SPAH
SCOTTISH PAEDIATRIC AND ADULT HAEMOGLOBINOPATHIES NETWORK

NHS SCOTLAND

Sickle Cell Disease (SCD) Information for GPs: Adult Patient Management in Primary Care

This guide is a basic resource for GPs caring for adult patients with sickle cell disease.

Medications

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

The following medications are recommended for adult patients with sickle cell disease:

- Folic Acid 5mg - Folic Acid supplementation aids production of new red blood cells, which is increased in SCD.
- Penicillin V 250mg bd or Erythromycin 250mg bd for Penicillin allergic patients - patients are more prone to infections like pneumococcal infection due to an absent or poorly functioning spleen. If patients don't want long term prophylaxis, please tell them to seek early medical attention if they develop symptoms of infection.

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 available (unless they have a history of GI ulceration or AKI/ CKD). You can offer weaker opiates like codeine or dihydrocodeine if they aren't already taking stronger ones like tramadol, oxycodone or oramorph.

If your patient requires opiates for sickle cell pain that their Haematology Consultant hasn't recommended, or if their pain persists for several days despite regular pain relief, contact their haematology team (details below).

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 with 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".

https://assets.publishing.service.gov.uk/government/uploads/system/uploads/attachment_data/file/566853/Green_Book_Chapter7.pdf

Additionally, they should receive hepatitis B vaccination because they may need blood products in their lifetime.

All adults with SCD should receive:

Vaccine	Frequency
Pneumococcal PPV23	Every 5 years
Influenza immunisation	Yearly
Covid immunisation	Currently yearly, though check green book as may change with viral prevalence
Primary Immunisation Hepatitis B (if not received in childhood – routine after 2017)	3 doses in total at 0, 1 and 2 months Booster 12 months

Note: If your patient has moved to the UK as an adult, 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.

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.

Make sure they take appropriate malaria prophylaxis if they are travelling to a high-risk malaria region.
For known G6PD deficient patients:

- Atovaquone-proguanil, doxycycline, mefloquine or proguanil prophylaxis are safe.
- No need to withhold chloroquine prophylaxis from those known to be G6PD-deficient.
- Primaquine is contraindicated in patients with G6PD deficiency.

https://assets.publishing.service.gov.uk/government/uploads/system/uploads/attachment_data/file/774781/ACMP_guidelines_2018.pdf

A travel guide for patients is available on the SPAH website.

Patients with sickle cell disease should be strongly encouraged to get travel insurance to cover them if they become unwell abroad. The Sickle Cell Society website has a list of insurance brokers that other patients have found helpful.

Travel insurance for sickle cell » Sickle Cell Society

Pregnancy Planning

6) What contraception methods are safe for my patients with sickle cell disease?

In accordance with UK Medical Eligibility Criteria (MEC) for contraception guidance 2016
(https://www.fsrh.org/standards-and-guidance/uk-medical-eligibility-criteria-for-contraceptive-use-ukmec/),

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

7) My patient with sickle cell disease 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 assess their fitness for pregnancy. They will discuss partner haemoglobinopathy screening to assess the likelihood of them having a child with a significant haemoglobin disorder.

Managing Infection

8) My patient has signs or symptoms of infection. What should I do?

Remember that your patient is at a higher risk of infection due to hyposplenism.

- See and assess your patient
- Treat them following local microbiology guidelines
- Tell them to stay hydrated when they are unwell

If they're systemically unwell, can't stay hydrated, have chest pain, or severe crisis pains or are hypoxic, **urgently** contact the haematology team**.** They will need assessment in secondary care.

You can find more resources on sickle cell disease at the links below:

- Sickle Cell Society Information for Health Professionals
- Sickle Cell Society - Inheritance of Sickle Cell Anaemia
- Sickle Cell Society - Day-to-Day Care
- Sickle Cell Society - Employment
- NHS Information on SCD Treatment

For information on sickle cell trait, see:

- Sickle cell disease - Carriers - NHS (www.nhs.uk)

Haematology Department Contact Details:

Consultant Haematologist (Name):	
Secretary (Name):	
Telephone No. (Routine enquiries):	
Telephone No. (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