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): _____

Clinical Nurse Specialist (Name): _____

Secretary (Name): _____

Telephone No. (Routine enquiries): _____

Telephone No. (Emergencies): _____

NSD610-017.27 V1
Developed January 2021
Review January 2024

SPAH
SCOTTISH PAEDIATRIC AND ADULT HAEMOGLOBINOPATHIES NETWORK

NHS SCOTLAND

Sickle Cell Disease Information for GPs: Patient management of Young Person in Primary Care

This guide is intended as a basic resource for GPs of young people with Sickle Cell disease (SCD).

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 the young person with sickle cell disease:

- Folic Acid– Folic acid supplementation aids production of new red blood cells, which is increased in SCD.
  Dosages: by mouth
  12 years and over 5mgs once daily

- Phenoxymethylpenicillin V (or Erythromycin for Penicillin allergic patients). Patients are more prone to infections, such as 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: https://bnfc.nice.org.uk/

- Vitamin D - Asymptomatic Vitamin D deficiency is common in the UK and is prevalent in the non-white population. Therefore supplementation to prevent rickets is recommended.
  Vitamin D dosages: by mouth
  12 years and over 600-800 units daily

  Ref:
  https://bnfc.nice.org.uk/treatment-summary/vitamins.html

2) My patient is asking for painkiller, 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 are given 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 (usually Ibuprofen) by repeat prescription. See link below for appropriate dosages.

Ref: https://bnfc.nice.org.uk/

If your patient's pain has persisted for several days despite regular analgesia, please discuss with the haematology team urgently (contact details below). Your patient may require admission to 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 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".

Patients should receive vaccinations in accordance with the UK Routine Childhood Vaccination programme.

They should also receive the Influenza vaccination from 6 months then annually.

If they are new to the UK / present late, they may require vaccinations in accordance with the "catch up" programme.

Ref:
https://www.gov.uk/government/collections/immunisation-against-infectious-disease-the-green-book

Patients should receive additional vaccinations from the list below. You may be asked to arrange these in the community or they may be delivered in the hospital setting. You will be advised of the same in writing.

PPV23 (Pneumovax) (First dose at 2 years then due every 5 years)

ACWY Meningitis (1 or 2 doses dependent on age at diagnosis / presentation)

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

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.

Patients with Sickle Cell disease are vulnerable to malaria. If your patient is travelling to a high risk region, please ensure they are prescribed appropriate malaria prophylaxis.

Details of paediatric drugs / dosages can be found via the links detailed below.

Ref:
https://bnfc.nice.org.uk/treatment-summary/antimalarials.html

https://assets.publishing.service.gov.uk/government/uploads/system/uploads/attachment_data/file/774781/ACMP_guidelines_2018.pdf (chapter 4 page 39)

Known G6PD deficient patients:

- Atovaquone-proguanil, 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.

Managing Infection

6) My patient has signs or symptoms of illness (i.e. infection of pain) What should I do?

If the patient is brought to the surgery, please see and assess your patient.

Pain

If he / she is systemically well with no signs of pyrexia, dehydration and does not have any severe sickle crisis pains, they can be managed in the community without haematology input. Please advise:

- Rest
- Increased oral fluid intake

- Regular analgesia (Paracetamol and Ibuprofen) for 24 hours

If you have concerns or if their condition persists beyond 24 hours or deteriorates, please inform the haematology team. Please advise your patient / or arrange for him / her to attend Haematology unit / ED for assessment.

Infection / other symptoms

- Pyrexia (temp >38 degrees)
- Unable to maintain their fluid intake
- Develops chest pain or sever crisis pains
- Any respiratory symptoms or signs e.g. cough or increased work of breathing.

Please inform the Haematology team urgently and advise your patient or arrange for him / her to attend Haematology Unit / ED for urgent assessment.

Pregnancy Planning

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

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 vaginal ring
- Copper IUS

8) 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.

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/