Spleen Australia

Asplenia/Hyposplenism – Paediatric Guidelines Age 0 to 18 years *(17/04/2023 v3)*

Disclaimer:

These guidelines have been produced to guide clinical decision making for the medical, nursing and allied health staff of specialist paediatric services and general practitioners. They are not strict protocols, and **they do not replace the judgement of a senior clinician**. Clinical common sense should be applied at all times. These clinical guidelines should never be relied on as a substitute for proper assessment with respect to the particular circumstances of each case and the needs of each patient. Clinicians should also consider the local skill level available and their local area policies before following any guideline.

Background

- Individuals with a non-functioning spleen are at increased risk of infection with encapsulated bacteria, most importantly *Streptococcus pneumoniae* (pneumococcus), the cause of invasive pneumococcal disease (IPD) which includes: meningitis (brain infection); septicaemia (blood infection) and pneumonia (chest infection).
- Infections with other bacteria, such as *Neisseria meningitidis* (meningococcus) and *Haemophilus influenzae* type b (Hib), *Capnocytophaga canimorsus* (after dog or cat bite) also occur at an increased rate in patients with asplenia/hyposplenism.
- The first two years following splenectomy are considered the highest risk for infection, although several reports indicate that the risk is lifelong.
- Children with congenital asplenia, cancer related asplenia/hyposplenism, haematological conditions requiring splenectomy and those with sickle cell anaemia are at greater risk of infection than those who have had a splenectomy for trauma. Some of these infections are vaccine preventable diseases and protection can be optimised by appropriate and timely immunisation.
- Immunisation status should be reviewed by GP or specialist whenever there is a change in immunisation recommendations
  e.g. new vaccine; or at least every 5 years. Yearly influenza vaccination is recommended.
- Individual case discussions should be undertaken with the treating specialist.
- Please note that meningococcal, pneumococcal, *Haemophilus influenzae* type b and influenza vaccines may be co-administered.
- More information regarding asplenia/hyposplenism or assistance with immunising your patients can be obtained from Spleen Australia www.spleen.org.au - T (03) 9076 3828 or if in Queensland 1800 SPLEEN (775336).

This guideline is written by paediatric infection specialists, vaccinology experts, immunisation nurses, paediatric pharmacists and Spleen Australia staff.
March 2023 *reviewed by Nigel Crawford [1,3], Rachael McGuire[2,3], Penelope Jones[4]*
[1] Immunisation Service, Department of General Medicine, MCRI, Melbourne
[2] SAEFVIC, Murdoch Children's Research Institute (MCRI), Melbourne
[3] The Melbourne Vaccine Education Centre (MVEC), Royal Children's Hospital
[4] Spleen Australia, A Clinical Service and Registry for people without a functioning spleen, Alfred Health, Melbourne

This guideline has been reviewed and is permitted for use in Queensland by Children's Health Queensland Infection Management and Prevention Service and Queensland Specialist Immunisation Service, and approved by the Western Australian Department of Health.

Additional vaccines and medical advice for people <18 years who have asplenia or hyposplenism

Table 1: Pneumococcal vaccination

Brand	**Prevenar 13®** *(Pneumococcal Conjugate Vaccine 13vPCV)*	**Pneumovax 23®** *(Pneumococcal Polysaccharide Vaccine 23vPPV)*
Age at diagnosis	**Dose**	**Dose**
≥ 6 weeks to ≤ 11 months	3 doses (min. 8 weeks apart) + 1 booster[¥β§]	Single dose at ≥ 4 years[^]
≥ 12 months	Single dose at diagnosis[Ω^β§]	Single dose at ≥ 4 years[^]
Additional booster doses		
	Not recommended	5 years following completion of initial dose[Σ]

¥ Doses as per NIP at 6 weeks, 4 months, 6 months (additional dose for those with a special risk condition) and routinely at 12 months of age.

β Booster doses are given at ≥ 12 months of age/8 weeks since previous dose (whichever is later).

§ If not up to date with NIP recommendations refer to https://immunisationhandbook.health.gov.au/catch-up-vaccination for catch up advice.

^ Ideally Prevenar 13® is administered first, followed by Pneumovax 23® at ≥ 4 years of age/minimum of 8 weeks later (whichever is later). If Pneumovax23® is inadvertently administered first, a minimum of 12 months should elapse before administering Prevenar 13®.

Ω To be given a minimum of 8 weeks following any previous doses.

Σ Maximum of 2 doses of Pneumovax 23® in a lifetime.

Table 2: Meningococcal vaccination

Brand	**Nimenrix®** *(Meningococcal Quadrivalent Conjugate Vaccine*	**Bexsero®**[β€] *(Meningococcal B Recombinant Multicomponent Vaccine MenBV)*
Age at diagnosis	**Dose**	**Dose**
≥ 6-weeks to ≤ 5 months	3 doses (min. 8 weeks apart) + 1 booster[¥¶]	3 doses (min. 8 weeks apart) + 1 booster[β¶§]
≥ 6-months to ≤ 11 months	2 doses (min. 8 weeks apart) + 1 booster[¥¶†]	2 doses (min. 8 weeks apart) + 1 booster[β¶§]
≥ 12 months	2 doses (min. 8 weeks apart)[¥]	2 doses (min. 8 weeks apart)[β§]
Additional booster doses		
≤ 6 years of age when primary course completed	Give a booster 3 years after completing of the primary course. Further booster doses every 5 years[†]	Give a single booster dose only 3 years after completing the primary course[§]
≥ 7 years of age when primary course completed	Give a booster dose every 5 years following the completion of the primary course[†]	Give a single booster dose only 5 years after completing the primary course[§]

¶ Booster doses are given at ≥ 12 months of age/8 weeks since previous dose (whichever is later)

A single dose of Nimenrix® is funded on the NIP at 12 months and for year 10 students and adolescents aged 15-19 years who missed receiving the vaccine at school

¥ Administration of prophylactic paracetamol is recommended for those < 4 years of age (15mg/kg per dose) 30 minutes prior to vaccination (or as soon as possible after), as well as 2 subsequent doses (4-6 hours apart) to reduce the likelihood and severity of fever

† Menveo® may be used interchangeably as an alternate brand to Nimenrix®. Where possible completing a course with the same brand is preferred

§ Bexsero® is registered for use from 6 weeks of age. Trumenba® (not funded by NIP) is an alternate meningococcal B vaccine available as a 3 dose course for individuals' with asplenia/hyposplenism **aged 10 years or older**. Meningococcal B vaccines are **not** interchangeable.
https://immunisationhandbook.health.gov.au/vaccine-preventable-diseases/meningococcal-disease

Additional vaccines and medical advice for people <18 years who have asplenia or hyposplenism

Table 3: *Haemophilus influenzae* type b (Hib) vaccination	
Brand	**Infanrix hexa®/ActHIB®**
Age at diagnosis	**Dose**
≥ 6-weeks	As per NIP^§
> 5 years of age	One dose

^ If not up to date with NIP refer to https://immunisationhandbook.health.gov.au/catch-up-vaccination for catch up recommendations.

§Additional booster doses not required.

Table 4: Influenza vaccination	
Brand	**Variable**- refer to http://www.mvec.vic.edu.au/immunisation-references/influenza-vaccine-recommendations/ for age-appropriate brands and details
Age at diagnosis	**Dose**
< 6-months	Not recommended
≥ 6-months	Recommended annually£€

£ Immunisation of family members recommended

€ 2 doses, given 4 weeks apart is recommended for those < 9 years of age in the 1st year of receiving the vaccine

all Aboriginal & Torres Strait Islander people ≥ 6 months of age

Table 5. COVID-19 vaccination
Further booster doses may be required for children with asplenia/hyposplenism and or additional risk factors for COVID-19. Refer to government websites.

Additional vaccines and medical advice for people <18 years who have asplenia or hyposplenism

Table 5. Antibiotics

	Recommendation	Duration
Antibiotic prophylaxis (Daily antibiotics)	Amoxicillin 20 mg/kg (up to 250 mg) orally, daily OR Phenoxymethylpenicillin 250 mg (child younger than 1 year: 62.5 mg to 5 years: 125 mg) orally, 12-hourly. If hypersensitivity to penicillins is reported: Assess whether the patient has immune-mediated hypersensitivity. Refer to the Australian Antibiotic Therapeutic guidelines (circa 2018) "Antimicrobial hypersensitivity – Types of antimicrobial hypersensitivity" and seek infectious disease specialist advice on testing and/or alternative prophylaxis options.	Required duration of antibiotic prophylaxis is difficult to determine – a reasonable recommendation for prophylaxis is up to 16 years of age. Minimum recommended duration is: • Up to the age of 5 years in children with asplenia • Up to the age of 5 years who are hyposplenic due to sickle cell anaemia or another congenital haemoglobinopathy • At least 3 years after splenectomy Consider lifelong prophylaxis for patients who: • are severely immunosuppressed (e.g. those with coexisting hypogammaglobinemia or advanced liver disease, solid organ transplant recipients) • have had a splenectomy for haematological malignancy, particularly those with graft-versus-host diseases (GVHD) or receiving ongoing immunosuppressive therapy. • have survived overwhelming post splenectomy infection, particularly invasive pneumococcal infection.
Emergency supply of antibiotics	Amoxicillin+clavulanate 22.5mg/kg/dose (amoxicillin component) (max 875mg/dose), orally, twice daily (use DUO preparation) For infants (over 3 months of age) and children with immediate non- severe or delayed non severe hypersensitivity to beta-lactams: Cefuroxime 15mg/kg/dose (up to 500mg/dose) orally 12-hourly For infants and children with immediate severe or delayed severe hypersensitivity to penicillins, seek Infectious Disease Specialist advice.	All patients should also have an emergency supply of antibiotics available at home or when away. An antibiotic course should be commenced if there is sudden onset of unexplained fever and immediate medical review is not available. Once the first dose is taken the patient must be reviewed by a medical practitioner.

Additional vaccines and medical advice for people <18 years who have asplenia or hyposplenism

Table 3.	Additional information – For Children with Asplenia/Hyposplenism aged 0 - 18 years of age
Antibiotics (refer to Table 2)	DAILY DOSE The required duration of antibiotic prophylaxis is difficult to determine and should be discussed with the treating physician and/or an infectious diseases physician (see Table 2). EMERGENCY SUPPLY If immediate access to medical care is not available (eg. whilst holidaying, patients living in remote areas or out of hours), a supply of antibiotics should be available for immediate use should symptoms of bacterial infection occur such as fever, rigors and/or malaise. Take this supply of antibiotics and see doctor as soon as possible either GP or emergency department in a hospital. (See Table 1 for dosing)
Timing of immunisations	Elective splenectomy- In persons undergoing an elective splenectomy, vaccination should be completed where possible, TWO weeks before the scheduled operation. In some cases, vaccination may need to commence 10-12 weeks before splenectomy to allow the administration of both conjugate and polysaccharide doses of the vaccines to maximise protection. Emergency splenectomy, or when vaccinations are not administered pre-operatively, patients should be immunised more than ONE week after surgery.
Multiple immunisations	It is safe to administer all required vaccines at the same time. However giving the vaccines over a few days might be better tolerated.
Chemotherapy	Patients who are receiving immunosuppressive chemotherapy and/or radiotherapy and require additional vaccines due to asplenia/hyposplenism should receive immunisation as per the attached table and then receive additional post-chemotherapy booster vaccines as recommended in the AIH. (see Australian Immunisation Handbook http://immunise.health.gov.au/internet/immunise/publishing.nsf/Content/Handbook10-home). Antibiotic chemoprophylaxis should be continued throughout this period.
Overseas travel	Seek specialist advice about travel vaccines needed for specific destinations. Patients should be aware of the increased risk of severe *Plasmodium falciparum* malaria and Babesiosis. It is recommended to adhere to strategies to avoid mosquito bites (e.g. applying insect repellent, wearing long sleeved shirts from dusk until dawn) as well as taking appropriate anti-malarial prophylaxis.
Animal bites	Dog or cat bites/severe scratches can result in bacterial infections with *Pasteurella sp*., *Capnocytophaga canimorsus, Streptococcus sp. and* anaerobic bacteria and therefore require prophylactic antibiotic treatment with Augmentin (Amoxycillin/clavulanate) or if beta-lactam hypersensitive - metronidazole plus Bactrim (trimethoprim/sulfamethoxazole).
Tick bites	Babesiosis is a rare tick-borne infection endemic to certain countries (including North and South America, Europe, Asia and Africa) and patients with asplenia or hyposplenism travelling to these areas should take precautions to avoid tick bites.
Other bacterial pathogens	Other organisms have been reported to cause severe infections in patients with asplenia including *Salmonella* species, *Staphylococcus aureus*, *Escherichia coli, Campylobacter* species, *Bacteroides* species, *Pseudomonas* species and *Plesiomonas shigelloides.*
Other procedures	Dental procedures or general surgery – additional antimicrobial prophylaxis is not required for people with asplenia/hyposplenism
Pregnancy	Underlying risk for overwhelming sepsis is not increased, but immunisation status should be clarified as per Tables above
Education	Parents and patient should be educated regarding the: • Need for early investigation and management of any febrile illness • Signs of bacterial infection and when to give emergency supply of antibiotics (after checking expiry of supply) • Recommendation to wear or carry a medi-alert bracelet or alert card to inform health professionals of risk of overwhelming infection • Requirement to update their vaccination cards when booster doses of vaccines are given. These cards are available from Spleen Australia.
Spleen Australia	Registration with Spleen Australia - Clinical Service & registry for people with non-functioning spleens, based in Melbourne at the Alfred Hospital, is recommended for people residing in Victoria, Tasmania, Queensland or Western Australia or if you have had treatment for a asplenia/hyposplenism in these states. This clinical service offers telephone advice, reminders and an education kit (with booklet, medical alerts, fridge magnets etc). www.spleen.org.au or call 03 9076 3828 or if reside in Queensland call 1800 SPLEEN (775336). "Spleen-ie App" is also available and it records vaccines plus additional health tips. Registered patients and their GPs receive regular updates that contains medical updates and the latest information on staying healthy.