

Living well with sickle cell disease

Information and advice for patients

Sickle Cell and Thalassaemia

What is Sickle Cell Disease (SCD)

Sickle cell disease is a serious and lifelong health condition that affects the red blood cells and the haemoglobin they contain. Healthy red blood cells are round, disc shaped, flexible and can move through small blood vessels without any problems. Haemoglobin is the protein that carries oxygen to all parts of the body. Sickle red blood cells are a different shape (often 'sickle shaped') and cannot change their shape to move through narrow blood vessels. The sickled red blood cells can block small blood vessels and prevent the blood flow, this can happen anywhere in the body.

The main symptoms of sickle cell disease:

- Painful episodes called sickle cell crisis, which can vary from mild to very severe pain and can last up to a week.
- An increased risk of serious infections.
- Anaemia
- Episodes of tummy (abdominal) pain can occur if sickle cells block blood vessels in your abdomen
- Chest pain, high temperature (fever) and shortness of breath or fast breathing when there are blocked blood vessels in the lungs and can sometimes occur with a lung infection. This is acute chest syndrome and is very serious and should be treated urgently in hospital.
- For males of all ages - a persistent, painful erection of the penis called priapism caused by lack of oxygen supply to the area.
- Can cause other problems such as delayed growth, strokes, kidney and lung problems.

It is important for people with sickle cell disease to learn about their condition so they know how to keep themselves well and avoid things that can cause a sickle cell crisis.

The triggers for a crisis include:

- Dehydration
- Not having your recommended medications and vaccinations
- Being too cold or too hot
- Excessive physical exertion/stress

- Emotional stress and anxiety
- Not eating a healthy balanced diet

What can you do to keep well?

Keep hydrated (drink plenty)

You can become dehydrated if you are not drinking enough fluids, especially on a hot day, or if you have diarrhoea and/or vomiting.

To keep hydrated:

- Drink 3-4 litres of fluid per day in the form of juice, squash or water.
- Only have tea and coffee in moderation as they increase the amount of urine you pass.
- Only drink alcohol in moderation as it can cause dehydration.

If you are unable to drink sufficient fluids because you are vomiting, please contact the Sickle Cell and Thalassaemia (SCAT) centre for advice.

Avoid getting too hot or cold

Extremes of temperature can trigger a sickle cell crisis. To avoid this make sure you wrap up warm in cold weather and try to keep cool in hot weather and drink plenty of fluids to make sure you keep hydrated.

Avoid excessive physical stress

It is important that you exercise regularly, but make sure you only exercise within your limits. Excessive exercise can trigger a sickle cell crisis.

Avoid emotional stress and anxiety

If you feel stressed or are finding it difficult to cope with the various challenges of work, relationships, finances etc. please talk to your specialist nurse, doctor, social worker or psychologist about this. It is important that you get the support you need to reduce emotional stress and anxiety.

Eat a healthy diet

Like anyone else, you should eat a healthy, balanced diet with plenty of fresh fruit and vegetables (these contain folic acid among other nutrients). Please do not take any iron supplements without talking to your specialist nurse or doctor first.

Medications

Have your recommended medications and vaccinations

If you have been prescribed medications it is important to take these as directed. You are also advised to take the following:

Folic acid

Folic acid is needed to help your body make red blood cells. A normal balanced diet should

contain adequate folic acid but it is recommended that you take a 5mg supplement of folic acid each day.

Penicillin V

People with sickle cell disease have a spleen that doesn't work properly. The spleen is a gland in the abdomen (tummy) and is part of the immune system which helps the body to fight infections. If your spleen isn't working properly you will be more prone to developing infections, particularly from certain types of bacteria, including pneumococcus bacteria.

To reduce the risk of developing an infection you should take 250mg of Penicillin twice a day (adults only) or 250mg of Erythromycin twice a day if you are allergic to penicillin. You can find further information about these medications, including the possible side effects, in the manufacturer's leaflet that comes with them. It is also important that you know the warning signs of an infection so that you can seek help quickly. Your specialist nurse or doctor will give you more information about these.

Vaccinations

You should have had all the immunisations in the childhood immunisation programme and should continue to have the following booster doses:

- **Pneumovax** - every 5 years to protect against the pneumococcus bacteria.
- **Meningivac** - This gives protection against meningococcus types A and C which cause meningitis.
- **Influenza (flu)** - once a year to protect against the flu virus.
- **Hepatitis B** – this is transmitted through infected blood and sexual contact with an infected person.

You can get these vaccinations from your GP/Practice Nurse.

Disease modifying treatments

Stem Cell or Bone Marrow Transplant

The only cure for sickle cell disease is a stem cell or bone marrow transplant but they are not done very often because of the risks involved and the availability of suitable donors.

Blood transfusions

Some patients with sickle cell disease have top up blood transfusions or red cell exchange transfusions for complications of their sickle cell disease. This treatment is planned with you and your team. A Red blood cell exchange (RBCx) is a potentially lifesaving procedure that removes the sickled red cells and replaces them with healthy red blood cells. This can be needed in an emergency for a complicated sickle cell crisis or can be done as a routine or planned exchange transfusion to prevent sickle cell crisis and its complications. A single exchange transfusion before any major surgery also reduces the risk of complications from the general anaesthetic and surgery.

It is really important to keep to your appointment times for your crossmatch as there are a lot of people involved in getting your blood ready for you and it takes time. We also need you to attend on time for your transfusion.

Hydroxycarbamide and Crizanlizumab are the 2 treatments we currently offer to improve your sickle cell disease. Though they work very differently they both reduce frequency and severity of sickle cell crises. They can also protect your organs (kidneys, liver, heart etc) from the longer-term damage sickling can cause.

If you are on these treatments already, please make sure you understand how they work and how you take them to get the best effect for you.

If you are interested in hearing more about them, please chat to any of the SCAT team.

Painkillers

We have a useful leaflet on managing pain but please make sure you have a supply at home of the pain killers (analgesia) that works for you. It can be useful to keep a copy of your regular medications and your individual pain plan together, so friends and family can help you when you are in pain. It can also be useful to have your key documents together in case you need to come to hospital in an emergency.

Clinic appointments

It is important that you attend your clinic appointment or have your telephone clinic appointment and follow up blood and urine tests. You will be seen by one of the team and keeping you well is the priority, we review your health and medications and monitor you for any complications. If you think you will miss an appointment and need to reschedule please call SCAT. If you change your contact details, have moved out of area or you have not had an appointment with SCAT for some time please also call us.

If you miss appointments this is called a DNA (did not attend), Hospital policy means that after two DNAs we have to discharge you from our service. You can see your Gp and they can refer you back to SCAT. It is essential that your sickle cell disease is reviewed by a specialist service.

Annual Review

Once a year one of the team will complete an annual review at your clinic appointment. This is like a checklist to ensure we have done everything that should be done and checked to keep you healthy. It also gives you the opportunity to ask questions. You will be given a copy of this in your GP letter.

Work Rest and Play

Sickle cell disease should not prevent you from achieving what you wish from the world of work study and leisure.

Further Education:

It is useful to think about where you will have your sickle cell care when you plan to go to University or college. You can talk about this with your SCAT team. Please do not leave it to the last minute, if possible, so we can communicate with your new team if you move area. We run University peer support groups which start every September.

Work:

An employee with a disability such as scd can expect their employer to make reasonable adjustments to ensure that they are not at a disadvantage at work.

You are not obliged to disclose your sickle-cell disease upon job application. However, an employer cannot make reasonable adjustments unless they are informed about your sickle disease. The employer cannot ask about disability as part of the application process. However, an applicant may ask for reasonable adjustments as part of the job application or interview process.

You can have a workers individual support plan drawn up with your employer. The sickle cell society have produced some really helpful information on this <http://sicklecellwork.dmu.ac.uk>.

Time off work may be required to attend routine hospital/doctor appointments for tests, assessments, treatments. Some patients may also require time off work because of unplanned hospital appointments.

Considerations for an employer; As an employer, knowledge of SCD can help you to understand how the employee is managing in the work environment and also what adjustments need to be considered to minimise ill health. For example, this would include avoidance of draughts, being able to decide what tasks can be done or avoided and being able to offer support. Other considerations would be the same as for non-sickle patients such as flexible working. Some individuals may not disclose their illness for fear of stigmatisation, or having their capabilities underestimated.

Advice on benefits

Information regarding benefit entitlements can be obtained, free of charge from a local job centre plus office, social security office or job centre. Leaflets on housing benefit and council tax benefit are available from local councils. www.gov.uk www.gov.uk/benefitsadviser. The Citizens Advice Bureau (CAB) will advise on a range of earnings-related and other benefits which a person may be entitled to, whether they are working or not. This may include housing, welfare and state benefits. They will advise on whether the person is entitled to family income support, invalidity care allowance, Personal independence allowance, family fund and a range of other benefits.

Travel advice

Travelling with sickle cell disorder is not usually a problem but it is important to plan your trip carefully. Before travelling abroad you will need to get a letter from the SCAT centre with the following information:

The type of sickle cell disease and any complications you have. The medications you are taking. Contact details for your SCAT team at home. Ensure you take a sufficient amount of medication on holiday with you as some pain medications may be difficult to get abroad.

There is some more information in the leaflet on 'preventing infections and travel advice'.

Contact details

If you have any questions or concerns please contact the Sickle Cell and Thalassaemia Centre.

Sickle Cell & Thalassaemia Centre

C3, 3rd Floor
Midland Metropolitan University Hospital (MMUH)
Grove Lane
Smethwick
B66 2QT

Telephone: 0121 507 6040

Opening Hours

Monday, 9am – 5pm

Tuesday, 9am – 6pm

Wednesday, 9am – 5pm

Thursday, 9am – 5pm

Friday, 9am – 4pm

Saturday blood transfusion service only

For Further information

You can find more information about sickle cell from the Sickle Cell Society:

www.sicklecellsociety.org (Accessed 23 July 2020).

For more information about health whilst travelling abroad visit the Fit for Travel website:

www.fitfortravel.nhs.uk (Accessed 23 July 2020).

For more information about our hospitals and services please see our websites

www.swbh.nhs.uk, follow us on X @SWBHnhs and like us on Facebook

www.facebook.com/SWBHnhs.

Sources used for the information in this leaflet

Sickle Cell Society (2018) Standards for the clinical care of adults with sickle cell disease in the UK. Available at: <https://www.sicklecellsociety.org/wp-content/uploads/2018/05/Standards-for-the-Clinical-Care-of-Adults-with-Sickle-Cell-in-the-UK-2018.pdf> (Accessed 31 May 2023).

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Incorporating the Midland Metropolitan University Hospital, City Health Campus, Sandwell Health Campus and Rowley Regis Hospital.

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ML6215

Issue Date: January 2024

Review Date: January 2027