Primary Care Information on Sickle Cell Disease (SCD)

Risk from infection in people with Sickle Cell Disease

People with sickle cell disease are more at risk from some infections, in particular bacterial infections, as spleen function is poor. Pneumococcus and Salmonella species are especially dangerous but any bacterial infection will affect a person with sickle cell more severely than other people.

**Steps to reduce the infection risk are:**

1. Having available at all times a supply of Penicillin V.

This can be taken EITHER at a dose of 250mg [one tablet] twice daily every day OR not taken every day but the patient should know to start taking a treatment dose – 500mg [2 tablets] FOUR times a day at the first hint of infection, fever, sore throat, cough, shivers etc. Even if they take it at a preventing, lower dose they should increase to this larger treatment dose if they develop symptoms. Penicillin allergic patients should have a supply Erythromycin 500mg bd.

We recommend that, if they are not feeling better within 24 hours, they should be directed to return to their GP or contact the Bristol Haematology & Oncology Centre (BHOC) Same Day Emergency Care (SDEC) Unit Telephone Helpline (0117 342 2011) for further assessment and consideration of broader spectrum antibiotics. If symptoms are not responding to usual antibiotics, please refer to BHOC **urgently via the SDEC Telephone Line** especially if there is any possibility of sepsis.

2. Appropriate vaccination.

Adults with SCD who have not received primary vaccination as part of the national schedule in the UK should be offered:

• Pneumococcal conjugated vaccine [PCV13] - given once.

• Pneumococcal Polysaccharide Vaccine (PPV23) - every 5 years.

• Meningococcal vaccines (Meningococcal ACWY] - given once.

• Meningococcal B – 2 doses 1 month apart.

• Haemophilus Influenza type B/Men C - given once.

• Hepatitis B vaccine if they have not previously received it and are non-immune.

• Influenza – yearly.

• COVID – as available.

3. If there is any suspicion of **food poisoning**, please refer to us for assessment, stool culture etc. Salmonella infection must be treated, even if symptoms are mild, or symptoms have settled but stool culture is positive, as it can become rapidly invasive and very serious.

4. Please advise antimalarial prophylaxis if you are aware she/he is travelling to a malarious area. People often think that if they have sickle cell disease they are protected against malaria – this is far from correct, and malaria can be especially dangerous in these patients.

Managing pain in sickle cell disease

Pain is common, but not universal, in people with HbSS and Sβᴼ thalassaemia. It is less common but still sometimes occurs in those with SC or Sβ⁺ thalassaemia. It can be of varying severity. Many uncomplicated pain episodes can be managed safely at home, taking oral paracetamol and ibuprofen, and plentiful fluid. Patients who have significant pain crises will usually have a supply of stronger analgesia: co-codamol, dihydrocodeine, tramadol and/or oral morphine.

‘Red-flag’ symptoms: significant fever, marked pallor, sleepiness, vomiting/diarrhoea so unable to keep up positive fluid balance, chest pain, breathing problems, any suggestions of limb weakness, anything UNUSUAL other than familiar limb or back pains. If these occur, with or without pain, the person must be assessed here at the hospital, and will often need admission for care of complicated episodes.

Watching for less common complications

Sickle cell can cause a host of complications, the range getting wider as the person gets older. Patients will have a comprehensive ‘annual review’ screening for some of the longer-term problems, as well as managing any current symptoms or problems.

**For male patients:** However, an acute complication to be watchful for from the start is **priapism**. This is a painful penile erection, lasting longer than normal. It can be ‘stuttering’ – coming and going, sometimes a couple of times a night, or can be ‘fulminant’ – an attack which starts and will not spontaneously resolve. If your patient has stuttering priapism, please let us know for an early clinic review. If he has a fulminant attack, he should be directed immediately to the Emergency Department (ED) for possible aspiration.

Sexual Health and Contraception

Progesterone-only contraceptives are associated with no significant adverse effects in women with SCD. Pills/dept or implants may be recommended. The WHO recommends that in the case of the low dose combined oral contraceptive pill (COCP), the risks of pregnancy are outweighed by the potential risks of thrombosis and this form of contraception may be a reasonable option in some women with SCD. An intrauterine contraceptive device (IUCD) can be considered, but in view of the potential risks of infection with IUCDs an IUD (Mirena) is the preferred option. Termination should be avoided, if possible, by contraception, as it carries a high risk of post-operative complications.

You and your team in primary care can help by:

1. Repeat prescribing Penicillin V 250 mg bd for those who take it regularly, and 500 mg qds [give a two week supply] for those who take it only for signs of infection. Please give Erythromycin 500 mg bd for people who are penicillin allergic.
2. Prescribing oral Paracetamol and Ibuprofen when requested, and any other medications including stronger analgesia, as indicated from clinic letters.
3. Remembering that symptoms which may be trivial in others [e.g. sore throat, fever of 38.0°C or higher] may warn of significant bacterial infection in those with sickle cell disease: please give broad spectrum antibiotics early, and refer to us promptly via SDEC if there is any possibility of sepsis.
4. Being aware of the side effects of some of the medications he/she may require in the future: for example ***Hydroxycarbamide*** [given as a disease modifier as it reduces the frequency and severity of pain crises] can cause neutropenia and ***Deferiprone***, sometimes used to reduce iron levels in patients on regular transfusions, can cause agranulocytosis, SO anyone on these medications should be referred immediately to hospital if febrile. ***Deferasirox*** is also used to reduce iron levels in transfused patients, and it can cause indigestion, even upper GI bleeding, rash, and kidney and liver function abnormalities. If people on ***Desferrioxamine*** develop acute abdominal pain we will need to assess them in case of Yersinia bowel infection.

**Please contact us immediately if you have any concerns about anyone on these medications.**

1. Encouraging/giving annual flu vaccine and other vaccinations as needed.

More information needed? Contact our Service directly or please refer to the national sickle cell disease guideline on https://www.sicklecellsociety.org/sicklecellstandards

Contact numbers University Hospitals Bristol & Weston NHS Foundation Trust - Haemoglobinopathy Service:

0117 342 2774 or 07825 948 038 – Nicole Paterson and Prisca Lux, Clinical Nurse Specialists (Monday – Friday, 9am to 5pm, not including bank holidays)

Out of hours:

Same Day Emergency Care (SDEC) Unit:

0117 342 2011 (24/7 including bank holidays)

OR

Haematology Registrar On-Call

0117 923 0000 (switchboard)