Patient information

Haemoglobinopathy Disorders - Living with Sickle Cell Disease Information and Advice for Patients and Carers

Haematology Liverpool

Haemoglobin is the substance in red blood cells that is responsible for the colour of the cell and for carrying oxygen around the body.

People with sickle cell disorder are born with the condition, it is not contagious. It can only be inherited from both parents each having passed on the gene for sickle cell.

The main symptoms of sickle cell disorder are anaemia and episodes of severe pain. The pain occurs when the cells change shape after oxygen has been released. The red blood cells then stick together, causing blockages in the small blood vessels.

These painful episodes are referred to as sickle cell crisis.

Home Treatment of painful Crisis

Reduce/minimise triggers if possible (See below).

Take analgesia as previously discussed by Haematology team

Examples of Common Pain Medicines

Paracetamol	**1g**	maximum of four times in 24 hours
Ibuprofen	**400mg**	maximum of three times in 24 hours
Co-codamol	**30/500mg**	maximum of four times in 24 hours
Codeine	**30mg/60mg**	maximum of four times in 24 hours
Tramadol	**50/100mg**	maximum of four times in 24 hours
Dihydrocodeine	**20mg/60mg**	maximum of four times in 24 hours

Your team will have discussed your individual needs with you.

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.

- Drink three to four 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 Team (SCT) for advice.

Have your recommended medications and vaccinations.

If you have been prescribed medications it is important that you 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

People with sickle cell disease have a spleen that doesn't work properly. This is called functional hyposplenism. 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 placing you at increased risk of pneumonia and meningitis.

There are measures mentioned below that may reduce your risk of infection.

Your doctor will recommend that you take phenoxymethylpenicillin (penicillin V) 250mg twice a day. This is an antibiotic that is very good at destroying the types of bacteria your spleen would normally destroy. This is aimed to reduce the risk of infection that these bacteria can cause.

If you are allergic to phenoxymethylpenicillin (penicillin V) then another type of antibiotic will be offered.

If you have any concerns or questions regarding taking these tablets, then please discuss it with you doctor or specialist nurse.

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 five years to protect against the pneumococcus bacteria.

Influenza (flu) - once a year to protect against the flu virus.

Hepatitis B - to protect against hepatitis B which can be transmitted through infected blood.

Meningivac - This gives protection against meningococcus types A and C which cause meningitis. A single dose should be given if not already received as part of child immunisation schedule.

Haemophilus Influenzae Type B - A single dose should be given if not already received as part of child immunisation schedule.

COVID 19 - As per Heamatology team advice

Your sickle cell team will liaise with your family doctor (GP) regarding your up-to-date vaccinations.

Hydroxycarbamide

Some patients with sickle cell disease may be prescribed hydroxycarbamide. Hydroxycarbamide is a medicine taken in capsule form. It causes changes in the blood, which reduce the frequency of sickle cell crises and the need for transfusion in some patients with sickle cell disease. It may also be called hydroxyurea.

Blood Transfusion

Some patients with sickle cell disease may require blood transfusions (exchange or top up) to reduce the number of sickled cells in their circulation and improve symptoms of anaemia.

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 or doctor 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.

Specialist Heamoglobinopathy Team:

Consultant Haematologist

Lead Clinical Nurse Specialist

Two Clinical Nurse Specialists

Haematology Specialist Registrar (Rotational Position)

Clinical Psychologist

Feedback

Your feedback is important to us and helps us influence care in the future.

Following your discharge from hospital or attendance at your Outpatient appointment you will receive a text asking if you would recommend our service to others. Please take the time to text back, you will not be charged for the text and can opt out at any point. Your co-operation is greatly appreciated.

Further Information

Contact Details:

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