

Sickle Cell

Haematology

What are Sickle Cell Disorders?

Sickle cell disorders is the umbrella name given to a group of hereditary blood conditions, which affects the haemoglobin (an oxygen carrying component that gives blood its red colour) Under certain conditions the red blood cells change shape to resemble that of a farmers sickle which can cause many problems and symptoms

People with sickle cell disorder have symptoms which include anaemia, severe pain and an increased risk of infection.

It is important for people with sickle cell disease to learn as much as possible 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

How you can avoid the triggers for a sickle cell crisis

Keep hydrated

You can become dehydrated if you are not drinking enough fluids, especially on a hot day, or 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 Haematology Day Unit /Haematology Ward for advice.

Have your recommended medications and vaccinations

If you have been prescribed medications it is important that you take these as directed.

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.

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

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 - to protect against hepatitis B which is transmitted through infected blood and sexual contact with an infected person

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 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.

How to Manage Pain at home

- Use prescribed pain medication only as directed by your doctor
- Rest and try to keep warm
- Increase your fluid intake
- Massage if appropriate
- Try to eat small amounts of food
- Relaxation- some people find that activities such as music, breathing exercises and massage useful

When to Call the Hospital For Advice

There are certain signs and problems you need to watch out for and contact the hospital for emergency advice. These are:

- Fever- If you have a temperature of 38 degrees or above, with or without shivering and shaking
- Chest discomfort- pain or trouble breathing
- Stomach discomfort- severe pain or swelling
- Headaches- severe headaches, blurred vision, dizziness
- Change in behaviour- Appears confused, drowsy or unable to speak
- Penis- painful erections
- Pain- sickle pain anywhere in the body that cannot be controlled at home with your usual pain relief and pain coping methods

Please note that this list is not a total list of all the problems and complications that can occur with your sickle cell. Please do not wait to contact the hospital for help and advice as early as possible, to avoid you becoming very unwell

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 your consultant with the following information:

- The type of sickle cell disease you have
- Any complications you have
- The medications you are taking
- Contact details for your Haematology 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. You also need to check whether you need to have any extra vaccines before you travel or take any extra medications such as anti-malaria tablets.

It is also vital that you get adequate travel insurance that will cover you if you have any medical problems related to your sickle cell disease whilst abroad.

If you are travelling by plane, make sure you drink plenty of non-alcoholic fluids, keep warm and go for short frequent walks up and down the aisle if your journey is long.

Whilst abroad it is important that you keep hydrated; if you are not sure about the cleanliness of the water then drink bottled water, particularly if you develop diarrhoea and/ or vomiting.

Contact details

If you have any questions or concerns please contact the **Haematology Unit**
Haematology ward (B11 Heart and Lung centre)
01902 694241 / 4242

Haematology Day Unit

0190 2695232 / 5045

Monday – Friday 9am- 5pm

Emergency Line 07920587036

Haematology Clinical Nurse Specialist

01902 695276

Monday- Friday 9am- 5pm

Further Information

For more information about sickle cell:

Sickle Cell Society

www.sicklecell society.org

For more information about our hospitals and services please see our website:

The Royal Wolverhampton NHS Trust

www.royalwolverhamptonhospitals.nhs.uk

Sources used for information in this leaflet

Sickle Cell Society, 'Standards for the clinical care of adults with sickle cell disease in the UK', 2nd ed, 2018

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。