

# Staying well with Sickle Cell Disorders

Directorate of Clinical Haematology

## Introduction

This leaflet has been written for patients with Sickle Cell Disorders to help them keep well.

A person with sickle cell disease can do a lot to help themselves keep well.

The first step is to learn as much as possible about the illness, get to know the things that commonly trigger a sickle cell crisis and sickle cell related illness and how to avoid those things and if a sickling crisis or illness does occur what to do to reduce their effect and if necessary when and how to seek medical help.

## What can cause a Sickle Cell Crisis?

It is known that certain things can increase the likelihood of having a sickle cell crisis including:

- Not having enough water in the body (dehydration)
- Not eating enough food (voluntary or involuntary fasting)
- Not taking recommended medications and vaccinations
- Being too hot or too cold
- Excessive physical exertion
- Emotional stress and anxiety
- Infection
- Smoking

## How can I help to reduce the risk of serious complications?

Trying to avoid these triggers can help but it is important to note that sometimes sickling crisis and illness does occur despite doing everything to avoid them. In these situations it is important to know what to do in order to avoid serious complications

- Dehydration is one of the most common causes of sickle cell crisis. The blood becomes thicker and sickle shaped red blood cells are more likely to stick together and cause a blockage in the blood circulation. It is therefore important for an adult to drink at least 3 litres of fluid daily. The fluid can be in the form of plain water, diluted squash tea and other fluids.

**The prevention of infection is a major priority in all healthcare and everyone has a part to play.**

- Please decontaminate your hands frequently for 20 seconds using soap and water or alcohol gel if available
- If you have symptoms of diarrhoea and/or vomiting, cough or other respiratory symptoms, a temperature or any loss of taste or smell please do not visit the hospital or any other care facility and seek advice from 111
- Keep the environment clean and tidy
- Let's work together to keep infections out of our hospitals and care homes.

Avoid alcohol if drunk it should be in very small quantities because alcohol increases dehydration and can lead to other complications. If for any reason a person with sickle cell disease is not able to drink normally, for example if they are vomiting losing fluid because they are sweating excessively due to a fever or they have diarrhoea it is important they seek medical attention promptly.

- A balanced diet is important because all body cells need food to keep healthy but the type of food eaten will determine how well the body tissues are able to work properly in order to prevent illness. In the UK it is recommended that in order to stay healthy people should eat five portions of fruit and vegetables a day, reduce the amount of fatty foods and salt and do regular exercise.
- The anaemia of sickle cell disease is caused by a rapid breakdown of red blood cells and not due to lack of iron necessarily. Therefore it is important that individuals should not self-medicate with iron and should only use iron medications if it has been prescribed by their doctor following a blood test that confirms they are genuinely iron deficient.
- Folic acid is a mineral that helps in the making of red blood cells. Some clinics recommend 5mg folic acid once per day, but this can vary between doctors so check with your doctor.
- Penicillin is recommended for people with sickle cell disease because their spleen does not work properly therefore they do not develop good immunity and are prone to getting infections especially pneumococcal infections. Taking penicillin twice a day is recommended on-going into adulthood.
- Extremes of temperature and sudden change from a warm to a cold environment can trigger a sickle cell crisis. When a person is cold the blood vessel walls thicken making the space in the middle smaller. When the red blood cells are sickle shaped they are more likely to cause an obstruction and sickling crisis.
- If a person is too hot they will sweat more losing more water from the body, when this happens the blood will become thicker. In order to prevent the body from becoming dehydrated the kidneys will reduce the amount of urine it is producing to make more concentrated urine. In people with sickle cell disease their kidneys are not able to concentrate the urine efficiently therefore even when they are too hot the kidneys continue to produce large amounts of diluted urine leading to dehydration increasing the risk of a sickle crisis.
- During excessive physical stress the body needs more oxygen in order to provide for active muscles. People with sickle cell disease have a chronic anaemia which reduces their ability to carry oxygen around the body and therefore during physical exercise they become breathless and tire easily. Although it is important to maintain physical activity this should be done in moderation. Each individual will need to identify their own physical tolerance level and maintain their own physical boundary. Physical stress includes things that put a greater physical burden on the body for example pregnancy. It is not uncommon for women to experience more frequent sickling events during pregnancy and in the early weeks after delivery.
- Emotional stress and anxiety are a contributing factor to trigger sickle crises from late childhood and early adulthood e.g. exams, stress at work and relationship stress. Financial pressures and other challenges of adulthood can compromise the wellbeing of someone with sickle cell disease. If these stressors cannot be managed by the individual speak to your GP haematology consultant or nurse specialist.
- Infection is the most common trigger for a sickle cell crisis. Although these can be difficult to avoid the most common cause is failing to take the daily prescribed penicillin and recommended vaccinations. Trying to avoid situations that place one at greater risk of infection including poor hygiene, improper preparation of food, lack of attention to treating wounds including insect bites and stings all helps.
- Cigarette smoke contains a chemical called carbon monoxide that affects the oxygen carrying capacity of the blood, which will already be lower in a person with sickle cell disease. Smoking also worsens symptoms including colds, flu and chest infections. These can all trigger crises if the symptoms are severe.

Speak to your GP or nurse specialist if you would like advice on quitting smoking.

## Useful contact numbers:

**Consultant in Haemoglobinopathies**

01902 695271

**Haematology Clinical Nurse Specialist**

01902 695276

## Where can I get further information?

**The Sickle Cell Society**

54 Station Road,  
London NW10 4UA

Tel 02089617795

web [www.sicklecellsociety.org](http://www.sicklecellsociety.org)

**Sickle Cell and Thalassaemia Support Project (Wolverhampton)**

2nd floor office St.Johns House

St. Johns Square

Wolverhampton

WV2 4BH

Tel 01902 444076

[www.sctsp.org.uk](http://www.sctsp.org.uk)

email [info@sctsp.org.uk](mailto:info@sctsp.org.uk)

**Sickle Cell Care (Wolverhampton)**

58 Bank Street

Wellington Road

Bilston

WV14 8PD

Tel 01902 498274

[www.sicklecellcare.net](http://www.sicklecellcare.net)

## English

If you need information in another way like easy read or a different language please let us know.

If you need an interpreter or assistance please let us know.

## Lithuanian

Jeigu norėtumėte, kad informacija jums būtų pateikta kitu būdu, pavyzdžiui, supaprastinta forma ar kita kalba, prašome mums apie tai pranešti.

Jeigu jums reikia vertėjo ar kitos pagalbos, prašome mums apie tai pranešti.

## Polish

Jeżeli chcieliby Państwo otrzymać te informacje w innej postaci, na przykład w wersji łatwej do czytania lub w innym języku, prosimy powiedzieć nam o tym.

Prosimy poinformować nas również, jeżeli potrzebowaliby Państwo usługi tłumaczenia ustnego lub innej pomocy.

## Punjabi

ਜੇ ਤੁਹਾਨੂੰ ਇਹ ਜਾਣਕਾਰੀ ਕਿਸੇ ਹੋਰ ਰੂਪ ਵਿਚ, ਜਿਵੇਂ ਪੜ੍ਹਨ ਵਿਚ ਆਸਾਨ ਰੂਪ ਜਾਂ ਕਿਸੇ ਦੂਜੀ ਭਾਸ਼ਾ ਵਿਚ, ਚਾਹੀਦੀ ਹੈ ਤਾਂ ਕਿਰਪਾ ਕਰਕੇ ਸਾਨੂੰ ਦੱਸੋ।

ਜੇ ਤੁਹਾਨੂੰ ਦੁਭਾਸ਼ੀਏ ਦੀ ਜਾਂ ਸਹਾਇਤਾ ਦੀ ਲੋੜ ਹੈ ਤਾਂ ਕਿਰਪਾ ਕਰਕੇ ਸਾਨੂੰ ਦੱਸੋ।

## Romanian

Dacă aveți nevoie de informații în alt format, ca de exemplu caractere ușor de citit sau altă limbă, vă rugăm să ne informați.

Dacă aveți nevoie de un interpret sau de asistență, vă rugăm să ne informați.

## Traditional Chinese

如果您需要以其他方式了解信息，如易读或其他语种，请告诉我们。

如果您需要口译人员或帮助，请告诉我们。