG autoantibodies and are then removed from the circulation. Many patients need no treatment if their platelet count remains over 50×10^9/l and no spontaneous bleeding occurs. If the count falls below 20×10^9/l the risk of bleeding increases. Spontaneous recovery occurs in under 10 per cent of patients and treatment is initially directed at decreasing the autoantibodies and the rate of platelet destruction. Eighty per cent of patients go into remission on steroid therapy (60 mg prednisolone/day). If this does not result in remission, or continuing high doses are needed, or the platelet count remains low (under $20-30 \times 10^9$/l), splenectomy is indicated, the spleen being the initial and major site of antibody production and also a major site of platelet destruction. Good results are usually obtained, with approximately 80 per cent of patients having a satisfactory outcome, no longer requiring steroids to maintain an adequate number of platelets. However, in more heavily sensitized patients, platelets can continue to be destroyed elsewhere in the reticuloendothelial system, including the liver, and after an initial good response a relapse may also occur secondarily to an acute bacterial or viral illness. A good response is most likely in patients under the age of 45 years, in those in whom the thrombocytopenia is less severe, and in those who have shown at least an initial response to steroid therapy. At operation, the spleen is usually of normal size and no special technique is needed for its removal, although a careful search must be made to ensure that no splenunculi remain. Although the preoperative platelet count may be very low, platelet transfusion is not begun until the splenic vessels have been ligated, when 6 to 10 units of platelets may be transfused. In addition, some patients may benefit from high doses of intravenous gammaglobulin preoperatively to raise the platelet count to an acceptable number (greater than 20×10^9/l).

Haemolytic anaemias

Hereditary spherocytosis

This is an autosomal-dominant hereditary disorder characterized by small, spherocytic red cells. Abnormalities in the cell membrane transform the red cells to a spherical shape, which results in a reduced ability to be deformed and therefore to entrapment in the spleen. The clinical presentation is commonly in childhood but may be delayed until later in life. Mild, intermittent jaundice is associated with mild anaemia, splenomegaly, and gallstones. The diagnosis is made on examination of a peripheral blood film, in which reticulocytes and spherocytes are seen. Serum bilirubin is commonly elevated.

If the disease is severe, splenectomy is the treatment of choice but surgery is normally

delayed until over the age of 6 years (and always until the age of 2 years), to minimize the risk of postsplenectomy infection. The red cells retain their spherical appearance but in the absence of the spleen they have a near-normal survival. The long-term results of the operation are excellent, relapses being attributed to missed splenunculi, but with no very convincing evidence.

The decision to operate for mild to moderate disease is more difficult and the treatment of patients with asymptomatic anaemia is controversial. The efficacy of partial splenectomy to relieve haemolysis but maintain phagocytic function is an attractive possibility currently under study.

Hereditary elliptocytosis

This condition is characterized by oval-shaped red cells caused by an abnormality in the cell membrane. There are three types, two inherited as an autosomal-dominant and one as a recessive trait. Most heterozygous carriers require no treatment, but if the anaemia is symptomatic, partial relief can be obtained from splenectomy.

β-Thalassaemia

The management of this condition involves regular blood transfusion, chelating agents to minimize the problem of iron overload, and the judicious use of splenectomy if hypersplenism occurs and the patient becomes transfusion dependent. Marked splenomegaly can occur (Figs 4, 5). If indicated, surgery should be delayed until at least 5 years of age.
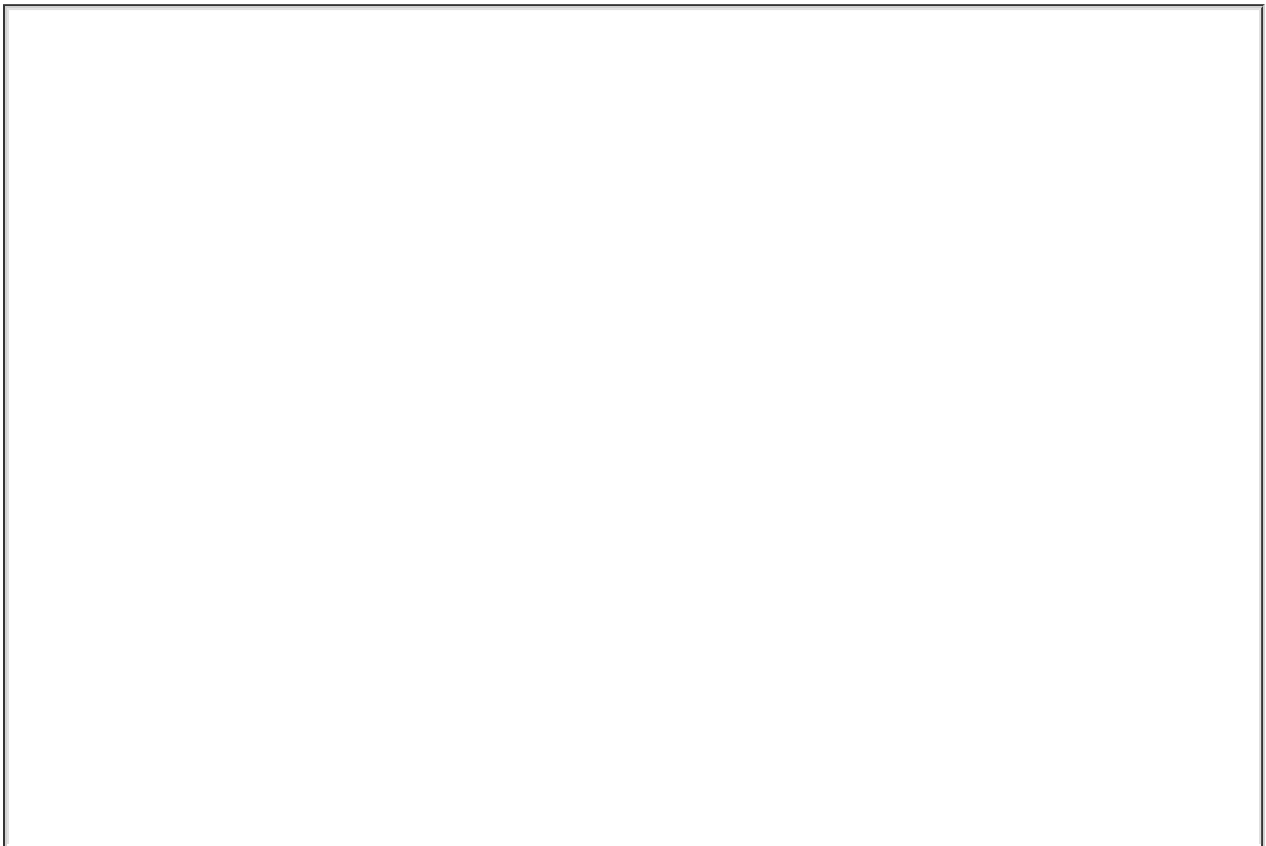
Fig. 5. The mobilized spleen of the patient shown in Fig. 4.

Sickle-cell disease

In this condition, splenic infarction can occur, and only in rare cases is removal of the spleen indicated. In cases of sequestration crisis there is rapid enlargement of the spleen or liver that may be fatal. If the infant or baby survives, these episodes have a tendency to recur and splenectomy may be required. In addition the spleen may enlarge to such a degree that hypersplenism develops, a problem most common in areas in which malaria is endemic.

Lymphoma

The surgeon may be required to remove the spleen for a variety of reasons in the management of patients with lymphoma (Fig. 6). Occasionally, splenectomy may be needed to achieve a diagnosis (in the absence of palpable nodes for biopsy) and this is becoming more frequent in human immunodeficiency virus-related disease, in which splenic lymphoma may develop. Splenectomy may also be indicated in hypersplenism (see above), or to relieve the symptoms of gross splenomegaly (such as satiety and discomfort in the

left upper quadrant). Staging laparotomy for either Hodgkin's disease or non-Hodgkin's lymphoma is now rarely performed. In Hodgkin's disease, accurate staging is important as it has a bearing on both prognosis

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and the selection of treatment for an individual patient, but no clear survival advantage has been demonstrated in patients who have been surgically staged. This probably relates both to the success of salvage chemotherapy for disease that relapses after radiotherapy, and an increased tendency to use chemotherapy for early, bulky disease. Hence CT is increasingly replacing surgical staging, although the technique is not without its limitations. It cannot detect disease in normal-sized nodes, differentiate diseased from reactive nodes, or demonstrate micronodular involvement of the spleen (Fig. 7). Magnetic resonance imaging can be useful when there is a suspicion of focal bone or marrow involvement.

Fig. 6. An enlarged spleen showing extensive involvement with disease in a patient with stage IV Hodgkin's disease. The patient had a pancytopenia due to hypersplenism, precluding chemotherapy. Splenectomy led to haematological remission allowing the initiation of chemotherapy, which produced a complete remission in disease.
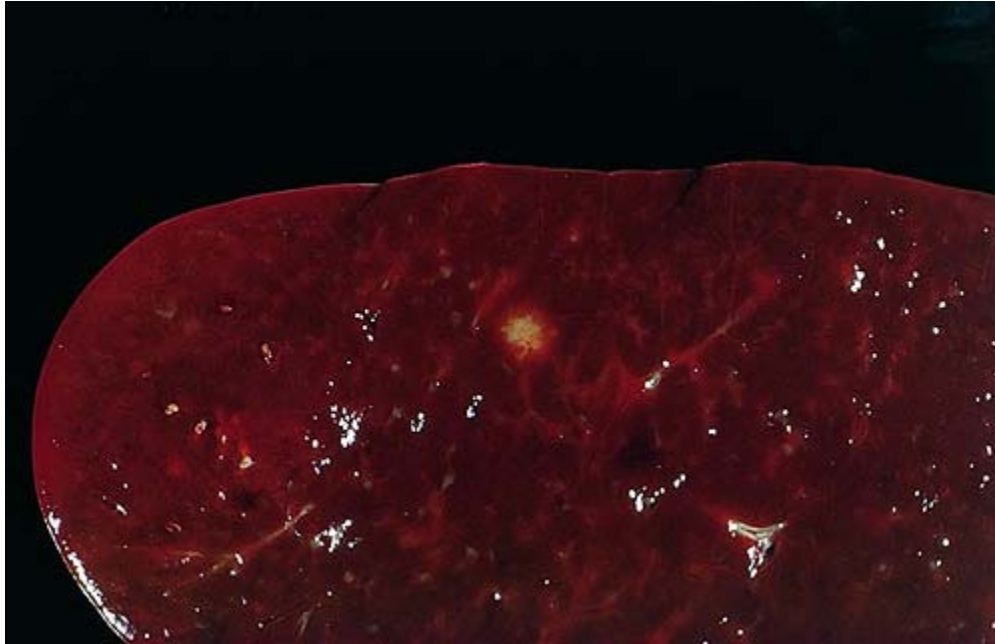
Fig. 7. The spleen of a patient with clinical stage 1A Hodgkin's disease who presented with nodes in the neck. No disease was evident in the abdomen at laparotomy but one positive nodule of disease was found on one cut surface of the spleen.

In low-grade non-Hodgkin's lymphoma, splenectomy may be helpful if the disease is largely confined to the spleen, especially if hypersplenism is a feature.

Chronic lymphatic leukaemia

In chronic lymphatic leukaemia, 50 per cent of patients will have splenomegaly of variable degree, and splenectomy can be of value in selected cases. Indications for surgery include those cases resistant to medical management in which splenomegaly is present, when hypersplenism develops, or if the patient develops autoimmune complications (such as haemolytic anaemia or thrombocytopenia) and this fails to respond to medical treatment. In patients with chronic lymphatic leukaemia where the spleen is the main organ affected, splenectomy

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can downstage the disease and can lead to an improvement in survival. It is worthwhile to remember that these patients have poor humoral immunity and so postsplenectomy prophylaxis against infection will rely mainly on antibiotics rather than immunization.

Hairy-cell leukaemia

In hairy-cell leukaemia, a low-grade, B-cell condition of middle-aged men, two-thirds of patients have splenomegaly, and premature destruction of blood cells occurs during their passage through the spleen. In the past, splenectomy has been the treatment of choice, but

is now reserved for patients with a very large spleen. Interferon-α improves the blood count and the bone marrow but rarely induces a complete remission; new drugs, including deoxycoformycin, lead to remission rates of approximately 75 per cent.

Myelosclerosis

This is a disease for which there is no definitive treatment. It may present with splenomegaly, which may be massive, but the place of splenectomy is controversial. If the spleen is removed early in the disease it will prevent the development of hypersplenism, but the operation may be technically difficult and followed by a massive thrombocytosis that may prove resistant to treatment. Splenectomy is probably best reserved for cases in which splenic pain or hypersplenism are major features.

Splenectomy for other reasons

1. Abscess: a relatively rare condition, generally requiring a splenectomy.

2. Hydatid disease (see Chapter 47.2.4)

3. As part of a cancer operation: because of the local extension of malignant tumours, the spleen may need to be resected as a part of operations to remove tumours of the greater curve of the stomach, the splenic flexure, or the tail of the pancreas. Splenectomy as part of radical gastric surgery for malignancy is no longer routinely practised.

4. Felty's syndrome (rheumatoid arthritis, splenomegaly, leucopenia): splenectomy may lead to a temporary rise in the neutrophil count but does not seem to provide reliable protection against recurrent infection. It is best reserved for when severe anaemia is secondary to hypersplenism.

5. Systemic lupus erythematosus: this may be associated with immune thrombocytopenic purpura and the role of splenectomy is controversial; it is most successful when combined with immunosuppression.

6. Human immunodeficiency virus infection: in the later stages of this disease, mild to moderate splenomegaly may develop and splenectomy may be necessary to exclude lymphoma or to diagnose opportunistic infection.

7. Immunosuppression: splenectomy will prolong renal allograft survival in experimental models in the rat, and has been shown to produce improved survival of human renal allografts in association with conventional immunosuppressive drugs. However, as time progresses there is an increased death rate from overwhelming infection; as a result, splenectomy is no longer practised as part of an immunosuppressive protocol in clinical renal transplantation.

Operations on the spleen

Historical background

The observation that the asplenic state was compatible with life was made by Aristotle and was confirmed by experiments in the seventeenth and eighteenth centuries by Wren and Morgagni. It is unknown when the first splenectomy was done but splenectomy as treatment for a disease process was possibly first performed in 1549 by Adriana Zaccarello (although there is dispute as to whether the removed organ was in fact an ovarian mass!). The first splenectomy for trauma was in 1678 by Nicholas Matthias. In 1928, William Mayo reported a series of 500 splenectomies, with a mortality rate of 10 per cent. In the absence of a clear understanding of splenic function, subsequent reports of healthy survivors led to the concept that there were no ill effects from splenectomy. Despite the fact that in 1919 Morris and Bullock had reported that asplenic rats were susceptible to infection and had a shorter lifespan than other rats, the caution advised by these investigators went unheeded for over 30 years. In 1953, a report from King and Schumacker demonstrated an increased susceptibility to infection and death from sepsis in infants who had undergone splenectomy for congenital spherocytosis. It had been noted by Billroth that haemostasis could occur in a traumatized spleen without removal and that non-operative management of splenic trauma was therefore feasible. Early in the twentieth century, initial attempts at non-operative management and splenorrhaphy had poor results and splenectomy was the rule for trauma. In the latter half of the twentieth century, with the recognition of the risk of overwhelming postsplenectomy infection, especially in children, non-operative management has become an option once more and the current management of trauma is usually dictated by the age of the patient, the experience of the institution, the individual surgeon, and the type of trauma.

Operations

Open splenectomy

General technique

Under general anaesthesia with endotracheal intubation and muscle relaxation, the patient is positioned supine on the operating table. After the intravenous administration of a prophylactic antibiotic (a penicillin or second-generation cephalosporin), the abdomen is opened through an upper midline, a left paramedian, a subcostal, or, rarely, a thoracoabdominal approach. Having opened the peritoneal cavity, a laparotomy is performed appropriate to the indication for surgery. Before mobilization of the spleen it is advisable to divide, if necessary after diathermy or ligation, any adhesions between its lower pole and the greater omentum or splenic flexure. The colon can then be retracted down with the aid of a moistened abdominal pack. The right-handed surgeon then retracts the spleen medially with the left hand in order to facilitate division of the posterior layer of the lienorenal

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ligament behind the spleen by sharp dissection. This is usually avascular unless splenomegaly has resulted in vascular adhesions at this site. The gastrosplenic ligament is then divided between ligatures on the short gastric vessels passing to the upper pole of the

spleen, care being taken not to include any gastric tissue in the ligatures. Having divided these two ligaments the spleen can be delivered medially into the wound and the splenic pedicle identified. The splenic artery and vein are then double ligated separately, without damaging the tail of the pancreas in the splenic hilum. After checking for haemostasis the abdomen is closed and a drain is not usually needed. In the presence of oozing, a suction drain can be left in the left upper quadrant for 24 to 48 h. If there has been suspected or definite damage to the tail of the pancreas, a tube drain should be inserted to drain a potential pancreatic fistula.

Special problems

When splenectomy is done for traumatic injury, an upper midline incision is recommended in order that a full laparotomy may be performed as necessary. In such cases the lienorenal ligament may have been avulsed and the spleen may deliver into the wound with little mobilization. However, if salvage surgery is planned (see below), then mobilization must be done with extra care.

When performing splenectomy for massive splenomegaly, a subcostal incision can be used if the costal margin is wide and if the spleen does not extend below the umbilicus, in which event a midline incision is preferable. In the absence of adhesions between the diaphragm and the spleen, the size of the spleen stretches the ligaments and mobilization is not usually difficult. However, in the presence of dense adhesions, these should ideally be divided under direct vision, if necessary after ligation of the splenic artery.

Laparoscopic splenectomy

Laparoscopic splenectomy was first described in 1992; in some centres, especially those with a large paediatric practice, it is now used for the elective removal of the spleen in selected cases, especially in idiopathic thrombocytopenic purpura. The operation involves the use of either four or five ports, usually with the patient in the lateral position. Whether the main splenic vessels should be identified and stapled as the first procedure or after mobilization of the spleen is a matter of debate, but following successful splenectomy the organ is either removed in a bag via the umbilical port (after maceration) or via a small Pfannensteil incision. A specific problem is that splenunculi may be missed and left *in situ*, which may result in a poor operative result when the splenectomy was for a haematological disease such as idiopathic thrombocytopenic purpura. The size of the spleen must also be assessed preoperatively, with most series quoting an upper limit of 20 cm for successful removal. It is an operation with a considerable 'learning curve', but in common with other laparoscopic techniques, advocates of the procedure report reductions in analgesia requirements and length of hospital stay, and a more rapid return to normal activities. To date there have been no randomized studies to compare morbidity between the open and laparoscopic operation.

Operations for splenic salvage

As the spleen is a delicate, easily traumatized organ, splenic trauma can result in haemorrhage. For the reason discussed below, salvage of the spleen is preferable to

splenectomy in certain situations. Various techniques have been described.

Haemostatic agents

For trivial trauma, pressure can be combined with topical agents to arrest bleeding. Fibrin glue is a highly concentrated form of human fibrinogen and clotting factors. It is used by spraying a thin layer directly on to the injury or injecting it into or over a fractured surface. For knife or bullet tracts it can be injected deep into the base of the injury and slowly withdrawn. This distends the tract slightly to allow for both a haemostatic and tamponade effect. The glue can also be used in association with sutures or to seal the splenic surface after a partial resection. It has advantage over cyanoacrylate adhesive in that it is less histiotoxic. Microfibrillar collagen acts by trapping and then activating platelets. It forms a firm, adherent coagulum with an affinity for moist surfaces. It has been shown to be hypoallergenic, exciting little in the way of tissue reaction, and is absorbed in 3 to 6 weeks. It should be applied with dry instruments and in sufficient amounts to cover the bleeding surface to a depth of several millimetres. Following application, pressure should be applied for about 5 min. Linear or stellate cracks can be packed and it is most successful if the surface blood flow can be reduced to a mild to moderate ooze. Other topical agents include gelatin foam, thrombin, and bovine collagen. These techniques are applicable to minor superficial lacerations or capsular tears without parenchymal damage.

Splenic artery ligation

The main splenic artery can be ligated in continuity in an attempt to reduce blood loss from a traumatized spleen; this can be done without inevitable splenic infarction and loss of function, provided the short gastric vessels are intact. Arterial ligation may also be used in addition to the other techniques described below. The branches of the splenic artery can be dissected at the hilum. The most constant is the superior polar artery, which is the first branch from the main artery before entering the hilum. The lower polar vessels may not be obvious outside the spleen and it may be necessary to ligate the main vessel distal to the superior polar branch. The branches to bleeding segments may be ligated individually, either with or without partial splenectomy. Ligation of the short gastric vessels may be indicated for bleeding from the upper pole. After ligating any of the vessels, the spleen should be carefully examined so that any devitalized tissue can be resected by sharp dissection, diathermy, or blunt finger dissection. During this technique, as vessels are encountered they may be clipped, tied, or under-run. When under-running with a stitch the parenchyma of the spleen may cut through, but the vessel wall will hold and residual bleeding can be controlled with the aid of haemostatic agents as described above. Stitches can be tied over pledgets of absorbable gelatin sponge (Gelfoam), oxidized regenerated cellulose (Surgicell), or compressed microfibrillar collagen.

Repair techniques (splenorrhaphy)

Before any repair is undertaken the spleen must be fully mobilized to assess the extent of damage. During this procedure, extra care must be taken not to strip the posterior capsule, as consequent blood loss may preclude a successful repair. Capsular avulsion is most likely to occur if the incision in the posterior peritoneum is made too close to the spleen.

Any peritoneal folds to the lower pole of the spleen from the greater omentum must be carefully divided. Following this incision, blunt dissection posteriorly is continued to elevate the spleen and the tail of the pancreas. If time permits, isolation of the splenic artery with a vascular sling is ideal to allow occlusion with a

vascular clamp if necessary. The veins are fragile and no attempt should be made to isolate them.

Splenorrhaphy should aim both to achieve control of bleeding and to avoid causing or leaving behind infarcted splenic tissue. Simple suture of the torn spleen often results in further bleeding, but the use of a buttress technique with collagen, omentum, or Teflon pledgets can be effective. Haemostatic agents can be used in addition to the sutures (see above). The spleen may be wrapped in the greater omentum to assist haemostasis, and Dexon and Vicryl mesh can be used to achieve a similar effect.

In view of the segmental nature of the vascular supply, if one pole or segment of the spleen has been extensively damaged, partial splenectomy can be successfully performed. Haemostatic agents or an omental buttress can then be applied to the raw splenic surface after resection.

In general, it is appropriate to drain the left upper quadrant after a repair procedure. Drainage, in addition to clinical assessment, can be used to determine the success of the procedure. These procedures are often technically more demanding than a simple splenectomy, and it must be constantly remembered that the short- and long-term morbidity of such a procedure must be less than that resulting from splenectomy in order for it to be an acceptable part of the treatment.

Complications of splenectomy

General

Certain complications are specific to the operation of splenectomy, as opposed to those seen after any abdominal operation. Atelectasis of the left lower lobe is common, and all patients should receive active physiotherapy from the first postoperative day. Gastric ileus is usually short lived and a nasogastric tube is not needed routinely. Postsplenectomy fever is described and cannot always be attributed to atelectasis or subphrenic haematoma; in the absence of a definite cause it is a self-limiting feature.

Thrombocytosis and thrombosis

The platelet count often increases to between 600 and 1000×10^9/l postoperatively, usually peaking between days 7 to 12. This rise is usually transitory but may last up to 3 months. If a count of 750×10^9/l is reached it is a sensible precaution to administer aspirin (150 mg/day) to prevent deep venous thrombosis.

Overwhelming postsplenectomy infection

Definition and cause

Splenectomy is the traditional treatment for the traumatized spleen, but it is now well recognized that there are disadvantages to the asplenic state, the most important of these being the potential for the development of overwhelming postsplenectomy infection, a term coined by Diamond in 1969. The clinical course consists of a fulminant bacteraemia, a frequent absence of a septic focus, coma, shock, consumptive coagulopathy, and adrenal haemorrhage. The bacteraemia is most commonly associated with the encapsulated organism *Streptococcus pneumoniae*, and less commonly with *Neisseria meningitidis, Escherichia coli*, and *Haemophilus influenzae*.

Removal of the spleen renders the patient more susceptible to infection for two reasons. Firstly, the absence of the red pulp results in impairment of phagocytosis and clearance of exogenous organisms. Secondly, antibody production and subsequent bacterial opsonization are reduced by the removal of the white pulp, and poorly opsonized bacteria are cleared less effectively by the liver.

Incidence

Estimates of the incidence of overwhelming postsplenectomy infection and its associated mortality vary, but large overviews found an overall incidence of approximately 1 to 5 per cent, with all series showing the incidence in children and adolescents to be consistently higher than in the adult population. The incidence of overwhelming postsplenectomy infection varies according to the indication for which the splenectomy is performed. Some 1.5 to 2.5 per cent of children develop overwhelming postsplenectomy infection when the spleen is removed for trauma, but the greatest risk is seen in patients undergoing splenectomy for congenital anaemias, portal hypertension, or lymphoreticular tumours. The time of development of overwhelming postsplenectomy infection is variable in relation to the splenectomy. Although it has been described up to 30 years after loss of the spleen, it usually occurs within 3 years after the operation. It is of particular importance, as this infection is associated with a very high mortality rate (25–75 per cent reported in many series). It is primarily to avoid this complication that a renewed interest in splenic preservation has arisen, especially in children. Attempts to preserve the spleen can be achieved by conservative, non-operative management, or by one or other of the splenic salvage operations described in the previous section.

Prophylaxis

Antibiotics

The effect of prophylactic antibiotics on overwhelming postsplenectomy infection is controversial. Traditionally, oral penicillin has been given postoperatively for periods that have varied from 6 weeks to 5 years, or until the onset of puberty if splenectomy is performed in a child. As the time of greatest risk is during the first 2 or 3 years after splenectomy, it would not be unreasonable to give all patients some form of antibiotic prophylaxis at least during that time. However, many septic episodes in asplenic patients are secondary to organisms not sensitive to penicillin and patient compliance is a problem.

As a compromise, it seems appropriate to treat patients with prophylactic antibiotics only if they are at high risk, such as children and immunosuppressed patients. For the prevention of infection, antibiotics may be given in reduced dosage to avoid undesirable side-effects. Penicillin V (250 mg daily) is a recommended prophylactic dose. It is likely that antibiotics are of more use when given promptly and appropriately when infection develops, and clinicians should ensure that asplenic patients are supplied with amoxycillin (amoxicillin) to take at the first sign of a respiratory illness or fever. Amoxycillin is recommended firstly because of the higher blood concentrations achieved with oral administration, and secondly because of its activity against *H. influenzae*.

Vaccines

Vaccines are available to the pneumococci, *H. influenzae*, and meningococci. Splenectomy results in a reduced antibody responsiveness and this is most marked in the young and those with malignancy or on immunosuppressive drugs. Although vaccination failures have been reported, in the case of elective surgery, vaccination is recommended and should be given as soon as a decision to operate has been taken. In a trauma victim, vaccination can be given in the postoperative period and the resulting amounts of antibody

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will be protective in the majority of cases, although they are less than 50 per cent of those achieved if vaccination is given in the presence of an intact spleen. Following vaccination, antibody titres remain high for at least a year before slowly declining, but this rate is increased in immunosuppressed patients and those with lymphoma. Although the incidence of pneumococcal infection has been shown to be reduced following vaccination, protection is not guaranteed. The reasons for this include the fact that about half the organisms causing postsplenectomy sepsis are not pneumococcal, and further more, 20 per cent of subtypes are not covered. Antigenic types also vary and therefore protection from a particular vaccine may be incomplete.

The pneumococcal vaccine consists of purified, capsular polysaccharide antigens of the 23 most prevalent serotypes of *S. pneumoniae*. Most healthy adults show at least a twofold rise in antibody titre within 2 weeks. Antibody titres of 256 ng immunoglobulin nitrogen/ml or more are indicative of immunity. Raised antibody titres have been demonstrated 5 years after vaccination but the exact duration of protection is not known. Current advice is to recommend revaccination every 6 years.

The vaccine against *H. influenzae* consists of bacterial polysaccharide conjugated to protein. The response to this preparation and its long-term efficacy are greater than the response to the polysaccharide vaccine alone and so its use is particularly important in children and when vaccination has to be given postoperatively.

Implantation of splenic tissue

Splenosis, or spontaneous regrowth of splenic tissue following splenectomy for trauma, possibly accounts for the lower incidence of overwhelming postsplenectomy infection in patients who have had splenectomy for trauma rather than for other reasons. Detection of splenosis by scintiscanning is complicated by the presence of splenunculi. The precise incidence of splenosis is unknown but may be as high as 50 per cent following trauma.

Function of the splenic tissue can be assessed using red cell morphology, tests of phagocytic function, the presence of Howell–Jolly bodies, assays of IgM, IgG, and IgA, and scanning. It is clear that the presence of splenosis does not eliminate the risk of developing overwhelming postsplenectomy infection, as there have been documented reports of deaths from this cause occurring in patients in whom splenosis has been demonstrated at autopsy. It has been estimated that 25 to 30 g of tissue is needed for protection against overwhelming postsplenectomy infection.

In an attempt to conserve functioning splenic tissue in cases in which splenunculi are not present and where splenorrhaphy is not possible, splenic tissue can be autotransplanted at the time of surgery by dicing the spleen into small pieces and implanting them into the abdominal wall or an omental pouch. Both techniques can be shown to result in functioning splenic tissue and it is clear from reports that such transplants can be shown to survive, but experimental work has shown a variable potential for decreasing the incidence of infection in asplenic animal models. Further evidence suggests that the mere presence of functioning splenic tissue in itself is insufficient to protect against overwhelming postsplenectomy infection (whether from splenosis or as a result of autotransplantation), and it seems likely that the normal splenic vasculature is crucial for maximum protection. Overwhelming postsplenectomy infection affects only 2.5 per cent of patients after splenectomy for trauma and clinical reports of 'successful' transplants are anecdotal, so it is unclear whether it should be routine practice, but as it is not associated with significant morbidity there seems little to be lost by performing it.

Further reading

Bickerstaff KI, Morris PJ. Splenectomy for massive splenomegaly. *British Journal of Surgery* 1987; **74**: 346–9. [Outcome review of 47 patients.]

Cooper MJ, Williamson RCN. Splenectomy: indications, hazards and alternatives. *British Journal of Surgery* 1984; **71**: 173–80. [Excellent review article.]

Foss Abramsen A *et al*. Systemic pneumococcal disease after staging splenectomy for Hodgkin's disease 1969–1980 without pneumococcal vaccine protection: a follow up study 1994. *European Journal of Haematology* 1997; **58**: 73–7. [Demonstrates 20-fold increased risk compared with the general population.]

Ghosh S, Symes JM, Walsh TH. Splenic repair for trauma. *British Journal of Surgery* 1988; **75**: 1139–40. [Thirteen cases of repair from a single centre.]

Holdsworth RJ, Irving AD, Cushchieri A. Postsplenectomy sepsis and its mortality rate: actual versus perceived risks. *British Journal of Surgery* 1991; **78**: 1031–8. [Literature review of postsplenectomy sepsis 1952–1987.]

Katkhouda N *et al.* Laparoscopic splenectomy: outcome and efficacy in 103 consecutive patients. *Annals of Surgery* 1998; **228**: 568–78. [Good account of operative technique.]

King H, Schumaacker HB. Splenic studies. I. Susceptibility to infection after splenectomy performed in infancy. *Annals of Surgery* 1952; **136**: 239–42. [Historical interest.]

Kram HB, del Junco T, Clark SR, Ocampo HP, Shoemaker WC. Techniques of splenic preservation using fibrin glue. *Journal of Trauma* 1990; **30**: 97–101. [Good account of use of glue.]

Malangoni MA, Levine AW, Droege EA, Aprahamian C, Condon RE. Management of injury to the spleen in adults. *Annals of Surgery* 1984; **200**: 702–5. [Review of 77 patients.]

Morgenstern L, Shapiro SJ. Techniques of splenic conservation. *Archives of Surgery* 1979; **114**: 449–54. [Good review of available techniques.]

Morris DH, Bullock FD. The importance of the spleen in resistance to infection. *Annals of Surgery* 1919; **70**: 513–21. [First experimental demonstration of overwhelming postsplenectomy infection.]

Morris PJ, Cooper IA, Madigan JP. Splenectomy for haematological cytopenias in patients with malignant lymphomas. *Lancet* 1975; **ii**: 250–3. [Review of 86 patients.]

Mucha P, Daly RC, Farnell MB. Selective management of blunt splenic trauma. *Journal of Trauma* 1986; **26**: 970–9. [Good discussion.]

Redmond HP, Redmond JM, Rooney BP, Duignan JP, Bouchier-Hayes DJ. Surgical anatomy of the human spleen. *British Journal of Surgery* 1989; **76**: 198–201. [A useful article when splenic salvage is to be considered.]

Rescoria FJ, Breitfeld PP, West KW, Williams D, Engum SA, Grosfeld JL. A case controlled comparison of open and laparoscopic splenectomy in children. *Surgery* 1998; **124**: 670–5. [Non-randomized comparison and good discussion.]

The Splenic Injury Study Group. Splenic injury: a prospective multicentre study on non-operative and operative treatment. *British Journal of Surgery* 1987; **74**: 310–13.

[Multicentre study of splenic salvage.]