NHS SCOTLAND

SPAH
SCOTTISH PAEDIATRIC AND ADULT HAEMOGLOBINOPATHIES NETWORK

Sickle Cell Disease (SCD)
Information for GPs: Patient management of a young person (over the age of 12) in Primary Care

This document has been prepared by NHS National Services Scotland (NSS) on behalf of SPAH. 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. The SPAH Network is a collaboration of stakeholders involved in care of patients with haemoglobinopathies, who are supported by an NSS Programme Team to drive improvement across the care pathway.

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This guide is a basic resource for GPs caring for young people with Sickle Cell Disease (SCD).

Medications

1) What medications should I regularly provide for my patient with SCD?

The following medications are recommended for a young person with SCD:

- **Folic Acid**
  Folic Acid supplementation aids production of new red blood cells, which is increased in SCD.

  Dosage: by mouth
  12 years and over 5mgs once daily

- **Phenoxymethylpenicillin V** (or Erythromycin for Penicillin allergic patients). Patients are more prone to infections like pneumococcal infection due to an absent or poorly functioning spleen.

  Penicillin dosages: by mouth
  12 years and over 250mgs twice daily
  OR
  Erythromycin dosages: by mouth
  12 years and over 500mgs twice daily

  Ref: BNFC (British National Formulary for Children) | NICE

- **Vitamin D**
  Many people in the UK have low vitamin D levels without showing any symptoms, especially among non-white groups. To help prevent conditions like rickets, it's recommended they take vitamin D supplements.

  Dosage: 12 years and over 400-800 units daily (varies as per local guidelines and BNFc)
  Ref: Vitamins | Treatment summaries | BNFC | NICE

- **Hydroxycarbamide**
  Many people with SCD take hydroxycarbamide to help reduce complications. The hospital will prescribe and provide this medication. Patients on hydroxycarbamide are at risk of cytopenia and this should be considered during their assessment.

2) My patient wants painkillers, including opiates, on repeat prescription. Is this, ok?

SCD can cause episodes called painful crises. These episodes can be unpredictable and severe. Taking painkillers early during an episode can help avoid hospital assessment or admission.

Make sure your patient has paracetamol and NSAIDs (usually Ibuprofen) by repeat prescription. See link below for appropriate dosages.
Ref: BNFC (British National Formulary for Children) | NICE

If your patient's pain has lasted for several days despite regular analgesia, please contact the haematology team **urgently** (contact details below). Your patient might need to be admitted to the hospital for treatment with strong opiates given under medical supervision. **We do not expect you to prescribe in the practice setting.**

3) Are there any medications that could trigger a sickle cell crisis?

No. Common triggers of sickle cell crises include dehydration, infection, hypoxia, stress, and extremes of temperature.

Vaccinations

4) What vaccinations should I give my patients SCD?

Patients are more prone to infections like pneumococcal infection due to an absent or poorly functioning spleen. To reduce this risk, give these vaccines in accordance with the "Green Book".

Patients should receive vaccinations in accordance with the UK routine childhood vaccination programme.

They should also receive the Influenza vaccination from six months then annually.

Note: If your patient has moved to the UK or has presented late, it's crucial to get their previous immunisation history. If it's unclear, refer them for "catch-up" vaccinations in addition to those required for their sickle cell disease.

Ref: Vaccination of individuals with uncertain or incomplete immunisation - GOV.UK

Patients should receive additional vaccinations from the list below. This will be organised by the Secondary Care team in hospital or via the immunisation service.
PPV23 (Pneumovax): First dose at 2 years then due every 5 years
ACWY Meningitis: 1 or 2 doses dependent on age at diagnosis or presentation

Ref: The Green book of immunisation - chapter 7 - Immunisation of immunocompromised individuals

5) What if my patient wants to travel abroad?

For foreign travel, your patient should get the standard recommended vaccinations relevant to their destination. Recommend a travel clinic if necessary.

Patients with SCD are vulnerable to malaria. Make sure they take appropriate malaria prophylaxis if travelling to a high-risk malaria region. Yellow Fever vaccination may also be required.

Details of paediatric drugs and dosages:
Malaria, prophylaxis | Treatment summaries | BNFC | NICE
UK Health Security Agency – Guidelines for Malaria Prevention in travellers from the UK (See Chapter 4, Page 39)

For known G6PD deficient patients:
Advice on safe antimalarials is available at the BNFC.org

Anaemias | Treatment summaries | BNFC | NICE

Managing infection

6) What should I do if my patient has signs or symptoms of illness (for example infection or pain)?

If the patient comes to the surgery, please assess them.

Pain: If they are systemically well with no signs of pyrexia, dehydration, or severe sickle cell crisis pain, they can be treated in the community without needing haematology input. In this case please advise:

- rest
- increased fluid intake
- regular analgesia (Paracetamol and Ibuprofen) for 24 hours.

If you're concerned or if their condition doesn't improve after 24 hours or gets worse, please contact the haematology team.

Also, advise the patient or arrange for them to go to the haematology unit, emergency department or local paediatric assessment unit for further evaluation.

Infection and other symptoms

- pyrexia (temp >38 degrees)
- unable to maintain their fluid intake
- develops chest pain or severe crisis pains
- any respiratory symptoms or signs, for example, cough or increased work of breathing

Please contact the haematology team **urgently** and advise them or arrange for them to attend the haematology unit, emergency department or local paediatric assessment unit for urgent assessment.

Pregnancy planning

7) What methods of contraception can I safely provide for my patients with SCD?

In accordance with UK Medical Eligibility Criteria (MEC) for contraception guidance 2016 https://fsrh.org/standards-and-guidance/external/ukmec-2016-digital-version/

There is no restriction (UK MEC 1) on:
- Subdermal implant (Nexplanon)
- IM / SC Depo-Provera
- Levonorgestrel-releasing intrauterine system (Mirena IUS)
- Progesterone only pills

The advantages generally outweigh risks (UK MEC 2) for:
- combined oral contraceptive pill
- combined transdermal patches
- combined vaginal ring
- copper IUS

8) My patient with SCD wants to have a baby. What advice should I give them?

Advise your patient to contact their haematology team for further discussion.

The haematology team may need to stop certain medications, such as ACEi or hydroxycarbamide. These are not safe in pregnancy or prior to conception, even in male patients. They will also need to assess your patient's overall health to access their fitness for pregnancy. They will discuss partner haemoglobinopathy screening to assess the likelihood of them having a child who also has a significant haemoglobin disorder.

Further Resources

Further resources on sickle cell disease:

Sickle Cell Society - Supporting People Affected by Sickle Cell Disorder

Inheritance of Sickle Cell Anaemia » Sickle Cell Society

Day to Day care to people with SCD » Sickle Cell Society

Sickle Cell Work and Employment » Sickle Cell Society

Sickle cell disease - Treatment - NHS

Further resources on sickle cell trait can be found at the link below:

Sickle cell disease - Carriers - NHS

Contact details

	Name	Contact number
Consultant Haematologist or Paediatrician		
Clinical Nurse Specialist		
Secretary		
Routine Enquiries		
Emergencies		

If you need an alternative format, contact nss.equalitydiversity@nhs.scot. or Scotland BSL contactscotland-bsl.org for British Sign Language.

Additional information can be found on the Scottish Paediatric and Adult Haemoglobinopathies Network (SPAH) website. spah.scot.nhs.uk