

Information about Sickle Cell Disease and Thalassaemia (for Adults)

This leaflet offers more information about the more common red cell disorders sickle cell disease and thalassaemia. If you have any further questions or concerns, please speak to a member of the team using the contact details listed towards the end of this leaflet.

What are sickle cell disease and thalassaemia?

Sickle cell disease and thalassaemia are lifelong, inherited (genetic) disorders which affect the red blood cells. They are sometimes called 'red cell disorders' or 'haemoglobin disorders' as they both affect the haemoglobin, a protein found in red blood cells.

Sickle cell disease (SCD) is a group of conditions which causes the red blood cells to change shape or 'sickle'. This means they can get stuck in small blood vessels and block them. This can happen quite suddenly, causing symptoms including pain which are known as a crisis. Repeated blockages can also lead to complications. For more information, see the leaflet 'Sickle cell disease' ('sickle cell anaemia') on patient.co.uk available on line (<https://patient.info/health/sickle-cell-disease-sickle-cell-anaemia#>) or ask a member of your haematology team for a copy.

SCD is diagnosed by a blood test. It is more common in people whose family origins are African, African-Caribbean, Asian or Mediterranean. In England, Scotland and Wales there is a screening programme to test pregnant women and newborn babies for SCD.

Thalassaemia is a condition that, in its most severe form, causes abnormalities in haemoglobin – the component of blood that carries oxygen in the blood. This can mean there is not enough normal haemoglobin and the red blood cells break down more easily. Those people with more severe thalassaemia often need blood transfusions to provide healthy red blood cells. For more information, see the leaflet 'Thalassaemia' on patient.co.uk available on line or from your haematology team (<https://patient.info/health/thalassaemia-leaflet#>).

Thalassaemia is diagnosed by blood test. It is more common in people whose family origins are Mediterranean or Asian.

What treatments are available?

People can have different forms of SCD or thalassaemia, with varying needs and possible complications. St George's Hospital offers treatment, support and advice for all forms of these conditions, including medical treatment in our outpatient clinics or on the hospital wards. Please see our leaflet 'Sickle Cell and Thalassaemia Service (Adults)' for more detail about our services.

Some people with sickle cell disease and thalassaemia may benefit from blood transfusions ('top ups') or red cell exchanges. This is something that your consultant will discuss with you if it could be appropriate or beneficial for you. You can also see the leaflet 'information for patients with sickle cell disease who may need a blood transfusion' from the NHS Blood and Transplant

service, found at <https://nhsbtdbe.blob.core.windows.net/umbraco-assets-corp/15546/blc7251-sickle-cell-disease.pdf> or from your haematology team for more detail.

For people having regular blood transfusions, the extra blood can lead to a build-up of iron in the body, which can be potentially dangerous. Chelation (iron-removing) treatment can often be needed. Your consultant will discuss this with you. For more information about medication which is used for removing iron, see the leaflets which come with your medicine (the commonly prescribed ones are desferal, deferiprone and exjade) or ask the haematology team for more information.

For those with SCD, a medication called hydroxycarbamide (also called hydroxyurea) can also be helpful for some people. Ask your consultant if this might be appropriate for you. Hydroxycarbamide helps to stimulate the production of foetal (baby) haemoglobin, which does not sickle, and so can reduce the frequency of crises and reduce the risk of longer-term complications for some people. Please see the trust leaflet 'Using Hydroxycarbamide' for more information.

Sickle cell disease/thalassaemia and pregnancy

Most women with sickle cell disease have a straightforward pregnancy and a healthy baby, although some extra care is needed. If you are planning a pregnancy, let your haematology team know before you become pregnant, if possible, so that they can review your medicines and make sure all your checks are up to date. Some medicines, such as hydroxycarbamide, will need to be stopped and you should discuss this with your Haematologist. For more information about pregnancy and sickle cell disease see the leaflet '**Sickle cell disease and pregnancy**' by the Royal College of Obstetricians & Gynaecologists, available from <https://www.rcog.org.uk/globalassets/documents/patients/patient-information-leaflets/pregnancy/pi-sickle-cell-disease-and-pregnancy.pdf> or ask the team for a copy.

Similarly with thalassaemia, it is important to discuss with your Haematologist before getting pregnant, so that they can help you to be in the best possible health, including looking at your iron levels and chelation. For more information about pregnancy and thalassaemia see the leaflet '**Beta thalassaemia and pregnancy**' by the Royal College of Obstetricians & Gynaecologists, available from <https://www.rcog.org.uk/globalassets/documents/patients/patient-information-leaflets/pregnancy/pi-beta-thalassaemia-and-pregnancy.pdf> or ask the team for a copy.

With both sickle cell disease and thalassaemia, it is important to be offered the opportunity to find out if your partner also has the condition or is a carrier of it (sometimes known as having the 'trait'), as this will affect whether any children you have could also have sickle cell disease/thalassaemia. We can refer you to our preconception counsellors for more information and testing.

Managing a sickle cell crisis at home

Many people will experience episodes of pain or feeling unwell that they manage at home. It is generally helpful to take your painkillers as prescribed, keep warm, rest and drink plenty of fluids. Talk to your GP if you have any concerns or call your nurse specialist.

When to seek medical advice

If you are in a lot of pain or feel very unwell, come to our emergency department (A&E) where you will be assessed and given rapid treatment. Indicators for when it can be important to seek medical advice include:

- When you have taken your usual painkillers as prescribed and they are not working
- If you have pain that is worse than usual and getting more severe even with your painkillers
- Severe pain that is different to your usual pain
- A high fever (sweating, chills)
- Difficulty breathing or shortness of breath or chest pains
- Slurred speech
- Any unusual weakness or dizziness or confusion
- Priapism that lasts more than two hours.

Potential difficulties with sickle cell disease

Priapism

Priapism is a long-lasting erection of the penis which is usually very painful. It occurs in 40% of men with sickle cell disease. Priapism occurs because the blood in the penis is trapped and unable to return to the circulation. It is important to let your consultant know if you experience priapism. If you have an episode that lasts for two hours or more, it is important to seek medical advice straight away (you should come to A&E). For more information on priapism, please see the trust leaflet on 'Priapism: information for people with sickle cell disease'.

Chronic pain

Some people with sickle cell disease experience chronic pain, as well as the acute (short-term) pain of a crisis. Chronic, or persistent pain, is pain that has been there for six months or more, and can be felt anywhere in the body. At St George's Hospital we have a specialist red cell pain management service for people with sickle cell and thalassaemia: please ask your consultant for more information or contact the team directly (see the details at the end of this leaflet). You can find more information in our leaflet entitled 'Red Cell Pain Management Service'.

Is there anything I can do to help myself?

For sickle cell disease, your haematologist will make recommendations to help you stay well. Coming to clinic is an important part of this management. These often include taking a preventative antibiotic daily, taking folic acid and ensuring that you stay up to date with vaccinations.

The following vaccinations are recommended for all patients with sickle cell disease and should be administered at your GP surgery:

Pneumococcal vaccine: you should receive this every five years.

Seasonal flu vaccine: you should arrange this with your GP annually.

Hepatitis B vaccine: advised for all patients likely to ever require blood transfusions. Your immunity will be checked and a booster dose may be required.

Meningitis ACWY and B

Haemophilus Influenza type B

You should also avoid smoking and excess alcohol.

Sickling can be triggered by cold, lack of oxygen, dehydration, infection and over-exertion, so it can help to drink plenty of fluids, avoid getting cold, treat infections and fevers quickly and seek medical advice if you feel unwell.

If you are planning on traveling abroad, you should ensure that you take medication to prevent malaria if needed and talk to your consultant about any measures that might be recommended before a long flight. You can read more in the leaflet 'Information on travel for adults with sickle cell disease'.

For patients with thalassaemia, keeping your appointments for clinic and blood transfusions and taking your medicines as prescribed is really important. Any difficulties with this should be discussed with your team to try to find ways to help make this easier. If feeling unwell, particularly if short of breath or suffering from infection (such as fevers) then you should come to the emergency department or speak to your team.

Contact us

If you have any questions about your condition, please discuss with a member of the red cell haematology team at your next appointment or using the details below.

Dr Elizabeth Rhodes (lead sickle cell and thalassaemia consultant)

Tel: 020 8725 0885

Dr Julia Sikorska (sickle cell and thalassaemia consultant)

Tel: 020 8725 0885

Carol Rose (clinical nurse specialist)

Tel: 07500 835735

Red Cell Pain Management and Psychology Service - Dr Jenna Love (clinical psychologist),

Ms Rebecca McLoughlin (specialist physiotherapist)

Tel: 07798 581198, e-mail: SCDpain@stgeorges.nhs.uk

Sickle cell and thalassaemia secretary

Tel: 020 8725 0885

Please see the leaflet '**Sickle Cell and Thalassaemia service (adults)**' for contact details of local support groups and other hospital departments and wards.

For more information leaflets on conditions, procedures, treatments and services offered at our hospitals, please visit www.stgeorges.nhs.uk

Additional services

Patient Advice and Liaison Service (PALS)

PALS can offer you on-the-spot advice and information when you have comments or concerns about our services or the care you have received. You can visit the PALS office between 9.30am and 4.30pm, Monday to Friday in the main corridor between Grosvenor and Lanesborough Wing (near the lift foyer).

Tel: 020 8725 2453 **Email:** pals@stgeorges.nhs.uk

NHS Choices

NHS Choices provides online information and guidance on all aspects of health and healthcare, to help you make decisions about your health.

Web: www.nhs.uk

NHS 111

You can call 111 when you need medical help fast but it's not a 999 emergency. NHS 111 is available 24 hours a day, 365 days a year. Calls are free from landlines and mobile phones.

Tel: 111



Reference: CLH_ISCDT_01 **Published:** August 2019 **Review date:** August 2021