**Primary Care Information on Sickle Cell Disease**

**Providing newborn screening results to parents**

All babies in the UK are screened for Sickle cell disease as part of the newborn screening bloodspot test. Any positive results are sent to the Paediatric Haematology team at Bristol Royal Hospital for Children (BRHC) as the coordinating centre for the region. The results will also be sent to the mothers GP. The team at BRHC have responsibility for ensuring these results are provided to the mother/carers within 28 days of the baby’s birth. If the baby is living within the Bristol area the team from BRHC will contact the family directly to inform them of the results. The team will liaise with nominated paediatricians within the rest of the region to inform mother/carers of the babies who don’t live with the Bristol area.

The team at BRHC will inform the baby/mother’s GP and Health visitor when they have informed the parents of the results.

**Outpatient care**

Any baby with a positive newborn screening result will be seen within the sickle cell clinic at BRHC within 90 days of birth. This appointment will be made and sent to the parents/carer by the team at BRHC.

**Risk from infection in people with Sickle Cell Disease**

A major aim of neonatal screening and follow-up care is to reduce the morbidity and mortality from preventable disease by antibiotic prophylaxis and immunisations.

Splenic hypofunction resulting from splenic infarction, usually from the first 6 months of life, means that children are at a greatly increased risk of infection by encapsulated organisms, such as Streptococcus pneumoniae (pneumococcus), Haemophilus influenzae type b and Neisseria meningitidis.

**Steps to reduce the infection risk:**

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

Babies will start on prophylactic Penicillin V within 90 days of birth, initially at a dose of 62.5mg twice daily. This will be usually started at their first sickle cell clinic outpatient review. The dose will increase at 1 year to 125mg twice day and at 5 years to 250mg twice daily. Penicillin allergic patients will receive Erythromycin.

A child with a persistent temperature or any other indications of infection should be reviewed promptly and treated aggressively. If in any doubt advise can be sought from the Paediatric Haematology SpR on bleep 3495 or the Paediatric Haematology CNS team (0117 342 8721). See bottom of document for all contact numbers during office hours as well as evening/weekend hours. If symptoms are not responding to usual antibiotics, please refer to Children’s Emergency Department (CED) for urgent review, especially if there is any possibility of sepsis.

2. Appropriate vaccination.

Routine childhood vaccinations are recommended for all children with sickle cell disease. In addition, according to the Standards and Guidelines for Clinical Care of Sickle Cell Disease in Childhood, 3rd edition November 2019, it is recommended that they have additional vaccinations as follows:

**Pneumovax** (23-valent polysaccharide pneumococcal vaccine) given at age 2 years and every 5 years thereafter during childhood, at least 2 months after PCV 13 (Prevenar 13).

**Influenza vaccination given annually from age 6 months.** For children aged 6 months to 2 years, give intramuscular flu vaccine and for those children aged 2 years to 17 years, Fluenz tetra nasal spray (a live attenuated vaccine).

**MenACWY** is required as per the Green Book for Asplenic children. For a child diagnosed in the first year of life, two doses of MenACWY 1 month apart (in practice this probably means giving two doses of MenACWY in the second year of life).

Further information can be found at Chapter 7 of the green book <https://assets.publishing.service.gov.uk/media/5e18a52940f0b65dc1918763/Greenbook_chapter_7_Immunsing_immunosupressed.pdf>

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.

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 occurs in those with SC or Sβ⁺ thalassaemia. It can be of varying severity. In infants and young children there may be visible swelling of small bones of hands and feet (dactylitis). 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 at home: 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 at BRHC for some of the longer-term problems, as well as managing any current symptoms or problems.

**Splenic sequestration**

Acute splenic sequestration has been defined as an acute fall of Hb and markedly elevated reticulocyte count, together with an acute increase in spleen size. It is a serious complication of SCD, occurring mainly in children < age 5 years and, if unrecognised, carries significant mortality. Mortality rates can be reduced substantially by parental education, regular palpation of the abdomen at home to detect early signs of splenic enlargement and prompt intervention with transfusion. If you have any concerns that a child is having a splenic sequestration, please refer them urgently to the Children's Emergency Department (CED).

**Acute Aplastic Crisis**

Transient red cell aplasia (TRCA) is characterised by a drop in Hb over a period of about a week, often to levels as low as 30 g/L, with a very low reticulocyte count. It may be associated with fever, headache and abdominal pain. In a young child, it may be difficult to differentiate between TRCA and acute splenic sequestration, as the spleen may still be palpable. In contrast to acute splenic sequestration, the reticulocyte count will be very low or absent and IgM/PCR for parvovirus B19 will be positive. It usually takes about 7 days for the reticulocyte to return to normal, and a top-up transfusion is often needed until this happens. If you have any concerns that a child is having a splenic sequestration, please refer them urgently to the Children’s Emergency Department (CED).

**Priapism**

**For male patients:** An acute complication to be watchful for is **priapism**. This is commonest around puberty but can in fact happen in boys of any age. It is a painful, unwanted penile erection, lasting longer than normal erections. 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 inform the Paediatric Haematology CNS team (0117 342 8721) or the Paediatric Benign Haematology SpR on 0788016927 for an early clinic review. **If he has a fulminant attack, he should be directed immediately to the Children’s Emergency Department (CED) for urgent management.**

**Sexual Health and Contraception**

Progestogen-only contraceptives (pills, injections and implants), progestogen-releasing intrauterine systems and barrier methods have no restrictions for use in women with SCD. The advantages of using low-dose combined hormonal contraceptives (pills, patches and rings) and intrauterine devices generally outweigh the theoretical or proven risks in women with SCD. Women should be informed that in the general population the risk of venous thromboembolism with use of combined hormonal contraception is approximately doubled compared to non-users, but that the absolute risk remains low. There is lack of evidence on whether this risk increases further due to their sickle cell disease.

Use of long-acting reversible contraceptive (LARC) methods such as injectables, implants and intrauterine devices are more effective in preventing pregnancies than user-dependant methods such as oral contraceptive pills and barrier methods. Due to the significant health risk associated with pregnancy or termination of pregnancy in women with SCD, women should be advised to consider LARC methods, which are highly reliable and effective.

**You and your team in primary care can help by:**

1. Repeat prescribing Penicillin V (as per clinic letters). Please prescribe either as liquid or tablet as per each child’s preference. **NB** reconstituted Penicillin V only lasts for 7 days, but parents/carers are able to make up new bottles at home, so please ensure adequate amounts are prescribed to prevent the need for numerous repeat prescription requests and interruptions in supply.
2. Repeat prescriptions of Folic acid and Vitamin D supplements as outlined in clinic letters.
3. Prescribing oral Paracetamol and Ibuprofen when requested, and any other medications including stronger analgesia, as indicated from clinic letters. Please prescribe as liquid or tablet as per each child’s preference.
4. 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 CED if there is any possibility of sepsis.
5. Being aware of the side effects of some of the medications he/she may be prescribed by the hospital:   
   ***Hydroxycarbamide,*** given as a disease modifier as it reduces the frequency and severity of pain crises, is prescribed and monitored by the hospital and may cause neutropenia. ***Deferasirox, the most common drug*** used to reduce iron levels in transfused patients can commonly cause abdominal pain, rash, and kidney and liver function abnormalities. These are usually monitored by the hospital but please be aware that patients with iron overload are more susceptible to infection and this may be exacerbated by chelation therapy. This is particularly the case in patients on ***Desferrioxamine;*** any patient on this drug who develops abdominal pain and diarrhoea will need to be assessed for possible 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/wp-content/uploads/2019/11/SCD-in-Childhood_Final-version-1.pdf>

**Contact details**

**Monday – Friday 9.00hrs – 17.00hrs**

Paediatric Benign Haematology CNS team - 0117 342 8721

Paediatric Haematology registrar - 0117 923 0000 (switchboard) Bleep 3495

Paediatric Benign Haematology SpR 0788016927

**Out of hours, weekends and bank holidays**

Paediatric registrar on call for Oncology/Haematology via switchboard – 0117 923 0000