Splenectomy, Hyposplenism and Asplenia

The spleen is involved in producing protective humoral antibodies, the production and maturation of B and T cells and plasma cells, removal of unwanted particulate matter (eg bacteria) and also acting as a reservoir for blood cells, especially white cells and platelets.

Splenectomy

Splenectomy may occur in three different ways:

- Planned, where prophylactic measures can be used to prevent later complications.
- Traumatic, due to an accident or during surgery.
- Autosplenectomy, which refers to the physiological loss of spleen function (hyposplenism), eg associated with sickle cell anaemia (chronic damage to the spleen results in atrophy), coeliac disease, dermatitis herpetiformis, essential thrombocytopenic purpura and ulcerative colitis.

Indications for splenectomy

- Trauma: 25% of injuries are iatrogenic.
- Spontaneous rupture: this usually occurs in patients with massive splenomegaly (eg infectious mononucleosis) and is often precipitated by minor trauma.
- Hypersplenism: hereditary spherocytosis or elliptocytosis, idiopathic thrombocytopenic purpura.
- Neoplasia: lymphoma or leukaemic infiltration.
- With other viscera: total gastrectomy, distal pancreatectomy.
- Other indications: splenic cysts, hydatid cysts, splenic abscesses.

Complications of splenectomy

- Thrombocytosis: platelet count usually peaks after 7-10 days. There is no evidence of an increased risk of thromboembolic disease but prophylactic aspirin may be considered for very high platelet counts.
- Overwhelming post-splenectomy infection:
  - Due to encapsulated bacteria such as Streptococcus pneumoniae, Haemophilus influenzae and Neisseria meningitidis.
  - Occurs post-splenectomy in 4% of patients without prophylaxis.
  - The greatest risk of mortality is in the first two years and is estimated at 50%.
  - Management: immunisation and antibiotic prophylaxis as outlined under ‘Management’, below.

Hyposplenism

Causes

- Operative splenectomy: for severe splenic trauma, splenic cysts, or as part of a resective procedure for an abdominal tumour.
- Functional hyposplenism: sickle cell anaemia (HbSS) disease and haemoglobin sickle-C (HbSC) disease, thalassaemia major, essential thrombocytopenic purpura, and lymphoproliferative diseases (Hodgkin’s lymphoma and non-Hodgkin's lymphoma, chronic lymphocytic leukaemia (CLL)), coeliac disease, inflammatory bowel disease. Often, Howell-Jolly bodies on peripheral blood film give an important clue to diagnosing hyposplenism.
- Bone marrow transplantation: (splenic irradiation or chronic graft-versus-host disease).
- Congenital asplenia: (associated with cardiac abnormalities and biliary atresia).

Investigations

- Blood film: features of hyposplenism include Howell-Jolly bodies, Pappenheimer bodies, target cells and irregular contracted red blood cells.
Complications of hyposplenism

- Fulminant, potentially life-threatening infection is a major long-term risk of hyposplenism; yet, such infection is largely preventable.
- The most common infection is pneumococcal infection (mortality up to 60%), followed by *H. influenzae* type b (less common but significant in children), and *N. meningitidis*.
- Other infections include *Escherichia coli*, malaria, babesiosis, and *Capnocytophaga canimorsus* (associated with dog bites).
- Asplenic patients should be strongly advised of the increased risk of severe falciparum malaria, should take all antimalarial precautions/prophylaxis and ideally avoid holidays in malaria-endemic areas.

Management

- National guidelines recommend the following:[1] [2]
  - Immunisations (all routine vaccines (including live vaccines, e.g. measles, mumps and rubella (MMR)) can be given safely to children or adults with an absent or dysfunctional spleen). Similarly, asplenia or hyposplenism is not a contraindication for live vaccinations prior to travel (e.g. yellow fever and live oral typhoid vaccine).[2]
  - Delay live vaccines if the patient has received immunosuppressants: delay vaccination for three months after stopping systemic steroids (e.g., adults 40 mg prednisolone per day for more than one week, children following a daily dose of 2 mg/kg/day for more than one week, or 1 mg/kg/day for more than one month), delay six months after treatment with chemotherapy/radiotherapy and/or other immunosuppressants (e.g., methotrexate, ciclosporin, etc.) and 12 months after stopping all immunosuppressants for bone marrow transplant (longer if there is evidence of graft-vs-host disease). Further details are in the Green Book (Chapter 6).[3]
  - All vaccines should be given at least two weeks before splenectomy if possible. Following splenectomy, functional antibody responses are better with delayed (14-day) vaccination. All other non-immunised patients at risk should be immunised at the first opportunity.
  - Re-immunisation of asplenic patients is currently recommended every five years. However, antibody levels may decline more rapidly, particularly in patients with sickle cell anaemia and lymphoproliferative disorders. Re-immunisation in these patients may be made on the basis of antibody levels.
  - Children under two years of age have a reduced ability to mount an antibody response to polysaccharide antigens and are, therefore, at particular risk of vaccine failure. The newer conjugate 7-valent vaccine (called PC7, as it affords protection against seven disease-causing strains of pneumococci), produces better response in the under-2s so should be used where available in this age group and in other individuals at particularly high risk.[4]
  - *H. influenzae* type b (Hib) vaccination - give to all unimmunised patients but re-immunisation is not currently recommended.

Summary of vaccinations[3][1]

- If splenic dysfunction occurs aged <2 years and unvaccinated or partially vaccinated - give all routine immunisations including boosters. One month after the Hib/meningitis C (Hib/MenC) and pneumococcal conjugate vaccine (PCV13) booster doses, a dose of MenACWY conjugate vaccine should be given and, after the child’s second birthday, further additional doses of Hib/MenC and pneumococcal polysaccharide vaccine (PPV) should be given.
- If splenic dysfunction occurs aged 2-5 years and fully vaccinated (with PCV7) including booster - give booster dose of Hib/MenC vaccine and PCV13, followed one month later with one dose of MenACWY conjugate vaccine and PPV (at least two months after PCV13).
- If splenic dysfunction occurs aged 2-5 years and fully vaccinated (with PCV13) including booster - give booster dose of Hib/MenC vaccine and PPV, followed one month later with one dose of MenACWY conjugate vaccine.
- If splenic dysfunction occurs aged 2-5 years and unvaccinated or partially vaccinated with PCV7 - give booster dose of Hib/MenC vaccine and PCV13, followed one month later with one dose of MenACWY conjugate vaccine. This should be followed later by a second dose of PCV13 and then PPV (at least two months after PCV13).
- If splenic dysfunction occurs aged >5 years and regardless of previous vaccination history - give booster dose of Hib/ MenC vaccine and a single dose of PPV followed at one month with a single dose of MenACWY conjugate vaccine.
Influenza vaccination - annual influenza vaccination is recommended.

If validated assays are available then it is recommended that response to pneumococcal vaccination and timing for repeat doses be checked.[1]

**Lifelong prophylactic antibiotics**

These are recommended in patients at high risk of pneumococcal infections and the antibiotics of choice are oral penicillin V or macrolides.[1] Patients developing infection, despite measures, must be given systemic antibiotics and admitted urgently to hospital.

- Risk factors for high risk in hyposplenism include: age <16 years or >50 years; poor response to pneumococcal vaccination; previous invasive pneumococcal illness; underlying haematological malignancy resulting in splenectomy (increased risk if immunosuppressed).
- Risk is greatest in the first two years post-splenectomy but continues throughout life (it certainly doesn't stop at age 16).
- Use penicillin V (adult 250-500 mg bd - although 500 mg od may be more realistic if compliance is a particular problem), amoxicillin (adult 250-500 mg daily), erythromycin (adult 250-500 mg daily) orally. Reduce the dose for children. Antibiotics may need to be altered due to differing local antibiotic sensitivities - on the advice of the local public health department.
- Consider recommending that the patient take a full therapeutic dose of antibiotics if they develop infective symptoms such as pyrexia, malaise, shivering, etc. and seek medical advice immediately.
- Allowing patients to have a reserve supply of antibiotics at home or on holiday may also seem appropriate.
- Pneumococcal resistance to penicillins remains low in the UK. Knowledge of local resistance patterns should be used to guide the choice of antibiotic.[1]
- If not deemed to be high risk then the pro's and con's of taking lifelong antibiotic prophylaxis needs to be discussed with each individual patient.

UK Primary Care seems rather bad at implementing these guidelines.[5][6] Only 54% of a sample of 974 post-splenectomy patients had received pneumococcal and Hib vaccinations and were taking antibiotic prophylaxis.[5] A case can therefore be made for the establishment of a disease register of hyposplenic patients and regular auditing.[1] The British Committee for Standards in Haematology recommends the following:[1]

- Patients should be given written information and carry a card to alert health professionals to the risk of overwhelming infection. Patients should wear an alert bracelet or pendant.
- Patients should be aware of the potential risks of overseas travel, particularly with regard to malaria and unusual infections, eg those resulting from animal bites.
- Patient records should be clearly labelled to indicate the underlying risk of infection. Vaccination and re-vaccination status should be clearly and adequately documented.

## Further reading & references

3. Immunisation against infectious disease - the Green Book (latest edition); Public Health England

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