Clinical haematology

About sickle cell disease

Information for patients, relatives and carers

Introduction

The purpose of this leaflet is to provide patients who have sickle cell disease (SCD) with a brief guide to the condition and an overview of the services provided by Imperial College Healthcare NHS trust.

What is SCD?

SCD is a disorder of the body's red blood cells that is genetically inherited and lifelong, meaning that people are born with SCD and it is passed on from parent to child. It is caused by an abnormality in haemoglobin (the part of red blood cells that carries oxygen round the body), which makes the red blood cells become inflexible and sticky. This in turn makes other blood cells as well as the vessels they travel in sticky. Blood flow becomes interrupted and inflammation occurs. This process may cause pain and problems in the organs and may affect the immune system. In SCD, this can happen suddenly and is often painful - these are known as sickle cell crises (sickling). The long-term effects of crises and background sickling (that may have no symptoms) are known as chronic complications. The approach we take to manage these complications is to limit and prevent both of these from happening as much as possible.

How common is SCD?

SCD is the most common inherited condition in the world and mostly affects people whose family origins are in the Middle East, Sub- Saharan Africa, parts of India and parts of the Mediterranean. Between 12,000 and 15,000 people in the UK have SCD and just fewer than 300 babies are born with the condition in this country every year. Parents of all babies born in the UK are given the opportunity to have their baby tested for SCD in the first week of life (the heel prick test).

What kind of problems can happen in SCD?

- Infection
- Gallstones
- Joint damage/destruction
- Loss of spleen function (making patients prone to infections)
- Damage to kidneys
- Leg ulcers
- Visual impairment
- Neurological damage which may lead to stroke

- Erectile dysfunction (priapism)

The signs and symptoms of crisis may include:

- fatigue (feeling tired or weak)
- pain
- jaundice (yellowing of the white of the eyes)
- paleness of skin, or inside the mouth or eyes
- dizziness
- headaches
- symptoms of infection, such as fever

If you develop any of the following symptoms please seek medical attention straight away:

- fever: 38°C or above
- shortness of breath or difficulty in breathing
- feeling very unwell
- abdominal pain
- pain that cannot be controlled with your usual painkillers
- pain which is more severe and/or is different from your usual sickle pain
- new weakness, particularly if felt more on one side of your body than the other
- if you think you are much more anaemic than usual
- priapism a painful erection of the penis lasting more than 30 minutes

Treatment

You may be able to manage a simple painful sickle cell crisis at home by taking painkillers, resting, drinking fluids and keeping warm. However, if the pain becomes worse you may need stronger painkillers in hospital. We can help to control the pain in hospital, and also treat the underlying cause (trigger) and any complications relating to your condition. Sometimes the crisis may not be particularly painful or, more importantly, you may feel unwell from the start. If you are unwell even if you have no pain, you should always contact us as you may need to come to hospital. We may also suggest other ways of managing your condition with preventative treatments. We usually recommend this if the frequency of sickle cell crises increases or if an episode was particularly severe. Treatment options include starting hydroxycarbamide or a long-term blood transfusion programme. Some people have had a bone marrow transplant that has cured their sickle cell disease.

What can trigger a sickle cell crisis?

Common triggers of a sickle cell crisis include: infection, stress, dehydration, cold/hot weather or sudden changes in temperature or nothing at all. Sometimes crises happen out of the blue.

What to do in an emergency

If you have a non-urgent medical problem or a medical problem unrelated to your sickle cell disease, for example a rash or a twisted ankle, you can contact your GP or go to your local Emergency Department (A&E). If necessary, they can discuss your problem with us. If you have a non-urgent red cell/iron problem, you can ask for your outpatient appointment to be brought forward. It may also be appropriate to have a review in day care or Renal and Haematology Triage Unit (RHTU) if you think you need urgent hospital assessment and treatment for your condition. You can call the 24 hours red cell patients number on **020 3311 7755**. .

If you think your condition may be life threatening call 999 and show your 'Patient Access Card' to the ambulance crew. If you attend an Emergency Department (A&E) in another hospital and are admitted, depending on your condition, it may be possible to transfer you to Hammersmith Hospital. Please be aware that although we are always happy to support other teams, we would not be able to manage your care at another hospital.

Note: If you have an infectious illness, such as cough, cold, diarrhoea (runny poo), vomiting or chicken pox, please inform the triage nurse when you contact the service and they will advise you what to do next.

Please do not come to clinic or Haematology Outpatient Department (HOPD or day care) or RHTU if you have any of the above symptoms.

How to manage pain at home

Get to know your body and how sickle cell affects you. This helps you to recognise the early signs that a sickle crisis is starting. Rest, keep warm, take your painkillers, and keep hydrated. Take a warm bath or apply a heat pad to the painful area. You could distract yourself by listening to music, doing puzzles or watch a movie.

Be mindful of the kind of pain you're experiencing. Is it your usual sickle crisis pain or chronic pain or non-sickle pain?

If the pain becomes worse you may need stronger painkillers in hospital. We can help to control the pain in hospital, and also treat the underlying cause (trigger) and any complications relating to your condition.

Medications

Medications you need to take at home are:

- penicillin V or erythromycin prophylaxis antibiotics
- folic acid for development of the red blood cells
- paracetamol, codeine, naproxen/ ibruprofen, e.g. painkillers

Certain medications for people with sickle cell disease need to be given or monitored in the clinic and will be prescribed in clinic. These medications may include stronger opiates (painkillers). This is prescribed on a case by case basis and will be prescribed from one place only (which may be clinic), as part of national guidance.

In addition, you may need to take:

- hydroxycarbamide (Hydroxyurea) to manage frequent pain crisis
- etilefrine for management of priapism
- deferasirox (Exjade®)
- deferiprone (Ferriprox™) for iron overload
- desferrioxamine (Desferal®) an infusion pump inserted under the surface of skin for iron overload

Other medications should be prescribed by your GP.

What can I do to stay well?

- Maintain a good fluid intake by drinking 2 to 3 litres water daily
- Eat healthily
- Exercise
- Make sure infections are treated quickly
- Maintain a good balance of nutrition and activity
- Ensure your vaccinations are up-to-date
- Avoid smoking
- Take penicillin twice a day for life
- It is important to avoid extremes of temperatures, wrapping up and wearing sufficiently warm clothing when it is cold
- Ensure that you attend your clinic appointments so that we can review your health and monitor you for any complications

Attending your clinic appointments is a requirement stipulated in the National Standards produced by the Sickle Cell Society together with the Department of Health. These appointments are every six to twelve months in healthy people but may be more frequent if you have health problems.

If you do not have an appointment or your appointment is inconvenient, please **change it.** We understand that you have busy lives with many commitments and we will do our best to schedule an appointment that is convenient for you. The red cell clinic telephone number is **020 3313 3297.**

Travel advice

Check any necessary travel vaccines needed to the country you are travelling to and arrange these through your GP practice or a travel clinic at least six weeks in advance. If you are visiting an area that requires Yellow Fever vaccination and are currently taking the drug hydroxycarbamide please discuss this with your hospital specialist as it may not be recommended.

For more information please see our leaflet: *Travelling abroad: advice for patients with sickle cell disease and thalassaemia*.

Protect yourself against infection

Make sure you are up to date with routine immunisations:

- pneumovax (given every 5 years)
- haemophilus influenzae type b and meningococcal C
- meningococcal ACWY conjugate
- influenza (given every year)

Carry a card with your vaccine record. This can be provided by your GP practice.

Transfusion and iron chelation

Due to complications of sickle cell disease some patients require regular blood transfusions or red cell exchanges. Blood contains iron. Accumulation of iron in the body is called iron overload.

Iron cannot be removed by the body naturally, resulting in complications affecting the heart, the liver, the endocrine glands (adrenal and pancreas). If allowed to accumulate, it will result in serious complications.

Regular blood tests and scans are done to monitor and detect when the iron levels are raised and require treatment.

Medication is needed to remove the iron from the body. This is called iron chelation therapy. They are used to remove iron from the body to reduce and prevent damage to organs.

There are three types of iron chelators which can be used alone or combined. These are:

1) Deferasirox (Exjade®) taken in tablet form by mouth
2) Deferiprone (Ferriprox™) taken in tablet form by mouth
3) Desferrioxamine (Desferal®) an infusion pump inserted under the surface of skin.

Genetic counselling and testing

Sickle cell disease is a genetically inherited condition. If you are planning to have a baby, you can arrange for genetic counselling and you should also arrange for your partner to be tested if they do not know whether they are a carrier for unusual haemoglobin. If this shows you are both at risk of having a baby affected with SCD, the option of prenatal diagnosis (testing) will be discussed.

If you or your partner becomes pregnant, please tell us immediately if you wish to have prenatal diagnosis.

Obstetric care

It is important that you receive the best care during your pregnancy as early as possible. You and your baby will be followed-up and monitored closely at regular appointments in the joint obstetric/haematology clinic.

What are the risks to me and my baby in pregnancy?

Many women with sickle cell disease have healthy babies and problem-free pregnancies. However, pregnancy may be a physically and emotionally demanding time for the mother, and complications may develop for a variety of reasons, some of which may or may not be related to the SCD. To ensure that you and your baby thrive during the pregnancy, and that any problems are identified early and managed properly, you will be seen by healthcare professionals and have more frequent visits and scans.

Things to be aware of:

- painful crises can be more common during pregnancy and can be brought on by cold weather, dehydration and doing too much physical activity. If you have morning sickness (which can lead to dehydration) or any other concerns, contact your maternity unit as soon as possible
- crises in pregnancy can also lead to worsening anaemia or acute chest syndrome. If you feel particularly tired, short of breath, or you think you are having a crisis, please contact your haematology team and the maternity unit as soon as possible
- pregnant women are generally at an increased risk of developing blood clots in the legs or lungs (venous thrombosis). SCD further raises this risk
- you are also at an increased risk of high blood pressure during the pregnancy. This is known as pre-eclampsia and tends to happen later in pregnancy. To minimise this risk, all women with SCD are offered low dose aspirin as soon as pregnancy is confirmed
- SCD may also affect the growth of your baby because it can affect how your placenta works
- you are more likely than women without SCD to go into labour early or need a caesarean section. If you do not go into labour early, your maternity team may advise you to have your labour started off (induced) at some point before your due date to reduce the risks of becoming unwell to you and your baby. Your obstetrician and midwife will talk to you about your options

National Haemoglobinopathy Registry (NHR)

The NHR is a database of patients with red cell disorders (mainly sickle cell and thalassaemia) living in the UK. This database collects data that is required by the Department of Health from haemoglobinopathy centres. The central aim of the registry is to improve patient care of these conditions. Your red cell team will speak to you about this in detail and offer you an information leaflet.

How do I make a comment about my visit?

We aim to provide the best possible service and staff will be happy to answer any of the questions you may have. If you have any **suggestions** or **comments** about your visit, please either speak to a member of staff or contact the patient advice and liaison service (**PALS**) on **020 3313 0088** (Charing Cross, Hammersmith and Queen Charlotte's & Chelsea hospitals), or **020 3312 7777** (St Mary's and Western Eye hospitals). You can also email PALS at imperial.pals@nhs.net The PALS team will listen to your concerns, suggestions or queries and is often able to help solve problems on your behalf.

Alternatively, you may wish to complain by contacting our complaints department:

Complaints department, fourth floor, Salton House, St Mary's Hospital, Praed Street
London W2 1NY

Email: ICHC-tr.Complaints@nhs.net

Telephone: **020 3312 1337 / 1349**

Alternative formats

This leaflet can be provided on request in large print or easy read, as a sound recording, in Braille or in alternative languages. Please email the communications team: imperial.communications@nhs.net

Wi-fi

Wi-fi is available at our Trust. For more information visit our website: www.imperial.nhs.uk

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