SPAH
SCOTTISH PAEDIATRIC AND ADULT HAEMOGLOBINOPATHIES NETWORK

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Sickle Cell Disease Information for GPs: Paediatric patient management in Primary Care

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

- Folic Acid– Folic acid supplementation aids production of new red blood cells, which is increased in Sickle Cell disease.

  Dosages: by mouth
  For child 1 month – 11 years 2.5mgs once daily
  For child 12 years– 17 years 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
  For child 1 – 11 months 62.5mgs twice daily
  For child 1 year - 4 years 125mgs twice daily
  For child 5 years – 17 years 250mgs twice daily

  OR if penicillin allergic or not tolerated

  Erythromycin dosages: by mouth
  For child 1 month – 23 months 125mgs twice daily
  For child 2 years – 7 years 250mgs twice daily
  For child 8 years – 17 years 500mgs twice daily

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

- Vitamin D- Asymptomatic Vitamin Deficiency is common in the UK and is prevalent in the non-white population. Therefore, supplementation to prevent rickets is recommended.

  Vitamin D dosages for maintenance: by mouth
  For child birth – 12 months 400 units daily
  For child 1 year - 12 years 600 units daily
  For child >12 years 600 - 800 units daily

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

2) My patient is asking for painkillers 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 children 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: Immunisation against infectious disease - GOV.UK (www.gov.uk)

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.

Please note Doxycycline is unsuitable for children under 12 years irrespective of their weight.

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

Ref: Malaria, prophylaxis | Treatment summaries | BNFC | NICE
https://bnfc.nice.org.uk/treatment-summaries/malaria-prophylaxis/

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 child 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 severe 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.

Resources

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

https://www.sicklecellsociety.org/
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:
Sickle cell disease - Carriers - NHS (www.nhs.uk)

Contact Details

Consultant Haematologist (Name): _____

Clinical Nurse Specialist (Name): _____

Secretary (Name): _____

Telephone No. (Routine enquiries): _____

Telephone No. (Emergencies): _____

Further Information

Scottish Paediatric and Adult Haemoglobinopathies Network (SPAH) through the website:
spah.scot.nhs.uk

If you require an alternative format, please contact nss.equalitydiversity@nhs.scot, telephone: 0131 275 600 British Sign Language, please contact Scotland
BSL: Contact Scotland (contactscotland-bsl.org)