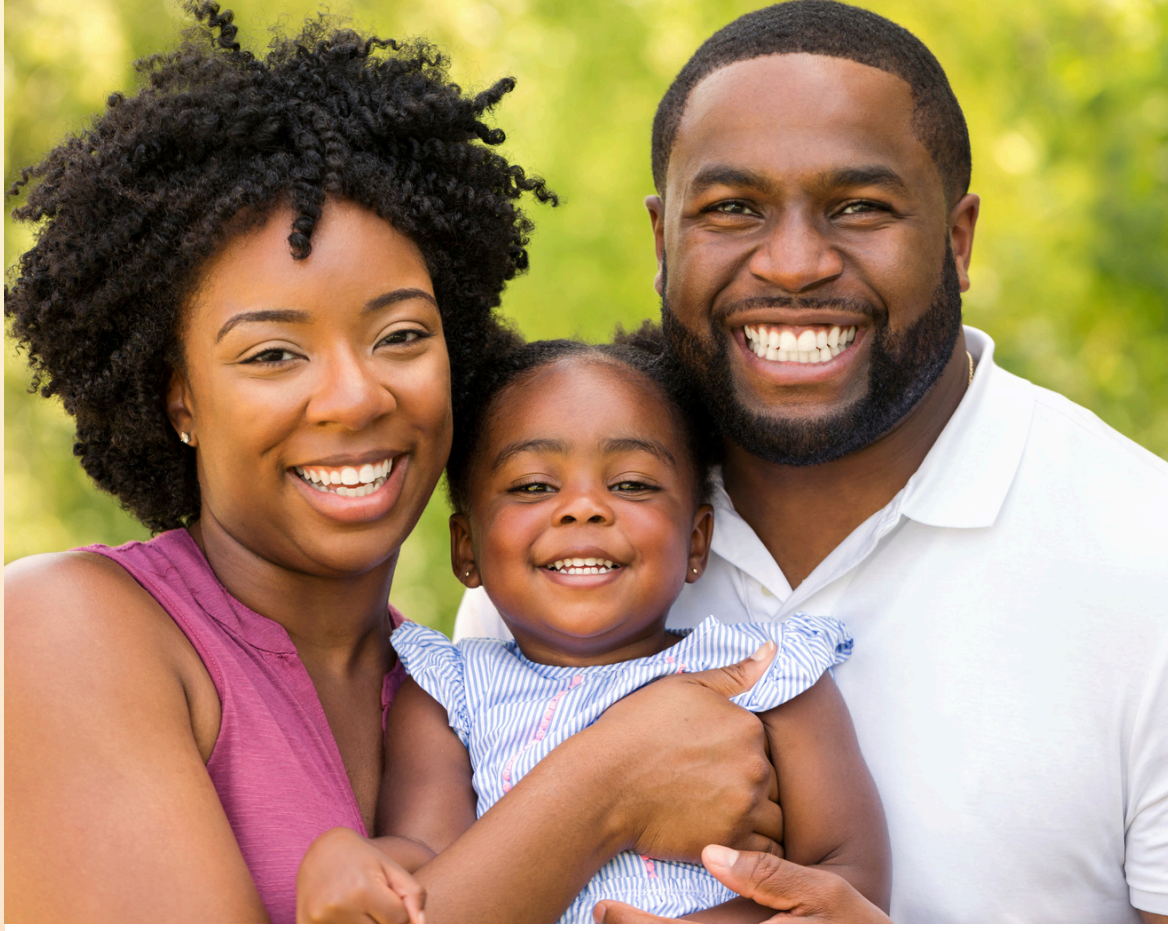
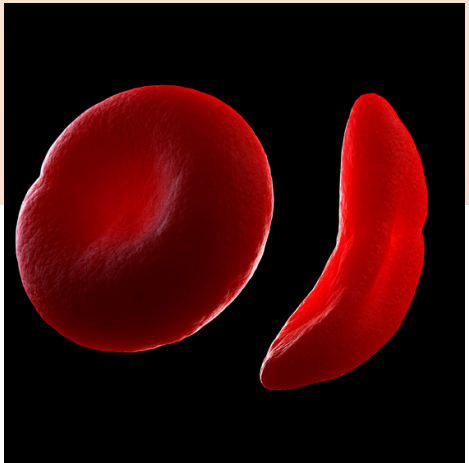
SICKLE CELL SOCIETY

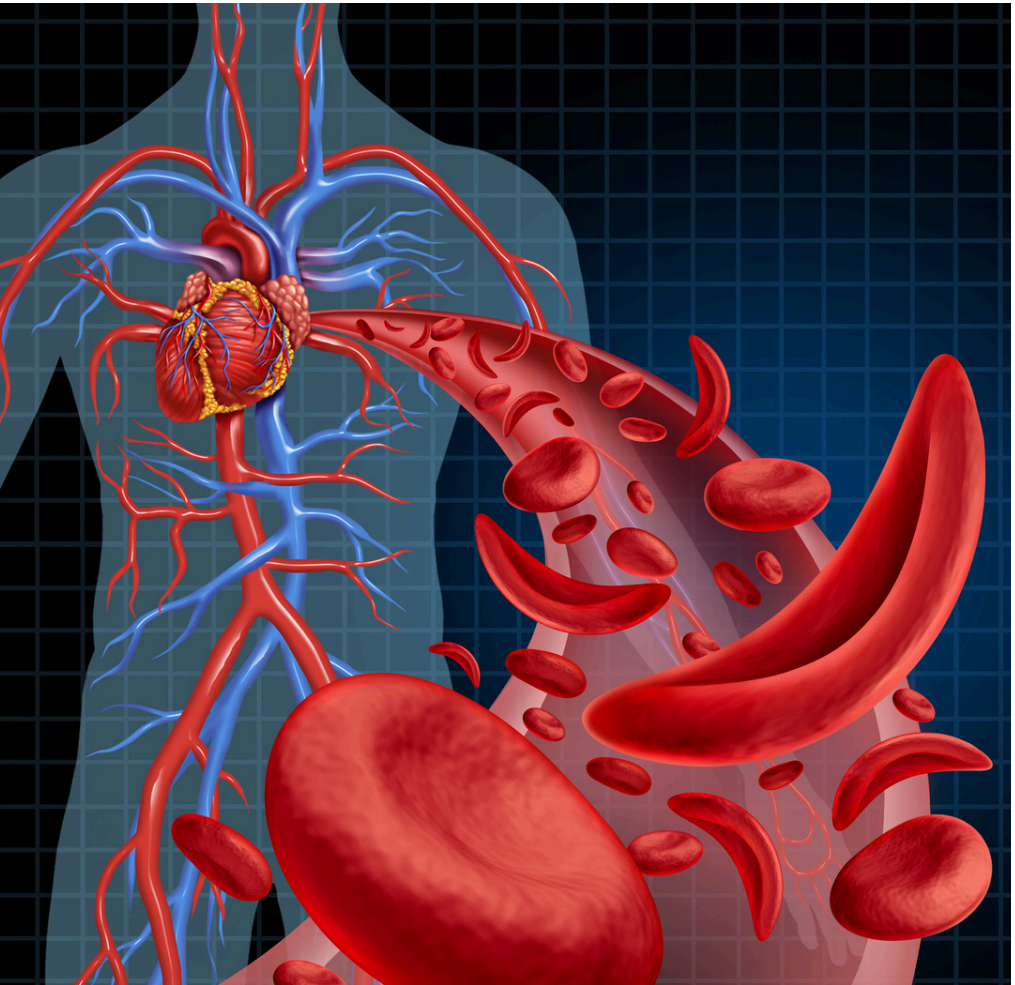
Guide to Sickle Cell Disorder





Sickle Cell Awareness Month
September 2024

About Sickle Cell Disorder



Sickle cell disorder is a genetic condition that affects the red blood cells, which carry oxygen throughout the body.

In people with this disorder, the red blood cells are shaped like sickles or crescent moons, rather than the usual round and flexible shape. They are sticky and don't last as long as normal round red blood cells.

The abnormal shape of the red blood cells can cause them to get stuck in small blood vessels, blocking blood flow and leading to pain and potential organ damage. The painful episodes are known as a 'crisis' and the pain can be severe and result in hospitalisation.

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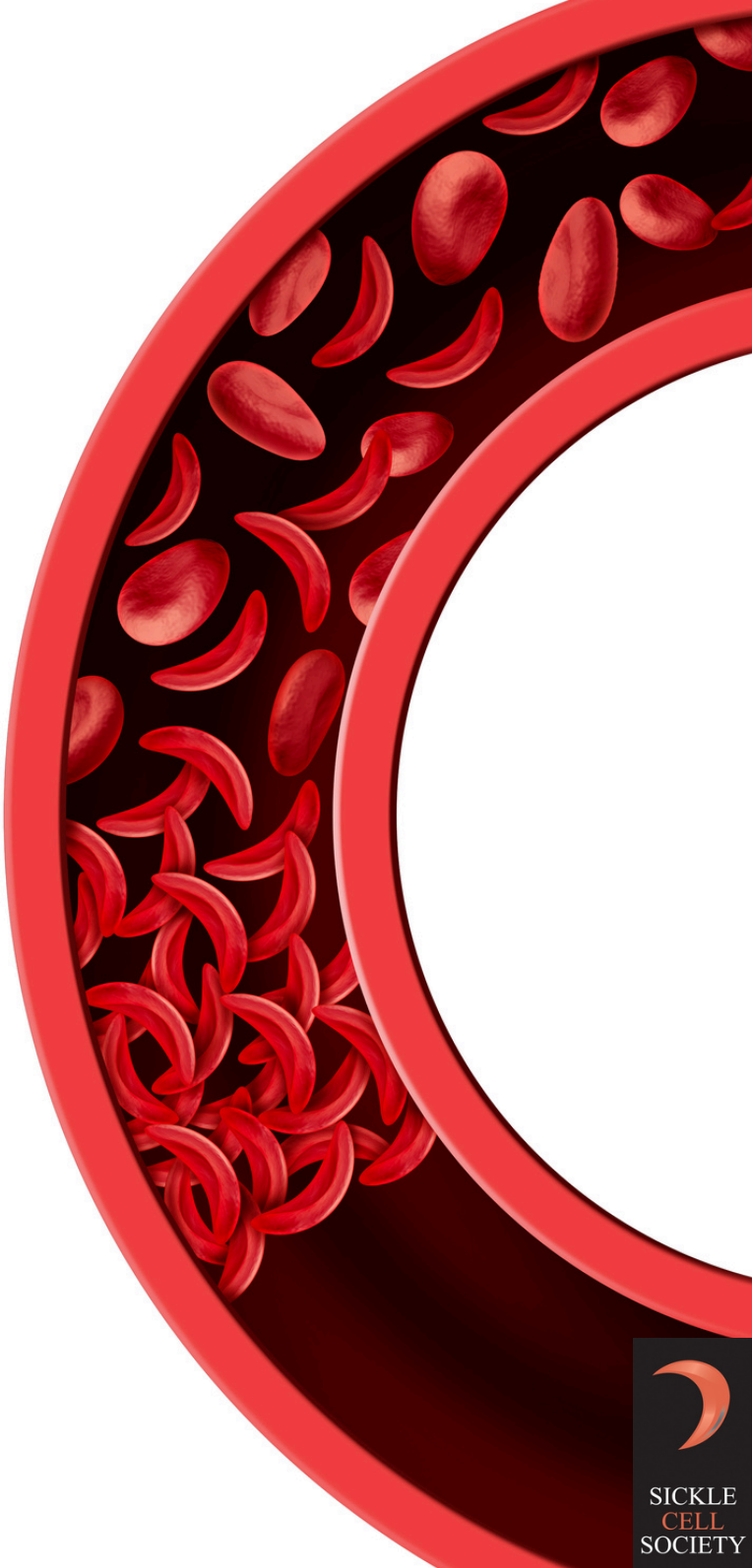
Challenges

The sickle-shaped cells break down more quickly than normal red blood cells, leading to a shortage of red blood cells, known as anaemia.

Living with sickle cell disorder can be challenging. A crisis can occur without warning and is one of the most common and distressing symptoms of sickle cell disorder. It can be fatal.

The pain can be very severe and last for several days or even weeks. A sickle cell crisis can affect any part of the body, but it is most common in the limbs and back. The frequency of pain episodes varies widely among people with sickle cell disorder. Some people may experience them as often as once a week, while others may have fewer than one episode per year.

The exact triggers for these painful episodes are not always clear, but they can sometimes be caused by factors such as weather (wind, rain, or cold), dehydration, stress, or strenuous exercise.



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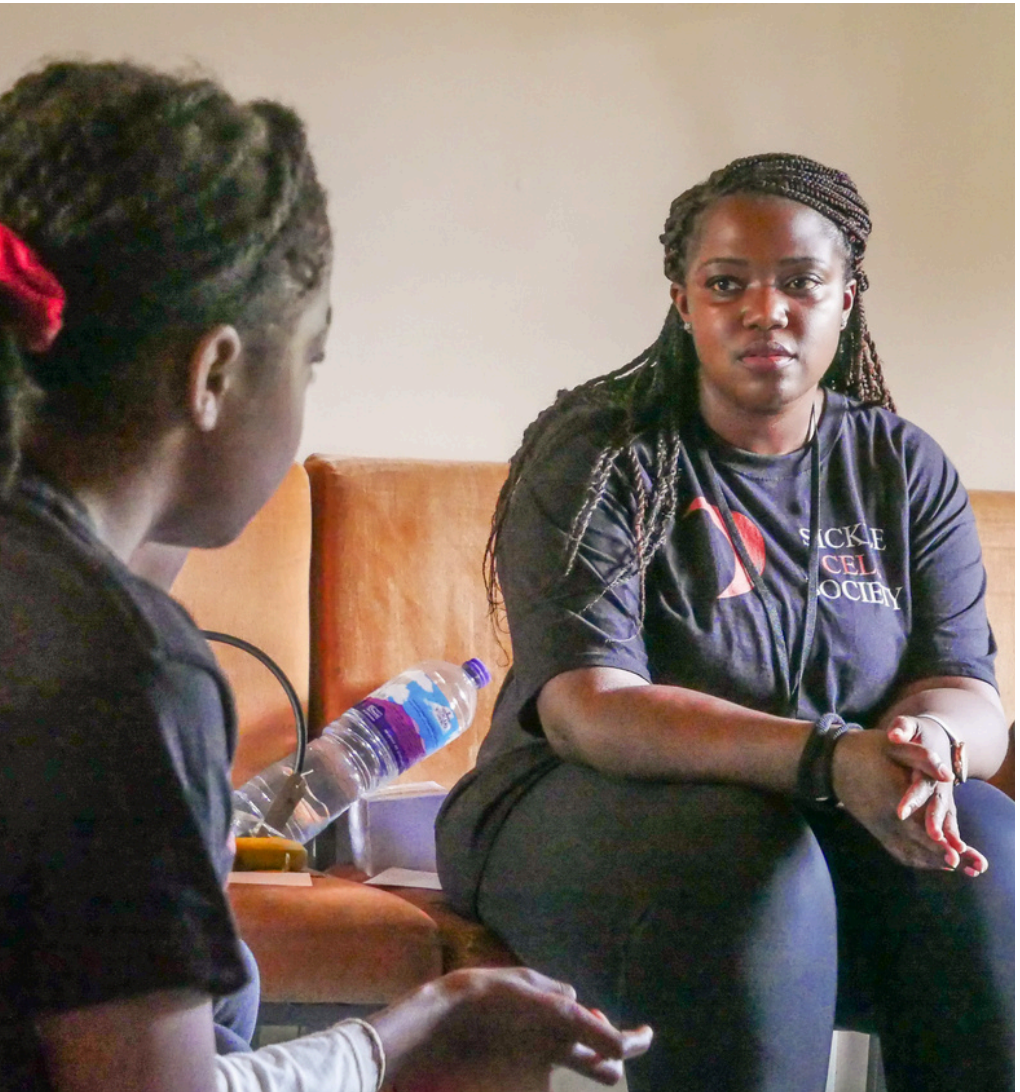
Complications



Frequent pain and medical visits can interrupt and disrupt daily activities, work, and education.

Anaemia can cause severe fatigue, making it hard for people affected to keep up with regular tasks and normal life.

Over time, the disorder can lead to complications such as infections, delayed growth in children, and damage to organs like the spleen, heart, liver, kidneys and eyes.

Other complications include stroke, acute chest syndrome and priapism – a serious condition where there is a prolonged and often painful erection of the penis that lasts for hours beyond or without sexual stimulation. Mental health can also be impacted.

Children with sickle cell disorder are also at increased risk for stroke. The risk is highest between the ages of 2 and 16.

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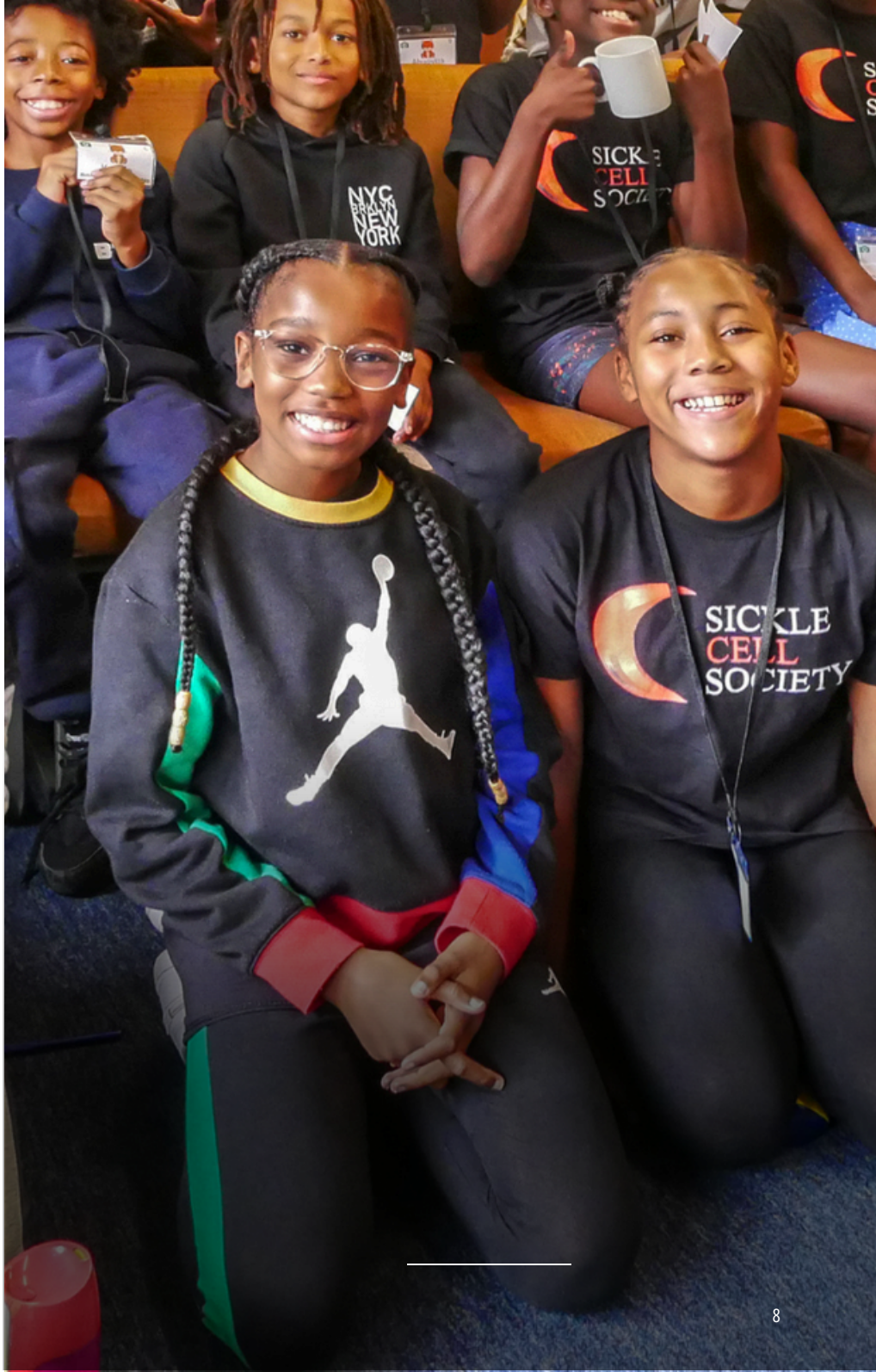
Management

Managing sickle cell disorder requires regular medical care. Treatments include medications to reduce pain and prevent complications, blood transfusions, and sometimes a bone marrow transplant.

People with the disorder need to stay hydrated, avoid extreme temperatures, and be careful about infections. Despite these challenges, many people with sickle cell disorder live full and active lives, especially with the support of family, friends, and healthcare providers.

Advances in treatment and increased awareness are helping to improve the quality of life for those affected by the condition.

Sickle cell disorder can affect anyone, although in the UK it predominantly affects people from African and Caribbean backgrounds. Around 17,500 people in the UK have the condition, and it is the fastest growing genetic condition in the country.



Inheriting Sickle Cell

People with sickle cell disorder are born with it, as it is determined by our genes.

In order to inherit the condition, both parents must have either full sickle cell disorder or sickle cell trait (where parents are carriers and have one sickle cell gene each).

A simple blood test will tell whether you have sickle cell trait or the disorder.

We successfully campaigned for newborn testing for sickle cell disorder, and in 2006 all babies born in England began having a sickle cell test in their first week of life.

Almost 300 babies are born in the UK with sickle cell each year, and approximately 1 in 79 babies born in the UK carry sickle cell trait.

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Treatments

The only possible cure for the disorder is bone marrow transplant – also known as a stem cell transplant. However, this is only possible for a limited number of affected individuals with severe sickle cell disorder, and who have a suitable donor.

Hydroxycarbomide has been the first line treatment in the NHS for many years. People with the condition have been in the position of having very limited treatment options for decades, while other conditions have been the subject of research, development and many new drug releases in that same time period.

In 2024 Voxelotor was approved for use in sickle cell disorder in England and Scotland. We continue to campaign for more safe and effective treatments for people living with the condition, and we encourage people living with the condition to consider taking part in medical trials.

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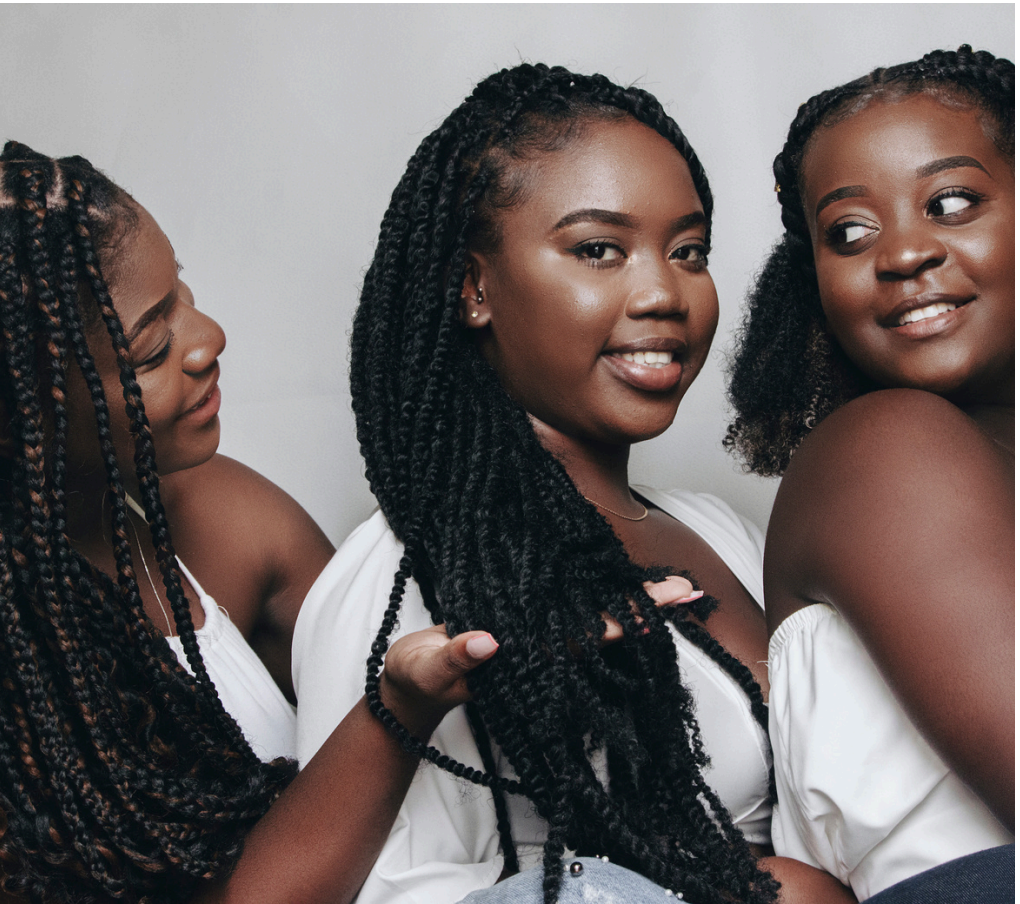
The Sickle Cell Trait



People with sickle cell trait are born with it, inheriting the trait when one parent passes on the sickle gene. It will never develop into full sickle cell disorder. People with sickle cell trait typically do not exhibit symptoms, so we advise blood testing to determine if the trait is present.

In those with sickle cell trait, the majority of red blood cells are normal and round, but some sickle-shaped cells may appear under certain conditions. Sickle cell trait is found in about 1 in 4 West Africans and 1 in 10 Afro-Caribbeans. It is also present in people from the Mediterranean, Asia, and the Middle East. While less common in white Europeans, the growing diversity of populations may change this distribution.

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Most people with sickle cell trait are healthy. However, anaesthetics can cause complications, so it is important to inform dentists or doctors before undergoing any treatment.

Additionally, people with sickle cell trait might experience pain at high altitudes (above 10,000 feet), during long-haul flights in unpressurized planes, or while mountain climbing. It is crucial to disclose having the trait before engaging in such activities, as supplemental oxygen might be needed.

Extreme exercise can also trigger issues, so professional athletes should follow a training program that accounts for this condition.

While sickle cell trait is not an illness, it requires consideration for those planning to have children. If one partner does not have the trait, their children will not have sickle cell disorder but have a 50% chance of inheriting the trait. If both partners have the trait, there is a 25% chance that any child conceived will have sickle cell disorder and a 50% chance the child will inherit the trait.

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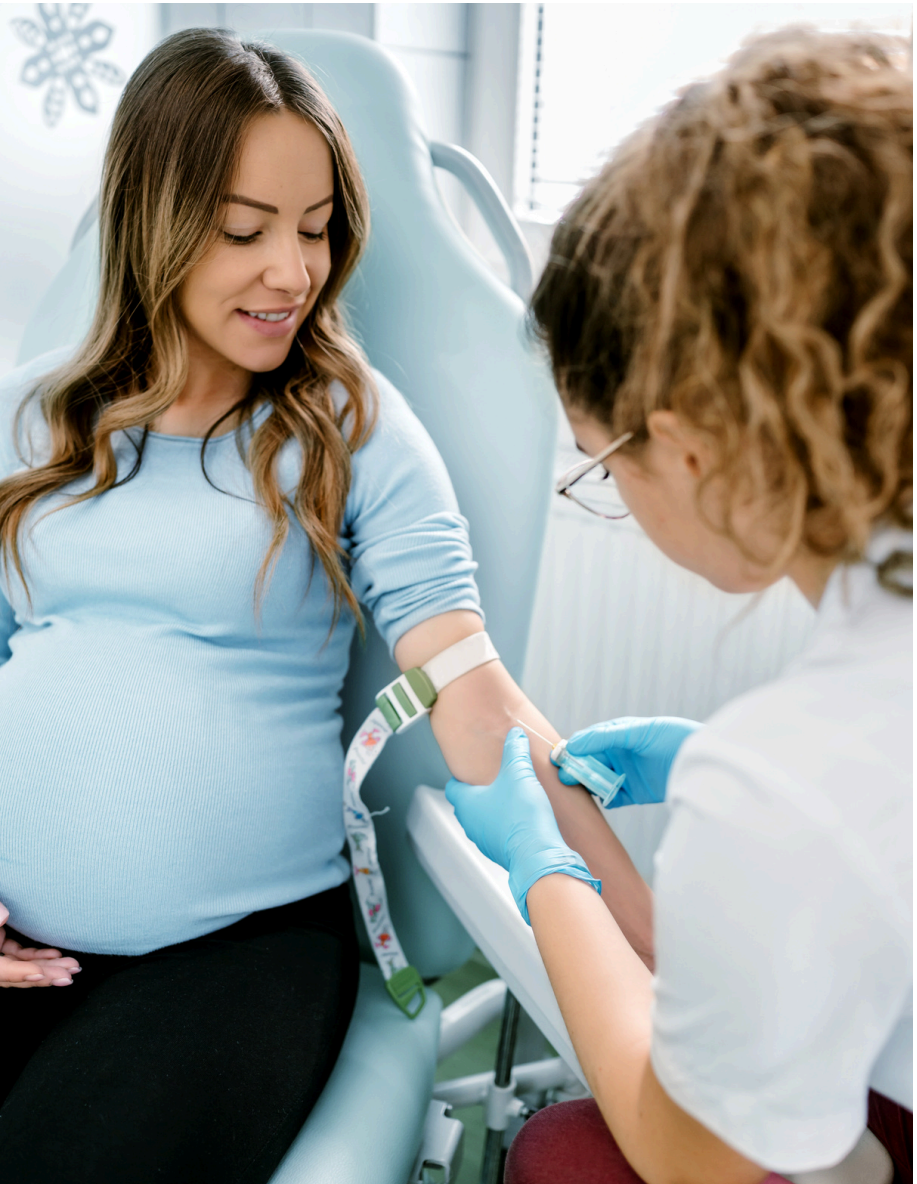
Sickle Cell Screening



If you want to know your sickle cell status, you can request a blood test from your GP. Additionally, local sickle cell centres in some parts of the country can arrange this test for you.

All newborn babies are offered screening for sickle cell disorder as part of the newborn bloodspot (heel prick) test when they are five days old. This early detection is crucial because babies with sickle cell disorder are vulnerable to serious infections. By identifying the condition early, babies can be prescribed penicillin and referred for specialist care to help them stay healthy. The screening also detects babies who carry the trait for sickle cell.

All pregnant women are offered screening for sickle cell disorder early in pregnancy, ideally by ten weeks. This screening identifies parents who carry the trait. If the mother is found to have the trait, the baby's father is also offered a screening test.

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ABOUT US

We are the UK's patient charity for people living with sickle cell disorder. We believe that people living with the condition have the right to quality care.

We support and represent people, patients and families affected by sickle cell disorder to improve their overall quality of life.

We work with health care professionals, pharmaceutical companies, researchers, parliamentarians, parents, and people living with sickle cell to support and advise, raise awareness of the disorder and campaign for improvements in treatments and care.

Our aim is to support those living with sickle cell, empowering them to achieve their full potential.

Reach out to us for :

- **Information and advice**. We run a telephone and email advice line (contact details on next page), and we have lots of information on our website, under FAQs.
- **Mentoring for young people**. We run programmes in London, Sheffield, and the North West.
- Advice about **sickle cell testing**. There is information on our website about inheriting the sickle cell gene and how you can get tested
- **Children's holiday.** We run a retreat each summer for families of children with sickle cell disorder. It offers some respite, connection with others and a chance to get advice and support
- Our campaigns include encouraging people from **black heritage communities to give blood**. To help those affected by Sickle Cell, 250 blood donations are needed each day.
- We also engage with **parliament** to apply pressure on making change in the NHS so that people living with sickle cell can have the quality care they deserve
- We work with Genomics England to ensure **researchers into sickle cell** will have an accurate and targeted set of data to use as a basis for their work going into the future, to give people living with sickle cell the best outcomes for generations to come.
- We offer **consultancy services** so that big organisations have accurate advice and information about sickle cell.

Contact

Sickle Cell Society Helpline

- Opening hours : 9am-5pm Monday -Friday (except bank holidays)
- Tel - Mon & Tue: 07842 245 980; Wed: 020 89617795; Thurs & Fri: 07809736089
- Email info@sicklecellsociety.org

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