

Trust Guideline for Patients with an Absent or Dysfunctional Spleen

A Clinical Guideline

For use in:	All clinical areas
By:	All clinical staff
For:	Adults, Paediatrics and Neonates
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If Yes – does the strategy/policy deviate from the recommendations of NICE? If so, why?	N/A

This guideline has been approved by the Trust's Clinical Guidelines Assessment Panel as an aid to the diagnosis and management of relevant patients and clinical circumstances. Not every patient or situation fits neatly into a standard guideline scenario and the guideline must be interpreted and applied in practice in the light of prevailing clinical circumstances, the diagnostic and treatment options available and the professional judgement, knowledge and expertise of relevant clinicians. It is advised that the rationale for any departure from relevant guidance should be documented in the patient's case notes.

The Trust's guidelines are made publicly available as part of the collective endeavour to continuously improve the quality of healthcare through sharing medical experience and knowledge. The Trust accepts no responsibility for any misunderstanding or misapplication of this document.

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1. INTRODUCTION

DEFINITIONS

This guideline applies to patients who have recently undergone a splenectomy or who have recently been diagnosed with a dysfunctional spleen.

Splenectomy:

Elective, e.g., for haematological disease or splenic abscess, cysts, mass and neoplasm.
Emergency, e.g., for traumatic injury to spleen or intra operative splenic injury

Dysfunctional Spleen: This includes conditions such as homozygous sickle cell disease and coeliac syndrome that may lead to splenic dysfunction

RISKS OF ASPLENIA OR DYSFUNCTIONAL SPLEEN:

Overwhelming infection is a major risk in patients with an absent or dysfunctional spleen and although uncommon, is associated with a high mortality. These infections are often due to encapsulated bacteria such as *Streptococcus pneumoniae*, *Haemophilus influenzae* type b and *Neisseria meningitidis* and more than half of those infected die. Other serious infections include malaria, babesiosis (caused by tick bite) and *Capnocytophaga canimorsus* (caused by dog bites) and secondary infections following influenza. For these reasons, it is imperative that all patients with an absent or dysfunctional spleen are appropriately immunised and receive appropriate antibiotic prophylaxis.

CHECKLIST

Appropriate vaccinations given

Appropriate antibiotic prophylaxis prescribed

Patient information leaflet and Splenectomy card given
(Available from pharmacy. It is the responsibility of the team looking after the patient to ensure this happens).

<http://www.christie.nhs.uk/media/245400/SplenectomyCard.pdf>

Advise patients that they may wish to invest in an alert bracelet or pendant

EDL informs GP that a splenectomy has been performed.

2. VACCINATION INFORMATION

Elective Splenectomy

Ideally start immunisation course at least TWO (ideally four to six) weeks prior to surgery.

Emergency Splenectomy

Ideally start immunisation course at least TWO weeks post surgery.

Given the changing pattern of routine vaccination, patients of different ages may have different "routine" vaccination histories. It is therefore absolutely essential to assess vaccination requirements against an individual's vaccination history.

First diagnosed at age ten years onwards

Older children and adults, regardless of previous vaccination, should receive :

- One dose of **PPV23**, **MenB** and **MenACWY** conjugate vaccine followed by
- One additional dose of **MenB** 4 weeks later
- Annual **influenza** vaccine each season

Vaccines:

Men ACWY Conjugate = Meningococcal A, C, W135 and Y conjugate vaccine (**Menveo**[®])

Men B = Meningococcal B vaccine (**Bexsero**[®]),

PPV23 = Pneumococcal polysaccharide vaccine

First diagnosed from two years to under ten years of age

Ensure children are immunised according to the national schedule, and they should also receive.

- One dose of **PPV23**, followed by
- One dose of **MenACWY** conjugate vaccine
- If not received the 2+1 schedule for **MenB**, ensure they have received two doses of **MenB** 8 weeks apart since their first birthday
- If they have not received any **PCV** previously they should receive a dose of **PCV13** followed by a dose of **PPV23** 8 weeks later
- Annual **influenza** vaccine each season

Vaccines:

Men ACWY Conjugate = Meningococcal A, C, W135 and Y conjugate vaccine (**Menveo**[®])

Men B = Meningococcal B vaccine (**Bexsero**[®]),

PCV13 = Pneumococcal conjugate vaccine (**Prevenar**[®] 13)

PPV23 = Pneumococcal polysaccharide vaccine

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First diagnosed at 12-23 months of age

If not yet administered, give the routine 12-month vaccines: **Hib/MenC, PCV13, MMR** and **MenB**, plus

- One additional booster dose of **PCV13** and one dose of **MenACWY** conjugate vaccine 8 weeks after the 12-month vaccinations; and
- One dose of **PPV23** after the second birthday and at least 8 weeks after the last dose of PCV13
- Annual **influenza** vaccine each season

Vaccines:

Men ACWY Conjugate = Meningococcal A, C, W135 and Y conjugate vaccine (**Menveo**[®])

Men B = Meningococcal B vaccine (**Bexsero**[®])

Hib/Men C = Haemophilus type B conjugate vaccine (**Menitorix**[®])

PCV13 = Pneumococcal conjugate vaccine (**Prevenar**[®] 13)

PPV23 = Pneumococcal polysaccharide vaccine

MMR=Measles, mumps and Rubella vaccine

First diagnosed under 1 year of age

Children should be fully immunised according to the national schedule, and should also receive

- Two doses of **MenACWY** vaccine at least 4 weeks apart during their first year
- An additional priming dose of **PCV13**, such as to receive a total of 2 priming doses at least 8 weeks apart (commencing no earlier than 6 weeks of age) in their first year
- One additional booster dose of **PCV13** and one dose of **MenACWY** conjugate vaccine 8 weeks after the 12-month vaccinations; and
- One dose of **PPV23** after the second birthday and at least 8 weeks after the last dose of PCV13
- Annual **influenza** vaccine each season for patients aged over 6 months

Vaccines:

Men ACWY Conjugate = Meningococcal A, C, W135 and Y conjugate vaccine (**Menveo**[®])

Men B = Meningococcal B vaccine (**Bexsero**[®])

PCV13 = Pneumococcal conjugate vaccine (**Prevenar**[®] 13)

PPV23 = Pneumococcal polysaccharide vaccine

Revaccination Schedule

- Offer annual influenza vaccine to all patients
- PPV23 vaccination every 5 years

3. ANTIBIOTIC PROPHYLAXIS

All patients should be offered lifelong antibiotic prophylaxis. The increased risk of infection in patients with hyposplenism is life long, but is highest early after splenectomy, the biggest risk being from pneumococcal infection.

Patients deemed to be at highest risk

- Aged <16 years or >50 years old
- Inadequate serological response to pneumococcal vaccination
- A history of previous invasive pneumococcal disease
- Splenectomy for underlying haematological malignancy, particularly those who have received splenic irradiation or who have ongoing GvHD are also at continuing high risk.
- Patients with active ongoing graft-versus-host disease

Adult Antibiotic Prophylaxis		
	Prophylaxis	Duration
First line	Penicillin V 250mg bd	Minimum 2 years but preferably lifelong. However, antibacterial prophylaxis may be discontinued in those over 5 years of age with sick-cell disease who have received pneumococcal immunisation and who do not have a history of severe pneumococcal infection
If penicillin allergy	Clarithromycin 250mg bd	
Child Antibiotic Prophylaxis		
	Prophylaxis	Duration
First line	Under 1 year Penicillin V 62,5mg bd 1 – 5years Penicillin V 125mg bd 5-18 years Penicillin V 250mg bd If cover also needed for <i>H.influenzae</i> in child give amoxicillin instead	Antibiotic prophylaxis should be continued until at least 16 years old (and for a minimum of 2 years and preferably lifelong) However, antibacterial prophylaxis may be discontinued in children over 5 years of age with sick-cell disease who have received pneumococcal immunisation and who do not have a history of severe pneumococcal infection.
If penicillin allergy	1 month – 2 years Erythromycin 125mg bd 2-8 years Erythromycin 250mg bd 8-18 years Erythromycin 500mg bd	

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Standby courses of Antibiotics

Patients may develop infection despite vaccination and antimicrobial prophylaxis; these patients require treatment with broad spectrum antibiotics as soon as possible. All patients should keep a supply of antibiotics at home, changing from prophylactic to therapeutic doses if they develop a febrile illness. This is particularly important for those who do not, or will not take prophylactic antibiotics or due to compliance problems. This should be discussed on an individual basis with the patient. If the patient requires a standby course of antibiotics, we recommend Amoxicillin 500mg tds or Clarithromycin 500mg bd (if penicillin allergy). Patients should be advised of the importance of seeking medical attention as soon as possible if they develop any signs of infection e.g. sore throat, fever, malaise, severe headache and flu-like symptoms.

4. SPECIAL PATIENT GROUPS/SITUATIONS

Chemotherapy and Radiotherapy (or other immunosuppressive treatment)

- Ideally, vaccinations should be given at **least TWO weeks** (ideally 4-6 weeks) before initiation of treatment such as chemotherapy or radiotherapy. Where it is not possible to vaccinate beforehand, splenectomy, chemotherapy or radiotherapy should never be delayed.
- If it is not practicable to vaccinate TWO weeks before the initiation of chemotherapy and/or radiotherapy, immunisation can be delayed until at **least THREE months** after completion of therapy in order to maximise the response to the vaccine, whilst ensuring adequate antibiotic cover is prescribed in the interim.
- Individuals with immunosuppression should be vaccinated in accordance with the standard schedule but it should be borne in mind that these individuals may not make a full antibody response.

Pregnancy/Breast-feeding

- All of the vaccines may be given during pregnancy and breast-feeding when protection is required without delay.

Travel

- Patients should be educated as to the potential risks of overseas travel, particularly with regards malaria and unusual infections, for example those resulting from animal bites and tick bites.

Animal Bites

- Human, dog or other bites may be fatal if untreated due to infection with *Capnocytophaga canimorsus* and other virulent organisms. All animal bites need to be treated quickly with antibiotics.

Tick bites

- Babesiosis is a rare tick borne infection that can cause moderate to severe disease, including haemolytic anaemias. Therefore it is essential to take precautions against being bitten in endemic areas.

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Clinical audit standards

All patients undergoing either elective or emergency splenectomy should be managed according to the Trust Guideline

All patients should be given written information and carry an 'I have no functioning spleen' card to alert healthcare professionals to the risk of overwhelming infection

This guideline will be updated in accordance with changes in the recommendations made in "Immunisation Against Infectious Disease – The Green Book".

Summary of development and consultation process undertaken before registration and dissemination process

The authors listed above on behalf of the Antimicrobial Subcommittee, which has agreed the final content, drafted the guideline. During its development it was circulated for comment to Haematology, Oncology, Surgical and Paediatrics Directorates.

Comments received from these listed parties have been addressed and incorporated into this guideline. This guideline has been extended to include neonates, as well as paediatrics and adults.

Distribution List

The Guidelines Assessment Panel has ratified this guideline. The Guideline has been distributed to all clinical directors and a copy is available on the Trust Intranet.

References

Department of Health – Immunisation against Infectious Disease 2013 – "The Green Book" – updated version available online:

https://www.gov.uk/government/uploads/system/uploads/attachment_data/file/309218/Green_Book_Chapter_7_v1_3.pdf