NHS SCOTLAND

SPAH
SCOTTISH PAEDIATRIC AND ADULT HAEMOGLOBINOPATHIES NETWORK

Paediatric guideline

Vaccination guideline

NOTE
This guideline is not intended to be construed or to serve as a standard of care. Standards of care are determined based on all clinical data available for an individual case and are subject to change as scientific knowledge and technology advance and patterns of care evolve. Adherence to guideline recommendations will not ensure a successful outcome in every case, nor should they be construed as including all proper methods of care or excluding other acceptable methods of care aimed at the same results. The ultimate judgement must be made by the appropriate healthcare professional(s) responsible for clinical decisions regarding a particular clinical procedure or treatment plan. This judgement should only be arrived at following discussion of the options with the patient, covering the diagnostic and treatment choices available. It is advised, however, that significant departures from the national guideline or any local guidelines derived from it should be fully documented in the patient's case notes at the time the relevant decision is taken.

This guidance has been prepared by NHS National Services Scotland (NSS) National Networks. Accountable to Scottish Government, NSS works at the heart of the health service providing national strategic services to the rest of NHS Scotland and other public sector organisations to help them deliver their services more efficiently and effectively. Working across professional and organisational boundaries, National Networks support the delivery of safe, effective healthcare that's designed around patients, carers and families.

Contents

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Stakeholders involved **Others consulted**	Consultant Paediatric Haematologists, Consultant Paediatricians, Haematology Nurse Specialists, Consultant Paediatric Radiologist L Jones, Paediatric Infectious Diseases and Immunology Consultant, K Templeton, Consultant Virologist
Methodology used	Review of The Green Book of Immunisation Reference to recent BSH guideline on Prevention and treatment of infection in patients with an absent or hypofunctional spleen:
Rationale	The network was established to support and develop haemoglobinopathy services throughout Scotland in improving standards of clinical care for patients. The network supports the delivery of evidence based, patient centred care through the development and implementation of clinical guidelines, care pathways and information resources utilising a once for Scotland approach.
Scope	Haematology specialists and general paediatrician including hospital staff managing children with haematological conditions in NHS Scotland
Approval process	The guideline was approved by the SPAH Paediatric Guideline and Protocol subgroup on 20 May 2025. This subgroup has the authority to develop, review and endorse guidelines. This has been agreed by the SPAH Steering Group and noted in its Terms of Reference. See appendix for list of Paediatric Guideline and Protocol subgroup members

Background

Patients with sickle cell disease (SCD) are at increased risk of infection due to multiple factors as outlined below:

- functional asplenia or hyposplenia, regardless of spleen size, age or types of sickle disease i.e. Hb SS, HbSC and Hb SB[1]
- Neutrophil function is also abnormal, although the exact mechanism for this is not clear it contributes to the increased risk of infection from streptococcus pneumonia, staphylococcus aureus and salmonella Neisseria

As a result, prevention of infection with both vaccination and prophylactic antibiotics is important.

Routine vaccination for children with sickle cell disease is recommended and should be given in accordance with the complete routine immunisation schedule - GOV.UK

Children and adults with sickle cell disease should receive all live vaccines according to national recommendation even while on hydroxycarbamide. [1,2]

Children that arrive in the UK with an incomplete vaccination record should also follow the guidance in the document, vaccination of individuals with uncertain or incomplete immunisation - GOV.UK

Specific protection in line with hyposplenia can be found in Immunisation of individuals with underlying medical conditions: the green book, chapter 7 - GOV.UK

Children with SCD should receive the following additional vaccinations to the routine vaccination schedule:

Box 7.1 Practical schedule for immunising individuals with asplenia, splenic dysfunction or complement disorders*

Note: Since these vaccines do not protect against all strains, antibiotic prophylaxis should also be strongly considered

First diagnosed or presenting 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 two priming doses of PCV13 with an 8-week interval in their first year
- a booster dose of MenACWY conjugate vaccine 8 weeks after the vaccinations scheduled at one year of age
- an additional booster dose of PCV13, to be administered at least 8 weeks after the routine PCV13 booster scheduled at 1 year of age, and
- one dose of PPV23 after the second birthday[¥] and at least 8 weeks after the last dose of PCV13

First diagnosed or presenting at 1 year to under 2 years of age

If not yet administered, give the routine vaccines due at 1 year of age: Hib/MenC, PCV13, MMR and MenB vaccines, plus:

- one dose of MenACWY conjugate vaccine at least 8 weeks after the vaccines scheduled at 1 year of age
- an additional booster dose of PCV13, to be administered at least 8 weeks after the routine PCV13 booster scheduled at 1 year of age, and
- one dose of PPV23[¥] after the second birthday

First diagnosed or presenting 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 MenACWY conjugate vaccine and
- one dose of PPV23[¥]
- If they have not received the routine 2+1 schedule for MenB, ensure they have received two doses of MenB 8 weeks apart since first birthday
- If they have not received any PCV previously, they should receive a dose of this first followed by the dose of PPV23 at least 8 weeks later

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
- an additional MenB vaccine dose 4 weeks later

All patients aged over 6 months

Annual influenza vaccine each season (see Chapter 19)

* Patients on complement inhibitor therapy (Eculizumab or Soliris®) are not at increased risk of pneumococcal disease and do not require PPV23 or additional doses of PCV13 (see Chapter 25).

¥ Patients with asplenia and splenic dysfunction should receive boosters of PPV23 at five yearly intervals.

Note: only one dose of MenACWY being recommended for children between 6 to 12 months

Prevenar 13, pneumococcal conjugate vaccine (PCV)

- in under 1y an additional PCV dose to be given at least 8 weeks after the 1st dose at 12 weeks.
- 1-2y, a second booster dose to be given 8 weeks after the dose given at 1year of age
- as this is a conjugate vaccine it will have produce a better immunogenic response and therefore all attempts should be made to give at least one dose prior to the Pneumovax vaccination

Pneumovax, 23 valent Polysaccharide pneumococcal vaccine (PPV)

- first dose at 2 years of age and 5 yearly thereafter
- should consider administering at least one dose of PCV 13 or 15 vaccine, 8 weeks before PPV23, if the patient has not received a PCV vaccine before
- there is evidence of a rise of infections causing invasive pneumococcal disease that are not prevented by vaccination and emphasis must remain on children continuing to take regular penicillin in addition to immunisations[3]
- Penicillin prophylaxis should be given for a minimum of 5 years from birth

Meningitis ACWY vaccine

- the standard UK vaccine guidance recommends this additional meningitis vaccine in all young people at around 14years of age
- in sickle cell disease, due to hyposplenism, this should be administered in the first year of life or at diagnosis
- one dose should be given between 6 and 12 months of age (different to the Green Book)
- a dose should also be given at 8 weeks after routine 1y vaccinations
- children diagnosed at 2y or older should receive one dose of meningitis ACWY vaccine
- Men ACWY should also be offered to anyone travelling to parts of the world where these serotypes are prevalent, e.g. sub-Saharan Africa and Saudi Arabia. This should be given at least 2 weeks before travel if a dose has not been given in the last 12 months. Check other at-risk areas for travel. Meningococcal Disease | CDC Yellow Book 2024. Also check on CDC website where patient is travelling, for other vaccinations that may be required

Meningitis B Vaccine (4CMenB, Bexsero)

- universal Meningitis B vaccine is now administered at 8 weeks and 16 weeks of age with a booster dose at 1year of age (2+1)
- for new diagnosis or first presentation and under 2 years of age, follow the UK guideline.
- if aged between 2-10 years of age at presentation and they have not received the routine 2+1, they should receive 2 doses of Meningitis B vaccine 4-8 weeks apart
- for new diagnosis or first presentation aged ten years onwards, administer two doses of Men B 4 weeks apart

Haemophilus influenzae type b vaccine

- additional booster vaccination against (Hib) is no longer recommended as current control of Hib is excellent
- one dose of Hib could be administered to patients with Sickle cell disease if they have never received Hib vaccine before, but the Hib/Men C vaccine will soon not be available

Influenza vaccine

- annually for all children from 6 months of age
- children aged 6 months to less than 2 years should be offered a suitable quadrivalent inactivated influenza vaccine
- older children should be offered the live attenuated influenza vaccine unless medically contraindicated or parental choice (as porcine based)
- children under 9 years of age who have not previously been vaccinated against influenza should be offered two doses of the influenza vaccine given at least 4 weeks apart

Hepatitis B vaccine

- recommended for all children with HbSS and HbSβ, to ensure that all children requiring blood transfusion, whether as an elective or emergency procedure are protected
- Hepatitis B vaccination has been part of the universal paediatric vaccination programme since Autumn 2017
- children born in the UK prior to this time or overseas may not have received this. Perform serological testing for Hepatitis B surface antibody. If the patient in non-immune, arrange vaccination via the local Vaccination centre
- Engerix B dosing stated below (**alternative brands also available - be aware that dosing may vary**), see BNF-C for specific dosing information.

Injection	Dose
1st dose	0.5 mL IM
2nd, 1 month after the first dose	0.5 mL IM
3rd, 6 months after the first dose	0.5 mL IM

- if they have not been routinely vaccinated in the UK, then antibodies to Hepatitis B surface antigen (anti-HBs) should be checked to ensure an adequate response of >10mIU/ml
- if there is any level detected below 10mIU/ml then there should be a full 3 dose vaccination course performed, after checking for other HBV markers to check they are not HBsAG or Core positive [4]

COVID vaccine

- children with sickle cell disease and no other risk factors have not been deemed at increased risk of severe COVID infection
- Government guidance will be updated seasonally and should be sought with regards to the recommendation of COVID vaccines for children

BCG vaccination

- BCG is recommended, preferably at birth, based on tuberculosis risk which is assessed by health visitor
- if this is not administered at birth, follow the national/RCPCH guidelines

Malaria Prophylaxis

- the formulation advised depends on area to be visited. Consult a travel clinic (based in GP practice or community pharmacy) or refer to the BNF link Malaria, prophylaxis | Treatment summaries | BNF | NICE
- this should be provided by GP/Hospital pharmacy
- check patient's G6PD status, as certain preparations are contraindicated in deficient patients. Guidelines for malaria prevention in travellers from the UK 2023
- patients going to live in malarial areas should be advised to stay on prophylaxis lifelong if possible
- general advice re preventing bites – mosquito nets, clothing, and repellents should be given

Yellow Fever[6]

- dose should be given at least 10 days before travel to an endemic area to allow protective immunity but can still be considered for last minute travellers Yellow Fever Vaccine & Malaria Prevention Information, by Country | CDC Yellow Book 2024
- also ensure patients and families are counselled about the importance of insect bite precautions
- must be given at least 28 days apart from the MMR vaccine
- consider a second re-immunising dose if they meet the criteria base on green book and the last dose was greater than 10 years ago [5]

HPV vaccine

- recent update in green book, July 2023, advising only one dose required for adolescent and young adults under 25 years of age
- for patient on Hydroxycarbamide a second dose can be considered but one dose would also be considered sufficient

References

1 Prevention and treatment of infection in patients with an absent or hypofunctional spleen: A British Society for Haematology guideline, Ladhani et al, British Journal of Haematology, February 2024
2 The Complete routine Immunisation Schedule, UKHSA, Sep 2023
3 Immunologic Effects of Hydroxyurea in Sickle Cell Anaemia, Lederman et al, Pediatrics. 2014 Oct;134(4):686-95.
4 Are booster immunisations needed for lifelong hepatitis B immunity? European Consensus Group on Hepatitis B immunity, THE LANCET • Vol 355 • February 12, 20005 The Green Book: Information for public health professionals on immunisation, UK Health Security Agency.
6 Tolerance and humoral immune response to the yellow fever vaccine in sickle cell disease children treated with hydroxyurea: a multicentre prospective study, Koehl et al, J Travel Med. 2021 Apr 14;28

Useful resources and links

Useful Resources and Links

Sickle Cell Disease in Childhood: Standard and Recommendations for Clinical Care, 3rd Edition- November 2019
EVIDENCE-BASED MANAGEMENT OF SICKLE CELL DISEASE: EXPERT PANEL REPORT, 2014. US department of Health and Human Services.
Sickle cell protocol, Haematology and Oncology Unit, RHC, Glasgow.
Guidelines for the Management of Sickle Cell disease in Paediatrics, West London Haemoglobinopathy Coordinating Centre, October 2023

Complete routine immunisation schedule from 1 September 2024 - GOV.UK

Screening of individuals with uncertain or incomplete screening status - GOV.UK

UK and international immunisation schedules comparison tool - GOV.UK

Appendix - Guidelines and Protocols subgroup membership

Name:		Job Title:	Organisation:
Katrina	Adams	Non-Malignant Haematology Nurse Specialist	NHS GG&C
Ruth	Allen	Consultant Paediatric Radiologist	NHS GG&C
Susan	Baird	Consultant Paediatric Haematologist	NHS Lothian
Nadia	Catherwood	Paediatric Haematology Nurse Specialist	NHS GG&C
Shahzya	Chaudhury	Consultant Paediatric Haematologist	NHS GG&C
Sarah	Clarke	Non-Malignant Paediatric Haematology Consultant	NHS GG&C
Emma	Cockburn	Consultant Paediatrician	NHS Tayside
Cheryl	Hill	Non-Malignant Haematology Nurse Specialist	NHS GG&C
Kirsten	Husselbee	Consultant Paediatrician	NHS Tayside
Jennifer	Milne	Paediatric Haematology Nurse	NHS Tayside
Lucy	Paterson	Clinical Nurse Specialist - Benign Haematology	NHS Lothian
Fernando	Pinto	Consultant Paediatric Haematologist	NHS GG&C
Gordon	Taylor	Consultant Paediatric Haematologist	NHS Grampian
Rosina	Shujaat	Consultant Paediatric Haematologist	NHS Lothian