Further resources on sickle cell disease can be found at the links below:

https://www.sicklecellsociety.org/resource/information-health-professionals/

https://www.sicklecellsociety.org/resource/inheritance-sickle-cell-anaemia/

https://www.sicklecellsociety.org/resource/day-day-care-people-scd/

https://www.sicklecellsociety.org/resource/employment/

https://www.nhs.uk/conditions/sickle-cell-disease/treatment/

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

https://assets.publishing.service.gov.uk/government/uploads/system/uploads/attachment_data/file/701031/SCT28_Hb_AS_carrier_leaflet_180418_web.pdf

Contact Details

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

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SPAH
SCOTTISH PAEDIATRIC AND ADULT HAEMOGLOBINOPATHIES NETWORK

NHS SCOTLAND

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

This guide is intended as a basic resource for GPs of adult patients with sickle cell disease.

Medications

1) Are there any medications that I should be providing routinely for my patients with SCD?

It has been standard practice to recommend the following medications routinely 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, such as pneumococcal infection, due to an absent or poorly functioning spleen. For patients not wishing to take long term prophylaxis, please advise them to seek early medical attention if they develop symptoms of infection.)

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

Sickle cell disease is associated with episodes called painful crises. These episodes can be unpredictable and severe. However, if patients take painkillers early during an episode, they may be able to avoid hospital assessment or admission.

Please ensure your patient is advised to have a ready supply of paracetamol and NSAIDs (provided there is no history of GI ulceration or AKI/ CKD). Weak opiates, including codeine/ dihydrocodeine, can be provided if your patient is not already taking stronger opiates, eg tramadol, oxycodone or oramorph.

If your patient is requiring strong opiates for sickle cell pain which is not already recommended by his/ her Haematology Consultant or if your patient's pain has persisted for several days despite regular analgesia, please discuss with the haematology team urgently (contact details below).

3) Are there any commonly prescribed drugs 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 sickle cell disease?

Patients are more prone to infections, such as pneumococcal infection, due to an absent or poorly functioning spleen. This risk can be reduced by administering the following 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

Recommended Vaccinations: (*Please confirm that patient has previously had these vaccinations, especially if new to UK.)

Vaccination	Date Given	Date due (If applicable)
PPV23 [Pneumovax] (due every 5 years)		
Influenza (please send annual reminder)		
*Haemophilus Influenza B		
*Hepatitis B		
* Meningitis C		
*Meningitis B		
* Meningitis ACWY		
*Meningitis C & Haemophilus Influenza B (Menitorix)		
* Prevenar 13		

5) What should I do if my patient is wishing to travel abroad?

Your patient should receive any additional recommended vaccinations for foreign travel. Please advise them to attend a travel clinic, if necessary.

If your patient is travelling to a high risk region for malaria, please ensure they take appropriate malaria prophylaxis.

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

Pregnancy Planning

6) What methods of contraception can I safely provide 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/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 baginal ring
- Copper IUS

7) My patient with sickle cell disease wants to have a baby. What advice should I give them?

Please advise your patient to contact his/ her haematology team to discuss this further (contact details below).

The haematology team may need to stop medications, such as ACEi or hydroxycarbamide, which 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 your patient having a child who also has a significant haemoglobin disorder.

Managing Infection

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

Please remember that your patient is at increased risk of infection due to hyposplenism.

Please see and assess your patient and treat him/ her in accordance with local microbiology policy. Please advise him/ her to stay well hydrated whilst they are unwell.

If he/she is systemically unwell, unable to maintain their fluid intake, develop chest pain or severe crisis pains, please inform the haematology team urgently and advise your patient/ or arrange for him/her to attend ED for urgent assessment.